DYSTONIA

A comprehensive and longitudinal study of the epidemiological, social, economic and psychological implications of dystonia within the population of the North East of England

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PUBLICATIONS

The following abstracts and publications have been published since the start of the research and form part of the ongoing relationship with other contributors to the overall research projects. None of the specific material used in previous publications is quoted in this research, except where noted.

Butler A.G. (1995) **The socio-economic implications of dystonia.** *The Dystonia Society Newsletter.* ; 19 ; 4-5 and 20 ; 4-5 and 21 ; 5-6. London. TDS.

Butler A.G. (1996) **The social and economic implications of dystonia**. *European Journal of Neurology* ; 3 : 79.

Butler A.G. (1997) **The epidemiology of Spasmodic Torticollis in North East England.** In: *The National Spasmodic Torticollis Association Annual Symposium* in Nashville, Tennessee on 9th November 1997.

Butler A.G, Duffey P.O.F. (1996a) **The epidemiological survey of dystonia in the North East of England**. *European Journal of Neurology* : 3 : 28

Butler A.G, Duffey P.O.F. (1996b) **An epidemiological survey of dystonia at Darlington Memorial Hospital**. *European Journal of Neurology* : 3 : 79

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Butler A.G, Duffey P.O.F. Hawthorne M.R, Barnes M.P. (1998) **The socioeconomic implications of dystonia**. In: Fahn S, Marsden C.D and DeLong M.R. eds., *Advances in Neurology, Vol. 78 : Dystonia 3.* Philadelphia : Lippincott-Raven. 349-358.

Butler A.G, Hawthorne M.R, Duffey P.O.F, Gudex C.M. (1995) **A comparison using a number of different rating scales measuring the effectiveness of Botulinum Toxin therapy in the treatment of dystonia and secondary dystonic spasms**. *Movement Disorders*; 10 : 398.

Duffey P.O.F, Butler A.G, Hawthorne M.R, Barnes M.P. (1998) **The epidemiology of primary dystonia in the north of England.** In: Fahn S. Marsden C.D. and DeLong M.R. eds. *Advances in Neurology, Vol. 78 : Dystonia 3.* Philadelphia : Lippincott-Raven. 121-126.

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Medd D.Y. (1999) **Clinical Report : A single-case study of generalised dystonia and hypnosis, with unexpected immobility and an untoward effect.** *Contemporary Hypnosis* : Vol 16, No 1, 45-48.

Medd D.Y. (1999) **Hypnosis with selected movement disorders.** *Contemporary Hypnosis* : Vol 16, No 2, 81-86.

Gudex C.M, Hawthorne M.R, Butler A.G, Duffey P.O.F. (1995) **A cost-utility analysis of Botulinum Toxin therapy in the treatment of dystonia.** *Movement Disorders* : 10 ; 373.

Gudex C.M, Hawthorne M.R, Butler A.G, Duffey P.O.F. (1997) **Measuring patient benefit from Botulinum Toxin in the treatment of dystonia : Feasibility of Cost-Utility Analysis.** *PharmacoEconomics* : 12 : 6 ; 675-684.

Gudex C.M, Hawthorne M.R, Butler A.G, Duffey P.O.F. (1998) **Effect of dystonia and Botulinum Toxin treatment on health-related Quality of Life.** *Movement Disorders* : 13 : 6 ; 941-946.

Whitaker J, Butler A.G, Barnes M.P. (1998) **Botulinum Toxin treatment for people with dystonia by an outreach nurse practitioner - a comparative study.** Report to the Northern and Yorkshire Regional Health Authority (NYRHA).

Whitaker J, Butler A.G, Semlyen J.K, Barnes M.P. (1999) **Botulinum Toxin treatment for people with dystonia treated by an outreach nurse practitioner - a comparative study.** Submitted to the *Archives of Physical Medicine and Rehabilitation* in 1999. Currently under peer review.

Note :

All of the enclosed is my own work except where specifically noted. Although a number of the above publications are jointly authored, these have been specifically noted when abstracts have been quoted in the text.

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1st September 1999

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ABSTRACT

Dystonia is a little known neurological disease of the central nervous system and consists of a group of related movement disorders, characterised by involuntary and prolonged spasms of muscle contraction. Although it is nearly 90 years since this neurological disorder was first named, relatively little research had been undertaken into dystonia for the first 65 years and it was not until the mid 1970's that researchers started to look at the disorder.

This particular programme of research has taken place exactly over a six year period, starting in May 1993, and relates to a large number of different areas of study. This research has proven that dystonia is far more prevalent than previously thought, it is next to Parkinson's Disease in degree of prevalence and is far more common than other better known neurological conditions, such as Motor Neurone Disease, and yet it remains largely unknown to most members of the medical profession and the general public at large.

Dystonia has been historically extremely difficult to diagnosis and this meant it has been previously very difficult to obtain sufficient numbers for study, which in turn has created a number of significant social and economic consequences, which has mainly meant that most cases of people with dystonia have remained undiagnosed or misdiagnosed for many years.

This research was designed to measure the severity and prevalence of dystonia in the northern part of the UK, the implication the disease has had on the working life and environment of each patient and how that person is coping with the various personal, social and family relationships caused by the onset and potential gradual deterioration of the disorder, as well as measuring the quality of life of each patient during a number of different therapies.

Although there has been research into other neurological disabilities, very little is known about the implications that dystonia can have on the affected person and their families. This is the first time that all types of dystonia have been studied and that certain related subjects have been specifically included.

This research has been enormously helped by the tremendous expansion in the use of Botulinum Toxin therapy and although an enormous amount of work has been completed and accomplished during this research programme, it should never be forgotten that the subjects of this thesis are real people and that the implications and results of this research have had, and will have, a tremendous impact on their lives and that of their families.

PART ONE : INTRODUCTION

CHAPTER 1

Introduction and Aim of the Research

What is Dystonia ?

Dystonia is a previously little known neurological disease of the central nervous system (CNS) and consists of a group of related movement disorders, characterised by involuntary and prolonged spasms of muscle contraction. It has been classically defined as " *a syndrome of sustained muscle contractions, frequently causing twisting and repetitive movements, or abnormal postures.* " (Fahn et al, 1987)

It can affect the whole body or a single part of the body or some combination between the two. Although it is nearly 90 years since this neurological disorder was first named (Oppenheim, 1911), relatively little research had been undertaken into dystonia for the first 65 years, apart from the odd periodical review (Herz, 1944).

Since the 1970's, a number of eminent medical people have started to research into dystonia and this has increased its 'popularity' amongst medical researchers. However, to date, there has not been a comprehensive longitudinal study of the disease within a specific population anywhere in the world.

What is the aim of this research ?

This research is designed to measure the severity and prevalence of dystonia in part of the UK, the implication the disease has had on the working life and environment of each patient and how that person is coping with the various personal, social and family relationships caused by the onset and potential gradual deterioration of the disorder, as well as measuring the quality of life of each patient in a number of different therapies.

Although there has been research into the prevalence of most neurological disabilities (Hewer, 1993), very little is known about the implications that dystonia can have on the affected person and their families, apart from limited circulation articles in The Dystonia Society (TDS) newsletters (Butler, 1995).

The purpose of this research was designed to measure the quality of life of each patient, the severity and prevalence of dystonia in the UK, the implication the disease has had on the working life and environment of each patient and how the patient is coping with the various personal, social and family relationships caused by the onset and potential gradual deterioration of the disorder.

It is known that dystonia causes considerable morbidity in terms of reduced mobility, pain, low self-confidence, embarrassment, poor social interaction and difficulties with employment (Marsden and Quinn, 1990) but this will be the first time that all aspects of life in people with the disorder has been comprehensively studied.

Why was it necessary ?

During private correspondence in July 1991 between the author and the late David Marsden, Professor of Neurology at the Institute of Neurology in London, David Marsden stated that " *no one knows how many cases of dystonia exist in the U.K.* "

When The Dystonia Society was first established, based on Marsden's patients' list in 1983, there were only 6 known dystonia patients in the North East, all of whom were Generalised and most of whom had Familial (ie, genetically inherited) Dystonia.

This is not to say that there were not other people correctly diagnosed at the time in the North East, but it does indicate that they were not known outside of their own medical centre nor were they on any formal (or even informal) epidemiological register.

When the first Self-Help Group (SHG) was established in the North East in 1990, there were 40 known people with dystonia on the initial mailing list, including 25 TDS members.

In a survey of 705 people with dystonia, it had been established that one third of all dystonia sufferers take 5 years or more to diagnose; 37% said that at least one doctor had suggested that their condition was "*all in the mind*" and 32% had been referred at one time to a psychiatrist; 66.7% of all sufferers needed at least 5 consultations before diagnosis and 65.7% were misdiagnosed at some stage (TDS, 1993).

At the time of the eventual start of this research, TDS (NE) had only 56 members, with only a further 87 people with dystonia known in the region. This meant that the research started officially on 6th May 1993 with only 143 known people displaying at least one of the various symptoms known as dystonia in the geographical area known as the North East of England.

Moreover as the author was not medically qualified, it was important for a detailed literature review and a limited training programme to be carried out prior to the start of this research. This took over two years of intensive study from 1991 until 1993 and included attending a number of research and training seminars throughout the U.K., some of which are noted throughout the following pages.

Finally, it was necessary as the author felt that the number of people with dystonia had always been under-estimated and therefore some form of in-depth investigation had to be carried out before others could categorically state the true relationships involved.

PART ONE : INTRODUCTION

CHAPTER 2

Background

<u>How did this research come about ?</u>

" *One of the main problems about dystonia in this country is that we have no really good idea about how common it is. It is likely that the majority of cases are undetected by the medical services and so research into the social and economic aspects of the cases we know about would not really reflect what is actually going on in the population. In fact, one of the things we really do need to know is how common dystonia is, and this is difficult to do without actually doing a door to door survey, which I do not think is the sort of thing you have in mind.*"

The above is an extract from a letter to the author by Dr Nick Fletcher, at the time Senior Registrar in Neurology at St Bartholomew's Hospital in London and the Medical Advisor to the Dystonia Society, dated 10th August 1992.

This was the initial stimulus which started the author on a task which has been far longer than at first envisaged and at times very difficult to accomplish, but it has since become "*one of the most important pieces of research into dystonia, outside of purely medical research to find a cure* " (Source : private correspondence with the Dystonia Medical Research Foundation in the USA).

<u>Why was it done in the way it was ?</u>

One of the main reasons why there has been a problem in the diagnosis of dystonia is mainly historical, as it was not even classified as a neurological disorder until quite recently. "*Until the 1970's most patients with dystonia were referred to psychiatrists in the belief that these curious motor disorders were an expression of an unhappy mind. Vigorous efforts over the past two decades have now, however, established that the various syndromes of dystonia are the result of abnormal brain function, usually in the basal ganglia.* " (Marsden and Quinn, 1990).

It was therefore necessary to firstly find out how many people had dystonia in order to obtain sufficient numbers of subjects from which to draw the necessary information on which to research for this project.

Access to people who have dystonia was considered to be the primary difficulty in carrying out any sort of dystonic research project (Butler et al, 1998) and as the very process of identifying those people with dystonia meant first having to obtain a correct diagnosis, the need was seen to start by firstly carrying out a comprehensive epidemiology of dystonia.

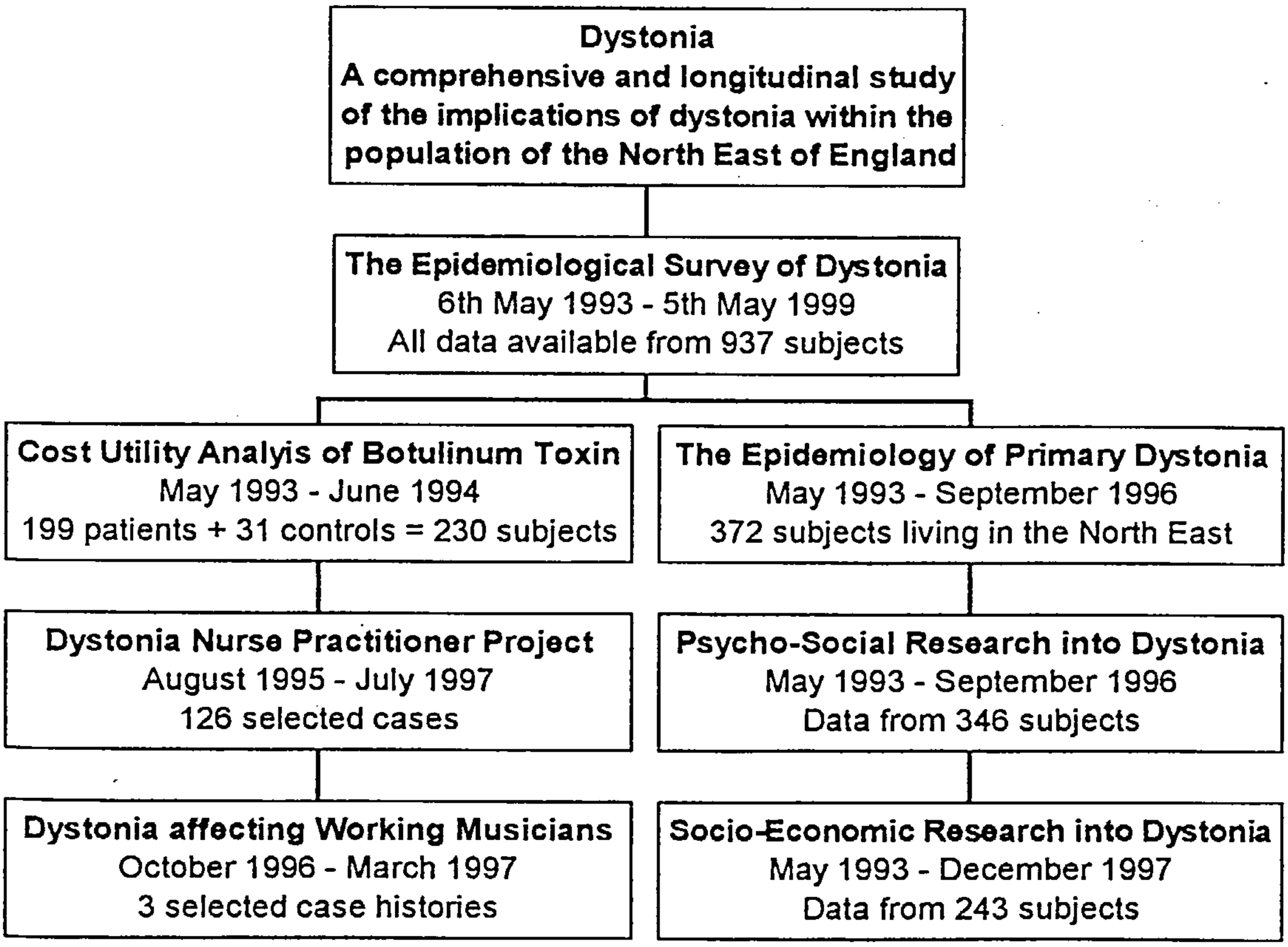
When did it start ?

The research started officially on 6th May 1993 and continued full time until 5th May 1999 for an inclusive period of 6 years exactly. However the reader should be aware that although the entire process was continuous there were, of necessity, a number of different research projects, which became part of the entire process, as follows :-

Oct 1996 - Mar 1997	: Dystonia affecting Working Musicians	= 0.5 years
May 1993 - June 1994	: A Cost Utility Analysis of Botulinum Toxin	= 1.2 years
Aug 1995 - July 1997	: The Dystonia Nurse Practitioner Project	= 2.0 years
May 1993 - Sept 1996	: The Epidemiology of Primary Dystonia	= 3.4 years
May 1993 - Sept 1996	: The Psycho-Social Research into Dystonia	= 3.4 years
May 1993 - Dec 1997	: The Socio-Economic Research into Dystonia	= 4.7 years
May 1993 - May 1999	: The Epidemiological Survey of Dystonia	= 6.0 years

Although each part of the above research is free standing as such, the whole was always designed to form the author's overall research for his Doctor of Philosophy degree thesis. The following chart shows the relationship between the various projects and the number of patients involved in each as well as the length of time taken.

CHART No 1. Schematic showing the different research projects undertaken.



There have been a number of publications, made as a direct result of the research done by the researcher and his fellow workers, which are all listed on pages vii and viii.

PART TWO : DYSTONIA

CHAPTER 3

Literature Review

General Review

Before 1976, dystonia was often considered to be a pyschogenic / psychological / psychiatric disorder and therefore, as it was a vital part of the initial literature review to define which classification system should be used in the research programme, it was necessary to examine what already existed.

International Classification of Diseases

The International Classification of Impairments, Disabilities and Handicaps (ICIDH) had been developed by the World Health Organisation (WHO) as a way of inter-relating diverse data on individuals to illuminate the impact of disease, to promote a more sensitive use of terminology and expose weaknesses in thinking about disability. There has been some criticism of the WHO definitions in saying that " *they pay insufficient attention to the external determination of handicap and are yet another means of labelling or stigmatising the handicapped* " (Wood, 1987). It also should be remembered that ICIDH is not a classification of persons but of the types of experiences to which they may be exposed. " *It is not a research or assessment tool but a means of organising data in a useful way. It was never designed as a basis for organising people into categories* " (Wood et al, 1989).

Further work was therefore needed to assess ICIDH's usefulness to these particular dystonia research programmes. Eventually all ICIDH codes were rejected, including ICD8 and ICD9 (as used in the NHS prior to the implementation of ICD10 in April 1995).

The reasons are obvious once one looks, for example, at the classification of dystonia in ICD9 (NHS Man. Exec, 1978). Code 333.6 shows *Idiopathic Torsion Dystonia*, with only Dystonia Musculorum Deformans or (Schwalbe) Ziehen - Oppenheim Disease as a sub category. Code 333.7 shows *Symptomatic Torsion Dystonia*, with only Athetoid Cerebral Palsy or Vogt's Disease and Double Athetosis as sub categories. And finally, all the focal dystonias are shown under code 333.8 as *Fragments of Torsion Dystonia*, with only Blepharospasm, Organic Writer's Cramp, Orofacial Dyskinesia and Spasmodic Torticollis shown as the sub categories.

Any person, who knows even the slightest amount about dystonia and its various sub-categories, will know that the above categories do not reflect either the current known different stages of dystonia nor the most up-to-date classifications of dystonia. Therefore it became quite obvious quite quickly that some other system as opposed to that currently in use within the NHS had to be used to classify dystonia.

The Read Codes

The author then studied and eventually rejected the new Read Codes, which are being gradually implemented in the NHS, as they merely follow the ICD model (NHS Man. Exec, 1993). For example; the three ICD9 categories shown above equal the Read Code categories of F136, F137 and F138. Even Spasmodic Dysphonia is **still** being categorised as R0444, i.e., under 'Symptoms, Signs and Ill-Defined Conditions'. The reason for all these rejections was on the grounds that they were either too general or not detailed enough for a proper dystonic classification.

Epidemiological Surveys of Dystonia

Any epidemiological survey of dystonia (ESD) is normally, by definition, quantitative in nature as its purpose is initially merely to identify the actual numbers of people with dystonia within a given area. However, as has been seen by earlier reference to Professor David Marsden and Dr Nick Fletcher, access to people who have dystonia was considered to be the primary difficulty in carrying out any sort of research programme and as the very process of identifying those people with dystonia meant first obtaining a correct diagnosis, the opportunity arose of having previously unknown (unidentified and undiagnosed) patients available for other dystonia research projects.

As can be proved all the research projects stem from the initial need to correctly identify sufficient numbers of people with dystonia. Therefore, in practice, the epidemiology was the tool which identified the subjects with dystonia and although it was also a piece of important research within itself, it led to the different and varied research projects and in particular the data for this research thesis.

However, in order to carry out an epidemiology of dystonia, a review of the literature on the subject revealed several important areas in both the design and implementation of such a study. According to one of the world's foremost neuroepidemiologists : " *Neuroepidemiology may be defined as the study of the distribution and dynamics of neurological diseases in human populations and the factors that affect those characteristics.* " (Schoenberg 1978)

It is precisely for these reasons that this ESD is of such importance because it attempts, as never before, *to identify all cases of* (dystonia) *in a well-defined population* (author's emphasis). Other studies (Nutt et al, 1988 and Nakashima, 1995) have been exclusively clinic based and therefore will have a definite referral bias and do not carry the same validity as one that attempts 100% identification. Those epidemiological studies that were not clinic based (Alter, 1976; Korczyn, 1980; Zilber, 1984 and Li, 1985) are flawed in that either the ascertainment of their diagnosis was incomplete or the nature of their criteria was limited in that they were not purely designed to just pick up dystonia and therefore are not as accurate as the data from the North East study.

Disability in General

Five separate, but linked, surveys into disability in Great Britain have been published as six separate reports (No 1: Martin et al, 1988; No 2: Martin and White, 1988; No 3: Bone and Meltzer, 1989; No 4: Martin et al, 1989; No 5: Smyth and Robus, 1989; No 6: Meltzer et al, 1989). The surveys were undertaken from 1984 to 1988 and are the most comprehensive and recent available from the Office of Population, Censuses and Surveys (OPCS).

Their aim was to find out the number of disabled people and the severity of their disability and they estimated that there were 6,560,000 people with disabilities in Great Britain in 1985, although it has proved impossible to accurately differentiate people with any form of dystonic spasm within this estimate from the data.

The types of disability and severity were distinguished according to ICIDH, a feature of the survey being the construction of the measure of severity of disability from 1 (low) to 10 (high). Of the six and a half million disabled people in the UK with at least one disability, only 400,000 (7%) were living in communal homes, and this number is related in direct proportion to the severity of the disability.

These reports contain a lot of very detailed findings but it has proved difficult to relate any of them to dystonia in particular or even other neurological movement disorders in general. However, one particular finding will and can be compared in this research ie, the fact that more women are affected than men.

Approximately one in every nine people in the U.K. is disabled in one form or another and one family in every six has a member who has a disability, nevertheless the OPCS reports define disability as medical criteria whereas WHO (ICIDH) classification of disability covers a range of medical conditions which may not be handicapping depending on the individual's ability to overcome the socio-economic and environmental obstacles (O'Brien and McFetridge. 1991). The views of disabled people in general on disability show that the use of terms like 'the disabled' to describe 'physically handicapped people' is often deplored and attempts by the medical profession to understand how disability affects a person's life are described in at least one journal as "*inadequat*e" and "*likely to promote stereotypical views*" (Brisenden 1986).

Other research has shown that the discrimination faced by disabled people in a society of 'normal' people acts as a catalyst to the perpetuation of passivity and dependence amongst people with disabilities. A lack of facilities, and inadequate designs, ensure that they constantly face problems ranging from gaining access to a building to facing constant medical interventions, regardless of their value (Barton 1989). One needs to discover methods by which disabled people can achieve greater independence and an equal place in society, exercising freedom of choice for independent living as a right, rather than a privilege (Brisenden 1986 and Oliver 1990). Whilst researching the field of disability, it became quickly evident that there were several schools of thought within the dystonic population in particular, and the disability movement in general, about the different theories regarding disability.

The Social Model of Disability.

One of these 'schools of thought' relates to the "*social model of disability*", which is a vast potential area of research in itself, well beyond the brief of this research thesis, nevertheless it should be mentioned and explained. In essence, this view of disability states that " *traditionally, disability has been seen as the problem of the individual and it has been the individual who has had to change or be changed by professionals through rehabilitation or cure* " (Michael Oliver, Professor of Disability Studies at the University of Greenwich, spoken at a seminar on disability in Newcastle-upon-Tyne on 3rd February 1994).

Furthermore, according to Professor Oliver, " *Now, disabled people and their organisations have described, from their own experience, how it is economic and social barriers which stop people with impairments participating fully in society. These barriers are so widespread that we are prevented from ensuring a good quality of life for ourselves*" (Oliver, 1990).

This explanation is known as the 'social model of disability', because it focuses on society's disabling environments and barriers of attitude, rather than on individuals with impairments. The social model was formulated by disabled people and now also has been accepted by a number of non-disabled academics and professionals. It stresses human rights and the equality of opportunities.

Mike Oliver has suggested that " *money spent on medical research into cures for disabling illnesses and conditions is wasted money and most funds raised for this purpose are allocated and administered by able-bodied people, and the obsessive pursuit of a cure can ruin as many lives as the illness or condition itself* " (Oliver 1990) and at the Newcastle Seminar, he further asserted that " *disabled people do not want to be cured and such (research) funds would be better utilised in removing some of the social barriers which limit the potential of disabled people.*"

Oliver predicts that " *the disability movement will continue to confront the disablism in service provision and professional practice, but the tactics of confrontation are beginning to change. In place of writing books and articles and speaking at conferences, the 1990's may well become the decade of direct action on disability. This direct action will build on the failures of the past, challenge the vested interests of the present and force a restructuring of state welfare that is so far-reaching that we may not even recognise the welfare state in the twenty-first century.*" (Oliver, 1991)

The author's personal experience is not compatible with this view of disability and conversations of an anecdotal nature, since February 1994, have suggested that most people with dystonia (which, for those with primary dystonia at least, is potentially a curable disease) want a cure, if one can be found - although most agree that more could be done by way of practical help now !

PART TWO : DYSTONIA

CHAPTER 4

Classification

Up until 1996, it was necessary to use the Ad Hoc Committee of the Dystonia Medical Research Foundation criteria (Fahn, 1988) which classifies dystonia in three ways.

TABLE 2. Classification of Dystonia

(I)	By cause	(a) Idiopathic	(Primary Dystonia)
		(b) Symptomatic	(Secondary Dystonia)
(II)	By age at onset	(a) Childhood-onset	: 0-12 years
		(b) Adolescent-onset	: 13-20 years
		(c) Adult-onset	: >20 years
(III)	By distribution	(a) Focal	- affects a single part of the body only
		(b) Segmental	- affects two or more connecting parts
		(c) Hemidystonia	- affects an ipsilateral arm and leg only
		(d) Multifocal	- affects two or more non-contiguous parts
		(e) Generalised	- defined as segmental, plus at least one leg.

There were generally considered two causes of dystonia; **idiopathic** (familial and sporadic) generally with an unknown cause or **symptomatic** with a known (or assumed) cause. There are many known causes for secondary dystonia which have been listed within the medical textbooks.

The age at onset classification is only normally used where it can give a fairly accurate measure of the distribution of the disorder over time. A rough rule of thumb guide is the earlier one gets this disorder the greater the chance of it spreading throughout the entire body. However, the opposite is also true, in that the later one develops a form of dystonia, the more localised (or focal) it remains.

The final classification is the position which the disorder currently occupies within the human body. These are generally described as above, although there are also a number of sub-divisions within these categories. It should be remembered that the disease will often move throughout the human body over time and therefore one can often see a young child developing dystonia in a limb, which will progress until it is present in most (if not all) parts of the entire body.

It is therefore often assumed that once one develops the disorder it is with one for the rest of ones life. This is generally true, but there have been a number of exceptions to this and indeed a number of the present cohort of patients have gone into remission, some for many years.

Each of these classifications will be discussed in greater detail later, but it must be realised from the start that the classification with regard to 'age by onset' and by 'distribution' has remained constant throughout this entire time.

Aetiology

The new aetiological classification has really come about as a direct result of our ever-changing and improving understanding of this disorder. As a result of a paper (Fahn et al, 1998) presented in Miami, Florida at the 3rd International Dystonia Symposium in October 1996, the aetiological classification of dystonia has greatly changed. However, it should be realised that this has not changed either its 'age by onset' nor essentially its 'distribution', only its aetiology. These main changes relate to the recent discoveries in the genetics of dystonia, which indicate that more than gene can cause idiopathic dystonia.

This major breakthrough owes much of its reasoning to the recent classification of parkinsonism, where it has proved useful to divide " *parkinson disorders into primary (usually called Parkinson's Disease), secondary (due to environmental or structural causes), Parkinson-plus syndromes (in which other neurologic features in addition to parkinsonism are also present) and heredodegenerative syndromes (such as Hallervorden-Spatz disease, Wilson's disease and Huntington's disease), which can be present with a parkinsonian picture* " (Fahn, 1995).

The idea is to divide dystonia into four main aetiological classification, each one relating to specific other " *recognised entities that appear to be distinct and each may be enlarged in the future as further genetic, pathological and biochemical advances are made*" (Fahn et al, 1998).

TABLE 3 : New Aetiological Classification

1 : Primary Dystonia	syndromes with dystonia as the sole phenotype
2 : Dystonia-Plus	distinct from the following two types and includes dystonia with parkinsonism, dopa-responsive dystonia, dopamine-agonist responsive dystonia, with myoclonic jerks and dystonia-myoclonus
3 : Secondary Dystonia	non-genetic, develops due to environmental factors
4 : Heredodegenerative Dystonias	neurodegenerations which produce dystonia as a prominent feature, e.g. Huntington's, Wilson's, etc

Whereas the previous aetiological classification of dystonia was essentially divided into two, ie primary (idiopathic) and secondary (symptomatic), recent discoveries into the genetics of dystonia have meant that whilst the original three categories for describing patients, namely age at onset, distribution and aetiology remain, the aetiological category can be expanded to include four sub-categories as shown in Table 3 above, which the genes being defined more specifically in Table 4 below.

The gene nomenclatures have been expanded rapidly and the acceleration has greatly increased over the past year and at the recent National Spasmodic Torticollis Conference in October, 1998 in San Diego, USA, the following genes were identified and discussed.

TABLE 4 : Gene Nomenclature for the Dystonias

DYT1	Gene : 9q34.1, Auto-dominant, early-onset and limb onset
DYT2	Gene : autosomnal-recessive dystonia
DYT3	Gene : Xq13, Lubag
DYT4	Gene : a whispering dysphonia family
DYT5	Gene : 14q22, dopa-responsive dystonia, GTP cyclohydrolase l gene
DYT6	Gene : 8p21-q22, mixed type dystonia
DYT7	Gene : 18p, familial torticollis
DYT8	Gene : 2q, PNKC, paroxysmal
DYT9	Gene : 1p, CSE, paroxysmal
DYT10	Gene : not imp., PNKC, paroxysmal
DYT11	Gene : myoclonic, Auto-dominant
DYT12	Gene : 19q, Rapid Onset Dystonia Parkinsonism, reduced penetration and Auto-dominant

These will <u>not</u> be discussed any further at this stage, but essentially they are placed here to show that genetics are beginning to play a more and more important part in the evaluation of dystonia.

<u>Distribution</u>

The distribution of the dystonias, as described in Table 2, has not changed but these need to be described in greater detail as one of the reasons why dystonia is so poorly acknowledged is often due to the reason that someone with a focal dystonia does not know they have a neurological condition, even though they might know the given name of what they have. The reason for this is often due to the various names given to, in particular, the focal dystonias.

The following table shows the distribution of the dystonias as well as the names given to these various conditions, which goes to explain why these different conditions are often misdiagnosed or more often not thought to be related, outside of medical circles.

TABLE 5. Nomenclature of the Focal Dystonias.

Blepharospasm	- affecting the eyes (or specifically the eyelids)
Dystonia Dysphagia	- affecting swallowing
Focal Dystonia of a Limb	- affecting an arm, a leg or a foot (a Peripheral Dystonia)
Lingual Dystonia	- affecting the tongue
Oromandibular Dystonia	- affecting the jaw and mouth
Spasmodic Dysphonia	- affecting the larynx and speech
Spasmodic Torticollis	- affecting the neck
Unilateral Blepharospasm	- affecting one eye (or one set of eyelids) only
Writer's Cramp	- affecting the hand and fingers (a Peripheral Dystonia)

Hemi-facial Spasm is a 'dystonic type' spasm affecting one side of the face only. It has been argued that **all** cases of Hemi-facial Spasm are **not** classified as being a focal dystonia (Elston, 1997). The results of this research, to be discussed later, could argue that this is a false assumption and that a number of so-called 'hemi-facial spasms' are in fact a focal dystonia of one side of the face, or in particular one set of eyelids, but in order to satisfy current medical opinion the term 'Unilateral Blepharospasm' has been included above to differentiate between the two definitions.

Some of the above can be defined even more, for example Cervical Dystonia (which is the technically correct term for what is generally known as Spasmodic Torticollis) can be defined in four distinct ways depending on how the head moves or in what direction the head turns.

TABLE 6. The Cervical Dystonias.

Torticollis	Where the head turns either left or right
Laterocollis	Where the head tilts either left or right
Retrocollis	Where the head leans backwards
Antecollis	Where the head goes forward, so that the chin touches the chest

Therefore where someone is described as having Spasmodic Torticollis to the right, eg., ST (R), it means their head turns to the right. These definitions have been used in the script, except that Laterocollis is not generally used and a turn or a tilt is usually both just called Spasmodic Torticollis and they are not differentiated.

The Segmental Dystonias have also a number of different terminologies to describe various different forms of the disorder as shown on the table below. Remember that Segmental Dystonia was originally described (Table 2, Page 9 above) as a dystonia which affects two or more connecting parts.

TABLE 7. The Segmental Dystonias.

Axial Dystonia	Affects the trunk and neck
Brachial Dystonia	Affects an arm + trunk, both arms +/- neck or trunk.
Crural Dystonia	Affects one leg and trunk, both legs +/- the trunk.
Cranial Dystonia	Affects two parts of the cranium +/- neck musculature.

Cranial Dystonia can be sub-divided and is often given other names, eg.,

Craniocervical Dystonia	Affects a part of the head plus the neck musculature.
Meige's Disease	Combination of Blepharospasm & Oromandibular
Brueghel's Syndrome	Combination of Blepharospasm & Oromandibular

The Hemi-dystonias affect an ipsilateral arm and leg only, ie., on the same side of the body

Multi-focal Dystonia affects two or more non-contiguous parts, eg., a left arm and right leg or a right hand and the neck, etc, etc.

Another way of describing Generalised Dystonia, which is defined as a combination of Segmental Crural Dystonia and any other segment, is to say at least one leg +/- the trunk and another segment, such as the head and the neck or the eyes and a hand, etc.

PART TWO : DYSTONIA

CHAPTER 5

Previous Epidemiologies

General Review

As previously defined : " *Neuroepidemiology may be defined as the study of the distribution and dynamics of neurological diseases in human populations and the factors that affect those characteristics* " (Schoenberg, 1978) and as firstly the author was not medically qualified and secondly no one knew how many people had dystonia in the North East of England, it was necessary to undertake some basic training in epidemiology in order to start the work.

One of the important areas of research in the literature review was to define the difference between prevalence and incidence. Prevalence and incidence are related to each other as prevalence approximately equals incidence multiplied by the average duration of the disease.

This was the point at which the research should start, ie., how many people had dystonia within a given population, because " *the two most important considerations for the neuroepidemiologist in the design of studies are the representativeness of the population selected for investigation and the accuracy of the diagnoses in that population* (and) *because of ..... limited access to neurologic expertise, some individuals with the disease of interest may never seek medical care or may never be correctly diagnosed. To avoid this problem, the neuroepidemiologist attempts to identify all cases of a particular neurologic disease in a well-defined population.* " (Schoenberg, 1986).

By 1990, research had shown that there had only been a few epidemiological studies of dystonia ; in the USA (Eldridge, 1970 and Nutt et al, 1988), Israel (Korczyn et al, 1980 and Zilber et al, 1984), the Republic of China (Li et al, 1985) and Spain - restricted to gypsies (Gimenez-Roldan et al, 1988).

Even then these did not accurately reflect what was required, as " *the two most important considerations for the neuro-epidemiologist in the design of studies are the representativeness of the population selected for investigation and the accuracy of the diagnoses in that population.* " (Schoenberg, 1986). Since then there have been a number of limited studies undertaken in the USA (Risch et al, 1995), Assuit in Egypt (Kandil et al, 1994) and Tottori in Japan (Nakashima et al, 1995).

However, the only research which was specifically related to a known and defined population which was similar to that present in the North East of England was that carried out at the Mayo Clinic in Rochester, Minnesota in the USA (Nutt et al, 1988).

Results of previous epidemiologies

The only epidemiological evidence available of estimated prevalence from an epidemiological survey of dystonia (and other neurological disorders) which took place in Rochester, Minnesota, USA at the Mayo Clinic, is taken from files collected from 1950 to 1982, based on an average population from 1960 to 1979 of just over 100,000 people, where 697 cases were examined and 34 cases of idiopathic dystonia found (Nutt et al, 1988) including 11 cases of Spasmodic Torticollis (Claypool et al, 1995). This small sample with very wide confidence intervals (17.2 to 47.9 per 100,000 and 0.41 to 12.4 per 100,000 for focal and generalised dystonia, respectively at the 95% interval) formed the basis on which all future prevalence predictions were based.

Thus the prevalence for both idiopathic and symptomatic dystonia was estimated at 391 per million, or " *20,000 people in Britain ... calculated on the basis that 45% of all cases of generalised dystonia and 10% of cases of focal dystonia are symptomatic*" (Marsden and Quinn, 1990), although Marsden himself was quoted in the BMJ article as saying that 20,000 may be a "*substantial underestimate*".

The only epidemiology which had been carried out in the population within the North of England, up to relatively recent times and in anything like a similar disorder, was the epidemiology of Parkinson's Disease (PD) which had been carried out in Carlisle, Cumbria between 1955 and 1961. Here incidence had been shown to be 12.1 new cases per 100,000 population / year with the prevalence rates as of January 1st, 1961 to be 112.5 per 100,000. (Brewis et al, 1966). The average duration of PD in Carlisle in the early 1960's was 9.3 years.

It had been reported as long ago as 1984 that " *there have, as yet, been no formal community-based surveys of torsion dystonia* " but that Huntington's Disease had a mortality for US Whites of 1.6 per 1,000,000 per year (Kurtzke et al, 1984) and that Wilson's Disease had a prevalence ratio of 1.6 per 100,000 and the incidence rate was 0.2 per 100,000 per year in the Icelandic population (Gudmundsson, 1969) and a world-wide prevalence of 3 per 100,000 (Scheinberg et al, 1984).

To gauge an idea about the prevalence of dystonia, a table showing the distribution pattern of patients with dystonia is reproduced below (Fahn et al, 1988). A total of 932 patients with dystonia were seen by the Movement Disorder Group in the 13 years between September 1, 1973 and March 31, 1986 at the Dystonia Clinical Research Centre located at Columbia-Presbyterian Medical Centre, New York City in the USA.

NB : It should be noted now that over the 6 years of the epidemiology in the North East of England from 6th May 1993 to 5th May 1999, one will see a total of 937 people diagnosed, as opposed to the 932 shown above which took 13.6 years to generate and complete. Therefore it will be interesting to see the differences in a purely regional epidemiology in the North East of England as opposed to that of a large hospital in New York which takes referrals only.

TABLE 8 : Distribution of Patients at Dystonia Clinical Research Centre in New York from 01.09.73 to 31.03.86.

Category of Patient	Idiopathic Number	Idiopathic Percentage	Symptomatic Number	Symptomatic Percentage
Childhood-onset				
Focal	11	1.2	5	0.5
Segmental	34	3.7	10	1.1
Multifocal	6	0.6	3	0.3
Generalised	70	7.5	54	5.8
Unilateral	2	0.2	15	1.6
Sub-Total	123	13.2	87	9.3
Adolescent-onset				
Focal	21	2.3	3	0.3
Segmental	8	0.9	10	1.1
Multifocal	5	0.5	2	0.2
Generalised	13	1.4	5	0.5
Unilateral	3	0.3	3	0.3
Sub-Total	50	5.4	23	2.4
Adult-onset				
Focal	271	29.1	33	3.5
Segmental	150	16.1	11	1.2
Multifocal	2	0.2	6	0.6
Generalised	11	1.2	9	1.0
Unilateral	1	0.1	8	0.9
Sub-Total	435	46.7	67	7.2
Paroxysmal dystonia	17	1.8	16	1.7
Tardive dystonia			90	9.6
Psychogenic dystonia			24	2.6
TOTAL	**625**	**67.1**	**307**	**32.8**

In the above chart :-

1) The categories of paroxysmal, tardive and psychogenic dystonia are shown removed from the idiopathic and symptomatic dystonias in order to list them as separate entities.

2) Their definitions were explained in the original text and no distinction was made as to age at onset for these last three categories.

3) Three patients with diurnal fluctuations are included among the childhood-onset idiopathic dystonia.

TABLE 9 : Distribution of patients at Dystonia Clinical Research Centre based on Onset and Type Categories.

Onset	Idiopathic	Percentage	Symptomatic	Percentage
Childhood	123	20.2%	87	49.2%
Adolescence	50	8.2%	23	13.0%
Adult	435	71.6%	67	37.8%
Total	608	100%	177	100%
Type	**Idiopathic**	**Percentage**	**Symptomatic**	**Percentage**
Generalised	94	15.5%	68	38.4%
Multifocal	13	2.1%	11	6.2%
Segmental	192	31.6%	31	17.5%
Focal	303	49.8%	41	23.2%
Hemi-dystonia	6	1.0%	26	14.7%
Total	608	100%	177	100%

The North East of England (including Cumbria)

The population of the five northern counties of England, ie Cumbria, Northumberland, Tyne & Wear, Co. Durham and Cleveland, with the Mayo clinic figures extrapolated, gave an estimated 1,009 people with idiopathic dystonia in the Northern Regional Health Authority (NRHA) area. These calculations were based on the population statistics from the mid-1991 census, which were supplied by each county's Research and Intelligence Units as updated in March 1993 and are shown in the following table.

TABLE 10. Prevalence of the various forms of dystonia in the North East region based on the Mayo Clinic estimates.

County (with) : Population	GD	BL	OMD	SD	ST	WC
Cleveland : 550,100	**19**	**28**	**19**	**29**	**49**	**38**
Co. Durham : 589,900	**20**	**30**	**21**	**31**	**53**	**41**
Tyne & Wear : 1,125,600	**38**	**57**	**39**	**58**	**100**	**78**
Northumberland : 304,700	**11**	**16**	**11**	**16**	**27**	**21**
Cumbria : 483,200	**16**	**25**	**17**	**25**	**43**	**33**
Totals = : 3,053,500	**104**	**156**	**107**	**159**	**272**	**211**

Index :-

- GD = Generalised Dystonia,
- BL = Blepharospasm,
- OMD = Oromandibular Dystonia,
- SD = Spasmodic Dysphonia,
- ST = Spasmodic Torticollis,
- WC = Writer's Cramp.

As previously identified on page 2, there were currently only 143 people identified with dystonia in May 1993, at the time of the start of the survey, therefore it was obvious that there would be little point in conducting any meaningful research with only 14% (143) of the estimated population of 1009 people potentially available, particularly as even those calculations were based on a "*substantial underestimate*" (Marsden and Quinn, 1990) and there was no way of establishing any sort of representativeness of the sample unless more people, than those currently known, were correctly identified.

Based on the Dystonia Society survey (TDS, 1993), already mentioned on page 2, and the author's own personal experience, it was deduced that a high proportion of sufferers remained undiagnosed in the U.K. in general and within the North East in particular.

Therefore it was essential that, before any research could be undertaken, some method was found to identify more people with dystonia in the region than currently available, who could then be asked to participate in the author's proposed research project.

Any epidemiological survey of dystonia would, by definition, be quantitative in nature as its purpose was initially merely to identify the actual numbers of people with dystonia within the NRHA (Northern Regional Health Authority).

As previously mentioned, access to people who have dystonia is considered to be the primary difficulty in carrying out any sort of research project (Butler et al, 1998) and as the very process of identifying those people with dystonia meant first obtaining a correct diagnosis, the opportunity arose of having previously unknown (unidentified and undiagnosed) patients available for other dystonia research programmes.

As there were only 12 clinical neurologists practising in the North East at the time, the average number of dystonia patients registered at any neurology clinic was estimated to be relatively small. However it was known that, in practice and based on the author's personal experience, a couple of consultants had registered the bulk of the diagnosed dystonic patients in the NRHA area, mainly due to the advent of Botulinum Toxin therapy in the treatment of dystonia and secondary dystonic spasms.

It was at this point in the negotiations with the Dystonia Society and local medical personnel that the idea of an expanded project base was first mooted by Mr Maurice Hawthorne, who is an Otolaryngologist and Consultant Surgeon at the ENT Department of the North Riding Infirmary in Middlesbrough and who runs the Botulinum Toxin clinic based there. The author had approached the heads of the two Botulinum Toxin clinics for access to their patients and through this initial contact in February 1993, the Cost Utility Analysis (CUA) of Botulinum Toxin therapy in the treatment of dystonia (Gudex et al, 1995) was started on 6th May 1993.

Although the CUA was the initial research, it was always envisaged as a method of firstly expanding the author's knowledge of dystonia and secondly training him in general research techniques and enabling an expansion of people with dystonia into a formal epidemiology of dystonia within the North East region.

PART TWO : DYSTONIA

CHAPTER 6

Treatments

Research of the literature has shown that treatments, prior to the development of Botulinum Toxin, have proven very hit and miss regarding a stable and useful treatment for dystonia. With the exception of the use of Levodopa in the treatment of dystonia in cases of Dopa-Responsive Dystonia, the use of other drugs has proved very variable. There have been a large number of different drugs used over the years with differing results, none of which have proved particularly effective. Until the pathogenesis of dystonia is understood, the treatment of the majority of dystonia will be non-specific, aimed at the symptoms but not the cause of the disease. The treatment of dystonia was often empirical and symptomatic, but there are a group of conditions which often do respond to treatment. There also are a number of conditions which often respond to specific drug treatments, which are listed below.

<u>Conditions with a Specific Rational Therapy</u>

1. Wilson's disease :

One of the first things to be eliminated, when patients with a dystonic disorder are evaluated, is Wilson's Disease where an excess of copper is stored in the body. This is eminently treatable, with the oldest case on record being about 53 years of age.

2. Acute drug-induced dystonia :

The acute form of this disease is where a person is given treatment for sickness and within minutes of being given an injection, the eyes are turning up into the head, often with an arching of the back, called an oculogyric crisis. This responds very well to intravenously given anticholinergics or Diazepam and is an acute disorder, ie., responding well to treatment and not being long term.

3. Chronic drug-induced dystonia :

Tardive Dystonia, which has only become recognised in recent years, represents one perspective of Tardive Dyskinesia. This occurs in patients who have received long term neuroleptics, often with a psychiatric diagnosis, and may look indistinguishable from an idiopathic dystonic person, although this is a drug-induced problem. The chronic form may respond to antidopaminergics, anticholinergics, Benzodiazepines and Baclofen, although it is the neuroleptics that have caused the problem and it is not always an easy condition to treat.

4. Parkinson's Disease :

Dystonia can often occur in Parkinson's Disease. It can occur in drug-naive Parkinson's Disease, where patients who have never received treatment for the condition and may have painful dystonic inversion of the feet which might well improve on introduction of treatment. Drug-induced dystonia in the long standing Parkinson's Disease patient is also a feature and is often very difficult to treat.

5. Lesions of the Basal Ganglia :

Lesions of the basal ganglia may often be part of the cause of hemi-dystonia. A lesion or a structural cause in the opposite basal ganglia, if it is amenable to surgery, can often lead to some resolution of the symptoms.

6. Dopa-Responsive Dystonia (DRD) :

5% to 10% of patients with childhood onset dystonia, beginning in the legs, respond dramatically to Levodopa in small doses with no long term complications, which does often coexist with elements of Parkinsonism. All childhood onset dystonia cases should have a trial of Levodopa as the treatment is effective indefinitely and not associated with the long term problems that Levodopa has in treating people with Parkinson's Disease. There is often an inherited component to it but it does not always express itself and DRD is often called "Segawa's Syndrome" in some literature.

7. Paroxysmal Kinesogenic Dystonia :

This form of dystonia occurs where, if the patient moves suddenly, they will adopt dystonic postures. This can occur multiple times in a day and every time they move suddenly, they will trigger the dystonia. These cases are really quite sensitive to anticonvulsants, so drug treatment can be very effective.

8. Paroxysmal Non-Kinesogenic Dystonia :

Its colleague is not related to movement, but if the patient has a heavy meal or is anxious or stressed or drinks alcohol, they can adopt a more sustained dystonic posture for a more sustained period of time. This condition responds reasonably well to Benzodiazepines.

9. Sandifer's Syndrome :

Sandifer's Syndrome is a condition peculiar to children, where after eating a meal the child will develop borborygmi and often their head will adopt the posture of a severe dystonia. The mechanism of this is thought to be related to oesophageal reflux of acid. By doing an operation to cure the reflux, one can cure the condition.

10. Autosomal Dominant Myoclonic-Dystonia Syndrome :

This is a mixture of myoclonus, so called jerking problems, together with dystonia which responds quite well to alcohol.

Surgery and Orthopaedic Procedures

1. Ablative CNS lesions

Prior to 1950, people like Horsley (in 1900 he first treated hyperkinesias by ablating the motor cortex in a boy with severe hemi-dystonia), Bucy and Case (1939) and Klemme (in 1940 he excised cerebral cortex for Parkinsonism tremor and dystonia), Putnam (in 1938 he incised the pyramidal tract in the upper cervical cord) and Walker (in 1952 he incised cerebral peduncle in the midbrain) were doing fairly crude procedures in an effort to try and stop involuntary movements contralaterally. A lot of the early patients had Parkinsonism, or Parkinson's Disease, or post encephalitic Parkinsonism, and they had dystonia associated with it, but some of these patients also had primary dystonia and the price that all of these patients had to pay was Hemiplegia. The patients sacrificed an abnormal posture for a loss of function to a profound degree. So really it is not surprising that these are not practised anymore.

However, by 1951, Meyers started to experiment with deep lesions and showed that by putting a lesion, deep down in the Basal Ganglia, the contralateral movement disorder could be improved without incurring the terrible price to pay of hemi-paresis. He showed globus pallidus lesions could reduce contralateral dyskinesias without spasticity and weakness in Parkinsonians, but with a 16% mortality.

Cooper was trying to copy Meyers when he inadvertently cut the anterior carotid artery. He quickly finished the operation having sealed off the artery. When the patient recovered, it actually turned out that he had achieved a better result than the patients that Meyers was treating. Cooper then started performing this lesion intentionally, rather than cutting the tract, he started to ligate the arteries. Between 1955 and 1974, he did 226 chemopallidectomies and chemothalamotomies for torsion dystonia. Most patients received 1 or 2 operations, some up to 7 in total. He retired in 1976 and reported a huge series of cases, where he had injected phenol or alcohol into the globus pallidus and into the thalamus to treat torsion dystonia. Cryothalamectomy (freezing of the thalamus) was eventually the most favoured technique and Cooper reported that 70% of his patients actually gained 'some benefit'. This is an unparalleled figure in the literature although several patients paid a price by having swallowing difficulties.

By 1983, Andrew found that limb dystonia, especially hemidystonia, responded best and that lesioning the posterior portion of the ventrolateral thalamus (sensory relay nuclei) gave the best results. This operation was reviewed by Andrews most recently who showed that, in bilateral thalamotomies, there really is a huge morbidity and mortality; over half the patients are left with major problems speaking and swallowing, compared with only 11% for the single side operation. Andrews also pointed out that limb dystonia, and hemidystonia particularly, responds best and lesioning the particular part of the thalamus, which is a sensory relay station, and passes on message to the brain, generally gives the best result.

2. CNS Stimulation

Cerebellar stimulation has been attempted by Siegfried and Hood but by 1982, Waltz's cervical cord stimulation claimed to benefit 72% of 110 cases with generalised and cervical dystonias, however further studies fail to substantiate this claim. Peripheral surgery for torticollis was first attempted in 1891 by Keen, where he divided the first three cranial nerves extraspinally. Then in 1915, Taylor attempted the first intraspinal approach which showed benefit in up to 80% from rhizotomy of anterior roots of C1-3 bilaterally but with a high complication rate.

Waltz in 1982 claimed amazing results by stimulating the cervical cord. He claimed that 72% of his patients in a large series did very well. After more critical evaluation, this procedure can no longer be recommended. Now peripheral surgery takes all sorts of forms. Basically it involves tackling the nerves and muscles extraspinally at the site of the problems.

Of the other problems that have been tackled, probably the main one is Spasmodic Torticollis. The idea has been to cut as many nerves as possible without leading to unacceptable side effects. A Rhizotomy sections cervical roots as they emerge from the spinal cord. Many patients undergoing this operation became quadriplegic, whilst others developed swallowing difficulties.

By 1989, Bertrand and Molina-Negro were doing selective extraspinal denervation using intra-operative nerve stimulation to identify branches and claim 87% success in a relatively small series by attacking the specific muscles that are involved in Spasmodic Torticollis.

3. Physical methods and behavioural modifications

There are certain things that make dystonia a lot worse, ie stress, lack of sleep, having no one to talk to, isolation and the feeling of being misunderstood. There are a lot of patients where there can be a huge delay in making a correct diagnosis and this can lead to a great deal of misery and can exacerbate the problem quite markedly. Doctors often when speaking to patients with dystonia are adopting some form of psychotherapy to make people realise that they are not alone and although they do not know the cause, they are trying to do something about the problem.

4. Relaxation therapy and self-hypnosis

Relaxation therapy can have some limited benefits as can self-hypnosis.

5. Sensory Biofeedback methods

This is where people can have an EMG or muscle monitors put on their muscles feeding back to themselves, so that they can see when a muscle is becoming over-active and they can adopt strategies where they can relax and make the activity die down, so that the muscle becomes less over-active. As will be seen, this has not been popular with North East dystonia patients with only 6 having tried this technique with no satisfactory results.

6. Mechanical devices / braces

These have had a varied response in the past. Generally, ill fitting braces and rigid appliances, which try and constrain the movement of dystonia, can actually be positively harmful. However imaginatively designed devices can be very useful. People who use sensory tricks for Spasmodic Torticollis or Retrocollis certainly have used devices where, with an appropriate brace and just a lightly touching device in the occipital area, can actually receive a very slight sensory stimulus and prevent the patient from showing too much of a dystonic pattern.

Drug Treatments

Systemic pharmacotherapy can overall give benefit to 30-40% of dystonic patients. Initial treatment is guided by use of drugs with low potential for adverse side effects and the anatomic distribution of the dystonia. About 5% of patients with any form of idiopathic dystonia may experience a spontaneous improvement or resolution of the movement disorder, this is most likely in the first five years and relapses are common.

1. Anticholinergic Agents

Drugs like Benzhexol or Artane may help approximately 50% of children and 40% of adults and very large does (60-100 mg) may be needed, as opposed to people with Parkinson's Disease who may be given just 6 mg to help with their tremor. Improvement is independent of age of onset, age at starting the therapy, severity of dystonia at start of the therapy and aetiological diagnosis. However improvement is dependant on dosage, side effects (more prominent in adults and a function of age not serum level), peripheral (dry mouth, urinary retention, constipation and blurred vision may need co-administration of philocarpine and pyridostigmine as muscarinic agonists) and central (may cause drowsiness, confusion, memory impairment and hallucinations).

The improvement is often independent of the facts listed above and is dependant upon the dosage, although that is very loosely related in the big trials done and some people respond to a much lower dose. The method is to start on a given low dose and to build up very slowly because there may be a gap of several months before people respond to a particular dosage. Certainly, in the series done by Professor Marsden, a large number of people, 65% of adults, had side effects and a large number of them were so bad they had to stop taking the drug.

2. Dopamine Agonists

This includes Levodopa, which has already been discussed. There is a query about whether there is a sub-set or sub-group of the torsion dystonias, because the same drug can actually worsen symptoms in about a third of adult cases. 5% to 10% of childhood onset dystonia beginning in the legs respond to low dose of Levodopa, however it may worsen symptoms in up to 35% of adult cases. Greene et al reported in 1987 that 12% of 41 patients who failed anticholinergic treatment responded to dopamine agonists eg Lysuride. This is the same sort of drug as Bromocriptine, a direct acting Dopamine Agonist.

3. Dopamine antagonists

These are the exact pharmacological opposite to Dopamine Agonists, yet 20% of people will respond to these drugs. No individual has responded to both dopamine agonists and antagonists suggesting again multiple pathophysiological mechanisms may be involved. These drugs are either Dopamine Depletors, eg Reserpine, Tetrabenazine, where they act to actually get rid of the Dopamine from the nerve before it is released or they are Dopamine Receptor Blockers, eg Pimozide and Haloperidol. They latch onto the receptors, post synaptically, and stop the Dopa from being active.

Response is variable : generalised / segmental = 0-78%, cranial dystonia = 3-34%, spasmodic torticollis = 9-46% and writer's cramp = 0%, which just shows that it is very difficult to have a consistent scale of assessment and assessment criteria vary. The side effects, certainly with Tetrabenazine, a dopamine depletor, are quite significant depression and that is what is so limiting. In addition, with the Dopamine Receptor Blockers, Parkinsonism may also be a problem.

4. Other Drugs

Inadequate dosing and inadequate duration of treatment may make a drug appear less effective than it really is. Here is the sort of treatment where people may rather idiosyncratically respond. These are not the first line drugs because they are often, in many cases, not very effective.

Baclofen : The physiotherapists deal with this drug as a spasmolytic. It may be effective in about 20% of generalised dystonia and is often successful in occasional segmental dystonia.

Benzodiazepines : There is no evidence one is better than the other. Clonazepam is the drug that people tend to have used a lot with 15% of patients responding. Carbamazepine, an anticonvulsant, helps between 10% - 11% of patients.

Alcohol : With the use of alcohol, intravenous infusion may help to reduce severe drug-unresponsive Spasmodic Torticollis, although this is clearly not a practical treatment to be walking around with an alcohol infusion running.

Lithium : This is a very toxic drug, has a limited use and may help blepharospasm with oromandibular dystonia, ie., Meige's Syndrome.

5. Triple Therapy

This is used in children and especially adults with severe (sometimes life-threatening) dystonia not responding to monotherapy trials, where the muscle spasm is so severe there is breakdown of muscle, increased body temperature, exhaustion, respiratory difficulties. It comprises of Tetrabenazine (75 mg), Pimozide (up to 12 mg daily) and Benzhexol (up to maximum tolerated and titrated against Pimozide). In a series reported by Marsden et al in 1984 this helped 1 of 2 children and 9 of 12 adults with severe dystonia

In conclusion, a small number of patients with dystonia may be helped with a specific treatment for a specific cause of the condition. Many patients with generalised dystonia may be helped by systemic pharmacological therapy. The treatment is empirical and roughly follows a 'ladder' scheme where the 'least toxic-most likely to be successful as monotherapy' is at the bottom and the 'most toxic-polytherapy' is at the top. In drug treatment the secret is to take a single drug to its tolerated limit (in most cases) and not to increase doses too quickly. All childhood and adolescent onset cases of dystonia should be given a trial of Levodopa treatment first. Centrally placed CNS lesions may be very effective in selected cases but surgery is regarded as the 'last gasp' treatment in most centres. Peripheral surgical procedures have now been supplanted by Botulinum Toxin therapy.

The Introduction of Botulinum Toxin Therapy

The advent of Botulinum Toxin therapy has revolutionised the treatment of dystonia. In May 1993, there were only 149 people who were getting Botulinum Toxin therapy in the North East of England, whereas by May 1999, 6 years later, the number was 722, a 385% increase or an average increase of 64.1% per year.

Botulinum Toxin injections can relieve the symptoms of dystonia and this is now considered as the best treatment for many adult onset focal dystonias such as blepharospasm (Grandas et al, 1988), writers cramp (Cohen et al, 1989) spasmodic torticollis (Stell et al, 1988) and spasmodic dysphonia (Ludlow et al, 1989) and it was this aspect in the treatment of dystonia which was the primary area covered originally by the research project when it commenced in 1993 (Gudex et al, 1995, 1997 and 1998).

" *... research into dystonia is more active now than ever before. As a result, physician awareness has increased greatly. A once obscure condition which was usually thought to be psychological in origin is now recognised as a relatively common reason for referral to movement disorder clinics. Although treatment remains difficult and often frustrating .... these factors allow individuals who care for these unfortunate patients to provide them with some hope that relief from their considerable disability eventually may be realised.* " (Riley and Lang, 1988)

There is a clinical syndrome called 'botulism', where a bacteria 'Clostridium Botulinum' overgrows in the gut. There was an outbreak in Preston, in the North West of England a few years ago, where cans of Hazel Nut puree which, at their site of origin, had become infected with this anaerobic bacteria. Being in an enclosed can, they came to multiply and survive very nicely. Then they were put in pots of yoghurt which were then sealed. Unfortunately this caused a large outbreak of clinical botulism. The cases presented with difficulty with swallowing, difficulty with speaking and respiratory problems. They required intensive care and management, but with appropriate treatment, including antibodies and ITU type approaches, they recovered quite well. These toxins, which are produced by these micro-organisms, are potent neurotoxins and very small quantities of this agent are needed to cause the above type of symptoms.

There are 7 serotype separate chemicals, which this group of bacteria produce. In terms of clinical use, it is serotype A which is currently used, although other serotypes are being investigated for their clinical usefulness. It is such a potent toxin that absolutely minuscule quantities of this agent are used. It effectively weakens the muscle into which it is injected and it can relieve spasms and involuntary movements. The quantities that are used do not usually cause profound weakness. There is only a percentage of the muscle motor end plates that are affected and thus it is able to dampen down the over activity, rather than causing actual weakness. One of the fundamental problems is it is an organic compound and that a proportion of people who are treated can develop antibodies. The effect of the antibodies is that they neutralise the effect of the toxin.

It effectively controls dystonia for between 8 to 12 weeks. It is a localized treatment with minimal systemic effects. It can be used on an out-patient or in-office procedure and it controls or relieves symptoms in up to 98% of patients. Everyone who is involved in treating dystonias with Botulinum Toxin is simplifying their injection techniques and as the numbers have increased so has the expertise in administering the injections. The following chart gives where the injections are usually placed.

TABLE 11 : Injections Sites for Spasmodic Torticollis

Headneck position	**Muscles involved**
Torticollis	SCM contralateral to the direction of turning, Ipsilateral trapezius and/or levator scapulae
Laterocollis	Ipsilateral splenius capitis, trapezius, scalenus and / or levator scapulae
Retrocollis	Bilateral splenius capitis, semispinalis and longissimus capitis
Anterocollis	Bilateral SCM, scalenil submental complex

In ordinary torticollis, the Sterno-Cleido-Mastoid (SCM) is injected, contralaterally to the direction of the turning and either the ipsilateral trapezius or the bilateral trapezii, depending on the relationships of the movements around the neck. The splenius capitis is often injected as well because that muscle is often active in Spasmodic Torticollis. For Retrocollis, bilateral splenius capitis muscles are injected and it is slightly deeper and more difficult to isolate these muscles. Anterocollis is very difficult to treat, requiring bilateral SCM injections. The scalenil do cause head problems on occasions.

PART THREE : METHODOLOGY

CHAPTER 7

Awareness Raising

As can be seen from the evidence in the literature review, no one knew how many cases of dystonia existed in the U.K. and, at the time of the start of the research in May 1993, there were only 143 people with dystonia known in the region. Therefore the initial problem was obviously going to be to see if sufficient numbers of people with dystonia could be correctly identified and this was predicted to be a mammoth task. Therefore a series of awareness raising campaigns were launched from 1993 to 1998, ie., during and throughout the entire research process. The author was involved in giving all or part of a number of separate lectures during this period as follows :-

<u>Twelve Awareness Raising Events in the UK</u>

1. **14th July 1993** : Dystonia Awareness Raising Day held in the Lecture Theatre at Hunters Moor Hospital, Newcastle, UK.

2. **20th April 1994** : Dystonia Awareness Raising Day held in the Lecture Theatre at Hunters Moor Hospital, Newcastle, UK.

3. **31st March 1995** : An International Workshop on the use of Botulinum Toxin for the treatment of dystonia held at Manchester University, UK.

4. **9th April 1995** : The Dystonia Society Conference at Warwick University, UK.

5. **24th - 28th June 1996** : The First World Congress in Neurological Rehabilitation held in Newcastle, UK.

6. **September 1996** : The Awareness Raising Campaign in Darlington, UK.

A total of 101,766 people living in 45,383 houses within the postal districts of Darlington, Co. Durham, ie DL1, DL2 and DL3, were given a pre-addressed leaflet describing dystonia and asking them to send it back to the author, if they felt they exhibited any of the symptoms described on the leaflet. A total of 41 people replied who were then visited and examined by a qualified neurologist.

7. **30th January 1997** : A Dystonia Orientation Day at Hunters Moor Hospital, Newcastle.

8. **31st January 1997** : A Dystonia Orientation Day at the North Riding Infirmary, Middlesbrough.

9. **23rd to 27th March 1997** : The "Health and the Musician" Conference held at York University.

10. **28th May 1998** : "The extended role of the nurse in a Botulinum Toxin clinic " at the North Riding Infirmary, Middlesbrough.

11. **22nd September 1998** : The North Devon District Hospital, Barnstaple, Devon.

12. **3rd December 1998** : A Dystonia Study Day held in the Conference Room at Hunters Moor Hospital, Newcastle.

Four Awareness Raising Events abroad

13. **15th June 1995** : The Botulinum Toxin Conference in Munich, West Germany.

Two poster presentations were made at the above international conference. One was entitled " The social and economic implications of dystonia " and has since been published in the European Journal of Neurology ; 3 : 79. The second was entitled " An epidemiological survey of dystonia in the North East of England " and was since published in the European Journal of Neurology ; 3 : 28.

14. **9th - 11th October 1996 :** The Third International Dystonia Symposium held in Miami, Florida, USA.

Two lectures were given, which have since been published as chapters and printed in Advances in Neurology, Volume 78 : Dystonia 3. They are " The Epidemiology of the Primary Dystonias in the North of England " given by Dr Phil Duffey and " The Socio-Economic Implications of Dystonia " given by the author.

15. **8th and 9th November 1997** : The National Spasmodic Torticollis Association Symposium held in Nashville, Tennessee, USA.

A joint lecture with John Whitaker, the Outreach Dystonia Nurse Practitioner, was given. The author's lecture was entitled " Some Social and Economic Implications of Spasmodic Torticollis in the North East of England. "

16. **7th and 8th November 1998** : The National Spasmodic Torticollis Association Symposium held in San Diego, California, USA.

A repeat of the previous year's series of lectures with John Whitaker, the Outreach Dystonia Nurse Practitioner. The author spoke again on "Some Social and Economic Implications of Spasmodic Torticollis in the North East of England."

PART THREE : METHODOLOGY

CHAPTER 8

Data Collection and Instruments

Data Collection

Only three sources of dystonic patients were initially available to the author in 1993 :-

a) the register of Dystonia Society members in the northern region, then defined as the five northern counties of Cumbria, Northumberland, Tyne & Wear, Co.Durham and Cleveland, which had the same boundaries as the NRHA (see note below).

b) the patients registered at the Movement Disorder Clinic of Hunters Moor Regional Rehabilitation Centre (HMRRC) in Newcastle.

c) the patients registered at the Botulinum Toxin clinic of the ENT Department of the North Riding Infirmary (NRI) in Middlesbrough.

Note : By April 1994, the NRHA's boundaries had been changed and the southern part of Cumbria was removed and Yorkshire was added and the Northern and Yorkshire Regional Health Authority (NYRHA) was formed. Henceforth all epidemiological research into Primary Dystonia was restricted to the four counties of Northumberland, Tyne & Wear, Co. Durham and Cleveland. Research into any social or economic implications was not however restricted geographically in the same way.

Having established the number of people with dystonia registered within these three primary sources, the rest of the research was initially carried out by a combination of interview, questionnaire and research through the clinical notes of the subjects at the two medical venues. The only criteria used in initially selecting subjects for the study was their referral to, or participation in, these two clinics administering Botulinum Toxin (Bot. Tox.) injections in the area. Subsequently a third Bot. Tox. clinic at the Sunderland Eye Infirmary (SEI) was used from 1995 onwards.

However applications were made (and approved) to the Local Research Ethics Committees (LREC) in January 1994, once the CUA fieldwork had been completed, to expand the ESD research in an attempt to identify further previously undiagnosed subjects. This was accomplished by dividing the data collection into distinct phases.

The first phase included personally attending the relevant clinics at HMRRC, NRI and SEI and interviewing **all** the patients attending. A very small number of patients were screened out of the study from the clinics at NRI and SEI, if their injection of Botulinum Toxin was post ophthalmic operation and non spasmodic. In particular, those patients who did not have any dystonic spasms and where either M.S. or Spasticity patients were removed from the lists.

All clinical data was collected and processed on a Clinical Information Form (CIF) and no verification of diagnosis was made until the differentiation between primary and secondary dystonia and spasms due to other neurological conditions was confirmed by the Neurological Registrar. The records of other hospitals in the area were reviewed and the examination of case notes was not restricted to those coded for dystonia. The review also included case notes coded for any unspecified disorder or movement gait not attributed to a specific disease.

An example of how detailed this work became and how this complicated procedure was accomplished can be seen by a quote from the Darlington Memorial Hospital (DMH) study (Butler and Duffey, 1996b), below :-

" a total of 20 visits were made from 8th February to 8th August 1995 during which time all 665 patients who had attended the DMH neurology clinic from 4th September 1991 (when computer records were commenced) to 29th March 1995 (when ICD 10 codes were introduced) were screened, a total of 4.5 years. A total of 10 primary and 10 secondary dystonia cases were identified together with a further 4 cases of hemi-facial spasm from the researcher's coding (ICD 9) of all 665 cases."

During 1995, the Sunderland Eye Infirmary (SEI), Sunderland General (SGH), Middlesbrough General (MGH) and the Dryburn Hospital (DHD) in Durham were also visited and varying degrees of success accomplished further identification of dystonic patients. For example, all cases of dystonia attending SEI have since been identified and staff continue to monitor all new patients attending their Botulinum Toxin clinic.

1996 saw the expansion of this programme to include continuous monitoring of all neurology clinics in the North East to establish all new patients attending, principally the Royal Victoria Infirmary (RVI) and the General Hospital (NGH) in Newcastle.

Liaison with the Family Health Services Authority (FHSA) in South West Durham established a database of the 310 individual doctors practising in County Durham. A series of GP Focus Groups established the best methodology of contacting and recruiting GP's onto the ESD. Dr Phil Duffey, at that time Hon. Senior Registrar at the RVI, undertook to contact those within the Darlington catchment area, ie with a Post Code DL1, DL2 or DL3, and visited them during 1996.

After discussions with several epidemiologists and other experts in the field, it was decided that Darlington, which is a town of 101,766 people, should be the subject of an intensive campaign in 1996 to establish the prevalence of dystonia within its known population. This was achieved by a combination of all the above phases, with the added impetus of two other distinct campaigns, run in conjunction with TDS throughout the North East. The first involved a publicity campaign whereby all the local media, including radio, television and the press were contacted and persuaded to run a short but intensive publicity campaign about the Epidemiology in general and diagnosis of those outside the net from the previous stages in particular, ie those as yet undiagnosed or not registered with a neurologist. This type of advertising has proved very successful and has resulted in some spectacular successes which will be discussed later in the thesis.

The second campaign was a mail shot of all 45,383 homes in the Darlington area, in which a short but effective leaflet asked householders to identify a number of specific muscular spasms, from which the screening process then commenced to diagnose all those potential people with a dystonic spasm. This took place during the autumnal months of 1996 and enabled the research team to attempt 100% validation within a relatively small geographical area. Finally, the research team attempted to identify any other dystonic subjects through tracing a number of known medications back from the subscriber / dispenser to the purchaser. This inevitably threw up a number of other neurological disorders which required screening out, but this, the final and most difficult stage, was effectively completed by the end of 1998. The ESD will now continue, using a number of different sources, at least until 2003 and hopefully beyond.

Instruments

The appendices include copies of all the actual questionnaires used in the research study and includes a number of documents used initially to gain informed consent from the patients, however it is important to realise that a number of different methodological techniques have been used throughout this research study and these have often been combined to produce the overall results.

All questionnaires were initially numbered from 1 through to 9 inclusive, however this was changed from 01 through to 99 to enable other instruments within the same area of study to be inserted, as required. The questionnaires used are listed below for reference only and detailed under their own individual sections.

Questionnaire No's 01 to 13 = Quality of Life and Health State Questionnaires

Chapter 9 outlines these questions in full.

Questionnaire No. 20 : Demographic and Dystonic Profile Questionnaire.

The questions were designed by the researcher and were agreed by the Dystonia Society's research sub-committee in March 1993 (Focus Group 1993). A postal version of the questionnaire has been used where the patient lived outside the NYRHA area, ie the West Indies, the Western Isles and London to name but three participants.

The appendices show the actual demographic questions and answers sheets used in the interview, which usually took place in the patient's home. It was soon realised that the clinic was not the best place for the interview to take place as the time available was too short and the circumstances of the clinic often too traumatic for reliable and in-depth answers. The questionnaire was designed to obtain purely demographic information as well as provide a demographic and dystonic profile of each subject. All the information was of a purely factual nature and therefore required no validation, except that in a number of areas, the patient's memory of events was verified by reference to, and inspection of, their clinical records, case notes or hospital files.

In total, 473 interviews took place over a 4 year period. The questions were divided into relevant sections as follows :-

a) *Demographic data*, including social and economic information.

Geographical location - Post Code for determining county, district and postal sector.
Age - date of birth, confirming that the Clinical Information Form (CIF) was correct.
Gender - male or female. There were no specific questions regarding ethnicity.
Marital status - noted single, married, divorced, separated, cohabiting or widowed.
Number and gender of children, including eldest's and youngest's year of birth.
Social Income Group - researcher's placement based on occupation and status.
Current Income - patient only (family income was noted where no income existed).
Number, range and amount of any benefits, allowances, pensions, etc.

b) *Dystonic Profile*

Current (and any other) Hospital Registrations - compared with CIF.
Travel time and method to and from current clinic - of patient and partner / friend.
Date (as near as possible) of Onset - subjective which was then compared with CIF.
Date (as near as possible) of Diagnosis - which was then compared with CIF date.
Clinical Diagnosis (by interview and observation) - confirmed with CIF diagnosis.
Dystonic type - confirmed with CIF diagnosis from clinical records.
Aetiology - mainly from CIF, obtained from clinical notes.

c) *Other Medical Information*

Possible trigger - subjective opinion based on past events
Family history - in particular familial dystonia or other neurological disorders.
Current medication, dosage and frequency - potential side effects.
Comorbidity - interview with subject, confirmed with CIF.
Previous treatments - from subject's memory, confirmed with CIF.
Side effects from previous treatments - including duration.
Botulinum Toxin injections - number, frequency and side effects.

Questionnaire No. 21 : TDS Questionnaire.

These were a number of questions specifically regarding the Dystonia Society, which were designed to be gradually omitted if the subject was not a member or had not heard of TDS before. They were designed to obtain factual information on :-

First contact with TDS - where and how
Contact with other people with dystonia - frequency, members or not
Possibility of starting a counselling service (which was eventually started in 1995)
Publicity - where first heard of TDS to measure effectiveness of awareness raising
Membership - current, dropped out, etc., plus other voluntary organisations
Which SHG - NE, Cumbria, Yorkshire, others in UK, or on the mailing list
Newsletter - most read part and why
National and local aims - open ended questions about the future.

Questionnaire No. 25 : Demographic Changes Questionnaire.

This questionnaire was administered at the end of the study (sent out with No's 80 & 81) to each subject to determine any changes, which were then included as part of the SPSS Coding for Questionnaire No 20, with the changes noted. It was designed to form part of each subjects' demographic profile and related specifically to those areas of factual information which could possibly have changed during the period of the study, ie ;-

a. Employment
b. Income and / or Benefits
c. Time off work, if appropriate
d. Change of hospital and method of travel or distance to and from the new clinic.

NB : Any changes in dystonic spasms were noted as part of the ESD on the CIF during the study. Of the 199 active participants in the CUA, only 9 (4.5%) had changed clinic during the period 1993 to 1994. With one exception, which is related to a geographical relocation, these were due to a referral from the general Movement Disorder clinic at HMRRC to the specific treatment available for Spasmodic Dysphonia and Oromandibular Dystonia at the ENT clinic at the NRI in Middlesbrough. However by the end of the study the number of this type of hospital change had risen to 23, but 258 No 25 Questionnaires were completed in total, noting other changes during the study period.

Questionnaire No. 30 : Clinical Profile of Dystonia Questionnaire.
Questionnaire No. 40 : Torticollis Questionnaire. (attached to Questionnaire No. 30)
Questionnaire No. 50 : Living with Dystonia Questionnaire : A Psychological Profile.
Questionnaire No. 60 : Impact of Dystonia Questionnaire.
Questionnaire No. 70 : Primary Carer's Questionnaire.

Chapter 11 details all of these questionnaires in full.

Questionnaire No. 80 : Environmental Questionnaire.

This questionnaire, as designed by the research team, was a result of all the questions which remained unanswered after the first three years of the research project. A number of specific questions, which related to areas previous unasked, had cropped up during demographic profile interviews with the patients and had been noted by the author or the Neurological Registrar.

The whole environmental issue had been raised by a TDS member who had developed an interest in the effect that environmental substances had on her dystonia, also a number of patients had indicated, during their interviews, that certain foods made their spasms worse. It was administered at the very end of the research study and was sent to every participant who had been registered in the ESD. A total of 300 completed questionnaires were returned.

The questionnaire was divided into 7 sections :-

1. *General Questions* - eg., favoured hand / foot, thyroid disease, life patterns.
2. *Effect on spasms* - of smoking, gas, sound, light, water, different actions, etc.
3. *Chemical Exposure* - 6 dichotomous variables
4. *Allergic reactions* - 17 dichotomous variables
5. *Food and drink* - listed are most types of food and drink
6. *Other Environmental substances* - natural inhalants and chemicals
7. *Final Comments* - why some days are good and others are bad ?

Questionnaire No. 81 : Diagnostic Questionnaire.

This questionnaire was designed by Dr Phil Duffey, who was at that time Hon. Senior. Registrar in Neurology at the RVI, as a vehicle to determine which and how many medical specialists had been seen by the subject before a correct diagnosis had been made. Remember the evidence that " *66.7% of all sufferers needed at least 5 consultations before diagnosis and 65.7% were misdiagnosed at some stage.* " (TDS, 1993)

In Dr Duffey's own words on the questionnaire itself : " *I hope that the information you have provided may make the diagnosis of future patients with dystonia easier. Dystonia can be very difficult to diagnose and it is not my intention to imply criticism of those involved in your particular case. Hindsight is a wonderful tool !* "

It had been discovered that the information was not always available, nor accurate, in the patient's clinical notes. This instrument was also used to determine how many and which alternative treatments had been sought before, during or after normal medical therapy. This questionnaire was issued with Questionnaire No. 80 at the end of the study, but a total of 361 were returned, 61 more than Questionnaire No. 80.

Finally the date of this questionnaire was also used to determine the length of participation in the study and the duration of each subject's personal involvement.

Questionnaire No's 91-94 : Dystonia Nurse Practitioner Project.

This series of questionnaires were designed by the DNP research team, consisting of the author, John Whitaker, the DNP, and Professor Mike Barnes. A total of 86 x No 91, 77 x No 92, 76 x No 93 and 76 x No 94 were eventually completed.

Although this thesis will not duplicate the work carried out under the DNP Project, it is necessary to understand what was asked and why. Therefore Chapter 13 will duplicate a number of points already published, however the questionnaires themselves were designed as part of the Monitor / Evaluator's role in monitoring and evaluating the work carried by the Dystonia Nurse Practitioner during his two year contract.

These will be described in detail under Chapter 13.

Clinical Information Form (CIF).

The Clinical Analysis Form (CAF) was designed and used by the researcher and the Neurological Registrar to record certain details from each patients' clinical records or hospital notes for the CUA research. It had space for up to 11 sets of questionnaires matched to the injection date, including Bot. Tox. dosage and the number of injection sites. All data was then transferred onto SPSS. After cleaning the data by frequency analysis, any anomalies were rechecked with data on the CIF or CAF. If required, the files were also re-examined to check the accuracy of any particular entry.

The Clinical Information Form (CIF) has been modified from the CAF, removing the need for detailed Bot. Tox. injection information in favour of the information to be used in the ESD generally. It has space for basic demographic information, including confirmation of DoB, as well as the patient's exact clinical diagnosis with dates of onset and diagnosis. Details of comorbidity and current medications were also obtained from the records, if available. Both forms are shown in the appendices and the CIF is still being used in the ESD research project as administered by the author and funded by a grant from the National Lottery Charities Board (NLCB).

Validation

Validation of all the survey data took several forms :-

1. Each case file (once identified) was examined by a suitably qualified neurologist (Dr Phil Duffey) to ensure that a correct diagnosis had been made by the author. Where necessary a personal visit was made by Dr Duffey to the subject's home.

2. A differentiation between primary and secondary dystonia and other neurological movement disorders was made by the author and confirmed by Dr Duffey.

3. The diagnostic data was then presented to an independent source for verification and feedback, usually Mr M.R.Hawthorne or Professor M.P.Barnes.

4. Repeated application of the instruments (questionnaires) determined their reliability, enabled an assessment of change over a defined period of time and allowed comparison with both internal and external control subjects.

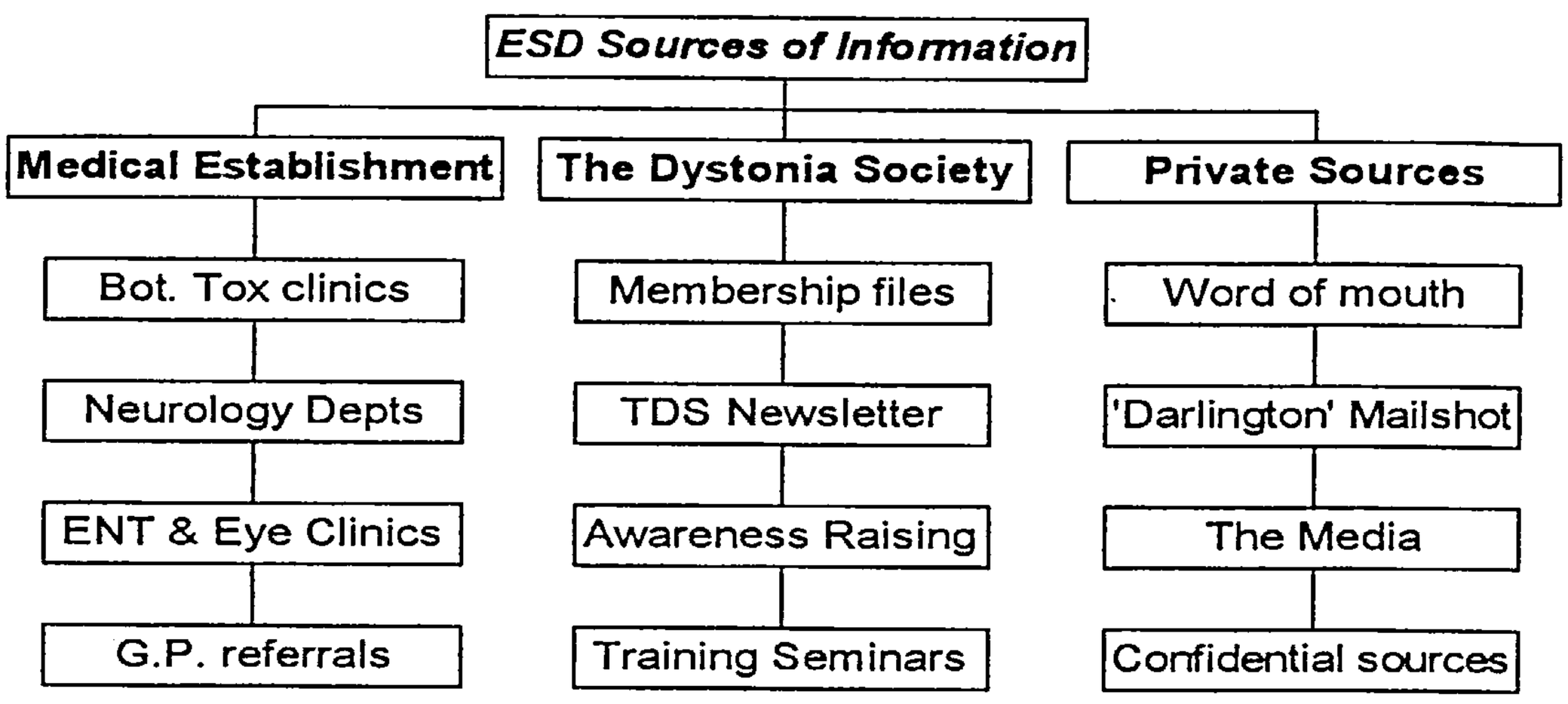
5. External controls (not receiving Bot. Tox. therapy) were matched in age, gender and type of dystonic spasm from the Dystonia Society membership. A small number of controls were selected to participate who were totally non-dystonic, but who matched other similar dystonic symptoms, ie., Generalised MS, Ulnar Nerve Palsy, Hereditary Spastic Paraplegia and Carpel Tunnel Syndrome.

   In the CUA, external controls completed n.l.t. 5 questionnaires, relating to two full treatment cycles, as did full participants in the study. The other non-dystonic controls completed all the remaining questionnaires for eventual comparison with similar dystonic symptoms such as Generalised Dystonia, Arm Dystonia, Leg Dystonia and Writer's Cramp respectively (see 5 above).

6. The technique known as capture-recapture (Laporte, 1994) was used to cross reference each new subject and check if he or she had already been identified from other sources.

The total number of sources was then noted, as below :-

CHART 12. Sources used in correctly identifying and diagnosing each subject.



PART THREE : METHODOLOGY

CHAPTER 9

The Cost Utility Analysis

This chapter relates specifically to the combined 'Quality of Life' and 'Health State' Questionnaire, which was numbered from 01 to 11 in the Cost Utility Analysis (CUA) of Botulinum Toxin therapy and was then numbered from 12 to 13 in the Epidemiological Survey of Dystonia (ESD). There was also a final section at the end which related specifically to Dystonia Self-Perception questions.

The CUA took place from May 1993 until June 1994, whereas the ESD was continuous until the final date of May 1999 and related to a total of 6 years continuous study, although only a total of 409 subjects completed these questionnaires in the ESD at the beginning and 319 subjects at the end of the study. 199 subjects completed 1,243 questionnaires in the CUA, thus a grand total of 1,772 of these questionnaires were completed and returned during the duration of the study.

During the CUA, this instrument was used as a postal questionnaire issued at the beginning (beg : Q.01), middle (mid : Q.02) and end (end : Q.03) of each patient's injection cycle, roughly every six weeks. During this particular project, the minimum number of cycles required was 3 (ie 7 questionnaires repeated), but a number of patients completed up to 5 cycles (ie 11 questionnaires repeated) as shown below :-

TABLE 13. Sequence of CUA questionnaires used during injection cycles.

Cycle	Q.01	Q.02	Q.03	Q.04	Q.05	Q.06	Q.07	Q.08	Q.09	Q.10	Q.11
1st	**beg**	**mid**	**end**								
2nd			**beg**	**mid**	**end**						
3rd					**beg**	**mid**	**end**				
4th							**beg**	**mid**	**end**		
5th									**beg**	**mid**	**end**

During the ESD, the same instrument was issued to all consenting subjects on entering (Questionnaire No 12) and at the end of the study (Questionnaire No 13) and the time between completing these questionnaires varied from 0.5 to 3.5 years.

The EuroQol Questionnaire : Questions 01 to 06.

This instrument has been used in several member states of the European Community and results can be compared with other research projects within the EEC. It was used in this study as a direct result of the collaboration with the Centre for Health Economics at York University. During the CUA it has proved to be very compatible with the SF36 (Butler et al, 1995), as has also been demonstrated by other research projects (Hollingworth et al, 1995).

The use of generic health status measures is desirable because they enable comparisons of results from treatment of one patient group with those from treatment of other patient groups or from the general population (Guyatt et al, 1991). The EuroQol (EuroQol Group, 1990) is a generic measure that can generate an overall index of health-related Quality of Life (HR-QOL), which can be used in a Cost Utility Analysis (Hurst et al, 1994 and Brooks, 1996). When completing the EuroQol instrument, respondents describe their own health on 5 dimensions, each with 3 levels of severity.

The 5 dimensions are :-

1. Mobility
2. Self Care
3. Usual Activities
4. Pain or Discomfort
5. Anxiety or Depression

The 3 levels of severity are none (1), moderate (2) and severe (3). The 5-dimensional and 3-level structure of the EuroQol classification describes 243 composite health states. The social tariffs used here were elicited from a representative sample of the UK population (n = 2997). Two sets of such health states preferences, the so-called 'social tariffs', were available. One tariff - the 'interim UK tariff' (MVH, 1994) - is based on the visual analogue scale (VAS) method of valuing health states and the other - the 'A1 tariff' (Dolan et al, 1995) based on the full study population data and a 10 year duration - is the time-trade-off (TTO) method (Dolan et al, 1996). This method requires respondents to choose between two certain health profiles : either to live in a less-than-full health state for a defined period of time (in this case 10 years) and then die, or to live in full health for a period of 10 years and then die. The period of time in full health is changed until the respondent is indifferent between the two options. In both tariffs, health state preferences range from 100 (full health) to 0 (death), with negative scores for states considered worse than death (Gudex et al, 1997).

<u>The SF 36 Health Status Questionnaire</u> : Questions 07 to 16.

The anglicised version has been previously used in various surveys in the UK and the results can be compared to a norm (Jenkinson et al, 1993). The project's use of SF36 was registered with the International Resource Centre for Health Care Assessment in Boston, Massachusetts, USA on 26th April 1993.

The instrument was conceptualised as a " *generic measure of health concepts relevant across age, disease and treatment groups. It provides a comprehensive, psychometrically sound and effective way to measure health from the patient's point of view by scoring standardised responses to standardised questions* (Ware and Sherbourne, 1992). According to the authors of the instrument: " *standardisation of content and scoring is what makes interpretation of the SF 26 scales possible. There are at least two good reasons to adhere to the standards of content and scoring described in this manual. First, they are most likely to produce scores with the same reliability and validity as those reported here and in other Medical Outcomes Study (MOS) publications. Second, comparisons of results across studies are made possible to the benefit of all who use these content and scoring standards.* " (Ware et al, 1993)

The SF36 (ie short form with 36 questions) is designed to measure eight health concepts and a self reported health transition. The standardised scoring system (Ware, 1988) yields a profile of eight scale scores as follows :-

PF : Physical Functioning, ie the extent to which health limits physical activities such as self-care, walking, climbing stairs, bending, lifting and moderate and vigorous exercises.

RP : Role Functioning - Physical, ie the extent to which physical health interferes with work or other daily activities, including accomplishing less than wanted, limitations in the kind of activities or difficulty in performing activities.

BP : Bodily Pain, ie intensity of pain and effect of pain on normal work, both inside and outside the home.

GH : General Health, ie personal evaluation of health, including current health, health outlook and resistance to illness.

VT : Vitality, ie feeling energetic and full of pep versus feeling tired and worn out.

SF : Social Functioning, ie extent to which physical health or emotional problems interfere with normal social activities.

RE : Role Functioning - Emotional, ie extent to which emotional problems interfere with work or other daily activities, including decreased time spent on activities, accomplishing less and not working as carefully as usual.

MH : Mental Health, general mental health, including depression, anxiety, behavioural-emotional control, general positive effect.

HT : Reported Health Transition, evaluation of current health compared to 1 year ago.

The Dystonia Self-Perception Questionnaire : Questions 17 to 24.

These questions were designed by the researcher, as a direct result of the March 1993 Focus Group, to obtain the subject's own qualitative view on their dystonic spasms, its presentation and their relationship to other people. The research sub-committee of the Dystonia Society had a direct input, but the Focus Group, made up of people who had dystonia, was the guiding force behind this concept and each section contained open ended questions to allow all respondents complete freedom in their answers. It was also designed so that the results could be both qualitative and quantitative. It was also scored and coded for SPSS by Dr Jay Holland, Psychiatric Registrar at the RVI in Newcastle, based on a 7 point scale, which was designed to evaluate any positive or negative changes over time as a consequence of the intervention of Botulinum Toxin therapy.

1 = Extremely positive	2 = Positive	3 = Slightly positive
	4 = Neutral	
5 = Slightly negative	6 = Negative	7 = Extremely negative

PART THREE : METHODOLOGY

CHAPTER 10

The Epidemiology of Primary Dystonia

An Epidemiology of Primary Dystonia (EPD) has already been written and published by Dr Philip Duffey, the author, Mr Maurice Hawthorne and Professor Michael Barnes in Advances in Neurology, Volume 78, Dystonia 3, Chapter 13. However the enclosed, although partly duplicating this work, also describes the detailed methodology in obtaining the original information.

The main difference in obtaining this information, as opposed to all the other information obtained, was in determining who had a 'primary' dystonia, which was originally described as being different to other forms of dystonia only in its cause. This was not always clear and therefore where there was any doubt as to cause, then it was **not** described as primary. In other words, only those cases which were definitely primary were included in this written work.

The methodology by which all dystonias are identified has changed slightly since that time, due to the introduction of the "new" classification of dystonia, which was first introduced at the 3rd International Dystonia Symposium held in Miami, Florida, USA in October 1998. As previously described, prior to this conference, all dystonias were categorised in two basic ways, either primary (idiopathic, familial and sporadic) or secondary (symptomatic with a known or assumed cause). Since then the categorisation has been expanded into a) primary dystonia, b) dystonia-plus, c) secondary dystonia and d) heredodegenerative diseases. These have been previously described previously and therefore are not reproduced here.

The catchment area for this study comprised the counties of Northumberland, Tyne and Wear, Durham and Cleveland in the north-east of England. The region contained both rural and urban areas with major conurbations existing around the cities of Newcastle, Middlesbrough and Sunderland. The sample population was calculated to be 2,605,100 (United Kingdom Office for National Statistics, Estimated Residential Population Mid-1995).

The survey was divided into three phases that have run from 1993 onward. In the first phase the records of neurologists, and otolaryngeal and ophthalmic surgeons in the region were reviewed. The examination of case notes was not restricted to those coded for dystonia; it also included case notes coded for any unspecified disease. Added to this list of patients were those known to the Dystonia Society, a charitable organisation representing individuals with dystonia and campaigning for increased awareness of the condition.

In the second phase of the study measures were taken to heighten awareness of dystonia in the region; articles describing dystonia were placed in local newspapers, a series of radio discussions on the subject were given and on two occasions a short video sequence illustrating the more common forms of dystonia was screened on the regions' independent television channel. In addition, seminars for family physicians and other medical practitioners on the subjects of dystonia and the use of botulinum toxin were organised, in conjunction with the Dystonia Society in 1993, 1994, 1996 and 1998.

Finally, a postal survey was undertaken in a well-defined area within the region already exposed to phases one and two of the study. A brochure containing pictorial representations of the various focal dystonias and information regarding the concept of dystonia was delivered to 45,383 households containing 101,766 individuals. The targeted addresses were contained within three postal codes; most were within a single town boundary, its suburbs and neighbouring villages. The demographic characteristics of this community were considered to be representative of the region as a whole. The aim of the postal survey was to gauge, albeit crudely, whether a significant number of individuals with dystonia remained to be identified after the first two stages of the study.

Patients identified by these means in whom a diagnosis of dystonia was thought possible were interviewed and examined. Demographic and medical details were obtained and if appropriate the dystonia was classified according to anatomical distribution. Individuals fulfilling the diagnostic criteria for primary dystonia as described by the Ad Hoc Committee of the Dystonia Medical Research Council (Fahn, 1988) and known to be both exhibiting their condition and resident in the catchment area on the designated date of January 1st, 1996 were included in the study. A history of exposure to neuroleptic drugs or other agents known to be capable of producing dystonia were cause for exclusion.

This gives a good and clear description of the methodology adopted throughout the investigation of all the primary dystonias and has continued in much the same way since the publication of the above passages in the autumn of 1998.

PART THREE : METHODOLOGY

CHAPTER 11

Psycho-Social Research

A large number of different questionnaires were used during this section of the study.

Questionnaire No. 30 : Clinical Profile of Dystonia Questionnaire.

This instrument was administered once to every consenting participant and was designed, based on the work of Dr Marjan Jahanshahi in 1988, and amended to the ESD format by the author in 1993. It had already been used by over 300 subjects in a number of studies (Jahanshahi and Marsden, 1988a & b and Jahanshahi, 1991). A total of 346 subjects completed this questionnaire. It asks the patient a number of factual questions, based on the following three areas :-

a. *Contact with Medical Services*
b. *Clinical Features of the dystonia*
c. *Nature of your accommodation*

Questionnaire No. 40 : Torticollis Questionnaire. (attached to Questionnaire No. 30)

Again designed and used (as above) by Dr Marjan Jahanshahi, a total of 155 of these questionnaires were completed and related to specific questions on torticollis, laterocollis, retrocollis and antecollis, ie :-

d. *Type and direction of torticollis*
e. *Geste Antagoniste*

'Geste Antagoniste' is the term used to describe any sensory trick used to keep the head in the midline position. It has been known for some time (Patterson and Little, 1943) and can be defined as : "*The severity of the abnormal posture and of the involuntary movements of the head is affected by body position, often being worse when walking and relieved in the supine position. Patients use 'trick' movements to keep the head in the midline*" (Jahanshahi and Marsden, 1989b).

Questionnaire No. 50 : Living with Dystonia Questionnaire : A Psychological Profile.

This questionnaire was administered once during the ESD to every consenting participant. A total of 335 of these questionnaires were completed during the course of the ESD. It is a combination of four instruments; the first two of which have been used in other torticollis studies (Jahanshahi and Marsden, 1990a and b; Jahanshahi, 1991; Jahanshahi and Marsden, 1992) and had been completed by 294 patients prior to the start of this study.

The second two have been used in countless studies and have been validated many times over (see below).

a. *Functional Disability Questionnaire (FDQ)*

This 27-item scale was devised to assess the effects of torticollis on activities of daily living. It has been previously shown (Jahanshahi and Marsden, 1990a and b) to have good internal consistency, construct validity and test-retest reliability. Each item is scored based on a 0 (not at all affected) to a 4 (severely affected). The total scores range from between 0 to 108, with higher scores indicating higher disability. Based on previous reliability and validity analyses, 3 of the 27 items were sometimes omitted.

b. *Body Concept Scale (BCS)*

Patients rated the concept, 'my body', on 22 seven point semantic differential scales. The internal consistency, construct and concurrent validity and test-retest reliability of the scale have been shown to be acceptable (Jahanshahi and Marsden, 1990a and b). Scores range between 21 and 154, with higher scores indicating a more negative body concept. Based on previous reliability and validity analyses, some items can be omitted.

c. *Beck Depression Inventory (BDI)*

This 21 item scale measures depression. Each item consists of four statements representing increasing severity of depression, scored 0 to 3. Scores range from 0 and 63, with higher scores indicating higher levels of depression (Beck, 1961).

d. *Rosenberg's Self-Esteem Scale (SES)*

The 10-items of this scale assess an individual's feelings of self-worth and self-deprecation (Rosenberg, 1965). The response categories constitute a 5-point agree-disagree format. Evidence (Warr and Jackson, 1984) suggests that the positively and negatively worded items of this scale reflect two independent aspects of self-esteem. Also, lack of positive self-esteem is reportedly less predictive of depression than self-deprecation (Brown et al, 1986). Therefore, a positive self-esteem and a self-deprecation score were derived, each with a range of 5-20.

<u>Questionnaire No. 60 : Impact of Dystonia Questionnaire</u>.

This instrument was administered once at the end of the CUA or during the ESD to every participant and is an Acceptance of Illness (AOI) questionnaire. This 8-item scale consists of statements relating to the patient's ability to accept the reality of his (or her) illness and to adjust to it. Patients indicate their extent of agreement / disagreement with each item on a 5 point scale. Total scores are derived by summing across and dividing by the number of items. Scores range between 1 and 5, with higher scores indicating less acceptance of illness. The last question measures which of four stages the patient has currently reached. These stages are generally accepted as being Shock, Anger, Despair or Acceptance (Felton et al, 1984).

This particular questionnaire also has a final open ended question, which will enable the researcher to determine how successful the research organisation has been and if there are any lessons to be learnt for future research programmes, in methodology, presentation or personal attitudes. A total of 338 questionnaires were completed and returned.

Questionnaire No. 70 : Primary Carer's Questionnaire.

" *Too often in the past, socio-medical research has tended to concentrate on the person to whom the disease or disorder has presented itself and little or no measurement has taken place with the family or carers of that person . This has been considered to be one of the most important areas of research which has been almost totally ignored by the modern social epidemiologist.* " (Mechanic, 1988)

Because of the sentiment behind the above statement, this instrument was administered once during the ESD to every participant's primary carer in the form of a Carer's Strain Index (CSI). There is space at the beginning to indicate the carer's relationship to each subject in question.

This 13-item scale consists of statements relating to the primary carer's ability to cope with the subjects' illness and how or if they have had to adjust their own life to it. Patients indicate their extent of agreement / disagreement with each item on a 4 point scale. Total scores are derived by adding up the individual scores. Scores range between 13 and 54, with higher scores indicating less strain on the carer (derived from the Caregiver Strain Index : Robinson, 1983).

There was an open-ended question added at the end to enable the carer to describe how dystonia has changed their life, if at all.

A total of 225 of these questionnaires were completed.

PART THREE : METHODOLOGY

CHAPTER 12

Socio-Economic Research

It is quite difficult to describe exactly which questions were asked in the field, apart from getting a general feel of the demographic data collected during questionnaire No 20, which was entitled the 'demographic and dystonic profile' questionnaire.

The important questions asked gave an insight specifically to anything which related to the patient's current and previous Social Income Group (SIG), their current and previous incomes and questions relating specifically to their current job status, their previous job status and their current methods and levels of income. The specific demographic data asked related specifically to social and economic information, such as :-

Geographical location - Post Code for determining county, district and postal sector.

Age - date of birth, confirming that the Clinical Information Form (CIF) was correct.

Gender - male or female. There were no specific questions regarding ethnicity.

Marital status - noted single, married, divorced, separated, cohabiting or widowed.

Number and gender of children, including eldest's and youngest's year of birth.

Social Income Group - researcher's placement based on occupation and status.

Current Income - patient only (family income was noted where no income existed).

Number, range and amount of any benefits, allowances, pensions, etc.

All of the above information was collected and placed on the SPSS file in order to maintain its confidentiality. The rest of the questions really related to getting a feel for the person's socio-economic status before the onset of the dystonia and any changes since that time. The whole point being able to discern whether the socio-economic status of that person had changed from the time of the onset of their dystonia up to the point of interview.

This was generally quite easy as the vast majority of patients were very open and responded very well to the reasons why these questions were being asked and their answers reflected the truth of the situation, in which they now found themselves. This was much appreciated by the researcher and often reflected the need for further discussions outside the current research data. The use of the Dystonia Counsellor was greatly appreciated here and several clients were referred across to him as they became formally identified.

PART THREE : METHODOLOGY

CHAPTER 13

The Dystonia Nurse Practitioner Project

The post of Dystonia Nurse Practitioner was first created in mid-1997 by Professor Mike Barnes for Nurse John Whitaker, who had been working within the dystonia clinic in Newcastle as the clinic nurse since it had first started in the mid 1980's.

The idea of the project was to see if " *a properly trained outreach nurse practitioner could provide a service that is as good as, or better than, that provided at a hospital outpatient clinic for people with dystonia requiring treatment with botulinum toxin.* "

In doing so, it was to see if an outreach nurse could administer the drug effectively to as good a standard of efficiency as the doctors were already doing in the clinic. The main motive behind the idea was that the doctors could see that once the patient was on a standard mode of injections there was little to be gained and much that could be achieved by having the injections administered by a nurse rather than a doctor, particularly if the injections were given in the patients' own home. Remember that, by 1997, the numbers of patients had increased considerably and whereas in the 'old days' the clinic was administered by a single doctor and a nurse being run once a month, by now the clinic was run weekly using three doctors and two nurses and was (to put it mildly) extremely busy !

The first part of this two year research post was training of the nurse in order to administer the Botulinum Toxin therapy without supervision, followed by a proposed 18 month administration of the drug within an arbitrarily selected group of half the clinic's patients, the other half of the patients being administered by the doctors at the clinic in the normal way.

Before the process could start, a very carefully selected criteria had to be adopted to ensure complete fairness and independence regarding the patient selection. The following was adopted by the research team of Professor Barnes, John Whitaker and the author and was used in the process to effectively monitor independently the work of the nurse practitioner.

The Inclusion Criteria were :-

- People attending the movement disorder clinic at Hunters Moor Regional Neuro-Rehabilitation Centre with a definite clinical diagnosis of Spasmodic Torticollis, Blepharospasm, Hemi-Facial Spasm or other Segmental Dystonia, Hemi-Dystonia or Generalised Dystonia.
- Treatment of dystonia with botulinum toxin injections on at least two preceding occasions, with a clinical need for such injections to continue.
- Willingness to give fully informed consent to participate in the study by the individuals concerned.

The Exclusion Criteria were :-

- Inability to travel on a regular basis to the outpatient clinic (as would be required as a commitment over the study period).
- Pregnancy or child bearing potential.
- Psychiatric or other psychological problems that made, in the opinion of the investigators, compliance with the study protocol unlikely.
- Previous known allergic susceptibility to botulinum toxin.
- Previous serious side effects or other reaction to botulinum toxin.
- Complex or variable dystonic movement disorder that required variation in muscles injected with botulinum or significant variations in other treatments on a clinic by clinic basis.

Having found out which patients were available, it was then necessary to decide which patients were to be in which group. This was done by using the patient's individual Identification Number as a three digit number being selected using random selection procedures. The patient's Clinical Reference Number could have been used but as it was 4 digits, it was agreed that their ESD reference number being 3 digits was much more efficient. Having selected which patients were to be in which group, a letter was then sent out to each patient inviting them to take part in the process. They were then individually contacted and the process explained to each one very carefully.

A total of 126 people met the initial criteria and were invited to take part in the process. Because there was a need to determine individual treatments for those with Spasmodic Torticollis, Blepharospasm, Writers' Cramp and Generalised Dystonia, a total of 64 people were invited to take part in the Home Group, whereas there were only 62 in the Clinic Group.

PART THREE : METHODOLOGY

CHAPTER 14

Data Input and Coding Frame

The data is input onto a computer in an SPSS (statistical package for social science) file entitled 'phddata.sav'. This file contains all the six years data from 6th May 1993 to 5th May 1999, but it has also been used on other associated research projects :-

ESD = Epidemiological Survey of Dystonia

This contains all the data available at the end of the input and will be the main file used for this thesis.

CUA = Cost Utility Analysis

This file represents all the data collected on subject's No 001 to 231 inclusive. However it is important to note that No's 148, 152, 159, 170, 175 and 210 were all non-dystonic CUA controls and have been since removed from the file after the completion of the Cost-Utility Analysis was completed and published (see publications)

EPD = Epidemiology of Primary Dystonia

This was used specifically in the work since published by Dr Philip Duffey and the author and it is vitally important to understand that only the data which confirmed that if the subject had a primary dystonia was used. All other data was not used at that time

DNP = Dystonia Nurse Practitioner

This data related only to those 126 subjects who were selected as part of the Dystonia Nurse Practitioner research project. They are identified by a specific 'C' for Clinic or 'H' for Home in that particular section of the programme. Everyone else has a 'N' for 'not in the dnp study' against their file reference in the column marked 'dnp location'.

PSR = Psycho-Social Research

This specifically relates only to all the subjects who have a number from 001 to 550 (whether they agreed to participate or not) plus three other subjects, ie No's 564, 594 and 677 - who have since completed all the necessary forms under this research project. This particular part of the research is strictly in collaboration with Dr Marjan Jahanshahi who, it is hoped, will be publishing further results later.

IFD = The Impact of Focal Dystonia on the Working Life of Musicians

This research project only related to three selected cases, ie No's 005, 542 and 594, who had been, or were musicians, and had a focal dystonia.

SER = Socio-Economic Research

This particular research project is discussed in detail in this thesis and relates specifically to all data available at the time.

Patient Numbering and Status.

The patient's identification (ID) number is automatically determined by the row number and the column shown as ESD is used for ranking purposes only.

NB : Six CUA patients have been removed and replaced by ESD patients between 04.12.95 and 04.03.96 - these were No's 148, 152, 159, 170, 175 and 210.

The rest of the numbering system is made up as follows :-

001 to 199 inclusive : were active CUA patients coded A, B or D
200 : is a new ESD patient coded M
201 to 231 inclusive : were external control CUA patients, re-coded to ESD
232 to 300 inclusive : are new ESD patients coded A, B or M
301 to 347 inclusive : were (1994) 500 Series patients re-numbered to 300
348 to 937 inclusive : are new ESD patients numbered and coded accordingly

Syntax Files

Syntax file copies have been enclosed separately in the appendices and no formulae have been included in this file. The 7 syntax files were run in the following order :-

phd-geog.sps : Recoding 'post codes' into 'county' and 'district'.
phd-all.sps : 21 computations re Age, DoB, Dates, Differences, etc
phd-dec.sps : 12 computations re defining decimal places in the above.
phd-sf36.sps : 29 computations re EuroQol and SF36 as per handbook.
phd-qual.sps : 3 computations re qualitative open-questions.
phd-psr.sps : 17 computations re PSR, as per Dr Jahanshahi's instructions.
phd-env.sps : 93 comps 'if env = 1, envaa = 0,' etc., & 'if alg = 2, alga = 2,' etc.

SPSS Coding Frame

The SPSS codes (up to No 275) were used in the Cost Utility Analysis (CUA) but have since been amended for the ESD, EPD, SER, PSR and DNP, as required.

All variables are entered as Numeric 4.0; missing values None; alignment Right and column width 4, unless otherwise shown.

The value label positions, ie their number, are numbered as per the list of variables on the working file.

The file contains 937 rows (subjects) x 842 columns (variables) and therefore has a total of 788,954 potential data sets.

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 15

Epidemiology, Prevalence and Incidence

There were a total of 937 subjects registered in the ESD with 937 separate variables encoded on the SPSS file, thus producing 788,954 data sets, at the time of the analysis after 6th May 1999.

The Epidemiology of Primary Dystonia (Duffey et al, 1998) has shown that the crude point prevalence ratios of all the primary dystonias, within the population of the 4 counties of North East England, is 14.28 per 100,000 people, with 95% confidence intervals from 12.8 to 15.75. This breaks down into 12.86 per 100,000, with 95% confidence intervals from 11.45 to 14.25 for Focal Dystonias and 1.42 per 100,000 (95% c.i. 0.95-1.89) for Generalised.

This prevalence shows that 1 in 7,000 people have Primary Dystonia in the region, but when adding the known and estimated figures for secondary dystonia (Marsden and Quinn, 1990) it goes to prove that dystonia is second only to Parkinson's Disease as the most prevalent movement disorder, more prevalent than other better known diseases such as MS and MND (Marsden and Fahn, 1998).

Whilst the Epidemiology of Primary Dystonias (EPD) restricted itself to the 2.5 million population of the four North Eastern Counties of England, this research had no such boundaries. Socio-Economic research makes no distinction between primary and secondary dystonias, because an abnormal posture or involuntary muscle spasm presents in exactly the same way in both cases. If the subject is unemployed due to their dystonia, it does not matter if it was induced, genetically inherited or idiopathic - they are still economically vulnerable.

In more general terms, the concentration on Darlington in 1996 has shown an even more dramatic increase in diagnosis and therefore the known prevalence of all forms of dystonia. Currently a total of 42 people display some form of dystonic spasm within the population of 101,766 in the Post Codes DL1, DL2 and DL3 - this gives a crude prevalence figure of 41.27 per 100,000 population (or 1 in 2,400) for this town alone.

Although prevalence in Durham, Tyne & Wear and Cleveland have approximately similar results, there is a distinct reduction in Northumberland, Cumbria and North Yorkshire. The author believes that the main explanation for this is due to the fact that awareness raising in these specific rural areas was carried out to a lesser degree of intensity than in the more urban areas.

During the research on the three musicians with dystonia (Butler and Duffey, 1997), it was established that the prevalence of dystonia within the ranks of professional musicians in Germany was reported as high as 1 in 500 (Altenmuller, 1997). The reason for this may be explained in that professional musicians, almost more than any other occupation, make very fine repetitive movements of the fingers and hands and that very small dystonic tremors or postures, that are not noticeable nor cause a problem in others, can be devastating for them.

According to Altenmuller : " *Focal limb dystonia of the upper extremities occurs more frequently in musicians (prevalence about 1:500) than in other professions requiring highly skilled movements of the hand (prevalence about 1:3400). The increased incidence of dystonia in musicians may be related to specific qualities of their sensory motor skills; 1) routines for stereotyped movements are rehearsed for extended periods of time with gradually increasing degrees of complexity, 2) via auditory feedback, the motor performance is extremely controllable by both performer and audience, 3) rehearsing and performance is closely linked to emotion.* "

This last point is uncomfortably close to suggesting that dystonia is still linked to a psychogenic disorder, nevertheless the significance of Altenmuller's research is well worth noting. However it is interesting to note :-

1. The EPD (Duffey et al, 1998) has a proven prevalence of 1 in 7,000 (for primary dystonias only)
2. Nutt (Nutt et al, 1988) talks of a prevalence of 1 in 3,000 in the USA (but based on just 34 people with dystonia)
3. The Darlington study (Butler and Duffey, 1996b) shows that 1 in 2,400 of the local population have some form of dystonic spasm.

What has been shown by all of the above is that the more research is undertaken the more the prevalence figure increases !

The point being that if dystonic musicians are truly shown to have a prevalence of 1 in 500, how much more work is required within the ESD to arrive at an absolutely valid figure ? There is no reason to suppose that musicians are more prone to dystonia than any other group of people, merely that the very nature of their occupation brings out the dystonia to a greater degree by the very nature of the fine repetitive movements required.

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 16

Diagnoses, previous and current

Diagnosis

Because diagnosis plays a major factor in getting sufficient numbers of people with dystonia available for research purposes, it was decided in 1992 that an awareness raising campaign would be run in conjunction with the local TDS SHG's. This has manifest itself in a number of seminars for medical professionals and a series of seven training videos made at Hunters Moor in Newcastle, as well as various radio and television broadcasts and newspaper articles being written over the 5 year period. All of this has helped to raise awareness of dystonia amongst the medical professionals and the general public which has dramatically increased the number of diagnoses made.

The total number of subjects diagnosed each year to date has seen a steady increase, but there were only 33 subjects diagnosed from 1937 to 1977, averaging just 0.83 per year for those 40 years. The average over the past 10 years has been 70.5 per year and the average over the last 5 years has been 95.0 per year or 1.83 per week. However this is only relevant in so far as it shows that the awareness raising campaigns have been working.

What is more significant is the number of onsets each year. Reference to the table below shows that the years 1997 and 1996 have less number of onsets than the 12 years previously to 1985. This can be explained in that dystonia can often take several years to develop from the initial onset and many people do not seek medical assistance until the problem is at least a couple of years old. Ignoring the data for the last 2 years, we see the average number of onsets for the 5 years from 1991 to 1995 as being 53.2 per year. The average is 44.2 per year for the period from 1986 to 1990 and 19.2 per year for the period from 1981 to 1985.

There is no neurological reason to suppose that the incidence will vary by this wide margin over three consecutive 5 year periods, therefore it could be argued that the reason why we see a gradual increase in onset data in this time frame is a direct result of the increased awareness in newly diagnosed cases. Therefore the epidemiology has not yet reached all those people with an onset date over 10 years ago, who either have yet to be diagnosed or who have been diagnosed and yet remain unknown to the epidemiological register.

The introduction of Botulinum Toxin (BT) therapy has had a tremendous impact on the rate of diagnosis and has helped correct some of the errors in diagnosis of the past. However regardless of the reasons, it is entirely feasible to assume that a large number of people with dystonia in the region still remain undetected by the ESD. As to how many, this depends on which 5 year incidence ratio one uses.

TABLE 14. Onset vs Diagnosis in each year.

Year	Onset in the year	Diagnosis in the year	Notes
1999	1	21	Project ended 5th May
1998	15	71	End 5th project year
1997	26	117	End 4th project year
1996	39	87	End 3rd project year
1995	47	103	End 2nd project year
1994	81	90	End 1st project year
1993	72	96	Project started 6th May
1992	53	69	
1991	54	60	
1990	69	49	TDS (NE) formed
1989	36	37	
1988	48	19	BT available in the NE
1987	39	12	
1986	42	11	
1985	25	10	
1984	24	5	
1983	15	10	TDS formed
1982	18	8	
1981	21	9	
1980	22	13	
1979	10	2	
1978	25	5	
1977	10	4	
1976	14	4	
1975	18	2	
1974	9	1	
1973	7	2	DMRF founded
1972	6	4	
1971	6	0	
1970	3	1	
1969	2	2	
1968	8	3	
1967	6	0	
1966	4	0	
1965	6	0	
1664	9	2	
1963	6	1	
1962	0	1	
1952 - 1961	20	4	
1942 - 1951	12	1	
1932 - 1941	6	1	
1921 - 1931	3	0	
Totals =	937	937	

The undetected population could be between 720 and 1810 people, assuming an average 7.6 years between onset and diagnosis and an average age of 39.4 years at onset (both statistics taken from the results of the ESD) and using the Life Tables (CSO, 1993) which show male life expectancy at birth to be 72.7 years (78.3 years for females) and at 40 years (the closest to 39.4) male life expectancy is 74.8 years (79.6 years for females). This means between 46.5% and 68.6% of the dystonic population remains undetected.

An alternative method of calculating the undetected population would be to extrapolate the various study figures for the entire region. The population of the four northern counties was 2,605,100 (U.K. Office for National Statistics, Estimated Residential Population) in mid-1995. The best set of prevalence data yet known in the region are the Darlington study figures deduced at 1 in 2,400, which gives a dystonic population for the four northern counties of the North East of 1,085. A total of 779 primary and secondary dystonias are already known in that geographical area (as of 05.05.99), thus the percentage remaining undetected is 28.2% or 306 people.

The lowest undetected percentage, based on the onset data, is 46.5% and the estimated undetected percentage, based on the epidemiological data, is 50.3%. Although both these sets of figures are relatively close, they indicate the tremendous amount of work still required to do.

The referrals to the Bot. Tox. clinics (including previously diagnosed as well as newly diagnosed) has remained fairly constant since the start of the study at 2.6 per week. This has seen the frequency of the clinics increase from once a month, to twice a month, to once a week during this period. The cost of supplying the Botulinum Toxin has also dramatically increased during the same period. The Hunters Moor clinic which spent less than £50,000 per year on Bot. Tox. at the start now has an annual bill of £510,000 for toxin alone (data from fiscal year 1997/98).

The diagnosis of individual cases of dystonia has been the single most difficult piece of research to undertake. The previous diagnoses have often been rough and ready, to say the least. The number of people who have previously been misdiagnosed is very high. The Dystonia Society in its 1993 survey of 705 people indicated that 463 (65.7%) were misdiagnosed at some stage. Our research indicates an even higher figure and with some profound results. In order to determine the exact nature of this problem, it is necessary to go back into time and find out exactly what the previous diagnoses were and see how this compares with the present day diagnoses.

In order to do this, we need first to establish what exactly were the previous diagnoses and how long it took to get a correct diagnosis. 338 people answered the question on the reasons for any delay in diagnosis. This was principally (according to the opinion of the patients) that their Doctor was unaware of the condition (37.0%). However the second most common cause was misdiagnosis (26.4%) and in this case 52 people were treated for this misdiagnosed condition together with a further 14 people who were misdiagnosed and treated for having PD, MS or CP. There was no delay in 13.9% of cases, 10.7% did not seek advice and a further 10.7% had an unknown cause or reason. Finally there were 4 people who were still waiting for an appointment at the time of the research and another person who had just gone into remission.

The actual diagnosis was carried out in the vast majority of cases by a hospital or privately consulted Dr or Consultant (78.2%). However, of the remainder, a few (6.7%) were diagnosed by their own G.P., mainly as a result of the campaign of awareness raising conducted during the course of the research in the region, with Ophthalmologists coming next with 4.4%. Self-diagnosis was the next highest at 3.8%, with Speech Therapists coming next for Spasmodic Dysphonia (1.5%). The balance were made up of 8 different medical professions diagnosing the dystonia, with 2 people being diagnosed through one of their own children having the disease.

Another area of concern was the length of time it has taken not only to obtain a correct diagnosis, but also is a few cases (35 people) to be actually told that they have dystonia. The majority are not specifically children, where one can understand (particularly many years ago) the medical profession not telling a child what they had. Nevertheless, this seems to have applied even to the present day.

There were 6 people who had never been told what they actually had, 2 of whom had been diagnosed in 1996, 3 in 1994 and one had originally been diagnosed in 1992 but had still not been told what she had. Of the remainder, the greatest difference between an actual diagnosis and being officially told what the disease was called was officially recognised as 18 years. These numbers are small, ie a total of less than 4%, nevertheless it does just show the extent of the prestige with which the medical profession is still held by the general public.

The extent of the present diagnosis is quite staggering when one considers that at the time of the commencement of the research programme there were less than 150 people known to have dystonia within the region. 'Known' meaning within the knowledge of the researchers, The Dystonia Society or anyone outside of the immediate family or medical professional attending the particular patient. As can be seen from the years of diagnosis above, one can easily see the growth and extent of this phenomenon.

A total of 585 people (62.4% of the final total of 937) were diagnosed within the 6 years that the research project has been running, although the percentage of onset during this period was only 30.0%. This shows the true extent of the research programme's value.

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 17

Treatments, previous and current

Table 15. Different treatments used and various effects recorded.

Treatments	Previous No	Current No	Effect (%) Better	Effect (%) Unchanged	Effect (%) Worse
Bot. Tox. Therapy	149	493*	471 (95.5)	20 (4.1)	2 (0.4)
Drugs / Medication	287	232	74 (31.9)	111 (47.8)	47 (20.3)
Physiotherapy	125	191	13 (6.8)	164 (85.9)	14 (7.3)
Acupuncture	43	101	11 (10.9)	90 (89.1)	-
Osteopathy	34	98	5 (5.1)	91 (92.9)	2 (2.0)
Hypnotherapy	24	73	6 (8.2)	66 (90.4)	1 (1.4)
Surgery	40	53	18 (34.0)	31 (58.5)	4 (7.5)
Psychologist	33	49	3 (6.1)	41 (83.7)	5 (10.2)
Counselling	18	46	46 (100)	-	-
Chiropractic	12	25	3 (12.0)	21 (84.0)	1 (4.0)
Yoga	3	20	6 (30.0)	11 (55.0)	3 (15.0)
Meditation	1	15	6 (40.0)	9 (60.0)	-
Alexander Technique	2	8	4 (50.0)	4 (50.0)	-
Aromatherapy	9	5	3 (60.0)	2 (40.0)	-
Speech Therapy	30	5	-	-	-
Healing Hands	8	5	-	-	-
Wearing a collar	37	4	-	-	-
Homeotherapy	7	4	-	-	-
Biofeedback	2	4	-	4 (100)	-
Relaxation Technique	8	3	-	-	-
Reflexology	6	3	-	-	-
Heat Treatment	35	2	-	-	-
Manipulation	23	1	-	-	-
Psychiatry	43	-	-	-	-
Rheumatology	12	-	-	-	-
Orthopaedic Surgery	7	-	-	-	-
Cortisone Injections	4	-	-	-	-
Ophthalmology	2	-	-	-	-
Phenol Injections	1	-	-	-	-
Immunology	1	-	-	-	-
Hydrotherapy	1		-	-	-
Totals =	1007	1440	-	-	-

NB : * There is a section in this category entitled "Too early to define" - there were a total of 80 patients still under this category at the time of closing the file and this number has not been included above nor in the % calculations.

Although this research programme has been built up mainly through the current treatment for dystonia, particularly Botulinum Toxin Therapy, nevertheless the previous treatments should be noted and some of the lessons explored. The above chart lists the treatments previously used as described during the interview, together with those treatments used since, together with their effect. Only 373 people detailed their 1007 previous treatments, averaging 2.7 each, however it does show the extent to which patients have tried to obtain some relief from their constant muscle spasms. As can be seen, the far most effective treatment is definitely the Botulinum Toxin therapy. Not only does it score by far the highest, but also its non-effect category is relatively small and the bad-effect category is very small. By far the most effective type of treatment is Counselling with 100% effectiveness - more of this later.

Botulinum Toxin Therapy and Side Effects

What can also essentially be seen is the reduction in other forms of therapy compared to the tremendous increase in Botulinum Toxin therapy. This is a measurable increase and this therapy is now currently the first line treatment for dystonia, although it must be remembered it does not 'cure' the disease, merely makes it more acceptable or comfortable to live with. As can be seen, not everyone with dystonia currently uses this toxin, for a number of different reasons. The total figures currently for its use within our own dystonic population is as shown below.

Of the current survey population of 937, exactly 22.9% (215 people) have never been injected. A further 10.2% (96) have only been injected once with a further 5.2% (49) people having had a 2nd injection as part of their current treatment phase. This means that a further 145 people are currently still in the interim treatment stage. Of the remaining 577 (61.7%) people, they currently belong to the largest regional group of dystonia patients and two of the three hospitals currently injecting the toxin within the region are currently first and third in supplying toxin in the UK. The way the toxin has been used during the past 6 years has also been monitored and checked. The following chart is not intended to be a measurement of effectiveness, merely of usage.

TABLE 16. Measurement of Botulinum Toxin Usage.

Bot. Tox.	Before start	6.5.93 - 31.3.94	1.4.94 - 31.3.95	1.4.95 - 31.3.96	1.4.96 - 31.3.97	1.4.97 - 31.3.98	1.4.98 - 31.12.98
0 inj	529	751	770	646	537	546	491
1 inj	87	34	9	53	56	46	104
2 inj	69	36	37	48	70	48	87
3 inj	43	41	38	65	113	89	95
4 inj	42	49	54	67	103	114	107
5 inj	33	20	23	39	37	59	42
6 inj	40	6	5	16	13	30	10
7 inj	20	-	1	2	6	5	1
8 inj	17	-	-	1	1	-	-
9+ inj	57	-	-	-	1	-	-
Ann =	-	186	167	291	400	391	446
Totals	937	937	937	937	937	937	937

The majority of people had their injections according to where in their body the spasms actually occurred. Occasionally the injection was not in the particular point of spasm but from where the greatest effect would be received. For example, there was one patient who had a few injections in her stomach musculature to try and reduce the pain occurring in her abdomen. Of the specific injection sites, 40.0% of injections were in the neck / shoulder position, 26.1% around the eye muscles, 13.0% specifically in the face musculature, 8.2% in the throat, 6.5% in the hand or arm, 4.3% in the leg or foot and 1.7% in the back or abdomen.

TABLE 17. Different side effects

Side Effect	**Previous** type of side effect	**Current** type of side effect
Drowsiness	35	0
Hallucinations	35	0
Nausea	33	8
Increased spasms	26	11
Felt ill / more pain	25	15
Tiredness / Lethargy	21	3
Depression	12	0
Double Vision	9	27
Dry Mouth / Throat	9	4
Lack muscle control	8	55
Loss of Weight	7	0
Dizziness	6	1
Allergic Reaction	6	1
Headaches	5	7
Difficulty Walking	5	3
Addiction	4	0
Dry Eyes	3	4
Constipation	3	0
Hospitalised	2	4
Skin Rash	2	2
Elation	2	0
Increased Weight	2	0
Memory Loss	2	1
Dysphagia	1	73
Ptoris	1	25
Insomnia	1	0
Bruising / Bleeding	0	23
Flu-like Symptoms	0	16
Coughing	0	7
Tickling Throat	0	4
Watery Eyes	0	3
Feeding by Tube	0	2
Sneezing	0	1
Phlegm	0	1
Totals =	265	301

These injections were only noted in full detail up until 1997, after which time the numbers and frequencies of the clinics got too busy in order to strictly document every patient. However they do give a strong indication of the spread of the injections and furthermore a strict record was maintained on any particular side-effects. It was noted that these gradually decreased as the injection teams became more practised at giving the injections, nevertheless the type and area of side effects are interesting, particularly when compared to those side effects experienced by the same patients previously during their various drug regimes. Some of the side-effects do not relate specifically to Bot. Tox. Injections, but they do give an overall impression of the type of side effects experienced.

The length of time that these various side-effects was also noted - in particular the difference in time between the two phases.

TABLE 18. Length of time affected by all treatments

Length of time affected :	Previous side effects	Current side effects
One day	5	9
A couple of days	6	44
About a week	5	35
Over a week	2	39
A month or over	3	40
Until treatment stopped	108	1
Undefined period	40	11
2 - 3 weeks	2	23
Up to 2 months	1	12
Totals =	172	214

The difference is obvious by comparing the two sets of figures. In the first set, the highest proportion by far (62.8%) is "until the treatment stopped" with the second highest figure being (23.3%) being for "an undefined period". This represents a positive resentment that the patient had to go through the sheer hell, at times, of trying to find a treatment that suited them. Although there is a tremendous spread in the current set of figures, one can see straight away the difference. The highest figure is for the period of "a couple of days" and that is only 44 people or 20.6%. The difference that this treatment has had on people with dystonia, in general, has been nothing short of miraculous. It has had a tremendous effect on their Quality of Life as well as improving the overall effect of medical interventions, as can be seen above.

<u>Previous treatments</u>

Another way of looking at previous treatments was to ask each patient to tell us which specialist he or she had actually consulted **before** they were actually diagnosed as having dystonia. This was carried out towards the end of the research and a total of 362 participated in this part of the research. Two main questions were asked; firstly who did you consult before you were finally diagnosed with dystonia and what alternative therapies had you undertaken before you were officially diagnosed ? The answers were quite interesting and are shown below.

TABLE 19. Different specialists consulted prior to diagnosis and alternative treatments given.

Specialists consulted prior to the diagnosis	No	%	Alternative Treatments	No	%
Neurologist	140	16.3	Physiotherapy	179	23.6
Physiotherapist	114	13.2	Acupuncture	100	13.2
Orthopaedic Surgeon	94	10.9	Osteopathy	95	12.5
Osteopath	92	10.7	Hypnosis	72	9.4
Psychiatrist	88	10.2	Counselling	52	6.9
Psychologist	59	6.9	Surgery	47	6.2
Rheumatologist	59	6.9	Psychotherapy	36	4.7
Unknown Consultant	48	5.6	Speech Therapist	34	4.5
Ophthalmologist	37	4.3	Heat/Shock Treat.	24	3.2
Speech Therapist	31	3.6	Chiropractic	19	2.5
Neuro-surgeon	28	3.3	Reflexology	12	1.6
ENT Surgeon	26	7.0	Traction	11	1.4
Chiropractor	20	2.3	Yoga	11	1.4
Orthodontic Surgeon	5	0.6	Relaxation Tech.	10	1.3
Paediatrician	4	0.5	Manipulation	10	1.3
Paediatric Neurologist	4	0.5	Faith Healing	9	1.2
Pain Specialist	3	0.3	Experimental drug	8	1.1
Haematologist	2	0.2	Homeopathy	8	1.1
Chiropodist	2	0.2	Hydrotherapy	8	1.1
Gynaecologist	1	0.1	Aromatherapy	7	0.9
Aromatherapist	1	0.1	Behave. Therapy	7	0.9
Plastic Surgeon	1	0.1	-	-	-
Geriatrician	1	0.1	-	-	-
Immunologist	1	0.1	-	-	-
Totals =	861	100%	Totals =	759	100%

This means that 429 people consulted an average of 2.0 specialists plus another 1.8 alternative therapies before getting a correct diagnosis.

<u>Medications</u>

A comprehensive list of all medications taken by the patients was logged at the time of interview. These were then placed into various categories, as best as possible in order to determine if the medication was given for dystonia primarily, to alleviate the conditions of dystonia secondarily or not for dystonia at all. Of course a number of medications could be placed in a number of different slots but this was determined in conjunction and with the advice of the medical people on the team. The number of medications given were as shown on the following page.

TABLE 20. Different Primary Medications given

Name of medication	Primary medication (n)
Diazepam	53
Benzhexol	45
Baclofen	24
Co-careldopa	22
Clonazepan	21
Tetrabenazine	11
Cannabis Resin	10 - note an illegal drug
Co-beneldopa	10
Orphenedrine	9
Dantrolene	5
Primidone	3
Selegiline	3
Benztropine	2
Pimozide	2
Thioridazine	2
Trifluperazine	2
Haloperidol	1
Chlorpromazine	1
Levodopa	1
Methixene	1
Lysuride	1
Totals =	229

Sub-Totals

Benzodiazepines	74	=	32.3%
Antimuscarinics	57	=	24.9%
Others	47	=	20.5%
Spasmolytics	29	=	12.7%
Neuroleptics	22	=	9.6%
	229	=	100%

The above drugs are considered the primary ones given to people with dystonia. There are also a number of other medications which may be prescribed which are considered by some as efficacious as these.

Of the 229 people noted as taking at least one of the above drugs, the vast majority (72.1%) can be seen to be taking just the first 5 on the list. The main drug taken is Diazepam closely followed by Benzhexol. Although a lot has been talked about the over-prescription of Diazepam, it is one of the most effect of the medications given for the various forms of dystonia.

The use of Baclofen has been quite controversial, especially with the introduction in the USA in particular, of Intrathecal Baclofen usage. This is where an internal pump is placed, usually to one side of the frontal stomach area, which feeds Baclofen directly into the spinal cord and it has proved useful, although the trials are still progressing, in limiting severe muscle spasms to do with the trunk itself.

The following drugs are considered more of a secondary nature or, in other words, they are not treating the symptoms but the effects of the disorder.

TABLE 21. Different Secondary Medications given.

Name of medication	Given to no. of people as secondary medication	
Co-proximal	35	
Amitripyline	32	
Co-codamol	24	
Carbamazepine	23	
Dothiepin	16	
Paracetemol	14	
Ibuprofen	13	
Temezepam	12	
Co-dydramol	12	
Diclofenac	11	
Dihydrocodeine	10	
Nitrazepam	9	
Fluoxetine Hydrochloride	8	
Propranolol	8	
Sodium Valproate	8	
Hypromellose	7	
Clomipramine	5	
Phenytoin	5	
Lorazepam	5	
Chlordiazepoxide	4	
Procyclidine	4	
Solpadol	4	
Lofepramine	3	
Lormetazepam	3	
Primidone	3	
Chlormethiazole & Clobazam	4	(2 each)
Diflunisal & Dipipanone	4	(2 each)
Fluvoxamine & Imipramine	4	(2 each)
Indomethacin & Naproxen	4	(2 each)
Pergolide & Piroxicam	4	(2 each)
Remoxipride & Solpadeine	4	(2 each)
Sulpriride & Zopiclone	4	(2 each)
Benorylate & Buprenorphine	2	(1 each)
Dipyridamole & Doxepin	2	(1 each)
Feldene Gel & Flurbiprofen	2	(1 each)
Methadone & Naftidrofuryl	2	(1 each)
Parafon & Paroxetine	2	(1 each)
Risperidone & Tiaprofenic Acid	2	(1 each)
Trimipramine	1	

Totals =	319 given to 145 patients - averaging 2.2 each	

There were then a further total of 115 different other drugs given to the patients for other problems not associated with their dystonia nor any of the associated problems related to it. These are available if required as are the number of dosages noted.

Prescriptions

The frequency of prescription were noted.

TABLE 22. Frequency of Prescription.

When taken	No. taken
As required (prm)	97
Once a day (od)	73
Twice a day (bd)	51
3 x a day (tds)	43
At night (nocte)	36
4 x a day (qds)	30
Injections (prm)	4
Taken weekly	3
Totals =	337

As were the number of methods of prescription noted.

TABLE 23. Different methods of Prescription

Prescribed Free of Charge	236
Patient paid NHS rates	174
Patient paid privately	9
Totals =	419

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 18

Comorbidity and The Quality of Life

Quality of Life was measured using the EuroQol and SF36 measurements. The results were analysed using SPSS techniques and are shown below, as are the various Comorbidity measures adopted. Each will be detailed separately.

Comorbidity

Comorbidity was measured using the standardised questions and can be stated as a definitive measure in the 442 people (47.2%) who responded.

The question asked was " What other conditions currently affect you " and the answers were divided into :

1. Those directly related to dystonia
2. Those indirectly related to dystonia
3. Those not related to dystonia.

45 people (4.8% of the entire total) died either before (11), during the study (17) or since completion (17) - this was noted in this section but only 2 peoples' deaths were in any way related (see later for further information).

A number of specific diseases were classed as being related to dystonia, or of primary consideration as a specific cause of their dystonia, in a total of 94 (10.0%) people.

TABLE 24. Comorbidity of patients - directly related to dystonia.

19 people had had a **Stroke** (related to dystonia or not)
13 have **Parkinson's' Disease** (related to dystonia or not)
12 have **Cerebral Palsy** (related to dystonia or not)
11 have **Multiple Sclerosis** (related to dystonia or not)
9 have been defined as having **Tardive Dyskinesia**
8 have **Benign Essential Tremor**
8 have **Spasticity**.
3 with an **Head Injury**
2 with **Multiple System Atrophy**
2 with **Anoxic Brain Damage**
6 with each having **Chiari Malformation, Gilles de Tourettes Syndrome, Leigh's Disease, Myoclonus, Shy Drager's Syndrome** or **Pneumococcal Meningitis**.

A further 16 diseases were defined as indirectly related to dystonia. These 105 people, ie 11.2% of the total, are listed below.

TABLE 25. Comorbidity of patients - indirectly related to dystonia.

21 people were officially diagnosed as having **Depression**
19 were diagnosed as having **Cervical Spondylosis**
14 had an **Anxiety Neurosis**
9 had **Epilepsy**
8 suffered from **Insomnia**
6 were officially recorded as **Drug Addicts**
6 had **Learning Difficulties**
5 had **Schizophrenia**
4 had a **Personality Disorder**
3 were **Alcoholics**
3 were **Manic Depressives**
2 people had **Agrophobia**
5 people, each with **Anorexia Nervosa, Cerebellar Ataxia, Parkinson's' Disease** and **Post "Psycho-Surgery"**.

It should be noted that each of these 105 cases were assessed as being indirectly related to dystonia but there were a further 492 not specifically related to dystonia but which were recorded to define the 74 other diseases which were present at the same time as dystonia was also present, of which only the most prevalent are shown below

TABLE 26. Comorbidity of patients - not related to dystonia

57 cases of **Hypertension** were recorded - this represented 6.1%
53 cases of **Arthritis** were also mentioned = 5.7%
52 cases of **Thyroid Disease** were specifically noted = 5.5% (more of this later)
24 cases of **Angina** were found, which represents 2.6%
21 cases of **Asthma**, this is equal to 2.2% of the total
19 cases of **Ischaemic Heart Disease** = 2.0%
19 cases of an **Ulcer** (peptic or duodenal) = 2.0%
12 cases of **Cerebrovascular Disease** = 1.3%
10 people have **Diabetes** = 1.1%
10 people have a **Hernia** = 1.1%
10 people have **Migraines** = 1.1%
10 people have **Glaucoma** = 1.1%

There were a further 169 cases exhibiting the remaining 61 different diseases or conditions, but these represent less than 1.0% of the total and have therefore been exempted from the list. They however do represent a number of generally more common conditions, such as **Anaemia, Diverticular Disease, Lumber Pain, Rheumatoid Arthritis** and **Valvular Heart Disease** with 6 cases each.

Major Illnesses

212 patients have had at least one major illness apart from dystonia in their life. 77 people had a second major illness, 33 a third and 16 a fourth. As over two thirds of the patients were women, it was not surprising that the major illness was of the female variety.

However, it was also interesting that other major operations were to do with the neck or the leg / arm or hand or eyelid surgery. These are listed below as the 20 most prolific diseases or illnesses.

TABLE 27. Other Major Illnesses.

Type of illness	Frequency
Hysterectomy	29
RTA	28
Growth removed	15
Stroke	14
Neck - Operation	14
Cancer - unspecified	13
Leg / Arm / Hand Operation	13
Eyelid Surgery	10
Heart attack	10
Cardiovascular Disease	10
Fractures - Bones	9
Appendicitis / ectomy	9
Brain Tumour / Operation	9
Gall stones / Bladder	8
Thyroidectomy	7
Arthritis	7
Unspecified Operation	7
Hiatus Hernia	6
TB	5
Ulcers	5
53 Other diseases (averaging 2.1)	111

Totals =	339

Movements

237 people registered 345 types of movement averaging 1.46 each.

TABLE 28. Types of Different Movements

Type of movement	Frequency	%
Slow pulling / turning	118	34.2%
Slight twitching	72	20.9%
Quick jerking	68	19.7%
Regular trembling	57	16.5%
Mixture of 1 to 4 above	8	2.3%
Other various movements	22	6.4%
Totals =	345	100%

Of the various other movements, the eyelids shut = 5, occasional trembling = 5, rhythmical / continuous movements = 4, cramps or cramping = 3, stiffness = 2, voice whispers = 1, sudden jerks (once) = 1 and Action Induced / Task Specific = 1.

The severity of these movements was measured as well as the degree of control and the pain levels as below. The first question asked was " Please indicate how severe you think your dystonia is at present (compared to others with dystonia that you have seen or compared to how it has been in the past) by circling the appropriate number of the scale below : " and the results are :-

TABLE 29. Severity of Movement.

Severity	Frequency	%
Not severe at all	16	4.8%
1	21	6.4%
2	28	8.5%
3	51	15.5%
4	48	14.5%
5	67	20.3%
6	28	8.5%
7	21	6.4%
8	26	7.9%
9	9	2.7%
Very severe	15	4.5%
Totals =	330	100%

The second question asked was " Please indicate the degree of control you presently have over your **muscle spasms** by circling the appropriate number on the scale below" and the results are :-

TABLE 30. Degree of control over muscle spasms.

Degree of control	**Frequency**	**%**
No control at all	52	15.8%
1	36	10.9%
2	26	7.9%
3	34	10.3%
4	21	6.4%
5	50	15.2%
6	27	8.2%
7	29	8.8%
8	24	7.3%
9	17	5.2%
Complete control	13	4.0%
	Total = 329	100%

The next question was " Please indicate the degree of control you presently have over the **involuntary movement / abnormal postures** of your affected body part(s), by circling the appropriate number of the scale : " and the results are :-

TABLE 31. Degree of control over involuntary movements / abnormal postures.

Degree of control	**Frequency**	**%**
No control at all	43	14.9%
1	33	11.5%
2	28	9.7%
3	28	9.7%
4	25	8.7%
5	32	11.1%
6	27	9.4%
7	22	7.6%
8	21	7.3%
9	10	3.5%
Complete control	19	6.6%
	Total = 288	100%

Pain levels

198 people indicated where the levels of pain were and they were measured based on a standard level in 14 different parts of the body. A total of 5 different parts of the body were available to be covered individually as follows. Remember this is to indicate where actual pain levels are located, not which muscles are affected.

TABLE 32. Where is the greatest pain ?

Where is the pain	**1st No**	**2nd No**	**3rd No**	**4th No**	**5th No**	**Total**
Neck muscles	106	14	4	4	0	128
Shoulder muscles	12	46	11	3	1	73
Arm muscles	12	12	18	1	2	45
Back muscles	6	15	8	6	3	38
Leg muscles	10	12	6	5	5	38
Head	12	16	5	1	1	35
Hand muscles	5	6	4	4	0	19
Foot muscles	4	5	5	3	2	19
Trunk muscles	6	3	2	2	3	16
Jaw muscles	5	2	2	2	0	11
Eye muscles	10	0	0	0	0	10
Face muscles	10	2	1	0	0	13
Mouth muscles	0	3	0	0	0	3
Throat / Larynx	1	0	1	0	0	2
Totals =	199	136	67	31	17	450

This gives a very good picture of the entire body and where in particular the greatest pain resides. As we can see the greatest number of people have pain in the neck, then shoulder and arm. This is mostly because the greatest number of people are affected with torticollis. However this is not the only area where pain resides as can be seen. There was a question about how frequent this pain was, the answers were 'often' (44.9%), 'continuously' (40.3%) and 'infrequently' only 14.8%. Which means that out of 196, 85.2% or 167 of them have pain either continuously or often.

Spontaneous Improvement

One of the questions most often asked of Doctors is "will this go away on its own". Therefore we asked it of the patients and the answers were very interesting as 63 people said Yes, they had had some form of spontaneous improvement. This was 18.1% of the 349 who answered the question so it appears there should be about a 20% chance of some form of remission during the lifetime of the disease.

We gave each person answering two opportunities to give us the year it started, how long it lasted and if it was partial or complete remission while it lasted. The answers were interesting as firstly of the 63 people who responded, the years of the improvement were a very wide spread starting in 1954 up until 1996, with the mean in mid-1990. Secondly there were 19 people who had had a second period of remission, it was much more concise from 1981 to 1996 and the mean was slightly earlier in 1989. The first time 56.5% said it was partial, the remainder said it was complete and the second time 57.9% said it was partial and the rest said it was a complete remission. The answers are tabulated below :-

TABLE 33. Length of time of Spontaneous Remission.

How long it lasted	1st time	2nd time	Total	%
A few days only	9	4	13	15.9
Several weeks	11	3	14	17.1
4 weeks or more	4	-	4	4.9
6 - 8 weeks	2	3	5	6.1
2 months	1	1	2	2.4
3 months	1	1	2	2.4
6 months to a year	6	1	7	8.5
Over a year	1	2	3	3.7
2 - 6 years	9	-	9	10.9
8 years	2	1	3	3.7
15 - 27 years	2	1	3	3.7
Still in remission	15	2	17	20.7
Total =	63	19	82	100%

As can be seen above, apart from those 17 people who were still in remission at the point we completed their study, there were a large number of people (43.9%) who had had a remission from a few days up to 4 years. What we need to know is what caused this remission. Unfortunately in every case it was different and there seems to be no connecting factor, except as one person, still in remission, stated " Don't question it, just accept it and pray it continues."

Quality of Life Results

The overall results generally compliment those already published under several different sources, therefore the results are not detailed below, although some conclusions can be drawn later. Even though the previously published results only relate to the data collected on the first 239 participants, the final results relate specifically to all the 400+ people who completed all the EuroQol and SF36 forms detailed, however the methodology has been published previously and the overall results do not differ at all from those previously published only in the detail of numbers. For these reasons, no data is shown here.

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 19

Genetics

Although genetics was never originally envisaged as being part of this research project; events have moved on since it was first started. At the recent International Symposium held in Miami, Florida in October 1996, a vast amount of new data was brought out regarding this particular subject.

At that point in time, a total of seven new gene nomenclatures had been discovered all relating to dystonia. Since then, as discussed in Chapter 4, a total of 12 genes are thought to be responsible for different types of dystonia throughout the world. Therefore it was decided quite late on during this research to attempt to discover how many people within the region had proof (documented or otherwise) of a genetic ancestry or at minimum a suspicion.

Therefore most people have been asked the question : " Are you aware of anyone else in your family with any form of muscle spasm ? " The results are quite interesting and are given herewith. A total of 556 people (59.3%) answered this question, in one form or another. A total of 210 people (37.8% of those responding) answered in the negative with "no definable trigger". Thus leaving 346 as the number who thought there might have a "trigger" for their dystonia. Although this is discussed in detail elsewhere, a total of 47 people answered positively regarding "a familial disease".

However a total of 173 people (28.7%) answered 'Yes' to the question : " Do you have any family members with dystonia ? " 423 (70.3%) answered No and a further 6 answered Unknown - as they were adopted. Adding in the people who had more than a single relation affected, a total of 265 different relations were affected as follows :-

TABLE 34. Relationship to others affected

53 had a **sister** affected in some way
45 had a **mother** affected
44 had a **maternal relation** affected
43 had a **paternal relation** affected
33 had a **father** affected
18 had a **brother** affected
14 had a **daughter** affected
10 had a **son** affected
4 had a **nephew** or **niece** affected and
1 had his **wife** affected (very unusual to have a married couple separately affected)

---

265 people had a relation affected in some way

This however did not show the whole picture as a number of people did not have a confirmed or diagnosed dystonia, as follows :-

TABLE 35. Type of spasm reported in the allegedly affected people.

56 people had a **focal dystonia**	)
39 people had **generalised dystonia**	) A total of 98 people had a definite
2 people had **segmental dystonia**	) 'diagnosed' dystonic spasm
1 person had a **hemi-dystonia**	)
71 people had an **undiagnosed spasm**	
63 people had **another neurological disorder**	
33 people had **undefined spasms**	
----	
265 different people with different types of spasms	

Therefore the only proven (at this time) number of people with a known dystonic family pattern was 98 and this figure is based on known and provable diagnoses and relates to 10.5% of the known dystonic population. It is however very likely that a figure approaching over 200 is more realistic. This relates to 16.6% of those who were able to answer the question. This figure is currently in excess of current known patterns of familial dystonia and is an area that should be explored and expanded further.

In particular, it would be very interesting to examine the 71 people with an undiagnosed spasm and the 63 people who were related to the patient with dystonia but who had another neurological disorder. This might well pick up a number of other dystonics who had been previously misdiagnosed and often treated as well.

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 20

Social Implications

Social and Economic Groupings

Social class can be defined in a number of different ways using a number of different classifications. A review of the common classifications of occupations in social classes used in empirical research in Britain shows that only one meets the requirements of the current research into dystonia. " *Most commercial, social, advertising and consumer research enterprises use the social grading of occupations, originating from the Institute of Practitioners in Advertising ie, Marketing Research : MR.*" (Reid, 1989). However the main reason that this classification has been used in this research project is that it is the only one in common usage that has the non-employed category of retired, on disability benefit, income support and other forms of allowance.

TABLE 36. Social and Economic Group (SEG) Categories.

A :	Professional - defined as successful business persons (eg, self-employed / manager / executive of large enterprise), higher professionals (eg, bishop, surgeon, medical specialist, barrister, accountant), senior civil servants (above Principal) and local government officers (chief executive, treasurer, town clerk)
B :	Managerial - defined as senior, but not the very top, people in the same area of employment as category A.
C1 :	White Collar Workers - defined as small trades people, non-manual, routine administrative, supervisory and clerical.
C2 :	Blue Collar Workers - defined as skilled manual workers, at the top of their trade or skill.
D :	Semi-skilled and unskilled workers - defined as all those people currently in work but not previously categorised.
E :	Unwaged - defined as those at the lowest levels of subsistence, including the retired, those on social security because of sickness or unemployment, and students.

By using these social gradings of occupations, this research has been able to give an insight into a number of different social and economic factors resulting in a large number of people with dystonia falling into the lowest category, ie, being unemployed, prematurely retired, on incapacity benefit or income support. However this group also includes a few wealthy retired folk with large occupational pensions which were noted.

The following table shows the SEG of the study population and demonstrates very clearly that even though the SEG of the Head of the Household (HoH) at his or her highest level is roughly the same as the National Average, there was greater weighting in SEG C_1 amongst the patients at their highest level.

This is due to the fact that 66.1% of the study population are female (confirming previous studies that dystonia tends to affect roughly double the number of women to men) and that the majority of female employment is often categorised as 'White Collar Worker' or with 'Clerical Status'.

The following table also shows there is a movement downwards in patient SEG status. Every category shows a reduction over time, from Questionnaire No 20 at the start of the study to Questionnaire No 25 at the end, except 70.2% of the study population start life in the lowest social and economic group (ie E - Unwaged) and this increased to 72.8%. All correlations across the table by Chi-Square Test come out as highly significant. These Chi-Square results are 'goodness of fit' tests against the national average distribution.

TABLE 37. Social and Economic Groupings (1996)

Social Economic Group	Q're 20 1st SEG	Q're 25 2nd SEG	Highest Patient	Highest HoH	Nat. Ave.
A : Professional	0.7%	0.8%	1.3%	3.1%	3%
B : Managerial	3.1%	3.0%	8.5%	17.9%	14%
C_1 : White Collar	14.6%	12.5%	38.9%	22.1%	22%
C_2 : Blue Collar	4.1%	2.3%	13.5%	32.4%	28%
D : Semi-skilled / Unskilled	5.2%	4.2%	21.6%	19.7%	18%
E : Unwaged, etc.	70.2%	72.8%	14.0%	2.6%	15%
Deceased	2.2%	4.5%	2.2%	2.2%	-
n	460	265	459	458	
Chi-Square	1214.82	757.45	325.47	269.08	
D.F.	6	6	6	6	
Significance p =	< .001	< .001	< .001	< .001	

The above figures were derived from the data available in 1996. A further comparison was made using the latest data available in 1998. The following table shows that the figures are not dissimilar. Furthermore when the first two columns were compared using a Spearman Correlation, the results indicated that 448 sets were significant, p <.0001.

TABLE 38. Social and Economic Groupings (1998)

Social Economic Group	Q're 20 1st SEG	Q're 25 2nd SEG	Highest patient	Highest HoH	Nat. Ave.
A : Professional	0.6%	0.4%	1.5%	3.2%	3%
B : Managerial	2.6%	1.8%	9.2%	19.2%	14%
C_1 : White Collar	10.8%	7.8%	39.0%	22.0%	22%
C_2 : Blue Collar	4.1%	1.6%	13.8%	32.4%	28%
D : Semi-skilled / Unskilled	4.6%	2.7%	21.7%	20.1.%	18%
E : Unwaged, etc.	75.6%	78.9%	15.0%	3.0%	15%
Deceased	1.7%	6.9%	-	-	-
n	655	450	480	463	
Chi-Square	2039.4230	1548.4535	236.3750	290.6263	
D.F.	6	6	5	6	
Significance p =	< .001	< .001	< .001	< .001	

Looking at the above table, one is struck by the similarity to the previous results two years previously, thus showing that there is little percentage change over time, allowing for the ageing process regarding retirements, deaths, etc and that the results are consistent with the hypothesis that dystonia moves people socially downwards over a period of time, as the disorder increases in severity and that the accepted prognosis, although sometimes slow, is almost inevitable.

Depression, Anxiety and Pain

In the CUA, a number of subjects (59.8%) were measured with moderate anxiety or depression and 8.8% had severe anxiety or depression. Over 60.4% had moderate pain or discomfort with 21.8% experiencing severe pain or discomfort (Gudex et al, 1997).

Comparing the EuroQol and SF36, the results were found to be very compatible (Butler et al, 1995). Self-rated health (VAS) and tariff scores were highly correlated with all SF36 dimensions. Self-rated health was most highly correlated with SF36 social functioning (SF), while the tariff score was most highly correlated with SF36 physical functioning (PF). Spearman correlation between self-rated health (VAS) and tariff score was 0.57 ($p < 0.001$).

The most significant figures are those relating to the Pain threshold levels compared to the general population. Only 3.8% of the general population (Dolan et al, 1995) suffer severe pain, whereas 21.8% of the injection group and 10.7% of the CUA control reported severe pain levels.

A comparison between the results can be shown thus :-

TABLE 39. Comparison of Pain and Discomfort to Anxiety and Depression levels on EuroQol vs SF36.

	Injection Group	Control Group	General Population
EuroQol	n = (102)	n = (28)	n = (3395) [1]
Pain / Discomfort	%	%	%
None = 1	17.8 (18)	14.3 (4)	67.0 (2275)
Moderate = 2	60.4 (61)	75.0 (21)	29.2 (991)
Severe = 3	21.8 (23)	10.7 (3)	3.8 (129)
Anxiety/Depression	%	%	%
None = 1	31.4 (32)	53.6 (15)	79.1 (2685)
Moderate = 2	59.8 (61)	39.3 (11)	19.1 (648)
Severe = 3	8.8 (9)	7.1(2)	1.8 (61)
SF36	%	%	n = (542) [2]
Bodily Pain	47.9 (102)	56.4 (28)	76.9 (542)
Mental Health	58.9 (101)	65.4 (28)	73.7 (542)

[1] Taken from the Measurement and Valuation of Health report 1994 - using adults, non-institutionalised, in Great Britain (not N.I.) aged 18+ years.

[2] taken from Jenkinson et al, 1993.

The SF36 chart above again confirms the EuroQol findings, in that people with dystonia (both injection group and control group) have a lower score in all areas as compared to the general population. Even though dystonia is known to be a debilitating disorder and patients have pain due to their muscle spasms, this is the first time those pain thresholds had been quantified.

Social Isolation

Previous research (Butler and Duffey, 1996a) had shown that most people with dystonia often felt a sense of isolation as their disorder is predominantly visual in presentation. This phenomenon was measured as part of the Nurse Practitioner Project and more particularly in the Psycho-Social Research programme. Although the author is not a qualified psychologist, the following is an interpretation of the results.

58.5% of subjects live with their spouse or partner. Of the remaining 41.5%, 17.5% are single, 14.4% are widowed, 8.1% are divorced and 1.5% are separated.

However 24.6% of this population live totally alone, which leaves 16.9%, who have someone else in the house with them, either children or parents. Eight people live with a sibling and one with an aunt.

Nevertheless these raw statistics do not show the whole picture. Social isolation relates as much to a 'feeling' of isolation. The Functional Disability Questionnaire (FDQ) social score results showed 11.4% not at all affected, a further 45.4% only mildly affected, 32.4% moderately affected and 10.8% severely affected.

Social isolation can affect other pursuits, the FDQ Leisure score results showed that, although 21.3% of the subjects' leisure activities were not affected by their Torticollis, 44.8% were mildly and 23.7% were moderately affected, whereas 10.2% were so severely affected that they had practically no leisure activities outside the home at all.

Feeling 'uncomfortable' in social situations was measured and 12.0% of subjects reported feeling 'very uncomfortable'. On a scale from 0 (not at all uncomfortable) to 10 (very uncomfortable) - the majority (54.4%) were in the 5 to 10 range.

Finally, the measure of 'disfigurement' showed that on a scale from 0 (not at all disfigured) to 10 (extremely disfigured) - a significant minority (24.8%) felt that they were in the 7 to 10 range. All of which has produced scores on the Self Esteem Scale (SES) which show that over 89.8% of subjects are in the Negative Self Esteem range of 20 to 40, with only 10.2% being in the 0 to 20 Positive Esteem range.

There needs to be a more detailed analysis of these results but the author does not feel competent nor qualified to attempt such an analysis, which will be done by Dr Marjan Jahanshahi, a Clinical Psychologist from the National Institute of Neurology at Queen Square, London at a later date and therefore do not form part of this dissertation.

The results of the social research have been previously described in a number of different publications, but this is the first time that they have been totally described en bloc. Because the work has been so wide ranging, it is best described in each section according to the method of obtaining the research.

Marital Status, Children and Hereditary

Of the 937 people in the study, only 757 (80.8%) described their marital status. Of these, 438 (57.9%) were married at the time of the interview, 126 (16.6%) were single, 111 (14.7%) had been widowed, 60 (7.9%) were divorced and 11 (1.5%) were each either cohabiting or separated.

The number of children averaged out at 1.8 per head, with a total of 287 male and 270 female children spread across those who answered the question about " how many children do you have of each sex ? " and " how many currently live with you " , as follows :-

TABLE 40. Number of children of the patients.

No of children	Male	Male (at home)	Female	Female (at home)
0	202	392	217	404
1	152	70	176	70
2	99	23	72	12
3	23	3	12	0
4	10	1	7	1
5	2	0	3	0
6	1	0	0	0
Total =	489	489	487	487

The current age of the children reflect the current age of their parents, thus the eldest first born was born in 1927 with the youngest in 1996 and the youngest last child was born in 1996 with the eldest of the youngest children being born in 1931. However there is an average birth year for the eldest (born in 1968) which equals the average for the youngest being also 1968. This greatly reflects the average age of the patients as a whole being 56.61 years (as of 1st January 1999) having been born in 1942 making them on average 25.5 years when they first became parents.

Time off work

Before the advent of the Dystonia Nurse Practitioner, a number of patients had to take time off work in order to attend the clinics and it was thought to be a worthwhile exercise to measure this. Therefore from May 1993 until 1996, over 500 people were asked if they had to take time off work because of their dystonia.

A total of 85 people had had to take time off work in order to attend the clinic and a further 64 helpers had also had to take time off work in order to either bring them to the clinic or to accompany them. It worked out thus :-

TABLE 41. Time off work taken to obtain treatment in previous 4 weeks

Time off work	No of people	No of helpers
One hour	5	0
Two hours	12	9
Three hours	2	3
Four hours	16	30
Five hours	1	0
Six hours	1	1
Seven hours	1	0
Eight hours	47	21
	---	-- ----
Totals =	85 (Ave 5.8 hours)	64 (Ave 5.0 hours)

The same question about the number of days they had had to have off work before visiting the clinic was then asked and if any of their partners had had to take time off work also. The answers were as follows :-

TABLE 42. Days off work for patients and partners.

Days off work	No of people	No of partners
One day	13	21
Two days	1	9
Three days	2	0
Four days	1	1
Five days	2	2
Six days	0	1
Seven days	1	0
Eight days	2	0
Fifteen days	0	1
Thirty days (all the time)	38	3
Total =	60 (Ave 20 days)	38 (Ave 4.2 days)

Travel to and from hospital

The distance the patient had had to travel to and from the hospital was then measured by asking the distance they lived away from the hospital one way only. The distance was then measured again after a period of time averaging two years and compared the difference, if any. The results were :-

TABLE 43. Distance travelled to hospital(s).

Distance travelled	To hospital	To hospital (later)
Home visits by DNP	0	42
1 - 5 miles	132	40
6 - 10 miles	88	27
11 - 15 miles	85	27
16 - 20 miles	41	24
21 - 25 miles	13	5
26 - 30 miles	11	3
31 - 50 miles	26	11
51 - 75 miles	19	4
76 - 100 miles	18	5
101 - 150 miles	12	2
Over 150 miles	10	4
Total =	455	197

The time it took to travel to the hospital and the method was also noted as follows :-

TABLE 44. Travel time to old and new hospitals

Time to hospital in mins	No. to old hospital	No. to new hospital
Home visits by the DNP	0	45
Up to 5 minutes	5	2
6 - 10 minutes	42	11
11 - 15 minutes	46	11
16 - 20 minutes	73	21
21 - 30 minutes	108	39
31 - 45 minutes	57	21
46 - 60 minutes	42	21
Up to 2 hours	53	17
Up to 3 hours	20	7
Over 3 hours (1 way only)	9	2
Total =	455	197

TABLE 45. Method of travel to old and hospitals

Method of travel	No. to old hospital	No. to new hospital
Own Car	105	31
Family Car	137	51
Friend's Car	28	7
Public - mainly Bus	82	35
Public - mainly Train	31	8
Ambulance / Hospital Car	42	14
Taxi	20	3
Bicycle	3	1
Walked or pushed in wheelchair	7	1
Aeroplane	1	1
Home visits by DNP	0	45
Totals =	456	197

Major events

One of the great things about this research is the fact that one has been able to ask a number of questions about quite personal things that might be considered 'major events'. This research has been able to ask these questions and probe into often personal events. An example of this were questions relating to major events in their lives that might have had a bearing on their getting dystonia. The answers are quite interesting.

TABLE 46. Type of major event and what they think caused I.T.D.

Major event	No. of people	What they think caused I.T.D.
Don't know	-	147
Stress - caused by ??	45	45
Accident at work / home	17	17
Other neuro disorder	17	17
Bereavement	27	15
Hereditary, familial	-	15
Use of drugs	-	13
RSI	10	10
RTA	9	9
Fall	13	7
Major head injury	7	6
Surgery	5	5
Born with it	-	5
Exhaustion at work	4	4
Alcoholism	4	4
Trapped nerve	-	3
Pregnancy	3	2
Chemical exposure	-	2
Viral infection	-	2
Minor head injury	17	-
Non-related condition	9	-
Stroke / Cerebellar / SAH	7	-
Menopause	3	1
Drug overdose	2	-
W.W.II birth / bomb	2	-
Heart attack	1	1
Gave up smoking	1	1
Measles	1	-
Flu injections	1	-
Divorce	1	-
Severe migraines	1	-
Schizophrenia	1	-
Cerebella Ataxia	1	-
Physical strain	-	1
Puberty	-	1
Slipped disc	-	1
Total =	209	336

The interesting things here will be discussed at length in the following section but there are at least 189 people with a definite answer as to why they think they got dystonia. Apart from the 15 who stated that it was definitely hereditary, a number have come up with the same ideas, which can all be brought back to a definite stress related period in their life, eg., 45 stress related, 17 an accident and 15 a bereavement = 77 (40.7%).

Then there are the 17 others for whom other people, ie doctors, relate their dystonia to another neurological condition, or the 13 who believe their dystonia was caused by the use of drugs. A couple of other questions also had quite interesting answers. *" Has anyone ever suggested this condition was caused by some mental problem ? "* brought out a very interesting response as 127 people (38.4%) answered Yes. When asked if they thought " *their own GP knew enough about dystonia* ", an interesting 16.5%, or 54 people, also responded with a Yes.

Living Standards

A total of 346 people answered the question " *Who do you live with ? "* as follows :-

TABLE 47. With whom do you live.

With spouse	200	57.8%
Alone	85	24.6%
With parents	35	10.1%
With children	10	2.9%
With sibling	8	2.3%
With boy / girl friend	7	2.0%
With aunt	1	0.3%
Total =	346	100%

The question was then asked " *in what type of accommodation* ", the answers were :-

TABLE 48. In what type of accommodation.

House	279	80.6%
Flat	40	11.6%
Bungalow	21	6.1%
Residential Home	3	0.9%
Terraced Cottage	1	0.3%
Bed and Breakfast	1	0.3%
Prison	1	0.3%
Total =	346	100%

Finally, the question was asked, *" Does this accommodation meet your needs, if not, why not ? "* and the answers were related mainly to the problem of having to cope with stairs, as the disease progressed. 29 people answered the question, 44.8% of whom stated that "*the stairs were the main problem*". Then three people said that "*there was no help at home*" and another three said they needed "*more things adapted*". Two people each said that "*there was no WC downstairs*", "*the house was too big*" or "*it was currently being adapted*". One person each complained that "*there was no shower and the house had stairs*", "*the doors were too small to accommodate the wheelchair*", "*there were outside steps making it difficult to get in or out*" and finally that "*they now had to sleep downstairs as they could no longer manage the stairs*".

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 21

Economic Implications

Employment Status

The full employment status of 671 (71.6%) of the 937 people in the survey are known and a further 450 had their employment status checked again after a period averaging 2.42 years. The results prove that dystonia is a debilitating disease and that it can have a devastating effect upon peoples life and in particular their employment status.

The following table gives an excellent perspective on the way in which dystonia can move people over a period of time regarding their employment status. There is a much higher proportion of retired people than their ages would indicate and a definite movement to various forms of retirement, particularly due to ill health. As can be seen over time, a number of areas have increased and are directly related to the decrease in others. For example, there is a 5.8% drop in full time employment, which is more than matched by the increase in retirements from 42.8% to 54.9%.

TABLE 49. Employment Status.

Employment Status	At the beginning	After an average 2.42 years
Full time employment	90 (13.4%)	34 (7.6%)
Part time employment	35 (5.2%)	23 (5.1%)
Unemployed	39 (5.8%)	14 (3.1%)
Self employed	15 (2.2%)	2 (0.4%)
Unwaged	57 (8.5%)	23 (5.1%)
Retired	287 (42.8%)	247 (54.9%)
Retired - ill health	58 (8.6%)	35 (7.8%)
On long term sick	79 (11.8%)	36 (8.0%)
Deceased	11 (1.6%)	36 (8.0%)
TOTAL =	671 (100%)	450 (100%)

Financial Income

There is a definite difference however when it comes to the income of dystonia patients compared to the general population. Dystonia patients have some of the lowest incomes within the population as a whole. Of those with a known and provable income (45.5%), the average income in 1994 was a little as £5,250 per annum and it had increased only to £5,550 by 1997. The extremes did not fair much better, with a low going from £200 to £500 a year (personal income only) to a high going from £39,000 to £42,900. It is important to remember however that the 75% figure related to only between £8,500 and £9,000 per annum. It can therefore be forcefully seen that the dystonic population is generally one of the poorest in the country as a whole.

This is well amplified by the following chart which shows the number of people within the various Social-Economic Groups (Reid, 1989).

TABLE 50. Socio-Economic Groups.

SEG	At the beginning	After an average 2.42 years
A : Professional	4 (0.6%)	2 (0.4%)
B : Managerial	19 (2.8%)	8 (1.8%)
C1 : White Collar	74 (11.0%)	35 (7.8%)
C2 : Blue Collar	28 (4.2%)	7 (1.6%)
D : Semi-Retired	31 (4.6%)	12 (2.7%)
E : Retired	503 (75.1%)	353 (78.3%)
Deceased	11 (1.6%)	34 (7.5%)
TOTAL =	670 (100%)	451 (100%)

The main factor is that when one takes the dystonic population as a whole and compares it with the general population there is no discernible difference.

TABLE 51. Comparison of Highest Patient Status to the General Population

SEG	Highest Patient Status	General Population
A : Professional	1.7%	3%
B : Managerial	9.1%	14%
C1 : White Collar	39.1%	22%
C2 : Blue Collar	13.7%	28%
D : Semi-Retired	21.5%	18%
E : Retired	14.9%	15%
TOTAL =	100%	100%

The subjects' income has been derived from four different sources. Firstly all consenting subjects were interviewed at the earliest opportunity and all (then current) forms of income were noted and calculated to produce an annual income in £ sterling for the patient themselves (Questionnaire No 20). Where possible family or joint income was not shown, which resulted in a few individuals being shown with zero income. In some cases only the benefit title, not the level of subsidy, was available.

Secondly, the published levels of state subsistence were used to calculate and confirm definite levels of income against subjective or unknown answers to previous questions.

Thirdly, a final questionnaire (Questionnaire No 25) was sent out in 1996/97 to all subjects in the study asking if there had been any changes in the past years to income, employment, treatment or attendance at a clinic or hospital and the results were correlated against previously obtained data.

Finally, the time between the original interview and the subsequent update was calculated for each person and where 'no change' had been noted by the subject, an increase based on H.M. Government's published inflation figures was produced. The criteria used is shown on the table below.

TABLE 52. Criteria used for inflation

The difference in time	Calculated inflation during that time
3.2 years to 2.7 years	Income increased by 10%
2.6 years to 1.6 years	Income increased by 7%
1.5 years to 0.6 years	Income increased by 3%
0.5 years to 0.0 years	Income increased by 0%

Because at least four fiscal year ends have passed where income tax and benefits are calculated from 5th April each year, there was an overlap and/or adjustment required for each individual case. The average increase over the period was 6.25%, however a number of persons had a dramatic reduction in income and this is reflected in the results.

A few had an increase above levels of inflation due to a promotion, a new job or regaining employment having been previously unemployed. With very few exceptions, all subjects co-operated in this economic aspect of the research and the degree of participation was 80.7% with only 16.2% actually declining (3.1% were deceased at the time).

It has proved very interesting comparing the patient's actual annual income over time. Despite the fact that most incomes have risen by an average 6.25% over the period of the study, there is a universal diminution of income up to the £15,000 per year band. Half the study population have less than £6,000 ($9,900) to live on and only one third have more than £8,000 p.a. ($13,000).

The mean income from 1993 (£7,467.08) to 1997 (£7,406.58) showed a difference over the 4 years of (minus) £527.19. No comparison was possible for income over the £30,000 per year band. However the Chi-Square Test results show all the comparable differences to be significant.

TABLE 53. Chi-Square Tests : Comparisons in Income over time.

Year	**1993**	**1997**
n	423	244
Chi-Square	438.3499	237.5328
D.F.	12	12
Significance	p = < .001	p = < .001

In the band £3,000 - £7,000 there are up to 5.5% of patients who have had significant changes in their income downwards and this, at a time, when they are already classed as one of the lowest income groups in UK society as a whole. There is however some indication that recent improvements in treatment therapies are bearing fruit amongst the study population.

Economic Benefits

This will be discussed in greater detail later on, but further sensitivity analysis has revealed that the majority of the subjects reap no direct economic benefit from any form of therapy or treatment. There is however a small but significant number of subjects who remain in work as a direct result of Botulinum Toxin therapy.

For example; in 1994, 19.6% of those patients in work had an average of 77.8% time off work before the start of Botulinum Toxin (BT) therapy, whereas only 14.2% had to have an average 48.7% time off work after BT therapy.

To interpret these figures in more detail, this means that during the six weeks prior to an interview, 19.6% of those patients who were still registered as employed (albeit most were on sick leave) and who had not yet received their first set of Bot. Tox injections, had had an average of 77.8% time off work during that six week period.

This same group of subjects, who were either seen at the clinic during the CUA or had completed a second interview, indicated that 14.2% of them had had to have only 48.7% time off work after the Bot. Tox therapy had become effective. This still means that a proportion of those registered for work, had eventually to stop work and either become retired due to ill health or go onto long term sick leave, but it also indicates that for a number of patients currently employed, they were able to return to work and continue to benefit financially from that position.

Treatments generally were found to only stabilise the condition and make it bearable. In 1993/94 very few subjects found any of the treatments worked sufficiently well to put them back into work, however this has changed as more people get diagnosed earlier. By 1999, more people are currently remaining in work as a result of Botulinum Toxin treatment.

This aspect will be discussed in detail later, but in essence it is as a direct result of the length of time it takes to get a correct diagnosis after the presentation of the first symptoms to the medical profession.

There was a direct correlation between early diagnosis and the ability to remain in work. The longer it takes for diagnosis, the further the condition deteriorates and therefore the greater the chance that the subject has to finish work and opt for either long term sickness benefit or, in most cases, eventual early retirement.

The results of the economic implications of this research have been published in many different journals, but this is the first time that all the results have been combined and shown within one document. Nevertheless it is important to show them in a constructive order, therefore they are shown in the order obtained.

The benefits received by 367 patients was specifically measured and the results are shown below. A total of 684 benefits are currently shown which represents each person receiving on average 1.9 benefits in their own right. The benefits shown in the following table are graded according to their order of priority.

TABLE 54. Different benefits received.

Benefit	Number	%
Retirement / Widows	157	23.0
Invalidity Benefit	105	15.4
Occupational Pension	100	14.6
Mobility Allowance	77	11.3
Income Support	49	7.2
Disabled Living (DLA)	44	6.4
Attendance Allowance	38	5.6
Severe Disability	28	4.1
Child Benefit	27	3.9
Sickness Benefit	24	3.5
Industrial Injury	9	1.3
MOD Disability	6	0.9
Invalid Care (ICA)	5	0.7
Disabled Working	4	0.6
Loss of Earnings	4	0.6
Family Credit	4	0.6
HM Prison Work	1	0.1
Cancer Victim	1	0.1
Rent Allowance	1	0.1
TOTALS =	684	100%

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 22

Psychological Profiles

The psychological profiles have been designed, as previously stated, by Dr Marjan Jahanshahi, nevertheless all the work undertaken here is the author's own, although patience should be envisaged as the author is not a psychologist. There were a number of specific instruments given to the author and therefore this chapter will merely detail the results of each instrument, as determined by Dr Jahanshahi.

Psychiatric Problems

51 people reported they had previously had some form of psychiatric problem. The vast majority were found to be suffering from depression (42.4%) with the next highest number from anxiety (19.7%). The rest had, in order to priority, the following disorders; 5 cases of nervous breakdown, 4 cases of 'imagined' dystonia, 3 cases of depression and anxiety, 2 cases of schizophrenia and 2 cases of agrophobia. Then there were single cases of obsessive neurosis, alcoholism, panic attacks, paranoia, amnesia, loss of speech, tremor of the head and anorexia. There seems to have been a fairly even spread over time, with the earliest case in 1951 and the last in 1995, with 7 cases in the 1950's, 9 in 1960's, 12 in 1970's, 17 in 1980's and with 20 cases reported in the first half of the 1990's.

The suggested remedies were not surprisingly 69.2% for drugs and medication. Then in order of priority came 6 cases for counselling, 6 cases for shock treatment (ECT), 4 cases for psychotherapy, 3 cases for injections (unknown type) and finally 1 case of speech therapy.

332 people answered the question on antidepressants and 42 people (12.7%) were specifically prescribed these drugs. Although only 35 people (10.5%) said they were for the control of anxiety.

Living with Dystonia Questionnaire

A total of 333 people completed this questionnaire in full, representing 35.9% of the total number of participants, although it should be remembered that no questionnaires have been issued since 1996, ie after the first 4 years and first 500 people, therefore the return rate is effectively 66.6%.

The results are shown in detail below, which will be discussed in the next section of this thesis in further detail.

TABLE 55. Social Scores.

Social Scores :	Not Applicable	Not at all affected	Mildly affected	Moderately affected	Severely affected
5 Transport	21	145	59	54	54
7 Conversation	2	100	83	96	52
9 Restaurant / Pub	7	109	70	85	62
16 Eating in public	2	189	68	42	32
17 Dinner Parties	37	102	62	52	80
20 Crossing Roads	2	148	64	64	55
22 Drinking	3	202	62	42	24
24 Going to the Theatre	38	119	61	52	63
Totals (2,664) =	112	1114	529	487	422

The Social Scores were in total :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
3	1	5	1	7	5	8	31
9	25	10	26	11	18	12	18
13	14	14	17	15	15	16	18
17	16	18	17	19	9	20	14
21	14	22	11	23	12	24	15
25	8	26	5	27	2	28	11
29	3	30	2	31	2	32	3

Total = 333 for all scores.

TABLE 56. Physical Scores.

Physical Scores :	Not Applicable	Not at all affected	Mildly affected	Moderately affected	Severely affected
2 Housework	10	132	70	74	47
4 Running	33	89	37	52	122
8 Carrying	1	121	63	94	54
12 Walking	4	122	64	75	68
13 Sexual Intercourse	88	161	34	18	32
19 Hobbies	15	101	73	54	90
26 Playing sports	91	72	32	36	102
27 Going up stairs	3	153	80	62	35
Totals (2,664) =	245	951	453	465	550

The Physical Scores were in total:-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
5	1	6	16	7	50	8	23
9	10	10	16	11	16	12	13
13	23	14	19	15	13	16	11
17	10	18	15	19	10	20	17
21	15	22	12	23	8	24	9
25	4	26	5	27	8	28	9

TABLE 57. Self-Care Scores.

Self-Care Scores :	Not Applicable	Not at all affected	Mildly affected	Moderately affected	Severely affected
1 Dressing oneself	1	223	56	38	15
10 Brushing ones teeth	8	238	44	27	16
15 Washing oneself	4	254	42	24	9
21 Shaving / making up	8	169	73	48	35
25 Self co-ordination	3	154	73	55	48
Total (1,665) =	24	1038	288	192	123

The Self-Care Scores were in total :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
3	1	4	9	5	101	6	53
7	35	8	32	9	18	10	9
11	12	12	15	13	6	14	8
15	9	16	12	17	4	18	3
19	4	20	2				

TABLE 58. Leisure Scores.

Leisure Scores :	Not Applicable	Not at all affected	Mildly affected	Moderately affected	Severely affected
3 Watching television	0	177	83	45	28
6 Writing	2	140	68	67	56
11 Reading	0	151	74	62	46
Total (999) =	2	468	225	174	130

The Leisure Scores were :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
3	71	4	42	5	54	6	53
7	29	8	25	9	25	10	19
11	7	12	8				

TABLE 59. Other Scores.

Other Scores :	Not Applicable	Not at all affected	Mildly affected	Moderately affected	Severely affected
14 Driving	150	73	40	27	43
18 Typing	153	86	37	23	34
23 Riding a bicycle	138	80	21	19	75
Total (1,332) =	532	311	130	105	254

These three scores were removed from the regular matrices as there were very high Not Applicable scores of 150 (45.0%), 153 (45.9%) and 138 (41.4%) respectively. The highest Not Applicable score otherwise is only 91 for Playing Sports and 88 for Sexual Intercourse.

Disabling, Uncomfortable and Disfigurement

Of the 332 people who answered these questions, the spread was as follows :-

TABLE 60. Disabling / Uncomfortable / Disfigurement.

	Disabling	Uncomfortable	Disfigurement
Not at all	26 (7.8%) 7th	28 (8.4%) 5th	42 (13.0%) 1st
Level 1	37 (11.1%) 4th	26 (7.8%) 6th	39 (12.1%) 3rd
Level 2	44 (13.3%) 1st	48 (14.4%) 1st	35 (10.8%) 5th
Level 3	38 (11.4%) 3rd	33 (9.9%) 4th	41 (12.7%) 2nd
Level 4	35 (10.5%) 5th	17 (5.1%) 10th	26 (8.0%) 8th
Level 5	43 (13.0%) 2nd	35 (10.5%) 3rd	37 (11.5%) 4th
Level 6	25 (7.5%) 8th	25 (7.5%) 7th	23 (7.1%) 9th
Level 7	29 (8.7%) 6th	25 (7.5%) 7th	35 (10.8%) 5th
Level 8	23 (6.9%) 9th	8 (11.4%) 11th	31 (9.6%) 7th
Level 9	13 (3.9%) 11th	18 (5.4%) 9th	9 (2.8%) 10th
Very / Extremely	19 (5.7%) 10th	40 (12.0%) 2nd	5 (1.5%) 11th
Total =	332 (100%)	333 (100%)	323 (100%)

As can be seen, Level 2 is the highest in both *Disabling* and *Uncomfortable*, but we can see where the difference is coming next, where 40 people consider the next highest level of uncomfortable is the most uncomfortable level. This is highly significant and should be borne in mind at all times. The pattern for *Disfigurement* is different, where generally speaking the pattern is gradually increasing as its preponderance decreases. Nevertheless in all cases there is an overall spread and generally none of the figures are particularly encouraging.

Body Concept Scales

This is a chart where between 305 and 309 patients (305 meaned) put where they think they are against 22 different opposites, for example, graceful vs awkward, or lethargic vs energetic, or swift vs sluggish, on a 7 point scale. The results are :-

TABLE 61. Speed / Strength Scales.

Speed / Strength Scale	-ve, very	-ve, fairly	-ve, slightly	Equally descrip.	+ve, slightly	+ve, fairly	+ve, well
3 Swift vs Sluggish	10	28	29	67	70	63	38
7 Fit vs Unfit	16	32	44	60	63	39	52
10 Weak vs Strong	27	41	50	67	57	34	30
13 Slow vs Fast	12	33	36	75	53	50	49
15 Healthy vs Sick	40	54	41	65	56	28	22
21 Active vs Passive	43	58	33	74	41	34	23
22 Delicate vs Robust	31	44	48	88	44	33	17
Total (2,142) =	179	290	281	496	384	281	231

The Speed / Strength Scores were :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
7	3	8	1	9	3	10	3
12	3	13	3	14	5	15	8
16	7	17	6	18	8	19	7
20	3	21	7	22	10	23	8
24	14	25	11	26	12	27	6
28	16	29	15	30	16	31	15
32	10	33	11	34	14	35	10
36	3	37	8	38	5	39	5
40	11	41	5	42	10	43	3
44	4	45	5	46	2	47	3
48	2	49	4	Total = 305			

TABLE 62. Postural / Movement Scale.

Postural / Movement **Scale**	**-ve, very**	**-ve, fairly**	**-ve, slightly**	**Equally descrip.**	**+ve, slightly**	**+ve, fairly**	**+ve, well**
1 Graceful /Awkward	11	29	33	87	56	38	52
6 Rigid vs Flexible	21	39	41	76	49	41	38
8 Unbal vs Balanced	43	47	33	50	45	48	43
9 Steady vs Unsteady	46	46	26	43	48	46	54
16 Clumsy /Well cood	37	49	33	50	46	46	44
17 Straight vs Twisted	70	36	22	55	44	37	42
18 Mobile v Immobile	84	55	35	49	40	22	20
20 Uncontr vs Control	41	54	43	54	48	27	38
Total (2,450) =	353	355	266	464	376	305	331

The Postural / Movement Scores were :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
7	3	8	2	9	4	10	7
11	5	12	5	13	6	14	5
15	5	16	5	17	8	18	6
19	2	20	8	21	9	22	7
23	5	24	12	25	14	26	11
27	12	28	16	29	10	30	12
31	9	32	10	33	7	34	5
35	8	36	6	37	10	38	5
39	12	40	6	41	4	42	7
43	6	44	4	45	8	46	4
47	4	48	3	49	8	Total = 305	

TABLE 63. Evaluative / Aesthetic Scale.

Evaluative / Aesthetic Scale	-ve, very	-ve, fairly	-ve, slightly	Equally descrip.	+ve, slightly	+ve, fairly	+ve, well
2 Lethargy vs Energy	20	32	41	72	60	52	29
5 Ugly vs Beautiful	14	21	60	126	45	21	18
14 Poised v Unpoised	17	41	40	75	54	41	38
19 Flawed vs Perfect	16	27	35	77	63	54	33
Totals (1,222) =	67	121	176	350	222	168	118

The Evaluative / Aesthetic Scores were :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
4	1	5	2	6	3	7	5
8	5	9	6	10	9	11	10
12	10	13	18	14	19	15	23
16	32	17	28	18	19	19	23
20	20	21	17	22	19	23	3
24	9	25	14	26	3	27	3
28	4	Total = 305					

TABLE 64. Tension Scale.

Tension Scale	-ve, very	-ve, fairly	-ve, slightly	Equally descrip.	+ve, slightly	+ve, fairly	+ve, well
4 Calm vs Agitated	36	38	39	62	48	50	34
11 Relaxed vs Tense	26	30	24	59	45	71	53
Totals (615) =	62	68	63	121	93	121	87

The Tension Scores were :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
2	21	3	9	4	20	5	16
6	16	7	20	8	45	9	17
10	36	11	28	12	36	13	16
14	27	Total = 307					

TABLE 65. Male / Female Scale.

Male / Female Scale	-ve, very	-ve, fairly	-ve, slightly	Equally descrip.	+ve, slightly	+ve, fairly	+ve, well
12 Male vs Female	156	63	38	41	4	2	3

This was the score of 307 people and is not included normally as very clearly the vast majority went with their normal sex. There were 9 people however that did not, this does not necessarily mean that 2.9% were homosexual but it is a good indication.

The Beck Depression Inventory

The BDI was used as it was one of the most respected instruments in the field of psychological profiling. A total of 329 people completed the BDI in full and the results are shown below.

TABLE 66. The Beck Depression Inventory.

Beck Depression Inventory	Not at all condition	Mild condition	Moderate condition	Severe condition
1 Sadness	174	125	28	7
2 Discouraged	180	107	38	8
3 Failure	225	70	31	7
4 Satisfaction	97	185	38	13
5 Guilty	265	44	17	7
6 Punished	263	39	2	29
7 Disappointed	193	125	4	11
8 Blame	168	132	26	5
9 Kill	272	55	3	1
10 Cry	230	77	2	22
11 Irritated	118	178	19	16
12 Lost Interest	226	85	16	4
13 Decisions	178	99	47	7
14 Look	151	86	85	10
15 Work	80	166	65	20
16 Sleep	112	131	51	36
17 Tired	56	187	71	17
18 Appetite	227	82	14	8
19 Weight	264	33	19	15
21 Worried	169	117	40	4
22 Sex	165	75	30	59
Totals (6,963) =	3813	2198	646	306

No 20 was related to the answer to Question 19 about weight, ie were you eating less? 92 said Yes (29.5%) whereas 220 said No (70.5%). The final total score was :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
0	13	1	9	2	7	3	14
4	13	5	13	6	13	7	16
8	16	9	22	10	6	11	20
12	15	13	14	14	9	15	14
16	16	17	14	18	7	19	10
20	5	21	9	22	7	23	2
24	9	25	3	26	1	27	1
28	5	29	1	30	5	32	5
33	1	34	1	35	1	36	3
37	1	39	1	40	2	41	3
42	1	57	1	Total = 329			

Self Esteem Scores

Between 325 and 330 people answered this questionnaire and were scored thus :-

TABLE 67. Self-Esteem Scores.

Self Esteem Scores	Strongly agree	Agree	Disagree	Strongly disagree
1 Satisfied	35	113	151	27
2 No good	26	102	121	77
3 Qualities	4	28	214	81
4 Do things	55	112	115	45
5 Proud	16	67	160	84
6 Useless	49	134	98	49
7 Person worth	14	56	187	69
8 Respect	25	100	142	59
9 Failure	15	54	147	111
10 "+" Value	17	70	185	54
Total (3,268) =	256	836	1520	656

The 325 Self Esteem Totals were drawn between 10 minimum and 40 maximum :-

Value	Frequency	Value	Frequency	Value	Frequency	Value	Frequency
10	1	13	3	14	1	16	5
17	3	18	6	19	6	20	8
21	12	22	8	23	16	24	12
25	14	26	19	27	37	28	22
29	30	30	23	31	23	32	8
33	19	34	9	35	12	36	6
37	9	38	4	39	4	40	5

Value	Positive Self Esteem	Negative Self Esteem
5	2	2
6	3	4
7	2	5
8	8	11
9	6	6
10	13	25
11	21	20
12	26	34
13	48	32
14	53	47
15	65	37
16	32	27
17	17	23
18	16	12
19	6	21
20	8 = 326	19 = 325

The Impact of Dystonia

TABLE 68. The Impact of Dystonia.

Impact of Dystonia	Strongly disagree	Mildly disagree	Not agree or disagree	Mildly agree	Strongly agree
1 Hard Time	45	32	53	96	95
2 Useless	67	43	27	93	90
3 Miss the things	62	43	33	76	107
4 More dependant	85	43	28	83	81
5 Uncomfortable	113	38	62	77	31
6 Self-sufficient	106	51	48	50	65
7 Inadequate	85	73	31	72	62
Totals (2,246) =	563	323	282	547	531

The 320 to 323 Impact of Dystonia people who scored are shown above :-

Impact of Dystonia :	No of people	%
7 Minimum Impact	12	3.8%
8	2	0.6%
9	7	2.2%
10	14	4.4%
11	11	3.4%
12	8	2.5%
12	4	1.3%
14 25% Impact	7	2.2%
15	10	3.1%
16	16	5.0%
17	14	4.4%
18	15	4.7%
19	11	3.4%
20	14	4.4%
21 50% Impact	13	4.1%
22	8	2.5%
23	13	4.1%
24	14	4.4%
25	15	4.7%
26	10	3.1%
27	16	5.0%
28 75% Impact	11	3.4%
29	10	3.1%
30	18	5.6%
31	13	4.1%
32	14	4.4%
33	6	1.9%
34	7	2.2%
35 Maximum Impact	6	1.9%
	---	---
Totals =	319	100%

The Acceptance Stages

The acceptance stages were answered by 318 people as follows :-

23 people are still in shock that they have contracted dystonia
25 people have moved to anger that they have contracted dystonia
12 people have moved to despair that they have contracted dystonia
258 people have moved to acceptance that they have dystonia !!

These are important stages, which regardless of how well anybody accepts that they have dystonia, they have to move through. Unfortunately a number of the 60 people (18.9%) have not just been diagnosed but have been in either shock, anger or despair for several years. This will need to be discussed later on.

The Primary Carer's Questionnaire

221 people answered this questionnaire, although 339 people responded, but 84 people (24.8%) had no primary carer, ie they lived alone and without any support of any description.

A further 25 people declined to answer any questions and another 9 failed to complete the questionnaire but of the remainder, 123 (36.3%) was the husband of the patient, 54 (15.9%) was the wife, 14 (4.1%) was the mother, 11 (3.2%) the daughter, 8 (2.4%) the father, 7 (2.1%) the boy or girl friend, 5 (1.5%) the son, 3 was the brother, 3 where the carer was not related to the person being cared for, 1 was the sister and 1 was the aunt of the person being cared for, ie 220 in total.

TABLE 69. Primary Carer's Questionnaire.

Question ?	Strongly agree (%)	Agree (%)	Disagree (%)	Strongly disagree (%)
Sleep	22 (10.0)	50 (22.6)	73 (33.0)	76 (34.4)
Inconvenience	20 (9.1)	69 (31.2)	76 (34.4)	56 (25.3)
Physical strain	16 (7.2)	61 (27.6)	80 (36.2)	64 (29.0)
Confining	20 (9.1)	64 (29.0)	77 (34.8)	60 (27.1)
Adjust at home	25 (11.3)	81 (36.6)	64 (29.0)	51 (23.1)
Change plans	25 (11.3)	56 (25.3)	85 (38.5)	55 (24.9)
Other demands	23 (10.4)	87 (39.4)	71 (32.1)	40 (18.1)
Emotional	27 (12.2)	88 (39.8)	66 (29.9)	40 (18.1)
Behavioural	22 (9.9)	93 (41.9)	61 (27.5)	46 (20.7)
Adjust at work	12 (5.5)	43 (19.5)	104 (47.3)	61 (27.7)
Financial strain	17 (7.7)	36 (16.4)	101 (45.9)	66 (30.0)
Overwhelmed	4 (1.8)	27 (12.3)	105 (47.7)	84 (38.2)
Distressing	48 (21.8)	69 (31.4)	60 (27.3)	43 (19.5)
Totals =	281 (9.8%)	824 (28.7%)	1023 (35.7%)	742 (25.8%)

The CSI total scores show the following :-

Score	No	Score	No	Score	No
13	1	15	1	16	1
17	2	18	1	19	4
20	2	21	2	23	2
24	2	25	6	26	6
27	5	28	10	29	10
30	5	31	7	32	9
33	12	34	8	35	11
36	12	37	12	38	4
39	20	40	4	41	5
42	2	43	7	44	3
45	3	46	8	47	2
48	4	49	2	50	4
51	1	52	20	Total = 220.	

Carer's Personal Statements

The following 271 statements were made by 110 primary carer's at the end of their questionnaire as a confidential message and not within the sight or knowledge of the patient's themselves. The entire questionnaire was administered confidentially.

TABLE 70. Personal Statements.

The message is :	**No**	**%**
He / She needs my support	45	16.6
This disease has changed my life	38	14.0
This disease has not changed my life	37	13.7
It has been very stressful	31	11.4
We are now limited socially	24	8.9
It has affected my emotions	20	7.4
I feel useless and helpless	15	5.5
It has improved with treatment	15	5.5
He / She has been very positive	14	5.2
The patient has changed	10	3.7
More publicity is required	7	2.6
I have had to stop work	5	1.8
The general public is ignorant of this disease	5	1.8
More research is needed	3	1.1
Doctors are ignorant of this disease	2	0.8
Totals =	271	100%

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 23

Environmental Profiles

This section was added in 1996 to see what environmental factors have what effect on different peoples' dystonia. This chapter is presented as written in the questionnaire and is divided into a number of different sections.

Extremities

The question " *Which hand do you write with ?* " is usually sufficient to determine the answer required, however as anyone who has worked with people with dystonia will know, this is not the correct question. Therefore the question was asked " *Which hand do you currently write with ?* " and " *Have you always used this particular hand ?* ". A similar question was also asked: "*Which foot would you usually use to kick a ball ?*"

The answers were quite interesting, in that out of a total of 330 people, 86.1% use their right hand and only 13.3% use their left (there was one person who could not write and another who was totally ambidextrous). However of this number 27 people, or 8.3%, used to use the other hand previously and had learnt to use this other hand to avoid their particular form of dystonia. This did not apply just to people with Writers Cramp only, but to many other forms of dystonia affecting their writing.

The question regarding which foot was used to kick a ball showed that 87.8% used their right and 10.2% used their left foot. However here there were 6 people who were also ambidextrous.

There was a section in one of the previous questionnaires regarding which different parts of the body had been affected and when. The answers to these questions were :-

TABLE 71. Which arm or hand is affected

Right arm (excluding hand)	69 people affected
Right hand only	76 people affected
Left arm (excluding hand)	61 people affected
Left hand only	58 people affected
Writing with R. hand	78 people affected
Writing with L. hand	7 people affected

However when asked the supplementary question *" Does the dystonia affect your hand only when writing ? "*, 17 people (20.2%) said Yes whereas 67 (78.8%) stated No, it was not just when writing.

The Thyroid Gland

A number of people had mentioned during the earlier part of the research programme that they had had some problems with their thyroids. Therefore it was decided that a specific question about this particular gland should be asked. The answers were quite interesting when the question " *please tell us about any illnesses you have had* " was asked and then went on to list 112 different specific diseases and illnesses which at least 442 of the patients had had previously - some still currently. However, when it came to the question of thyroids, a total of 52 out of 442 (11.8%) indicated they had had previous problems in this area. Therefore when the question about the thyroid was specifically asked, a total of 52 out of a grand total of 318 (16.4%) answered Yes and specifically described their specific problems.

TABLE 72. Self-reported Thyroid Gland activity

27 had an over-active thyroid, equivalent to 8.5%,	) Thyroid gland activity in the
10 had an under-active thyroid, equated to 3.1%,	) general population is estimated
7 had had their thyroids removed,	) at 4.7 per 1,000 per annum
6 had enlarged or swollen glands,	) for hypothyroidism
1 had had a nodule on their thyroid and	) and 0.8 per 1,000 per annum
1 had had a thyroglossal cyst.	) for hyperthyroidism.

Life patterns

The next question asked if they felt that their dystonia had been affected or caused by any particular change in their life pattern. Of the 303 people who answered, 48 (15.8%) said Yes. The largest number related this change to a recent bereavement; but apart from this single cause relating to 18 people (37.5%) the rest related it in small numbers, eg., divorce = 4, family stress = 4, neuro-surgery = 3, Road Traffic Accident = 3, working stress = 3 and stroke = 2. The other 9 causes were written up by one person each. These changes have taken place over the past 50 years from 1946 to 1995, nevertheless the vast majority (75%) said it had occurred in the past 10 years. A similar question had been asked previously which related to any particular trigger which people thought might have started off their dystonia. 556 responded and over 37% stated there was nothing, the rest had responded as follows :-

TABLE 73. What triggered your dystonia ?

Trauma or stress	78	14.0%
Accident	65	11.7%
Alcohol / Drug Induction	41	7.4%
Medical Operation	28	5.0%
Bereavement	22	4.0%
Action Induced / Task Specific	13	2.3%
Childhood Disease	8	1.4%
Pregnancy	5	0.9%
Industrial Injury	5	0.9%
Stroke	4	0.7%
Repetitive Strain Injury	4	0.7%
Heart Attack / Flu Inj / Cancer Radiotherapy	1 each	0.6%

Smoking

When asked " *Do you smoke regularly ?* " only 72 people (23.5%) replied in the affirmative, the remainder (234) replied in the negative, however when asked "*Have you ever smoked ?* ", a total of 165 (54.5%) replied Yes. The point here was not to determine how many people smoked but if they were affected by other people's smoke. In answer to this question, there were 119 people (39.1%) who said Yes, they were.

Gas Appliances

However, when the same question was applied to Gas Appliances, the affirmation was tremendously high, ie 260 people (85.2%) stated Yes, they were affected. This was really quite a significant figure and should be discussed in greater detail later.

External Influences

1. Sound :

25.5% of the 306 people questioned answered that they were affected by certain sounds. These were loud noise or music (46.2%), screeching or shrill noise (28.2%) or a sudden noise (10.3%). There were other noises affecting people but to a much lesser degree, eg., buzzing (7.7%) and long banging (5.1%).

2. Light :

45.2% of those 305 people who answered this question, had a positive response to bright light, or in particular, to sunlight. These were mostly people with Blepharospasm, but not exclusively, and those who were photo-sensitive probably had a very slight form of Blepharospasm as well but it was not significant and therefore remained undiagnosed.

TABLE 74. How light affected 138 people

My eyes clamp shut (all Blepharospasm)	35	25.4%
It causes headaches or migraine	30	21.7%
I am photosensitive	30	21.7%
It causes blinking (all Blepharospasm)	11	8.0%
It makes spasms worse (all Blepharospasm)	11	8.0%
I get blurred vision (all Blepharospasm)	7	5.1%
I get dizziness (all Blepharospasm)	7	5.1%
An allergic reaction occurs	3	2.2%
My eyes water	3	2.2%
Warmth helps my spasms	1	0.7%

3. Drinking Water :

89.8% of the 305 people questioned drank the normal mains water, however there were some exceptions. For example, 5.2% drank just bottled water, 3.3% drank filtered water, 1.0% drank well water and just 2 people (0.7%) drank spring water.

However when asked specifically what type of water their main drinking water was; 101 people of the 302 questioned (33.4%) did not know. Of the remainder, 27.2% drank fluorinated water, 22.2% drank soft water and 16.2% drank hard water. Only 1.0%, ie 3 people, drank specially softened water.

Spasm Tests

The following shows the results of the set of tests conducted in this part of the research designed to indicate which of the following " *make your spasms worse* ".

TABLE 75. Spasm Test Results.

Type of involvement	No	Better	Unchanged	Worse
Stress	274	0.4%	15.0%	84.7%
Relaxation	246	61.8%	35.8%	2.4%
Fatigue	255	1.2%	23.1%	75.7%
Emotion	252	1.2%	29.8%	69.0%
Distraction	229	9.2%	69.4%	21.4%
Self-consciousness	251	0.4%	31.1%	68.5%
Social situations	228	1.8%	36.4%	61.8%
Heat	233	19.7%	65.2%	15.0%
Cold	247	1.6%	58.3%	40.1%
Walking	242	3.3%	49.6%	47.1%
Running	216	1.9%	42.1%	56.0%
Carrying objects	233	0.4%	47.2%	52.4%
Writing	246	1.2%	51.6%	47.2%
Sleep	239	41.8%	47.7%	10.5%
Lying on my back	230	20.0%	63.5%	16.5%
Lying on my side	235	19.6%	63.4%	17.0%
Pre-menstrual cycle	96	1.0%	68.8%	30.2%
During menstrual cycle	94	2.1%	73.4%	24.5%
Between menstruation	92	5.4%	89.1%	5.4%
On wakening	231	26.0%	50.6%	23.4%
During the morning	235	14.0%	67.7%	18.3%
During the afternoon	231	5.6%	69.7%	24.7%
During the evening	248	4.4%	48.8%	46.8%

As can be seen, there are definite patterns arising from these series of questions. Firstly there is a tremendous upsurge in '*stress*' levels making people feel worse, at 84.7% it is the highest level of any above and should be taken specific notice of.

The second highest level is immediately below on the opposite side, ie '*relaxation*', makes people feel so much better by 61.8%. It should also be positively noted that '*emotion*', '*fatigue*', '*self-consciousness* ' and '*social situations* ' all have a very negative effect on people. '*Heat*' has a more positive effect than '*cold*', as does '*running*' and '*carrying objects* ' in general, although the more severe the disability the greater the difficulty here. '*Sleep*' generally has some good results, although how or where one rests has little impact. The '*pre-menstrual cycle* ' and '*during menstruation*' also has a slightly worse effect on about a third of the 90 odd people who responded. Finally, one can definitely see how the length of the day has on most people with dystonia. One can see a definite pattern of worsening as the evening progresses from 18.3% in the morning, to 24.7% by the afternoon but up to 46.8% by the evening. This will be discussed at greater length later.

Exposure to Chemicals

57 people out of 301 (18.9%) had been previously exposed to various chemicals. 13 due to crop spraying, 24 due to chemical pollution, 7 through an industrial accident, 2 from a chemical fire and 36 due to a number of different reasons. 5 had worked in a chemical factory and 3 had been exposed to paint. Of the rest, the reasons were numerous and varied and amounted to 8 reasons for 2 people and 12 different reasons for individuals. Although they varied from exposure to chemical weapons or chlorine to working in a science laboratory or a steel mills, they were too numerous and varied to be of any significance.

Allergy Problems

The whole problem of different allergies was next explored and some interesting answers were received, with 201 (66.6%) answering positively.

TABLE 76. Different Allergies.

Symptoms	Responded	Yes	%
Arthritis	302	79	26.2
Persistent Fatigue	302	71	23.5
Wind / Bloating	302	69	22.8
Migraines	302	61	20.2
Puffy Ankles / Hands / Face	302	59	19.5
Mouth Ulcers	302	45	14.9
Diarrhoea	302	40	13.2
Asthma	302	39	12.9
Eczema	302	36	11.9
Hay Fever	302	36	11.9
Stomach Ulcers	302	26	8.6
Rhinitis	302	22	7.3
Allergy to others not above	302	18	6.0
Hyperactivity	302	11	3.6
Uticaria / Hives	302	8	2.6
Blood relative - Yes allergic	289	150	51.9

Apart from the last figure, where over 50% of our respondents had a blood relative who was allergic to something, there was not a tremendous response to any of the different substances above. The most prevalent was '*Arthritis*' with over a quarter of people responding, followed quite closely by '*Persistent Fatigue* '. These different points will be discussed later.

Food and Drink

49 people out of 299 (16.4%) responded positively as to whether they were affected by what they ate or drank. But even more, 76 out of 217 (35.0%) said " *Yes, our spasms are made worse by what we eat or drink* ", as follows :-

TABLE 77. Food and Drink affected.

Food or Drink	Not at all affected	Mildly affected	Moderately affected	Severely affected
Pork	207	6	0	4
Beef	202	5	3	7
Red meat	203	5	2	7
White meat	214	1	1	1
Fish or shellfish	207	**3**	**3**	**4**
Wheat	212	1	2	2
Rice	215	0	1	1
Corn	215	1	0	1
Oats	214	0	1	2
Other Cereals	211	**0**	**2**	**4**
Milk	206	5	4	2
Egg	207	3	3	4
Cheese	194	8	8	7
Yoghurt	208	2	5	2
Other Dairy	214	**1**	**0**	**2**
Coffee	191	9	12	5
Tea	209	5	1	2
Other Liquids	201	**0**	**6**	**10**
Citrus Fruit	203	6	3	5
Potato	214	1	1	1
Other Fruit, Veg	211	**1**	**4**	**1**
Yeast	210	2	3	2
Chocolate	182	12	14	9
Sugar	200	11	5	1
Other Foods	214	**1**	**1**	**1**

Of those **highlighted**, further explanation is required. Of the 10 fish or shellfish, there was 1 person who could not eat fish, 6 who could not eat shellfish and 3 who could not eat either.

Of the 6 people who could not eat other cereals, 2 people could not eat Brazil Nuts, 2 who could not eat Pastry, 1 who could not eat too much fat and another who could not eat 3 different types of wheat. In the Other Dairy section, there were 2 who could not eat fresh cream and 1 who could not eat cottage cheese.

In the Other Liquids section, there were 5 who could be drink any form of Alcohol, plus another 4 who could not drink Wine and 1 other who could not drink Gin. There were a further 2 who could not drink Orange Juice and then single individuals who had difficulty with Low C Chocolate and Vinegar, Tap Water, Soft drinks in cans and fruit drinks and vinegar also. Finally, there were 6 people who had difficulty with other fruits and vegetables. They were all individually allergic to strawberries, carrots and tomatoes, parsnip, banana, oranges and finally some vegetables.

24 people out of 295 (8.1%) were on an exclusion diet of one form or another and 48 out of 292 (16.4%) had a particular craving for certain foods or drinks. The highest number (19) craved chocolate with the next (14) craving alcohol. Of the remainder, 6 craved tea, 5 craved tea and the rest individually craved such foods as Ribena, cream, bananas, crisps, Lucozade, hot spicy foods, milk pudding or anything sweet.

The number of cups of tea or coffee consumed in a day was asked and the answers proved quite interesting. The chart below shows the number consumed per day by individuals. Interestingly enough, 97 people (10.4%) said they didn't drink coffee but only 8 (2.7%) said they drank only decaffeinated because undecaffinated made their spasms worse. Only 45 people (15.0%) said they did not drink tea.

TABLE 78. Number of cups of tea and coffee drunk per day.

No of cups drank per day	Tea	Coffee
1	10	62
2	27	42
3	39	37
4	46	22
5	32	15
6	46	12
7	18	4
8	16	5
9	4	0
10	10	5
12 - 16	3	0
20	5	0

Therefore the average number of cups of tea drunk per day were 5.2 and coffee was 3.0. But it was interesting the number of abstainers, 15% for tea and 32.2% for coffee. Some people had definitely mentioned that in particular coffee had been removed from their diets and this had seemed to have a beneficial effect on their dystonia. This is an interesting point and perhaps more should be made of this very simple recommendation to see if the degree of spasms lessened in the dystonic population.

Wine was also mentioned in particular. 300 people answered this question, 38.0% drank wine, 57.3% did not drink wine but a further 4.7% did not drink alcohol at all. The 114 who did drink wine were then asked a supplementary question as to how they felt it helped their spasms. 51 people (44.7%) felt it relaxed their spasms and a further 33 people (28.9%) thought it mildly affected them. 9 people thought alcohol severely affected them, 5 felt they had more confidence, 4 said it gave them a slight headache, 3 were alcoholic and another 3 said it made them slightly sick or nauseated. Finally 2 people said it helped them go to sleep, 2 more said alcohol reacted with their drugs and one person said it loosened their bowels and another said it dried up their mouth.

Finally Cola was asked about. 18 people said Yes, they did think that Cola affected them, 91.0% said they were not affected and 3.0% (9 people) said they did not drink Cola at all. Of the 18 people who had said Yes, 4 said they got wind or bloating, another 4 said they either got worse spasms or they got spasms specifically in the stomach, 3 got an upset stomach and 2 felt edgy or nervous. Of the remaining 5 people they individually got ; slight hiccups, choked when drinking, a sensitive tongue, it dried up their mouth or it aided their digestion.

Environmental Factors

A total of 304 people answered these questions, 99 of which said No to everyone, which left 205 who said Yes to some - these are laid as follows in order of priority :-

TABLE 79. Types of material and degree of affectedness.

Type of Material	Not at all affected	Mildly affected	Moderately affected	Severely affected
Cigarette Smoke	177	50	39	38
New Paint	185	65	30	24
Exhaust Fumes	199	50	34	21
Pipe	202	41	29	32
Cigars	202	44	32	26
Cigarettes	205	42	29	28
Dust	215	35	35	19
Paint Stripper	216	42	24	22
Petrol Fumes	217	41	31	15
Diesel Fumes	222	36	29	17
Varnish	225	41	22	16
Ammonia	227	38	20	19
Glues	229	40	14	21
Turpentine	233	41	18	12
Hairsprays	234	35	28	7
Damp humid days	238	22	36	8
House dust	239	25	20	20
Air-freshener	241	28	21	14
Insecticides	242	33	17	12
Chlorine	242	30	14	18
Burning Rubber	242	30	19	13

Fixative	243	33	16	12
Swimming Pools	243	32	14	15
Adhesive	246	35	15	8
Bleaches	248	32	13	11
Dry cleaning fluid	248	27	13	16
Deodorants	249	27	22	6
Wasp stings	250	21	20	13
Pesticides	250	26	18	10
Perfume	250	28	15	11
Methylated Spirits	253	29	10	12
Bee stings	255	19	17	13
Calor / Butane	257	25	16	6
Lighter Fuel	257	26	11	9
Polishes	258	23	18	5
Carbon Tetrachlo.	258	24	8	14
Surgical Spirit	258	25	12	9
Weedkillers	259	21	11	13
Paraffin	259	24	9	12
Deodorants	259	24	11	10
Detergents	259	28	6	11
Mothballs	260	18	16	10
Other pollens	260	22	16	6
Burning Tar	260	24	11	9
Disinfectants	260	27	10	7
Oils	260	28	8	8
White spirit	260	29	7	8
Rotting vegetation	262	21	15	6
New mown grass	262	22	13	7
Oil	263	21	14	6
Plastics	263	22	10	9
Bitumen Tar	264	16	14	10
Mites	266	17	12	9
Hay	267	16	15	6
Sealer	270	19	8	7
Silage	270	20	6	8
Old houses	271	17	10	6
Pollutants in water	271	19	5	9
Asphalt	273	12	10	9
Fungus	273	15	13	3
Degreasers	273	17	8	6
Moulds	274	14	12	4
Make-up	274	18	7	5
Furniture Polish	276	14	9	5
Pesticide in food	276	16	5	7
New fabric	276	21	2	5
Food Additives	277	13	5	9

Feathers	277	13	9	5
Powder	278	14	7	5
Cats	279	8	9	8
Long grass	279	9	12	4
Newsprint	280	11	8	5
Silver Polish	280	13	7	4
Liquid Polishers	281	12	5	6
New carpets	281	13	6	4
Manure	282	13	5	4
Tartrazine	283	9	2	10
Coated paper	283	12	6	3
Heating Oils	283	14	2	5
Shampoo	283	14	4	3
Phenolic Resin cans	289	11	1	3
Formaldehyde	280	11	4	9
Carpet Shampoo	284	14	3	3
Dogs	285	8	8	3
Rodents	286	3	8	7
Creams	286	11	2	5
Other Smells 2	287	1	2	14
Waxes on fruit etc	287	7	5	5
After-shave	287	10	3	4
Birds	288	5	7	4
Coal Fire	288	8	6	2
Charcoal	288	11	2	3
Houseplants	288	12	2	2
Car Upholstery	289	8	3	4
Pine Products	290	8	4	2
Odour of pines	291	7	3	3
Trees	292	7	5	0
Cushion	293	8	2	1
Other Smells 1	294	1	2	7
Carpet Backing	295	5	3	1
Furniture Upholst.	296	7	1	0
Linoleum	296	7	1	0
Padding	297	5	1	1
Horses	297	5	2	0
Other Smells 3	298	0	4	2
Floor Tiles	299	3	2	0

The ten "Other Smells 1" is 2 burning smells, 2 fishy smells and the other 6 are rubbish bins, carrots, wood smoke, creosote, onions and rubefacient cream. The 17 "Other Smells 2 " is 2 are Aspirin and 2 are Garden Soil with the other 13 are bath cleaner, lager, body odour, food colouring, penicillin, maxolon, essential oils, reaction to a drug, eels, scotchguard tape, lanolin, algipan spray and straw dust. The 6 " Other Smells 3 " is individually unwashed bodies, tomatoes, flowers, aspirin; rats and hair dye.

The Analysis of Conduct of Research

An analysis of the conduct of the research was planned at the end of questionnaire 60 asking for criticism of the study and at the end of questionnaire 90 asking for comments on the research study. Criticism of the study was asked for from the respondents at the end of questionnaire 60 as a way of monitoring the way the research was going at the end of the 2nd year. Herewith are the results :-

TABLE 80. Responses from the subjects on the conduct of this research project.

A precise of the words used :	**No of respondents**	**%**
Mainly positive responses :		
Completing the research has helped me	76	
It has been much appreciated	64	
Done with great understanding	33	
It has been conducted very professionally	32	
Done with great consideration	23	
Done with great courtesy and respect	17	
It has helped expressing my feelings	9 = 245	55.3%
No comment nor criticism	82	18.5%
Mainly negative responses :		
Only some questions were relevant	28	
Difficult to complete without help	19	
More research is needed	16	
Some questions were duplicated	11	
It found quite personal and disquieting	8	
It has been too much trouble at times	8	
I have needed more space to complete	7	
Need to inform more GP's	6	
I prefer writing than just answering questions	5	
I hope this research helps young people	4	
Now in remission	3	
Please use simpler words in the future	1 = 116	26.2%
Totals =	443	100%

One of the main things, apart from general thanks for doing the research, has been the quite positive responses and thanks by over 50% of the participants, such as "*completing the research has helped me*" and "*it has helped expressing my feelings*". Also the personal comments are much appreciated, particularly the comments such as "*much appreciated*", "*done with great understanding*", "*conducted very professionally*", "*done with great consideration and with great courtesy and respect*". On the other hand, even the criticisms were positive and comments such as "*more research is needed*", "*need to inform more GP's*" and the comment that it was hoped that "*this research helps young people*" were all quite constructive.

Even the more personal criticisms were quite useful and it helped to change the systems in the future whilst at the same time keeping the questions exactly the same to ensure continuity.

The second question regarding how the patients felt about certain things outside of themselves yet definitely affecting their own dystonia has resulted in the following comments being made at the end of questionnaire 90.

TABLE 81. Responses reacting to outside stimuli.

A precise of the words used :	**No of respondents**	**%**
Better		
Better - when there is no stress	45	
Better - with sun and warmth	22	
Better - with positive thoughts	17	
Better - when I am busy	6	
Better - after drinking alcohol	4	
Better - helped by surgery / counselling	3	= 28.0%
Neutral		
Definite pattern - but why ?	23	
No comment either way	11	
No definite pattern	8	
Unaffected by anything	7	
Currently in remission	2	= 14.7%
Worse		
Worse - with activity	46	
Worse - when tired	34	
Worse - missing injections	23	
Worse - self conscious	19	
Worse - with worry	12	
Worse - over time	11	
Worse - link with drugs, etc	9	
Worse - in climatic changes	9	
Worse - on windy days	7	
Worse - when reading or watching TV	7	
Worse - with depression and anxiety	5	
Worse - in bright sunlight	4	
Worse - suspect hormonal link	4	
Worse - before my periods	4	
Worse - on my affected side	4	= 57.2%
Totals =	346	100%

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 24

The Dystonia Society

The Dystonia Society was formed in 1983 in London by the patients of the late Professor Marsden. It is with deep gratitude that the author thanks the Society in the North East for encouraging him in this research and for allowing him access to their membership as well as allowing him access to a number of controls used in the Cost Utility Analysis work from the membership throughout the rest of the UK. Unfortunately there was a disagreement with the National Society in 1999 and currently there is now no Dystonia Society organisation in the North East of England, it being now being called ADDER (Action for Dystonia, Diagnosis, Epidemiology and Research). However in all the work that has been done in the North East, the people in the Dystonia Society in the North East has been of primary importance for which all those members and people in position of power who have helped are herewith heartily thanked.

The research into the Dystonia Society was really to see how many people were members and how those people with dystonia viewed organisations that purported to represent them, etc. The questions were asked and the answers received as follows.

Contact

Firstly one needed to establish what sort of contact there was between people with dystonia. Therefore 452 people were asked how often they talked to others with dystonia, the answer was particularly interesting, as over half (51.1%) had never talked to another person with a similar disorder. Of the remainder, 81 had spoken occasionally, 56 rarely, 51 once only, 17 frequently, 10 had only ever spoken to one person but 6 people had spoken often. Therefore these 221 people were then asked where or how they spoke to other people with dystonia. The vast majority, ie 90 people (40.7%), spoke at the Bot. Tox. clinic, 60 (27.1%) attended TDS meetings, 32 (14.5%) on the phone, 22 at home to other family members, 14 in the hospital and 3 at work.

Counselling

The question was asked starting on 6th May 1993, *" Do you think counselling should be provided and when ? "*. The vast majority (77.2%) said Yes, only 11.4% said No and the rest were either "*Don't know*" or "*Not for me*". However 285 people answered as to when this should occur. The vast majority (57.2%) thought it should be offered when they were *first diagnosed*, 22.1% thought it should be available *if required* and the final 59 people (20.7%) thought it should be there *when it was required.* Finally the question was then asked "*Who should do this counselling ? "*

The replies were equally interesting in that 287 people made positive suggestions, the majority (26.5%) thought it should be someone who knew about dystonia, then came three equal suggestions, all at 19.5% each, someone in the NHS, someone from TDS and by a doctor. 21 people (9.1%) suggested it should be done by someone with it, 13 people suggested by a nurse and 3 people by a Social Worker.

Interestingly enough, the Dystonia Counselling Service was started in the North East in 1995 by the funding of a trained Counsellor by an anonymous sponsor and it was then funded by a National Lottery Charities Board award. The Dystonia Counsellor is a qualified Counselling Psychologist and his wife assists him by making his appointments at the Botulinum Toxin clinics and generally talking to and being confined in, by the patients attending. In the past 4 years, both have been a tremendous asset to the dystonia patients in the North East.

TDS Membership

As membership of TDS was not a pre-requisite in any way in order to qualify for entry into this research project, it was decided in the early days of the project that we could use the research to also enquire as to why people joined or did not join TDS. The first question raised was " *how had you heard of TDS ?* " The vast majority were evenly divided between through their own doctor at the hospital or clinic and through this research. This took up over 64% of those 485 who answered the question. Of the remainder, 41 people had read about TDS in a newspaper or magazine, 31 people had heard about it on the radio, as had another 31 through leaflets or advertisements. 24 people had heard about TDS through a relative or friend and the rest (14) in a variety of different ways. Interestingly enough, the specific awareness raising campaign on the television run by the group had also informed 9 people in 1994 and 22 other people in 1997, which meant they were then correctly diagnosed and treated.

The membership of TDS is also very interesting to study to compare its success to the amount of advertising and awareness raising carried out during this 6 year period. It was a definite policy of the researcher not to deliberately recruit patients into TDS during this period but if people asked to join then he would give out the literature, etc. The following chart shows the extent of TDS membership in the entire survey population.

TABLE 82. Membership of TDS

No, not a member	630	
Joined since the study started	123	)
Yes, a member before the survey	101	)
Re-joined or joined through RC	40	) Total membership : 296 (31.6%)
No longer a member, dropped out	21	)
Deceased member	11	)
A member of MSS, PDS, etc	11	

Total =	937	

Questions were asked as to why people joined TDS. Only 123 were able to answer this question at the time, but of them, 37 joined to find out more about it, 35 to get specific information, 22 joined to meet and talk to others, 11 people could not answer why they had joined, 6 people each said they did it to get answers and to give support, 4 to read the literature and 2 to receive news. They felt the most useful aspect of being a member was to be in contact with one another (27.4%), to receive the newsletter (21.0%) and to get information (20.2%). 10 people wanted news about research, 9 liked being part of the Self-Help Group and another 6 liked being able to get answers to their questions. The year in which these 296 people joined is also indicative of the effort the researchers have put into the research and making these people feel part of the team.

TABLE 83. When did people join TDS.

	No of Members	Specific Event
Joined in 1983	1	Formation of TDS in London
Joined in 1984	3	
Joined in 1985	2	
Joined in 1986	2	
Joined in 1987	6	
Joined in 1988	10	
Joined in 1989	8	
Joined in 1990	8	Formation of TDS in North East
Joined in 1991	1	
Joined in 1992	23	
Joined in 1993	51	Start of research in North East on 6.5.93
Joined in 1994	32	
Joined in 1995	36	
Joined in 1996	52	
Joined in 1997	25	
Joined in 1998	36	End of research in North East on 5.5.99

Totals =	296 members - not all current.	

All of the 937 people were placed in a group, either as a member of a particular self-help group or merely being on the mailing list, as follows .-

TABLE 84. No. of members of TDS.

Tyneside SHG -	formed in 1990	113	
Teesside SHG -	formed in 1993	88	
Cumbria SHG -	formed in 1995	41	
Darlington SHG -	formed in 1998	6	
Sunderland SHG -	formed in 1998	10	
Yorkshire SHG's		9	
Member of another SHG		20	
Member of TDS -	no contact requested	9	Total no. = 296 (31.6%)
Not a member, on mailing list only		556	
No contact requested		85	Total no. = 641 (68.4%)

In order to complete the research into TDS, it was necessary to ask a few more pertinent questions. The first related to what people got most out of being a member. The highest number who answered did not know (19.3%). Of the remaining 92 people the answers in descending order were ; talking to others (19), more knowledge (17), sharing the experience (14), reading the news (13), sharing a feeling of belonging (11), the social contact (11) and finding out about other peoples experience (7).

The Dystonia Society national newsletter was discussed and of the 110 people who answered, their comments showed that 39 people preferred the "*Letters to the Editor's*" section, followed very closely by "*all of it*" (36). The other people liked the medical articles (21), the latest research (10), news from other groups (3) and one person liked particularly the other contact addresses.

Finally a question was asked about what people felt the national aim of the society should be and what its local aim should be, if different. Of the 249 people who answered the first question, the highest number thought it should be to raise awareness of dystonia (76), followed by finding a cure (50), giving information (43), funding research (42) and contact with others (25). As the research got going, it was noticeable that gradually a few different answers were creeping in, for example counselling, welfare, raising funds, getting specific treatment and getting more Dystonia Nurse Practitioners were these latest ideas, but all too small to be significant.

The second question referred to the local aims of the local group. The answers were mostly reflective of the work currently going on within the group. For example, the greatest number (128) stated that contact with others was the most important local aim. This was followed by raising awareness (37), giving information (26), counselling and welfare (12), funding research (12) and raising funds (11). Getting treatment and getting more Dystonia Nurse Practitioners was a later and smaller consideration.

The Welfare Benefits Service

The Welfare Benefits Service was an additional service set up in mid-1998 by the local branch to look after the welfare arrangements of branch members. This was the start of the services which were restricted to branch members only, as all previous services of TDS (NE) were open to members or not alike.

The members were referred through a number of different people, 8 through the author, 6 through Peter Williams, the Regional Co-ordinator from March 1996 until July 1997, 2 through David Medd, the Dystonia Counsellor, and 1 each through John Whitaker; the Dystonia Nurse Practitioner, Maurice Hawthorne, the ENT Consultant Surgeon and Harry Crow, the new Regional Co-ordinator from 1st January 1999.

During the period between April 1998 when the service started and January 1999 when we closed the records the following availed themselves of the Welfare Benefits Co-ordinator's services.

TABLE 85. The Welfare Benefits Service Usage.

April 1998	2 members	
May	3 members	
June	1 member	
July	1 member	
August	1 member	
September	4 members	
October	2 members	
November	2 members	
December 1998	2 members	
January 1999	1 member	= 19 members in total

Although the details of the service are totally confidential, it is worth noting an outline of the results of the problems encountered :-

TABLE 86. Results of Welfare Benefit Advice

6 were still ongoing at the time of this report,
5 had had a positive result,
2 had been spoken to and dealt with on the phone,
2 had currently an unknown result (still pending).
1 had been referred to an adviser
1 had been seen at home but with no further result
1 had had a partial result
1 had been referred on to a different adviser.

However, although this service is breaking new ground in this field in the region, it is considered a very worth while service and one that should continue to benefit members through the area.

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 25

SPASMODIC TORTICOLLIS

A specific questionnaire related to distinct questions about Spasmodic Torticollis, or Cervical Dystonia to give its correct medical terminology (Jahanshahi, 1990). Because the largest number of people in this study have Spasmodic Torticollis, this was easy to administer and the results are taken directly below from this two page questionnaire, of which a total of 152 were completed as follows :-

TABLE 87. What is the position of your head ?

Chin turns to the side	79	52.0%
Ear tilts sidewards	38	25.0%
Head bends backwards	22	14.5%
Heads bends forward	13	8.6%
Total =	152	100%

TABLE 88. What is the direction to which your head moves ?

Right turn and tilt	34	22.4%
Left turn and tilt	27	17.8%
Left turn only	19	12.5%
Right and backwards	17	11.2%
Left and backwards	13	8.6%
Right and forwards	8	5.3%
Right tilt only	7	4.6%
Forwards only	6	3.9%
Left and forwards	6	3.9%
Backwards only	5	3.3%
Right turn only	5	3.3%
Left tilt only	5	3.3%
Totals =	152	100%

Do you have a method of holding your head called a 'Geste Antagoniste' ?

151 people answered, 107 people (70.9%) said Yes with 44 (29.1%) answering No.

Is it still effective ?

102 people answered, 78 people (76.7%) said Yes with 22 (21.6%) answering No and with 2 people saying it was not necessary now as there were currently in remission.

How many years has it remained effective ?

22 people answered, 7 said an unknown number of years, 7 more said 2 years, 4 said one year and then 1 each said 3, 4, 5 and 7 years. 7 years was the longest recorded time it lasted without change.

Describe in detail (in terms of hand used, area of head, neck or face touched), the gesture that you use(d) to keep your head straight. Perform this gesture in front of a mirror before answering and write your answers on a separate piece of paper : This question produced verbatim answers from 54 subjects which are reproduced exactly in Appendix F.

Which hand is used to touch your head ?

109 people answered this question, with 53 (48.6%) saying the *right hand* and 46 (42.2%) using *the left.* A further 10 people (9.2%) could use *either or both hands.*

Where do you place your hand ?

83 people (78.3%) place their hand *in front of their body*, 18 people (17.0%) at the *back of their body*, 5 people (4.7%) use *different places on different occasions.*

How do you place your hand ?

52 people (46.0%) *lightly touch*, 42 people (37.2%) *push hard*, 16 people (14.2%) *pull hard* and a further 3 people use *different techniques on different occasions.*

My head starts moving when :-

45 people (42.5%) answered *after*, 34 (32.1%) *before*, 25 (23.6%) *at the same time* and 2 people stated they moved *sometimes before and sometimes after.*

I feel that my head is :-

142 people answered with 126 people (88.7%) stating *being pulled*, 14 (9.9%) stating *being pushed* and the remaining 2 people said *both at times.*

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 26

The Outreach Nurse Practitioner Project (ONPP)

Based on the criteria set out in Chapter 13, a total of 126 were selected to participate in this study based on the random selection procedure. Each subject was placed in one of the groups before being approached by the Outreach Nurse Practitioner (ONP) to obtain their informed consent. The subject was not informed into which group he or she had been placed until after informed consent was obtained. This was done in order not to influence the potential decisions of the subjects in the selection process and to ensure that the results obtained were not subject to preference for one type of treatment or another.

The Pilot Scheme

As the ESD, at that time, had nearly 500 patients throughout the region, it was decided that a pilot scheme should be able to be carried out on subjects who were not registered at Hunters Moor Hospital in Newcastle, thus ensuring further independence and impartiality. A total of 10 subjects, 4 male and 6 female, were selected by the author as meeting the ONPP Admission Criteria, who were registered at the North Riding Infirmary in Middlesbrough.. These were interviewed over a three day period and represented 2 Hemi-facial Spasms, 2 Blepharospasms, 2 Writers Cramps and 4 Spasmodic Torticollis. The interviews were conducted by the Nurse Practitioner, with the author, the Evaluator / Monitor (M/E), making the initial introduction and remaining in attendance. All questionnaires from the ONP and the M/E, not previously administered in ESD, were tested and validated during the Pilot Scheme.

The M/E tested out the series of evaluation and monitoring questionnaires regarding the evaluation process and although the basic questions were initially quantitative in nature, it was soon realised that qualitative questions got the most accurate results, although this meant more difficulty in coding onto SPSS later, however adjustments were made accordingly. During the Pilot Scheme there were a number of complaints about injections being given behind screens at Hunters Moor, thus resulting in a lack of confidentiality during treatment. It was also realised that allergies were not recorded on any of the ONP or M/E questionnaires. Therefore questionnaire 81 was administered to all subjects in the ONPP by the author.

Finally, it soon became clear that sufficient time should be allowed during the interviews for counselling and emotional outbursts. Although the author had seen most of the subjects during the ESD demographic interviews, it was obvious that the presence of a Nurse Practitioner in the patient's home often released a pent-up stream of emotion. This manifest itself in many different ways but always in long answers to the qualitative open-ended questions and often in tears of relief at being listened to at last - consequently time allotted for interviews was extended during the initial stages.

The Interviews

A total of 126 people, who met the Inclusion Criteria, were approached by the ONP to participate in the study. At the time of the first interview by the M/E, 16 had declined in the Clinic Group and another 12 in the Home Group. Therefore a total of 98 people were available to be visited for the first interview.

However, over the period of the study, 4 people deceased, 2 from each group and a further 28 subjects (12 in the Home Group and 6 in the Clinic Group) dropped out of the project for differing reasons. The final numbers, as of 14th May 1997, were :-

TABLE 89 : Numbers participating in the research project

Status at end of study	No of patients	%
Home - completed	38	30.2
Home - dropped out	12	9.5
Home - declined	12	9.5
Home - deceased	2	1.6
Clinic - completed.	38	30.2
Clinic - dropped out	6	4.8
Clinic - declined	16	12.7
Clinic - deceased	2	1.6
Total	126	100%

The numbers of people completing in each group were equal, ie 38, as were the number of people who died, ie 2 in each group, and the numbers not completing plus those declining to participate, ie 24 in each group.

The following charts are of the Home and Clinic Groups showing those available for interview at the time of the start of the study. In practice, more people were seen than actually participated as a number asked for home interviews before finally declining.

The charts have been taken directly from their 'Access' files and have been made to fit the page and therefore the each year's date has been missed off. Please anticipate that the first dates will be early in 1996 , eg 1st appointment, and the last dates will be in mid 1997.

All 38 subjects who completed the study in the Home Group also completed all the required questionnaires and interviews. This represents a 100% record, with no exceptions, in the Home Group.

CHART No 90. The Home Group

No	HMH	Diag	ESD	Post	Status	App.	Q10	Q20	Q30	Q91	Q92	93/94	Final
1	65	ST	90	NE66	Yes	23.04	x2	Yes	Yes	10.04	13.08	29.04	29.04
2	167	ST	52	DH03	OOP	25.03	Yes	Yes	Yes	06.03	14.09	13.05	17.02
3	173	ST	122	DH08	OOP	25.03	Yes	Yes	Yes	06.03	Out	of the	Proj.
4	189	ST	24	NE34	Yes	29.04	x2	Yes	Yes	24.03	14.08	12.05	14.04
7	276	ST	65	NE05	Yes	27.03	x2	Yes	Yes	14.03	13.08	22.04	16.04
8	283	ST	68	CA20	Yes	18.04	x2	Yes	Yes	09.04	14.10	09.04	10.04
9	284	ST	55	NE33	OOP	19.02	Yes	Yes	Yes	17.02	13.08	Out -	Proj.
10	286	ST	61	NE38	Yes	29.04	x2	Yes	Yes	02.04	13.08	22.04	24.03
11	347	RT	140	NE15	Yes	27.03	x2	Yes	Yes	06.03	13.08	07.05	12.03
12	369	ST	76	CA28	Yes	18.04	x2	Yes	Yes	27.04	23.10	09.04	24.04
13	372	ST	87	DH01	Yes	24.06	x2	Yes	Yes	20.04	13.08	05.05	24.02
14	373	ST	82	CA14	Yes	24.04	x2	Yes	Yes	20.04	14.10	09.04	24.04
15	374	RT	79	NE21	Yes	15.07	x2	Yes	Yes	20.04	23.10	13.05	18.03
16	433	ST	146	DH08	Yes	25.03	x2	Yes	Yes	20.03	13.08	14.04	17.03
17	545	ST	186	NE38	Yes	29.04	x2	Yes	Yes	20.03	14.09	06.05	24.03
18	602	ST	306	CA02	Yes	20.05	x2	Yes	Yes	20.05	14.10	09.04	17.04
19	616	ST	311	NE34	OOP	01.04	Yes	Yes	Yes	Out	of	the	Proj.
21	673	ST	319	NE27	Yes	06.03	x2	Yes	Yes	23.02	13.08	01.05	16.04
23	808	ST	443	NE03	Yes	28.08	x2	Yes	Yes	26.02	14.10	13.05	23.04
24	861	ST	389	NE34	Yes	18.03	x2	Yes	Yes	26.02	14.09	12.05	24.03
26	909	ST	384	DH03	Yes	26.02	x2	Yes	Yes	23.02	13.08	13.05	14.04
27	955	ST	382	SR03	Yes	18.03	x2	Yes	Yes	23.02	13.08	08.05	17.02
28	1030	ST	395	SR05	Yes	25.03	x2	Yes	Yes	20.03	13.08	14.04	14.04
30	1109	ST	400	NE04	OOP	08.05	Yes	Yes	No	15.05	Out	of the	Proj.
31	1160	RT	445	DH02	Yes	15.04	x2	Yes	Yes	26.02	14.10	13.05	10.03
32	1329	ST	466	DH01	OOP	18.03	Yes	Yes	No	Out	of	the	Proj.
33	1350	ST	450	SR03	Yes	15.04	x2	Yes	Yes	13.02	13.08	08.05	24.02
34	1361	ST	433	NE25	Yes	21.02	x2	Yes	Yes	29.02	14.10	01.05	18.03
36	540	BL	185	NE25	Yes	28.05	x2	Yes	Yes	20.05	23.10	01.05	19.03
38	715	BL	442	CA02	OOP	06.06	No	Yes	No	16.05	Out	of the	Proj.
40	1067	BL	402	NE17	Yes	29.04	x2	Yes	Yes	26.02	13.08	13.05	28.04
42	1166	BL	452	CA06	Yes	04.07	x2	Yes	Yes	14.06	10.11	09.04	17.04
43	1199	BL	454	DH01	OOP	01.07	No	Yes	No	22.05	Out	of the	Proj.
44	1339	BL	474	DL05	OOP	20.05	No	Yes	No	Out	of	the	Proj.
45	51	HFS	62	NE36	Yes	05.02	x2	Yes	Yes	04.02	14.08	12.05	01.04
46	56	HFS	94	NE15	OOP	26.02	x2	Yes	Yes	23.02	Out	of the	Proj.
47	183	BL	19	NE12	Yes	15.05	x2	Yes	Yes	24.03	14.08	22.04	05.03
48	186	HFS	115	NE23	Dec.	-	Yes	Yes	Yes	Out	of	the	Proj.
50	421	HFS	187	DH06	OOP	-	Yes	Yes	No	Out	of	the	Proj.
51	507	HFS	144	NE24	OOP	-	Yes	Yes	No	Out	of	the	Proj.
52	598	HFS	197	DH07	Dec.	-	Yes	Yes	Yes	Out	of	the	Proj.
53	623	HFS	318	NE05	Yes	08.05	x2	Yes	Yes	06.03	23.10	07.05	10.04
54	677	HFS	36	NE35	Yes	11.04	x2	Yes	Yes	02.04	23.10	12.05	24.02
55	691	HFS	377	NE16	Yes	01.04	x2	Yes	Yes	30.05	14.10	15.04	17.03
56	790	HFS	444	NE07	OOP	-	No	No	No	Out	of	the	Proj.
57	913	BL	408	NE49	Yes	28.03	x2	Yes	Yes	28.03	14.10	02.05	20.03
58	1164	HFS	449	DH05	Yes	25.03	x2	Yes	Yes	14.03	13.08	05.05	24.02
60	1227	HFS	471	CA04	Yes	29.02	x2	Yes	Yes	23.02	13.08	09.04	24.04
61	1271	HFS	462	DH01	Yes	22.04	x2	Yes	Yes	26.02	13.08	06.05	10.03
62	470	WC	199	TS16	Yes	13.05	x2	Yes	Yes	23.02	13.08	04.04	24.03
63	277	GD	53	NE03	Yes	13.03	x2	Yes	Yes	24.03	14.08	07.05	26.02
64	652	GD	313	NE22	Yes	19.03	x2	Yes	Yes	23.02	13.08	02.05	05.03

CHART No 91. The Clinic Group

No	HMH	Diag	ESD	Post	Status	App.	Q10	Q20	Q30	Q91	Q92	93/94	Final
1	170	ST	117	NE15	Dec.	17.04	Yes	Yes	Yes	Out	of	the	Proj.
2	180	ST	16	CA16	Yes	10.04	x2	Yes	Yes	14.04	25.09	09.04	11.03
3	182	ST	20	NE37	Yes	14.04	x2	Yes	Yes	18.03	13.08	05.05	18.03
4	200	RT	121	NE08	Yes	17.04	x2	Yes	No	30.03	13.08	02.05	15.04
5	244	ST	26	SR06	Yes	19.04	x2	Yes	Yes	14.04	25.10	12.05	08.04
6	266	ST	49	DL14	Yes	14.02	x2	Yes	Yes	27.02	25.09	15.04	15.04
7	268	ST	51	SR02	Yes	27.03	x2	Yes	Yes	02.03	25.09	08.05	04.03
8	269	ST	54	NE37	Yes	23.07	x2	Yes	Yes	02.03	13.08	22.04	22.04
9	270	ST	67	NE33	Yes	17.06	x2	Yes	Yes	10.04	13.08	12.05	18.02
10	272	ST	64	NE09	Yes	17.04	x2	Yes	Yes	10.04	13.08	29.04	22.04
11	280	ST	56	NE15	Yes	17.03	x2	Yes	Yes	18.03	25.09	07.05	18.03
12	282	ST	57	CA02	Yes	24.06	x2	Yes	Yes	24.04	14.12	09.04	11.03
13	285	ST	60	NE10	Yes	03.04	x2	Yes	Yes	10.04	13.08	06.05	18.03
14	367	ST	83	NE15	Yes	03.07	x2	Yes	Yes	10.04	25.10	07.05	04.03
15	376	RT	85	NE24	Yes	08.05	x2	Yes	Yes	10.04	27.10	02.05	11.02
16	394	ST	86	NE23	Yes	17.04	x2	Yes	No	10.04	27.10	02.05	01.04
17	431	ST	145	NE13	Yes	17.04	x2	Yes	Yes	24.04	27.10	14.05	06.05
18	440	ST	142	NE23	Yes	24.04	x2	Yes	Yes	24.04	27.10	02.05	29.04
19	572	RT	190	NE61	Yes	24.04	x2	Yes	Yes	24.04	27.10	02.05	08.04
21	795	RT	329	SR05	Yes	29.03	Yes	Yes	No	27.03	27.10	14.05	18.03
22	816	AT	390	NE46	Yes	06.03	x2	Yes	Yes	08.03	29.09	02.05	25.02
23	818	RT	399	NE34	Yes	03.04	x2	Yes	No	27.02	20.09	12.05	15.04
24	884	ST	338	NE09	OOP	10.04	Yes	Yes	No	28.02	Out	of the	Proj.
27	1092	ST	422	DH01	OOP	27.03	No	Yes	No	14.02	Out	of the	Proj.
30	1103	RT	394	SR07	Yes	27.03	x2	Yes	Yes	13.03	20.09	08.05	08.04
31	1128	ST	469	NE32	Yes	10.04	x2	Yes	Yes	26.02	27.10	12.05	04.03
32	1266	ST	461	SR03	Yes	10.04	x2	Yes	Yes	24.04	27.10	08.05	25.03
33	1349	ST	489	NE34	Yes	06.03	x2	Yes	Yes	27.02	14.10	12.05	07.05
35	166	BL	98	NE25	OOP	06.03	Yes	Yes	Yes	Out	of	the	Proj.
37	375	BL	78	DH07	Yes	12.06	x2	Yes	Yes	10.04	23.10	13.05	08.04
40	750	BL	330	NE03	Yes	24.04	x2	Yes	Yes	27.03	27.10	07.05	26.02
42	1056	BL	379	DL07	Yes	13.03	x2	Yes	Yes	13.03	20.09	04.04	19.03
43	1154	BL	451	NE06	OOP	17.04	Yes	Yes	Yes	21.02	Out	of the	Proj.
45	187	HFS	118	NE46	Dec.	17.04	Yes	Yes	No	30.07	Out	of the	Proj.
47	264	HFS	69	NE30	Yes	22.05	x2	Yes	Yes	10.04	27.10	01.05	22.04
48	265	HFS	47	NE06	Yes	13.03	x2	Yes	Yes	02.03	25.09	07.05	04.03
49	267	HFS	48	NE33	Yes	27.03	x2	Yes	Yes	18.03	27.10	12.05	11.03
51	270	HFS	50	NE13	Yes	26.02	x2	Yes	Yes	26.02	20.09	22.04	22.04
52	447	HFS	120	CA06	OOP	27.03	Yes	Yes	Yes	Out	of	the	Proj.
53	503	HFS	189	NE03	OOP	03.07	Yes	Yes	No	Out	of	the	Proj.
54	506	BL	147	NE13	Yes	01.05	x2	Yes	Yes	24.04	27.10	02.05	04.02
55	584	HFS	305	CA01	Yes	27.03	x2	Yes	Yes	27.03	27.10	09.04	25.02
57	1110	HFS	414	NE06	Yes	22.05	x2	Yes	Yes	13.06	27.10	07.05	18.03
58	1210	HFS	456	CA01	Yes	10.04	x2	Yes	Yes	15.05	27.10	01.04	01.04
59	1260	HFS	472	CA18	Yes	03.04	x2	Yes	Yes	03.04	14.10	09.04	18.03
60	1288	HFS	465	NE34	Yes	03.04	x2	Yes	Yes	27.02	13.08	12.05	08.04

89% of the 38 subjects who completed the study, also completed all the required questionnaires and interviews. 100% completed the required evaluation and monitoring questionnaires but a few have missed other (non obligatory) questionnaires. One person had still not completed the 2nd SF36 Q're (despite many reminders) and 4 people did not complete all the remaining questionnaires. The difference in overall response rates between the two groups (100% v 89%) suggests that visitations in the patient's own home obtains a better response rate than requests at clinics, by post or in phone calls.

Application of the Questionnaires

Close examination of the above data will show that, generally speaking, the first interviews took place at, or slightly before, the date of the first appointment of the ONP. Evaluation and monitoring interviews did not take place with the ONP present but they were conducted as close as possible to the first ONP visit. There were exceptions to this policy but this was not considered to affect any of the answers or results. The drop out rate was evenly spread across both groups, as was morbidity, and the statistics for the completion of the questionnaires are shown below :-

TABLE 92. No of people completing each Questionnaire

	Q're 91	Q're 92	Q're 93	Q're 94	Dropped Out
Home Group	44	39	38	38	6
Clinic Group	42	38	38	38	4
Totals	86	77	76	76	10

The following shows the number of subjects and the length of time from completing the first evaluation Q're No 91 to the last Q're No 94. The average period for all 76 subjects was 1.09 years, ie 1 year and 34 days, but as can be seen, over 90% of the subjects were evaluated over a period of 1 year or more.

TABLE 93. No of subjects and length of time to completion

Number of years	0.8	No of subjects	1	Percentage	1.3
	0.9		5		6.6
	1.0		14		18.4
	1.1		34		44.7
	1.2.		21		27.6
	1.3		1		1.3

A total of 239 interviews took place from 4th February 1996 to 14th May 1997 spread over a period of 1 year, 3 months and 10 days. There were a further 5 home visits to subjects who eventually declined to participate or died before further interviews could take place. A total of 11,820 miles were travelled in connection with the monitoring and evaluation of this project.

Finally, it should be mentioned that the author received a tremendous welcome and a high level of co-operation in all the different homes visited and it was noted that everyone, without exception, were extremely grateful that the research project was taking place in their part of world, even those who, eventually for one reason or another, declined to participate themselves.

Comparison between the two groups

A number of different comparisons between the two groups were made based on factual evidence from either clinical files or personal interview and conclusions drawn :

1) Doctor

The doctor who first diagnosed and treated the condition in each case was named and cross-referenced between the two groups. Although there was a predominance of doctors based at Hunters Moor, the RVI and Newcastle General for obvious reasons, there was very little difference between the two groups. Therefore no bias or preference could be shown for one doctor or another between the groups.

2) Geographical Location

A comparison between the geographical location of the two groups showed that the two groups were drawn from roughly the same geographical areas. This showed that there should be no difference between the responses based on location alone.

TABLE No 94. Geographical Location of patients in ONPP.

Location	Home Group	Clinic Group
Co. Durham	12	5
Cleveland (Teesside)	1	0
N. Yorkshire	0	1
Tyne & Wear	37	36
Northumberland	5	10
N. Cumbria	7	8
S. Cumbria	2	2
Total	64	62

3) No of injections before the study

A frequency analysis indicated that the mean of the number of injections before being interviewed for the ESD was 3.94 in the Home Group and 5.03 in the Clinic Group. This also equated well with 8 people in the Home Group and 10 in the Clinic Group never having been injected before the start of the ESD. Although, on average, the Home Group had been injected 1.09 times less than the Clinic Group before being interviewed this was not considered significant to make any difference to future results.

4) No of injections as of 03.07.97

Likewise, by frequency analysis, the average number of injections in the Home Group by 3rd July 1997 was 14.39 and 15.87 for the Clinic Group. Therefore, on average, the Home group had been injected 1.48 times less than the Clinic Group. Therefore the increase from baseline was minimal (0.39 times) and was again not significant.

5) TDS Membership, as of May 1997

TABLE 95. TDS Membership as of May 1997

Value Label	Value	Clinic Frequency	Home Frequency	Clinic Percent	Home Percent	Difference
Yes, a member	1	10	6	16.1	9.4	+6.7
No, not a member	2	37	37	59.7	57.8	+1.9
No longer a member	3	2	3	3.2	4.7	- 1.5
Joined since study	4	12	17	19.4	26.6	- 7.2
Deceased member	6	1	1	1.6	1.6	0.0
Total		62	64	100.0	100.0	0.0

Although there was a slight difference in TDS membership before the start of the study, where 10 of the Clinic group were already members, as opposed to 6 of the Home group, this was more than compensated for in the number of people joining TDS since the start of the study. Therefore current membership, allowing for drop outs and deceased members is 22 for the clinic group and 23 for the home group. This represents a 20% increase in members for the Clinic group as opposed to 183% increase for the Home group.

6) Year of Onset

The earliest year of onset was 1938 in the Home Group and 1944 in the Clinic Group. The latest year of onset was 1994 in both groups.

Year of onset - Clinic Group : Mean 84.871 Median 88.000 Mode 90.000
Valid cases 62 Missing cases 0
Year of onset - Home Group : Mean 83.875 Median 88.500 Mode 90.000
Valid cases 64 Missing cases 0

The mean year of onset is 1984.871 for the Clinic Group and 1983.875 for the Home Group. The median being 1988.00 and 1988.50 respectively. The Mode in both groups was 1990, therefore statistical analysis showed that there was no difference between the two groups regarding onset.

7) Year of Diagnosis

A similar exercise was undertaken regarding the year in which each subject's diagnosis was confirmed. The earliest year of diagnosis was 1944 in the Home Group but 1975 in the Clinic Group. The latest year of diagnosis was 1995 in both groups. The mean year of diagnosis is 1991.403 for the clinic group and 1990.359 for the home group. The median being 1992.50 and 1993.00 and the mode was 1992 and 1993 in both groups respectively. Again there was no significant difference between the two groups.

8) Aetiology of Dystonia

There were 47 subjects in the Home Group and 46 in the Clinic Group with Primary (Idiopathic) Dystonia, 4 in the Home Group and 2 in the Clinic Group had Secondary (Symptomatic) Dystonia and the figures for Hemi-facial Spasm were 13 and 14 respectively. These figures were considered so close as to be almost equal.

9) Secondary Cause

Of the 6 secondary causes known to exist; 2 in the Home Group and 1 in the Clinic Group had dystonia caused by a Cardiovascular Accident (CVA); 1 in the Home Group was drug-induced and another was caused by MS and finally 1 in the Clinic Group had dystonia as a result of Parkinson's Disease. Again, these figures were considered to be almost equal. There was no significant difference in aetiology or secondary cause between the two groups.

10) Socio-Economic Group (SEG) of the patient at time of Questionnaire 20

As can be seen, there is no significant difference between the two groups as far as the SEG of the patients were concerned. The Home Group had slightly more in the 'retired' category, but the number was small (5) and the overall balance was maintained.

TABLE 96. The SEG of both groups

Value Label	Value	Clinic Frequency	Home Frequency	Clinic Percent	Home Percent	Difference
B : Managerial	2	3	2	5.0	3.3	+1.7
C1 : White Collar	3	12	10	20.0	16.4	+3.6
C2 : Blue Collar	4	2	4	3.3	6.6	- 3.3
D : Semi-skilled	5	6	3	10.0	4.9	+5.1
E : Retired	6	37	42	61.7	68.9	- 7.2
Total		60	61	100.0	100.0	- 0.1

11) Marital Status

There is a slight difference in married patients in the home group (5) and widowed patients in the clinic group (5), but there was no overall significant difference between the two groups.

TABLE 97. Martial Status of the two groups

	Clinic	Home
Single	6	8
Married	35	40
Cohabiting	1	2
Separated	2	0
Divorced	4	5
Widowed	12	7
Total	60	62

12) Age

The mean age (at the start of the DNP study) was 54.202 years for the Clinic Group and 57.886 years for the Home Group. The median was 52.000 years and 59.550 years respectively. However there was a large difference in the mode from 33.6 in the Clinic group to 63.4 in the Home group.

Age (as of the date of the 1st Questionnaire, No 91) for the Clinic Group

Mean 54.202 Median 52.000 Mode 33.600
Sum 2276.500
Valid cases 42 Missing cases 20

Age (as of the date of the 1st Questionnaire, No 91) for the Home Group

Mean 57.886 Median 59.550 Mode 63.400
Sum 2547.000
Valid cases 44 Missing cases 20

13) Gender

There is a 15% difference between the sexes in the two groups, ie 10 more males in the home and 8 more females in the clinic group. In general, we know that dystonia affects more females than males, but the difference above was not considered likely to affect the results.

A summary comparing the two groups over the 13 different areas shows :-

1) Doctor Preference - no bias shown to one doctor or another
2) Geographical Location - no difference between the respondents based on location
3) No of injections before the study - not considered significant to make any difference
4) No of injections as of 03.07.97 - the increase from baseline was minimal
5) TDS Membership, as of May 1997 - there was a slight difference before the start
6) Year of Onset - there was no difference between the two groups regarding onset
7) Year of Diagnosis - there was no significant difference between the two groups
8) Aetiology of Dystonia - the figures were considered so close as to be almost equal
9) Secondary Cause - there was no significant difference in secondary cause
10) Socio-Economic Group - there is no significant difference between the two groups
11) Marital Status - there was no overall significant difference between the two groups
12) Age - the difference in mean ages between the two group was only 3.684 years
13) Gender - there was a 15% difference between the sexes in the two groups

It was therefore agreed, at a further meeting of the Management Team, that as there was no significant difference between the two groups that data input and analysis could commence without fear of skew or bias in the results.

The data was input onto SPSS on a coding frame designed by the M/E based on his previous experience and professional qualifications. There was no considered need to ask for other statistical or consultancy assistance.

Input and analysis of data

The SPSS coding frame for the input and analysis of the evaluation and monitoring data was designed as an extension of the existing data on the 'ESD' file. After all the data had been entered and cleaned, the sort option was engaged and a new file 'ONP' was created. It retained all the data obtained from 5th May 1993 as part of the ESD with the added information obtained as part of the ONPP with only 126 cases shown.

For the record, there are 825 variable columns x 126 cases in the 'ONP' SPSS file, thus totalling 103,950 data sets of which over 75% are completed. Missing data has been ignored for statistical purposes or treated as '0' if computations are essential.

The input and coding of data was strictly in the chronological order in which the information appeared in the questionnaires. For the purposes of this chapter, questionnaires 91 through 94 are of paramount importance in the evaluation and monitoring process and data obtained from questionnaires 10 through 81 was used either for demographic purposes, for comparisons across the two groups or as part of the Nurse Practitioners report to validate and confirm other results obtained in his own data collection.

Therefore the next section will deal with the following items in the following order :-

1) Number of injections - before and during the study
2) Number of injectors - before and during the study
3) Time available for treatment - before and during the study
4) Number of injections hurt - before and during the study
5) Rate injectors knowledge - at their best and at their worst
6) Rate injectors attitude - at their best and at their worst
7) Service received at HMH - before and during the study
8) Patients own knowledge - before and during the study
9) What affects the spasms - define and compare the two groups
10) Treatment compared to that received a year ago
11) Service compared to that received a year ago
12) What patients think about the whole project regarding the ONP
13) Preference for injections at home or in the clinic and why
14) Anything else they wanted us to know about this project

<u>The results</u>

01) Number of injections - before and during the study

A comparison has already been made between the two groups in order to ensure there was no bias going into the study. The results showed that "*the mean of the number of injections before being interviewed was 3.94 in the Home Group and 5.03 in the Clinic Group*" ..... and "*the average number of injections in the Home Group by 3rd July 1997 was 14.39 and 15.87 for the Clinic Group*".

The actual average number of injections at the first interview was 9.27 for the Home Group and 12.5 for the Clinic Group. This increased by 2.36 and 2.16 (respectively) by the 2nd and by 2.53 and 2.16 by the 3rd interview. Therefore by the end, the Clinic Group had had, on average, 16.82 injections and the Home Group 14.16. Therefore the difference had reduced slightly over the period of the study. This will be discussed later, but it is noted that this increase in Home Group average injections is almost certainly due to the ONP injecting within the optimum time span rather than being restricted to the appointed dates within the clinic structure.

02) Number of injectors - before and during the study

The same question was asked three times, at the beginning, in the middle and at the end of the study.

A comparison of the two groups across the three interviews shows :-

TABLE 98. No of injectors during the study

		Clinic 1st Interview	Home 1st Interview	Clinic 2nd Interview	Home 2nd Interview	Clinic 3rd Interview	Home 3rd Interview
No of injectors	1	0	2	14	39	21	38
	2	5	4	19	0	14	0
	3	6	10	4	0	3	0
	4	13	12	1	0	0	0
	7	7	9	0	0	0	0
	6	10	7	0	0	0	0
	7	1	0	0	0	0	0
	Totals	42	44	38	39	38	38

The baseline measurement shows that on average each patient had been injected by 4.2 different doctors since they started their Botulinum Toxin therapy. There was very little difference between the two groups, 4.3 for the Clinic and 4.0 for the Home Group. As, by definition, only the Dystonia Nurse Practitioner could inject the Home Group, what is interesting is the number of different injectors in the Clinic Group, starting at 4.3 injectors over 13.2 injections (Ratio 1 : 3.1), continuing with 1.8 injectors over 2.2 injections (Ratio 1 : 1.2) and ending with 1.5 injectors over 2.2 injections (1 : 1.5).

The above might be construed as showing that a patient used to change doctor for his or her injection every 3.1 visits and that went down to every 1.2 visits in the middle of the study and to every 1.5 visits by the end of the study. In practice, however, what it does show is an improvement in the number of doctors injecting the same number of patients and that patients were more likely to get the same doctor (particularly if they ask for him or her) than in the past.

For a long period of time from 1988 to 1991, there was normally only one doctor injecting, ie Dr M.P.Barnes. As the clinic grew in stature and the numbers of patients seeking treatment increased, two doctors were available to inject patients. However, since 1994 when the clinic became a weekly event, the number of doctors available to inject increased to 4 at any one clinic and this did not include the doctors who attended the clinic by way of training in Botulinum Toxin injection techniques.

However, the majority of patients were able to see the same doctor for at least two injections in a row and only two doctors were normally available throughout the period of the study. 50.0% of clinic patients reported being injected by only 2 doctors in the 6 months to the middle of the study and this increased to 55.3% clinic patients had seen only one doctor for their injections in the 6 months to the end of the study.

03) Time available for treatment - before and during the study

Although the same question was asked at the beginning, in the middle and at the end of the study, the answers were subjective based on the patients' memory but they were noted verbatim. The ONP noted exactly the time he spent with each patient but these results are based on the M/E interviews.

TABLE 99. A comparison over time

		Clinic	Home	Clinic	Home	Clinic	Home
		1st Interview		2nd Interview		3rd Interview	
Time in minutes	2	1	1	0	0	0	0
	3	0	2	1	0	1	0
	4	2	2	0	0	0	0
	5	14	14	11	0	7	0
	7	0	3	0	0	0	0
	10	19	18	20	0	20	0
	12	0	2	0	0	0	0
	15	4	1	4	1	5	0
	20	2	1	1	4	3	6
	25	0	0	0	1	0	0
	30	0	0	1	5	2	8
	40	0	0	0	3	0	4
	45	0	0	0	4	0	6
	60	0	0	0	12	0	12
	75	0	0	0	1	0	0
	90	0	0	0	7	0	2
	120	0	0	0	1	0	0
	Totals	42	44	38	39	38	38

The average time overall that the doctors were able to spend with each patient, prior to the start of the study, was 8.3 minutes. This could be rounded up to a statement of "not more than (nmt) 10 minutes per patient". There was no significant difference in this average between the two groups, being 8.8 minutes for the Clinic Group and 7.9 minutes for the Home Group. The main difference between the two groups was that the Home Group had 9 categories from 2 to 20 minutes, whereas the Clinic Group had 7 categories within the same time span. The baseline measurement for both groups should therefore be 8.3 minutes (or nmt 10 minutes). In a few cases (8 in total) the doctor was able to spend between 15 and 20 minutes with each patient. This increase of 50% and 100% over the average should be noted and compared again at the end. The averages for the Clinic Group were 8.8 mins Baseline, 9.7 mins mid-study and 9.6 mins by the end of the study. The averages for the Home Group were 7.9 mins Baseline, 54.2 mins mid-study and 44.5 mins at the end (a considerable increase).

There were 6 people in the Clinic Group who were seen for a longer period in mid-study and this had gone up to 10 people seen for between 15 and 30 minutes by the end. Nevertheless, the overall average time with the doctor remained at "*nmt 10 minutes per patient*" throughout. The average time which the DNP was able to spend with the patient was significantly more than this, although the research itself took up a considerable amount of time, nevertheless the time allotted for treatment remained higher than that allowed for in the clinic group.

04) Number of injections hurt - before and during the study

The comparison was made based on the answers to the question "*How many times has their (his) technique in giving you the injections hurt or been painful ?*", which was asked at the beginning, in the middle and at the end of the study.

TABLE 100. No of injections hurting

		Clinic 1st Interview	Home 1st Interview	Clinic 2nd Interview	Home 2nd Interview	Clinic 3rd Interview	Home 3rd Interview
Never hurt	0	11	8	24	31	19	25
No of times =	1	14	11	8	5	12	9
	2	10	7	5	0	5	3
	3	1	3	1	3	2	0
	4	0	3	0	0	0	1
	5	0	2	0	0	0	0
	6	1	2	0	0	0	0
	8	8	2	0	0	0	0
	9	0	2	0	0	0	0
	10	0	1	0	0	0	0
	12	1	0	0	0	0	0
	14	0	2	0	0	0	0
	15	0	1	0	0	0	0
	18	1	0	0	0	0	0
Hurts everytime	30	1	0	0	0	0	0
	Totals	42	44	38	39	38	38

The two groups compared favourably in the number of times they report the injection technique having hurt or been painful. One person in the clinic group reported it had been painful everytime, but it could be suggested that this person was particularly sensitive to the injection as she had been injected by a great number of different people over a long period of time. Both groups reported over 50% of patients either never having experienced pain or only on one or two occasions. The average over the two groups was every 2.7 occasions. The difference between the groups was every 2.8 occasions for the clinic group and slightly more, ie every 3.5 occasions, for the home group. What can be seen very clearly from the above table, is that (regardless of the reasons why - which will be discussed in detail later) there was a tremendous increase in the number of patients reporting that the technique in injecting them did not hurt at all and was not painful compared to the baseline measurement.

The percentages are :-

	Clinic Group		Home Group	
	Never	Once only	Never	Once only
Baseline	26.2	33.3	18.2	25.0
Mid study	63.2	21.1	79.5	12.8
End study	50.0	31.6	65.8	23.7

	Overall	Overall
Baseline	59.5	43.2
Mid study	84.3	92.3
End study	81.6	89.5

The largest increase was in the Home Group going from 43% to 90% overall in patients reported either never any pain or once only, but interestingly enough there was also a significant increase in the Clinic Group as well from 60% to 80%. In other words, the very fact that the ONP was being assessed independently on his technique in giving the injections seemed to have a remarkable effect on the doctors in the clinic. Whereas the ONP's patients reported a 300% increase in injections free from pain, the clinic patients reported a 116% increase as well.

05) Rate injectors knowledge - at their best and at their worst (combined with),
06) Rate injectors attitude - at their best and at their worst.

The comparison was made based on the answers to a series of six questions, the same two repeated three times, ie., at the beginning, in the middle and at the end of the study. The questions, in relation to the person treating their dystonia, were a) "*How would you rate their knowledge of you and your particular condition* ?" and b) "*How would you rate their personal attitude towards you and your condition* ?" The answers were graded by the subject on a 5 point scale from Very Good to Very Poor but, because there were a number of injectors, it was not possible for the subject to give an opinion on all of them. Therefore the best and worst in each case were identified (anonymously) and graded accordingly. Thus the results show a best knowledge and a worst knowledge, as well as a best attitude and a worst attitude. Where there was only one injector in the previous period, eg the ONP, the answer was sometimes marked the same in best and worst alike.

The results are shown in tabulation form :-

The categories are scored Very Good(1), Good(2), Average(3), Poor(4),Very Poor(5)

The numbers of answers are shown with their overall (percentage).

C = Clinic Group and H = Home Group. 1 : Baseline, 2 : Mid-study, 3 : End-study.

The Total column also shows the (Mean) of the scores (1 to 5) ie V.Good to V.Poor.

TABLE 101. Comparison of injectors knowledge and attitude.

	V. Good	Good	Average	Poor	V. Poor	Total
H1:Best Knowledge	25 (56.8)	11 (25.0)	7 (15.9)	1 (2.3)	0	44 (1.64)
H1: Worst Know.	0	9 (20.9)	16 (37.2)	11 (25.6)	7 (16.3)	43 (3.36)
H1: Best Attitude	27 (61.4)	13 (29.5)	2 (4.5)	2 (4.5)	0	44 (1.52)
H1: Worst Attitude	1 (2.3)	20 (46.5)	10 (23.3)	6 (14.0)	6 (14.0)	43 (2.93)
C1:Best Knowledge	20 (47.6)	17 (40.5)	5 (8.1)	0	0	42 (1.64)
C1: Worst Know.	0	5 (11.9)	17 (40.5)	12 (28.6)	8 (19.0)	42 (3.55)
C1: Best Attitude	24 (57.1)	14 (33.3)	4 (9.5)	0	0	42 (1.52)
C1: Worst Attitude	2 (4.8)	17 (40.5)	15 (35.7)	4 (6.5)	4 (6.5)	42 (2.79)
H2:Best Knowledge	25 (64.1)	12 (30.8)	2 (5.1)	0	0	39 (1.41)
H2: Worst Know.	24 (61.5)	13 (33.3)	2 (5.1)	0	0	39 (1.44)
H2: Best Attitude	32 (82.1)	6 (15.4)	1 (2.6)	0	0	39 (1.21)
H2: Worst Attitude	31 (79.5)	7 (17.9)	1 (2.6)	0	0	39 (1.23)
C2:Best knowledge	15 (39.5)	13 (34.2)	8 (21.1)	2 (5.3)	0	38 (1.92)
C2: Worst Know.	6 (15.8)	9 (23.7)	19 (50.0)	4 (10.5)	0	38 (2.55)
C2: Best Attitude	24 (63.2)	9 (23.7)	3 (7.9)	1 (2.6)	1 (2.6)	38 (1.58)
C2: Worst Attitude	9 (23.7)	14 (36.8)	12 (31.6)	2 (5.3)	1 (2.6)	38 (2.26)
H3:Best Knowledge	27 (71.1)	11 (28.9)	0	0	0	38 (1.29)
H3: Worst Know.	26 (68.4)	12 (31.6)	0	0	0	38 (1.32)
H3: Best Attitude	30 (78.9)	8 (21.1)	0	0	0	38 (1.21)
H3: Worst Attitude	29 (76.3)	9 (23.7)	0	0	0	38 (1.24)
C3:Best knowledge	15 (39.5)	12 (31.6)	9 (23.7)	2 (5.3)	0	38 (1.95)
C3: Worst Know.	8 (21.1)	9 (23.7)	16 (42.1)	5 (13.2)	0	38 (2.47)
C3: Best Attitude	24 (63.2)	8 (21.1)	4 (10.5)	1 (2.6)	0	38 (1.61)
C3: Worst Attitude	11 (28.9)	15 (39.5)	10 (26.3)	1 (2.6)	1 (2.6)	38 (2.11)

There is no baseline differentiation overall between the two groups in either 'best' score being 1.64 and 1.52 respectively but there is a slight drop in the Clinic Group score in worst knowledge (0.19) which is matched by a drop in the Home Group score in worst attitude (0.14). Therefore one can say that the two groups are equal overall at baseline.

However, by the end of the study, there is a very significant swing towards the ONP in the Home Group. There was also a small but definite betterment in the worst scores in the Clinic group, possibly showing the effect that the study has had on the doctors in the clinic.

07) Service received at HMH - before and during the study

The subjects were initially asked to "*tell me (in your own words) what you think about the service you get from HMH*". The second interview asked, "*How do you think the service you currently get could be improved* ?" and the final interview asked the first question again. The answers, often lengthy and contradictory, have been categorised and condensed down from 36 different variables. The results are shown below broken down into positive, neutral and negative comments. Out of the 363 answers, 203 (55.9%) could be considered to be positive, 54 (14.9%) were neutral (being merely comments or suggestions) and 106 (29.2%) were negative.

TABLE 102. Comments on the service received

Variable label	Home 1st	Clinic1st	Home 2nd	Clinic 2nd	Home 3rd	Clinic 3rd
Positive Comments : (203)						
Seen promptly	8	2	0	0	0	0
Staff efficient / polite	6	8	2	0	0	0
No complaints	9	7	0	0	0	0
Good service / v. happy	24	20	9	6	22	15
Receptionist VG	5	2	0	0	0	0
Friendly atmosphere	4	5	0	2	0	0
Can't be improved	0	0	19	6	9	1
Better with same Dr	0	2	0	7	0	0
HMH better than others	1	2	0	0	0	0
Neutral comments : (54)						
Nothing / No comment	0	0	17	11	10	9
Increase no of ONP's	0	0	1	1	0	0
DSS Expenses at HMH	0	0	0	1	0	0
Counselling Service	0	0	0	0	1	1
Annual Review at HMH	0	0	2	0	0	0
Negative comments : (106)						
Occasional waiting	1	3	0	4	1	5
Consultation too short	3	3	1	5	2	3
Like a production line	3	6	0	0	0	1
Better before rec. moved	2	4	0	0	0	0
BT - bad batches / waste	1	0	2	3	0	2
Screen / no privacy	1	1	0	4	0	2
Car parking !	2	1	0	1	0	0
Ambulance service	0	2	0	0	1	0
Canteen (no signs)	0	1	0	0	0	0
Cancelled / delayed app.	1	2	0	0	2	13
More flexibility required	0	0	0	5	0	0
Improve Dr's attitudes	0	0	0	2	0	4
Improve Dr's training	0	0	0	4	0	2
Total = 363 answers	71	71	53	62	48	58

08) Patients own knowledge - before and during the study

The comparison was made based on the answers to a series of eight questions, each set getting progressively harder over the three questionnaires. The first set related to each patient's own dystonia, ie what type of dystonia did they have and what caused it. The second set related to the differences between primary and secondary dystonia, whilst the final questions used more technical (medical) terminology about the brain, etc. The answers were then coded by the M/E on a 5 point scale from very good to very poor.

111 What type of muscle spasm(s) do you have ?
112 Do you know what is the cause of this type of spasm ?
211 Do you have primary or secondary dystonia ?
212 What is thought to cause of primary dystonia ?
213 What are one of the causes of secondary dystonia ?
214 What is the main difference between the two types ?
311 What is meant by idiopathic dystonia ?
312 What is the Basal Ganglia ?

TABLE 103. Patients' own knowledge of their own dystonia

	V.G. = 1	Good = 2	Ave. = 3	Poor = 4	V.P. = 5	Total
111:clinic	22 (52.4)	12 (28.6)	3 (7.1)	1 (2.4)	4 (9.5)	42
111:home	20 (45.5)	14 (31.8)	3 (6.8)	3 (6.8)	4 (9.1)	44
112:clinic	15 (35.7)	9 (21.4)	7 (16.7)	5 (11.9)	6 (14.3)	42
112:home	14 (31.8)	7 (15.9)	9 (20.5)	7 (15.9)	7 (15.9)	44
211:clinic	11 (28.9)	3 (7.9)	11 (28.9)	5 (13.2)	8 (21.1)	38
211:home	16 (42.1)	3 (7.9)	8 (21.1)	3 (7.9)	8 (21.1)	38
212:clinic	11 (28.9)	6 (15.8)	4 (10.5)	6 (15.8)	11 (28.9)	38
212:home	7 (18.4)	9 (23.7)	5 (13.2)	7 (18.4)	10 (26.3)	38
213:clinic	9 (23.7)	6 (15.8)	2 (5.3)	9 (23.7)	12 (31.6)	38
213:home	4 (10.5)	4 (10.5)	3 (7.9)	12 (31.6)	15 (39.5)	38
214:clinic	4 (10.5)	3 (7.9)	6 (15.8)	13 (34.2)	12 (31.6)	38
214:home	2 (5.3)	1 (2.6)	3 (7.9)	16 (42.1)	16 (42.1)	38
311:clinic	3 (7.9)	0 (0.0)	4 (10.5)	10 (26.3)	21 (55.3)	38
311:home	1 (2.6)	0 (0.0)	5 (13.2)	8 (21.1)	24 (63.2)	38
312:clinic	9 (23.7)	7 (18.4)	3 (7.9)	5 (13.2)	14 (36.8)	38
312:home	7 (18.4)	11 (28.9)	1 (2.6)	7 (18.4)	12 (31.6)	38
Totals =	155 (24.7)	95 (15.1)	77 (12.3)	117 (18.6)	184 (29.3)	628

At baseline, ie 111 and 112, one can see that there was a slight difference between the two groups, ie one or two people in each category. The clinic group was more knowledgeable in the 'very good' category and they were overall 9.4% higher over the average giving them an initial slight advantage, ie their baseline knowledge was slightly higher than the home group. The frequency numbers however were relatively small in each category, ranging from a high of 22 to a low of 1.

As we progress, through 211, 212, 213 and 214 to 311 and 312, we see the results of the Outreach Nurse Practitioner (ONP) being able to spend more time with the home group reflected in the fact that there was a slight improvement in this group's scores.

Taking the overall figures in Questionnaires 92 and 93 for the categories over average, the clinic group scored 4.8% higher than the home group. As their baseline measurement was 9.4% higher, this shows the home group having made an 4.6% improvement. However the frequencies in each category for each question were small and roughly similar, the maximum difference being 5 people. Therefore one can draw the conclusion that, although there was a slight improvement in the home group, the difference was small and in fact statistical analysis shows no significant difference.

What was interesting (and surprising to the M/E) was the number of patients who initially did not know what type of dystonia they had nor what caused it, ie 9.3% and 15.1% respectively scoring *'very poor'* ie., they knew absolutely nothing. However anecdotal evidence has proved that this had changed by the end of the study and if the same questions had been asked at the end as at the beginning, then the results would have shown considerable improvement in both groups. The original idea of increasing the difficulty of the question was correct in its conception, but the designer was at fault in not repeating the earlier questions as well. During each interview, if the subject got the answer to a question wrong or was unable to answer at all, the correct explanation was given at the end of the session. However, in retrospect, the next interview should have repeated the questions from the previous interview in order to test the subject's retentive powers.

09) What affects the spasms - define and compare the two groups

TABLE 104. What makes the spasms worse ?

Answers	Clinic	Percent	Home	Percent
Nothing	2	3.1	7	10.8
Other peoples smoke	0	0.0	1	1.5
Social situations	5	7.7	3	4.6
Stress / Anxiety / Worry / Dep.	20	30.8	26	40.0
Angry, Upset, Tense	8	12.3	8	12.3
Cold	3	4.6	3	4.6
Physical activity	11	16.9	6	9.2
Tiredness	9	13.8	2	3.1
BT cycle too long	0	0.0	1	1.5
Sneezing	0	0.0	1	1.5
Reading / TV / Bright lights	3	4.6	5	7.7
Wind	1	1.5	1	1.5
Migraines	0	0.0	1	1.5
Agoraphobia	1	1.5	0	0.0
Loud Noise	1	1.5	0	0.0
Caffeine	1	1.5	0	0.0
Totals	65	100.0	65	100.0

TABLE 105. What makes the spasms better ?

Answers	Clinic	Percent	Home	Percent
In remission or an operation	1		1	
Medication	1		3	
Sleep	3		3	
Heat / Warmth	5		3	
Bot Tox	17		22	
Rest / Relaxation	19		13	
Smoking Cannabis	1		1	
Positive thoughts	4		2	
Nothing (except BT)	8		5	
Alcohol	4		5	
Good days / bad days	2		0	
Geste Antagoniste or Chewing	1		1	
TENS machine	0		1	
Totals	66		60	

No comparisons can be drawn as the individual numbers are not large enough to have any statistical significance and there is no real difference in any of the answers. As could have been expected; stress, anxiety, worry and depression was the largest cause of increasing spasms, with physical activity, tiredness and emotions a close second. Likewise Botulinum Toxin and relaxation are the major players in helping the spasms.

10) Treatment compared to that received a year ago (combined with),
11) Service compared to that received a year ago.

TABLE 106. Treatment and service compared to a year ago

Question ?	Answers :			Question ?	Answers :		
Treatment improved ?	Yes	No	Same	Service improved ?	Better	Worse	Same
Clinic Group = 38	11	5	22		13	5	29
(%)	28.9	13.2	57.9		34.2	13.2	52.5
Home Group = 38	28	0	10		30	0	8
(%)	73.7	0.0	26.3		78.9	0.0	21.1

Over 50% of the Clinic Group felt that the treatment and service had remained the same over the period of the study, with approx. 30% feeling that both had improved or got better in some way. Exactly 13.2 felt that both treatment and service had not improved or had indeed got worse. However, the Home Group overwhelmingly voted the study a success in that approx. 75% felt both treatment and service had improved or got better, with only between 21% and 26% thinking they had remained the same or not improved. Nobody felt that either had not improved or got worse in the Home Group. This was very significant with the statistical analysis showing $p < 0.001$.

12) What patients think about the whole project regarding the ONP

TABLE 107. Patients' comments about the whole project

Comments	Home	Percent	Clinic	Percent
Less stressful at home	6	6.2	3	3.2
DNP is the best injector	5	5.2	3	3.2
Better now diagnosed	5	5.2	3	3.2
Good idea / scheme	21	21.6	27	28.7
You are listened to	8	8.2	6	6.4
DNP very effective	6	6.2	5	5.3
Good - Very good	5	5.2	1	1.1
Saves travelling time	9	9.3	7	7.4
I have control !	1	1.0	1	1.1
DNP - same person	11	11.3	7	7.4
DNP - easy cases	4	4.1	10	10.6
Annual review	3	3.1	1	1.1
Dr's time limited	5	5.2	2	2.1
HMH - hard cases	4	4.1	9	9.6
Fall back - HMH	2	2.1	0	0.0
Wish I was on Home List	0	0.0	4	4.3
Staff - vg - pressure now off	2	2.0	5	5.3
Totals	97	100.0	94	100.0

The above is purely a frequency analysis based on the cumulative responses given as the questions was open-ended; "*Please, tell me in your own words, what you think about the whole project regarding the Nurse Practitioner.*"

13) Preference for injections at home or in the clinic and why

31 of the Home Group (81.6%) preferred home visits, none preferred the clinic, but 7 (18.4%) declared that they would not mind either. The figure dropped to 21 subjects (55.3%) in the Clinic Group preferring home visits, with 12 (31.6%) saying they would not mind either. Only 5 people (13.2%) actually stated they preferred the clinic.

TABLE 108. Preference for home or the clinic

Why home ?	Home	Clinic	Why clinic ?	Home	Clinic
More relaxed	11	3	Need Dr's advice	2	4
More convenient	20	25	Fixed time	1	0
More time	7	4	Happy either venue	3	5
Same person	11	3	No travel problems	3	9
DNP listens / explains	8	1	Coffee is free	2	1
Personal attention	6	3	See other people	1	3
Self conscious at clinic	3	4	Other things to do	1	5
Diff. walk./ travelling	4	4	Fits in with work	1	1
Time off work	2	6	Others need NP more	1	3
Saves money	1	5	Family unaware	0	1
Total	73	58		15	32

14) Anything else they wanted us to know about this project

The total of 26 variables were established from the answers given and are listed below. A Content Analysis was conducted on the written answers and will be discussed later, together with a statistical analysis of the following.

TABLE 109 (a). Final comments on the ONP project from the Home Group

Comments	The Home Group	Percent
Hope it continues	10	13.9
Very satisfied/happy	8	11.1
No comment	8	11.1
Many thanks	6	8.3
Personal relationship	5	6.9
Organised well	4	5.6
Clinic too busy	4	5.6
Offer research	4	5.6
Results should be published	3	4.2
HMH better NRI	3	4.2
NRI - waiting long	3	4.2
NRI - mixed clinic	3	4.2
Awareness - GP's	2	2.8
No Diff in treatment	2	2.8
Home more personal	2	2.8
Your family / Medical Prize	2	2.8
Clinic patients / Explanations	2	2.8
Longer injection cycle	1	1.4
Totals	72	100.0

TABLE 109 (b). Final comments on the ONP project from the Clinic Group

Comments	The Clinic Group	Percent
No comment	15	22.4
Very satisfied/happy	7	10.4
Clinic improved	7	10.4
Offer research	7	10.4
Hope it continues	6	9.0
Counselling Service	4	6.0
Home more personal	3	4.5
Personal relationship	2	3.0
Many thanks	2	3.0
More explanation	2	3.0
Ambulance travel	2	3.0
BT batch strength	2	3.0
App's cancelled why?	2	3.0
Organised well / Results published	2	3.0
Clinic too busy / No Diff in treatment	2	3.0
HMH better NRI / More ONP's needed	2	3.0
Totals	67	100.0

PART FOUR : ANALYSIS OF FINDINGS

CHAPTER 27

The Impact of Focal Dystonia on the Working Life of Musicians

Although something was known about the prevalence of dystonia in general, there was originally little reliable information about the incidence of individual types of dystonia, however, during the Epidemiological Survey of Dystonia (ESD), a small number of musicians were identified with different forms of focal dystonia.

The author had an opportunity to address a group of mainly medical doctors at an International Conference held in York, England during 23rd to 27th March 1997 entitled " Health and the Musician. " This conference was organised by the British Association of Performing Arts Medicine and the author was able to present three case histories to the conference.

By the time of the conference in 1997, this research project had already a good idea that social isolation in patients breeds a lack of diagnostic skill in Family Practitioners, which in turn leads to non or misdiagnosis and thus increases the isolation felt by most sufferers. Therefore the onset of dystonia for a musician can have a particularly debilitating effect, especially if that musician develops a focal dystonia affecting the hands or neck.

The three case histories involved a professional 'rock' guitarist, a semi-professional 'club' guitarist and singer and an amateur 'classical' pianist (please note these were all male, despite dystonia having a 2:1 female to male ratio in the ESD). The case histories show how the onset of dystonia has affected the working life of each musician. Each case history has four sections ; a) the chronological advance of the disorder, b) how this affected their playing, c) any successful treatments and d) the psychological impact of the disorder on the subject.

The impact of focal dystonia makes no distinction between primary and secondary causation, because an abnormal posture or involuntary muscle spasm presents in exactly the same way in both cases. If the subject is unemployed, due to their dystonia, it does not matter if it was induced, genetically inherited or idiopathic - they are still economically vulnerable. The dystonic lives of these three musicians are as described below :-

<u>CASE HISTORY No 1 :</u>

A single male, aged 32, who is a self-employed musician with no familial history of dystonia. He developed focal dystonia in his left (fretboard) hand in 1991, which was diagnosed 2 years later, but by 1994 had developed a dystonic tremor in his right hand also.

Initially he only had a feeling that his fretboard fingering was not as fluent as before, until one particular practice session when his 5th finger was observed to extend involuntarily. Over the next 18 months, this same digit became effectively useless, even for the most simple chord shapes. However his first three fingers were unaffected and he continued to play, simply refingered everything by altering four finger chords to their important components. Eventually the extension of the 5th finger became incessant whilst his 4th finger began to flex. Then his 3rd finger flexed completely, so that when gripping the guitar neck, the pressure could break open the skin under the nail. His 1st finger then began to extend and remained in extension, whilst his thumb developed a tremor. His left hand fingers also began to shake slightly and he could feel this sensation right up to his elbow.

Throughout this period a number of alternative treatments were tried with no success and drugs, such as Artane and Clonazepam, had no positive effect. Since September 1994, the patient has been successfully treated with Botulinum Toxin therapy, although it has taken some time to find the optimum injection sites and dosage. Currently he has a regular set of injections every three months, which reduces the involuntary flexion of the 3rd and 4th fingers and the extension of the index finger. A further injection to reduce the extension of the 5th finger is repeated every six months. A side effect of the injections is a slight weakness which makes it difficult to grip the neck of the guitar and he has to 'pace' his playing activity or risk running out of sufficient grip before the end of a night's playing. There is usually a 2 to 3 week gap after injections before playing gigs. He has also had to change some electrical equipment so that everything can be carried with only one hand. He considers to have regained, on average, about 60% of his former ability and usually, within the 3 month injection cycle, there is a substantial amount of time when he has full control again. His biggest psychological problem has been that his life's ambition, ie., to play music professionally, was at one time taken away from him completely. He has, however, coped with it very well and measures only as moderate on the anxiety / depression scales administered during the survey. He has met a number of other dystonia patients at the injection clinic and, in his own opinion, has less obvious disability than any of them, nevertheless dystonia has created a severe handicap and had extremely debilitating effect on his life as a musician!

CASE HISTORY No 2 :

A 52 year old widower, who had had a semi-professional career as a singer / guitarist working on the northern club circuit. He developed Spasmodic Torticollis in 1985 and was diagnosed in 1991. He eventually took early retirement as a Plumbing and Heating Engineer and has finished playing in public entirely. He has a Laterocollis to the right but it is not clear if this is as a result of, or coincidental to, his alcoholism. According to strict criteria he was not included in the Epidemiology of Primary Dystonia (EPD), as it has not been possible to determine primary causation, but he remains in the ESD.

Although his focal dystonia is in his neck and does not present in any other part of his body, it nevertheless has had a very profound effect of his playing. He found it difficult to stand for long periods with his guitar sling around his neck, which created great difficulties whilst standing on stage playing a long set.

Even when playing within a group situation, sitting was difficult as his chair was turned in a different direction to other band members. Because his pain / discomfort levels have been measured as severe, it was difficult to find a comfortable position to play any instrument, but mainly his guitar. It was particularly painful when learning a new song or rehearsing as he could not turn his head to the left to look at the fretboard for any length of time. Eventually he found that he could not even sit for a whole evening and therefore had to take frequent breaks in order to walk around and exercise his neck thus disrupting band and audience. He started Botulinum Toxin therapy in January 1993, two years after diagnosis, and has been regularly injected with between 240 and 360 units of the Speywood Product approximately every 12 weeks with an 80% degree of success. He has a Peptic Ulcer and Hypertension, but has remained very cheerful and stoic. He no longer drinks alcohol and therefore his whole way of life has changed as he no longer frequents his usual social establishments nor plays any longer in public. Nevertheless he has coped with the disorder and its effects extremely well, with the support of his current partner and family.

CASE HISTORY No 3 :

A divorced male, aged 53, who is a prematurely retired teacher. He developed action-induced dystonia in the fingers of his right hand in 1981, but it was not correctly diagnosed until 1995. He is a very keen amateur pianist, who also plays the clarinet. It is possible the trigger for the onset of the dystonia was severe emotional stress due to his marriage break up, combined with a fracture of the 5th metacarpal and torn ligaments between the 3rd, 4th and 5th fingers in the right hand and several subsequent other physical traumas.

The first indications in 1981 concerned the index finger being 'triggered' when playing sequential configurations with his 3rd, 4th and 5th fingers in his right hand. It was also noticed that the 4th and 5th fingers were flexing when not in use whilst playing. Initially exercises were developed to counter this trait, but this helped only at the expense of speed of movement.

Eventually playing proved extremely difficult because, when the 3rd finger went down onto the keyboard, it remained clamped whilst flexing strongly into the key, meanwhile the index, 4th and 5th fingers triggered backwards, with flexors and extensors locked against each other and seemingly always set off by the 'glued' middle finger. He has been unable to play sequences comfortably faster than mm. 44-50 since. He had tried a number of different therapies before Botulinum Toxin in 1996, which weakens the flexing of his right 1st and 2nd digits.

Throughout this entire period, he has experienced no pain in his right hand, but in his own words :-

" *This co-ordination problem has meant that I have lost the facility to play comfortably the music that gave me the deepest satisfaction - any Mozart / Haydn piano sonata, Scarlatti, Bach's Goldberg variations - linear music. I have still not accepted in some way that this has happened and I often sit down to play feeling that I might play naturally this time !* "

PART FIVE : DISCUSSION

CHAPTER 28

Epidemiology

As the previous chapters clearly show, the primary difficulty in doing any research into dystonia in the North East, or indeed in any part of the UK, was correctly identifying sufficient numbers of people with the disorder. It should not be underestimated the tremendous effort and resources required to successfully accomplish this task. To start in May 1993 with so few numbers and to have proved by 1998 that it was a much more prevalent movement disorder in the UK has been of tremendous significance.

The Epidemiology of Primary Dystonia (EPD) in the North of England (Duffey et al, 1998) was described at the 4th International Dystonia Symposium held in Miami, Florida in October 1996 as *"one of the important pieces of research in its field"* by the late Professor C.D. Marsden, who was one of the most eminent authorities on dystonia research (Marsden & Fahn, 1998). This research, along with its associated Epidemiological Survey of Dystonia (ESD) and the Epidemiology of Spasmodic Torticollis (EST), will have far reaching implications for many years to come.

It should be remembered that the epidemiology was not only the tool which enabled a number of other research projects to be carried out, but was also an essential and important piece of research in its own right. However, none of the epidemiological evidence could have been brought to light, without firstly correctly identifying and diagnosing a large number of previously unknown (and thus undiagnosed) people.

As previously shown, there could be as many as 68.6% of the current dystonic population remaining undiagnosed, currently undetected by the ESD. Even if this figure is as low as 46.5%, this still means that, to date, approximately another 400 people are yet to be discovered in the North East by the researchers. This is the one of the next areas of additional research proposed from the year 2000 onwards.

Distribution

The following figures show how prevalent dystonia really is, after all the extensive work carried out in the North East of England. If one takes the four Northern Eastern counties, excluding Cumbria and North Yorkshire, the overall number is 779 dystonics within a population of just 2,542,448, which equates to 1 in 3,264. Furthermore the Darlington figures show how high it can go with an individual prevalence of 1 in 2,400 therefore just imagine if the ESD were registering that relevant number throughout the 4 north eastern counties, one would see a total of 1059, or an additional 280 people remaining undiagnosed within this region. The figures for the geographical distribution of dystonia is shown next and one will see how the different figures correlate or not.

TABLE 110. The Geographical Distribution of Dystonia within the ESD.

County	District	No. of people	%	Pop'n	No. per 100,000
	Chester Le Street	11	1.2		20.90
	Darlington	50	5.3		50.55
	Derwentside	18	1.9		20.92
	Durham	30	3.2		37.19
	Easington	26	2.8		26.58
	Sedgefield	37	3.9		40.87
	Teesdale	6	0.6		24.29
	Wear Valley	21	2.2		33.47
Co. Durham	=	199	21.2%	631,437 =	31.52
	Hartlepool	23	2.5		25.47
	Middlesbrough	53	5.7		37.14
	Redcar & Cleveland	50	5.3		34.79
	Stockton-on-Tees	51	5.4		29.41
Cleveland	=	177	18.9%	570,207 =	31.04
	Gateshead	67	7.2		32.97
	Newcastle	86	9.2		31.47
	North Tyneside	56	6.0		28.66
	South Tyneside	39	4.2		24.79
	Sunderland	82	8.8		27.67
Tyne & Wear	=	330	35.2%	1,061,513 =	31.09
	Alnwick	3	0.3		9.97
	Berwick-upon-Tweed	3	0.3		11.22
	Blyth Valley	24	2.6		30.16
	Castle Morpeth	10	1.1		19.88
	Tynedale	19	2.0		33.17
	Wansbeck	14	1.5		23.06
Northumberland	=	73	7.8%	279,291 =	26.14
	Allerdale	18	1.9		18.58
	Carlisle	27	2.9		26.85
	Eden	11	1.2		15.43
North Cumbria	=	56	6.0%	267,560 =	20.93
	Barrow	3	0.3		4.10
	Copeland	14	1.5		30.71
	South Lakeland	11	1.2		11.35
South Cumbria	=	28	3.0%	215,603 =	12.99
North Yorkshire		20	2.1%		n/a
The rest of Yorkshire		22	2.3%		n/a
Other Areas in the UK		32	3.4%		n/a
Grand Totals =		**937**	**100%**		

863 people within the North East and Cumbria gave an overall prevalence of 28.54 per 100,000 people based on an overall population of 3,0235,611 people.

TABLE 111. The spread of dystonic spasms in the region

Type of dystonia	Durham	Teesside	T & W	N' land	Cumbria	Total
Oppenheim's Dys.	3	2	4	1	4	14
Fam. Torticollis	4	5	4			13
Fam. Cerv-Cran	2		2			4
Other Familial	8	10	8	2	5	33
Sporadic	103	86	200	44	56	489
D.R.D.	3	1	3			7
Myoclonic		1	4			5
DRD - familial		4	3			7
Myo. - familial			5			5
2'ndary cause	2	1				3
Levo-Induced	2		1			3
Tardive Dyst.	6	3	6			15
C.P.	1	2	3	1	2	9
M.S.	3	1	2	1	2	9
Infectious			4			4
B.E.T.			2			2
Cerebrovascular	2	3	5			10
Metabolic ?	1		2			3
Head Trauma	1	1	3			5
M.S.A.		1				1
Shy Drager's	1					1
Leigh's Disease	1					1
P.D.	1		1	1		3
Tardive Dyskinesia	2	2	1	1		6
Parox. Ch-Athotos.	1					1
Parox. Dystonia	2		1			3
Parox. Familial	1					1
HSP	1	1				2
CP		1				1
Myoclonus						0
B.E.T.		1	1	1		3
Per. Neuropathy			1			1
Uln. Nerv. Palsy			1			1
Chorea	1					1
ME	1					1
MS			1			1
Arthrogryposis				1		1
Spasticy 2nd Stroke		4	2	2		8
Hemi-facial Spasm	40	44	57	18	12	171
Undefinable	5	2	3		2	12
Psychogenic	1	1			1	3
Total =	199	177	330	73	84	863

The above chart defines all the different categories found in the region as a whole.

Northumberland is an area that needs a lot more work carrying out before a true prevalence can be established. However it gives a reasonably good prevalence for Blyth Valley and Tynedale of 30.16 and 33.17 respectively. The problem is the relative sparseness, particularly in Alnwick and Berwick-upon-Tweed with just 3 people diagnosed in both districts. Based on the work done elsewhere, particularly if one uses the figures for Blyth Valley one could anticipate a prevalence of at least 30 per 100,000 which means there are at least another 8 unknown people in each district.

The prevalence figures for Cumbria reflect that certain areas are better covered than others. Carlisle has a prevalence of 26.55 per 100,000 whereas Barrow-in-Furness has just 4.10 per 100,000. However most of this discrepancy can be explained by the fact that most known dystonics in Carlisle are treated by the North East medical professionals, whereas the majority of people in Barrow travel to Preston or Liverpool for their treatment and are therefore not easily identified within a North Eastern prevalence study.

TABLE 112. The spread of dystonic spasms in the rest of the ESD

Type of dystonia	North Yorkshire	Rest of Yorkshire	Rest of UK	Total
Oppenheim		1		1
Fam.Tort.	1		1	2
Fam Cer-C		1		1
Oth. Fam.	1	2	2	5
Sporadic	9	16	27	52
D.R.D. fam	1	2		3
Tard. Dyst.	2		1	3
Cerebrovas	1			1
P.D.	1			1
Parox. Dyst	1			1
Myoclonus	1			1
HFS	2		1	3
Total =	20	22	32	74

These are included as shown, however one should be aware that the majority of those shown in North Yorkshire are often treated or seen at the North Riding Infirmary in Middlesbrough. The rest of Yorkshire are generally also seen there as well because it is only one of two or three hospitals in the country treating Spasmodic Dysphonia. The problem is now to see how many people remain "undiagnosed" within the North Eastern population.

The following map shows the geographical spread of dystonia across the North of England and defines the distinctive areas where more work is required and in particular highlights those areas which reflect where the highest prevalence is shown.

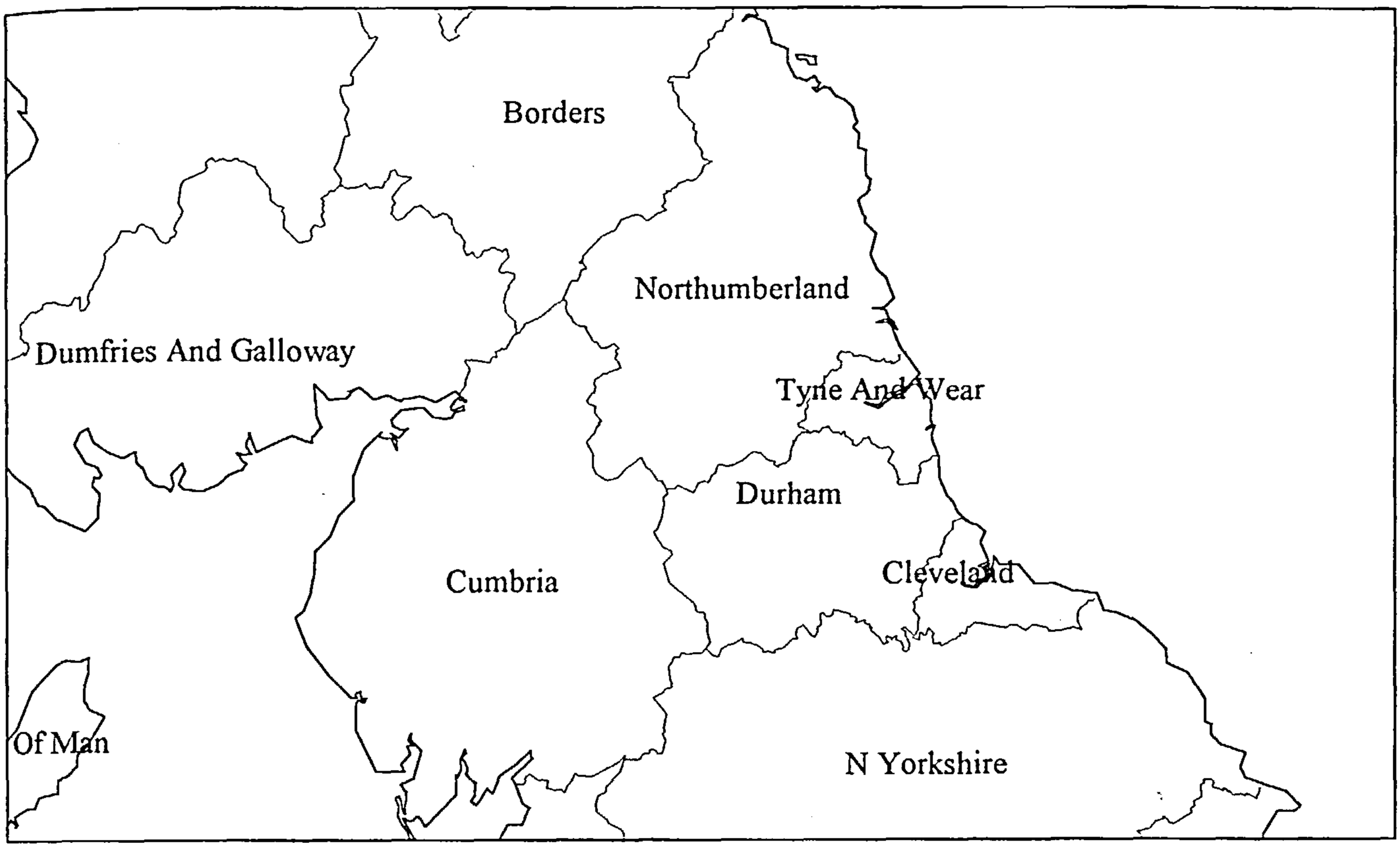


CHART 113. A map of the North of England showing the prevalence of dystonia.

County or Area	Population	Dystonics	Other related disorders	Dystonic proportion
Tyne and Wear	1,061,513	264	66	1 in 4,021
Co. Durham	631,437	150	49	1 in 4,210
Cleveland	570,207	123	54	1 in 4,636
Cumbria	483,163	69	15	1 in 7,002
Northumberland	279,291	51	22	1 in 5,476
N. Yorkshire	-	17	3	-
Rest of Yorkshire	-	22	0	-
The rest of the UK	-	31	1	-
Totals	3,025611	727	210	Average = 1 in 4,605

This represents the current figures for the Epidemiological Survey of Dystonia based on Primary and Secondary subjects only - removing other related disorders, including all the Hemi-facial Spasms - some of which will have a Primary Dystonia.

There are however, apart from certain sparseness of diagnosis, certain types of dystonia which show markedly difference diagnoses between certain geographical areas.

Spasmodic Dysphonia

An example of this "undiagnosis" can be shown by examining the data collected on Spasmodic Dysphonia and examining where the people with this disorder live.

There were 53 people with Spasmodic Dysphonia (SD) in the ESD. 3 of whom have it as secondary to a stroke or an aneurysm, in 2 cases it was considered to be pyschogenic, 1 person has it, as well as Parkinson's Disease (thought not to be related) and 1 person has it with Writer's Cramp, ie dystonia of the hand. However the remaining 46 people have just SD or dystonia of the larynx.

There are 45 North Riding Infirmary patients, 2 more are registered at Hunters Moor Hospital in Newcastle, 3 others are local to the North East but do not go to any hospital currently and 3 are from afar and were part of the research controls in previous research into the use and effectiveness of Botulinum Toxin therapy in its treatment. Using Darlington, a typical North Eastern town with approximately 100,000 population, as a testing ground for the research, it has currently the highest concentration of dystonia anywhere in the world, with currently 48.14 people per 100,000 people with dystonia. There were 4 Spasmodic Dysphonias in Postal Codes DL1, DL2 and DL3, thus making an average 3.92 cases of SD per 100,000 people in the town.

The definition of Adductor as opposed to Abductor Dysphonia is " *Patients with adductor SD exhibit a choked, strained-strangled voice quality with abrupt initiation and termination of voicing resulting in short breaks in phonation. Patients with abductor SD exhibit a breathy, effortful voice quality with abrupt termination of voicing resulting in aphonic whispered segments of speech* " (Brin et al, 1998). Between Mitchell Brin, Andrew Blitzer and Celia Stewart, they have 639 Adductor patients (85.5%) and 108 Abductor patients (14.5%) in New York, USA. The former are easier to treat than the latter and generally produce better results. In the past, Spasmodic Dysphonia was often considered a psychological disorder, this research has shown that in less than 4% of cases was it considered to be pyschogenic and this is not proven, only thought to be the only reason the larynx does not respond to various treatments, as all other larynxes do.

The North Riding Infirmary's Otolaryngeal Department has 45 SD patients, 6 of whom have *Abd*uctor Dysphonia (13.3%) with unusual combinations, whereas the remaining 39 have *Add*uctor Dysphonia (86.6%), which is generally much better at responding to Botulinum Toxin. Although the ESD did not specifically differentiate between Adductor and Abductor Dysphonia during the research, it is known there are tremendous differences when treating, particularly when injecting Botulinum Toxin into the larynx. These figures compare very well with the New York data (within 1.2%) and there was no statistically significant difference

The following shows where all 53 (SD) ESD people live, according to Post Code:-

14 live in Teesside area	TS
11 live in Darlington area	DL
5 live in Leeds area	LS
3 live in Tyne & Wear and Northumberland	NE
3 live in York area	YO
3 live in Bradford area	BD
2 live in London	TW & SE
2 live in Sunderland area	SR
2 live in Wakefield area	WF
2 live in Carlisle area	CA
1 lives in Lanchester area in Cumbria	LA
1 lives in Durham area	DH
1 lives in Hull area	HU
1 lives in Halifax area	HX
1 lives in Sheffield (Rotherham)	S
1 lives in Birmingham area	B

---

53 in total in the ESD

This just goes to show how much more work there is to do. Why are there only 2 people with Spasmodic Dysphonia in Sunderland, 3 in Tyne & Wear and Northumberland and yet 13 on Teesside ? The Speech Therapists and Otolaryngeal Surgeons in these areas need to be contacted as there must be statistically far more in these areas.

If Darlington's statistics are used again, being the most reliable as a base, with 3.92 per 100,000 people and based on a population of 2.6 million in the North East alone, there should be 102 people with SD in this region - the ESD shows us just 33 - or roughly a third. So the question must be asked again, where are they ?

The reason is perhaps quite obvious. People with SD do not normally get to see a Neurologist or an Otolaryngeal Surgeon, but another doctor who often does not recognise this condition unless they are very lucky. Where are the other 69 people ? How long will it take them to get some effective treatment ?

This is the next stage, which must be pursued, to find these people and ensure that they get the opportunity to be able to be diagnosed properly and receive treatment which might help their situation.

Although this must be the next stage of the operation, in the meantime, the above gives the results of where all the people who registered for the ESD in the research live. There are 45 people who have since deceased but who are included in the overall numbers.

This is the first time the full epidemiological survey of dystonia has been seen in such detail and it will remain a useful tool for all future research in the region and indeed elsewhere in the UK.

PART FIVE : DISCUSSION

CHAPTER 29

Diagnosis

Beyond doubt, this series of research projects has shown that increased research dramatically increases the number of correctly diagnosed dystonia patients and one of the lessons of that has been that the greater the number of new correctly diagnosed dystonia patients, the greater the pressure becomes on the medical establishment to provide better and more effective treatment centres. This research is extremely popular with those patients diagnosed through the various awareness raising campaigns run over the past eight years, but remains less popular with the NHS financial managers, who have seen a ten-fold increase in the cost of the Botulinum Toxin to the NHS in this region.

Prior to the start of this research, the Botulinum Toxin clinics were run monthly and with one or two medical staff in attendance. The increased diagnosis has meant an increase to weekly clinics, an increase in medical staff hours required to cover the additional workload and the provision of an Outreach Dystonia Nurse Practitioner to reduce this increased cost to the NHS.

Having proved that dystonia is far more prevalent than previously thought, next to Parkinson's Disease, dystonia must be placed on the curriculum at **all** medical schools. It is still not officially on the neurological curriculum at Newcastle University Medical School, although it is taught unofficially and the local group have established an essay prize for 4th year students to fund travel bonds during their electives. This has proved very successful and is currently being extended to student nurses at The School of Nursing at Teesside University as well.

The results show a definite improvement in not only the rate of correct diagnosis but also the length of time it takes to obtain a correct diagnosis. The few extreme cases of many years from onset to correct diagnosis should (hopefully) be relegated to a different age of uninformed and inadequate diagnosis. Recent interviews have shown that most cases have a correct diagnosis within a year, nevertheless it would be wrong to be complacent and sit back on ones' laurels. The number of new cases coming to light, through the epidemiology, is growing weekly and needs to be monitored closely.

There is no reason to suppose (there is demographic evidence even to support it) that the North East is any different in its dystonic population to any other part of the country. There are certain parts of the country that have an older population, mainly due to their popularity as retirement areas, the West Country being an example. However, this would mean an increased incidence of the focal dystonias which are more prevalent in the older population with onset in later life.

One can assume that a large number of patients with onset in their 50's, 60's and 70's in the interval during the wars, ie 1919 to 1938 have since died, but where are all those undiagnosed people who had onset since the end of the Second World War ?

Only 3 subjects are known to have been developed dystonia in each year, 1950, 1960 and 1970. If we assume an average incidence (taken from 1990 to 1994) of 60 people per year developing one of the forms of dystonia, where are the remaining 57 people. Reference to Table 14 on page 53 shows the amount of people who remain undiagnosed throughout the 1950's, 1960's and 1970's. It is not until 1980 that we even reach double figures for diagnoses. The answer therefore must be that they remain undiagnosed, certainly untreated with Botulinum Toxin therapy, which has proved to be the most effective treatment for many years.

The membership of the Dystonia Society could be used as a rough guide as to the success or otherwise of both awareness raising and consequently diagnosis. The membership of TDS locally remained at approximately 25% of those identified as having dystonia in the North East. Its membership nevertheless has grown steadily and with increasing numbers since the start of the Awareness Raising campaigns in 1992.

The aim must now be to turn these new research subjects into active (or even passive) members and thus ensure that the work started in 1993 continues with the assistance the National Lottery Charities Board award and to continue for many years to come.

The formal diagnosis of the people was actually quite straight forward once they were identified. The problem was one of finding them in the first place. When the research was started there were less than 150 people formally identified, therefore it could be said that this research has helped to get over 800 people diagnosed. Although that is not correct, it is true to say that 584 people have been diagnosed since the research started and only 281 had an onset during this period, therefore it is true to say that at least 300 people have been diagnosed because of this research. Furthermore this research can claim credit for getting the largest number of people formally identified with a form of dystonia within the regional database, that is for sure.

There have been a number of problems with getting people diagnosed because, although the medical people have been truly wonderful in working so hard to get this tremendous number formally identified and diagnosed, there have been some serious problems in the first place and some very sceptical people particularly in both the medical profession and unfortunately also in The Dystonia Society, but not in the North East. The level of co-operation has been truly tremendous and praise should especially go to those people in the North East who have supported and encouraged this research.

PART FIVE : DISCUSSION

CHAPTER 30

Treatment

The treatment of dystonia and all of its ancillary conditions has been totally changed by the advent of Botulinum Toxin therapy. Not everybody is on Botulinum Toxin, only 77.1% of the total number of people in this research have even tried it, but nevertheless it has totally revolutionised the treatment of this condition, which can be extremely painful and disabling for the majority of those with it.

Currently out of our 892 alive sample, just a fraction under 60% (532) are still receiving their Bot. Tox. injections every 8 to 12 weeks and some can go up to 3 or 4 months before requiring a further injection. A tiny few can go just over a year or so but these are exceptional and do not have severe spasms. A few, fortunately very few, have developed antibodies against the toxin. These are then put onto another manufacturers toxin and in some cases it works, in others the antibodies are in effect.

We are fortunate in that, although historically we have been using the British Bot. Tox. as our major injection source in the area, we have been able to use the American version, currently manufactured in Westport, Co. Mayo in the Republic of Ireland. Fortunately and coincidentally both John Whitaker, the Outreach Dystonia Nurse Practitioner, and the author were holidaying in that part of Ireland in September 1998 and were able to avail ourselves of a personal invitation to tour the factory in the company of the Manager.

The use of the American version was only mentioned in the early days of the research, when it was running at between 15 and 20 patients in 1995, but that number has increased considerably since then, however unfortunately any details for the research were stopped after 1996. Table 16 on page 57 shows that of the current treatments being used, 493 currently were being injected with Bot. Tox. and of those over 95% felt the effect of this treatment was better than without it. Only 2 people (at that time) felt worse because of it and this is also very significant. 20 people were currently unchanged and this could mean these were the potential number who were developing antibodies to the treatment - this is another area that needs to be followed up and discussed in more depth in another research paper.

60% of all the current patients in the Epidemiology are still receiving Botulinum Toxin injections and this leaves 40% who are not currently receiving any at all, about 20% of whom have <u>never</u> received any Bot. Tox. These people fall mostly into two distinct categories, those who are quite happy not to receive regular injections as their dystonia is not very severe and they always have the option to start treatment if their spasms get worse and those with generalised or quite severe dystonia who would not benefit from Botulinum Toxin treatment at all.

The list of the different drugs currently being used is very large with a total of 20 prescribed drugs described in Table 20 on Page 61 as being a primary medication, a further 52 different medications in Table 21 on Page 62 being described as secondary and another 115 different drugs being described as not associated with their dystonia.

Of these primary medications, the two main ones are Diazepam and Benzhexol with nearly 100 people being prescribed these two. Regarding the prescription of Levodopa, there is some confusion here as far more than 1 person is currently prescribed this drug and furthermore a number are prescribed either as Co-Beneldopa or Co-careldopa. The total number of people on some form of dopamine was 33, thus making it third in this list.

Again the use of Baclofen has been seen as quite controversial with 24 people being prescribed it, but one person since the research was concluded has reported the use of Intrathecal Baclofen usage, which is currently being researched in the USA. This person has had the pump placed in her abdomen in the UK and is currently one of those people running as part of an experimental trial.

PART FIVE : DISCUSSION

CHAPTER 31

Genetics

One of the most rewarding results of this research has been the positive identification of 28.7% of people with definite and proven genetic connections within the dystonic family. Previous research had figures in the region of 7% to 10% (Fletcher, 1990 and Fletcher et al, 1990) therefore this research has shown a possible genetic link in 173 people within this research programme. There is a further possibility of it increasing as more people become involved, therefore one of the strongest reasons for urgent future research programmes must be to test the blood of all people in this research programme to identify who has which dystonic genetic gene involved.

In response to the question " Do you have any family members with dystonia ? " a total of 173 people answered positively. Added to this, there were a further 67 people answering Yes in the 2nd wave and another 25 in the 3rd wave of answers - thus making a total of 265 people affected in this way. 173 people responds to a total of 28.7% of those answering this question.

Reference to Table 4 : Gene Nomenclature for the Dystonias on Page 11 lists the twelve genes thought to be responsible for different forms of dystonia around the world. Although this was not discussed in detail, it does show that this form of research must be given more priority in the future.

Further reference to Table 111 : The spread of dystonic spasms in the region shows a definite expansion in this field. There are 14 people with Oppenheim's Dystonia, 13 with a familial Spasmodic Torticollis, 4 with a familial cervical cranial dystonia and 33 with other forms of familial dystonia, thus totalling 64 people with proven forms of genetic dystonia. Added to these figures there were 7 people with Dopa Responsive Dystonia known to be familial plus a further 5 with myoclonic dystonia also proven to be familial. This totals 76 people with known and proven forms of dystonia within the total dystonic population, thus equating to 8.8% of the 863 people in this chart. It should be noted here that slightly less than 60% of the 937 people in the study answered the question on genetics, therefore the research should take the higher figure of 28.7% as being a more realistic figure.

As more and more research is done in this field, one can anticipate a tremendous growth in the numbers being identified with proven genetic and therefore a familial connection to their dystonia. This must be the route for the immediate future and hopefully will be the subject of future North Eastern research projects.

PART FIVE : DISCUSSION

CHAPTER 32

The Quality of Life

This is one area of research which has been explored more than any other within this research programme with a total of three full papers published on this related subject. The first looked at a Cost-Utility Analysis of Botulinum Toxin therapy and the second specifically measured the benefit of Botulinum Toxin therapy in the treatment of dystonia, whereas the third published in Movement Disorders in 1998 looked at the effect of dystonia and Botulinum Toxin treatment on health-related Quality of Life (HRQoL) and the results from this will be the subject of this chapter.

The background was that all adult participants with dystonia registered between 1993 and 1994 at the Botulinum Toxin Clinic at the North Riding Infirmary in Middlesbrough and the Movement Disorder Clinic at Hunters Moor Regional Rehabilitation Centre in Newcastle were invited to participate in this study. There were 199 patients with dystonia registered at these two clinics at this time. Of these, 25 declined to participate and a further 4 died before the study started. A further 31 people were recruited through The Dystonia Society's newsletter and therefore a final total of 201 people took part in this study and completed a series of questionnaires over the next 6 - 15 months. Five people stayed in remission throughout this period, thus leaving 195 people who represented an overall response rate of 85%. Of these, 102 were currently receiving BT injections whereas 28 formed the non-BT group. A further 66 people either did not fit into these groups or had partial data for less than a full cycle and were denoted as partial. Of these, 7 were receiving BT but their injections were too infrequent, 4 stopped receiving injections as they went into remission, 16 were new patients who were still in the early stages of receiving BT, 32 had a missing questionnaire at either injection or mid-cycle and 7 had missing EuroQol data. Each participant completed two generic questionnaires (the SF36 and EuroQol - see Appendix B) at 4 to 6 weekly intervals over a minimum of 6 months. Respondents also rated their current health status on a 0 to 100 visual analogue scale (VAS).

Dystonia appears to have a considerable effect on HRQoL, affecting physical, social and emotional aspects. Substantial differences on all EuroQol and SF36 dimensions were found between the study participants and general population samples. Participants in the study also rated their own health state (on the EuroQol VAS) approximately 30 points lower than the general population. Although care needs to be taken in making direct comparisons with the general population samples in view of their differing profiles (for instance, 53% of the EuroQol general population were employed and 68% were married or cohabiting), comparisons with respondents of similar age show the same trends. However all of these reflect considerably better HRQoL than that reported by the patients with dystonia.

People with non-focal dystonia differed from those with focal dystonia in physical aspects of HRQoL, : the non-focal group reported significantly more problems with usual activities and more had problems with mobility and self-care. However the two groups reported similar levels of pain and emotional well-being.

These findings are in line with previous findings of significant levels of depression and negative body image in patients with torticollis (Jahanshahi et al, 1988 and 1990). As far as is known, no previous study has assessed broader HRQoL for people with dystonia as a whole. It is also unknown how typical the study participants are of people who have dystonia in Britain, because there are many people with dystonia who do not present for diagnosis or who are misdiagnosed.

Both the EuroQoL and the SF36 performed well in the study. Both were well accepted by participants and there were few missing data on the questionnaires that were returned. Both instruments showed discrimination validity in identifying large differences in study participants and the general population; the EuroQoL also discriminated between the focal and non-focal dystonia groups. The EuroQoL and SF36 also correlated well with each other, providing evidence for construct validity. Perhaps surprisingly for generic instruments both identified expected improvements in HRQoL after the administration of BT, although these findings can only be suggestive because few of these changes were statistically significant.

The results of this particular study show that people with dystonia have a significantly worse HRQoL than a general population and tentatively suggests some of the problems experienced. In view of the considerable impact that dystonia has on HRQoL, it is likely that the demand for BT will continue to grow and this will require further monitoring of the effect of BT on HRQoL.

PART FIVE : DISCUSSION

CHAPTER 33

Social and Economic Implications

Economic Consequences

It has clearly been shown that in both employment and Social Economic Group (SEG) those with dystonia in general are at an economic disadvantage once the onset of the disorder commences, almost regardless from where in the 'social pecking order' they started.

There are some distinct advantages for those fortunate enough to have had Occupational Pensions, but this only applies to those having had to retire early due to their dystonia. The vast majority of dystonia patients eventually find themselves claiming some form of disability benefit, particularly as the disorder progresses over time.

There is evidence however that early diagnosis is leading to earlier treatment and this is being proved to lead to a better early prognosis. Although dystonia also can move through the body and most generalised dystonias start in a limb before progressing throughout the body, most focal dystonias have a later onset date and tend to remain in that part of the body in which they first presented.

It is suggested that evidence from the latest cases being referred to the various clinics shows that the earlier the intervention the better, at least, is the short term prognosis. This is shown particularly with those subjects still in employment. The majority of cases previously diagnosed and referred to Bot. Tox. clinics had had the disorder for many years and by the time the diagnosis had been made correctly, the condition was often so severe as to have forced that person to have given up work, either completely or on long term sick leave.

The evidence shows that once out of the job market because of a disability, it appears to be very difficult to get back into it again. Therefore one of the conclusions of this limited research study is not to leave work unless absolutely essential due to ones dystonia.

There are many examples of where people have regretted having had to give up their previously paid employment. The safety aspects of continuing their work with severe limb and body tremors were often overwhelming. Nevertheless in hindsight, many people should have insisted on re-training to work at a less physically demanding job which might have entailed primarily sitting and talking rather than doing a job entailing constant physical movements. This has been repeated again and again in 473 personal interviews undertaken by the author during the first four years of this research programme.

Social Consequences

It should be remembered that, " *the sense of isolation felt by most sufferers has never been measured and as the disorder is predominantly visual in presentation, a number of sufferers feel severe social and psychological pressure to remain hidden from view*" (Butler, 1996) and " *social isolation in patients breeds a lack of diagnostic skill in G.P.'s which in turn leads to non or misdiagnosis and thus increases the isolation felt by most sufferers.* " (Butler and Duffey, 1996a)

The author is not a qualified psychologist and therefore unable to draw significant conclusions on the results of the psycho-social research data, but nevertheless certain areas can be discussed as the results are apparent for anyone to see.

One of the main areas for discussion could be the so-called 'Vicious Circle Syndrome'. This scenario has come about as a result of 473 personal interviews undertaken by the author. The theory has been primarily publicised by the author, as a result of the fieldwork interviews, during a number of lectures given to dystonic and medical audiences on the subject.

TABLE 114. Vicious Circle Syndrome - The Theory

Event - Beginning of Muscular Spasms	*Result* - Patient lives with it for a while
Event - The Spasms get worse	*Result* - Visits Dr, "it will get better soon"
Event - Anxiety starts / Spasms get worse	*Result* - Visits to Dr become frequent
Event - Depression starts / Spasms worsen	*Result* - Dr suggests "it is all in the mind"
Event - Anxiety and Depression sets in	*Result* - This causes spasms to get worse
Event - Anxiety / Depression / Spasms	*Result* - Dr treats anxiety and depression
Event - Treatment makes Spasms worse	*Result* - Dr treats symptoms, not cause
Event - The Spasms continue to worsen	*Result* - Locked into 'the vicious circle'

Although the evidence for the theory is mainly anecdotal, the author has received overwhelming support from patient groups whenever he has spoken about his theory. Therefore some time should be spent now in examining this theory in greater detail and discovering whether it is in fact a genuine 'new' theory.

However, if one assumes that the scenario for a Spasmodic Torticollis patient can go something like the following, then evidence can be produced to back up the theory similar to the example showed overleaf.

TABLE 115. Vicious Circle Syndrome - The Evidence

1. At first a slight spasm is felt with either a turning or tilting of the head or an intermittent tremor. It does not last very long and goes away. The patient thinks no more about it (473 Fieldwork Interviews).

2. The spasm returns or gradually (usually very gradually) gets worse until eventually the patient decides to go and see his or her (GP) family practitioner (473 Fieldwork Interviews).

3. The doctor is probably unsure of what it is or what is causing it and usually suggests that there is nothing wrong and that it will get better on its own accord (473 Fieldwork Interviews).

4. The spasm gets worse and the visits to the doctor become more frequent and more insistent. The patient naturally gets slightly anxious as no diagnosis is forthcoming from the doctor. There is a great deal of evidence to suggest that anxiety (Jahanshahi and Marsden, 1990b) makes the spasms worse. As the anxiety grows so does the corresponding intensity of spasm. The prognosis of dystonia is generally associated with increased intensity of spasm at some period in the early stages of the disorder (Butler, 1997).

5. 37% of doctors will suggest it is "*all in the mind*", ie it is a psychological or psychogenic disorder. The patient knows that there is something physically wrong, but the doctor (who should know better) is suggesting something mental ? This tends to increase anxiety, often depression, as the lack of diagnosis and effective treatment also sets in (TDS, 1993).

6. Depression, combined with anxiety, is known to intensify muscle spasms, thus the vicious circle is starting. The more the spasms increase without a correct diagnosis, the more that anxiety and depression will increase (Butler, 1997).

7. 32% of doctors will refer dystonia patients to a psychiatrist, although the evidence from the literature review has proven that dystonia is a neurological, not a psychological, disorder of the central nervous system. Thus 68% of patients will be misdiagnosed-diagnosed and are referred to over 5 different consultants before getting a correct diagnosis (TDS, 1993).

8. A large number of subjects in the study were locked into this 'vicious circle'. The author often refers to it as a 'vicious spiral', ever getting worse, ever getting lower in spirit and depression, ever going round and round (Butler, 1997).

What is required is an intervention to break this circle and the evidence from the study is that it actually does not matter what form that intervention takes - in fact any intervention can work, depending on the individual, as shown on the following page below.

TABLE 116. Interventions that break the Circle !

- Contact with others with dystonia
- Education of the medical profession
- Early diagnosis correlating with early treatment
- Early treatment often equating with better prognosis
- Any treatment can have a positive effect on social coping strategies
- Intervention by the Dystonia Counselling Service
- Intervention by the Dystonia Nurse Practitioner

People with dystonia in the North East have been able to fund a Dystonia Counselling Service at both weekly Botulinum Toxin clinics at HMRRC and the NRI. Added to this, they now have a person funded full-time who is available to visit all clinics on a regular basis and supply literature and contacts as required. These services are a direct result of the increased awareness raised as a result of the research projects.

An interesting part of analysing where and how the ESD subjects came to be correctly diagnosed showed that 4.1% of people were not registered at any clinic or hospital and this figure was originally 10.7%, before the research study started in 1993. It could be argued that a figure of over 10% of subjects not being currently registered at any clinic or hospital was because these people had given up on the medical profession's ability to help them in any way. In fact, the words "*given up*", "*doctors can't help*", "*they don't understand*" and "*they don't know about dystonia*" were used on a significant number of occasions (qualitative data - content analysis) during the research.

It has been proved that dystonic patients score over 23.7 points below the general population, based on the SF36 Social Functioning (SF) dimension (Gudex et al, 1997). A number of patients have been measured as severely (clinically) depressed and have been referred for treatment. The Dystonia Nurse Practitioner and the Dystonia Counsellor, who is a qualified Counselling Psychologist, work very closely together and have identified a high number of depressed patients. However, the depression is the result of their dystonia and not the cause of their dystonia, although as has been seen above, there is a strong correlation between dystonia and psychological factors affecting the spasms.

Without exception, those subjects drawing disability allowances do so as a direct result of their dystonia. The incapacity to work can have a devastating effect on any person and it was particular so for the three musicians mentioned in the last chapter, one of whom has Spasmodic Torticollis. This is often the main reason for depression setting in.

A large number of subjects have indicated that they could cope much better with the disorder itself, once they knew what it was and what the prognosis was likely to be.

Finally, the Epidemiological Survey of Dystonia (ESD) has had a tremendous impact on the lives of people with dystonia, not least of which is for people with dystonia trying to claim any one of a number of disability allowances from the UK Benefits Agency. Up until 1996, this agency (nationally) had not even recognised that dystonia was a legitimate neurological disorder and claimants had often to individually go through the process of establishing their condition from scratch.

It was seen as " *an extremely rare condition* " which fluctuated in severity, so that claimants were often told to wait a few weeks as it would go away and they would then not require any form of disability or social security. This research has shown that not to be the case and furthermore has resulted in the Disability Handbook - the reference book for the Benefit Agency, Social Services and many other government agencies - having a section on dystonia included in it for the first time in 1997. This has greatly helped subsequent claimant applications.

PART FIVE : DISCUSSION

CHAPTER 34

Psychological Profiles

The psychological profiles reveal quite a lot about the people who have dystonia. Although there is a broad spectrum of different types of people and with differing levels of dystonia, what seems quite clear is the tremendous spirit which seems to pervade the entire cohort of people with dystonia. The psychological profiles will become the province of Dr Marjan Jahanshahi eventually on which to write a number of research papers, however with the reservations as previously mentioned herewith a discussion of the results.

The fact that a high percentage of patients were found to be suffering from depression (42.4%) with a further 10.7% suffering from anxiety is not really surprising when one remembers the length of time for most people to have been correctly diagnosed Those with serious psychiatric problems were quite small in number and will be examined in detail by Dr Jahanshahi, however it was not surprising that only 12.7% were specifically prescribed anti-depressants and only 35 people said they were for the control of anxiety.

The Living with Dystonia Questionnaire was very helpful in describing those people who were severely, or even moderately, affected by various different events. The highest scores were obtained, not unsurprising, in the physical events, such as running or playing sports, mainly due, one must say, to the average age of the participants, however other high scores were also obtained in continuing to take part in a hobby (90 people), having dinner parties (80), walking (68), going to the theatre (63) or a restaurant or pub (62). There were high scores also obtained in writing (56), crossing roads (55), transport and carrying objects (54) as well as holding a conversation (52). All of this just goes to show how debilitating some of the various forms of dystonia can be.

From the opposite point of view, those activities with the lowest number of people who were not affected at all were also interesting to examine. The lowest numbers were again holding a conversation (100) and having a hobby (101) which goes to show how social interaction seems to be the most debilitating part of dystonia.

The disabling, uncomfortable and disfigurement table on page 91 gives an interesting example of the different levels affecting a high proportion of the 332 people who answered these questions. The fact that the highest level of uncomfortability is the 2nd highest amongst those answering this question is highly significant. The numbers of disfigurement decrease as the level increases, which is interesting but only reflects the different levels of disfigurement amongst the study population.

The Body Concept Scales fit fairly well into the predispositions one would anticipate from the study population. The highest negative scores being in the area of mobility vs immobility and straight vs twisted whereas the highest positive scores were in the fit vs unfit and graceful vs awkward scores.

The Beck Depression Inventory (BDI) is one of the most respected instruments in the field of psychological profiling. A total of 331 people completed the BDI but the results only relate to 329 people. The highest number of people recorded a severe condition against sex (59) and sleep (36). This is the first time that sex has rated so highly and does show that this is an area which should be explored more. Although people are quite reluctant to discuss their sexual problems, it does appear that a relatively high number of people do have to make quite a severe moderation to their sexual drive and instincts.

The Self Esteem Scores showed that the highest number strongly disagreeing was in the section entitled " *All in all, I am inclined to feel that I am a failure* ". which was the highest positive self-esteem figure. The highest number in strongly agree (negative self-esteem) side was 49 people strongly agreeing and 134 agreeing to the statement "*I certainly feel useless at times* ".

The Impact of Dystonia certainly reflected these views in that 113 people strongly disagreed with the statement " *People are often uncomfortable around me because of my illness* " whereas the highest number of people (107) strongly agreed with the statement " *Because of my illness, I miss the things I like to do most.* "

The Acceptance Stages were answered by 318 people and although 81.1% had reached the final acceptance that they had dystonia, 7.2% were still in shock that they had contracted dystonia, 7.9% had moved to anger and 3.8% had moved to despair that they had contracted dystonia. This means that 60 people (18.9%) have not just been diagnosed but have been in either shock, anger or despair for several years. This is very dependant on the individuals own perception of their illness and not upon the severity of that illness. There were several cases of people who were mildly affected having been in stage one for many years whereas others more severely affected having reached final acceptance in a relatively short period of time.

The final questionnaire used in this section was that for the Primary Carer. 221 people answered this questionnaire as 84 people (24.8%) had no primary carer, ie they lived alone and without any support of any description. As previously mentioned a further 25 people declined to answer any questions and another 9 failed to complete the questionnaire. The descriptions of each primary carer is described on page 97. The question with which the highest number (52%) agreed was " *I have had to make emotional adjustments* " whereas the highest question of disagreement was " *I am completely overwhelmed* " with 85.9% disagreeing with this statement. The carer's personal statement quietly reflected this opinion with the highest agreement being " *My partner needs my support* " with almost equal division between " *This disease has changed my life* " (14.0%) and " *This disease has not changed my life.* "

PART FIVE : DISCUSSION

CHAPTER 35

Environmental Factors

Similar to the last chapter, the environmental factors also reveal quite a lot about the people who have dystonia. Starting with the extremities, one can see that although 86.1% use their right and 13.3% use their left hand, 8.3% have changed hands because of their dystonia. This is the first time this has ever been exclusively measured, which also affected the relative questions on hand writing, etc.

The questions on the thyroid gland were added at the end of the research programme due to some patients reporting problems with their thyroids. Although 52 out of 442 (11.8%) indicated they had had previous problems in this area this was not found to be significant, although further research is this area is indicated.

There were a relatively large number of people (15.8%) who felt that their dystonia had been affected or caused by a particular change in their life pattern. However these remained quite small in overall terms and therefore this was not found to be at all statistically significant. However 14.0% thought that trauma or stress had triggered their dystonia, which was almost equalled by those reporting it was triggered by an accident, ie 11.7%. Of the 556 who responded, over 37% stated there was nothing but this does leave over 200 people who thought there was a definite trigger.

Smoking did not gain any significant responses apart from the 39.1% (119 people) who thought their dystonia was affected by other peoples smoke, however 85.2% thought they were affected by gas from gas appliances. This was a tremendously significant figure but unfortunately further research would need to be done to see if this related to their dystonia or was just a general observation.

The same applies to external influences, such as sound, light and drinking water, except that it was significant that over 45% of those answering these questions were affected in some way by light. This particularly affected those with blepharospasm but not exclusively so.

There were however some very significant results on what affected individuals spasms. In particular 84.7% of peoples' spasms being made worse by stress, 75.7% being affected by fatigue, 69.0% by emotion, 68.5% by self-consciousness and 61.8% in social situations was particularly revealing. On the other side, 61.8% of those answering responded well to relaxation and 41.8% felt that sleep helped them. Ignoring any changes taking place during menstruation, the highest percentage remaining unchanged was during the afternoon (69.7%) and morning (67.7%), by distraction (69.4%), by heat (65.2%) and by lying either on the back (63.5%) or on their side (63.4%). Nevertheless, one can see quite clearly that *stress* again plays a very significant factor in dystonic life and this fact should be strongly emphasised and taken significant note of.

There could be no significance reached about the exposure to chemicals measured. Although 57 people experienced this phenomena the numbers were too small to reach any definite conclusions. The same conclusion was also reached regarding the different allergies experienced. 23.5% had persistent fatigue which could be explained as part of their dystonia, being in constant movement, etc.
There was not found to be any definite reaction to food and drink apart from those people who did find that their drinking of coffee definitely affected their dystonia but these numbers were far too small to be at all significant. However perhaps this could be examined to see if this very simple recommendation could lessen the spasms in the dystonic population. Apart from the 51 people who thought that alcohol relaxed their spasms, no significance could be found in this topic.

The whole field of environmental factors thought to affect dystonic spasms was found to be very interesting but no one factor was found to be significant. Of the 205 people who found some particular factor affected them, the highest numbers related to cigarette smoke, new paint, exhaust fumes and pipe, cigars and cigarettes. These did not seem to relate just to people with dystonia and they also affect the general population as well. However this part of the research should be expanded and developed to see if there are any clues to dystonic reactions to any of these factors.

The analysis of the conduct of the research was undertaken as planned and it was found that the majority of those taking part found the experience not unpleasant (73.8%)although 26.2% found some points of constructive criticism. What was more significant was the responses reacting to outside stimuli. There were a number of good and bad responses which should be noted.

The fact that 28.0% of the 346 people responding found that their dystonia was better when there was no stress, with sun and warmth, with positive thoughts, when they were busy and after drinking alcohol, etc was very interesting and gives some positive guidelines for future recommendations. Equally helpful was the fact that only 14.7% found no definite pattern, etc. The majority were able to point to a number of highly significant things that made their spasms worse, such as activity, when tired, missing their injections, with worry, etc - all of which should be mentioned in future booklets, etc on dystonia.

Although the majority of these points are self evident, they should be mentioned as being taken from the largest survey of dystonic patients yet undertaken and might possibly help future dystonia patients in their own personal struggle.

PART FIVE : DISCUSSION

CHAPTER 36

Conclusion

This research has taken exactly 6 years to complete and is a comprehensive and longitudinal study of the epidemiological, social, economic and psychological implications of dystonia within the population of the North East of England.

It was known in 1993 that :-

- dystonia had been historically extremely difficult to diagnosis,
- which had meant that it had previously been very difficult to obtain sufficient numbers for research studies,
- and this had created a number of significant social and economic consequences.

However between May 1993 and May 1999, it has been shown and proven that :-

- dystonia is far more prevalent than previously thought,
- which, with the advent of Botulinum Toxin therapy, is treatable in a large number of cases,
- and this has also shown that any form of intervention has a positive effect on the patient.

Early diagnosis resulting in early treatment of most dystonic conditions has resulted in better and more effective results. Treatment of any kind has a definite positive effect on social coping strategies and this has been seen again and again as a direct result of the increased activity in the region. Due to a combined effort between the local people with dystonia and the medical professionals, a number of unique developments have taken place in the North East of England as a direct result of this research and its associated spin-offs.

An Outreach Dystonia Nurse Practitioner has been funded as part of a previous research programme to administer Botulinum Toxin therapy in the patients' own homes and the effects and benefits were measured over a two year period and compared to a group of matched patients who continued to receive therapy at their local clinic. This post has since been fully funded as part of NHS mainstream clinic and out patient care.

All of this increased awareness has seen the numbers of people with dystonia receiving Botulinum Toxin therapy steadily increase over the past six years at an almost constant rate of nearly 2.6 new referrals per week. Because there is a strong correlation between early onset meaning more severe the eventual condition and late onset resulting in a more focal dystonia, prognosis can now be better explained, and planned for, than in the past. This research project has been extremely difficult to complete at times but nevertheless has produced an enormous amount of data, not least of which has been the new material, never before collated on dystonia patients.

This research has been both a rewarding and yet a very exacting experience. On the negative side and in the light of this experience, a number of different issues would have been started at different times and indeed the entire process would have been planned differently. However this is taken as being part of the learning curve and is not intended as a severe criticism of this research study.

On reflection, although the entire research study was not intended as such, the limited number of controls used in the Cost Utility Analysis on the effectiveness of the use of Botulinum Toxin therapy should have been continued beyond the end of that study. Although there are a very small number of controls in the overall study, they were too small in number to have any beneficial effect on the study as a whole. Nevertheless, they were important in that they were able to focus the input onto what was important and enabled a number of different issues to be explored. For example, although the total numbers were relatively small, the whole issue of the effects of environmental factors on various people's dystonia was an important issue and well worth exploring. It is envisaged that further research into this particular aspect of dystonia could reveal some rewarding and interesting results, which could have further impact of the future treatment of this disease.

Furthermore, it was realised by the end of the study how much more effective it would have been if the numbers had been increased over those that actually completed all the research papers. This was as much because of external factors as much as lack of commitment. The entire study was due to be finished within five years of the research starting and therefore the numbers of people completing all the research papers had been completed after the end of year four or after 500 plus people had been effectively identified. Due to the writer's sudden illness at the end of 1998, this process was extended until September 1999 when this thesis was finally completed. However, on reflection those joining the Epidemiology towards the end of 1998 and throughout 1999 should have been invited to complete the research questionnaires. This has since been done but the results do not form any part of this research thesis.

It is intended once the whole process of completing this thesis has been achieved to write a book based on the evidence of the research completed to date, including the results from the additional questionnaires issued and returned. Anecdotally, the evidence from these additional questionnaires does not appear to contradict any of the evidence gained from the previous research papers, they merely add to the evidence already produced and detailed in the enclosed.

DYSTONIA

A comprehensive and longitudinal study of the epidemiological, social, economic and psychological implications of dystonia within the population of the North East of England

PART SIX :

APPENDICES

APPENDIX A

ACKNOWLEDGEMENTS

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- **Athena Neurosciences (UK) Ltd**
- **REMEDI - Medical Trust**
- **ENTER - Ear, Nose, Throat and Eye Research, NRI, Middlesbrough**
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- North Riding Infirmary - ENT Department, Pharmacy and Medical Records
- Hunters Moor Hospital - Movement Disorder Clinic and Clerical Staff
- Sunderland Eye Infirmary - Botulinum Toxin Clinic
- Darlington Memorial Hospital - Neurology Clinic and Medical Records
- Sunderland General Hospital - Neurology Department
- Dryburn Hospital, Durham - Neurology Clinic
- Middlesbrough General Hospital - Neurology Department and Medical Records

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And finally, although I am told this is not usual for a PhD thesis, I would like to thank my wife for just being there but more particularly for being the inspiration as to why I chose dystonia for my topic and for her support over the six years which have been at times very difficult and exasperating, but so tremendously enjoyable and rewarding.

APPENDIX B

INSTRUMENTS

Actual working copies of a complete set of questionnaires have been included in this section.

Please note these were often produced as double sided sheets of A3 and are all individually numbered pages - ie they are not in sequence numbering.

SPSS Code	Name or purpose of Questionnaire	No of pages
Code 10 :	ESD : Quality of Life - Health State Questionnaire	8
Code 20 :	Demographic and Dystonic Profile Questionnaire	
	Postal Version - Questions sheet	4
	Postal Version - Answers sheet	2
Code 21 :	TDS Questionnaire	incl.
Code 25 :	Demographic Changes Questionnaire	1
Code 30 :	Clinical Profile of Dystonia Questionnaire	6
Code 40 :	Torticollis Questionnaire (attached to Q're 30 as pages 7 and 8)	2
Code 50 :	Living with Dystonia Questionnaire : A Psychological Profile	8
Code 60 :	Impact of Dystonia Questionnaire	2
Code 70 :	Primary Carer's Questionnaire	2
Code 80 :	Environmental Questionnaire	8
Code 81 :	Diagnostic Questionnaire	1
Code 91 :	Dystonia Nurse Practitioner Evaluation - 1st Questionnaire	1
Code 92 :	Dystonia Nurse Practitioner Evaluation - 2nd Questionnaire	1
Code 93 :	Dystonia Nurse Practitioner Evaluation - 3rd Questionnaire	1
Code 94 :	Dystonia Nurse Practitioner Evaluation - Final Questionnaire	1

A RESEARCH STUDY INTO THE EPIDEMIOLOGY OF DYSTONIA
The answers will help to measure your present health state

00) Completed and returned on ..... (date) Q. No | 10 |

Please indicate which statements best describe your own health state <u>today</u>. **Please tick one circle ( ) in each group below.**

For office use

01) <u>Mobility</u>

: I have no problems in walking about ( )

: I have some problems in walking about ( )

: I am confined to bed ( )

02) <u>Self-Care</u>

: I have no problems with self-care ( )

: I have some problems washing or dressing myself ( )

: I am unable to wash or dress myself ( )

03) <u>Usual Activities</u>

: I have no problems with performing my usual activities (work, housework, family, leisure) ( )

: I have some problems with performing my usual activities ( )

: I am unable to perform my usual activities ( )

04) <u>Pain / Discomfort</u>

: I have no pain or discomfort ( )

: I have moderate pain or discomfort ( )

: I have extreme pain or discomfort ( )

05) <u>Anxiety / Depression</u>

: I am not anxious or depressed ( )

: I am moderately anxious or depressed ( )

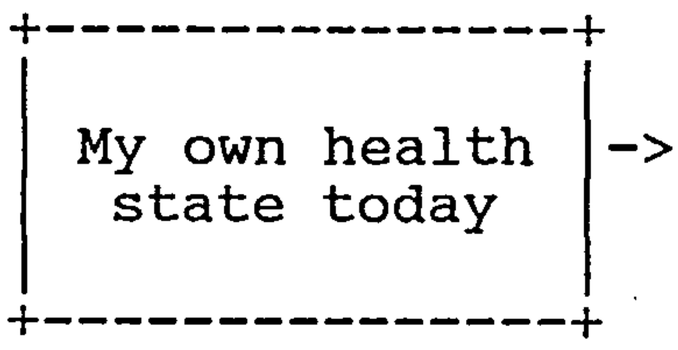
: I am extremely anxious or depressed ( )

06) Health State

To help people say how good or bad a health state is, we have drawn a scale (rather like a thermometer) on which the best state you can imagine is marked by 100 and the worst state you can imagine is marked by 0.

We would like you to indicate on this scale how good or bad is your own health **today**, in your opinion.

Please do this by drawing a line from the box below to whichever point on the scale indicates how good or bad your current health state is.



For office use

HEALTH STATUS QUESTIONNAIRE (SF-36)

The following questions ask for your views about your health, how you feel and how well you are able to do your usual activities. If you are unsure about how to answer any question, please give the best answer you can and make any comments in the space at the end.

Please tick one circle ( ) in each question

07) In general would you say your health is :

	Excellent	( )
	Very good	( )
	Good	( )
	Fair	( )
	Poor	( )

08) <u>Compared to one year ago</u>, how would you rate your health in general <u>now</u> ?

	Much better now than one year ago	( )
	Somewhat better now than one year ago	( )
	About the same	( )
	Somewhat worse now than one year ago	( )
	Much worse now than one year ago	( )

For office use

HEALTH AND DAILY ACTIVITIES

09) The following questions are about activities you might do during a typical day. Does your health limit you in these activities ? If so, how much ?

	Please tick one circle ( ) on each line Yes, limited a lot	Yes, limited a little	No, not limited at all	
a. Vigorous activities, such as running, lifting heavy objects, strenuous sports	( )	( )	( )	
b. Moderate activities, such as moving a table, pushing a vacuum cleaner, bowling	( )	( )	( )	
c. Lifting/carrying groceries	( )	( )	( )	
d. Climbing several flights of stairs	( )	( )	( )	
e. Climbing one flight stairs	( )	( )	( )	
f. Bending, kneeling, stooping	( )	( )	( )	
g. Walking more than a mile	( )	( )	( )	
h. Walking half a mile	( )	( )	( )	
i. Walking 100 yards	( )	( )	( )	
j. Bathing/dressing yourself	( )	( )	( )	

10) During the past 4 weeks, have you had any of the following problems with your work or your other regular daily activities as a result of your health ?

Answer Yes or No to each question	YES	NO	
a. Cut down on the amount of time you spent on work or other activities	( )	( )	
b. Accomplished less than you would like	( )	( )	
c. Were limited in the kind of work or other activities	( )	( )	
d. Had difficulty performing the work or other activities (eg it took extra effort)	( )	( )	

For office use

11) During the <u>past 4 weeks</u>, have you had any of the following problems with your work or your regular daily activities <u>as a result of any emotional problems</u> (such as feeling depressed or anxious) ?

Answer Yes or No to each question	YES	NO
a. Cut down on the <u>amount of time</u> you spent on work or other activities	( )	( )
b. <u>Accomplished less</u> than you would like	( )	( )
c. Didn't do work or other activities as <u>carefully</u> as usual	( )	( )

12) During the <u>past 4 weeks</u>, to what extent has your physical health or emotional problems interfered with family, friends, neighbours or groups ?

Please tick one circle ( ) only

- Not at all ( )
- Slightly ( )
- Moderately ( )
- Quite a bit ( )
- Extremely ( )

13) How much <u>bodily</u> pain have you had during the <u>past 4 weeks</u> ?

- None ( )
- Very mild ( )
- Mild ( )
- Moderate ( )
- Severe ( )
- Very severe ( )

14) During the <u>past 4 weeks</u>, how much did <u>pain</u> interfere with your normal work both outside and outside home

- Not at all ( )
- A little bit ( )
- Moderately ( )
- Quite a bit ( )
- Extremely ( )

For office use

YOUR FEELINGS

15) These questions are about how you feel and how things have been with you <u>during the past month</u>. (For each question, please indicate the one answer that comes closest to the way you have been feeling)

Please tick one circle ( ) on each line

How much time during the past month :	All of the time	Most of the time	A good bit of time	Some of the time	A bit of the time	None of the time	
a. Did you feel full of life	( )	( )	( )	( )	( )	( )	
b. Have you been very nervous	( )	( )	( )	( )	( )	( )	
c. Have you felt so down in the dumps that nothing could cheer you up ?	( )	( )	( )	( )	( )	( )	
d. Have you felt calm and peaceful ?	( )	( )	( )	( )	( )	( )	
e. Did you have a lot of energy ?	( )	( )	( )	( )	( )	( )	
f. Have you felt downhearted and low ?	( )	( )	( )	( )	( )	( )	
g. Did you feel worn out ?	( )	( )	( )	( )	( )	( )	
h. Have you been a happy person	( )	( )	( )	( )	( )	( )	
i. Did you feel tired ?	( )	( )	( )	( )	( )	( )	
j. Has your health limited your normal social activities (eg visiting)	( )	( )	( )	( )	( )	( )	

For office use

HEALTH IN GENERAL

16) Please choose a statement that best describes how <u>true</u> or <u>false</u> each of the following statements is for you.

	Definitely true	Mostly true	Not sure	Most false	Definitely false	
	Please tick one circle ( ) on each line					
a. I seem to get ill more easily than other people	( )	( )	( )	( )	( )	+-+ | | +-+
b. I am as healthy as anybody I know	( )	( )	( )	( )	( )	+-+ | | +-+
c. I expect my health to get worse	( )	( )	( )	( )	( )	+-+ | | +-+
d. My health is excellent	( )	( )	( )	( )	( )	+-+ | | +-+

The following questions are a bit more personal. They ask you about how you feel and how things have been with you **during the past week.** Please feel free to give as much information as you would like, use extra paper if needed.

17) If you have a partner or spouse, how has your relationship been with them, during the past week ? +-+ | | +-+

18) & how have your relationships been with other people? +-+ | | +-+

For office use

During the past week.....

19) Have you felt a burden to other people ? +-+ | | +-+

20) Have you felt embarrassment when with other people ? +-+ | | +-+

21) How have you felt about your appearance in general ? +-+ | | +-+

22) Have you felt that you lack control over your body ? +-+ | | +-+

In general.....

23) How do you feel you are coping with life ? +-+ | | +-+

24) Do you feel you are receiving enough support from family and friends ? +-+ | | +-+

Demographic Questionnaire - Questions Sheet

PLEASE COMPLETE THESE QUESTIONS BY WRITING YOUR ANSWERS ON THE ANSWERS SHEET AND SEND IT BACK TO ME IN THE S.A.E. PROVIDED.

00) Today's date is ? The time is ? I will have already filled in your Personal Questionnaire Code Number for you.

PLEASE NOTE : IF ANY QUESTION DOES NOT APPLY TO YOU, PLEASE WRITE "N/A" IN THE SPACE PROVIDED. IF YOU NEED ADDITIONAL SPACE TO ANSWER THE QUESTIONS, WRITE THE QUESTION NUMBER AND ANSWER ON A SEPARATE PIECE OF PAPER AND ATTACH TO THE ANSWER SHEET.

Firstly some questions about you personally

01) Are you male or female ?

02) What is your date of birth ?

03) What is the full postal code of your home address ?

04) What is your current marital status ?

Prompt : Are you single, married, cohabiting, separated, divorced or widowed ?

05) Do you have any children, what is their sex, year of birth of eldest and youngest and which still live with you ?

Next some questions about work

06) What is your current working situation ?

Are you in full time employment, part time employment, unemployed, self employed, unwaged (ie a housewife, student, etc), retired, or retired through ill health ?

07) What is or was your actual occupation ?

08) What is the occupation of the head of your household, if you are not that person or it is different from your own ?

09) What is your personal current annual income calculated to the nearest hundred pounds per annum ?

10) Do you receive currently any benefits or pension and if so, what are they ?

Prompt : Do you receive sickness benefit, disability allowance, attendance allowance, mobility allowance, occupational pension, retirement pension, any combination of these, no benefits at all ?

11) How long in hours, do you have to take off work to go to the hospital you normally attend for dystonia treatment ?

12) If somebody goes with you to the hospital, do they have to take time off work and if so, how long in hours ?

13) How much time have you had to have off work (in number of days) as a result of your dystonia in the last 6 weeks ?

14) How much time has your spouse or partner had to have off work (in number of days) as a result of your dystonia in the last 6 weeks ?

Now some questions about travelling

15) How many miles (one way only) do you travel to go to the hospital you normally attend for treatment for dystonia ?

16) How long does it take you, in number of minutes ?

17) What form of transport do you use ?

Prompt : Do you travel in your own car, a member of your family's car, a friend's car, by public transport (ie bus and/or train), by other private transport (ie in a taxi or mini-cab), by bicycle, walk or by some other form ?

Next some questions about Dystonia

18) Which type or types of dystonia do you have and which parts of the body does it affect ?

Torticollis / Blepharospasm / Oromandibular / Cranial / Spasmodic Dysphonia / Writer's Cramp / Focal Dystonia / Generalised Dystonia / + Any Other Neurological Disorder

19) In what year, did you first notice your dystonic symptoms?

20) In what year, were you first diagnosed as having dystonia? Please write below this answer if there was any particular event which happened just before your first symptoms appeared, which could be a <u>trigger</u> for the start of them.

21) Are you aware of any other family members who have displayed symptoms of muscle spasm or contractions ? Who are they, what relationship are they to you and, if you know, what type of dystonia do or did they have ?

22) Do you currently take any regular medication for your dystonia, what is the name of the drug (s), dosage and, if so, who pays for this medication ? Is it paid for by yourself privately or by prescription, by your G.P. as part of a prescription, by the hospital without charge to you or by other source ?

23) How much does this medication cost per dose ?

Prompt : Do you know the cost in pounds ? Do not worry if you do not, as long as we know the name of the medication, we can find out its cost later.

24) Have you had any previous treatment for Dystonia, before the current treatment, and if so, what types of treatment were they ?

Prompt : Have you previously been prescribed other drugs and what were their names, have you had surgery for dystonia and what type, have you been seen by a psychiatrist or had psychotherapy, have you receive any allergy or physiotherapy treatment, etc ?

25) Did any of these treatments cause any side effects and if so, how long did they last ?

26) Do you take any regular medication for anything else ?

Prompt : If yes, what for and what is the medication ?

27) If yes, who pays for this medication ?

Prompt : Is it paid for by yourself or family privately or by prescription, by your G.P. as part of a prescription, by the hospital without charge to you or by other source ?

28) How much does this medication cost per dose ?

Prompt : Do you know the cost in pounds ? Do not worry if you do not, as long as we know the name of the medication, we can find out its cost later.

29) Have you had Botulinum Toxin injections before today, how many times, where in your body and if there have been any side effects, what are they and how long do they last ?

E = Finally some questions about the social aspect of Dystonia

30) Have you ever talked to other people <u>with</u> dystonia ?

Prompt : Never, rarely, occasionally, often, frequently.

31) Under what circumstances do you most commonly talk to other people <u>with</u> dystonia ?

Prompt : Do you talk to people with dystonia at the clinic, during stays in hospital, at home socially, at meetings of the Dystonia Society, I have never talked to any other person <u>with</u> dystonia ?

32) Do you think that some form of counselling, other than strictly medical, should be provided either when you are first diagnosed or on a regular basis or when you want that service and by whom should this counselling be given?

33) Were you aware, before you received your Doctor's letter about this study, that there was a charity dedicated to helping people with Dystonia ? And if so, what was the source of this original information about TDS ?

Prompt : Was it another member, relative or friend, T.V. or radio, newspaper or magazine, doctor or hospital worker, social worker, other source , or can't remember ?

34) Are you a member of the Dystonia Society and if so, in what year did you join ?

35) If you are a member, why did you join originally ?

Prompt : To find out more about the disorder, to meet and talk to others, to read their literature, to attend their meetings, some other reason ?

36) If you are a member, what particular aspect of TDS's activities do you find most useful ?

Prompt : Is the the quarterly newsletter, information in general, being able to get answers to your questions, the literature, self-help group activities, or other reasons ?

37) Are you a member of a local Self-Help Group, if so, which one and what do you get most from this affiliation ?

Prompt : A feeling of belonging, sharing the experience, talking to others, social contact, other reasons ?

38) If you receive the Society's quarterly Newsletter, what aspect of this publication, do you find most interesting or useful ?

Prompt : The medical articles, the information about the latest research, the letters and experiences from other members, the contact addresses, the news from other areas and groups, the overseas information, or all of it ?

39) What would you want primarily from the Dystonia Society in general and a local self-help group in particular ?

THANK YOU FOR ANSWERING THESE QUESTIONS. I HOPE THAT WAS NOT TOO TEDIOUS - WE WILL ASK YOU TO COMPLETE THIS QUESTIONNAIRE AGAIN AT THE END OF THE STUDY TO SEE IF THERE HAVE BEEN ANY CHANGES IN YOUR CIRCUMSTANCES.

Demographic Questionnaire - Answers Sheet

ANSWER SHEET TO BE USED IN CONJUNCTION WITH QUESTIONS SHEET

#)	Questions	Write Answers Here	Office
00)	Date	Time ..... Questionnaire No	
01)	Male / Female	.....	
02)	Date of Birth	.....	
03)	Home Post Code	.....	
04)	Marital Status	.....	
05)	No of child/gender	.....	
	Still live with you	.....	
	Eldest Birth Year	.....	
	Youngest Birth Year	.....	
06)	Current Work Sit.	.....	
07)	Occupation of pat.	.....	
08)	Occ. Head of House	.....	
09)	Current Ann Income	.....	
10)	Benefits received	.....	
11)	Off work, hospital	.....	
12)	Off work, helper	.....	
13)	How long off work	.....	
14)	Off work, partner	.....	
15)	How many miles	.....	
16)	How long in minutes	.....	
17)	Form of transport	.....	
18)	Type of Dystonia	.....	
19)	Year : symptoms	.....	
20)	Year : diagnosed	.....	

21) Any in your family .....

Relationship to you .....

Type(s) of Dystonia .....

22) Current medication .....

Who pays .....

23) Cost per dose .....

24) Previous Treatments .....

(before current) .....

25) Any Side Effects .....

how long lasts .....

26) Other Medications .....

27) Who pays .....

28) Cost per dose .....

29) Bot Tox before .....

Where in your body .....

Any Side effects .....

How long lasts .....

30) Talked to others .....

31) What circumstances .....

32) Counselling .....

33) Heard of TDS where .....

34) Member of TDS, when .....

35) Why did you join .....

36) Most useful aspect .....

37) Self-help group .....

38) TDS Newsletter .....

39) Primary function .....

FINAL DEMOGRAPHIC QUESTIONNAIRE

Please answer the following questions in the space provided :

#)	Questions	Write or Circle Answers	Office
00)	Today's Date is :	Questionnaire No	25
01)	Has your current working situation changed since the start of the study ?	YES NO	
	If YES, How has it changed ?		
	.....		
02)	Has your personal current annual income changed ?	YES NO	
	If YES, what is your current annual income ?		
	.....		
03)	Do you <u>now</u> receive any benefits or pension which you did not receive when first interviewed ?	YES NO	
	If YES, what are the <u>new</u> benefits or pension ?		
	.....		
04)	Have you had to take time off work in the past 6 weeks because of your dystonia ?	YES NO	
	If YES, how much time ?		
05)	Has your spouse spouse or partner had to have off work because of your dystonia in the last 6 weeks?	YES NO	
	If YES, how much time ?		
06)	Have you changed hospital since the study started?	YES NO	
	If YES, how far is the hospital in miles ?		
	How long does it take you to get there ?		
	and by what method do you travel there ?		

QUESTIONNAIRE ON THE CLINICAL FEATURES OF DYSTONIA

The purpose of this questionnaire is to obtain information about the clinical features of your dystonia, that is the specific symptoms that you personally have. Other questions relate to your experiences of the medical services and the type of accommodation you live in. Throughout the questionnaire, when the word "dystonia" is mentioned , please assume it also means torticollis, blepharospasm, dysphonia, writer's cramp, hand or leg dystonia and secondary dystonic movements eg hemi-facial spasm, as well as secondary dystonias caused by other neurological disorders eg, Stroke, Multiple Sclerosis, Cerebral Palsy, Parkinsonism, etc.

#) Questions	Write Answers Here	Office
00) Today's Date is	Your code number is	30

Contact with Medical Services

01) Please give the date when your dystonic spasms first started : .....

02) Please give the date when you were first told by a doctor that you had dystonia (or another type of neurological disorder) : .....

03) If there was a delay between the start of your dystonia and its diagnosis (being told by a doctor that you had dystonia) what was this delay due to ?

.....

.....

04) Who first diagnosed your dystonia or other neurological disorder ? Tick one

1. G.P. .....
2. Hospital Doctor / Neurologist .....
3. Privately consulted Doctor / Neurologist .....
4. Psychiatrist .....
5. Other, please specify who : .....

05) Were there any major events before the start of your dystonia, that you think triggered its onset, such as an accident, fall or injury, illness, stress, loss of a relative, etc ?

YES NO

If YES, please give details, including dates :

.....

06) What do you think caused your dystonia ?

Office

.....

.....

07) Were you ever (wrongly) told by a doctor that your dystonia was psychological ? Circle answer YES NO

08) Do you feel your G.P. knows enough about dystonia ? YES NO

09) Below is a list of treatments. Please indicate which treatments you have personally had for your dystonia by ticking the appropriate column. For those treatments that you have had, please give the date you started the treatment and specify whether you found it helpful in controlling your particular type of dystonia (or muscle spasm) :

Treatment	Date when started	Effect of treatment Better	Unchanged	Worse
01 Medication / Drugs				
02 Surgery				
03 Physiotherapy				
04 Osteopathy				
05 Chiropractic				
06 Biofeedback				
07 Counselling				
08 Psychotherapy				
09 Acupuncture				
10 Yoga				
11 Meditation				
12 Hypnotherapy				
13 The Alexander Technique				
14 Other, please specify which:				

#) Questions	Circle answer	Office

10) Have you suffered from any major physical illness that has required treatment or hospitalisation ?

YES NO

If YES, please give details :

Type of physical illness	Date started
1.	
2.	
3.	
4.	

11) Have you suffered from any major psychiatric/psychological problems that have required treatment or hospitalisation ? YES NO

If YES, please give details :

Type of psychiatric / psychological problem	Date of onset	Treatment received
1.		
2.		
3.		
4.		

12) Are you currently taking any antidepressant medication ? YES NO

13) Are you currently taking any medication for the control of anxiety ? YES NO

<u>Present clinical features of your dystonia</u>

14) Please indicate (on the page overleaf) the parts of your body that are affected by dystonia, (that is muscle cramps, abnormal postures or involuntary movements) and give the date, as near as you can remember, when it started in each part of your body :

Tick part of body affected by dystonia. Date started

1. Eyelids (right, left or both)* ....
2. Face (right, left or both)* ....
3. Mouth / jaw / tongue* ....
4. Vocal chords, speech* ....
5. Right arm / hand* ....
6. Left arm / hand* ....
7. Writing hand (R or L)* ....

   Is this only during writing ? YES NO

8. Trunk (body) ....
9. Right leg / foot* ....
10. Left leg / foot* ....

(* specify which or delete as appropriate)

15) In addition to the muscle spasms, do you have involuntary movements of the affected body part ?

YES NO

If YES, please tick the answer(s) that best describe the type of involuntary movements

1. Regular trembling .....
2. Quick jerking .....
3. Slight twitching .....
4. Slow pulling / turning .....
5. None of the above, please specify .....

16) Please indicate how severe you think your dystonia is at present (compared to others with dystonia that you may have seen or compared to how it has been in the past) by circling the appropriate number on the scale below :

Not severe at all — Very severe

0 1 2 3 4 5 6 7 8 9 10

17) Please indicate the degree of control you presently have over your **muscle spasms**, by circling the appropriate number on the scale below :

Office

No control at all — Complete control

0 1 2 3 4 5 6 7 8 9 10

18) Please indicate the degree of control you presently have over the **involuntary movement / abnormal postures** of your affected body part(s), by circling the appropriate number on the scale :

No control at all — Complete control

0 1 2 3 4 5 6 7 8 9 10

19) At present, do you have any pain as a result of your **muscle spasms** ?

YES NO

If YES, please specify in which parts of your body you feel pain :

.....

.....

If YES, please indicate the severity of this pain, by circling the appropriate number on the scale below :

No pain — Very Severe pain

0 1 2 3 4 5 6 7 8 9 10

If YES, please indicate how frequently you experience this pain in the particular area(s), by ticking one of the categories below :

Infrequently .....
Often .....
Continuously .....

20) Since the start of your dystonia, have you had periods of substantial **spontaneous improvement or recovery**, which was <u>not</u> related to any particular form of treatment ?

YES NO

Office

If YES, please give details :-

Date of spontaneous improvement/recovery	How long did it last ?	Was it partial or complete ?
1.		
2.		
3.		

<u>**Nature of your Accommodation**</u>

21) Who do you live with ? Tick one answer

1. Alone .....
2. With husband / wife .....
3. With parents .....
4. With children .....
5. With sister or brother .....
6. With other relative, specify relationship : .....
7. Other, please specify : .....

22) What type of accommodation do you live in ? Tick one answer

1. House .....
2. Flat .....

23) Is the accommodation suitable for the problems caused by your dystonia ? YES NO

If NO, please specify the major ways in which your flat / house does <u>not</u> meet the needs of your type of dystonia :

.....

.....

.....

.....

QUESTIONNAIRE ON TORTICOLLIS

The following questions should only be answered by those of you who have a **torticollis, antecollis or retrocollis.** The purpose of this questionnaire is to be able to link the findings from our North East research to an on-going survey being conducted into Torticollis by Dr Marjan Jahanshahi at the Institute of Neurology in London.

Office 40

24) Please indicate the **main position** that your head / chin adopts at present by ticking the appropriate category :

1. Chin turns to the side .....
2. Ear tilts sideways towards shoulder .....
3. Head bends forward (chin pointing down) .....
4. Heads bends backwards (chin pointing up) .....

25) Please indicate the main direction towards which your head / chin turns or tilts at present, by ticking the appropriate category :

1. Right turn / tilt * .....
2. Left turn / tilt * .....
3. Backwards (chin pointing up) .....
4. Forwards only (chin pointing down) .....
5. Right and backwards .....
6. Right and forwards .....
7. Left and backwards .....
8. Left and forwards .....

* delete as appropriate

Geste Antagoniste

26) Since the start of your torticollis, have you ever had a 'trick' method or gesture (so-called geste antagoniste) to keep your head straight and upright in the body midline position (eg: touching the left side of your chin with your right hand) ?

YES NO

If YES, is this gesture still effective in keeping your head straight ?

1. YES, still effective

2. NO, no longer effective. Please specify how many years after the start of your torticollis the geste remained effective .....

27) Describe in detail (in terms of hand used, area of head, neck or face touched), the gesture that you use(d) to keep your head straight: Perform this gesture in front of a mirror before answering and write your answers on a separate piece of paper :

Office

+----+
| |
+----+

28) While performing this gesture to keep your head straight, which hand do you use ?

1. Right hand ..... 2. Left hand .....

+----+
| |
+----+

29) While performing this gesture to keep your head straight which statement is most true in your case

1. The area of my head/face/neck that I touch is in the <u>front</u> of my body. .....

2. The area of my head/face/neck that I touch is in the <u>back</u> of my body. .....

+----+
| |
+----+

30) While performing this gesture to keep your head straight which statement is most true in your case

1. I just <u>lightly touch</u> my head / face / neck to keep my head straight .....

2. I have to <u>pull hard</u> on my head / face / neck to keep my head straight .....

3. I have to <u>push hard</u> on my head / face / neck to keep my head straight .....

+----+
| |
+----+

31) While performing this gesture to keep your head straight which statement is most true in your case

1. My head starts moving towards the body midline position <u>before</u> my hand touches my head / face neck .....

2. My head starts moving towards the body midline position <u>at the same time</u> as my hand touches my head / face neck .....

3. My head starts moving towards the body midline position <u>after</u> my hand touches my head / face neck .....

+----+
| |
+----+

32) Which statement best describes your torticollis :

1. I feel that my head is being <u>pulled</u> to the abnormal position .....

2. I feel that my head is being <u>pushed</u> to the abnormal position .....

+----+
| |
+----+

QUESTIONNAIRE ON "LIVING WITH DYSTONIA"

The purpose of this questionnaire is to find out how you feel about yourself and how dystonia has affected your life. Our findings will be linked to an on-going survey being conducted at the Institute of Neurology in London. When "dystonia" is mentioned during this questionnaire, assume it also means torticollis, blepharospasm, dysphonia, writer's cramp and secondary dystonic movements eg hemi-facial spasm, as well as secondary dystonias caused by other neurological disorders eg, Stroke, Multiple Sclerosis, Cerebral Palsy, Parkinsonism, etc.

#) Questions — Write Answers Here — Office

00) Today's Date is ..... Your code number is 50

Below are a list of activities. Please indicate the extent to which your dystonia affects your engagement in, performance or enjoyment of these activities <u>at the present time</u>. Answer each question by circling the appropriate number :

If you have never driven , your answer to question 14, ie driving, should be: Not applicable = 0
Others vary from Not at all affected = 1
to Mildly affected = 2
to Moderately affected = 3
to Severely affected = 4

1.	Dressing/undressing yourself	0	1	2	3	4
2.	Doing housework (vacuuming,ironing)	0	1	2	3	4
3.	Watching television	0	1	2	3	4
4.	Running	0	1	2	3	4
5.	Use of public transport	0	1	2	3	4
6.	Writing	0	1	2	3	4
7.	Have a face-to-face conversation	0	1	2	3	4
8.	Carrying objects	0	1	2	3	4
9.	Going to restaurants or pubs	0	1	2	3	4
10.	Brushing teeth	0	1	2	3	4
11.	Reading	0	1	2	3	4
12.	Walking	0	1	2	3	4

(cont)

						Office
13. Having sexual intercourse	0	1	2	3	4	
14. Driving a car	0	1	2	3	4	
15. Washing face	0	1	2	3	4	
16. Eating, using knife and fork	0	1	2	3	4	
17. Going to or giving dinner parties	0	1	2	3	4	
18. Typing	0	1	2	3	4	
19. Engagement in hobbies (knitting, sewing, carpentry, gardening, etc)	0	1	2	3	4	
20. Crossing roads	0	1	2	3	4	
21. Shaving face, if male, and putting on make up, if female	0	1	2	3	4	
22. Drinking from a cup	0	1	2	3	4	
23. Riding a bicycle	0	1	2	3	4	
24. Going to the theatre/cinema/concert	0	1	2	3	4	
25. Activities requiring visual/manual coordination, such as pouring tea or using a screwdriver	0	1	2	3	4	
26. Engagement in sports (tennis, squash, jogging, swimming, golf, table tennis, etc)	0	1	2	3	4	
27. Walking up or down stairs	0	1	2	3	4	

Please indicate how <u>disabling</u> your dystonia is in terms of interfering with your daily activities <u>at present</u>, by circling the appropriate number on the scale below:

Not at all disabling — Very disabling

0 1 2 3 4 5 6 7 8 9 10

Please indicate how <u>uncomfortable</u> you feel in <u>social situations</u> because of your dystonia <u>at present</u> :

Not at all uncomfortable — Very uncomfortable

0 1 2 3 4 5 6 7 8 9 10

On page 4 (overleaf) is a list of adjectives that can describe the physical body. The adjectives at each end of a scale are opposites. Please place a cross on each scale at the point that best describes your own body :

Here is an **example** to show you the different ways to mark X :-

If you feel that your body is **very well described** by the adjective at the end of the scale, place a cross as follows :

Strong ___X___ : ___ : ___ : ___ : ___ : ___ : ___ Weak

OR

Strong ___ : ___ : ___ : ___ : ___ : ___ : ___X___ Weak

If you feel that your body is **fairly well described** by the adjective at the end of the scale, place a cross as follows :

Strong ___ : ___X___ : ___ : ___ : ___ : ___ : ___ Weak

OR

Strong ___ : ___ : ___ : ___ : ___ : ___X___ : ___ Weak

If you feel that your body is **only slightly described** by the adjective at the end of the scale, place a cross as follows :

Strong ___ : ___ : ___X___ : ___ : ___ : ___ : ___ Weak

OR

Strong ___ : ___ : ___ : ___ : ___X___ : ___ : ___ Weak

If you consider both adjectives to be **equally descriptive** of your body <u>or</u> both to be **completely irrelevant** to your description of your body then place the cross in the middle space as follow :

Strong ___ : ___ : ___ : ___X___ : ___ : ___ : ___ Weak

<u>PLEASE REMEMBER</u>:

1. Place the cross in the spaces, not on the boundaries.

2. Do not forget to put a cross on every scale.

3. Do not put more than one cross on a single scale.

Make each scale a separate and independent judgement. Work at fairly high speed and do not worry or puzzle over individual scales. Now turn the page and place your crosses accordingly.

MY BODY

Office

Graceful	___:___:___:___:___:___:___	Awkward
Lethargic	___:___:___:___:___:___:___	Energetic
Swift	___:___:___:___:___:___:___	Sluggish
Calm	___:___:___:___:___:___:___	Agitated
Ugly	___:___:___:___:___:___:___	Beautiful
Rigid	___:___:___:___:___:___:___	Flexible
Fit	___:___:___:___:___:___:___	Unfit
Unbalanced	___:___:___:___:___:___:___	Balanced
Steady	___:___:___:___:___:___:___	Unsteady
Weak	___:___:___:___:___:___:___	Strong
Relaxed	___:___:___:___:___:___:___	Tense
Masculine	___:___:___:___:___:___:___	Feminine
Slow	___:___:___:___:___:___:___	Fast
Poised	___:___:___:___:___:___:___	Unpoised
Healthy	___:___:___:___:___:___:___	Sick
Clumsy	___:___:___:___:___:___:___	Well-coordinated
Straight	___:___:___:___:___:___:___	Twisted
Mobile	___:___:___:___:___:___:___	Immobile
Flawed	___:___:___:___:___:___:___	Perfect
Uncontrollable	___:___:___:___:___:___:___	Controllable
Active	___:___:___:___:___:___:___	Passive
Delicate	___:___:___:___:___:___:___	Robust

Please indicate how disfigured you believe you appear to others as a result of your condition, by circling the appropriate number on the scale below :

Not disfigured at all — Extremely disfigured

0 1 2 3 4 5 6 7 8 9 10

B.D.I.

Office

On this page are groups of statements. Please read each group of statements carefully. Then pick out the one statement in each group which best describes the way you have been feeling the PAST WEEK INCLUDING TODAY Circle the number beside the statement you picked. If several statements in the group seem to apply equally well, circle each one. Be sure to read all of the statements in each group before making your choice

1 0 I do not feel sad
  1 I feel sad
  2 I feel sad all the time and can't snap out of it
  3 I am so sad or unhappy that I can't stand it

2 0 I am not particularly discouraged about the future
  1 I feel discouraged about the future
  2 I feel I have nothing to look forward to
  3 I feel that the future is hopeless and things cannot improve

3 0 I do not feel like a failure
  1 I feel I have failed more than the average person
  2 As I look back on my life, all I can see is a lot of failures
  3 I feel I am a complete failure as a person

4 0 I get as much satisfaction out of things as I used to
  1 I dont enjoy things the way I used to
  2 I dont get real satisfaction out of things anymore
  3 I am dissatisfied or bored with everything

5 0 I don't feel particularly guilty
  1 I feel guilty a good part of the time
  2 I feel quite guilty most of the time
  3 I feel guilty all of the time

6 0 I don't feel I am being punished
  1 I feel I may be punished
  2 I expect to be punished
  3 I feel I am being punished

7 0 I don't feel disappointed in myself
  1 I am disappointed in myself
  2 I am disgusted with myself
  3 I hate myself

B.D.I.(cont)

Office

8 0 I don't feel I am worse than anyone else
1 I am critical of myself for my weaknesses/mistakes
2 I blame myself all the time for my faults
3 I blame myself for everything bad that happens

9 0 I don't have any thoughts of killing myself
1 I have thoughts of killing myself, but would not carry them out
2 I would like to kill myself
3 I would kill myself if I had the chance

10 0 I don't cry anymore than usual
1 I cry more now than I used to
2 I cry all of the time now
3 I used to be able to cry, but now I can't cry even though I want to

11 0 I am no more irritated now than I ever am
1 I get annoyed/irritated easier now than I used to
2 I feel irritated all of the time now
3 I don't get irritated at all by the things that used to irritate me

12 0 I have not lost interest in other people
1 I am less interested in other people than I used to be
2 I have lost most of my interest in other people
3 I have lost all of my interest in other people

13 0 I make decisions about as well as I ever could
1 I put off making decisions more than I used to
2 I have greater difficulty in making decisions than before
3 I can't make decisions at all anymore

14 0 I don't feel I look any worse than I used to
1 I am worried that I am looking old / unattractive
2 I feel that there are permanent changes in my appearance that make me look unattractive
3 I believe that I look ugly

15 0 I can work about as well as before
1 It takes extra effort to get started on doing something
2 I have to push myself very hard to do anything
3 I can't do any work at all

B.D.I.(cont)

Office

16 0 I can sleep as well as usual
1 I don't sleep as well as I used to
2 I wake up 1 or 2 hours earlier than usual and find it hard to get back to sleep
3 I wake up several hours earlier than I used to and cannot get back to sleep

17 0 I don't get any more tired than usual
1 I get tired more easily than I used to
2 I get tired from doing almost anything
3 I am too tired to do anything

18 0 My appetite is no worse than usual
1 My appetite is not as good as it used to be
2 My appetite is much worse now
3 I have no appetite at all now

19 0 I haven't lost much weight, if any, lately
1 I have lost more than 5 pounds
2 I have lost more than 10 pounds
3 I have lost more than 15 pounds

Question : I am purposely trying to lose weight by eating less

Answer : (tick one) YES ..... or NO .....

20 0 I am no more worried about my health than usual
1 I am worried about my physical problems, such as aches and pains, upset stomach or constipation
2 I am very worried about my physical problems and it is very difficult to think about much else
3 I am so worried about my physical problems that I cannot think of anything else

21 0 I have not noticed any recent change in my interest in sex
1 I am less interested in sex than I used to be
2 I am much less interested in sex now
3 I have lost interest in sex completely

S. E. S.

Off.

Below are a number of statements which people sometimes make about themselves. Read each statement and tick the box which indicates how much you agree or disagree with what it says.

	strongly agree	agree	disagree	strongly disagree	
1. On the whole, I am satisfied with myself	☐	☐	☐	☐	☐
2. At times I think I am no good at all	☐	☐	☐	☐	☐
3. I feel that I have a number of good qualities	☐	☐	☐	☐	☐
4. I am able to do things as well as most other people	☐	☐	☐	☐	☐
5. I feel I do not have much to be proud of	☐	☐	☐	☐	☐
6. I certainly feel useless at times	☐	☐	☐	☐	☐
7. I feel that I am a person of worth, at least on an equal plane with others	☐	☐	☐	☐	☐
8. I wish I could have more respect for myself	☐	☐	☐	☐	☐
9. All in all, I am inclined to feel that I am a failure	☐	☐	☐	☐	☐
10. I take a positive attitude towards myself	☐	☐	☐	☐	☐

QUESTIONNAIRE ON " THE IMPACT OF DYSTONIA"

Put a tick in the box which best describes how you feel about your dystonia, at present.

00) Today's Date is .....

Your code number is

Office 60

	Strongly disagree	Mildly disagree	Neither agree or disagree	Mildly agree	Strongly agree	
01. I have a hard time adjusting to my illness.						
02. My illness makes me feel useless at times.						
03. Because of my illness I miss the things I like to do most.						
04. Health problems make me more dependant on others.						
05. People are often uncomfortable around me because of my illness.						
06. I will never be self-sufficient enough to make me happy.						
07. My illness does NOT make me feel inadequate.						P.T.O.

Please indicate which of these four statements best describes your feeling to your dystonia at present, by circling one of the numbers :

Office

01. I find it hard to believe that I have to live with dystonia for the rest of my life. I am shocked by the implications of dystonia for myself and my family.

+----+
| |
+----+

02. Why have I developed dystonia ? Why me ? What have I done to deserve this ?

03. My life is ruined by dystonia. My future is grim because of dystonia.

04. I have come to terms with the reality of my dystonia and am trying to cope with its effects as best as I can. I am trying to get on with my life, despite having dystonia

Finally, in thanking you for completing what, in some cases, has been a very large number of questionnaires, can I ask you one final question (and **please feel free to speak openly**).

Are there any criticisms or comments about the way you feel the research has been carried out, either on a personal level or from an organisational point of view ?

+----+
| |
+----+

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+----+
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+----+

+----+
| |
+----+

MAY I SINCERELY THANK YOU FOR ALL YOUR ASSISTANCE AND COOPERATION IN THIS IMPORTANT RESEARCH PROJECT

QUESTIONNAIRE FOR " YOUR PRIMARY CARER "

This form is to be completed by the person who is most involved in looking after (or who lives with) the person with dystonia (the so-called "primary carer"). All answers will be treated in the utmost confidence and this form should be placed, after completion, directly into an envelope marked C.S.I.

Below (and overleaf) are a list of statements, which have been made by other people who have to live with someone who has dystonia. You may feel that some of the statements also apply to you, whilst others do not.

Please indicate how much you agree or disagree with each of these statements, by ticking the appropriate box. Answer according to how you have felt over the **past month.**

#) Questions					Office
00) Today's Date is ..... Your code number is					70
01) What is your relationship to the person with dystonia ?					
	Strongly Agree	Agree	Disagree	Strongly Disagree	
Living with someone who has dystonia, I feel that ..					
02) My sleep is disturbed	☐	☐	☐	☐	☐
03) It is inconvenient	☐	☐	☐	☐	☐
04) It is a physical strain	☐	☐	☐	☐	☐
05) It is confining	☐	☐	☐	☐	☐
06) There have had to be family adjustments	☐	☐	☐	☐	☐
07) I have had to change my personal plans	☐	☐	☐	☐	☐

P.T.O.

(cont)

	Strongly Agree	Agree	Disagree	Strongly Disagree	Office
08) There have been other demands on my time	☐	☐	☐	☐	☐
09) I have had to make emotional adjustments	☐	☐	☐	☐	☐
10) Some of his / her behaviour is upsetting to me	☐	☐	☐	☐	☐
11) I have had to make adjustments at work	☐	☐	☐	☐	☐
12) It is a financial strain	☐	☐	☐	☐	☐
13) I am completely overwhelmed	☐	☐	☐	☐	☐
14) It is distressing that (name of person) has changed from his / her former self.	☐	☐	☐	☐	☐

15) If you would like to write a statement about how you feel that dystonia has changed your life in relation to the person with dystonia, feel free to do so here

☐

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☐

☐

☐

Remember : everything you say is treated in confidence.

QUESTIONNAIRE ON EXTERNAL FACTORS AFFECTING DYSTONIA

The purpose of this questionnaire is to obtain information about environmental factors which might affect your dystonic spasms. Several people in the study have informed us that certain external factors affect either the severity of their spasms or increase pain levels. As you know yourself some days are good and others are bad. Why ? Is there a common thread ?

We are now able to compare a large number of people with different dystonias and to see if there is any correlation between a number of different factors. The results of this questionnaire will help us to understand if there are any common features to any particular phenomenon.

#) Questions — Write answers .....

00) Today's Date is ..... Your code number is

Office: 80

<u>General Questions</u> — Please circle answer

01) Have there been any changes in your life pattern, apart from major operations, illnesses, etc. of which the research study is already aware, that you think may have contributed to your illness ?

YES NO

02) If YES, please describe these, with dates :

.....

.....

.....

03) Do you currently smoke regularly ? YES NO

04) Are you affected by other's smoking ? YES NO

05) Do you have gas appliances in your house? YES NO

06) Are you affected by certain sounds ? YES NO

07) If YES, please describe the type of sound(s)

.....

08) Are you affected by bright light/sunlight YES NO

09) If YES, please describe how it affects you:

.....

10) Is your drinking water ?

	Tick which	Office
Spring	.....	
Well	.....	
Mains	.....	
Filtered	.....	
Bottled	.....	

11) Is your mains water ?

Fluoridated	.....	
Soft	.....	
Hard	.....	
Softened	.....	
Don't know	.....	

The following sections are divided into :-
a) Allergic reactions b) Exposure to chemicals
c) Food and Drink d) Environmental substances

We would like you to complete the next two pages, but if you do not think you are affected by any of the above, you can finish this questionnaire after Page 4. Read on, just in case any of it makes you think !

12) Please write down any other factors (apart from the above) which you think affects your dystonia ?

.....

.....

.....

.....

a) <u>Allergic reactions</u>

13) Have you ever had any allergic problems? YES NO

14) If YES, please tick the type(s) shown below :

Asthma	.....	
Migraines	.....	
Persistent fatigue	.....	
Hay Fever	.....	
Arthritis	.....	
Hyperactivity	.....	
Rhinitis	.....	
Mouth Ulcers	.....	
Stomach Ulcers	.....	
Wind / Bloating	.....	
Eczema	.....	
Puffy ankles / hands / face	.....	
Uticaria / Hives	.....	
Diarrhoea	.....	
Other (describe)	.....	

15) Have any close blood relatives ever had any of the allergic problems shown ? YES NO

b) **Chemical Exposure**

16) Have you ever been exposed to chemicals? YES NO

17) If YES, was it during crop straying ? YES NO

18) Repeated exposure to industrial pollution YES NO

19) Was it in an industrial accident ? YES NO

20) Exposure to fumes from a chemical fire ? YES NO

21) Other (describe) ..... ? YES NO

c) **Food and Drink**

22) Do you think that your symptoms fluctuate as a result of the foods you eat or the beverages you drink ? YES NO

23) If YES, are there any foods that you know make you ill now, made you ill as a child or make your spasms worse ? YES NO

24) If YES, please check the following and indicate which of them affects you by circling the number :

If you are not affected at all, circle 0
If you are only mildly affected, circle 1
If you are moderately affected, circle 2
If you are severely affected, circle 3

Meat / Fish	a. Pork	0	1	2	3
	b. Beef	0	1	2	3
	c. Red meat	0	1	2	3
	d. Plaice	0	1	2	3
	e. Other	0	1	2	3
Cereals	f. Wheat	0	1	2	3
	g. Rice	0	1	2	3
	h. Corn	0	1	2	3
	i. Oats	0	1	2	3
	j. Other	0	1	2	3
Dairy Products	k. Milk	0	1	2	3
	l. Egg	0	1	2	3
	m. Cheese	0	1	2	3
	n. Yoghurt	0	1	2	3
	o. Other	0	1	2	3

						Office
Liquids	p. Coffee	0	1	2	3	
	q. Tea	0	1	2	3	
	r. Other	0	1	2	3	
Fruit / Vegetables	s. Citrus Fruit	0	1	2	3	
	t. Potato	0	1	2	3	
	u. Other	0	1	2	3	
Other Foods	v. Yeast	0	1	2	3	
	x. Chocolate	0	1	2	3	
	y. Sugar	0	1	2	3	
	z. Other	0	1	2	3	

25) Have you ever been on an exclusion diet? YES NO

26) Are there any foods or drinks you have frequently, crave for and actually make you feel better after consumption ? YES NO

27) If YES, please name them and the affect they have:

.....

.....

.....

28) How many cups of tea do you drink a day .....

29) How many cups of coffee drunk in a day ? .....

30) Does decaffeinated coffee or tea make any difference to your spasms ? YES NO

31) Do wines, spirits or beer affect you in any way ? YES NO

32) If YES, describe how they affect you :

.....

.....

33) Are you affected by Colas or other fizzy drinks, in any way ? YES NO

34) If YES, describe how they affect you :

.....

.....

THANK YOU FOR COMPLETING THIS QUESTIONNAIRE. THE NEXT 3 PAGES NEED ONLY BE COMPLETED IF YOU THINK YOU OR YOUR SPASMS ARE AFFECTED BY OTHER ENVIRONMENTAL SUBSTANCES

d. <u>Other environmental substances</u>

Office

Below are a list of substances. Please indicate the extent to which you are affected by any of them, by circling the appropriate number, as follows :-

If you are not affected at all, circle 0
If you are only mildly affected, circle 1
If you are moderately affected, circle 2
If you are severely affected, circle 3

NATURAL INHALANTS

35) Pollens	aa. New mown grass	0	1	2	3	
	ab. Trees	0	1	2	3	
	ac. Long Grass	0	1	2	3	
	ad. Hay	0	1	2	3	
	ae. Other pollens	0	1	2	3	
36) Moulds	af. Damp humid days	0	1	2	3	
	ag. Old houses	0	1	2	3	
	ah. Fungus	0	1	2	3	
	ai. Moulds	0	1	2	3	
	aj. Dust	0	1	2	3	
37) Animals	ak. Dogs	0	1	2	3	
	al. Cats	0	1	2	3	
	am. Horses	0	1	2	3	
	an. Rodents	0	1	2	3	
	ao. Birds	0	1	2	3	
	ap. Feathers	0	1	2	3	
38) Insects	aq. Bee stings	0	1	2	3	
	ar. Wasp stings	0	1	2	3	
	as. House dust	0	1	2	3	
	at. Mites	0	1	2	3	
39) Plants	au. Odour of pines	0	1	2	3	
	av. Pine products	0	1	2	3	
	aw. Houseplants	0	1	2	3	
	ax. Manure	0	1	2	3	
	ay. Silage	0	1	2	3	
	az. Rotting vegetation	0	1	2	3	

CHEMICALS

40) Aerosols	ba. Degreasers	0	1	2	3	
	bb. Deodorants	0	1	2	3	
	bc. Hairsprays	0	1	2	3	
	bd. Polishes	0	1	2	3	
	be. Insecticides	0	1	2	3	
	bf. Pesticides	0	1	2	3	
	bg. Air freshener	0	1	2	3	

If you are not affected at all, circle 0
If you are only mildly affected, circle 1
If you are moderately affected, circle 2
If you are severely affected, circle 3

							Office
41) Cosmetics	bh. After-shave	0	1	2	3		
	bi. Creams	0	1	2	3		
	bj. Deodorants	0	1	2	3		
	bk. Perfume	0	1	2	3		
	bl. Powder	0	1	2	3		
	bm. Make-up	0	1	2	3		
	bn. Shampoo	0	1	2	3		
42) Flooring	bo. New carpets	0	1	2	3		
	bp. Linoleum	0	1	2	3		
	bq. Floor tiles	0	1	2	3		
	br. Sealer	0	1	2	3		
	bs. Adhesive	0	1	2	3		
43) Foam Rubber	bt. Carpet backing	0	1	2	3		
	bu. Cushion	0	1	2	3		
	bv. Upholstery	0	1	2	3		
	bw. Padding	0	1	2	3		
44) Cleaners	bx. Ammonia	0	1	2	3		
	by. Bleaches	0	1	2	3		
	bz. Detergents	0	1	2	3		
	ca. Liquid polishers	0	1	2	3		
	cb. Silver polish	0	1	2	3		
	cd. Furniture polish	0	1	2	3		
	cc. Carpet shampoo	0	1	2	3		
45) Fuels	cd. Heating oil	0	1	2	3		
	ce. Gas	0	1	2	3		
	cf. Paraffin	0	1	2	3		
	cg. Calor/Butane	0	1	2	3		
	ch. Coal Fire	0	1	2	3		
	ci. Charcoal	0	1	2	3		
	cj. Burning tar	0	1	2	3		
	ck. Burning rubber	0	1	2	3		
46) Motoring	cl. Petrol fumes	0	1	2	3		
	cm. Oil	0	1	2	3		
	cn. Diesel fumes	0	1	2	3		
	co. Upholstery	0	1	2	3		
	cp. Exhaust fumes	0	1	2	3		
47) Paints	cq. New paint	0	1	2	3		
	cr. Paint stripper	0	1	2	3		
	cs. Turpentine	0	1	2	3		
	ct. Varnish	0	1	2	3		
	cu. Oils	0	1	2	3		
	cv. Fixative	0	1	2	3		

If you are not affected at all, circle 0
If you are only mildly affected, circle 1
If you are moderately affected, circle 2
If you are severely affected, circle 3

						Office
48) Tobacco	cw. Cigarettes	0	1	2	3	
	cx. Cigars	0	1	2	3	
	cy. Pipe	0	1	2	3	
	cz. Cigarette Smoke	0	1	2	3	
49) Solvents	da. Lighter fuel	0	1	2	3	
	db. Carbon Tetrachloride	0	1	2	3	
	dc. Glues	0	1	2	3	
	dd. Newsprint	0	1	2	3	
	de. White Spirit	0	1	2	3	
	df. Dry Cleaning Fluid	0	1	2	3	
	dg. Methylated Spirits	0	1	2	3	
	dh. Surgical Spirit	0	1	2	3	
50) Others	di. Chlorine	0	1	2	3	
	dj. Bitumen Tar	0	1	2	3	
	dk. Asphalt	0	1	2	3	
	dl. Weedkillers	0	1	2	3	
	dm. Mothballs	0	1	2	3	
	dn. Disinfectant	0	1	2	3	
	do. Swimming Pools	0	1	2	3	
	dp. Food Additives	0	1	2	3	
	dq. Pesticide Residues in foods	0	1	2	3	
	dr. Pollutants in Water	0	1	2	3	
	ds. Waxes (on some fruit and vegetables)	0	1	2	3	
	dt. Tartrazine	0	1	2	3	
	du. Phenolic Resin in cans	0	1	2	3	
	dv. Formaldehyde	0	1	2	3	
51) Smells given of by	dw. Plastics	0	1	2	3	
	dx. Coated Paper	0	1	2	3	
	dy. New Fabric	0	1	2	3	
	dz. Others	0	1	2	3	

The above list although very long is not comprehensive. If there any other elements which affect you, please add them below and circle the appropriate number :

52) Any other substance	0	1	2	3	
.....	0	1	2	3	
.....	0	1	2	3	

(cont)

52) Any other substance 0 1 2 3

.... 0 1 2 3

.... 0 1 2 3

.... 0 1 2 3

.... 0 1 2 3

.... 0 1 2 3

53) Any there any other comments that you may wish to write about this whole area of external factors which you think might affect either your dystonic spasms or your pain threshold levels ? Why, for example, do you think that some times are better than others ? Do you have a theory about this ?

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Office

QUESTIONNAIRE ABOUT DIAGNOSIS

This questionnaire contains a number of specific questions which Dr Phil Duffey, Neurological Consultant at York General Hospital, and who used to work at the Royal Victoria Infirmary in Newcastle would like to ask you about your past consultations, treatments and diagnoses. Dr Duffey has worked closely with the research team since 1993 on all the programmes. He is interested to learn how your diagnosis of dystonia came to be made and in particular who you were advised to see when your condition first arose.

#) Questions — Tick answer or comment here .....

Office: 81

00) What is today's Date : ..... Your code number is

01) To which of the following medical specialists were you referred, before your dystonia was diagnosed ?

- Orthopaedic Surgeon .....
- Rheumatologist .....
- Psychiatrist .....
- Other (please specify) .....

02) What physical or alternative treatments for your condition have been advised or tried ?

- Use of an Osteopath .....
- Acupuncture .....
- Hypnosis .....
- Physiotherapy .....
- Psychotherapy .....
- Other (please specify) .....

03) How long before the correct diagnosis was made did your symptoms begin ?

.....

Thank you for your help. I hope that the information you have provided may make the diagnosis of future patients with dystonia easier. Dystonia can be very difficult to diagnose and it is not my intention to imply criticism of those involved in your particular case. Hindsight is a wonderful tool !

Dr P. Duffey, MB.BS., MRCP.

FIRST QUESTIONNAIRE - DNP EVALUATION

Office
91

00) Today's Date is ..... Your code number is
Write your answers below :

01) Can you estimate how many times you have been injected since you first started ? .....

02) How many different people have given you those injections since you first started ? .....

03) How much time are they generally able to spend with you (in minutes) ? .....

04) How many times has their technique in giving you the injections hurt or been painful ? .....

Rating Scale to answer Questions Numbers 5 and 6 :
1 = Very good 2 = Good 3 = Average, 4 = Poor, 5 = Very Poor

05) How would you rate their knowledge of you and your particular condition ?
At their best .....
At their worst .....

06) How would you rate their personal attitude towards you and your condition ?
At their best .....
At their worst .....

07) Would you like to tell me (in your own words) what you think about the **service** you get from Hunters Moor :

.....

.....

.....

08) What type of muscle spasm(s) do you have ?

.....

09) Do you know what is the cause of this type of spasm ?

If, YES, what is it ? .....

10) Is there anything which makes these spasms worse ?

.....

11) Is there anything which makes these spasms better ?

.....

SECOND QUESTIONNAIRE - DNP EVALUATION

Office 92

00) Today's Date is ..... Your code number is
Write your answers below :

01) How many times you have been injected since you last answered this questionnaire ? .....

02) How many different people have given you those injections since then ? .....

03) How much time are they generally able to spend with you (in minutes) ? .....

04) How many times has their technique in giving you the injections hurt or been painful ? .....

Rating Scale to answer Questions Numbers 5 and 6 :
1 = Very good 2 = Good 3 = Average, 4 = Poor, 5 = Very Poor

05) How would you rate their knowledge of you and your particular condition ?
At their best .....
At their worst .....

06) How would you rate their personal attitude towards you and your condition ?
At their best .....
At their worst .....

07) Would you like to tell me (in your own words) how you think the *service* you **currently** get could be improved :

.....

.....

.....

08) Do you have primary or secondary dystonia ?

.....

09) What is thought to cause primary dystonia ?

.....

10) What are one of the causes of secondary dystonia ?

.....

11) What is the main difference between the two types ?

.....

THIRD QUESTIONNAIRE - DNP EVALUATION

Office 93

00) Today's Date is ..... Your code number is
Write your answers below :

01) How many times you have been injected since you last answered this questionnaire ? .....

02) How many different people have given you those injections since then ? .....

03) How much time are they generally able to spend with you (in minutes) ? .....

04) How many times has their technique in giving you the injections hurt or been painful ? .....

Rating Scale to answer Questions Numbers 5 and 6 :
1 = Very good 2 = Good 3 = Average, 4 = Poor, 5 = Very Poor

05) How would you rate their knowledge of you and your particular condition ? At their best ..... At their worst .....

06) How would you rate their personal attitude towards you and your condition ? At their best ..... At their worst .....

07) Would you like to tell me (in your own words) what you think about the service you get from Hunters Moor :

.....

.....

.....

08) What is meant by idiopathic dystonia ?

.....

09) What is the basal ganglia ?

.....

10) Do you think your treatment has improved compared to the treatment you got a year ago ? YES / NO

11) How do you rate the service compared to the service you received a year ago ?

Circle one answer : BETTER / ABOUT THE SAME / WORSE

FOURTH QUESTIONNAIRE - DNP EVALUATION

Office
+---+
|94
+---+

00) Today's Date is ..... Your code number is
Write your answers below:

01) Please tell me in your own words, what you think about the whole project regarding the Nurse Practitioner :

.....

.....

.....

.....

02) If you have the independent choice, would you prefer to be injected at home or in the clinic ? .....

03) If you prefer to be injected at home, why ? .....

.....

.....

04) If you prefer to be injected at the clinic, why ? ....

.....

.....

05) Is there anything else you would like to talk about or anything you would like us to know about this project ?

.....

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APPENDIX C

SPSS CODING FILE

The PhD SPSS file entitled 'phddata.sav' contains 6 years data from 6th May 1993 to 5th May 1999, which has also been used on other associated research projects :-

ESD = Epidemiological Survey of Dystonia - all data available at the time
CUA = Cost Utility Analysis - No 001 to 231 inclusive (see note re patient numbering)
EPD = Epidemiology of Primary Dystonia - 'if aet = 1' - all data at the time
DNP = Dystonia Nurse Practitioner - all data on 126 selected cases
PSR = Psycho-Social Research - all data to 550 + 564, 594 and 677
IFD = Dystonia affecting working musicians - only 3 selected cases
SER = Socio-Economic Research - all data available at the time
EST = Epidemiology of Spasmodic Torticollis - 001 - 766 inclusive

Patient Numbering and Status used in 'phddata.sav'.

The patient's ID number is automatically determined by the row number and the column shown as ESD is used for ranking purposes only. **NB** : Six CUA patients have been removed and replaced by ESD patients between 04.12.95 and 04.03.96. They were No's 148, 152, 159, 170, 175 and 210, who were all non-dystonic CUA controls.

001 to 199 inclusive : were active CUA patients coded A, B or D
200 : is a new ESD patient coded M
201 to 231 inclusive : were external control CUA patients, re-coded to ESD
232 to 300 inclusive : are new ESD patients coded A, B or M
301 to 347 inclusive : were (1994) 500 Series patients re-numbered to 300
348 to 937 inclusive : are new ESD patients numbered and coded accordingly

Syntax Files

Syntax file copies have been enclosed separately but most detailed formulae have been included in the coding file. The 7 syntax files were run in the following order :-

phd-geog.sps : Recoding 'post codes' into 'county' and 'district'.
phd-all.sps : 21 computations re Age, DOB, Dates, Differences, etc
phd-dec.sps : 12 computations re defining decimal places in the above.
phd-sf36.sps : 29 computations re EuroQol and SF36 as per handbook.
phd-qual.sps : 3 computations re qualitative open-questions.
phd-psr.sps : 17 computations re PSR, as per Dr Jahanshahi's instructions.
phd-env.sps : 93 comps 'if env = 1, envaa = 0,' etc., & 'if alg = 2, alga = 2,' etc.

SPSS Coding Frame

The SPSS codes (up to # 275) were used in the Cost Utility Analysis (CUA) but have since been amended for the ESD, EST, IFD, EPD, SER, PSR and DNP, as required. All variables are entered as Numeric 4.0; missing values None; alignment Right and column width 4, unless otherwise shown. The value label positions, ie #, are numbered as per "the list of variables on the working file". The file contains 885 rows (subjects) x 842 columns (variables) and therefore has a total of 745,170 potential data sets.

#	Name	Variable Label	Additional Info
001	esd	**ESD** Patient ID **number**	
002	geog1	**County** of Residence of ESD subject calc. from post codes - see syntax file : 'phd-geog.sps' 10 = Co. Durham 20 = Cleveland (Teesside) 30 = Tyne & Wear 40 = Northumberland 50 = N. Cumbria 60 = S. Cumbria 70 = N. Yorks 80 = Rest of Yorkshire 90 = Other Areas in UK	
003	geog2	**District** (Administrative) - see syntax file : 'phd-geog.sps' 10 = Co. Durham consists of :- 11 = Chester Le Street 12 = Darlington 13 = Derwentside 14 = Durham 15 = Easington 16 = Sedgefield 17 = Teesdale 18 = Wear Valley 20 = Cleveland (Teesside) consists of :- 21 = Hartlepool 22 = Middlesbrough 23 = Redcar & Cleveland 24 = Stockton on Tees 30 = Tyne & Wear consists of :- 31 = Gateshead 32 = Newcastle 33 = North Tyneside 34 = South Tyneside 35 = Sunderland 40 = Northumberland consists of :- 41 = Alnwick 42 = Berwick upon Tweed 43 = Blyth Valley 44 = Castle Morpeth 45 = Tynedale 46 = Wansbeck	

50 = N. Cumbria consists of :-
51 = Allerdale
52 = Carlisle
53 = Eden

60 = S. Cumbria consists of :-
61 = Barrow
62 = Copeland
63 = S Lakeland
64 = Lanchester

70 = N. Yorks
80 = Rest of Yorkshire
90 = Other Areas in UK

004 hosp1 **Main Hospital (alpha)** - Patient registered at String 4

A = NRI North Riding Infirmary, Middlesbrough
B = HMH Hunters Moor Hospital, Newcastle
C = CUA Patient in CUA living outside the region, unknown hospital
D = More Patient registered at more than 1 Hospital in the region
E = ESD Patient in ESD living outside the region, unknown hospital
F = SSG South Shields General Hospital
G = MGH Middlesbrough General Hospital
H = HGH Hartlepool General Hospital
J = FHN Freeman Hospital, Newcastle
K = BAH Bishop Auckland Hospital
L = LGH Leeds General Hospital
M = DMH Darlington Memorial Hospital
N = NGH Newcastle General Hospital
R = RVI Royal Victoria Infirmary, Newcastle
S = SGH Sunderland General (& Royal) Hospital
T = SEI Sunderland Eye Infirmary
X = UNK Patient in ESD living inside the region, unknown hospital
Y = DRY Dryburn Hospital, Durham

005 hosp2 **Second Hospital** (ie. patient's notes also available from)
Variable codes as above

006 hosp3 **Combined Hospitals** (for 135 'D' patients as of 31.12.98)
NB : 'D' patients were only shown in the CUA, but the details are :-

A+B = 7	A+G = 11	A+H = 2	A+K = 1	A+L = 2
A+M = 2	A+R = 1	B+A = 16	B+J = 1	B+K = 1
B+N = 7	B+R = 39	B+S = 5	B+Y = 8	G+A = 2
G+N = 3	K+A = 3	K+B = 1	M+A = 3	M+B = 1
M+G = 1	M+N = 1	N+B = 3	R+B = 9	T+B = 1
T+S = 1	T+Y = 2	S+N = 1	Total = 135	

007 hosp4 **Main Hospital (num)**
As per # 004 but numeric for filter purposes, etc.

1 = NRI North Riding Infirmary, Middlesbrough
2 = HMH Hunters Moor Hospital, Newcastle
3 = CUA Patient in CUA living outside the region, unknown hospital
4 = More Patient registered at more than 1 Hospital in the region
5 = ESD Patient in ESD living outside the region, unknown hospital
6 = SSG South Shields General Hospital
7 = MGH Middlesbrough General Hospital
8 = HGH Hartlepool General Hospital
9 = BAH Bishop Auckland Hospital
10= LGH Leeds General Hospital
11= DMH Darlington Memorial Hospital
12= NGH Newcastle General Hospital
13= RVI Royal Victoria Infirmary, Newcastle
14= SGH Sunderland General (and Royal) Hospital
15= SEI Sunderland Eye Infirmary
16= UNK Patient in ESD living inside the region, unknown hospital
17= DRY Dryburn Hospital, Durham
18= FHN Freeman Hospital, Newcastle

008 crn1 **Clinical Registration Number** of the patient at their **1st hospital** (if known) String 8

010 crn2 **Clinical Registration Number** of the patient at their **2nd hospital** (if known) String 8

012 surv **Survey Participation** Numeric 4.0

1 = Full participation - completed everything possible
2 = Part participation - completed most, but not all q'res
3 = Non participation - declined, only ESD data collected
4 = ESD data only - not asked to participate
5 = After closing date - the ESD closing date was 31.12.1998
6 = Died before (non) - no interview - ESD data only
7 = Died during (part) - some q'res completed
8 = Died since (full) - most q'res completed, if not all.
9 = Died since (non) - no interview - ESD data only

EuroQol Questionnaire : Q'res No. 1 or 10

013 date10 **Date of Q're 1** or date of first contact or date entered on SPSS — dd.mm.yy 8

014 age10 **Age** (of patient) **at date10** — Numeric 4.1

Compute age10 = YRMODA (xdate.year(date10), xdate.month(date10), date.mday(date10)) / 365.25 - YRMODA (date.year(dob),xdate.month(dob), xdate.mday(dob))/ 365.25. Execute.

Compute age10 = (age10 + 0.5) - (MOD((age 10 + 0.5),1)). Execute.
NB : This formula computes and shows age10 with no decimal places.

015 stat1 **Status of Q're** — Numeric 4.0

1 = Injected today date
2 = Mid-Injection Cycle
3 = Inj. stopped during (study) - used in CUA only
4 = Never (been) injected
5 = Inj. stopped before (study)
6 = In remission
7 = Q're not returned
8 = Unknown status
9 = Q're not issued - used in ESD only

016 mob1 relates to Q. 01: **Mobility**

1 = I have no problems (in walking)
2 = I have some problems (in walking)
3 = I am confined to bed (wheelchair)

017 self1 relates to Q. 02: **Self-care**

1 = I have no problems (re self-care)
2 = I have some problems (washing)
3 = I am unable to wash (or dress)

018 act1 relates to Q. 03: **Usual activity**

1 = I have no problems
2 = I have some problems
3 = Am unable to perform (usual activity)

019 disc1 relates to Q. 04: **Pain / discomfort**

1 = I have no pain (or discomfort)
2 = I have moderate pain (or discomfort)
3 = I have extreme pain (or discomfort)

020 anx1 relates to Q. 05: **Anxiety / depression**

1 = I am not anxious (or depressed)
2 = I am moderately anxious (or depressed)
3 = I am extremely anxious (or depressed)

021 ostate1 **EuroQol Own Status 1** (EuroQol calculation)
see syntax file : 'phd-sf36.sps' for formula

11111 = All 1 - no problems
22222 = All 2 - some problems
33333 = All 3 - severe problems

022	m1	**Mobility Calcs 1**	(Missing Value = 9.000)
023	s1	**Self-care Calcs 1**	(Missing Value = 9.000)
024	a1	**Usual Act Calcs 1**	(Missing Value = 9.000)
025	d1	**Pain / Disc. Calcs 1**	(Missing Value = 9.000)
026	x1	**Anx. / Dep. Calcs 1**	(Missing Value = 9.000)

027 euro1 **EuroQol Score 1** (HRQoL = Health-related Quality of Life)

100 = 11111 Best HRQoL score
46.81 = 22222 Average HRQol score
- 17.94 = 33333 Worst HRQoL score

028 heal1 relates to Q. 06: Health State
Visual Analogue Scale (VAS) from 0 to 100

SF 36 Questionnaire : Q'res No. 1 or 10

029 gen1 relates to Q. 07: **General Health** Numeric 4.0

NB : Where variable labels are shown with (numbers), it indicates that the variable has been recoded prior to SF36 calculations in accordance with the manual "How to score the SF-36 Health Survey."

5 = Excellent (1)	= 100	Recoded a second time as
4 = Very Good (2)	= 84	per instructions on Page 17
3 = Good (3)	= 61	of the above manual.
2 = Fair (4)	= 25	
1 = Poor (5)	= 0	(Final result 0 - 100)

030 comp1 relates to Q. 08: **Compared to 1 yr.**

1 = Much better
2 = Better
3 = About same
4 = Worse
5 = Much worse

031 vig1 relates to Q. 09a: **Vigorous Activity**

1 = Yes limited a lot
2 = Yes limited a little
3 = Not limited at all

032 mod1 relates to Q. 09b: **Moderate Activity** Template = Vigorous
033 lift1 relates to Q. 09c: **Lifting / carrying**
034 sev1 relates to Q. 09d: **Climbing several**
035 one1 relates to Q. 09e: **Climbing one**
036 bend1 relates to Q. 09f: **Bending, kneeling**
037 walk1 relates to Q. 09g: **Walking more mile**
038 walb1 relates to Q. 09h: **Walking half mile**
039 walc1 relates to Q. 09i: **Walking 100 yards**
040 bath1 relates to Q. 09j: **Bathing / dressing**

041 pf1 **Physical Functioning 1** (syntax : 'phd-sf36.sps')
0 = Low Physical Functioning
100 = High Physical Functioning

042 cut1 relates to Q. 10a: **Cut down** Template = Yes / No
1 = Yes
2 = No

043 acc1 relates to Q. 10b: **Accomplished less** Template = Yes / No
044 lim1 relates to Q. 10c: **Limited in kind**
045 had1 relates to Q. 10d: **Had difficulty**

046 rp1 **Role - Physical 1** (syntax : 'phd-sf36.sps')
0 = Worse Role - Physical
100 = Better Role - Physical

047 time1 relates to Q. 11a: **Amount of time** Template = Yes / No
048 less1 relates to Q. 11b: **Accomplished less**
049 work1 relates to Q. 11c: **Didn't do work**

050 re1 **Role - Emotional 1** (syntax : 'phd-sf36.sps')
0 = Worse Emotional Functioning
100 = Better Emotional Functioning

051 ext1 relates to Q. 12: **To what extent has** Template = Extent

5 = Not at all (1)
4 = Slightly (2)
3 = Moderately (3)
2 = Quite a bit (4)
1 = Extremely (5)

052 howl relates to Q. 13: **How much bodily** Template = How

6 = None (1)
5 = Very mild (2)
4 = Mild (3)
3 = Moderate (4)
2 = Severe (5)
1 = Very severe (6)

053 pain1 relates to Q. 14: **How much did pain** Template = Extent
Recoded as extent #051

054 bp1 **Bodily Pain 1** (syntax : 'phd-sf36.sps')

0 = Most Bodily Pain
100 = Least Bodily Pain

055 life1 relates to Q. 15a: **Full of life** Template = Life

6 = All of the time (1)
5 = Most of the time (2)
4 = Good bit of the time (3)
3 = Some of the time (4)
2 = A bit of the time (5)
1 = None of the time (6)

056 nerv1 relates to Q. 15b: **Very nervous** Not recoded
057 down1 relates to Q. 15c: **Have you felt down** Not recoded
058 calm1 relates to Q. 15d: **Have you felt calm** Recoded
059 lot1 relates to Q. 15e: **Did you have a lot** Recoded
060 low1 relates to Q. 15f: **Have you felt low** Not recoded
061 worn1 relates to Q. 15g: **Did you feel worn** Not recoded
062 happ1 relates to Q. 15h: **Are you happy** Recoded

063 mh1 **Mental Health 1** (syntax : 'phd-sf36.sps')

0 = Worse Mental Health
100 = Better Mental Health

064 tir1 relates to Q. 15i: **Did you feel tired** Not recoded

065 vt1 **Vitality 1** (syntax : 'phd-sf36.sps')

0 = Least Vitality
100 = Most Vitality

066 soc1 relates to Q. 15j: **Social activities** Not recoded

067 sf1 **Social Functioning 1** (syntax : 'phd-sf36.sps')
0 = Worse Social Functioning
100 = Better Social Functioning

068 ill1 relates to Q. 16a: **I get ill more** Template = Ill

1 = Definitely true
2 = Mostly true
3 = Not sure
4 = Mostly false
5 = Definitely false

069 any1 relates to Q. 16b: **As anybody** Recoded as below

5 = Definitely true (1)
4 = Mostly true (2)
3 = Not sure (3)
2 = Mostly false (4)
1 = Definitely false (5)

070 wor1 relates to Q. 16c: **Health to get worse** Not recoded
071 exc1 relates to Q. 16d: **Health is excellent** Recoded

072 gh1 **General Health 1** (syntax : 'phd-sf36.sps')
0 = Worst General Health
100 = Best General Health

073 type1 **Type of Bot Tox** used - Info from Clinical Analysis Form (CAF)

1 = Speywood - Dysport
2 = Allergan - Botox
3 = Both types used
7 = No longer injected
8 = Unknown type
9 = Never been injected

074 dose1 **Dosage of Bot Tox** - from CAF

075 site1 **Number of sites** - on nearest date to Q're from CAF

076 part1 relates to Q. 17: **With partner** Template = Partner

1 = Extremely positive
2 = Positive
3 = Slightly positive
4 = Neutral (missing) Missing Variable = 4
5 = Slightly negative
6 = Negative
7 = Extremely negative

077 peop1 relates to Q. 18: **With people** Template = Partner
078 burd1 relates to Q. 19: **Felt a burden**
079 emb1 relates to Q. 20: **Embarrassed**
080 app1 relates to Q. 21: **Appearance**
081 con1 relates to Q. 22: **Control**
082 cop1 relates to Q. 23: **Coping with life**
083 sup1 relates to Q. 24: **Support**

084 qual1 **Qualitative Data 1**

8 = Maximum	+ve
20 = Average	+ve
32 = Neutral	0
44 = Average	-ve
56 = Maximum	-ve

Compute qual1 = part1 + peop1 + burd1 + emb1 + app1 + con1 + cop1 + sup1.
Execute.

EuroQol Questionnaire : Q'res No. 2 or 11

085 date11 **Date of Q're 2** or date of last known injection. dd.mm.yy

086 time **time from Q're 10 to Q're 11** Numeric 4.1

Compute time = YRMODA (xdate.year(date11),xdate.month(date11),
xdate.mday(date11)) / 365.25 - YRMODA (xdate.year(date11),xdate.
month(date11),xdate.mday(date11)) / 365.25. Execute.

Compute time = (((time * 10)+0.5)-(MOD(((time * 10)+0.5), 1)))/10. Execute.

087 recode2 **time between Q're 10 and 11**.

1 = Over 3.50 years	
2 = 3.00 - 3.49 yrs	
3 = 2.50 - 2.99 yrs	
4 = 2.00 - 2.49 yrs	
5 = 1.50 - 1.99 yrs	
6 = 1.00 - 1.49 yrs	
7 = 0.50 - 0.99 yrs	
8 = 0.00 - 0.49 yrs	
9 = Negative years	

All variables are then repeated for the rest of Q're No. 11 (ie EuroQol 2 and SF36 2) and duplicates all the variables as for Q're No. 10 from # 088 until # 157.

158 qual3 **Qualitative difference over time**

Compute qual3 = qual1 - qual2. Execute.

A positive score means the subject is feeling better and a negative score means they are feeling worse over the period of time between 10 and 11, defined by #087.

Demographic Questionnaire : Q're No. 20

159 date20 **Date of Q're** No 20 dd.mm.yy 8

160 age20 **Age of patient at date20** Numeric 4.1

Compute age20 = YRMODA (xdate.year(date20),xdate.month(date20), xdate.mday(date20)) / 365.25 - YRMODA (xdate.year(dob),xdate.month (dob),xdate.mday(dob)) / 365.25. Execute.

Compute age20 = (((age20 * 10) + 0.5) - (MOD(((age20 * 10) + 0.5), 1)))/10. Execute.

161 gend relates to Q. 01: **Gender** Numeric 4.0

1 = Male
2 = Female

162 dob relates to Q. 02: **Date of Birth** dd.mm.yy 8

163 age1 **Age at 01.01.99** (ie end of epidemiology)

Compute age1 = YRMODA (98,13,00) / 365.25 - YRMODA (xdate.year(dob) , xdate.month(dob),xdate.mday(dob)) / 365.25. Execute.

Compute age1 = (age1 + 0.5) - (MOD((age1 + 0.5), 1)). Execute.

164 post relates to Q. 03: **Home Post Code** String 4
(2 alpha + 4 numeric, eg DL2 1 = DL02 1) Align left

NB : Post Codes within EPD and Cumbria to Sector Level (see list at end)

165 mar relates to Q. 04: **Marital Status** Numeric 4.0

1 = Single
2 = Married
3 = Cohabiting
4 = Separated
5 = Divorced
6 = Widowed

166 male1 relates to Q. 05: **No of male children**
167 fem1 relates to Q. 05: **No of female children**
168 male2 relates to Q. 05: **Male kids in house**
169 fem2 relates to Q. 05: **Female kids in house**

170 eld relates to Q. 05: **Eldest's birth year**
00 = 1900, then includes every year to 96 = 1996.
171 yng relates to Q. 05: **Youngest's birth year**
00 = 1900, then includes every year to 96 = 1996.

includes **Demographic Questionnaire : Q're No 25**
Q're updating demographic information

172 date25 **Date of Q're 25** dd.mm.yy 8

173 age25 **Age at date25** Numeric 4.1

Compute age25 = YRMODA (xdate.year(date25),xdate.month(date25), xdate.mday(date25)) / 365.25 - YRMODA (xdate.year(dob),xdate.month (dob),xdate.mday(dob)) / 365.25. Execute.

Compute age25 = (((age25 * 10) + 0.5) - (MOD(((age25 * 10) + 0.5), 1)))/10. Execute.

174 diff5 **Time from 20 to 25** (in years) Numeric 2.1

Compute diff5 = age25 - age 20. Execute.

Compute diff5 = (((diff5 * 10) + 0.5) - (MOD(((diff5 * 10) + 0.5), 1)))/10. Execute.

175 emp1 relates to Q. 06: **Employment Status at 20** Numeric 4.0

1 = Full time employment
2 = Part time employment
3 = Unemployed
4 = Self Employed
5 = Unwaged
6 = Retired OAP
7 = Retired ill health
8 = On long term sick
9 = Deceased

176 emp2 relates to Q. 06: **Employment Status at 25** Temp = Employ

177 seg1 relates to Q. 07: **SEG - Patient at 20** Template = SEG

1 = A : Professional
2 = B : Managerial
3 = C_1: White Collar (Skilled Non-Manual)
4 = C_2: Blue Collar (Skilled Manual)
5 = D : Semi-skilled (Unskilled)
6 = E : Retired (Unwaged, Student or Unemployed)
9 = Deceased

178 seg2 relates to Q. 07: **SEG - Patient at 25**
179 seg3 relates to Q. 07: **SEG - Patient (highest ever)**
180 seg4 relates to Q. 08: **SEG - Head of House (current)**
taken to be a 'male' Head of House, if widow = 9
181 seg5 relates to Q. 08: **SEG - Head of House (highest ever)**
taken to be a 'male' Head of House, if widow, not = 9

The above SEG codes are defined according to Reid (1989) as :-

A : Upper Middle Class. Defined as successful business persons (eg, self-employed / manager / executive of large enterprise), higher professionals (eg, bishop, surgeon, medical specialist, barrister, accountant), senior civil servants (above Principal) and local government officers (eg, chief, treasurer, town clerk). Known as Professional

B : Middle Class. Defined as senior, but not the very top, people in the same classification as A. Known as Managerial

C_1: Lower Middle Class. Defined as small tradespeople, non-manual, routine administrative, supervisory and clerical (sometimes referred to as 'white-collar' workers).

C_2: Skilled Working Class (often referred to as 'blue-collar')

D : Semi-skilled and unskilled working class

E : Those at the lowest levels of subsistence, including the retired, those on social security because of sickness or unemployment, and casual workers. I have also included students in this category.

The national percentages of informants falling into these classes are :-

A = 3%
B = 14%
C_1 = 22%
C_2 = 28%
D = 18%
E = 15%.

The reason that this classification has been used is it is the only one is common usage that has the non-employed category of retired, on disability benefit or income support.

182	inc1	relates to Q. 09: **Patient's Income at 20**	Comma 6.0
183	inc2	relates to Q. 09: **Patient's income at 25**	

NB : If there is no change in income shown on Q're 25, then the following formula is used to calculate for inflation :-

If diff5 =	3.2 to 2.7 years	then inc1 x 1.10 = inc2	with 10% added	- 38.4% of study.
	2.6 to 1.6	x 1.07	7%	28.3%
	1.5 to 0.6	x 1.03	3%	5.8%
	0.5 to 0.0	x 1.00	0%	27.5%
				100.0%

184 benel relates to Q. 10: **Benefits - 1st** Numeric 4.0

1 = Sickness Benefit
2 = Invalidity Benefit (Incapacity)
3 = Attendance Allowance
4 = Mobility Allowance
5 = Occupational Pension
6 = Retirement / Widows (Pension)
7 = Income Support
8 = Disabled Working (Allowance)
9 = Loss of Earnings (Benefit) include in Unemployment Benefit
10 = Invalid Care (ICA)
11 = Disabled Living (DLA)
12 = Severe Disability (Allowance)
13 = MOD Disability (War Pension)
14 = Cancer Victim (Allowance)
15 = Child Benefit (Family Allowance)
16 = Family Credit
17 = Industrial Injury (Benefit)
18 = Wages in prison
19 = Rent Allowance

185 bene2 relates to Q. 10: **Benefits - 2nd** Template = Benefits

186 bene3 relates to Q. 10: **Benefits - 3rd**

$benetot **Total of Benefits** - variables bene1, bene2 and bene3
Multiple Response Set - Range 1 to 19

187 hour1 relates to Q. 11: **Patient - hours off** to attend hosp.
188 hour2 relates to Q. 12: **Helper - hours off** to accompany. pat.
189 day1 relates to Q. 13: **Patient - days off** in last 6 weeks.
190 day2 relates to Q. 14: **Partner - days off** in last 6 weeks.
191 mile1 relates to Q. 15: **No of miles - 1 way** (old hospital)
192 mile2 relates to Q. 15: **No of miles to new hospital**
0 = Home visit by DNP (Dystonia Nurse Practitioner)
193 mins1 relates to Q. 16: **No of minutes - 1 way** (old hospital)
194 mins2 relates to Q. 16: **No of minutes to new hospital**
0 = Home visit by DNP (Dystonia Nurse Practitioner)

195 trav1 relates to Q. 17: **Method of travel** Template = Travel

1 = Own Car
2 = Family Car
3 = Friend's Car
4 = Public (Transport) mainly Bus
5 = Public (Transport) mainly Train
6 = Ambulance / Hosp Car (Prison Van)
7 = Taxi
8 = Bicycle
9 = Walked or pushed (in wheelchair)
10 = Home visits - DNP
11 = Aeroplane

196 trav2 relates to Q're 25 : **Method of travel** Template = Travel

197 dr1 (Name of) **Dr who diagnosed** Template = Doctor

0 = Not yet diagnosed
1 = Hawthorne - NRI
2 = Barnes - HMH
3 = Duffey - ESD et al
4 = Allchin - SEI
5 = Burns - RVI / DRY
6 = Tilley - DMH / MGH
7 = Newman - MGH
8 = Saunders - MGH et al
9 = Unknown at HMH
10 = Unknown at NRI
11 = Prof. Walton
12 = DNP study patient
13 = Bates - RVI / Cumbria
14 = Cartlidge - NGH
15 = G-Medwin - NGH
16 = Hudgson - NGH
17 = Jordan - RVI / HMH
18 = Prof. Marsden
19 = Foster - RVI
20 = Unknown Dr
21 = Cleland - SGH
22 = Shakir - MGH / HGH
23 = Shaw - RVI
24 = Jones - SGH
25 = Walls - NGH
26 = Prof. Nattrass / Seggar
27 = Unknown at MGH / HGH
28 = Unknown at NGH
29 = Unknown at RVI
30 = Field - HMH
31 = Bindoff - MGH

198 dr2 (Name of) **2nd Dr to diagnose** Template = Doctor

199 aet1 relates to Q. 18: **Aetiology of Dystonia (old)**
1 = Primary (Idiopathic)
2 = 2ndary (Symptomatic)
3 = Not dystonia (control)
4 = HFS

The aet1 classification was in existence at the start of the study in 1993 and used throughout all the fieldwork. It was amended in October 1996 at the 3rd International Dystonia Symposium in Miami. The aet2 classification was added to ESD in January 1998 and all the 838 subjects were re-classified according to the new codes.

200 aet2 relates to Q. 18: **Aetiology of Dystonia (new)**

10 = Primary Dystonia ie
11 = Oppenheim's
12 = Fam. Torticollis
13 = Fam. Cerv-Cran.
14 = Other Fam. (familial)
15 = Sporadic
20 = Dystonia-Plus ie
21 = D.R.D.
22 = Myoclonic
23 = D.R.D. (familial)
24 = Myoclonic (familial)
30 = Secondary Dystonia - caused by
31 = Levo-Induced
32 = Tardive Dystonia (including neuroleptic induced)
33 = C.P.
34 = M.S.
35 = Infectious
36 = B.E.T.
37 = Cerebrovascular
38 = Metabolic ?
39 = Head Trauma (Head Injury)
40 = Heredodegenerative Diseases ie
41 = M.S.A.
42 = Shy Drager's (Syndrome)
43 = Leigh's (Disease)
44 = P.D.
50 = Other Dystonias (not shown above) ie
51 = Tardive Dyskinesia
52 = Paroxysmal Choreo-athetosis
53 = Paroxysmal Dystonia
54 = Paroxysmal (Familial)
60 = Not Dystonia, ie controls with :-
61 = HSP
62 = CP
63 = Myoclonus
64 = BET
65 = Peripheral Neuropathy
66 = Ulnar Nerve Palsy
67 = Chorea
68 = ME
69 = MS
70 = PD
71 = Arthrogryposis
72 = SSS (Spasticity secondary to Stroke)
80 = HFS
90 = Undefinable (as this time)
91 = Psychogenic (in nature)

201 prim relates to Q. 18: **Primary Dystonia** (old aetiology)

1 = Idiopathic (ITD)
2 = Dopa Responsive (DRD)
3 = Paroxysmal
4 = Myoclonic

202 sec relates to Q. 18: **Secondary Cause** (old aetiology)

1 = Cerebrovascular Acc (CVA)
2 = Arnold Chiari (Malformation)
3 = Aneurysm
4 = Cerebral Palsy
5 = Drug Induced
6 = MS
7 = MSA
8 = Parkinson's (Disease)
9 = Psychogenic (in nature)
10 = Meningitis / Encephalomyelitis
11 = Shy Drager's (Syndrome)
12 = Stroke (Spasticity)
13 = Tardive Dyskinesia (Dystonia)
14 = Leigh's Disease
15 = Metabolic ??
16 = BET
17 = HFS (unknown cause)
18 = Undiagnosed (or undefinable at this time)
19 = HFS (2nd operation)
20 = SAH (Subarachnoid Haemorrhage)
21 = Stroke (dystonic)
22 = HFS (2nd CVA)
23 = HFS (Aneurysm)
24 = HFS (2nd D.I.)
25 = HFS (2nd MS)

203 notdys Relates to conditions which are **not dystonia**

61 = HSP
62 = CP
63 = Myoclonus
64 = BET
65 = Peripheral Neuropathy
66 = Ulnar Nerve Palsy
67 = Chorea
68 = ME
69 = MS
70 = PD
71 = Arthrogryposis
72 = SSS (Spasticity secondary to Stroke)

204	dist1	relates to Q. 18: **Distribution**	Temp = Distribution

1 = Focal dystonia — FD (abbreviation)
2 = Segmental dystonia — Seg
3 = Multi-focal dystonia — MF
4 = Generalised dystonia — GD
5 = Hemi-dystonia — HD
6 = Undefined spasms
7 = Other neuro disorder
8 = Undiagnosed spasms

205	foc1	relates to Q. 18: **Focal dystonia 1**	Template = Focal

1 = Blepharospasm — BL (abbreviation)
2 = Oromandibular — OMD
3 = Spasmodic Dysphonia (Laryngeal Dystonia) — SD
4 = Spasmodic Torticollis — ST
5 = Retrocollis — RT
6 = Antecollis — AT
7 = Hand (Writers) Cramp — HC or WC
8 = Lingual Dystonia (Tongue not Laryngeal) — LD
9 = Facial Dystonia — Fac
10 = Dystonic Dysphagia — DD
11 = Leg (Foot) Dystonia — FD - leg / ft
12 = Arm (Hand) Dystonia — FD - arm/hd
13 = Hemi-facial Spasm — HFS
14 = Orofacial dystonia — Orofac
15 = Trunk — Trnk

206	foc2	relates to Q. 18: **Focal dystonia 2**	Template = Focal
207	foc3	relates to Q. 18: **Focal dystonia 3**	
208	foc4	relates to Q. 18: **Focal dystonia 4**	

$foctot **Total of Focal Dystonias** - variables foc1, foc2, foc3 and foc4
Multiple Response Set - Range 1 to 15

209	sgmt	relates to Q. 18: **Segmental dystonia**

1 = Cranial
2 = Axial
3 = Brachial
4 = Crural
5 = Craniocervical
6 = Meige's Syndrome (Cranial)
7 = Breughel's Syndrome (Cranial)
8 = Cervical (both focal dystonias in the neck area)

210 side relates to Q. 18: **Side affected**
1 = Left
2 = Right
3 = Both sides
4 = Forward (neck)
5 = Backward (neck)
6 = Tremor (shaking)
7 = Diagonally opp sides
8 = Undefined side

211 onset1 relates to Q. 19: **Year of onset from the interview**
00 = 1900, then includes every year to 98 = 1998

212 age2 **Age at onset**
Compute age2 = onset1 + 1900 - xdate.year(dob). Execute.

213 diag relates to Q. 20: **Year of diagnosis from the interview**
00 = 1900, then includes every year to 98 = 1998

214 age3 **Age at diagnosis**
Compute age3 = diag + 1900 - xdate.year(dob). Execute .

215 diff1 **Difference onset-diag**, ie Years to correct diagnosis
Compute diff1 = age3 - age2. Execute .

NB : The difference in years should be correlated with the original diagnosis. Some patients did not seek early advice as their dystonia was not severe at first. See variable label #286 'del' - ie the reason for any delay in diagnosis. 'diff1' should not be used in isolation but must be seen in context.

216 trig` relates to Q. 20: **Trigger** (this question was asked additionally to Q. 20)
1 = Trauma or Stress
2 = Bereavement
3 = Stroke
4 = Accident (RTA / fall, etc)
5 = Medical (or Dental) Operation
6 = Alcohol or Drug Induced
7 = Other neuro disorder
8 = No definable trigger (Unknown)
9 = Pregnancy / Change of life
10 = Familial
11 = Childhood (type) disease (eg glandular fever)
12 = Industrial injury
13 = R.S.I. (Repetitive Strain Injury)
14 = Heart Attack
15 = Flu or Cortisone Injections
16 = Cancer / Radiotherapy
17 = Action induced / Task Specific

217 fam relates to Q. 21: **Family members**

1 = Yes (possible)
2 = No (definite)
3 = Adopted

218 who2 relates to Q. 21: **Which family members** Temp = Who

1 = Father
2 = Mother
3 = Brother
4 = Sister
5 = Paternal relation
6 = Maternal relation
7 = Son
8 = Daughter
9 = Spouse (wife)
10 = Nephew or Niece

NB : There is no who1 label
who2 relates to dist2
who3 relates to dist3
who4 relates to dist4

219 dist2 relates to Q. 21: **Distribution** Temp = Distribution

1 = Focal dystonia
2 = Segmental dystonia
3 = Multi-focal dystonia
4 = Generalised dystonia
5 = Hemi-dystonia
6 = Undefined spasms
7 = Other neuro disorder
8 = Undiagnosed spasms

220 who3 relates to Q. 21: **Which family** Template = Who
221 dist3 relates to Q. 21: **Distribution** Template = Dist
222 who4 relates to Q. 21: **Which family** Template = Who
223 dist4 relates to Q. 21: **Distribution** Template = Dist

$famtot **Total of Family Members** - variables who2, who3 and who4
Multiple Response Set - Range 1 to 10

$disttot **Total of Family Distribution** - variables dist2, dist3 and dist4
Multiple Response Set - Range 1 to 8

224 med1 relates to Q. 22 & 26 : **Current Medication** BNF page No.
No. 22 : 1991

100 = Drugs which are **Directly related to dystonia**

101 = Amanatadine (Symmetrel, Mantadine)	179
102 = Baclofen (Lioresal)	342
103 = Benzhexol (Artane, Broflex)	181
104 = Benztropine (Cogentin)	180

105 = Bromocriptine (Paroldel)	180
106 = Cannabis Resin (not Nabilone - Cesamet)	xxx / 157
107 = Chlorpromazine (Largactil)	134
108 = Clonazepam (Rivotril)	173
109 = Co-beneldopa (Madopar)	179
110 = Co-careldopa (Sinemet)	179
111 = Dantrolene (Dantrium)	343
112 = Diazepam (Valium, Stesolid, Diazemuls)	343
113 = Haloperidol (Serenace, Haldol, Dozic)	182
114 = Levodopa (Brocadopa, Larodopa)	178
115 = Lysuride Maleate (Revanil)	180
116 = Methixene (Maleate) (Tremonil)	181
117 = Orphenedrine (Biorphen, Disipel)	181
118 = Pimozide (Orap)	138
119 = Primidone	182
120 = Selegiline (Eldepryl)	180
121 = Tetrabenazine (Nitoman)	182
122 = Thioridazine (Melleril)	139
123 = Trifluperazine (Stelazine)	139

200 = Drugs which are **Indirectly related to dystonia**

201 = Amitripyline (Lentizol, Tryptizol)	144
202 = Benorylate (Benoral)	328
203 = Biperiden (Akineton)	181
204 = Buprenorphine (Temgesic)	165
205 = Carbamazepine (Tegretol)	172
206 = Clomipramine (Anafranil)	146
207 = Chlordiazepoxide (Librium)	130
208 = Chlormethiazole (Heminervin)	176
209 = Chlormezanone	131
210 = Clobazam (Frisium)	130
211 = Clomipramine Hydrochloride (Anafranil)	146
212 = Co-codamol (Paracodomal)	161
213 = Co-codaprin	162
214 = Co-dydramol	162
215 = Co-proxamol (Distalgesic)	161
216 = Dexamphetamine (Dexedrine)	151
217 = Diclofenac (Voltarol)	330
218 = Diflunisal (Dolobid)	331
219 = Dihydrocodeine (DF-118)	166
220 = Dipipanone Hydrochloride (Diconal)	167
221 = Dipyridamole (Persantin)	94
222 = Dothiepin (Prothiaden)	146
223 = Doxepin (Sinequan)	146
224 = Equagesic (not NHS)	162
225 = Feldene Gel	344
226 = Flurbiprofen (Froben)	331
227 = Fluvoxamine (Faverin)	151

228 = Hyoscine (Scopodrem)	156
229 = Hypromellose (eye drops, Isopto)	355
230 = Ibuprofen (Nurofen , Brufen, Fenbid)	330
231 = Imipramine Hydrochloride (Tofranil)	146
232 = Indomethacin	331
233 = Lofepramine (Gamanil)	147
234 = Lorazepam	131
235 = Lormetazepam	127
236 = Methadone Hydrochloride	183
237 = Naftidrofuryl (Praxilene)	87
238 = Naproxen (Naprosyn, Nycopren, Synflex)	333
239 = Nitrazepam	126
240 = Paracetamol (Calpol, Disprol, Paldesic)	160
241 = Parafon (Cilag -in KSA)	new
242 = Paroxetine (Seroxat)	151
243 = Pergolide	xii
244 = Phenytoin (Epanutin)	174
245 = Piroxicam (Feldene)	333
246 = Primidone (Mysoline)	174
247 = Procyclidine (Arpicolin, Kemadrin)	181
248 = Propranolol (Inderal)	67
249 = Prozac (Fluoxetine Hydrochloride)	150
250 = Remoxipride (Roxiam) - withdrawn	XII
251 = Risperidone (replacement - Feb '94)	new
252 = Solpadeine (not NHS)	162
253 = Solpadol (Tylex)	163
254 = Sodium Valproate (Epilim)	175
255 = Sulpiride (Dolmatil, Sulpitil)	139
256 = Temazepam	127
257 = Tiaprofenic Acid (Surgam)	334
258 = Trimipramine (Surmontil)	147
259 = Tryptophan	150
260 = Zopiclone (Zimovane)	129

300 = Drugs which are **Not related to dystonia**

301 = Acebutolol (Sectral)	68
302 = Alverine Citrate (Spasmonal)	30
303 = Aminophylline	109
304 = Amlodipine Besylate (Istin)	83
305 = Amoxycillin (Almodan)	191
306 = Antihistamines	360
307 = Aspirin	159
308 = Atenolol (Tenormin, Kalten, Tenif)	68
309 = Beclomethasone (Beconase, Becotide)	360 / 111
310 = Bendrofluazide (thiazide diuretic)	54
311 = Betahistine Hydrochloride (Serc)	155
312 = Betaxolol Hydrochloride (eye drops)	353
313 = Bismuth Chelate (De-nol)	34

314 = Bisoprolol Fumarate (Monocor)	68
315 = Brompheniramine Maleate (Dimotane)	115
316 = Budesonide (Pulmicort)	112
317 = Chlorambucil (Leukeran)	284
318 = Chlorpheniramine Maleate (Piriton)	115
319 = Cholestyramine (Questran)	97
320 = Clonidine (Dixarit)	170
321 = Co-codamol	161
322 = Co-codaprin	162
323 = Codeine Phosphate	37
324 = Co-dydramol	162
325 = Co-proxamol (Distalgesic)	161
326 = Cimetidine (Dyspamet, Tagamet)	32
327 = Cinnarizine (Stugeron)	155
328 = Cyclosporin (Sandimmun)	291
329 = Cyproterone Acetate (Dianette)	388
330 = Cytamen injections	302
331 = Danazol (Danol)	264
332 = Diclofenac (Voltarol)	330
333 = Digoxin (Lanoxin)	52
334 = Diltiazem Hydrochloride (Tildiem)	83
335 = Disodium Etidronate (Didronel)	261
336 = Diuretics (Dyazide, Frusene)	58 / 59
337 = Diuretics (Burinex, Neo-NaClex, Lasix)	60
338 = Domperidone (Motilium)	155
339 = Enalapril (Innovace)	77
340 = Expectorants (various)	123
341 = Fenbufen (Lederfed)	331
342 = Ferrous Gluconate/Sulphate (Ferrocontin)	298
343 = Folic Acid	302
344 = Frebusin	504
345 = Frusemide (Lasix)	56
346 = Gaviscon (Algicon)	27
347 = Glibenclaminde	238
348 = Glyceryl Trinitrate (Suscard, Nitro-lin)	80
349 = HRT (General)	250
350 = Hydroflumethiazide (Hydrenox)	55
351 = Hypromellose (eye drops, Isopto)	355
352 = Ibuprofen (Nurofen , Brufen, Fenbid)	330
353 = Indapamide (Natrilix)	55
354 = Indomethacin (Indocid-R)	331
355 = Isosorbide Mononitrate (Elanten, Ismo)	82
356 = Ispaghula Husk (Isogel,Metamucil,Fybogel)	41
357 = Itraconazole (Sporanox)	220
358 = Ketoprofen (Oruvail)	332
359 = Lactulose	45
360 = Lisinopril (Carace, Zesizil)	78
361 = Loperamide (Imodium)	37
362 = Maxijul	506

363 = Mefenamic Acid (Ponstan)	332
364 = Mesalazine (Asacol)	39
365 = Metformin Hydrochloride	239
366 = Methotrexate Sodium (Arthitrex)	ABPI
367 = Metoclopramide Hydrochloride (Maxolon)	156
368 = Metopropol Tartrate (Betaloc, Lopresor)	69
369 = Migraleve (Migravess, Paramax)	169
370 = Naproxen (Naprosyn, Nycopren, Synflex)	333
371 = Neomycin Sulphate (Otomize)	358
372 = Nifedipine (Adalat, Coracten)	84
373 = Norethiseterone (Primolut N) HRT	255
374 = Oestrogens (Premarin, Prempak)	252
375 = Omeprazole (Losec)	35
376 = Oxazepam	131
377 = Oxitropium Bromide (Oxivent)	107
378 = Oxprenolol (Trasidrex)	70
379 = Oxybutynin Hydrochloride (Ditropan)	279
380 = Oxytetracycline	202
381 = Paracetamol (Calpol, Disprol, Paldesic)	160
382 = Penicillin	188
383 = Phenylbutazone (Butacote)	333
384 = Philocarpine (eye drops)	352
385 = Pindocol (Viskew)	71
386 = Pizotifen (Sanomigran)	170
387 = Pravastatin (Lipostat)	99
388 = Prednisolone (Enteric)	248
389 = Prochlorperazine (Stemetil)	158
390 = Propranolol (Inderal)	67
391 = Quinine sulphate	342
392 = Ranitidine (Zantac)	34
393 = Salbutamol (Ventolin)	103
394 = Senna (Manevac, Senokot)	43
395 = Sertaline (Lustral)	151
396 = Simvastatin (Zocor)	100
397 = Sodium Valproate (Epilim)	175
398 = Sotalol Hydrochloride (Sotacom)	71
399 = Steroids (Nandrolone)	256
400 = Drugs which are **Not related to dystonia**	
400 = Sucralfate (Antepsin)	35
401 = Sulphasalazine (Salazopyrin)	39
402 = Sumatriptin (Imigran - injection)	ABPI
403 = Tamoxifen (Nolvadex)	296
404 = Terbutaline Sulphate (Bricanyl)	104
405 = Tenoxicam (Modiflex)	334
406 = Testosterone (Restandol, Virormone)	255
407 = Theophycline (Slo-phyllin)	109
408 = Thymoxamine (Opilon)	86
409 = Thyroxine (Eltroxin) [micrograms]	242

410 = Tiaprofenic Acid (Surgam)		334
411 = Timolol Maleate (Timoptol)		354
412 = Verapamil (Berkatens, Cordilox, Securon)		83
413 = Vitamins		320
414 = Warfarin (Marevan)		93
415 = Zopiclone (Zimovane)		129

225 dpd1 relates to Q. 22/26: **Dosage (mg) per day**

226 when1 relates to **when** the medication is **taken**

1 = Once a day	(od)
2 = Twice a day	(bd)
3 = 3 x a day	(tds)
4 = 4 x a day	(Qds)
5 = At night	(nocte)
6 = As required	(Prm)
7 = Taken weekly	
8 = Taken monthly	
9 = Injections	(Prm)

227 med2 relates to Q. 22 & 26: **Medication** Template = Med

228 dpd2 relates to Q. 22 & 26: **Dosage (mg) per day** Template = Dose

229 when2 relates to **when** the medication is **taken** Template = When

230 med3 relates to Q. 22 & 26: **Medication** Template = Med

231 dpd3 relates to Q. 22 & 26: **Dosage (mg) per day** Template = Dose

232 med4 relates to Q. 22 & 26: **Medication** Template = Med

233 dpd4 relates to Q. 22 & 26: **Dosage (mg) per day** Template = Dose

234 med5 relates to Q. 22 & 26: **Medication** Template = Med

235 dpd5 relates to Q. 22 & 26: **Dosage (mg) per day** Template = Dose

$medtot **Total medications** - variables med1, med2, med3, med4 and med5
Multiple Response Set - Range 101 to 415

$dpdtot **Total of dosage per day** - variables dpd1, dpd2, dpd3, dpd4 & dpd5
Multiple Response Set - Range 1 to 988

$whentot **Total of when taken** - variables when1 and when2
Multiple Response Set - Range 1 to 9

236 pay relates to Q. 22: **Pay for medication**

1 = Private Prescription
2 = Patient paid NHS
3 = Prescribed FOC (to patient)

237 prev1 relates to Q. 24: **Previous treatments** Template = Prev.

1 = Drugs / Medication
2 = Surgery
3 = Psychiatry
4 = Osteopathy
5 = Chiropractic
6 = Biofeedback
7 = Psychologist (Counselling / Stress Clinic)
8 = Physiotherapy
9 = Aromatherapy
10 = Acupuncture
11 = Yoga
12 = Meditation
13 = Hypnotherapy
14 = Alexander Technique
15 = Wearing a collar
16 = Heat Treatment (Electric Treatment / TENS Machine)
17 = Speech Therapy
18 = Healing Hands (Spiritualism)
19 = Reflexology
20 = Orthopaedic Surgery
21 = Manipulation (under anaesthetic)
22 = Homeopathy (Herbal remedies)
23 = Rheumatology
24 = Relaxation technique
25 = Cortisone injections (or Lignocaine)
26 = Phenol injections
27 = Immunology
28 = Ophthalmology
29 = Hydrotherapy

238 prev2 relates to Q. 24: **Previous treatments** Template = Prev.

239 prev3 relates to Q. 24: **Previous treatments**

240 prev4 relates to Q. 24: **Previous treatments**

241 prev5 relates to Q. 24: **Previous treatments**

$prevtot **Total of previous treatments** - variables prev1 to prev5 inclusive
Multiple Response Set - Range 1 to 29

242 sept1 relates to Q. 25: **Side effects** previous treatments Template = Side ...

1 = Felt ill / more pain
2 = Increased spasms
3 = Lack muscle control
4 = Nausea
5 = Drowsiness
6 = Flu like symptoms
7 = Dysphagia
8 = Feeding by tube
9 = Headaches
10 = Tickling throat
11 = Coughing
12 = Sneezing
13 = Phlegm
14 = Dry eyes
15 = Depression
16 = Ptosis
17 = Difficulty walking
18 = Double vision (blurred vision)
19 = Hallucinations (Confused)
20 = Constipation
21 = Loss of weight
22 = Dry mouth / throat
23 = Elation
24 = Tiredness / Lethargy
25 = Watery eyes
26 = Skin rash
27 = Increased weight
28 = Bruising / bleeding
29 = Dizziness
30 = Memory Loss
31 = Allergic reaction
32 = Insomnia
33 = Addiction
34 = Hospitalisation

243 sept2 relates to Q. 25: **Side effects (prev)** Template = Side ....

$septtot **Total side effects from previous treatments** - variable sept1, sept2
Multiple Response Set - Range 1 to 34

244 last1 relates to Q. 25: **How long it lasts** Template = Last

1 = One day
2 = A couple of days
3 = About a week
4 = Over a week
5 = A month or over
6 = Until stopped taking
7 = Undefined period
8 = 2 - 3 weeks
9 = Up to 2 months

245 com1 relates to Q. 00: **Co-morbidity 1** Info from CAF

100 = Co-morbidity **Directly related to dystonia** Temp = Comorbidity

101 = Anoxic Brain Damage
102 = BET (Benign Essential Tremor)
103 = Chiari Malformation
104 = Chorea
105 = Cerebral Aneurysm (or Carcinoma)
106 = Cerebral Palsy
107 = Death (not related to dystonia)
108 = Gilles de Tourette (Syndrome)
109 = Head Injury
110 = Leigh's Disease
111 = Meningoencephalitis / Encephalomyelitis
112 = Multiple Sclerosis
113 = MSA (Multiple System Atrophy)
114 = Myoclonus
115 = Parkinson's Disease
116 = Pneumococcal Meningitis
117 = Shy Drager Syndrome
118 = Stroke
119 = Spasticity (Subarachinoid Haemorrhage)
120 = Tardive Dyskinesia

200 = Comorbidity **Indirectly related to dystonia**

201 = Agoraphobia
202 = Alcoholism
203 = Anorexia Nervosa
204 = Anxiety Neurosis
205 = Bell's Palsy
206 = Cerebella Ataxia
207 = Depression
208 = Drug Addiction
209 = Drug Induced PD
210 = Epilepsy

211 = Insomnia
212 = Learning Difficulties
213 = Manic Depressive (Psychotic)
214 = PD (as well as dystonia)
215 = Personality Disorder
216 = Post 'psycho surgery' (thalomotomy)
217 = Schizophrenia (psychosis)
218 = Spondylosis, Cervical

300 = Co-morbidity **Not related to dystonia**

301 = Acne
302 = Addison's Disease
303 = Alcoholism
304 = Allergic Rhinitis
305 = Alzheimer's Disease
306 = Anaemia (Iron Deficiency)
307 = Angina
308 = Anxiety Neurosis
309 = Arthritis (unspecific or osteo)
310 = Asthma
311 = Atria Fibrillation
312 = Breast Carcinoma
313 = Bronchial Carcinoma
314 = Bronchitis (Chronic)
315 = Cardiac Failure
316 = Cerebrovascular Dis(ease)
317 = Cervical Myelopathy
318 = Cholecystectomy
319 = Chronic Fatigue Syndrome (ME)
320 = Constipation
321 = CTS (Carpel Tunnel Syndrome)
322 = Deafness
323 = Depression
324 = Diabetes
325 = Diverticular Disease
326 = Endometriosis
327 = Epilepsy
328 = Glaucoma (plus 1 case of Iritis)
329 = Gout
330 = Hay Fever (incl Urticaria)
331 = Hernia (Oesophageal or Hiatus)
332 = Hepatitis - Viral
333 = HRT (Hormone Replacement Therapy)
334 = Hypercholesterol Anaemia
335 = Hypertension
336 = Hysterectomy
337 = IBS (Irritable Bowel Syndrome)
338 = Inflammatory Bowel Disease

339 = Insomnia
340 = Ischaemic Heart Disease
341 = Kidney Transplant (or Renal problems)
342 = Leg Cramps
343 = Lumbar pain
344 = Lymphocytic Leukaemia (Chronic)
345 = Lymphoedema
346 = Lymphoma
347 = Muscular Degeneration
348 = Meniere's (and / or Tinnitus including Vertigo)
349 = Menstrual Symptoms (Pain)
350 = Migraine
351 = Osteoporosis
352 = Paget's Disease
353 = Parotid Tumour (Adenoma)
354 = Peripheral Neuropathy
355 = Peripheral Vascular Disease
356 = Pernicious Anaemia
357 = PMT (Pre-menstrual Tension)
358 = Polymyalgia Rheumatica
359 = Progressive Systemic Sclerosis
360 = Prolactinomaectomy
361 = Prostastic Carcinoma
362 = Psoriasis
363 = Raynaud's Syndrome
364 = Rheumatoid Arthritis
365 = Sandhoff's Disease
366 = Sick Sinus Syndrome
367 = Sinusitis
368 = Surgery, Knee or Hip
369 = Splenectomy
370 = Thrush (Candidiasis)
371 = Thyroid Disease
372 = Ulcer (Peptic also Duodenal)
373 = Ulnar Nerve Palsy
374 = Valvular Heart Disease

246 com2 relates to Q. 00: **Co-morbidity 2** Temp = Comorbidity
247 com3 relates to Q. 00: **Co-morbidity 3** Temp = Comorbidity
248 com4 relates to Q. 00: **Co-morbidity 4** Temp = Comorbidity

$comtot **Total co-morbidity** - variable com1, com2, com3 and com4
Multiple Response Set - Range 101 to 374

249 bef relates to Q. 29: **Bot Tox before**
No of times before the interview or the start of the research
0 = Never been injected before

250	inj1	**Injections from beginning to 31.03.94**	from hospital records
251	inj2	**01.04.94 - 31.03.95**	from files, odd missing data
252	inj3	**01.04.95 - 31.03.96**	from hospital records
253	inj4	**01.04.96 - 30.11.96**	from files, odd missing data
254	inj5	**04.12.96 - 25.03.97**	from hospital records
255	inj6	**01.04.97 - 25.06.97**	from hospital records
256	inj7	**01.07.97 - 31.12.98**	from hospital records

257 curr relates to Q. 29: **Current No. Bot Tox**
No of times up to 01.01.99 - from clinical records up to subject no 885, from subject no 885 onwards - the number of times at date of interview.

258 bod1 relates to Q. 29: **Where in body 1** Template = Body1

1 = Eye Muscles
2 = Face Muscles
3 = Neck or Shoulder (Muscles)
4 = Throat / Larynx
5 = Arm / Hand
6 = Leg / Foot
7 = Abdomen (Trunk or Back)
8 = Mouth
9 = Jaw

259 bod2 relates to Q. 29: **Where in body 2** Template = Body1

$bodtot **Total sites in body** - variable bod1 and bod2
Multiple Response Set - Range 1 to 9

260 sebt1 relates to Q. 29: **Side effects (Bot Tox) 1** Temp = Side ...
Same codes used as in # 242 'sept1'

261 sebt2 relates to Q. 29: **Side effects (Bot Tox) 2** Temp = Side ...

$sebttot **Total Side Effects (Bot Tox)** - variable sebt1 and sebt2
Multiple Response Set - Range 1 to 34

262 last2 relates to Q. 29: **How long it lasts** Temp = Last

1 = One day
2 = A couple of days
3 = About a week
4 = Over a week
5 = A month or over
6 = Until treatment stop
7 = Undefined period
8 = 2 - 3 weeks
9 = Up to 2 months

TDS Questionnaire : Q're No. 21

263 talk relates to Q. 30: **Talked to others**

1 = Never
2 = Rarely
3 = Occasionally
4 = Often
5 = Frequently
6 = Once only
7 = One person only

264 what relates to Q. 31: **Under what circumstances**

1 = At BT clinic
2 = In hospital
3 = At home
4 = On the phone
5 = At TDS meetings
6 = Never talked
7 = At work

265 coun1 relates to Q. 32: **Counselling**

1 = Yes
2 = No
3 = Don't know

266 coun2 TDS Counselling Service **used as of 31.12.98**

1 = Full counselling
2 = With family members
3 = One-off session only
4 = One-off with family

267 when relates to Q. 32: **When counselling**

1 = When first diagnosed
2 = If required / needed
3 = When required / need
4 = Not required by me
5 = Don't know

268 whom relates to Q. 32: **Who counsels**

1 = By the doctor (neurologist)
2 = By a nurse (practitioner)
3 = By a social worker (Social Services)
4 = By someone who knows (about it)
5 = By someone with it
6 = By someone in NHS
7 = By someone from TDS
8 = Don't know (not answered previously)

269 tds relates to Q. 33: **Heard of TDS**

1 = Another member
2 = Relative or friend
3 = National T.V. or Radio
4 = Newspaper / magazine
5 = Doctor or Hospital or Clinic
6 = Thru this research
7 = Leaflets / Adverts (North East only)
8 = Can't remember
9 = Disability Group
10 = Library or CAB (Citizens Advice Bureau)
11 = TTTV 1 in 1994
12 = TTTV 2 in 1997
13 = BBC TV (NE) in 1997

270 mem relates to Q. 34: **Member of TDS**

1 = Yes
2 = No
3 = No longer a member (dropped out)
4 = Joined since study (interviewed)
5 = Member of MSS, PDS (etc)
6 = Deceased member
7 = Dropped out and re-joined
8 = Joined by RC 1
9 = Joined for WBS
10= Joined by RC 2

271 join1 relates to Q. 35: **Year & Month when joined** (or re-joined)
From 8302 onwards only valid

272 join2 relates to Q. 35: **Year joined TDS**
From 83 to 99 only

273 numb **TDS Membership Number** (from HQ records)
0 = Number unknown

274 why relates to Q. 35: **Why joined TDS**

1 = To find out more (about dystonia)
2 = To meet and talk (to others / support)
3 = To read the literature
4 = To attend TDS (meetings)
5 = To receive the newsletter
6 = To get information (in general)
7 = To get answers (to my questions)
8 = Don't know
9 = To give support

275 most relates to Q. 36: **Most useful aspect**

1 = The newsletter
2 = Information (in general)
3 = Getting answers (to my questions)
4 = Being part of SHG (activities)
5 = Contact with others (people)
6 = News about research
8 = Don't know

276 shg relates to Q. 37: Self-Help Groups

1 = Teesside SHG
2 = Tyneside SHG
3 = Cumbria SHG
4 = Yorkshire Area (Hull, Leeds, Sheffield, etc)
5 = Member of another SHG
6 = TDS member, not SHG
7 = On mailing list only
8 = No contact (requested)
9 = Darlington SHG - 1998
10 = Sunderland SHG - 1998
11 = Durham SHG - 1998

277 get relates to Q. 38: **Get most out of TDS**

1 = Feeling of belonging
2 = Sharing experience
3 = Talking to others
4 = Social contact
5 = Reading the newsletter
= Finding out others (about people)
= More knowledge (about condition)
= Don't know

278 news relates to Q. 38: **Newsletter**

1 = Medical articles
2 = Latest research
3 = Letters to Editor (other people)
4 = Contact addresses
5 = News - other groups
6 = Overseas information
7 = All of it
8 = Don't know
9 = Can't read

279 aim1 relates to Q. 39: **National aim** Template = Aim

1 = Contact with others (help and support)
2 = Raising awareness (medical and public)
3 = Raising funds
4 = Finding a cure
5 = Funding research
6 = Giving information
7 = Counselling / Welfare
8 = Don't know
9 = Getting treatment
10 = DNP (more required)

280 aim2 relates to Q. 39: **Local aim** Template = Aim

Clinical Features Questionnaire : Q're No. 30

This questionnaire on the "Clinical Features of Dystonia" is divided into five parts :-

1. Contact with Medical Services - Questions 01 to 13
2. Clinical Features of your Dystonia - Questions 14 to 20
3. Nature of your Accommodation - Questions 21 to 23
4. Questionnaire on Torticollis - Questions 24 to 25
5. Geste Antagoniste - Questions 26 to 32

281 date30 **Date of Q're 30** dd.mm.yy 8

282 onset2 relates to Q. 01: **Year of onset from the questionnaire**
24 = 1924, then includes every year to 98 = 1998

283 diff2 **Difference between onset1 and onset 2** (in years)

Onset 1 date was obtained by interview (records)
Onset 2 date was obtained by questionnaire

-5 shows a questionnaire bias
0 shows no difference
+5 shows an interview bias

Compute diff2 = onset1 - onset2. Execute .

284 told relates to Q. 02: **Told of diagnosis** (year)
Different to being diagnosed, ie being told - compare to # 213 'diag'

285 diff6 **Difference between being diagnosed and being told**

-18 = 18 years before being told
- 8 = 8 years before being told
- 3 = 3 years before being told
0 = Diagnosed and told in same year
1 = 1 years error
3 = 3 years error
6 = 6 years error
92 = Never told (of diagnosis) since 1992
94 = Never told (of diagnosis) since 1994
96 = Never told (of diagnosis) since 1996

Compute diff6 = diag - told. Execute .

286 del relates to Q 03: **Delay in diagnosis**

1 = Dr, unaware of condition / lack of knowledge
2 = Mis-diagnosed
3 = Mis-diagnosed & treated for .....
4 = Mis-diagnosed CP/MS/PD
5 = Did not seek advice
6 = Diagnosed other (neuro disorder)
7 = Unknown Cause / Reason
8 = No (long) delay (NHS waiting list)
9 = Waiting for appointment (to be diagnosed)
10 = Went into remission

287 doc relates to Q 04: **Who diagnosed** (dystonia)

1 = Own G.P.
2 = Hospital Dr / Neuro.
3 = Private Dr / Neuro.
4 = Psychiatrist
5 = ENT Surgeon
6 = Neuro Surgeon
7 = Ophthalmologist
8 = Psychotherapist
9 = Self-diagnosed (family)
10 = Psychopharmacologist
11 = Speech Therapist
12 = Rheumatologist
13 = Physiotherapist
14 = Thru' child 1st
15 = Urologist

288 evnt1 relates to Q 05: **Major event** Template = Yes / No

1 = Yes
2 = No

289 evnt2 relates to Q 05: **Type of major event** Template = Event

1 = Major head injury
2 = Other neuro disorder
3 = Cerebella Ataxia
4 = Minor head injury
5 = Stress caused by .....
6 = Fall
7 = Stroke / Cere / SAH
8 = Non related condition
9 = Bereavement
10 = Surgery
11 = Divorce

12 = Accident (work / home)
13 = Severe Migraines
14 = RTA (Road Traffic Accident)
15 = Menopause
16 = RSI (Repetitive Strain Injury)
17 = Heart Attack
18 = Anxiety
19 = Measles
20 = Pregnancy (incl one still birth)
21 = Exhaust / stress at work
22 = Drug Overdose (Tranquillisers)
23 = Flu Injections
24 = Gave up smoking
25 = W.W.II (difficult birth / bomb blast)
26 = Schizophrenia
27 = Alcoholism
28 = Hereditary / familial
29 = Don't know
30 = Chemical exposure
31 = Physical strain
32 = Trapped nerve
33 = Born with it
34 = Puberty
35 = Use of drugs
36 = Slipped disc
37 = Viral infection

290	caus	relates to Q 06: **What caused it** (your dystonia)	Template = Event
291	ment1	relates to Q 07: **Suggest it was mental ?**	Temp = Yes / No
292	gp	relates to Q 08: **G.P. knows enough ?**	Temp = Yes / No
293	drug	relates to Q 09: **Drugs - Yr started** 0 = Unknown year	
294	eff1	relates to Q 09: **Effect of drugs** 1 = Better 2 = Unchanged 3 = Worse	Template = Effect
295	surg	relates to Q 09: **Surgery - Yr started**	Template = Year
296	eff2	relates to Q 09: **Effect of surgery**	Template = Effect
297	physio	relates to Q 09: **Physiotherapy - Yr started**	
298	eff3	relates to Q 09: **Effect of physiotherapy**	
299	osteo	relates to Q 09: **Osteopathy - Yr started**	
300	eff4	relates to Q 09: **Effect of osteopathy**	
301	chiro	relates to Q 09: **Chiropractic - Yr started**	

302	eff5	relates to Q 09: **Effect of chiropractic**	Template = Effect
303	biof	relates to Q 09: **Biofeedback - Yr started**	Template = Year
304	eff6	relates to Q 09: **Effect of biofeedback**	
305	couns	relates to Q 09: **Counselling - Yr started**	
306	eff7	relates to Q 09: **Effect of counselling**	
307	psycho	relates to Q 09: **Psychotherapy - Yr started**	
308	eff8	relates to Q 09: **Effect of psychotherapy**	
309	acup	relates to Q 09: **Acupuncture - Yr started**	
310	eff9	relates to Q 09: **Effect of acupuncture**	
311	yoga	relates to Q 09: **Yoga - Yr started**	
312	eff10	relates to Q 09: **Effect of yoga**	
313	medi	relates to Q 09: **Meditation - Yr started**	
314	eff11	relates to Q 09: **Effect of medication**	
315	hypn	relates to Q 09: **Hypnotherapy - Yr started**	
316	eff12	relates to Q 09: **Effect of hypnotherapy**	
317	alex	relates to Q 09: **Alexander Tech - Yr started**	
318	eff13	relates to Q 09: **Effect of Alexander Technique**	
319	botox	relates to Q 09: **Bot Tox therapy - Yr started**	
320	eff14	relates to Q 09: **Effect of Bot Tox therapy**	

0 = Too early to define
1 = Better
2 = Unchanged
3 = Worse

321 other1 relates to Q 09: **Other treatments - Yr started**

322 other2 relates to Q 09: **Type of treatment**

1 = Faith Healing (2 x Spirit; 2 x Faith; 1 x Prayer)
2 = Personal Method (1 x Drugs; 2 x Unknown, 1 x Routine
3 = Collar (Wearing) and 1 x Champissape ??)
4 = Homeopathy
5 = Aromatherapy
6 = Manipulation
7 = Speech Therapy
8 = Herbal remedies
9 = Reflexology
10 = Traction
11 = Relaxation Tapes
12 = Heat / Elec Stimulation (1 x Heat and 1 x Elec)
13 = TENS Machine

323	eff15	relates to Q 09: **Effect of other treatments**	Template = Effect
324	maj	relates to Q 10: **Major (physical) illness**	Template = Yes / No

325 phys1 relates to Q 10: **Type of physical illness 1**

1 = Achilles Tendon
2 = Adrenal glands (removed)
3 = Alcoholism
4 = Appendicitis (ectomy)
5 = Arthritis (unspecified and osteo)
6 = Asthma
7 = Bomb Blast
8 = Bones - fractures, etc
9 = Brain - operation
10 = Brain Tumour
11 = Bronchitis
12 = Cancer (unspecified)
13 = Cardiovascular Dis. (inc Angina, Bypass, etc)
14 = Cerebella Ataxia
15 = Cervical Spondylosis
16 = CP
17 = Crohn's Disease
18 = Dental Operation
19 = Diphtheria
20 = Diverticulitis
21 = Ectopic Pregnancy
22 = Endometriosis
23 = Epilepsy
24 = Eyelid surgery
25 = Gall stones / bladder (removed)
26 = Gastro-enteritis
27 = Glandular Fever
28 = Glaucoma
29 = Growth removed (various)
30 = Guillam Barre Syndrome
31 = Heart attack
32 = Hepatitis
33 = Hernia - Hiatus
34 = Hip Replacement
35 = Hydrocephalus
36 = Hypertension
37 = Hysterectomy (incl Tubal Tie)
38 = Inf. Bowel Disease
39 = Infantile Paralysis (Polio)
40 = Kidney Transplant (stones)
41 = Knee (Joint) Operation
42 = Leg / Arm / Hand Operation
43 = Malnutrition
44 = Mastoid Operation
45 = Meningitis
46 = Migraine
47 = MS

48 = Neck - Operation (incl HFS operation)
49 = Osteoporosis (brittle bones)
50 = Otitis Media
51 = Paralysed - both legs
52 = PD
53 = Pneumonia
54 = Prolapse Operation
55 = Prostate removed
56 = Rheumatic Fever
57 = RTA (incl stabbed, ladder fall, assault, accident)
58 = SAH
59 = Sarcoid problems
60 = Scarlet Fever
61 = Shingles
62 = Skin Complaint
63 = Slipped Disc
64 = Stroke
65 = Thyroid (ectomy)
66 = TB
67 = Throat Cancer
68 = Throat Operation
69 = Tinnitus / Vertigo
70 = Tonsillitis
71 = Tracheotomy
72 = Ulcer
73 = Unspecified Operation

326	year1	relates to Q 10: **Year of Illness 1**	Template = Year
327	phys2	relates to Q 10: **Type of physical illness 2**	Template = Physical
328	year2	relates to Q 10: **Year of Illness 2**	Template = Year, etc
329	phys3	relates to Q 10: **Type of physical illness 3**	
330	year3	relates to Q 10: **Year of Illness 3**	
331	phys4	relates to Q 10: **Type of physical illness 4**	
332	year4	relates to Q 10: **Year of Illness 4**	

$phystot **Total physical illnesses** - variable phys1 to phys4 inclusive
Multiple Response Set - Range 1 to 73

$yr1tot **Total Year of Onset** - variable year1 to year4 inclusive
Multiple Response Set - Range 0 to 98

333	ment2	relates to Q 11: **Psychiatric problems ?**	Temp = Yes / No

334 psyc1 relates to Q 11: **Type of psychiatric problem 1** Temp = Psychiatric

1 = Depression
2 = Anorexia
3 = Anxiety
4 = 'Nervous Breakdown'
5 = Tremor (of head)
6 = Imagined Dystonia
7 = Paranoia
8 = Schizophrenia
9 = Amnesia
10 = Agoraphobia
11 = Loss of speech
12 = Epilepsy
13 = Panic Attacks
14 = Depression and Anxiety
15 = Alcoholism
16 = Obsessive Neurosis

335 year5 relates to Q 11: **Year of illness 5** Template = Year

336 tret1 relates to Q 11: **Treatment received 1** Template = Treat.

1 = Medication / Drugs
2 = Surgery
3 = Physiotherapy
4 = Osteopathy
5 = Chiropractic
6 = Biofeedback
7 = Counselling
8 = Psychotherapy
9 = Acupuncture
10 = Yoga
11 = Meditation
12 = Hypnotherapy
13 = Alexander Technique
14 = Bot Tox Therapy
15 = Shock treatment (ECT)
16 = Injections
17 = Speech Therapy

337 psyc2 relates to Q 11: **Type of psychiatric problem 2** Temp = Psychiatric
338 year6 relates to Q 11: **Year of illness 6** Temp = Year
339 tret2 relates to Q 11: **Treatment received 2** Temp = Treatment

$psyctot **Total psychiatric illnesses** - variable pysc1 and pysc2
Multiple Response Set - Range 1 to 16

$trettot **Total psychiatric treatment** - variable tret1 and tret2
Multiple Response Set - Range 1 to 17

$yrstot **Total date (year) of illnesses** - variable year5 and year6
Multiple Response Set - Range 0 to 98

340	anti	relates to Q 12: **Antidepressants**	Template = Yes / No
341	anxi	relates to Q 13: **Control of anxiety**	Template = Yes / No
342	eyel	relates to Q 14: **Year affected (Left eye)**	Template = Year
343	eyer	relates to Q 14: **Year affected (Right eye)**	
344	eyes	relates to Q 14: **Year affected (Both eyes)**	
345	facer	relates to Q 14: **Year affected (R side of face)**	
346	facel	relates to Q 14: **Year affected (L side of face)**	
347	faces	relates to Q 14: **Year affected (Both sides of face)**	
348	mouth	relates to Q 14: **Year affected (mouth)**	
349	jaw	relates to Q 14: **Year affected (jaw)**	
350	tongue	relates to Q 14: **Year affected (tongue)**	
351	vocal	relates to Q 14: **Year affected (vocal chords)**	
352	speech	relates to Q 14: **Year affected (speech)**	
353	armr	relates to Q 14: **Year affected (right arm)**	
354	handr	relates to Q 14: **Year affected (right hand)**	
355	arml	relates to Q 14: **Year affected (left arm)**	
356	handl	relates to Q 14: **Year affected (left hand)**	
357	writ1	relates to Q 14: **Year affected (right hand)**	
358	writ2	relates to Q 14: **Year affected (left hand)**	
359	writ3	relates to Q 14: **Affected only when writing ?**	Template = Yes / No
360	trunk	relates to Q 14: **Year affected (trunk)**	
361	legr	relates to Q 14: **Year affected (right leg)**	
362	footr	relates to Q 14: **Year affected (right foot)**	
363	legl	relates to Q 14: **Year affected (left leg)**	
364	footl	relates to Q 14: **Year affected (left foot)**	
365	neck	add variable : **Year affected (neck)** - deduced from Q'res	
366	move1	relates to Q 15: **Involuntary movements**	Template = Yes / No
367	move2	relates to Q 15: **Type of movement 2**	Temp = Movement

1 = Regular trembling
2 = Quick jerking
3 = Slight twitching
4 = Slow pulling / turning
5 = Eyelids shut
6 = Occasional trembling
7 = Cramps / Cramping
8 = Mixture of 1 to 4
9 = Rhythmical (continuous)

10 = Voice Whispers
11 = Stiffness (Spasticity / Rigidity)
12 = Sudden Jerk (once only at a time)
13 = Action Induced / Task Specific

368 move3 relates to Q 15: **Type of movement 2** Temp = Movement
369 move4 relates to Q 15: **Type of movement 3**

$movetot **Total types of movement** - variable move2, move3 and move4
Multiple Response Set - Range 1 to 13

370 seve relates to Q 16: **How severe** is your dystonia

0 = Not severe at all
10 = Very severe

371 cont1 relates to Q 17: **Degree of control** Template = Control

0 = No control at all
10 = Complete control

372 cont2 relates to Q 18: **Degree of control** Template = Control

373 pain3 relates to Q 19: **Pain at present** Template = Yes / No

374 pain4 relates to Q 19: **Where pain 1** Template = Body2

1 = Eye Muscles
2 = Face Muscles
3 = Neck Muscles
4 = Shoulder Muscles
5 = Arm Muscles
6 = Hand Muscles
7 = Leg Muscles
8 = Foot Muscles
9 = Trunk Muscles
10 = Back Muscles
11 = Mouth Muscles
12 = Jaw Muscles
13 = Head
14 = Throat / Larynx

375 pain5 relates to Q 19: **Where pain 2** Template = Body2
376 pain6 relates to Q 19: **Where pain 3**
377 pain7 relates to Q 19: **Where pain 4**
378 pain8 relates to Q 19: **Where pain 5**

$paintot **Total where pain** - variable pain4 to pain8 inclusive
Multiple Response Set - Range 1 to 13

379 pain9 relates to Q 19: **Severity of pain**

0 = No pain
10 = Very severe pain

380 pain10 relates to Q 19 : **How frequent pain**

1 = Infrequently
2 = Often
3 = Continuously

381 spon1 relates to Q 20: **Spontaneous improvement** Template = Yes / No

382 spon2 relates to Q 20: **Year of improvement 1** Template = Year

383 spon3 relates to Q 20: **How long** Template = Long

1 = A few days only
2 = Several weeks
3 = 4 weeks
4 = More than a month
5 = 6 - 8 weeks
6 = 2 months
7 = 3 months
8 = 6 months to a year
9 = Over a year
10 = 2 years
11 = Several years
12 = 4 years
13 = Over 5 years
14 = 6 years
15 = 8 years
16 = 15 years
17 = 27 years
18 = Still in remission

384 spon4 relates to Q 20: **Partial or complete 1**

1 = Partial
2 = Complete

385 spon5 relates to Q 20: **Year of improvement 2** Temp = Year
386 spon6 relates to Q 20: **How long** Temp = Long
387 spon7 relates to Q 20: **Partial or complete 2**

$yeartot **Total years of improvement** - variable spon2 and spon5
Multiple Response Set - Range 00 to 98

$longtot **Total how long** - variable spon3 and spon6
Multiple Response Set - Range 1 to 18

$parttot **Total partial / complete** - variable spon4 and spon7
Multiple Response Set - Range 1 or 2

388 live1 relates to Q 21: **Who do you live with ?**

1 = Alone
2 = With spouse
3 = With parents
4 = With children
5 = With sibling
6 = With aunt
7 = Girl / Boy Friend (Live in Partner)

389 live2 relates to Q 22: **Type of accommodation**

1 = House
2 = Flat
3 = Bungalow
4 = Terraced Cottage
5 = B & B
6 = Prison
7 = Residential Home

390 live3 relates to Q 23: **Suitable** for dystonia Template = Yes/No

391 live4 relates to Q 23: Ways it does **not meet your needs**

1 = Stairs (too many or difficult)
2 = No shower + stairs (too many)
3 = No WC downstairs
4 = No help at home
5 = More adaptations needed
6 = Doors too small (for wheelchair)
7 = House too big (to keep clean)
8 = Outside Steps (Wheelchair)
9 = Being adapted (lift / handrails, etc) now
10 = (have to) Sleep Downstairs

Torticollis Questionnaire : Q're No 40

392 date40 **Date of Q're 40** dd.mm.yy 8

393 pstn1 relates to Q 24: **Position of head** Numeric 4.0

1 = Chin turns to the side
2 = Ear tilts sideways towards shoulder
3 = Head bends forward (chin pointing down)
4 = Heads bends backwards (chin pointing up)

394 pstn2 relates to Q 25: **Direction of head**

10 = Right turn or tilt
11 = Right turn
12 = Right tilt
20 = Left turn or tilt
21 = Left turn
22 = Left tilt
3 = Backwards (chin pointing up)
4 = Forwards (chin pointing down)
5 = Right and backwards
6 = Right and forwards
7 = Left and backwards
8 = Left and forwards

395 gest1 relates to Q 26: **Geste Antagoniste** Temp = Yes / No

396 gest2 relates to Q 26: **Still effective ?**

1 = Yes
2 = No
3 = Not necessary now (in remission)

397 gest3 relates to Q 26: If No, **how many years ?**
0 = Unknown no of years

398 gest4 relates to Q 27: **Describe in detail**

1 = Written description (attached / enclosed)

399 hand1 relates to Q 28: **Which hand** (do you use) ?

1 = Right hand NB : Other questions are Left (1) then Right (2)
2 = Left hand
3 = Either (both) hand

400 true1 relates to Q 29: **Where ?**

1 = Front of my body
2 = Back of my body
3 = Side of face
4 = Different (on different) occasions

401 true2 relates to Q 30: **How ?**

1 = Lightly touch
2 = Pull hard
3 = Push hard
4 = Different (on different) occasions

402 true3 relates to Q 31: **Moving when ?**

1 = Before
2 = At the same time
3 = After
4 = 1 or 3 (sometimes)

403 best relates to Q 32: **Feeling ?**

1 = Being pulled
2 = Being pushed
3 = Both at times

Living with Dystonia Questionnaire : Q're No. 50

This questionnaire is a composite of 4 distinct questionnaires :-

1. A Functional Disability Questionnaire (FDQ): Pages 1 and 2
2. A Body Concept Scale (BCS) : Pages 3 and 4
3. The Beck Depression Inventory (BDI) : Pages 5, 6 and 7
4. Rosenberg's Self-Esteem Scale (SES) : Page 8

404 date50 **Date of Q're 50** dd.mm.yy 8

405 fdq1 relates to Q 01: **Dressing** (Self Care) Temp = Affected

0 = Not applicable
1 = Not at all affected
2 = Mildly affected
3 = Moderately affected
4 = Severely affected

406 fdq2 relates to Q 02: **Housework** (Physical)
407 fdq3 relates to Q 03: **Television** (Leisure)
408 fdq4 relates to Q 04: **Running** (Physical)
409 fdq5 relates to Q 05: **Transport** (Social)
410 fdq6 relates to Q 06: **Writing** (Leisure)
411 fdq7 relates to Q 07: **Conversation** (Social)
412 fdq8 relates to Q 08: **Carrying** (Physical)
413 fdq9 relates to Q 09: **Restaurants or pubs** (Social)
414 fdq10 relates to Q 10: **Brushing teeth** (Self Care)
415 fdq11 relates to Q 11: **Reading** (Leisure)
416 fdq12 relates to Q 12: **Walking** (Physical)
417 fdq13 relates to Q 13: **Intercourse** (Physical)
418 fdq14 relates to Q 14: **Driving** (Physical)
419 fdq15 relates to Q 15: **Washing** (Self Care)
420 fdq16 relates to Q 16: **Eating** (Social)
421 fdq17 relates to Q 17: **Dinner parties** (Social)
422 fdq18 relates to Q 18: **Typing** (Physical)
423 fdq19 relates to Q 19: **Hobbies** (Physical)
424 fdq20 relates to Q 20: **Crossing roads** (Social)

425 fdq21 relates to Q 21: **Shaving** face (if male - Self Care)
or putting on **make up** (if female - Self Care)

426 fdq22 relates to Q 22: **Drinking** (Social)
427 fdq23 relates to Q 23: **Riding a bicycle** (Physical)
428 fdq24 relates to Q 24: **Theatre** (Social)
429 fdq25 relates to Q 25: **Coordination** (Self Care)
430 fdq26 relates to Q 26: **Sports** (Physical)
431 fdq27 relates to Q 27: **Stairs** (Physical)

NB : 3 subjects had data removed, as they were incomplete and skewing scores.

ESD No. 74 - variable fdq7 was removed.
ESD No. 171 - variables fdq1, fdq2 and fdq3 were removed.
ESD No. 433 - variables fdq1 to fdq12 (inclusive) were removed.

432 fdqall FDQ - **Total all scores**

Compute fdqall = fdq1 + fdq2 + fdq3 + fdq4 + fdq5 + fdq6 + fdq7 + fdq8 + fdq9 + fdq10 + fdq11 + fdq12 + fdq13 + fdq14 + fdq15 + fdq16 + fdq17 + fdq18 + fdq19 + fdq20 + fdq21 + fdq22 + fdq23 + fdq24 + fdq25 + fdq26 + fdq27. Execute .

27 = All are 'not at all affected'
54 = Average mildly affected
81 = Average, moderately affected
108 = Average, severely affected

433 fdqsoc FDQ - **Social Scores**

Compute fdqsoc = fdq5 + fdq7 + fdq9 + fdq16 + fdq17 + fdq20 + fdq22 + fdq24. Execute .

8 = All are 'not at all affected'
16 = Average mildly affected
24 = Average, moderately affected
32 = Average, severely affected

434 fdqphy FDQ - **Physical Scores**

Compute fdqphy = fdq2 + fdq4 + fdq8 + fdq12 + fdq13 + fdq19 + fdq27 . Execute .

7 = All are 'not at all affected'
14 = Average, mildly affected
21 = Average, moderately affected
28 = Average, severely affected

NB : Physical Scores has removed 14 (Driving), 18 (Typing), 23 (Riding a bicycle) and 26 (Sports) from the other categories, as experience has shown these generate high numbers of 0 (not applicable) scores.

435 fdqsel FDQ - **Self Care Scores**

Compute fdqsel = fdq1 + fdq10 + fdq15 + fdq21 + fdq25. Execute .

5 = All are 'not at all affected'
10 = Average, mildly affected
15 = Average, moderately affected
20 = Average, severely affected

436 fdqlei FDQ - **Leisure Scores**
Compute fdqlei = fdq3 + fdq6 + fdq11. Execute .

3 = All are 'not at all affected'
6 = Average, mildly affected
9 = Average, moderately affected
12 = Average, severely affected

437 fdqoth FDQ - **Other Physical Scores**

Compute fdqoth = fdq2 + fdq4 + fdq8 + fdq12 + fdq13 + fdq14 + fdq18 + fdq19 + fdq23 + fdq26 + fdq27 . Execute .

11 = All are 'not at all affected'
22 = Average, mildly affected
33 = Average, moderately affected
44 = Average, severely affected

NB : Other Physical Scores has had 14 (Driving), 18 (Typing), 23 (Riding a bicycle) and 26 (Sports) included along with the other categories. This sub-set must <u>not</u> be used in averaging scores and comparing with other sub-set averages.

In 'phddata.sav', the 'not applicable' scores are :-

fdq #	Number	out of	%
14 (Driving)	150	333	45.0%
18 (Typing)	153	333	45.9%
23 (Riding a bicycle)	138	333	41.4%
26 (Sports)	91	333	27.3%

438 disa relates to Q 27 : **Disabling** (re daily activities)

0 = Not at all disabling
10 = Very disabling

439 unco relates to Q 27 : **Uncomfortable** (in social situations)

0 = Not at all uncomfortable
10 = Very uncomfortable

440 bcs1 relates to Graceful vs Awkward Temp = bcs

1 = Negative, very well described
2 = Negative, fairly well described
3 = Negative, only slightly described
4 = Equally descriptive or irrelevant
5 = Positive, only slightly described
6 = Positive, fairly well described
7 = Positive, very well described

441	bcs2	relates to Lethargic	vs	Energetic
442	bcs3	relates to Swift	vs	Sluggish
443	bcs4	relates to Calm	vs	Agitated
444	bcs5	relates to Ugly	vs	Beautiful
445	bcs6	relates to Rigid	vs	Flexible
446	bcs7	relates to Fit	vs	Unfit
447	bcs8	relates to Unbalanced	vs	Balanced
448	bcs9	relates to Steady	vs	Unsteady
449	bcs10	relates to Weak	vs	Strong
450	bcs11	relates to Relaxed	vs	Tense
451	bcs12	relates to Masculine	vs	Feminine (opposite if feminine subject)
452	bcs13	relates to Slow	vs	Fast
453	bcs14	relates to Poised	vs	Unpoised
454	bcs15	relates to Healthy	vs	Sick
455	bcs16	relates to Clumsy	vs	Well-coordinated
456	bcs17	relates to Straight	vs	Twisted
457	bcs18	relates to Mobile	vs	Immobile
458	bcs19	relates to Flawed	vs	Perfect
459	bcs20	relates to Uncontrollable	vs	Controllable
460	bcs21	relates to Active	vs	Passive
461	bcs22	relates to Delicate	vs	Robust

462 bcsall **BCS - Total of all scores**

Compute bcsall = bcs1 + bcs2 + bcs3 + bcs4 + bcs5 + bcs6 + bcs7 + bcs8 + bcs9 + bcs10 + bcs11 + bcs12 + bcs13 + bcs14 + bcs15 + bcs16 + bcs17 + bcs18 + bcs19 + bcs20 + bcs21 + bcs22. Execute .

22 = Extremely negative
88 = Equally distributed
154 = Extremely positive

463 bcsspe **BCS - Speed / Strength Scores**

Compute bcsspe = bcs3 + bcs7 + bcs10 + bcs13 + bcs15 + bcs21 + bcs22. Execute .

7 = Extremely negative
28 = Equally distributed
49 = Extremely positive

NB : The Body Concept Scale Coding Frame is shown on page 53 (overleaf)

BCS Coding Frame

Graceful	1 : 2 : 3 : 4 : 5 : 6 : 7	Awkward
Lethargic	7 : 6 : 5 : 4 : 3 : 2 : 1	Energetic
Swift	1 : 2 : 3 : 4 : 5 : 6 : 7	Sluggish
Calm	1 : 2 : 3 : 4 : 5 : 6 : 7	Agitated
Ugly	7 : 6 : 5 : 4 : 3 : 2 : 1	Beautiful
Rigid	7 : 6 : 5 : 4 : 3 : 2 : 1	Flexible
Fit	1 : 2 : 3 : 4 : 5 : 6 : 7	Unfit
Unbalanced	7 : 6 : 5 : 4 : 3 : 2 : 1	Balanced
Steady	1 : 2 : 3 : 4 : 5 : 6 : 7	Unsteady
Weak	7 : 6 : 5 : 4 : 3 : 2 : 1	Strong
Relaxed	1 : 2 : 3 : 4 : 5 : 6 : 7	Tense
Masculine	1 : 2 : 3 : 4 : 5 : 6 : 7	Feminine
Slow	7 : 6 : 5 : 4 : 3 : 2 : 1	Fast
Poised	1 : 2 : 3 : 4 : 5 : 6 : 7	Unpoised
Healthy	1 : 2 : 3 : 4 : 5 : 6 : 7	Sick
Clumsy	7 : 6 : 5 : 4 : 3 : 2 : 1	Well-Coordinated
Straight	1 : 2 : 3 : 4 : 5 : 6 : 7	Twisted
Mobile	1 : 2 : 3 : 4 : 5 : 6 : 7	Immobile
Flawed	7 : 6 : 5 : 4 : 3 : 2 : 1	Perfect
Uncontrollable	7 : 6 : 5 : 4 : 3 : 2 : 1	Controllable
Active	1 : 2 : 3 : 4 : 5 : 6 : 7	Passive
Delicate	7 : 6 : 5 : 4 : 3 : 2 : 1	Robust

464 bcspos **BCS - Postural / Movement related**

Compute bcspos = bcs1 + bcs6 + bcs8 + bcs9 + bcs16 + bcs17 + bcs20. Execute .

7 = Extremely negative
28 = Equally distributed
49 = Extremely positive

465 bcseva **BCS - Evaluative / Aesthetic Scores**

Compute bcseva = bcs2 + bcs5 + bcs14 + bcs19. Execute .

4 = Extremely negative
16 = Equally distributed
28 = Extremely positive

466 bcsten **BCS - Tension scores**

Compute bcsten = bcs4 + bcs11. Execute .

2 = Extremely negative
8 = Equally distributed
14 = Extremely positive

467 disf relates to **Disfigurement**
0 = Not disfigured at all
10 = Extremely disfigured

468 bdi1 relates to Q 01: **Sadness** Template = BDI

0 = Not at all condition (described)
1 = Mild condition
2 = Moderate condition
3 = Severe condition

469 bdi2 relates to Q 02: **Discouraged**
470 bdi3 relates to Q 03: **Failure**
471 bdi4 relates to Q 04: **Satisfaction**
472 bdi5 relates to Q 05: **Guilty**
473 bdi6 relates to Q 06: **Punished**
474 bdi7 relates to Q 07: **Disappointed**
475 bdi8 relates to Q 08: **Blame**
476 bdi9 relates to Q 09: **Kill**
477 bdi10 relates to Q 10: **Cry**
478 bdi11 relates to Q 11: **Irritated**
479 bdi12 relates to Q 12: **Lost interest**
480 bdi13 relates to Q 13: **Decisions**
481 bdi14 relates to Q 14: **Look**
482 bdi15 relates to Q 15: **Work**
483 bdi16 relates to Q 16: **Sleep**
484 bdi17 relates to Q 17: **Tired**
485 bdi18 relates to Q 18: **Appetite**
486 bdi19 relates to Q 19: **Weight**
487 bdi20 relates to Q 19: Losing weight by **eating less ?** Temp = Yes / No
488 bdi21 relates to Q 20: **Worried** Temp = BDI
489 bdi22 relates to Q 21: **Sex**

490 bditot **Beck Depression Inventory - Total scores**

Compute bditot = bdi1 + bdi2 + bdi3 + bdi4 + bdi5 + bdi6 + bdi7 + bdi8 + bdi9 + bdi10 + bdi11 + bdi12 + bdi13 + bdi14 + bdi15 + bdi16 + bdi17 + bdi18 + bdi19 + bdi21 + bdi22. Execute .

0 = No depression measured
66 = Extremely (clinically) depressed

491 ses1 relates to Q 01 : **Satisfied** Template = SES

1 = Strongly agree (4)
2 = Agree (3)
3 = Disagree (2)
4 = Strongly disagree (1)

recoded according to the coding frame on page 56 (overleaf)

492 ses2 relates to Q 02: **No good**
493 ses3 relates to Q 03: **Qualities**
494 ses4 relates to Q 04: **Do things**
495 ses5 relates to Q 05: **Proud**
496 ses6 relates to Q 06: **Useless**
497 ses7 relates to Q 07: **Person of worth**
498 ses8 relates to Q 08: **Respect**
499 ses9 relates to Q 09: **Failure**
500 ses10 relates to Q 10: **Positive attitude**

SES Coding Frame

	strongly agree	agree	disagree	strongly disagree
1. On the whole, I am satisfied with myself	4	3	2	1
2. At times I think I am no good at all	1	2	3	4
3. I feel that I have a number of good qualities	4	3	2	1
4. I am able to do things as well as most other people	4	3	2	1
5. I feel I do not have much to be proud of	1	2	3	4
6. I certainly feel useless at times	1	2	3	4
7. I feel that I am a person of worth, at least on equal	4	3	2	1
8. I wish I could have more respect for myself	1	2	3	4
9. All in all, I am inclined to feel that I am a failure	1	2	3	4
10. I take a positive attitude towards myself	4	3	2	1

501 sestot **Self Esteem Scale - Total scores**

Compute sestot = ses1 + ses2 + ses3 + ses4 + ses5 + ses6 + ses7 + ses8 + ses9 + ses10. Execute.

10 = Minimum score
40 = Maximum score

502 pesteem **Positive Self Esteem**

Compute pesteem = ses1 + ses3 + ses4 + ses7 + ses10. Execute.

5 = Minimum score
20 = Maximum score

503 nesteem **Negative Self Esteem**

Compute nesteem = ses2 + ses5 + ses6 + ses8 + ses9. Execute.

5 = Minimum score
20 = Maximum score

Impact of Dystonia Questionnaire : Q're No. 60

504 date60 **Date of Q're 60** dd.mm.yy 8

505 iod1 relates to Q 01: **Hard time** Template = IOD

1 = Strongly disagree
2 = Mildly disagree
3 = Neither agree or disagree
4 = Mildly agree
5 = Strongly agree

506 iod2 relates to Q 02: **Useless** Template = IOD

507 iod3 relates to Q 03: **Miss the things**

508 iod4 relates to Q 04: **More dependant**

509 iod5 relates to Q 05: **Uncomfortable**

510 iod6 relates to Q 06: **Self-sufficient**

511 iod7 relates to Q 02: **Inadequate** Template = IOD

5 = Strongly disagree (1) recoded
4 = Mildly disagree (2)
3 = Neither agree or disagree (3)
2 = Mildly agree (4)
1 = Strongly agree (5)

512 iodtot **Impact of Dystonia** - Total scores

7 = Minimum impact
14 = 25% impact
21 = 50% impact
28 = 75% impact
35 = Maximum impact

Compute iodtot = iod1 + iod2 + iod3 + iod4 + iod5 + iod6 + iod7. Execute.

513 iod8 relates to Q Page 2: **Acceptance Stages**

1 = Shock
2 = Anger
3 = Despair
4 = Acceptance

514 crit1 relates to Questions : **Criticism of study 1** Temp = CRIT.

1 = It has helped me (hope it has helped you)
2 = Difficult without help
3 = Expressed my feelings (emotions)
4 = Courtesy / Respect
5 = Consideration
6 = Understanding
7 = Much appreciated (Thank you)
8 = Very professional
9 = "No comment" or No criticisms
10 = Too much trouble
11 = Need more space (to write)
12 = Questions duplicated
13 = Only some relevant (some not)
14 = Personal / disquieting
15 = Prefer writing (than talking about it)
16 = Now in remission (can't / difficult to answer)
17 = (Hope it) helps young people
18 = More research needed
19 = Inform more GP's (Dr's)
20 = Use simpler words

515 crit2 relates to Questions: **Criticism of study 2** Temp = CRIT.

516 crit3 relates to Questions: **Criticism of study 3** Temp = CRIT.

$crittot **Total criticisms of study** - variable crit1, crit2 and crit3
Multiple Response Set - Range 1 to 20

Primary Carer Questionnaire : Q're No. 70

517	date70	**Date of Q're 70**	dd.mm.yy 8
518	csi1	relates to Q 01: **Relationship** to person	Numeric 4.0

1 = Husband
2 = Wife
3 = Mother
4 = Father
5 = Son
6 = Daughter
7 = Carer - not related
8 = No primary carer
9 = Girl / Boy friend (Live in partner)
10 = Sibling (sister)
11 = Sibling (brother)
12 = Person declined
13 = Aunt

519 csi2 relates to Q 02: **Sleep** Template = SES

1 = Strongly agree
2 = Agree
3 = Disagree
4 = Strongly disagree

520 csi3 relates to Q 03: **Inconvenient**
521 csi4 relates to Q 04: **Physical strain**
522 csi5 relates to Q 05: **Confining**
523 csi6 relates to Q 06: **Adjust at home**
524 csi7 relates to Q 07: **Plans**
525 csi8 relates to Q 08: **Other demands**
526 csi9 relates to Q 09: **Emotional**
527 csi10 relates to Q 10: **Behaviour**
528 csi11 relates to Q 11: **Adjustments at work** (if missing - deduce 2 or 3)
529 csi12 relates to Q 12: **Financial strain**
530 csi13 relates to Q 13: **Overwhelmed**
531 csi14 relates to Q 14: **Distressing**

532 csitot **Carer's Self Inventory - Total scores**

13 = Minimum - 'strongly agree'
26 = Average - 'agree'
32 = Average score)
33 = Average score)
39 = Average - 'disagree'
52 = Maximum - 'strongly disagree'

Compute csitot = csi2 + csi3 + csi4 + csi5 + csi6 + csi
csi12 + csi13 + csi14. Execute.

533 csi15 relates to Q 15: **Statement 1**

1 = Has changed my life
2 = Has not changed life
3 = Patient has changed
4 = Affected my emotions
5 = Treatment improved it
6 = Been very positive
7 = Limited socially
8 = Had to stop work
9 = Been very stressful
10 = He / She needs my support
11 = More publicity required
12 = General public ignorant
13 = I feel useless (unable to help)
14 = Dr's ignorant
15 = More research needed

534 csi16 relates to Q 15: Statement 2

535 csi17 relates to Q 15: Statement 3

$stattot **Total of Statements** - variable csi15, csi16 and csi17
Multiple Response Set - Range 1 to 15

Environmental Factors Questionnaire : Q're No 80

This Questionnaire is divided into five different sections :-

1. General Questions	Questions 01 to 18 - Pages 1 to 2
2. Chemical Exposure	Questions 19 to 24 - Page 2
3. Allergic Reactions	Questions 25 to 27 - Page 3
4. Food and Drink	Questions 28 to 41 - Pages 3 to 4
5. Other Environmental Substances	Questions 42 to 60 - Pages 5 to 8

536 date80 **Date of Q're 80** dd.mm.yy 8

537 hand2 relates to Q 01: **Which hand ?** Template = Hand

1 = Left
2 = Right
3 = Can't write (illiterate)

538 hand3 relates to Q 02: **Always same hand ?** Temp = Yes/No

539 kick relates to Q 03: **Kick a ball, which foot ?**

1 = Left
2 = Right
3 = Ambidextrous

540 thy1 relates to Q 04: Abnormalities of **thyroid gland** Temp = Yes/No

541 thy2 relates to Q 05: Details

1 = Under-active (Myxedema)
2 = Over-active
3 = Thyroid removed
4 = Thyroglossal Cyst
5 = Nodule on thyroid
6 = Enlarged / swollen

542 chng1 relates to Q 06: **Life pattern** Template = Yes/No

543 chng2 relates to Q 07: **Describe**

1 = Drug dependency
2 = Bereavement
3 = (Neuro) Surgery
4 = Diabetic
5 = RTA (Car / Cycle)
6 = Divorce
7 = Redundancy
8 = Family Stress

9 = Pregnancy
10 = Working stress (changes)
11 = Neighbour problems
12 = Industrial injury
13 = Severe Migraine
14 = Depression
15 = Eyesight impaired
16 = Childhood diseases
17 = Stroke

544 chng3 relates to Q 07: **Year change took place**
0 = Unknown year

545 smok1 relates to Q 08: **Smoke regularly** Temp = Yes / No
546 smok2 relates to Q 09: **Ever smoked** (regularly)
547 smok3 relates to Q 10: **Affected by other's** (smoking)
548 gas relates to Q 11: **Gas appliances**
549 soun1 relates to Q 12: **Certain sounds**

550 soun2 relates to Q 13: **Describe sound**

1 = Long banging
2 = Screeching / Shrill Noise
3 = Sudden Noise
4 = Buzz, Hiss, etc (Vibrating, tapping, strong winds)
5 = Loud Noise / Music
6 = Have Tinnitus

551 sun1 relates to Q 14: **Bright light / sunlight** Temp = Yes / No

552 sun2 relates to Q 15: **Describe how**

1 = (Causes) Blinking
2 = Eyes shut (screw up)
3 = Headaches / migraines (pain behind eyes)
4 = Blurred vision
5 = Photosensitive
6 = Makes spasms worse
7 = Dizzy (walking in bright light)
8 = Warmth helps spasms
9 = Allergic reaction (to sunlight)
10 = Eyes water

553 wat1 relates to Q 16: **Drinking water**
1 = Spring
2 = Well
3 = Mains
4 = Filtered
5 = Bottled

554 wat2 relates to Q 17: **Mains water**

1 = Fluoridated
2 = Soft
3 = Hard
4 = Softened
5 = Don't know

555 spma relates to Q 18a: **Stress** Template = spm

1 = Better
2 = Unchanged
3 = Worse

556 spmb relates to Q 18b: **Relaxation**
557 spmc relates to Q 18c: **Fatigue**
558 spmd relates to Q 18d: **Emotion**
559 spme relates to Q 18e: **Distraction**
560 spmf relates to Q 18f: **Self-Consciousness**
561 spmg relates to Q 18g: **Social Situations**
562 spmh relates to Q 18h: **Heat**
563 spmi relates to Q 18i: **Cold**
564 spmj relates to Q 18j: **Walking**
565 spmk relates to Q 18k: **Running**
566 spml relates to Q 18l: **Carrying objects**
567 spmm relates to Q 18m: **Writing**
568 spmn relates to Q 18n: **Sleep**
569 spmo relates to Q 18o: **Lying on back**
570 spmp relates to Q 18p: **Lying on side**
571 spmq relates to Q 18q: **Pre-Menstrual Cycle**
572 spmr relates to Q 18r: **During Menstrual Cycles**
573 spms relates to Q 18s: **Between Menstrual Cycle**
574 spmt relates to Q 18t: **On wakening**
575 spmu relates to Q 18u: **During morning**
576 spmv relates to Q 18v: **During afternoon**
577 spmw relates to Q 18w: **During evening**

578 chem1 relates to Q 19: **Exposed to chemicals** Temp = Yes / No
579 chem2 relates to Q 20: **Crop spraying**
580 chem3 relates to Q 21: **Industrial pollution**
581 chem4 relates to Q 22: **Industrial accident**
582 chem5 relates to Q 23: **Chemical fire**
583 chem6 relates to Q 24: **Other type**

584 chem7 relates to Q 24: **Other types** of chemicals exposed to

1 = Photocopying
2 = Treated grass
3 = Fibre glass resin
4 = Woodworm / dry rot (timber treatment)
5 = Mercury
6 = Science Lab (worked in)
7 = Chlorine
8 = Shipyard Chemicals
9 = Asbestos
10 = Major Tranquillisers
11 = Paint (worked in factory or worked with)
12 = Pharmacy (worked in)
13 = Chemical Factory (worked in)
14 = Traffic pollution (garage forecourt)
15 = Embalmers (worked in)
16 = Building Chemicals (DPC, Sealocrete, etc)
17 = Military Service
18 = Concrete Cleaning Fluids
19 = Military Warfare (Chemical Weapons)
20 = Nuclear power
21 = U/G Explosives
22 = Steel Mills (worked in)

585 alg relates to Q 25: **Allergic problems** Temp = Yes / No
Syntax file : 'phd-env.sps' = 14 calculations

IF (alg EQ 2) alga = 2. IF (alg EQ 2) algb = 2. [etc, continues until]
IF (alg EQ 2) algn = 2. Execute .

This formula only used if no allergic problems at all have been notified.

586 alga relates to Q 26a: **Asthma**
587 algb relates to Q 26b: **Migraines**
588 algc relates to Q 26c: **Persistent fatigue**
589 algd relates to Q 26d: **Hay Fever**
590 alge relates to Q 26e: **Arthritis**
591 algf relates to Q 26f: **Hyperactivity**
592 algg relates to Q 26g: **Rhinitis**
593 algh relates to Q 26h: **Mouth Ulcers**
594 algi relates to Q 26i: **Stomach Ulcers**
595 algj relates to Q 26j: **Wind / Bloating**
596 algk relates to Q 26k: **Eczema**
597 algl relates to Q 26l: **Puffy ankles / hands / face**
598 algm relates to Q 26m: **Uticaria / Hives**
599 algn relates to Q 26n: **Diarrhoea**

600	algo	relates to Q 26m: **Allergic to ...** 1 = Metal (incl Zinc) 2 = Drugs (incl Penicillin, Clonazepam, etc) 3 = Skin Allergies (incl Dermatitis, Psoriasis, etc) 4 = Not an allergy (incl Heart Burn, Constipation) 5 = Eggs (severe)	
601	algp	relates to Q 27: **Blood relatives**	Temp = Yes / No
602	fad1	relates to Q 28: **Food and Drink**	Temp = Yes / No
603	fad2	relates to Q 29: **Spasms worse**	Temp = Yes / No
604	fada	relates to Q 30a: **Pork**	Temp = fad
		0 = Not affected at all 1 = Mildly affected 2 = Moderately affected 3 = Severely affected	
605	fadb	relates to Q 30b: **Beef**	
606	fadc	relates to Q 30c: **Red meat**	
607	fadd	relates to Q 30d: **White meat**	
608	fade	relates to Q 30e: **Fish or Shellfish 1**	
609	fad3	relates to Q 30e: **Fish or Shellfish 2** 1 = Fish 2 = Shellfish 3 = Both	
610	fadf	relates to Q 30f: **Wheat**	Temp = fad
611	fadg	relates to Q 30g: **Rice**	
612	fadh	relates to Q 30h: **Corn**	
613	fadi	relates to Q 30i: **Oats**	
614	fadj	relates to Q 30j: **Other Cereals 1**	
615	fad4	relates to Q 30j; **Other Cereals 2** 1 = (Brazil) Nuts 2 = Pastry 3 = (too much) Fat 4 = 3 types of Wheat	
616	fadk	relates to Q 30k: **Milk**	Temp = fad
617	fadl	relates to Q 30l: **Egg**	
618	fadm	relates to Q 30m: **Cheese**	
619	fadn	relates to Q 30n: **Yoghurt**	
620	fado	relates to Q 30o: **Other Dairy Products 1**	

621	fad5	relates to Q 30o: **Other Dairy Products 2** 1 = Cottage cheese 2 = (fresh) Cream	
622	fadp	relates to Q 30p: **Coffee**	
623	fadq	relates to Q 30q: **Tea**	
624	fadr	relates to Q 30r: **Other liquids 1**	
625	fad6	relates to Q 30r: **Other liquids 2** 1 = Alcohol - undefined 2 = Alcohol - Gin 3 = Alcohol - Wine 4 = Orange Juice 5 = Low C Choc & Vinegar 6 = Tap Water 7 = Soft drinks (cans) 8 = Fruit drinks & Vinegar	
626	fads	relates to Q 30s: **Citrus fruit**	
627	fadt	relates to Q 30t: **Potato**	
628	fadu	relates to Q 30u: **Other Fruit & Veg 1**	
629	fad7	relates to Q 30u: **Other Fruit & Veg 2** 1 = Strawberries 2 = Carrots & Tomatoes 3 = Parsnip 4 = Banana 5 = Oranges 6 = 'Some' vegetables ?	
630	fadv	relates to Q 30v: **Yeast**	
631	fadx	relates to Q 30x: **Chocolate**	
632	fady	relates to Q 30y: **Sugar**	
633	fadz	relates to Q 30z: **Other Foods** 0 = Not affected at all 1 = Mildly affected by curries 2 = Moderately affected by curries 3 = Severely affected by red wine	
634	excl	relates to Q 31: **Exclusion diet**	Temp = Yes / No
635	crav1	relates to Q 32: **Crave** (foods)	Temp = Yes / No

636 crav2 relates to Q 33: **Foods craved** (which)

1 = Chips & Fruit cake
2 = Choc & Alcohol
3 = Ribena
4 = Alcohol
5 = Tea & Coffee
6 = Chocolate
7 = Tea, Choc & Cream
8 = Bananas & Crisps
9 = Choc & Butter
10 = Choc & Sweet Food
11 = Anything Sweet
12 = Lucozade - Dry mouth
13 = Hot - Spicy Foods
14 = High Fat Foods (Chips, Cheese, etc)
15 = Garlic
16 = Fruit Juice (Orange Juice)
17 = Tea, Milk Pudding (Ice Cream)
18 = Coffee
19 = Choc and Yoghurt (Drinks)
20 = Horlicks
21 = Choc and Coffee

637 tea relates to Q 34: **No of cups of tea** (in a day)
638 coff relates to Q 35: **No of cups of coffee** (in a day)
639 deca1 relates to Q 36: **Decaffeinated**

1 = Yes
2 = No
3 = Don't drink coffee

640 deca2 relates to Q 37: **Describe how ?**

1 = Undec makes spasms worse
2 = Dec makes spasms better

641 wine1 relates to Q 38: **Wines**

1 = Yes
2 = No
3 = Don't drink alcohol

642 wine2 relates to Q 39: **Describe how ?**

1 = Have more confidence
2 = Relaxes spasms
3 = Severely affected (adversely)
4 = Alcoholic (recovering)
5 = Mildly affected (adversely)
6 = Slightly sick
7 = Dries up mouth
8 = Go to sleep
9 = Slight headache
10 = Loosens bowels (Colitis)
11 = Reacts with drugs

643 cola1 relates to Q 40: **Colas**
1 = Yes
2 = No
3 = Don't drink them

644 cola2 relates to Q 41: **Describe how ?**

1 = Choke when drinking
2 = Slight hiccups
3 = Feel edgy / nervous
4 = Tongue - sensitive / sore
5 = Dries up mouth
6 = Aids digestion
7 = Spasms in stomach
8 = Wind and bloating
9 = Upset stomach (nausea)
10 = Worse spasms (next day)

645 env1 **Environmental factors** (method of entering data only)
Syntax file : 'phd-env.sps' = 79 calculations
1 = No data for rest of series

IF (env EQ 1) envaa = 0. IF (enc EQ 1) envab = 0. [etc, continues until]
IF (env EQ 1) envdz = 0. Execute .

646 envaa relates to Q 42aa: **New mown grass** Template = fad
647 envab relates to Q 42ab: **Trees**
648 envac relates to Q 42ac: **Long grass**
649 envad relates to Q 42ad: **Hay**
650 envae relates to Q 42ae: **Other pollens**

651 envaf relates to Q 43af: **Damp humid days**
652 envag relates to Q 43ag: **Old houses**
653 envah relates to Q 43ah: **Fungus**
654 envai relates to Q 43ai: **Moulds**

Template = fad

655	envaj	relates to Q 43aj: **Dust**
656	envak	relates to Q 44ak: **Dogs**
657	enval	relates to Q 44al: **Cats**
658	envam	relates to Q 44am: **Horses**
659	envan	relates to Q 44an: **Rodents**
660	envao	relates to Q 44ao: **Birds**
661	envap	relates to Q 44ap: **Feathers**
662	envaq	relates to Q 45aq: **Bee stings**
663	envar	relates to Q 45ar: **Wasp stings**
664	envas	relates to Q 45as: **House dust**
665	envat	relates to Q 45at: **Mites**
666	envau	relates to Q 46au: **Odour of pines**
667	envav	relates to Q 46av: **Pine products**
668	envaw	relates to Q 46aw: **Houseplants**
669	envax	relates to Q 46ax: **Manure**
670	envay	relates to Q 46ay: **Silage**
671	envaz	relates to Q 46az: **Rotting vegetation**
672	envba	relates to Q 47ba: **Degreasers**
673	envbb	relates to Q 47bb: **Deodorants**
674	envbc	relates to Q 47bc: **Hairsprays**
675	envbd	relates to Q 47bd: **Polishes**
676	envbe	relates to Q 47be: **Insecticides**
677	envbf	relates to Q 47bf: **Pesticides**
678	envbg	relates to Q 47bg: **Air Freshener**
679	envbh	relates to Q 48bh: **After-shave**
680	envbi	relates to Q 48bi: **Creams**
681	envbj	relates to Q 48bj: **Deodorants**
682	envbk	relates to Q 48bk: **Perfume**
683	envbl	relates to Q 48bl: **Powder**
684	envbm	relates to Q 48bm: **Make-up**
685	envbn	relates to Q 48bn: **Shampoo**
686	envbo	relates to Q 49bo: **New carpets**
687	envbp	relates to Q 49bp: **Linoleum**
688	envbq	relates to Q 49bq: **Floor Tiles**
689	envbr	relates to Q 49br: **Sealer**
690	envbs	relates to Q 49bs: **Adhesive**
691	envbt	relates to Q 50bt: **Carpet backing**
692	envbu	relates to Q 50bu: **Cushion**
693	envbv	relates to Q 50bv: **Upholstery**
693	envbw	relates to Q 50bw: **Padding**

695	envbx	relates to Q 51bx: **Ammonia**	Template = fad
696	envby	relates to Q 51by: **Bleaches**	
697	envbz	relates to Q 51bz: **Detergents**	
698	envca	relates to Q 51ca: **Liquid polishers**	
699	envcb	relates to Q 51cb: **Silver polish**	
700	envcd	relates to Q 51cd: **Furniture polish** (Note - out of sequence)	
701	envcc	relates to Q 51cc: **Carpet shampoo** (Note - out of sequence)	
702	enved	relates to Q 52cd: **Heating Oil** (Note - add reference number)	
703	envce	relates to Q 52ce: **Gas**	
704	envcf	relates to Q 52cf: **Paraffin**	
705	envcg	relates to Q 52cg: **Calor / Butane**	
706	envch	relates to Q 52ch: **Coal Fire**	
707	envci	relates to Q 52ci: **Charcoal**	
708	envcj	relates to Q 52cj: **Burning Tar**	
709	envck	relates to Q 52ck: **Burning Rubber**	
710	envcl	relates to Q 53cl: **Petrol fumes**	
711	envcm	relates to Q 53cm: **Oil**	
712	envcn	relates to Q 53cn: **Diesel fumes**	
713	envco	relates to Q 53co: **Car Upholstery**	
714	envcp	relates to Q 53cp: **Exhaust fumes**	
715	envcq	relates to Q 54cq: **New paint**	
716	envcr	relates to Q 54cr: **Paint stripper**	
717	envcs	relates to Q 54cs: **Turpentine**	
718	envct	relates to Q 54ct: **Varnish**	
719	envcu	relates to Q 54cu: **Oils**	
720	envcv	relates to Q 54cv: **Fixative**	
721	envcw	relates to Q 55cw: **Cigarettes**	
722	envcx	relates to Q 55cx: **Cigars**	
723	envcy	relates to Q 55cy: **Pipe**	
724	envcz	relates to Q 55cz: **Cigarette smoke**	
725	envda	relates to Q 56da: **Lighter fuel**	
726	envdb	relates to Q 56db: **Carbon Tetrachloride**	
727	envdc	relates to Q 56dc: **Glues**	
728	envdd	relates to Q 56dd: **Newsprint**	
729	envde	relates to Q 56de: **White Spirit**	
730	envdf	relates to Q 56df: **Dry Cleaning fluid**	
731	envdg	relates to Q 56dg: **Methylated Spirits**	
732	envdh	relates to Q 56dh: **Surgical Spirit**	
733	envdi	relates to Q 57di: **Chlorine**	
734	envdj	relates to Q 57dj: **Bitumen Tar**	
735	envdk	relates to Q 57dk: **Asphalt**	
736	envdl	relates to Q 57dl: **Weedkillers**	
737	envdm	relates to Q 57dm: **Mothballs**	

738	envdn	relates to Q 57dn: **Disinfectants**	Template = fad
739	envdo	relates to Q 57do: **Swimming Pools**	
740	envdp	relates to Q 57dp: **Food Additives**	
741	envdq	relates to Q 57dq: **Pesticide Residues in Food**	
742	envdr	relates to Q 57dr: **Pollutants in Water**	
743	envds	relates to Q 57ds: **Waxes on fruit and veg**	
744	envdt	relates to Q 57dt: **Tartrazine**	
745	envdu	relates to Q 57du: **Phenolic Resin in cans**	
746	envdv	relates to Q 57dv: **Formaldehyde**	

747	envdw	relates to Q 58dw: **Plastics**
748	envdx	relates to Q 58dx: **Coated paper**
749	envdy	relates to Q 58dy: **New fabric**
750	envdz	relates to Q 58dz: **Other 1**
751	env2	relates to Q 58dz: **What other smells**

1 = Rubbish bins
2 = Carrots
3 = Burning smells
4 = Fishy smells
5 = Wood smoke
6 = 'Creosote'
7 = Onions
8 = Rubefacient cream

752	envea	relates to Q 59dz: **Other 2**	Temp = fad
753	env3	relates to Q 59dz: **What others 2 ?**	

1 = Bath cleaner
2 = (some) Lagers
3 = Body odour
4 = Food colouring
5 = 'Penicillin'
6 = 'Maxolon'
7 = 'Aspirin'
8 = Essential Oils
9 = Drug (reaction)
10 = Eels
11 = 'Scotchguard'
12 = 'Lanolin'
13 = 'Algipan' Spray
14 = (Garden) Soil
15 = Straw dust

754	enveb	relates to Q 59dz: **Other 3**	Template = fad

755 env4 relates to Q 59dz: **What others 3 ?**

1 = Unwashed bodies
2 = Tomatoes
3 = Flowers
4 = Aspirin
5 = Rats
6 = Hair Dye

756 comm1 relates to Q 60: **Comment 1**

1 = Worse, activity/walking : Better resting/relaxed
2 = Worse, self-conscious
3 = Better, busy
4 = Better, no stress : Worse with stress
5 = Worse, reading / TV
6 = Worse, worry
7 = Better, sun / warmth : Worse, cold / damp
8 = Currently in remission
9 = Counselling helps
10 = Worse, miss inj. : Better with inj / medication
11 = High Voltage Electricity : o/h lines ??
12 = Worse, tired : Better, sleep / in mornings
13 = Helped by surgery
14 = Worse, over time
15 = Worse, bright (sun)light
16 = Worse, Depression / Anxiety
17 = Worse, (severe) climatic changes
18 = Worse lie on affected side : Posture affects spasms
19 = Poss. link with drugs / solvents
20 = Worse (on) windy days
21 = "No comment / Don't know"
22 = Better (with) positive thoughts (or laughter)
23 = Unaffected (by) anything
24 = Definite pattern (but) why ?
25 = Better (with) alcohol
26 = No definite pattern
27 = Suspect hormonal link
28 = Worse, before period : Better, during period

757 comm2 relates to Q 60: **Comment 2**
758 comm3 relates to Q 60: **Comment 3**

$commtot **Total - Comments** - variable comm1, comm2 and comm3
Multiple Response Set - Range 1 to 28

Diagnosis Questionnaire : Q're No 81

759 date81 **Date of Q're 81 or date of death** dd.mm.yy 8

760 surv81 **Participation in Q're 81**

1 = participated
2 = not returned
3 = deceased
4 = not issued
5 = Info from interview

761 spec1 relates to Question 1 : **Specialist 1** Template = Specialist

1 = Orthopaedic Surgeon
2 = Rheumatologist
3 = Psychiatrist
4 = Neurologist
5 = ENT Surgeon
6 = Physiotherapist
7 = Ophthalmologist / Surgeon
8 = Neuro Surgeon
9 = Aromatherapist
10 = Osteopath
11 = Meditation Spec.
12 = Psychologist
13 = Chiropractor
14 = Speech Therapist
15 = Plastic Surgeon
16 = Chiropodist
17 = Gynaecologist
18 = Haematologist
19 = Paediatrician
20 = Paediatric Neuro.
21 = Pain Spec.
22 = Orthodontic Surgeon
23 = Unknown Consultant
24 = Geriatrician
25 = Immunologist

762 spec2 relates to Question 1 : **Specialist 2**
763 spec3 relates to Question 1 : **Specialist 3**
764 spec4 relates to Question 1 : **Specialist 4**

$spectot **Total - Specialists** - variable spec1, spec2, spec3 and spec4
Multiple Response Set - Range 1 to 25

765 alt1 relates to Question 2 : **Alternative treatment 1** Temp = Alternative

1 = Osteopathy
2 = Acupuncture
3 = Hypnosis / Hypnotherapy
4 = Physiotherapy
5 = Psychotherapy
6 = Speech Therapy
7 = Homeopathy
8 = Counselling
9 = Experimental drugs (Guinea Pig)
10 = Traction
11 = Yoga
12 = Chiropractic
13 = Surgery
14 = Behavioural Therapy
15 = Reflexology
16 = Hydrotherapy
17 = Aromatherapy
18 = Faith Healing (Healing Hands)
19 = Relaxation techniques
20 = Heat treatment / Elec shock
21 = Manipulation (under anaesthetic)

766 alt2 relates to Question 2 : **Alternative treatment 2**
767 alt3 relates to Question 2 : **Alternative treatment 3**
768 alt4 relates to Question 2 : **Alternative treatment 4**

$alttot **Total - Alternative treatments** - variable alt1, alt2, alt3 and alt4
Multiple Response Set - Range 1 to 21

769 diff3 relates to Question 3 : **Time to obtain a diagnosis**
Compared to diff between onset and diagnosis

770 diff4 **Compare diff1 & diff3** (in years)

Compute diff4 = diff1 - diff3 . Execute .

Dystonia Nurse Practitioner Questionnaires : Q'res No. 91 to 94

771 stat3 **dnp status** Numeric 4.0

1 = Home - completed
2 = Home - incomplete
3 = Home - declined
4 = Home - deceased
5 = Clinic - completed
6 = Clinic - incomplete
7 = Clinic - declined
8 = Clinic - deceased
9 = Not in study

772 dnp **dnp location** String 4

C = Clinic
H = Home
N = Not in dnp study

773 dnpno **dnp project number** String 5

C01 to C66 inclusive
H01 to H64 inclusive

774 date91 **Date of Q're 91** dd.mm.yy 8

775 age91 relates to **Age** (of patient) **as of date91** Numeric 4.1

```
Compute age91 = YRMODA (xdate.year(date91),xdate.month(date91),
xdate.mday(date91)) / 365.25 - YRMODA (xdate.year(dob),
xdate.month(dob),xdate.mday(dob))/ 365.25 .  Execute.

Compute age91 = (((age91 * 10) + 0.5) - (MOD(((age91 * 10) + 0.5),1)))/10.
Execute.
```

776 dnp101 **No of injections ?** (since first started)

777 dnp102 **No of injectors ?** (since first started)

778 dnp103 **How much time ?** (in minutes)

779 dnp104 **How often hurt ?** (number of times)

0 = Never hurt
30 = Hurt everytime

780 dnp105 **Best Knowledge 1** Temp = dnp-rating

1 = Very good
2 = Good
3 = Average
4 = Poor
5 = Very Poor

) Subjective ranking
) by the respondent
) in answering the
) questions relating
) to the injectors

781 dnp106 **Worst Knowledge 1**
782 dnp107 **Best Attitude 1**
783 dnp108 **Worst Attitude 1**

784 dnp109 **HMH Service now 1** Temp = dnp-service

1 = Seen promptly (usually)
2 = Staff efficient
3 = No complaints
4 = Good - Very good (excellent)
5 = Occasional waiting
6 = Receptionist VG.
7 = Friendly atmosphere (tea / coffee)
8 = Consultation (too) short
9 = Production Line (Cattle Market)
10 = Better before (ie reception moved / generally)
11 = Occasional inj. hurt
12 = Screen (no privacy)
13 = Car parking (difficult)
14 = Ambulance (travel time)
15 = Canteen (no signs / no drop in facilities)
16 = Different injectors
17 = Not (well) organised
18 = Explain clearly (needed)
19 = HMH better than other (hospitals)
20 = Better with same Dr.

785 dnp110 **HMH Service now 2** Temp = dnp-service

$hmhtot **Total - HMH Service now** - variable dnp109 and dnp110
Multiple Response Set - Range 1 to 20

786 dnp111 **Type of spasms ?** Temp = dnp-rating
787 dnp112 **What is the cause ?**

788 dnp113 **Spasms worse 1** Temp = dnp-worse

1 = No, nothing
2 = Smoking
3 = Social Situations
4 = Stress / Anxiety

5 = Angry / Upset / Tense
6 = Cold
7 = Physical activity
8 = Tiredness
9 = Worry
10 = BT cycle (too long)
11 = Sneezing
12 = Reading / (Watching) TV
13 = Bright lights
14 = Wind
15 = Migraines
16 = Agoraphobia
17 = Loud Noise
18 = Depression
19 = Caffeine

789 dnp114 **Spasms worse 2**

$wortot **Total - Spasms worse** - variable dnp113 and dnp114
Multiple Response Set - Range 1 to 19

790 dnp115 **Spasms better 1** Temp = dnp-better

1 = Operation
2 = Medication
3 = Sleep
4 = Heat (Warmth)
5 = Bot. Tox.
6 = Rest (Relaxation)
7 = In Remission
8 = Smoking (Cannabis)
9 = Positive Thoughts (Happy disposition)
10 = Nothing (except BT)
11 = Alcohol
12 = Chewing gum
13 = Good days / bad days
14 = TENS Machine
15 = Geste Antagoniste

791 dnp116 **Spasms better 2**

$betttot **Total - Spasms better** - variable dnp115 and dnp116
Multiple Response Set - Range 1 to 15

792 date92 **Date of Q're 92** dd.mm.yy 8

793 age92 **Age as of date92** Numeric 4.1

Compute age92 = YRMODA (xdate.year(date92),xdate.month(date92),
xdate.mday(date92)) / 365.25 - YRMODA (xdate.year(dob),
xdate.month(dob),xdate.mday(dob))/ 365.25 . Execute .

Compute age92 = (((age92 * 10) + 0.5) - (MOD(((age92 * 10) + 0.5),1)))/10.
Execute.

794 dnp201 **x injected last q're ?**
795 dnp202 **x injectors last q're ?**

796 dnp203 **How much time ? (in minutes)**
797 dnp204 **x injections hurt ?**
0 = Never hurt

798 dnp205 **Best Knowledge 2** Temp = dnp-rating
799 dnp206 **Worst Knowledge 2**
800 dnp207 **Best Attitude 2**
801 dnp208 **Worst Attitude 2**

802 dnp209 **Service improved 1** Temp = dnp-improved
1 = Can't be improved
2 = More flexible (appointments date and time)
3 = No, nothing
4 = Change inj. cycle (too long)
5 = Good - Very good
6 = Very polite / efficient
7 = Bot Tox (bad batches)
8 = Annual Review (for each patient)
9 = Time with the doctor
10 = No comment / complaints
11 = Same doctor (each time)
12 = Environment (of the clinic)
13 = Screen (more privacy)
14 = It has improved
15 = Time waiting (-ve)
16 = Waste of toxin (Availability)
17 = Doctor's attitude
18 = DSS Expenses (paid at clinic)
19 = Increase no of DNP's (or Dr's)
20 = (Wish I was / Glad I am) on Home List
21 = Car Parking
22 = Waiting - too long
23 = Short staffed (occasionally

803 dnp210 **Service improved 2**

$imptot **Total - Service improved** - variable dnp209 and dnp210
Multiple Response Set - Range 1 to 23

804 dnp211 **Aetiology of dystonia ?** Temp = dnp-rating
805 dnp212 **Cause of primary** (dystonia) ?
806 dnp213 **Cause of secondary** (dystonia) ?
807 dnp214 **Difference between 1st and 2nd ?**

808 date93 **Date of Q're 93** dd.mm.yy 8

809 age93 **Age as of date93** Numeric 4.1

```
Compute age93 = YRMODA (xdate.year(date93),xdate.month(date93),
xdate.mday(date93)) / 365.25 - YRMODA (xdate.year(dob),
xdate.month(dob),xdate.mday(dob))/ 365.25 .  Execute .

Compute age93 = (((age93 * 10) + 0.5) - (MOD(((age93 * 10) + 0.5),1)))/10.
Execute.
```

810 dnp301 **x injected last q're ?**
811 dnp302 **x injectors last q're ?**

812 dnp303 **How much time ? (in minutes)**
813 dnp304 **x injections hurt ?**
0 = Never hurt

814 dnp305 **Best Knowledge 3** Temp = dnp-rating
815 dnp306 **Worst Knowledge 3**

816 dnp307 **Best Attitude 3**
817 dnp308 **Worst Attitude 3**

818 dnp309 **Service at end 1** Temp = dnp-end

1 = No, nothing (no comment)
2 = Very satisfied (happy)
3 = Good - Very good (service)
4 = Second to none / Excellent
5 = Changed days (Wed to Tue)
6 = Changed time (am to pm)
7 = Train Dr's (talk to patients)
8 = Continue / Finance
9 = Self-conscious (open plan)
10 = Counselling service
11 = Screen (no longer up)
12 = Talk to Dr (time)

13 = Waiting occasionally long
14 = Cancelled appointments
15 = Delayed appointments
16 = Improved attitudes
17 = Improved time (Dr)
18 = Saves time / money
19 = Clinic busier now

819 dnp310 **Service at end 2**

$servtot **Total - Service from HMH** - variable dnp309 and dnp310
Multiple Response Set - Range 1 to 19

820 dnp311 **Idiopathic dystonia ?** Temp = dnp-rating
821 dnp312 **Basal ganglia ?**

822 dnp313 **Treatment improved ?**

1 = Yes
2 = No
3 = About the same

823 dnp314 **Service Improved ?** (compared to one year ago)

1 = Better
2 = About the same
3 = Worse

824 dnp401 **Whole project 1** Temp = dnp-project

1 = Less stress at home (more relaxed)
2 = DNP is the best (Dr Barnes / cant be bettered)
3 = Better now diagnosed (happier now settled)
4 = Good idea (scheme / brilliant)
5 = You are listened to
6 = DNP effective (Condition explained)
7 = Good - very good
8 = Save travel time / saves money
9 = Dr's time (limited)
10 = I have control
11 = Same person / DNP personal (relationship)
12 = DNP for easy cases (long distance)
13 = HMH for hard cases (short distance)
14 = Fall back (ie HMH)
15 = Wish (I had been) on home list
16 = Annual (HMH) review
17 = Staff - good at clinic
18 = Clinic now busier
19 = (DNP takes) pressure off clinic

825 dnp402 **Whole project 2**
826 dnp403 **Whole project 3**

$wholtot **Total - Whole project** - variable dnp401, dnp402 and dnp403
Multiple Response Set - Range 1 to 19

827 dnp404 **Home or clinic ?**

1 = Home
2 = Clinic
3 = Either

828 dnp405 **Why home 1 ?** Temp = dnp-home

1 = More relaxed (secure)
2 = More convenient (time / travel / money)
3 = More time (to discuss problems)
4 = Same person (continuity of treatment)
5 = DNP listens (trust)
6 = DNP explains (condition / treatment)
7 = Personal attention (of DNP)
8 = Self conscious (in clinic / on bus)
9 = Difficulty walking (travelling)
10 = Time (off work) Husband / Patient / My
11 = No reason given
12 = Save money / time / work (NHS Ambulance)

829 dnp406 **Why home 2**

$hometot **Total - Why home** - variable dnp405 and dnp406
Multiple Response Set - Range 1 to 12

830 dnp407 **Why clinic 1** Temp = dnp-clinic

1 = Dr's advice (needed sometimes)
2 = See other people (less isolated)
3 = Fixed time (better for me)
4 = Day out (able to do other things / shopping. etc)
5 = Fits (in) with work (times)
6 = Happy for either method
7 = No travel problems
8 = Family unaware of problems
9 = Others need DNP more than me
10 = Coffee / tea (Free of charge / facilities good)

831 dnp408 **Why clinic 2**

$clintot **Total - Why clinic** - variable dnp407 and dnp408
Multiple Response Set - Range 1 to 10

832 dnp409 **Other comments 1** Temp = dnp-comments

1 = Results (published)
2 = Very satisfied (happy)
3 = Clinic (became) too busy
4 = Personal relationship (with DNP)
5 = Your family also
6 = Many thanks
7 = Awareness (amongst G.P's)
8 = Medical Prize (students)
9 = Organised well
10 = Clinic patients (some resentful)
11 = More explanation (at clinic needed)
12 = Counselling Service (VG.)
13 = Clinic improved (over past year)
14 = No comment
15 = Offer research (for future)
16 = Hope it continues (want it to)
17 = Ambulance travel (long time)
18 = No difference in treatment (home / clinic)
19 = Home - more personal
20 = HMH better than NRI (other hospitals)
21 = NRI - long waiting
22 = NRI - mixed clinic
23 = Longer inj. cycle (now)
24 = BT strength (batches not standard)
25 = More DNP's (needed)
26 = appointments cancelled - why ?

833 dnp410 **Other comments 2**
834 dnp411 **Other comments 3**

$othtot **Total - Other comments** - variable dnp409, dnp410 and dnp410
Multiple Response Set - Range 1 to 26

835 diff7 **DNP Duration** (of study)

Compute diff7 = age93 - age91 . Execute .

Compute diff7 = (((diff7 * 10) + 0.5) - (MOD(((diff7 * 10) + 0.5),1)))/10. Execute .

836 date98 **End of survey or death** dd.mm.yy8

End of survey = 01.01.99, being 5.56 years from the start

837 age98 **Age at end or death** Numeric 4.1

Compute age98 = YRMODA (xdate.year(date98),xdate.month(date98), xdate.mday(date98)) / 365.25 - YRMODA (xdate.year(dob), xdate.month(dob),xdate.mday(dob))/ 365.25 . Execute .

Compute age98 = (((age98 * 10) + 0.5) - (MOD(((age98 * 10) + 0.5),1)))/10. Execute.

838 time3 **Time in study (years)** Numeric 4.1
ie time between beginning & end.

Compute time3 = YRMODA (xdate.year(date98),xdate.month(date98), xdate.mday(date98)) / 365.25 - YRMODA (xdate.year(date10), xdate.month(date10),xdate.mday(date10))/ 365.25 . Execute.

Compute time3 = (((time3 * 10) + 0.5) - (MOD(((time3 * 10) + 0.5),1)))/10. Execute.

839 recode1 **Time in study**

Recoded time 3 into categories as below :-

1 = Over 5.50 years
2 = 5.00 - 5.49 years
3 = 4.50 - 4.99 years
4 = 4.00 - 4.49 years
5 = 3.50 - 3.99 years
6 = 3.00 - 3.49 years
7 = 2.50 - 2.99 years
8 = 2.00 - 2.49 years
9 = 1.50 - 1.99 years
10 = 1.00 - 1.49 years
11 = 0.50 - 0.99 years
12 = 0.00 - 0.49 years
13 = Negative years

840 wbs1 **Welfare Benefits Service**

1 = Referred to Welfare Benefits Adviser
2 = Advised by WBA on telephone
3 = Advised by WBA in person
4 = Completed Benefit Application
5 = Successful application
6 = Applied for review / appeal
7 = Succesful review / appeal
8 = Unsuccessful review / appeal
9 = Unknown result

841 wbs2 **WBS - Referral Date**

9804 = April 1998 - start of WBS during ESD
9812 = December 1998 - end of WBS in ESD
9901 = January 1999 - continuation of WBS

842 wbs3 **WBS - Referral** (by whom)

DM = David (or Connie) Medd
GB = Ginger Butler
HC = Harry Crow
JW = John Whitaker
MH = Maurice Hawthorne
PW = Peter Williams

Templates Lists

EuroQol and SF 36 Questionnaire No 10

Order	Alpha	Name of Template	Columns using this template
1	46	MOBILITY	16, 89.
2	55	SELF-CARE	17, 90.
3	1	ACTIVITY	18, 91.
4	16	DISCOMFORT	19, 92.
5	5	ANXIETY	20, 93.
6	37	HEALTH	28, 101.
7	35	GEN. HEALTH	29, 102.
8	13	COMPARED	30, 103.
9	63	VIGOROUS	31-40, 104-113
10	67	YES-NO	42-45,46-48,115-118,120-122
11	32	EXTENT	51, 53, 124, 126.
12	38	HOW	52, 125.
13	43	LIFE	55-62,64,66,128-135,137,139
14	39	ILL	68-71, 141-144.
15	62	TYPE	73, 146.
16	49	PARTNER	76-83, 149-156.
17	53	QUALITATIVE	84, 157.

Demographic Questionnaire No 20

18	30	EMPLOY	175, 176.
19	54	SEG	177-181.
20	8	BENEFITS	184-186.
21	28	DOCTOR	197, 198.
22	17	DISTRIBUTION	204, 219, 221, 223.
23	34	FOCAL	205-208.
24	65	WHO	218, 220, 222.
25	45	MEDICATION	224, 227, 230, 232, 234.
26	64	WHEN	226, 229.
27	51	PREVIOUS	237-241.
28	57	SIDE-EFFECTS	242, 243, 259, 260.
29	42	LAST	244, 261.
30	12	COMORBIDITY	245-248.
31	40	INJECTIONS	250-255.
32	9	BODY1	257, 258.
33	3	AIM	277, 278.

PSR Questionnaires : Nos. 30 to 70

Order	Alpha	Name of Template	Columns using this template
34	68	YES-NO	286,289,290,322,331,338,339, etc
35	31	EVENT	287, 288.
36	66	YEAR	291-318,324,326,328,330, etc
37	29	EFFECT	292-319, 321.
38	50	PHYSICAL	323, 325, 327, 329.
39	52	PSYCHIATRIC	332, 335.
40	61	TREATMENT	334, 337.
41	47	MOVEMENTS	365-367.
42	14	CONTROL	369, 370.
43	10	BODY2	372-376.
44	44	LONG	381, 384.
45	48	PARTIAL	382-385.
46	2	AFFECTED (FDQ)	403-429.
47	6	BCS	438-459.
48	7	BDI	466-487.
49	56	SES	489-498.
50	41	IOD	503-509.
51	15	CRITICISM	512-514.
52	60	STATEMENT	531-533.

Environmental and Diagnostic Questionnaires : No 80

53	36	HAND	535, 537.
54	69	YES-NO	536,538,540,543-547,549, etc
55	59	SPM	553-575.
56	33	FAD	602-630, 644-752.
57	11	COMMENTS	754-756.
58	58	SPECIALIST	759-762.
59	4	ALTERNATIVE	763-766.

DNP Questionnaires : Nos. 91 to 94

60	25	DNP - RATING	778-781,784,785, 796-799, etc
61	26	DNP - SERVICE	782, 783.
62	27	DNP - WORSE	786, 787.
63	18	DNP - BETTER	788, 789.
64	23	DNP - IMPROVED	800, 801.
65	21	DNP - END	816, 817.
66	24	DNP - PROJECT	822-824.
67	22	DNP - HOME	826, 827.
68	19	DNP - CLINIC	828, 829.
69	20	DNP - COMMENT	830-832.

'phddata.sav' DATA SETS

Q're No	Name of Category	No of variables
Q're 00 :	ID, Geography, Hosp, CRN and Status	12
Q're 10 :	EuroQol and SF36 Health State Q'res (1st)	72
Q're 11 :	EuroQol and SF36 Health State Q'res (2nd)	74
Q're 20 :	Demographic Profile Questionnaire	103
Q're 21 :	TDS Questionnaire	17
Q're 30 :	Clinical Profile Questionnaire	111
Q're 40 :	Torticollis Questionnaire	12
Q're 50 :	Psychological Profile Questionnaire	100
Q're 60 :	Impact of Dystonia Questionnaire	13
Q're 70 :	Primary Carer's Questionnaire	19
Q're 80 :	Environmental Questionnaire	223
Q're 81 :	Diagnostic Questionnaire	12
Q're 91 :	Dystonia Nurse Practitioner Questionnaires	69
Q're 00 :	End of Study and WBS Variables	5

Therefore 745,170 data sets = 885 cases x 842 variables

35 Multiple Response Sets

$Foctot = Total of Focal Dystonias
$Famtot = Total of Family Members
$Disttot = Total of Family Distribution
$Medtot = Total Medication
$Dpdtot = Total Dosage per Day
$Whentot = Total of When Medication Taken
$Prevtot = Total of Previous Treatments
$Septtot = Total Side Effects from Previous Treatments
$Comtot = Total Comorbidity
$Bodtot = Total Sites in the Body
$Sebttot = Total Side Effects of Bot. Tox.
$Phystot = Total Physical Illnesses
$Yr1tot = Total Years of Onset of Physical Illnesses
$Psyctot = Total Psychiatric Illnesses
$Trettot = Total Treatments for Psychiatric Illnesses
$Yr5tot = Total Years of Onset of Psychiatric Illnesses
$Movetot = Total Types of Movement
$Paintot = Total Where Pain
$Yeartot = Total Years of Spontaneous Improvement
$Longtot = Total Length of Spontaneous Improvement
$Parttot = Total of Partial or Complete Improvement
$Crittot = Total Criticisms of Study
$Stattot = Total of Statements from Carer's Strain Index (CSI)
$Commtot= Total - Comments
$Spectot = Total - Specialists
$Alttot = Total - Alternative Treatments
$Hmhtot = Total HMH Service Now
$Wortot = Total of What Makes Spasms Worse
$Bettot = Total of What Makes Spasms Better
$Imptot = Total - Service Improved
$Servtot = Total - Service from HMH
$Wholtot = Total - Whole Project
$Hometot = Total - Why Home
$Clintot = Total - Why Clinic
$Othtot = Total - Other Comments.

258 VARIABLE NAMES USED

AGE	ACT	ANX	A	ACC	ANY	APP	AET	AIM	ACUP
ALEX	ANTI	ANXI	ARMR	ARML	ALG	ALGA to	ALGP	ALT	
BEND	BATH	BP	BURD	BENE	BEF	BOD	BIOF	BOTOX	BEST
BCS	BCSALL	BCSSPE	BCSPOS	BCSEVA	BCSTEN	BDI	BDITOT		
CRN	COMP	CUT	CALM	CON	COP	COM	CURR	COUN	CAUS
CHIRO	COUNS	CONT	CRIT	CSI	CSITOT	CHNG	CHEM	CRAV	COFF
COLA	COMM								
DATE	DISC	D	DOWN	DOSE	DOB	DIFF	DAY	DR	DIST
DIAG	DPD	DEL	DOC	DRUG	DISA	DISF	DECA	DNP	
ESD	EURO	EXT	EXC	EMB	ELD	EMP	EVNT	EFF	EYEL
EYER	EYES	EXCL	ENV	ENVAA to		ENVEB			
FEM	FOC	FAM	FACER	FACEL	FACES	FOOTR	FOOTL	FDQ	
FDQALL	FDQSOC	FDQPHY	FDQSEI	FDQLEI	FDQOTH	FAD	FADA to	FADZ	
GEOG	GEN	GH	GEND	GET	GP	GEST	GAS		
HOSP	HEAL	HAD	HOW	HAPP	HOUR	HYPN	HANDR	HANDL	HAND
ILL	INC	INJ	IOD	IODTOT		JOIN	JAW		KICK
LIFT	LIM	LESS	LIFE	LOT	LOW	LAST	LEGR	LEGL	LIVE
MOB	M	MOD	MH	MAR	MALE	MILE	MINS	MED	MEM
MOST	MENT	MEDI	MAJ	MOUTH	MOVE				
NERV	NOTDYS	NUMB	NEWS	NECK	NESTEEM				
OSTATE	ONE	ONSET	OSTEO	OTHER					
PF	PAIN	PART	PEOP	POST	PRIM	PAY	PREV	PHYSIO	
PSYCHO	PHYS	PSYC	PAIN	PSTN	PESTEEM				
QUAL		RP	RE	RECODE					
SURV	STAT	SELF	S	SEV	SOC	SF	SITE	SUP	SEG
SEC	SGMT	SIDE	SEPT	SEBT	SHG	SURG	SPEECH	SEVE	SPON
SES	SESTOT	SMOK	SOUN	SUN	SPMA to	SPMW	SPEC		
TIME	TIR	TYPE	TRAV	TRIG	TALK	TDS	TOLD	TRET	TONGUE
TRUNK	TRUE	THY	TEA		UNCO		VIG	VT	VOCAL
WALK	WALB	WALC	WORK	WORN	WOR	WHO	WHEN	WHAT	WHOM
WHY	WRIT	WAT	WINE		X		YNG	YOGA	YEAR

POST Home Post Code

Value Label	Value	Frequency	Percent	Valid Percent	Cum Percent
Welwyn Garden, Herts	AL08	1	.1	.1	.1
Birmingham	B26	1	.1	.1	.2
Cleckheaton, W Yorks	BD19	2	.2	.2	.4
Keighley, W Yorks	BD20	1	.1	.1	.5
Bolton, Lancs	BL06	1	.1	.1	.6
Bury, Lancs	BL08	1	.1	.1	.7
Worthing, Sussex	BN12	1	.1	.1	.9
Carlisle, Cumbria	CA01 1	1	.1	.1	1.0
Carlisle, Cumbria	CA01 2	5	.5	.5	1.5
Carlisle, Cumbria	CA01 3	1	.1	.1	1.6
Carlisle, Cumbria	CA02 4	2	.2	.2	1.8
Carlisle, Cumbria	CA02 6	2	.2	.2	2.0
Carlisle, Cumbria	CA02 7	6	.6	.6	2.7
Carlisle, Cumbria	CA03 0	1	.1	.1	2.8
Carlisle, Cumbria	CA03 9	2	.2	.2	3.0
Scotby, Carlisle	CA04 0	2	.2	.2	3.2
Low Hesket, Carlisle	CA04 8	1	.1	.1	3.3
Aspatria, Cumbria	CA05 3	1	.1	.1	3.4
Linstock, Carlisle	CA06 4	2	.2	.2	3.6
Wigton, Cumbria	CA07 0	1	.1	.1	3.7
Brampton, Cumbria	CA08 1	2	.2	.2	3.9
Penrith, Cumbria	CA10 1	2	.2	.2	4.2
Penrith, Cumbria	CA10 3	1	.1	.1	4.3
Penrith, Cumbria	CA11 0	2	.2	.2	4.5
Penrith, Cumbria	CA11 8	2	.2	.2	4.7
Penrith, Cumbria	CA11 9	1	.1	.1	4.8
Keswick, Cumbria	CA12 4	1	.1	.1	4.9
Cockermouth, Cumbria	CA13 0	1	.1	.1	5.0
Cockermouth, Cumbria	CA13 9	2	.2	.2	5.2
Workington, Cumbria	CA14 1	5	.5	.5	5.8
Workington, Cumbria	CA14 2	1	.1	.1	5.9
Workington, Cumbria	CA14 3	1	.1	.1	6.0
Workington, Cumbria	CA14 4	2	.2	.2	6.2
Workington, Cumbria	CA14 5	1	.1	.1	6.3
Maryport, Cumbria	CA15 6	1	.1	.1	6.4
Maryport, Cumbria	CA15 7	1	.1	.1	6.5
Appleby, Cumbria	CA16 6	1	.1	.1	6.6
Kirby Stephen, Cumb.	CA17 4	2	.2	.2	6.8
Ravenglass, Cumbria	CA18 1	1	.1	.1	6.9
Holmbrook, Cumbria	CA19 1	1	.1	.1	7.0
Seascale, Cumbria	CA20 1	3	.3	.3	7.4
Cleator Moor, Cumb.	CA25 5	2	.2	.2	7.6
Frizington, Cumbria	CA26 3	1	.1	.1	7.7
Whitehaven, Cumbria	CA28 6	1	.1	.1	7.8
Whitehaven, Cumbria	CA28 7	1	.1	.1	7.9
Whitehaven, Cumbria	CA28 8	1	.1	.1	8.0
Whitehaven, Cumbria	CA28 9	2	.2	.2	8.2
Gravesend, Kent	DA12	1	.1	.1	8.3
Montrose, Scotland	DD10	1	.1	.1	8.4
Dumfries, Scotland	DG02	1	.1	.1	8.5
Canonbie, Scotland	DG14	1	.1	.1	8.6
Gretna, Scotland	DG16	1	.1	.1	8.8
Durham City	DH01 1	2	.2	.2	9.0
Durham City	DH01 2	2	.2	.2	9.2
Durham City	DH01 3	2	.2	.2	9.4
Durham City	DH01 4	2	.2	.2	9.6
Durham City	DH01 5	5	.5	.5	10.1
Ch Le St, Durham	DH02 1	4	.4	.4	10.6
Ch Le St, Durham	DH02 2	1	.1	.1	10.7
Ch Le St, Durham	DH02 3	2	.2	.2	10.9
Birtley, Ch Le St.	DH03 1	5	.5	.5	11.4
Birtley, Ch Le St.	DH03 2	2	.2	.2	11.6
Ch Le St, Durham	DH03 4	4	.4	.4	12.1
Houghton Le Spring	DH04 4	2	.2	.2	12.3
Burnmoor, H. Le Sp.	DH04 5	1	.1	.1	12.4

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Value Label	Value	Frequency	Percent	Valid Percent	Cum Percent
Fenceho, H. Le Sp.	DH04 6	1	.1	.1	12.5
Penshaw, H. Le Sp.	DH04 7	3	.3	.3	12.8
Hetton Le Hole	DH05 0	1	.1	.1	12.9
Houghton Le Spring	DH05 8	2	.2	.2	13.1
E Rainton, Durham	DH05 9	2	.2	.2	13.3
Sherburn, Durham	DH06 1	2	.2	.2	13.6
Shotton Coll, Durham	DH06 2	4	.4	.4	14.0
Wheatley, Durham	DH06 3	3	.3	.3	14.3
Coxhoe, Durham	DH06 4	4	.4	.4	14.7
Bowburn, Durham	DH06 5	3	.3	.3	15.0
Lanchester, Durham	DH07 0	1	.1	.1	15.2
Sacriston, Durham	DH07 6	7	.7	.7	15.9
Langley Park, Durham	DH07 9	4	.4	.4	16.3
Consett, Co. Durham	DH08 0	1	.1	.1	16.4
Consett, Co. Durham	DH08 6	1	.1	.1	16.5
Consett, Co. Durham	DH08 7	1	.1	.1	16.6
Stanley, Co. Durham	DH09 0	1	.1	.1	16.8
Stanley, Co. Durham	DH09 6	5	.5	.5	17.3
Stanley, Co. Durham	DH09 7	1	.1	.1	17.4
Anfield Plain, Co. D	DH09 8	1	.1	.1	17.5
Dipton, Stanley	DH09 9	1	.1	.1	17.6
Darlington, Co. D	DL01 1	6	.6	.6	18.2
Darlington, Co. D	DL01 2	8	.9	.9	19.1
Darlington, Co. D	DL01 3	2	.2	.2	19.3
Darlington, Co. D	DL01 4	8	.9	.9	20.2
Darlington, Co. D	DL01 5	2	.2	.2	20.4
M. St. G, Co. Durham	DL02 1	2	.2	.2	20.6
Hurworth, Co. Durham	DL02 2	4	.4	.4	21.0
Darlington, Co. D	DL03 0	3	.3	.3	21.3
Darlington, Co. D	DL03 6	4	.4	.4	21.8
Darlington, Co. D	DL03 8	7	.7	.7	22.5
Darlington, Co. D	DL03 9	3	.3	.3	22.8
Shildon, Co. D	DL04 1	1	.1	.1	22.9
Shildon, Co. D	DL04 2	5	.5	.5	23.5
Newton Ay., Co. D	DL05 4	3	.3	.3	23.8
Newton Ay., Co. D	DL05 5	2	.2	.2	24.0
School Ay., Co. D	DL05 6	2	.2	.2	24.2
Newton Ay., Co. D	DL05 7	3	.3	.3	24.5
N'allerton, N. Yorks	DL06 2	2	.2	.2	24.8
Osmotherley, N Yorks	DL06 3	1	.1	.1	24.9
N'allerton, N. Yorks	DL07 8	1	.1	.1	25.0
N'allerton, N. Yorks	DL07 9	1	.1	.1	25.1
Bedale, N. Yorks	DL08 2	2	.2	.2	25.3
Leyburn, N. Yorks	DL09 3	1	.1	.1	25.4
Catterick, N. Yorks	DL10 7	1	.1	.1	25.5
Gunn.Richmond, N.Yks	DL11 6	1	.1	.1	25.6
B Castle, Durham	DL12 8	3	.3	.3	25.9
B Castle, Durham	DL12 9	3	.3	.3	26.3
Stanhope, B Castle	DL13 2	1	.1	.1	26.4
Wolsingham, Co. D	DL13 3	1	.1	.1	26.5
Tow Law, Co. D	DL13 4	1	.1	.1	26.6
B Auckland, Co. D	DL14 0	1	.1	.1	26.7
B Auckland, Co. D	DL14 6	3	.3	.3	27.0
B Auckland, Co. D	DL14 7	1	.1	.1	27.1
B Auckland, Co. D	DL14 8	2	.2	.2	27.3
B Auckland, Co. D	DL14 9	3	.3	.3	27.6
Crook, Co. Durham	DL15 0	3	.3	.3	28.0
Crook, Co. Durham	DL15 9	5	.5	.5	28.5
Spennymoor, Durham	DL16 6	3	.3	.3	28.8
Spennymoor, Durham	DL16 7	4	.4	.4	29.2
Ferryhill, Co Durham	DL17 0	1	.1	.1	29.3
Ferryhill, Co Durham	DL17 8	4	.4	.4	29.8
Ferryhill, Co Durham	DL17 9	2	.2	.2	30.0
Doncaster, S Yorks.	DN07	2	.2	.2	30.2
Scunthorpe, Lincs	DN17	1	.1	.1	30.3
Cinderford, Glos	GL14	1	.1	.1	30.4

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Value Label	Value	Frequency	Percent	Valid Percent	Cum Percent
Farnborough, Hants	GU14	1	.1	.1	30.5
Harrow, Middx	HA02	1	.1	.1	30.6
Stornoway, Lewis	HS01	1	.1	.1	30.7
Springfield, Hull	HU05	1	.1	.1	30.8
Sutton-on-Hull	HU07	1	.1	.1	30.9
Woodmansey, Hull	HU17	1	.1	.1	31.1
Halifax, W Yorks	HX02	1	.1	.1	31.2
Jersey, C.I.	JE03	1	.1	.1	31.3
Milnthorpe, Cumbria	LA07 7	1	.1	.1	31.4
Kendal, Cumbria	LA08 8	1	.1	.1	31.5
Kendal, Cumbria	LA09 5	1	.1	.1	31.6
Kendal, Cumbria	LA09 7	3	.3	.3	31.9
Cartmell, Cumbria	LA11 6	1	.1	.1	32.0
Ulverston, Cumbria	LA12 0	1	.1	.1	32.1
Barrow in Furness	LA13 9	1	.1	.1	32.2
Barrow in Furness	LA14 4	1	.1	.1	32.3
Dalton in Furness	LA15 8	1	.1	.1	32.4
Millom, Cumbria	LA18 5	1	.1	.1	32.6
Ambleside, Cumbria	LA22 0	1	.1	.1	32.7
Ambleside, Cumbria	LA22 9	1	.1	.1	32.8
Windermere, Cumbria	LA23 2	1	.1	.1	32.9
Colwyn, Clwyd, Wales	LL29	1	.1	.1	33.0
Lincoln	LN04	1	.1	.1	33.1
Leeds	LS05	1	.1	.1	33.2
Leeds	LS07	1	.1	.1	33.3
Leeds	LS08	1	.1	.1	33.4
Leeds	LS12	1	.1	.1	33.5
Leeds	LS17	1	.1	.1	33.6
Manchester	M28	1	.1	.1	33.7
Newcastle upon Tyne	NE01 8	1	.1	.1	33.8
Newcastle upon Tyne	NE02 1	3	.3	.3	34.2
Newcastle upon Tyne	NE02 3	2	.2	.2	34.4
Newcastle upon Tyne	NE02 4	2	.2	.2	34.6
Gosforth, Newcastle	NE03 1	9	1.0	1.0	35.5
Ouseburn, Newcastle	NE03 2	4	.4	.4	36.0
N Kenton, Newcastle	NE03 3	3	.3	.3	36.3
Kenton, Newcastle	NE03 4	3	.3	.3	36.6
Gosforth, Newcastle	NE03 5	2	.2	.2	36.8
Elswick, Newcastle	NE04 6	2	.2	.2	37.0
Cruddas, Newcastle	NE04 7	2	.2	.2	37.2
Grainger, Newcastle	NE04 8	2	.2	.2	37.5
Fenham, Newcastle	NE04 9	2	.2	.2	37.7
Chapel Pk, Newcastle	NE05 1	4	.4	.4	38.1
Fenham, Newcastle	NE05 2	4	.4	.4	38.5
Blakelaw, Newcastle	NE05 3	4	.4	.4	39.0
Newbiggin, Newcastle	NE05 4	1	.1	.1	39.1
Byker, Newcastle	NE06 2	4	.4	.4	39.5
Walker, Newcastle	NE06 3	2	.2	.2	39.7
Bensham, Newcastle	NE06 4	7	.7	.7	40.4
Heaton, Newcastle	NE06 5	2	.2	.2	40.7
High Heaton, Newc.	NE07 7	5	.5	.5	41.2
Gateshead, T & W	NE08 0	1	.1	.1	41.3
Teams, Gateshead	NE08 2	2	.2	.2	41.5
Deckham, Gateshead	NE08 3	3	.3	.3	41.8
Bensham, Gateshead	NE08 4	6	.6	.6	42.5
Sheriff Hill, Gates.	NE09 5	4	.4	.4	42.9
Low Fell, Gateshead	NE09 6	4	.4	.4	43.3
Gateshead, T & W	NE09 7	2	.2	.2	43.5
Pelaw, Gateshead	NE10 0	3	.3	.3	43.9
Wardley, Gateshead	NE10 8	7	.7	.7	44.6
Felling, Gateshead	NE10 9	3	.3	.3	44.9
Lobley Hill, Gates.	NE11 0	1	.1	.1	45.0
Dunston, Gateshead	NE11 9	2	.2	.2	45.3
Forest Hall, N Tyne	NE12 0	4	.4	.4	45.7
Longbenton, N Tyne	NE12 8	4	.4	.4	46.1
Palmersville, N Tyne	NE12 9	3	.3	.3	46.4

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Value Label	Value	Frequency	Percent	Valid Percent	Cum Percent
Seaton Burn, N Tyne	NE13 6	3	.3	.3	46.7
Wideopen, Newcastle	NE13 7	2	.2	.2	47.0
Horsley, Newcastle	NE15 0	1	.1	.1	47.1
Pendower, Newcastle	NE15 6	3	.3	.3	47.4
Newby, Newcastle	NE15 7	4	.4	.4	47.8
Lemington, Newcastle	NE15 8	3	.3	.3	48.1
Throckley, Newcastle	NE15 9	3	.3	.3	48.5
Swallwell, Newcastle	NE16 3	1	.1	.1	48.6
Whickham, Newcastle	NE16 4	2	.2	.2	48.8
Marley Hill, Newc.	NE16 5	2	.2	.2	49.0
Burnopfield, Co. D	NE16 6	1	.1	.1	49.1
Chopwell, Gateshead	NE17 7	2	.2	.2	49.3
Otterburn, N'land	NE19 1	1	.1	.1	49.4
Kirkheaton, N'land	NE19 2	1	.1	.1	49.5
Ponteland, N'land	NE20 9	1	.1	.1	49.6
Blaydon, T & W	NE21 4	2	.2	.2	49.8
Blaydon, T & W	NE21 5	2	.2	.2	50.1
Winlaton, T & W	NE21 6	2	.2	.2	50.3
Bedlington, N'land	NE22 5	2	.2	.2	50.5
Cramlington, N'land	NE23 6	5	.5	.5	51.0
Cramlington, N'land	NE23 7	4	.4	.4	51.4
Cramlington, N'land	NE23 8	1	.1	.1	51.5
Cramlington, N'land	NE23 9	5	.5	.5	52.1
Blyth, N'land	NE24 2	1	.1	.1	52.2
Blyth, N'land	NE24 3	2	.2	.2	52.4
Blyth, N'land	NE24 5	4	.4	.4	52.8
Whitley Bay, T & W	NE25 0	4	.4	.4	53.3
Whitley Bay, T & W	NE25 8	4	.4	.4	53.7
Whitley Bay, T & W	NE25 9	7	.7	.7	54.4
Whitley Bay, T & W	NE26 3	4	.4	.4	54.9
Whitley Bay, T & W	NE26 4	2	.2	.2	55.1
Backwood, Newcastle	NE27 0	3	.3	.3	55.4
Howden, Wallsend	NE28 0	1	.1	.1	55.5
Wallsend, T & W	NE28 6	1	.1	.1	55.6
Holycross, Wallsend	NE28 7	1	.1	.1	55.7
High Farm, Wallsend	NE28 8	3	.3	.3	56.0
Hadrian Pk, Wallsend	NE28 9	1	.1	.1	56.1
North Shields, T & W	NE29 0	1	.1	.1	56.2
New York, N Shields	NE29 8	2	.2	.2	56.5
North Shields, T & W	NE29 9	2	.2	.2	56.7
North Shields, T & W	NE30 1	1	.1	.1	56.8
North Shields, T & W	NE30 2	2	.2	.2	57.0
North Shields, T & W	NE30 3	5	.5	.5	57.5
Hebburn, S Tyne	NE31 1	1	.1	.1	57.6
Hebburn, S Tyne	NE31 2	4	.4	.4	58.1
Jarrow, S Tyne	NE32 4	2	.2	.2	58.3
Jarrow, S Tyne	NE32 5	4	.4	.4	58.7
S Shields, S Tyne	NE33 2	1	.1	.1	58.8
S Shields, S Tyne	NE33 3	2	.2	.2	59.0
S Shields, S Tyne	NE33 5	1	.1	.1	59.1
S Shields, S Tyne	NE34 0	3	.3	.3	59.4
S Shields, S Tyne	NE34 6	1	.1	.1	59.6
S Shields, S Tyne	NE34 7	7	.7	.7	60.3
S Shields, S Tyne	NE34 8	5	.5	.5	60.8
S Shields, S Tyne	NE34 9	1	.1	.1	60.9
Boldon Coll, S Tyne	NE35 9	3	.3	.3	61.3
East and West Boldon	NE36 0	2	.2	.2	61.5
Donwell, Washington	NE37 1	2	.2	.2	61.7
Washington, T & W	NE37 2	2	.2	.2	61.9
Washington, T & W	NE37 3	1	.1	.1	62.0
Ayton, Washington	NE38 0	2	.2	.2	62.2
Biddick, Washington	NE38 7	4	.4	.4	62.6
Fatfield, Washington	NE38 8	3	.3	.3	63.0
High Spen, T & W	NE39 2	1	.1	.1	63.1
Ryton, T & W	NE40 3	5	.5	.5	63.6
Crawcrook, Ryton	NE40 4	3	.3	.3	63.9

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Value Label	Value	Frequency	Percent	Valid Percent	Cum Percent
Prudhoe, N'land	NE42 5	4	.4	.4	64.4
Prudhoe, N'land	NE42 6	2	.2	.2	64.6
Riding Mill, N'land	NE44 6	1	.1	.1	64.7
Corbridge, N'land	NE45 5	1	.1	.1	64.8
Hexham, N'land	NE46 1	2	.2	.2	65.0
Hexham, N'land	NE46 2	2	.2	.2	65.2
Haydon Br., N'land	NE47 6	1	.1	.1	65.3
Allendale, Hexham	NE47 9	2	.2	.2	65.5
Haltwhistle, N'land	NE49 9	2	.2	.2	65.7
Morpeth, N'land	NE61 2	1	.1	.1	65.8
Morpeth, N'land	NE61 5	3	.3	.3	66.2
Choppington, N'land	NE62 5	2	.2	.2	66.4
Ashington, N'land	NE63 0	2	.2	.2	66.6
Ashington, N'land	NE63 8	2	.2	.2	66.8
Ashington, N'land	NE63 9	5	.5	.5	67.3
Newbiggin, N'land	NE64 6	1	.1	.1	67.4
Amble, N'land	NE65 0	1	.1	.1	67.6
Hepple, Morpeth	NE65 7	1	.1	.1	67.7
Longhorsley, Morpeth	NE65 8	1	.1	.1	67.8
Felton, Morpeth	NE65 9	2	.2	.2	68.0
Alnwick, N'land	NE66 1	1	.1	.1	68.1
Bolton, Alnwick	NE66 2	1	.1	.1	68.2
Bamburgh, N'land	NE69 7	1	.1	.1	68.3
Mansfield, Notts	NG20	1	.1	.1	68.4
Norwich, Norfolk	NR05	1	.1	.1	68.5
Plymouth, Devon	PL07	1	.1	.1	68.6
Totland, I. O. W.	PO39	1	.1	.1	68.7
Wokingham, Berks	RG11	1	.1	.1	68.8
Bracknell, Berks	RG12	1	.1	.1	68.9
Sheffield	S21	1	.1	.1	69.1
Sheffield	S25	1	.1	.1	69.2
Rotherham, S Yorks	S65	1	.1	.1	69.3
Barnsley, W Yorks	S73	1	.1	.1	69.4
W Norwood, London	SE27	1	.1	.1	69.5
Buxton, Lancs	SK17	1	.1	.1	69.6
Hendon, S'land	SR01 2	1	.1	.1	69.7
Ashbrooke, S'land	SR02 7	2	.2	.2	69.9
Hendon, Sunderland	SR02 8	1	.1	.1	70.0
Grangetown, S'land	SR02 9	2	.2	.2	70.2
Silksworth, S'land	SR03 1	7	.7	.7	71.0
Doxford, Sunderland	SR03 2	5	.5	.5	71.5
Herrington, S'land	SR03 3	3	.3	.3	71.8
Springfield, S'land	SR03 4	3	.3	.3	72.1
S Hylton, Sunderland	SR04 0	3	.3	.3	72.5
Millfield, S'land	SR04 6	1	.1	.1	72.6
Sunderland, T & W	SR04 7	5	.5	.5	73.1
High Barnes, S'land	SR04 8	2	.2	.2	73.3
Grindon, Sunderland	SR04 9	3	.3	.3	73.6
Fulwell, Sunderland	SR05 1	2	.2	.2	73.9
Southwick, S'land	SR05 2	1	.1	.1	74.0
Hylton Castle S'land	SR05 3	2	.2	.2	74.2
Town End Farm S'land	SR05 4	2	.2	.2	74.4
Witherwack, S'land	SR05 5	5	.5	.5	74.9
Monkwearmouth S'land	SR06 0	1	.1	.1	75.0
Whitburn, Sunderland	SR06 7	2	.2	.2	75.2
Seaburn Dene, S'land	SR06 8	3	.3	.3	75.6
Fulwell, Sunderland	SR06 9	3	.3	.3	75.9
Seaham, Co. Durham	SR07 0	2	.2	.2	76.1
Seaham, Co. Durham	SR07 8	3	.3	.3	76.4
Seaham, Co. Durham	SR07 9	2	.2	.2	76.6
Peterlee, Co. Durham	SR08 1	1	.1	.1	76.7
Peterlee, Co. Durham	SR08 2	4	.4	.4	77.2
Peterlee, Co. Durham	SR08 3	1	.1	.1	77.3
Peterlee, Co. Durham	SR08 4	1	.1	.1	77.4
Peterlee, Co. Durham	SR08 5	2	.2	.2	77.6
Duns, Berwickshire	TD11	1	.1	.1	77.7

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Value Label	Value	Frequency	Percent	Valid Percent	Cum Percent
Berwick upon Tweed	TD15 1	1	.1	.1	77.8
Berwick upon Tweed	TD15 2	2	.2	.2	78.0
Penzance, Cornwall	TR18	1	.1	.1	78.1
Middlesbrough	TS01 2	1	.1	.1	78.2
Middlesbrough	TS01 3	1	.1	.1	78.3
Middlesbrough	TS01 4	2	.2	.2	78.5
Middlesbrough	TS01 5	1	.1	.1	78.7
Pallister Pk, M'bro	TS03 0	3	.3	.3	79.0
N Ormesby, M'bro	TS03 6	3	.3	.3	79.3
Berwick Hills, M'bro	TS03 7	2	.2	.2	79.5
Pallister Pk, M'bro	TS03 8	3	.3	.3	79.8
Middlesbrough	TS04 2	4	.4	.4	80.3
Middlesbrough	TS04 3	3	.3	.3	80.6
Linthorpe, M'Bro	TS05 5	2	.2	.2	80.8
Acklam, M'Bro	TS05 6	3	.3	.3	81.1
Acklam, M'Bro	TS05 7	1	.1	.1	81.2
Acklam, M'Bro	TS05 8	7	.7	.7	82.0
Normanby, M'Bro	TS06 0	3	.3	.3	82.3
South Bank, M'Bro	TS06 6	2	.2	.2	82.5
Grangetown, M'Bro	TS06 7	4	.4	.4	82.9
Eston, M'Bro	TS06 9	5	.5	.5	83.5
Nunthorpe, M'Bro	TS07 0	2	.2	.2	83.7
Marton, M'Bro	TS07 8	4	.4	.4	84.1
Ormesby, M'Bro	TS07 9	4	.4	.4	84.5
Coulby Newham, M'Bro	TS08 0	4	.4	.4	85.0
Hemlington, M'Bro	TS08 9	4	.4	.4	85.4
Stokesley, M'Bro	TS09 5	1	.1	.1	85.5
Great Ayton, M'Bro	TS09 6	1	.1	.1	85.6
Redcar in Cleveland	TS10 1	1	.1	.1	85.7
Dalton Park, Redcar	TS10 2	1	.1	.1	85.8
Redcar in Cleveland	TS10 3	4	.4	.4	86.2
Redcar in Cleveland	TS10 4	1	.1	.1	86.3
Marske-by-the-sea	TS11 6	2	.2	.2	86.6
Marske-by-the-sea	TS11 7	1	.1	.1	86.7
Saltburn-by-the-sea	TS12 1	7	.7	.7	87.4
Skelton, Saltburn	TS12 2	6	.6	.6	88.0
Loftus, Saltburn	TS13 4	2	.2	.2	88.3
Runswick Bay, N Yks	TS13 5	1	.1	.1	88.4
Guisborough, Cleve.	TS14 6	4	.4	.4	88.8
Guisborough, Cleve.	TS14 7	4	.4	.4	89.2
Hutton Rudby, Yarm	TS15 0	1	.1	.1	89.3
Yarm, Stockton	TS15 9	2	.2	.2	89.5
Eaglescliffe, Stock	TS16 0	3	.3	.3	89.9
Egglescliffe, Stock	TS16 9	1	.1	.1	90.0
Thornaby, Stockton	TS17 0	4	.4	.4	90.4
Ingleby Barwick, St.	TS17 5	1	.1	.1	90.5
Thornaby, Stockton	TS17 8	3	.3	.3	90.8
Thornaby, Stockton	TS17 9	1	.1	.1	90.9
Stockton-on-Tees	TS18 1	1	.1	.1	91.0
Stockton-on-Tees	TS18 3	1	.1	.1	91.1
Oxbridge, Stockton	TS18 4	1	.1	.1	91.2
Hartburn, Stockton	TS18 5	3	.3	.3	91.6
Stockton-on-Tees	TS19 0	5	.5	.5	92.1
Newtown, Stockton	TS19 6	1	.1	.1	92.2
Fairfield, Stockton	TS19 7	5	.5	.5	92.7
Hardwick, Stockton	TS19 8	1	.1	.1	92.8
Ragworth, Stockton	TS19 9	1	.1	.1	93.0
Norton, Stockton	TS20 1	2	.2	.2	93.2
Norton, Stockton	TS20 2	6	.6	.6	93.8
Stillington, Co. D	TS21 1	1	.1	.1	93.9
Sedgefield, Co. D	TS21 2	1	.1	.1	94.0
Sedgefield, Co. D	TS21 3	3	.3	.3	94.3
Fishburn, Stockton	TS21 4	1	.1	.1	94.5
Stillington, Co. D.	TS21 7	1	.1	.1	94.6
Billingham, Stockton	TS22 5	3	.3	.3	94.9
Billingham, Stockton	TS23 2	2	.2	.2	95.1

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Value Label	Value	Frequency	Percent	Valid Percent	Cum Percent
Billingham, Stockton	TS23 3	1	.1	.1	95.2
Hartlepool, Cleve.	TS24 0	4	.4	.4	95.6
Hartlepool, Cleve.	TS24 8	1	.1	.1	95.7
Hartlepool, Cleve.	TS24 9	1	.1	.1	95.8
Hartlepool, Cleve.	TS25 3	2	.2	.2	96.1
Hartlepool, Cleve.	TS25 4	2	.2	.2	96.3
Hartlepool, Cleve.	TS25 5	6	.6	.6	96.9
Hartlepool, Cleve.	TS26 0	1	.1	.1	97.0
Hartlepool, Cleve.	TS26 8	1	.1	.1	97.1
Hartlepool, Cleve.	TS26 9	3	.3	.3	97.4
Hartlepool, Cleve.	TS27 3	2	.2	.2	97.7
Blackhall Coll Co. D	TS27 4	2	.2	.2	97.9
Wingate, Co. Durham	TS28 5	1	.1	.1	98.0
Trimdon, Co. Durham	TS29 6	2	.2	.2	98.2
Twickenham, Middx	TW02	1	.1	.1	98.3
Wakefield, W Yorks	WF03	1	.1	.1	98.4
Wakefield, W Yorks	WF04	1	.1	.1	98.5
Ossett, W Yorks	WF05	2	.2	.2	98.7
Pontefract, W Yorks	WF08	1	.1	.1	98.8
Castleford, W Yorks	WF10	1	.1	.1	98.9
City of York	YO03	1	.1	.1	99.0
Scarborough, N Yorks	YO13	1	.1	.1	99.1
Filey, N Yorks	YO14	1	.1	.1	99.3
Pickering, York	YO18	1	.1	.1	99.4
Wheldrake, York	YO19	1	.1	.1	99.5
Whitby, N Yorks	YO21	2	.2	.2	99.7
Whitby, N Yorks	YO22	1	.1	.1	99.8
Acomb, York	YO24	1	.1	.1	99.9
Langtoft, N Yorks	YO25	1	.1	.1	100.0
	Total	937	100.0	100.0	

Valid cases 937 Missing cases 0

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TITLE  "PHD-GEOG.SPS = SYNTAX FILE FOR ESD / SER / PSR / DNP / PHD"
SUBTITLE  'updated as of 19.11.98 - BOUNDARIES FROM POST CODES'

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('HU07'=80) ('HU17'=80) ('HX02'=80) ('JE03'=90) ('LL29'=90) ('LN04'=90)
('LS08'=80) ('LS12'=80) ('LS17'=80) ('M28'=90) ('NG20'=90) ('NR05'=90)

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('PL07'=90)  ('PO39'=90)  ('RG11'=90)  ('RG12'=90)  ('S65'=80)   ('S73'=80)
('SE27'=90)  ('SK17'=90)  ('TR18'=90)  ('TW02'=90)  ('WF03'=80)  ('WF05'=80)
('WF08'=80)  ('WF10'=80)  INTO  geog1 .
VARIABLE LABELS geog1 'county'.
EXECUTE .



RECODE post
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('DA12'=90)  ('DD10'=90)  ('DG02'=90)  ('DG14'=90)  ('DG16'=90)  ('DN07'=80)
('DN17'=90)  ('GL14'=90)  ('GU14'=90)  ('HA02'=90)  ('HS01'=90)  ('HU05'=80)
('HU07'=80)  ('HU17'=80)  ('HX02'=80)  ('JE03'=90)  ('LL29'=90)  ('LN04'=90)
('LS08'=80)  ('LS12'=80)  ('LS17'=80)  ('M28'=90)   ('NG20'=90)  ('NR05'=90)
('PL07'=90)  ('PO39'=90)  ('RG11'=90)  ('RG12'=90)  ('S65'=80)   ('S73'=80)
('SE27'=90)  ('SK17'=90)  ('TR18'=90)  ('TW02'=90)  ('WF03'=80)  ('WF05'=80)
('WF08'=80)  ('WF10'=80)  INTO  geog2 .
VARIABLE LABELS geog2 'district'.
EXECUTE .
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TITLE  "PHD-ALL.SPS = SYNTAX FILE FOR ESD / SER / PSR / DNP / PHD / "
SUBTITLE  'as of 01.01.99 - calculations for age, etc.'

COMPUTE age10 = YRMODA(xdate.year(date10),xdate.month(date10)
 ,xdate.mday(date10)) / 365.25 - YRMODA(xdate.year(dob),xdate.month(dob)
 ,xdate.mday(dob)) / 365.25 .
EXECUTE .

COMPUTE time = YRMODA(xdate.year(date11),xdate.month(date11)
 ,xdate.mday(date11)) / 365.25 - YRMODA(xdate.year(date10),xdate.month(date10)
 ,xdate.mday(date10)) / 365.25 .
EXECUTE .

RECODE
  time
  (3.50 thru 4.00=1)  (3.00 thru 3.49=2)  (2.50 thru 2.99=3)  (2.00
  thru 2.49=4)  (1.50 thru 1.99=5)  (1.00 thru 1.49=6)  (0.50 thru
  0.99=7)  (0.00 thru 0.49=8)   INTO  recode2 .
VARIABLE LABELS recode2 'recode2'.
EXECUTE .

COMPUTE age20 = YRMODA(xdate.year(date20),xdate.month(date20)
 ,xdate.mday(date20)) / 365.25 - YRMODA(xdate.year(dob),xdate.month(dob)
 ,xdate.mday(dob)) / 365.25 .
EXECUTE .

COMPUTE age1 = YRMODA(98,13,00) / 365.25 - YRMODA(xdate.year(dob),
xdate.month(dob),xdate.mday(dob)) / 365.25 .
EXECUTE .

COMPUTE age25 = YRMODA(xdate.year(date25),xdate.month(date25)
 ,xdate.mday(date25)) / 365.25 - YRMODA(xdate.year(dob),xdate.month(dob)
 ,xdate.mday(dob)) / 365.25 .
EXECUTE .

COMPUTE diff5 = age25 - age20 .
EXECUTE .

COMPUTE age2 = onset1 + 1900 - xdate.year(dob) .
EXECUTE .

COMPUTE age3 = diag + 1900 - xdate.year(dob) .
EXECUTE .

COMPUTE diff1 = age3 - age2 .
EXECUTE .

COMPUTE diff2 = onset1 - onset2 .
EXECUTE .

COMPUTE diff6 = diag - told .
EXECUTE .

COMPUTE diff4 = diff1- diff3 .
EXECUTE .

COMPUTE age91 = YRMODA(xdate.year(date91),xdate.month(date91)
 ,xdate.mday(date91)) / 365.25 - YRMODA(xdate.year(dob),xdate.month(dob)
 ,xdate.mday(dob)) / 365.25 .
EXECUTE .

COMPUTE age92 = YRMODA(xdate.year(date92),xdate.month(date92)
 ,xdate.mday(date92)) / 365.25 - YRMODA(xdate.year(dob),xdate.month(dob)
 ,xdate.mday(dob)) / 365.25 .
EXECUTE .

COMPUTE age93 = YRMODA(xdate.year(date93),xdate.month(date93)
 ,xdate.mday(date93)) / 365.25 - YRMODA(xdate.year(dob),xdate.month(dob)
 ,xdate.mday(dob)) / 365.25 .
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```
EXECUTE .

COMPUTE diff7 = age93 - age91 .
EXECUTE .

COMPUTE age98 = YRMODA(xdate.year(date98),xdate.month(date98)
 ,xdate.mday(date98)) / 365.25 - YRMODA(xdate.year(dob),xdate.month(dob)
 ,xdate.mday(dob)) / 365.25 .
EXECUTE .

COMPUTE time3 = YRMODA(xdate.year(date98),xdate.month(date98)
 ,xdate.mday(date98)) / 365.25 - YRMODA(xdate.year(date10),xdate.month(date10)
 ,xdate.mday(date10)) / 365.25 .
EXECUTE .

RECODE
  time3
  (6.00 thru 6.4999=1) (5.50 thru 5.9999=2)  (5.00 thru 5.4999=3)
  (4.50 thru 4.9999=4) (4.00 thru 4.4999=5)  (3.50 thru 3.9999=6)
  (3.00 thru 3.4999=7) (2.50 thru 2.9999=8)  (2.00 thru 2.4999=9)
  (1.50 thru 1.9999=10) (1.00 thru 1.4999=11) (0.50 thru 0.9999=12)
  (0.00 thru 0.4999=13) (-38.0 thru -0.0999=14) INTO  recode1 .
VARIABLE LABELS recode1 'recode1'.
EXECUTE .


TITLE  "PHD-DEC.SPS = SYNTAX FILE FOR ESD / SER / PSR / DNP / PHD / "
SUBTITLE  'as of 01.05.98 - decimal places'

COMPUTE age10 = (age10 + 0.5) - (MOD((age10 + 0.5),1)) .
EXECUTE .

COMPUTE time = (((time * 10) + 0.5) - (MOD(((time * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE age20 = (((age20 * 10) + 0.5) - (MOD(((age20 * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE age1 = (age1 + 0.5) - (MOD((age1 + 0.5),1)) .
EXECUTE .

COMPUTE age25 = (((age25 * 10) + 0.5) - (MOD(((age25 * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE diff5 = (((diff5 * 10) + 0.5) - (MOD(((diff5 * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE age91 = (((age91 * 10) + 0.5) - (MOD(((age91 * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE age92 = (((age92 * 10) + 0.5) - (MOD(((age92 * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE age93 = (((age93 * 10) + 0.5) - (MOD(((age93 * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE diff7 = (((diff7 * 10) + 0.5) - (MOD(((diff7 * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE age98 = (((age98 * 10) + 0.5) - (MOD(((age98 * 10) + 0.5),1)))/10 .
EXECUTE .

COMPUTE time3 = (((time3 * 10) + 0.5) - (MOD(((time3 * 10) + 0.5),1)))/10 .
EXECUTE .
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TITLE  "PHD-SF36.SPS = SYNTAX FILE FOR ESD / SER / PSR / DNP / PHD / "
SUBTITLE  'SF36 & EuroQol COMPUTATIONS as of 06.07.97'

COMPUTE ostate1=anx1 + (10*disc1) + (100*act1) + (1000*self1) + (10000*mob1).
EXECUTE .

COMPUTE m1=9.
if (mob1 eq 1)m1=0.
if (mob1 eq 2)m1=0.0665.
if (mob1 eq 3)m1=(0.0665*2)+0.0678.
COMPUTE s1=9.
if (self1 eq 1)s1=0.
if (self1 eq 2)s1=0.0834.
if (self1 eq 3)s1=0.0834*2.
COMPUTE a1=9.
if (act1 eq 1)a1=0.
if (act1 eq 2)a1=0.0953.
if (act1 eq 3)a1=0.0953*2.
COMPUTE d1=9.
if (disc1 eq 1)d1=0.
if (disc1 eq 2)d1=0.0522.
if (disc1 eq 3)d1=(0.0522*2)+0.1369.
COMPUTE x1=9.
if (anx1 eq 1)x1=0.
if (anx1 eq 2)x1=0.0629.
if (anx1 eq 3)x1=(0.0629*2)+0.0825.
missing values m1 s1 a1 d1 x1 (9).
COMPUTE euro1=((1-(m1+s1+a1+d1+x1+0.1716))*100).
FORMAT euro1(F5.2) .
if (mob1 eq 1 & self1 eq 1 & act1 eq 1 & disc1 eq 1 & anx1 eq 1)
euro1=100.00 .
EXECUTE .

COMPUTE ostate2=anx2 + (10*disc2) + (100*act2) + (1000*self2) + (10000*mob2).
EXECUTE .

COMPUTE m2=9.
if (mob2 eq 1)m2=0.
if (mob2 eq 2)m2=0.0665.
if (mob2 eq 3)m2=(0.0665*2)+0.0678.
COMPUTE s2=9.
if (self2 eq 1)s2=0.
if (self2 eq 2)s2=0.0834.
if (self2 eq 3)s2=0.0834*2.
COMPUTE a2=9.
if (act2 eq 1)a2=0.
if (act2 eq 2)a2=0.0953.
if (act2 eq 3)a2=0.0953*2.
COMPUTE d2=9.
if (disc2 eq 1)d2=0.
if (disc2 eq 2)d2=0.0522.
if (disc2 eq 3)d2=(0.0522*2)+0.1369.
COMPUTE x2=9.
if (anx2 eq 1)x2=0.
if (anx2 eq 2)x2=0.0629.
if (anx2 eq 3)x2=(0.0629*2)+0.0825.
missing values m2 s2 a2 d2 x2 (9).
COMPUTE euro2=((1-(m2+s2+a2+d2+x2+0.1716))*100).
FORMAT euro2(F5.2) .
if (mob2 eq 1 & self2 eq 1 & act2 eq 1 & disc2 eq 1 & anx2 eq 1)
euro2=100.00 .
EXECUTE .

COMPUTE pf1=(((vig1 + mod1 + lift1 + sev1 + one1 + bend1 + walk1 + walb1
 +walc1 + bath1)-10)/20)*100.
EXECUTE .

COMPUTE pf2=(((vig2 + mod2 + lift2 + sev2 + one2 + bend2 + walk2 + walb2
 + walc2 + bath2)-10)/20)*100.
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EXECUTE .

COMPUTE rp1=(((cut1 + acc1 + lim1 + had1)-4)/4)*100.
EXECUTE .

COMPUTE rp2=(((cut2 + acc2 + lim2 + had2)-4)/4)*100.
EXECUTE .

COMPUTE bp1=(((how1 + pain1)-2)/9)*100.
EXECUTE .

COMPUTE bp2=(((how2 + pain2)-2)/9)*100.
EXECUTE .

COMPUTE gh1=(((gen1 + ill1 + any1 + wor1 + exc1)-5)/20)*100.
EXECUTE .

COMPUTE gh2=(((gen2+ ill2 + any2 + wor2 + exc2)-5)/20)*100.
EXECUTE .

COMPUTE vt1=(((life1 + lot1 + worn1 + tir1)-4)/20)*100.
EXECUTE .

COMPUTE vt2=(((life2 + lot2 + worn2 + tir2)-4)/20)*100.
EXECUTE .

COMPUTE sf1=(((ext1 + soc1)-2)/8)*100.
EXECUTE .

COMPUTE sf2=(((ext2 + soc2)-2)/8)*100.
EXECUTE .

COMPUTE re1=(((time1 + less1 + work1)-3)/3)*100.
EXECUTE .

COMPUTE re2=(((time2 + less2 + work2)-3)/3)*100.
EXECUTE .

COMPUTE mh1=(((nerv1 + down1 + calm1 + low1 + happ1)-5)/25)*100.
EXECUTE .

COMPUTE mh2=(((nerv2 + down2 + calm2 + low2 + happ2)-5)/25)*100.
EXECUTE .



TITLE  "PHD-QUAL.SPS= SYNTAX FILE FOR ESD / SER / PSR / DNP / PHD / "
SUBTITLE  'qualitative data as of 04.07.97'

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COMPUTE qual3 = qual1 - qual2 .
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SUBTITLE  'for analysis as of 23.06.97 re Marjan'

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  fdq18 + fdq19 + fdq20 + fdq21 + fdq22 + fdq23 + fdq24 + fdq25 + fdq26 +
  fdq27 .
EXECUTE .

COMPUTE fdqsoc = fdq5 + fdq7 + fdq9 + fdq16 + fdq17 + fdq20 + fdq22 + fdq24 .
EXECUTE .

COMPUTE fdqphy = fdq2 + fdq4 + fdq8 + fdq12 + fdq13 + fdq19 + fdq27 .
EXECUTE .

COMPUTE fdqsel = fdq1 + fdq10 + fdq15 + fdq21 + fdq25 .
EXECUTE .

COMPUTE fdqlei = fdq3 + fdq6 + fdq11 .
EXECUTE .

COMPUTE fdqoth = fdq14 + fdq18 + fdq23 + fdq26 .
EXECUTE .

COMPUTE bcsall = bcs1 + bcs2 + bcs3 + bcs4 + bcs5 + bcs6 + bcs7 + bcs8 +
  bcs9 + bcs10 + bcs11 + bcs12 + bcs13 + bcs14 + bcs15 + bcs16 + bcs17 +
  bcs18 + bcs19 + bcs20 + bcs21 + bcs22 .
EXECUTE .

COMPUTE bcsspe = bcs3 + bcs7 + bcs10 + bcs13 + bcs15 + bcs21 + bcs22 .
EXECUTE .

COMPUTE bcspos = bcs1 + bcs6 + bcs8 + bcs9 + bcs16 + bcs17 + bcs20 .
EXECUTE .

COMPUTE bcseva = bcs2 + bcs5 + bcs14 + bcs19 .
EXECUTE .

COMPUTE bcsten = bcs4 + bcs11 .
EXECUTE .

COMPUTE bditot = bdi1 + bdi2 + bdi3 + bdi4 + bdi5 + bdi6 + bdi7 + bdi8 +
  bdi9 + bdi10 + bdi11 + bdi12 + bdi13 + bdi14 + bdi15 + bdi16 + bdi17 +
  bdi18 + bdi19 + bdi21 + bdi22 .
EXECUTE .

COMPUTE sestot = ses1 + ses2 + ses3 +  ses4 + ses5 + ses6 + ses7 + ses8 +
  ses9 + ses10 .
EXECUTE .

COMPUTE pesteem = ses1 + ses3 + ses4 + ses7 + ses10 .
EXECUTE .

COMPUTE nesteem = ses2 + ses5 + ses6 + ses8 + ses9 .
EXECUTE .

COMPUTE iodtot = iod1 + iod2 + iod3 + iod4 + iod5 + iod6 + iod7 .
EXECUTE .

COMPUTE csitot = csi2 + csi3 + csi4 + csi5 + csi6 + csi7 + csi8 + csi9 +
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EXECUTE .
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IF (env EQ 1) envcm = 0.
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IF (alg EQ 2) algm = 2.
IF (alg EQ 2) algn = 2.
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APPENDIX E

OTHER DOCUMENTATION

The following documents are enclosed.

1. Letter from the Doctor in the Cost Utility Analysis inviting patients to participate.
2. The Consent Form - completed by the patient
3. The Clinical Information Form - completed by the researcher for the Epidemiology
4. The Clinical Analysis Form - completed after the Cost Utility Analysis

DYSTONIAS, BOTULINUM TOXIN AND QUALITY OF LIFE

Dystonias are rather unusual illnesses and our understanding of them is far from complete. For instance, it is not known exactly how common they are, what sort of people are affected or how having a dystonia alters their lives.

In the past it has also proved difficult to treat dystonia, although thankfully in the last few years many patients have found Botulinum Toxin injections into or around the overactive muscles to be of great help.

Because of these uncertainties, we are attempting to study all the people with a dystonia who attend the ..... and to examine the effect of the Botulinum Toxin injections upon their quality of life.

If you choose to participate in our study, you will be asked to complete a series of questionnaires. These would ask questions about how you feel and attempt to measure the effectiveness of the treatment. The questionnaire would need to be completed just before a set of injections, once again when the injections are fully active and then again when the next set of injections are due. In total, you will be asked to complete the questionnaire approximately every six weeks for the next six to nine months. The questionnaire would be sent to you in the post with a stamped addressed envelope and it will take no longer than fifteen minutes to complete.

In addition, upon entering into the study, we would like to ask some detailed questions about what sort of person you are. This would be done by means of a short informal interview with Mr Ginger Butler, a researcher for the Dystonia Society, which is a national charity whose aim is to provide support to people affected by dystonia.

All the information gathered by the study will remain absolutely confidential and the medical treatment you are given will be identical whether or not you are involved in it. Hopefully the results of the study will be published in the medical literature in order to guide others involved in the treatment of dystonia, however they would be displayed as statistics and not reveal the identity of any of the individuals involved.

Would you be willing to take part in this study? I would be grateful if you could complete the enclosed form giving your decision and bring it with you on your next appointment. If you would like to discuss the matter further both Mr Butler and I would be happy to oblige when I next see you.

On hospital notepaper,signed by the doctor treating the patient

DYSTONIAS, BOTULINUM TOXIN AND QUALITY OF LIFE

I ..... (NAME)

OF ..... (ADDRESS)

.....

.....

..... (Post Code)

..... (Telephone No)

* AGREE / DO NOT AGREE TO TAKE PART IN THIS STUDY.

SIGNED .....

DATE .....

* PLEASE DELETE WHAT DOES NOT APPLY TO YOU

PLEASE REMEMBER TO BRING THIS FORM WITH YOU WHEN YOU NEXT ATTEND THE CLINIC FOR YOUR BOTULINUM TOXIN INJECTIONS AND EITHER GIVE IT TO YOUR DOCTOR OR ASK TO SEE MR BUTLER WHO WILL BE ABLE TO ANSWER ANY OF YOUR QUESTIONS ABOUT THE STUDY.

Epidemiological Survey of Dystonia (ESD)
CLINICAL INFORMATION FORM

Please complete the following information for **every new patient** who arrives at your clinic throughout 1996. **NB** : This is the minimum information required and it is *essential* for the Epidemiology to have true validity and accuracy that all new patients are logged. Please forward to A.G.Butler, Dystonia Research, Hunters Moor Regional Rehabilitation Centre, Hunters Road, Newcastle. NE2 4NR.

ESD Number : ..... (to be completed by research team)

Hospital No : ..... Hospital : .....

Patient's Full Name : .....

DoB : ..... Marital Status : .....

Address : ..... Gender : .....

..... Tel No : .....

..... Post Code .....

What is the patient's exact Clinical Diagnosis : .....

.....

.....

Date of Onset : ..... Date of Diagnosis .....

Details of any Co-morbidity (if known) include onset dates if poss.

.....

.....

.....

.....

Details of any known medications relating to their dystonia include dosage and frequency (this is <u>not</u> essential information)

.....

.....

.....

Any other comments you would wish to make, eg Dystonia Society info

.....

CLINICAL ANALYSIS FORM

Survey Number ..... Hospital No : ..... Hospital : NRI/HMH

First Interview : ..... Final interview : .....

Dates of Questionnaires	Injection Dates	Dosage	Number
01	.....	.....	.....
02	.....	.....	.....
03	.....	.....	.....
04	.....	.....	.....
05	.....	.....	.....
06	.....	.....	.....
07	.....	.....	.....
08	.....	.....	.....
09	.....	.....	.....
10	.....	.....	.....
11	.....	.....	.....

Exact Clinical Diagnosis : .....

.....

.....

Details of any Co-morbidity : .....

.....

.....

.....

Drug Name	Dosage	Frequency	Cost per dose	Total cost
.....				
.....				
.....				
.....				

APPENDIX F

VERBATIM ANSWERS

The following are the verbatim answers to the question : *"Describe in detail (in terms of hand used, area of head, neck or face touched), the gesture that you use(d) to keep your head straight. Perform this gesture in front of a mirror before answering and write your answers on a separate piece of paper.* "

The numbers in the brackets, eg. (016), relate to the number of the anonymous patient.

(016) I use the tips of the fingers of my left hand to the left side of my chin. I only use light pressure. My head starts to move a little before the fingers touch my chin. I am unable to bend or tilt my neck to the left now. I just turn my whole torso to compensate. My right shoulder is higher than my left and I do not stand as straight as I used to.

(026) On different occasions I would use different geste's, ie., when driving etc., I would have to put my left hand around the back of my neck and kept a continuous strong pull. When socialising, I would smoke cigarettes as the right hand movement to my mouth would help for short periods. I would move my left index finger side to the centre of my chin and lightly touch. None of these geste's are now needed due to the Botulinum Toxin injections, although this period has not been as good as in the past.

(040) By placing my hands behind my neck and pulling neck a little forward.

(046) Left hand on left side of face.

(054) I no longer need to do this as my neck has much improved with the Botulinum Toxin injections.

(057) Depending on the situation ; back of head when driving and crossing road and trying to look left ; right hand on chin when trying to sit still and also when trying to look at someone on the left side of me.

(061) Touch left side of chin with the back of the fingers of the right hand.

(072) Put my right hand against my chin and try to push my head back.

(073) I sometimes walk holding my chin with my right hand.

(075) Hand to mouth.

(077) Using the fingers of right hand on the chin to push the head to the front.

(082) I put my left hand up to my neck and hold it from my jaw line and as far back as my hand will go and push hard and then hold for a couple of seconds and then stop, otherwise it starts pulling over harder and then I find it hard to stop holding my neck.

(083) I used to, before injections, put my left hand down the left side of my face and push hard to keep my head straight.

(088) Right hand to right side of face or drinking (same side).

(090) I feel it's like a heavy weight on my left shoulder, so by lifting my left arm up towards my face, I know my head will turn to the centre and into a straight line with my body. It turns before I touch my face, in fact just thinking about my hand touching my face seems to work. (PS : I hope this makes sense.)

(092) Using right or left hand, raise index finger to mid point of chin, touching very lightly (any pushing causes fighting against it). This gesture moved my head back to normal mid-body position for very short periods. I used this method to enable me to relax in bed and so go to sleep.

(097) I have often tried to keep my head straight with my hand but it never works.

(109) Right hand to right side of face.

(117) When I tried to keep my head straight I used to use my left hand to hold my face at the front and push hard to keep my head straight. My head goes towards the mid-line when my hand pushes hard. My head was normally pulling towards the left.

(122) I sit or stand with my left hand on the left side of my face or chin and push my head or neck towards the right.

(124) Touching left side of chin with left hand.

(142) Left hand on left side of face.

(188) When walking I am almost constantly blowing my nose. This helps to keep my head straight. When in a sitting position, I cradle my head with my right hand.

(207) When sitting upright, if I can lean my elbow on a table I can keep my head still. When sitting upright my head trembles slightly at the moment. I can press with thumb or third finger half way up my neck and it will stop the shaking for a little while or press against the cheek bone near the ear on the right side. When laying in bed, I have the most trouble. I can lay on my right side - no trouble. I can lay on my left side when I have been in bed for half an hour or so. On my back, I always have to put my arm over my head to hold it in place.

(209) In addition to touching, when bad, I had to sit so my head was against a wall or similar (this lady is now in remission).

(221) This patient's answers were defined according to body part :-

1. Right hand :
a) Touching, brushing or resting in the right side of the face, jaw rear
b) Brushing forehead and / or hair (pulls it back)
c) Resting forehead in head
d) Almost any movement across upper part of body
e) Resting walking stick or shopping bag over right shoulder.

2. Left hand :
a) Similar to above, but less effective except when hand is applied to left side of face
b) If I wish to look to the right, eg when about to cross a road or turn towards someone, I tend to force my head around by cupping my chin in my left hand and pushing fairly hard (hence I no longer drive a car).

3. Both hands : Clasped behind my head, especially when seated

Body position :
a) If walking with someone or speaking to someone I always attempt to stand to their right even to the extent of talking to them across my left shoulder as far as possible
b) By standing 'militarily' upright some relief is obtained
c) Deliberate 'floppy' relaxation of all muscles helps temporarily

(230) At present, maybe as a result of BT injections, I can turn my head to centre without using my hand. However, in the past, I've used my right hand on my jaw and pushed gently to the centre.

(252) I use my right hand to cup my chin between my thumb and first fingers and support under my chin with my other fingers. This enables me to keep my face frontal, so I can see where I am walking, but I still don't walk correctly, I seem to sway.

(256) I hold the left side of my face and push with my left hand to put my face straight. I touch my left cheek. I put both my hands on my neck to bring my neck forwards.

(257) I used to hold my chin or side of my face with my right hand for approximately 2 years. I now occasionally hold my head in a more central position by placing my head on top of my head, this gives me slight support.

(263) All I do is lightly touch my neck with my right hand to bring it straight.

(272) I use my left index finger against my chin in order to keep my neck straight.

(279) Right hand to try to push chin back from twisting right and outwards. Left hand to push left temple away from left shoulder.

(307) I would place my left index finger straight against my left cheek with my thumb supporting under my chin and the other fingers of my left hand curled up into a ball.

(317) Whilst sitting down at a table, I need to place my hand under my chin with my elbow on the table for comfort and also to disguise it. I have been wearing a surgical collar for about six years, which I have to wrap scarves around for extra comfort. This enables me to keep my head higher. I have recently been to the clinic in Newcastle and they are developing a better collar for me.

(340) Join both hands behind back initially or half nelson with right hand around the back of neck.

(344) When sitting watching TV, my left hand supports my chin which helps the head from going forward.

(381) Apart from pain, I have tension in my neck that makes my head shake but it doesn't fall to any side. I find that placing my hand on my jaw steadies this tremor.

(382) I use my knuckles in a half clenched fist positioned on the left side of my face to keep my head straight.

(384) Right hand used to push right hand side of face and chin to keep head straight.

(392) I place my middle three fingers of my right hand on the side of my chin and push slightly, this makes my neck feel straight, but I have over the past year discovered that it actually isn't straight though the pulling sensation goes.

(395) Sometimes I use right hand, put straight fingers under cheekbone and thumb on right side of jaw next to chin. Sometimes I just use the back of folded index finger under cheekbone, sometimes it's my open hand with three fingertips on jaw below my chin, with thumb supported on neck. Looking down to read or in between writing, I put closed fist on right side of mouth. Watching television I lock my hands behind my head for a while. I used to put my elbows on my knees and cup my hands around my chin and face to watch television or while sitting in company, but had to stop because shoulders were playing up too much through doing it.

(409) I always had to wear a collar to keep my head up for years, nothing I did would help me. I learned to breathe in, then when I turned my head to the middle I breathed out. It was easier but slow, but it does work when no one is there watching you.

(433) I use my right hand, gently touching or resting against the right hand side of my face, not all of the time but most times.

(440) I put my left hand to the left side of my face.

(443) I place my right hand on my chin when in company, in what is supposed to appear to be a thoughtful manner, to check on my head position. I then correct accordingly. If alone, I occasionally put my right hand on my right jaw line and my left hand on the upper part of the left hand side of my skull to check alignment and I adjust accordingly.

(460) I use my right hand to touch the right side of my face which helps to a certain degree.

(468) Hemi-facial spasm tends to affect the left side of the face more often than the right. But it is my right side which is injected - I am left handed - is there any significance in this ? (This note has been answered by letter)

(484) I touch my neck on the right side with my right hand.

(497) I circle my left arm around the back of my head and hold my left hand on the right side of my neck.

(505) I place my left hand on my left cheek with my fingers touching my ear and my chin resting in the palm of my hand.

(510) Propping up right side of head with my right hand.

(538) Right hand on right side of face with elbow resting on table / desk prevented twitching and jerking of neck for first eighteen months.

(549) By gripping the back of my neck with my right hand and using my left hand when trying to write.

APPENDIX G

REFERENCES

Bibliographical references shown in this thesis use the **Harvard** system of referencing. Whilst every effort has been made to ensure full compliance, some of the medical references have been transferred from other referencing systems, mainly the format of a British Medical Journal 'Original Article' in accordance with **BMJ** 1996: 312: 41-43., notwithstanding this, all references comply to **B.S. 5605** : Bibliographical references.

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