Scottish Needs Assessment Programme

Huntington's Disease, Acquired Brain Injury and Early Onset Dementia

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EXECUTIVE SUMMARY

1. This report considers the health and social care needs of younger adults affected by a group of chronic disabling conditions: Huntington's disease, (HD), Early Onset Dementia (EOD) and Acquired Brain Injury (ABI).

2. In Scotland for every 100,000 population there are approximately 10 to 12 individuals with Huntington's disease; 300 with acquired brain injury and 60 with Early Onset Dementia together with a large number of carers and family members.

3. These conditions have a heavy impact on patients, families and carers with complex health and social needs disproportionate to the numbers of cases. Together they exemplify many of the health and social care needs of those in the community with chronic or progressive disabling neurological disorders and this is one reason why they are considered together in this report.

4. Current services are poorly designed to address many of their needs particularly in the case of children and adolescents. The costs and burden of caring are largely borne by patients, families and carers with the help of the voluntary services.

5. There *are* examples of good, well managed care. However, this is patchy and this report identifies some gaps in services, notably:

   - lack of access to early expert multi-disciplinary diagnostic assessment
   - poor co-ordination of clinical and social care
   - mental health needs are often not recognised or met
   - failure to ensure access to skilled, sustained rehabilitation
   - inappropriate long term care placement and poor access to respite care

6. Detailed information on the numbers of patients in need of care and on their levels of need and access to services is lacking and this prevents effective planning and provision for patients.

7. People with these conditions do not easily fit into current service patterns so that there is usually no natural leader of services for the patients as a group or as individuals. This makes the role of the specialist voluntary agencies particularly important in care planning and provision.

8. The main need is for well co-ordinated services from diagnosis through acute care to rehabilitation with better access to counselling, patient and carer information and advice on services. This means more effective joint working between health, social and voluntary services. Two ways in which this could be improved are:

   - Effectively implemented Care Pathways

- Agreed standards of care

9. The nature of these conditions and the lack of appropriate care pathways mean that most professionals in health and social care do not develop expertise in their management. Better access to expert clinical advice on management and on appropriate Care Pathways is therefore needed to ensure patients have access to high quality health and social care from an early stage.

RECOMMENDATIONS

10. The specialist voluntary agencies should be more involved in planning services and integrating their own services with those of the statutory sector. More involvement of patients and carers in service development could be achieved in this way. Health Boards, Trusts and Social Services should improve communication with these agencies.

11. Agreed Care Pathways are needed by both professional and lay carers. These should be piloted at a national or regional level with input from clinical, social and voluntary services.

12. Agreed standards of care are needed to complement Care Pathways. This should again be developed jointly between health and social services and the voluntary agencies. These standards should be made available to patients and carers along with advice on how to access care. This would create a more focused and informed demand to stimulate development of more effective services.

13. Professionals *and* patients need access to better information (e.g. on Care Pathways, on patient and carer needs, on available services and on care standards) This could be made much more accessible through the use of local area IT networks, wide area networks such as the NHS Net or more widely through the Internet.

14. Patients and carers urgently need help with the choices and decisions involved in moving through a complex care system. Advocacy provided by the specialist voluntary services and by more generic advocacy services is an important method of improving care for patients and should be developed and supported.

15. Mental Health is a priority for the NHS in Scotland and these conditions have a major impact on the mental health of patients and carers. This could start with local audits of current service patterns based on the checklists provided in the appendix to this report and carried out in conjunction with local Social Services and specialist voluntary agencies.

16. Regional centres with expertise in the management of these conditions should be identified as sources of advice and training to extend the availability of their expertise. This could be particularly valuable if planned in conjunction with the voluntary agencies.

17. The use of regional advisers to improve access to expert advice is another effective approach; one taken by the Scottish Huntington's Association and may be more widely applicable.

18. SIGN should consider whether the development of guidelines for the management of these cases is feasible.

19. Many cases of Traumatic Brain Injury could be prevented through better co-ordination of current injury prevention programmes to bring Scotland into line with the lower rates of injury experienced in other areas such as Scandinavia.

20. The data currently available to allow planning of services needs to be improved. This will require dedicated surveys of people with these conditions and their carers and/ or visits to existing facilities to determine the numbers of individuals affected, their current access to services and their preferences for future service development.

CHAPTER 1

INTRODUCTION

The Care Group

21. This report considers the health and social needs of adults and young people effected by three disabling conditions:

- Huntington's disease (HD)
- Acquired Brain Injury (ABI).
- Early Onset Dementia (EOD)

Details of the conditions are given in Chapter 2.

22. These conditions are all primarily neurological. They exhibit many similarities and predominantly affect people under the age of 65 but each also has unique features. For example HD has a dominant genetic basis and is progressive, whilst ABI has no genetic basis and has a recovery process during which relearning and new learning can take place. EOD has a weak genetic element and is progressive.

23. The three conditions, despite the differences, share similarities especially from the perspective of patients, the burden of care in each falling largely on their family and carers over long time periods. They generate major requirements for support and information and their diverse effects on patients mean that many agencies need to be involved in care so that multi-agency working is an important issue. These features are covered in more detail in the report.

24. There are other neurological conditions with a similar impact on patients and carers such as multiple sclerosis and motor neurone disease. These three conditions, however, exemplify many of the health and social care needs of patients with these kinds of disorders. It is likely that, if services for these patients are well designed and managed then patients with similar conditions and needs will benefit and this is another important reason for considering these patient groups together in this report.

Origins of the Report

25. In April 1998 the Scottish Affairs Committee of the House of Commons heard evidence from a number of organisations representing these patient groups including Alzheimer Scotland – Action on Dementia, the Scottish Huntington's Association and the Scottish Head Injury Forum. These bodies joined forces to express concern about certain aspects of the services provided in Scotland for the

patients and carers they represent, particularly those of appropriate long-term and respite care. That meeting led to this report.

26. In 1996 a similar report was prepared on the same groups of patients by the Health Advisory Service in England and this report draws on some of the information provided in that document.

Aims of the Report

27. This report attempts to summarise these patients' health and social care needs, to describe the current state of services in Scotland, highlight where needs are not being met and suggest some ways in which services could be improved.

28. The main audience for SNAP reports is the multi-disciplinary commissioning teams in Health Boards and this report is addressed to them. However, the needs of these patients and their carers are broad and a wide range of services is required. We hope therefore that the report will be useful to those in local authorities and in the voluntary sector who have a key role in this field. Health and Social Services have joint responsibilities for these groups and there is a real need for better joint planning and working.

29. We were particularly fortunate to have on the group which drew up this report representatives of voluntary associations and organisations representing the patient groups in Scotland. They have an intimate knowledge not only of the problems of people with these conditions and carers but also of the current development of services in Scotland.

CHAPTER 2

NATURE OF THE CONDITIONS

HUNTINGTON'S DISEASE

30. This disease bears the name of a physician who described it succinctly in a paper given in 1872, in Ohio, USA. There is evidence in the literature that the disorder was well known to doctors for at least three centuries before that, but as a rare and horrific disease that the families involved rarely discussed.

31. Huntington's disease (HD) is an autosomal dominant disorder and therefore affects both sexes equally and can be inherited by male and female offspring. Each child of an affected person is at 50% risk. In 1993 the mutation was described and confirmation of diagnosis, or prediction of status, can now be provided by using gene technology.

Features of the disease

32. The clinical diagnosis of HD is dependent upon a classical triad of neurological features of choreiform movements, psychological disturbance, and a dementing process. The disease is progressive and tends to begin in the fourth decade, leading to death 15 - 20 years later. The disease can affect children in its juvenile form, and can present as late as the 9th decade. There is no cure.

33. The clinical features result from neuronal loss in the cerebral cortex and deep grey matter. Although chorea has been recognised as a characteristic of HD - and until recently the disease was known as Huntington's chorea - there is widespread neurological disturbance with dystonic and myoclonic features. These features may worsen with time, but often plateau, and can lead to diagnostic difficulties in a family where the diagnosis has been withheld or hidden.

34. Abnormalities of gait are common, with staggering and falling a frequent presentation. Patients are often thought to be drunk, with consequent social difficulties. Ultimately the patient becomes totally dependent on others for care and mobility.

35. Speech difficulties (dysarthria) are universal, and relentlessly progressive. Communication difficulties lead to increasing management problems, as the needs of the patient cannot be understood.

36. Dysphagia (difficulty in swallowing) can lead to choking and in some cases death results from asphyxia or aspiration pneumonia. Weight loss is a common sign in later stages of the disease. Patients eat very slowly because of their motor difficulties, and the movement disorder can cause an increase in calorie requirements, with the need for dietary supplements. There is an additional

unknown mechanism causing the weight loss that can be unresponsive to high calorie foods.

Psychiatric Disorders in HD

37. Many psychiatric symptoms have been associated with HD, most commonly irritability, depression and personality changes which can precede the motor symptoms of neurological symptoms in a quarter to a half of cases (Hayden 1981). Anxiety disorders, panic disorder, sleep disturbance, paranoid illness, schizophrenia, suicide, substance misuse, hallucinations and delusions can all occur at any time during the course of the progression of HD. Mendez gives frequencies of psychiatric behaviours as follows:

Diagnosis	**Range**	**Average**
Depression	9.0 – 44.0%	23.0%
Mania	2.0 – 12.5%	4.8%
Personality	10 – 56.3%	20.2%
Schizophrenia	3.4 – 12.0%	7.8%
Irritability	18.8 – 50.0%	41.5%
Suicides	0.5 – 12.7%	2.6%
Sexual Dysfunction	2.5 – 29.4%	10.8%

Source: Mendez 1994

38. Impaired cognitive functioning often complicates the psychiatric treatment. Irritability, depression and paranoia have impacts on the assessment and treatment of the non-psychiatric components of HD. There is a risk that depression, in particular, is accepted as a consequence of having a diagnosis of HD and not treated sufficiently vigorously. Ringel found only 2% of 306 affected patients were prescribed antidepressant medication. (Ringel 1973)

39. There is also evidence to suggest up to fourfold increase in suicide as well as an increase in attempted suicide. Suicide risk is also elevated in at-risk individuals.

40. Families frequently describe irritability, or poor control over aggressive impulses manifested as shouting, throwing objects, slamming doors or assaulting others. The patients themselves rarely complain of this and it has been under reported in the past. Obsessive, ritualistic behaviour patterns are common, with the patient becoming irritable and distressed if there are changes to his/her routine.

41. Sexual disorders are common but often not volunteered by the partner unless asked directly. Increased libido and disinhibition are common. The divorce rate in

the families is higher than average, most occurring in the years before diagnosis is formally made.

42. The changes in personality and behaviour which occur in HD are often misdiagnosed and misinterpreted by families and professionals thus magnifying the feelings of guilt, anger and regret expressed by carers when they eventually obtain factual information and advice on HD

ACQUIRED BRAIN INJURY

43. The term acquired brain injury (ABI) implies damage to the brain that was sudden in onset and occurred after birth and the neonatal period. It is thus differentiated from birth injuries, congenital abnormalities and progressive or degenerative diseases affecting the central nervous system.

Classification by cause

44. There are several categories of ABI which are generally recognised, of which some are common, others relatively rare. Traumatic Brain Injury (TBI) is often used as the paradigm for ABI and will be in this report but it is essential that other causes, briefly outlined here, are recognised as resulting in similar consequences for the individual affected. The causes are summarised in Table I.

Table 1: Causes of Acquired Brain Injury

Type	Cause
Traumatic (TBI)	Head Injury
Haemorrhagic (HBI)	Bleeding in or around the brain (haemorrhagic stroke)
Vascular (VBI)	Blocked blood supply to brain (ischaemic stroke)
Hypoxic/Metabolic (HMBI)	Interruption of oxygen or glucose supply to brain
Infected (IBI)	Encephalitis, meningitis and brain abscess
Other	Poisons and uncommon complications of other diseases

Traumatic Brain Injury (TBI)

45. To distinguish injuries to the head which involve only the face, scalp etc. from those associated with impact on the brain itself the term traumatic brain injury is increasingly used. Such injuries are further subdivided into

- those in which the skull has been broken and the underlying brain exposed (open TBI)
- those in which a missile or other foreign body has entered the brain substance (penetrating TBI)
- those in which the skull has not been penetrated but damage to the underlying brain has occurred (closed TBI).

Haemorrhagic Brain Injury (HBI)

46. Approximately 15% of cases of stroke are the result of bleeding (haemorrhage) either into or around the brain, the remainder resulting from a blockage of blood supply to part of the brain. Haemorrhages into or around the brain do occur in association with trauma but it is usual to describe these non-traumatic bleedings as spontaneous - either spontaneous subarachnoid haemorrhage (around the brain) or spontaneous intracerebral haemorrhage (into the brain substance)

Hypoxic/Metabolic Brain Injury (HMBI)

47. A number of circumstances can result in the brain being denied oxygen and/or glucose of which both are essential to normal function. The brain is the most sensitive organ in the body in terms of oxygen need and glucose which is vital to metabolism. Lack of oxygen (hypoxia or anoxia) may result from cardiac or respiratory arrest, choking, strangulation, drowning, carbon monoxide poisoning or drug overdose. The commonest cause of lack of glucose (hypoglycaemia) is insulin overdose in people with diabetes. Metabolic disturbances other than lack of glucose include sudden disruption to the body chemistry in association with serious disorders of the liver or kidneys. Finally toxic chemicals may act by interfering with oxygen or glucose supply or poison the brain by another mechanism. For convenience these uncommon cases are often grouped with hypoxic metabolic damage.

Infected Brain Injury (IBI)

48. Infection of the brain (encephalitis or brain abscess) or its surrounding tissues (meningitis) may cause serious disruption to the functioning of the brain. While this may be short-lived and temporary some individuals are left with significant residual problems and are regarded as having 'infected brain injuries'. People whose body defences are impaired (e.g. those with HIV or immunosuppresant drugs) are particularly liable to these conditions.

Vascular Brain Injury (VBI)

49. Possibly the most common non-traumatic form of brain injury is stroke. Stroke results from the interruption of the blood supply to part of the brain as a result of blockage usually related to disease of the arteries.

50. Partly because it is so common and because the pattern of deficits resulting from stroke are generally well recognised and differ somewhat from other forms of brain injury, people with stroke are often catered for in designated stroke services. There is some overlap between those classified as stroke and the category of HBI described above.

51. Some individuals who suffer from stroke, other than HBI, may in fact benefit from being considered as having suffered a brain injury, hence the use of the term vascular brain injury.

52. No classification is without fault or critics. The above is not necessarily fully comprehensive. It is important to emphasise that often more than one mechanism of injury may occur at the same time. Thus a person who sustains a traumatic injury in a road traffic accident may subsequently develop meningitis or suffer a cardiorespiratory arrest. The key is that the individual has suffered a sudden insult to the brain and the classification provides a framework of reference to the types of damage.

Classification by Severity

53. Irrespective of the cause of ABI, it is usual to attempt to describe the extent of damage in terms of severity.

Glasgow Coma Scale

54. In the early hours after injury the depth of coma is considered to reflect the severity of injury and this is measured by the Glasgow Coma Scale (GCS) (Teasdale, 1974). This describes the individual's level of consciousness in relation to stimulation in terms of eye opening, verbal and motor responses. Table 2 gives a brief summary of the classification of severity according to GCS.

Table 2 : Glasgow Coma Scale - Classification of severity

GCS	SEVERITY	OUTCOME
3-8	Severe	Considerable risk of death. Survivors often left with permanent and significant disabilities
9-12	Moderate	Recovery likely but often protracted and residual problems likely
13-15	Minor	Usually make a rapid and complete recovery but significant exceptions with residual problems

55. The GCS only provides a crude indication of the likely longer-term effects. It is increasingly recognised that many classified as having minor or moderate TBI by this scale have significant problems (Hoff 1989). The GCS has been shown to be very useful in predicting early morbidity and mortality but is not so good at indicating the longer-term functional outcome.

Post-traumatic Amnesia

56. In cases of TBI there is usually a period of time after injury when the person is disorientated. This period of loss of memory between injury and the time of regaining continuous memory of day-to-day events is referred to as post-traumatic amnesia (PTA). It is difficult to measure accurately especially if estimated at a later date.

57. However the PTA has been shown to be predictive of the rate of recovery of mental (cognitive) function although less reliable in forecasting outcome at twelve months and beyond (Hall 1999). Nevertheless, as Table 3 suggests, it gives a useful classification of severity and some guidance as to likely outcome although even those classified as minor may have some persisting problems. Measurement of the equivalent period of confusion after other forms of ABI may be of some predictive value although this has not been fully demonstrated**.**

Table 3: Classification of severity of brain injury

Length of PTA	Severity	Outcome
<1 hour	Minor	Full recovery likely in most cases
1 hour – 1 day	Moderate	Full recovery possible but may take months and cognitive problems may persist
1 day – 1 week	Severe	Recovery may be prolonged with cognitive problems but may be full recovery
1 –2 weeks	Very severe	Less chance of full recovery even after many months
2-4 weeks	Extremely severe	Recovery may take a year or longer and permanent deficits increasingly likely
>4 weeks	Extremely severe	Permanent deficits almost certain and chance of full recovery very unlikely

Size, site & nature of brain damage

58. The development of modern brain scanning techniques (CT & MRI brain scans) has made it possible to show parts of the brain which have been damaged and can give clues to the nature of the damage. These scans represent an advance in directing early medical and surgical management of all forms of ABI.

59. These scans can also explain some of the physical and psychological consequences of brain injury by demonstrating that particular areas of the brain responsible for certain functions have been damaged. Extensive brain damage is usually associated with poorer outcome.

60. While the GCS, PTA and brain scan findings allow brain damage to be classified as minor, moderate and severe they only provide general guidance to longer-term outcomes.

The consequences of ABI

61. As the brain is the organ that controls what we think, feel and do, the effects of damage can be complex. ABI may result in impairments of movement (motor function); of sensation (sensory function); of vision, hearing and balance, smell

and taste (special senses); and of heart beat, respiration, the control of bowel and bladder (autonomic function). In addition, however, how the person perceives the world and the abilities to remember, concentrate, reason and judge (cognitive functions) may also be impaired. The person's emotional state may be disturbed and the attributes that constitute their individual personality, and so how others see them, are also frequently altered as a result of ABI.

62. Table 4 lists some of the common consequences of ABI. It is far from comprehensive as the possible effects of brain damage are so wide-ranging.

Table 4: Common consequences of ABI

Function/activity	Examples of difficulty
Physical	Defective vision, hearing, balance, sense of smell Paralysis (loss of movement) of limbs Ataxia (lack of co-ordination) Loss of sensation in a part of the body Difficulty controlling bowels or bladder
Communication	Dysphasia (disordered language- expression or understanding) Dysarthria (disordered articulation – speech intelligibility affected)
Cognition	Memory & orientation Attention & concentration Reasoning & judgement
Emotion	Depression, anxiety, lability. Mood swings, short temper
Behaviour	Apathy- lack of initiative Disinhibition- may be aggressive or sexually inappropriate

Impairments, Disabilities & Handicaps

63. In order to describe the consequences of disease or injury more clearly the World Health Organisation (WHO) published the International Classification of Impairments, Disabilities and Handicaps (ICIDH) in 1980. The definitions are as follows:

"In the context of health experience…."

Impairment: "any loss or abnormality of psychological, physiological or anatomical structure or function"

> *Disability:* "any restriction or lack (resulting from an impairment) of ability to perform an activity in a manner or within the range considered normal for a human being"
>
> *Handicap:* "a disadvantage for a given individual, resulting from an impairment or a disability, that limits or prevents the fulfilment of a role that is normal (depending on age, sex, and social and cultural factors) for that individual"

64. Most people have a mixture of impairments which interfere in various ways with their ability to carry out everyday self-care tasks i.e. result in disabilities. The WHO has recently revised their definitions and avoids the term disability replacing it with the concept of activity limitation. The person with an ABI may be limited in many everyday activities such as dressing and bathing or doing their own shopping.

65. However what is of even more significance is the resulting handicap. This term, which implied social disadvantage, has been supplanted by the concept of participation restriction by the WHO. This emphasises the fact that often other people or physical barriers restrict the ABI individual resuming as full a life as possible rather than this being a direct consequence of their impairments and activity limitations.

66. These consequences of ABI result in particular needs for specially designed services and this is dealt with in Chapter 5.

EARLY ONSET DEMENTIA

67. Dementia affects the brain causing a progressive loss of mental powers. It is formally defined by the World Health Organisation as "a syndrome due to disease of the brain, usually of a chronic or progressive nature, from which there is impairment of multiple higher cortical functions including memory, thinking, orientation, comprehension, calculation, learning capacity, language and judgement. Consciousness is not clouded. The cognitive impairments are commonly accompanied, and occasionally preceded, by deterioration in emotional control, social behaviour or motivation." (World Health Organisation, 1993).

68. Dementia is caused by the death of brain cells in critical areas of the brain. Although it is less common in people under 65, when the illness occurs in younger people it combines with family circumstances to generate particular difficulties for those with the illness and their families. McLennan (1999) explains that 'younger people with dementia are observed as experiencing

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- Declining short term memory
- Symptoms of anxiety and/ or depression
- Personality change with blunting of emotional perception and responsiveness
- Lack of interest and withdrawal from normal activities, including family relationships
- Forgetting appointments
- Inability to perform routine tasks to the same standard as previously
- Poor concentration
- Word finding difficulties
- Repetitive conversation
- The development, at times, of paranoid ideas'

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69. Table 5 summarises the main causes of dementia in people below the age of 65. Alzheimer's disease is the most common cause and with vascular dementia, mixed vascular dementia/Alzheimer's disease and Lewy Body Disease, account for approximately 90% of all causes.

Table 5: Causes of Dementia

Progressive Causes
Alzheimer's Disease
Vascular (Multiple infarction)
Pick's Disease and other lobar dementias
Lewy Body Disease
Huntington's Disease
Creutzfeldt-Jakob Disease (spongiform encephalopathy)
Infection (HIV, neurosyphilis)
Wilson's Disease
Kuf's Disease
Demyelinating Disease

Dementia Due to Brain Damage
Traumatic Brain Injury
Brain Haemorrhage
Brain infection (meningitis and encephalitis)
Hypoxia and hypoglycaemia
Poisoning (lead and other heavy metals)
Alcohol misuse

Potentially Reversible Conditions that may present as Dementia
Hypothyroidism and other endocrine disorders
Drug toxicity
Liver and kidney failure (chronic)
Vitamin deficiencies (particularly B vitamins)
Hydrocephalus
Severe depression

70. An accurate clinical diagnosis is important to ensure the patient does not have one of the potentially reversible conditions that may appear to be dementia and because some treatments depend on the type of dementia in question.

71. Dementia is the fourth most common cause of death in Scotland and is no respecter of social, family or intellectual status. People with dementia slowly lose their memory and understanding. They become confused and often frustrated as they become unable to do the things they used to do. Gradually they become less able to look after themselves and have to rely on others for help. Eventually even dressing, eating or going to the toilet may be impossible without help. Dementia places immense pressure and strain on the members of family who look after relatives with the illness. The impact of this when the person may still be employed and have a young family can be devastating.

72. Cox and McLennan (1994) describe the increased financial loss and disruption, the impact of dementia on adolescents, the effect on social networks and the pronounced physical activity of the younger person with dementia. Some studies have shown that younger people with dementia have a high incidence of depression, anxiety and challenging behaviour. This can lead to high levels of use of services when compared to older people with dementia.

73. As well as looking at the pathological and social consequences of early onset dementia, researchers have also begun to look at the experience of dementia from the perspective of the person with the illness. Keady (1995) has described a nine stage process:

1. Slipping
2. Suspecting
3. Covering Up
4. Revealing
5. Confirming
6. Maximising
7. Disorganisation
8. Decline
9. Death

The importance of this model is that it directs attention to the earlier stages of illness before diagnosis is normally sought.

74. There has been little research into the needs of carers of those with early onset dementia. However, Harvey (1998) has confirmed previous findings of stress and deleterious effects on their health and family life for carers of older people with dementia for younger people with dementia.

75. He points out that family caregiving is a dynamic process that extends over many years and that the following framework can be used to describe the different stages that caring can go through.

- Recognising the need
- Building on the past
- Taking it on
- Working through it
- Reaching the end
- A new beginning

CHAPTER 3

EPIDEMIOLOGY

76. The patterns of disease, their causes and effects is important in assessing the nature of health problems, their scope and severity, the trends over time (are they worsening and if so how fast) and their variability across Scotland. We are particularly interested in incidence - the number of new cases arising in a given time-period, and prevalence- the number of cases in a given population at a stated moment. We also need to be aware of the 'epidemiology of care' i.e. the pattern and intensity of provision of services and this is dealt with in Chapter 6.

Information Sources

77. Sources of epidemiological information available in Scotland include routinely collected health service activity data such as the Scottish Morbidity Record series (SMR) collected by the NHS in Scotland (NHSiS) and that collected in special surveys e.g. the Scottish Health Survey or the decennial census.

78. Routinely collected NHS data are an important source of information for needs assessment but it is important to be aware of the limitations of this kind of information. In particular, they are subject to errors in coding, variations in coding practice in different areas, double counting and inflation. These make interpretation of routine data difficult. Ideally we would know the incidence and prevalence of each condition by place, time, age, sex and severity. In practice we have to take a pragmatic approach drawing on data of variable quality and making informed judgements.

HUNTINGTON'S DISEASE

79. Huntington's Disease is caused by a specific genetic defect. Because of this the best guide to the frequency of the disorder is the systematic study of complete families rather than surveys. HD affects those diagnosed with the condition but also has implications for family members. HD has a disproportionate impact because of the large number of relatives at risk. This has been estimated to be in the order of eight to ten times the number of cases (Simpson 1989).

80. Variations in prevalence of HD between different countries are recognised. This is partly due to population spread from what is thought to have been a common northern European origin. In the UK, HD has been studied more intensively than anywhere else in the world and estimates of prevalence are available for a number of areas of the country. Higher rates are described in areas that have been more intensively studied. Currently quoted rates may still be underestimates because

the social consequences of the condition often cause families to conceal their plight.

81. In Scotland Huntington's Disease (HD) has been studied in detail in Grampian region. This region has a population of 528,400 and a quoted prevalence of HD of 12.49/100,000 (Simpson 1995). This is one of the highest described in the world literature. It reflects an area that is well studied, but suggests that other regions in the UK may have greater numbers of families than is currently recognised.

82. Estimates of prevalence of Huntington's Disease in the UK range from 25 - 100 cases per million population and the accepted prevalence rate is probably around 100 per million. The total number of cases in Scotland is on this basis likely to be around 600.

83. HD affects men and women equally although a number of studies seem to indicate a slight excess of women. The disease affects all age groups and is not one of old age (see additional data given in the appendix). The development of symptoms is often insidious so that the date of onset of the condition in an individual may be difficult to determine. The duration of HD is estimated to be between approximately 11 and 16 years. Simpson found 7 out of 46 cases were diagnosed before the age of 35 and 23 out of 46 were alive over the age of 55. A small proportion of cases (around 5%) develops in childhood very occasionally before the diagnosis is made in a parent.

Routine hospital data

84. Scottish Morbidity record data (SMR 1) are a poor indicator of the incidence or prevalence of the condition but give some idea of the degree of identifiable hospital contact and of trends over time. Table 6 shows the figures for Scotland for the period 1991-96

Table 6. Numbers of discharges from acute general hospitals in Scotland with any mention of a diagnosis of Huntington's Disease

Huntington's Disease by Year of Discharge	Scotland
All Ages	
1991	92
1992	84
1993	114
1994	148
1995	200
1996	184

Source: SMR1/01

ICD coding used: Huntington's Disease ICD9 333.4; ICD10 G10

ACQUIRED BRAIN INJURY

Definition and causes

85. There are particular problems in assessing incidence of prevalence for head injury (Jennet, 1998). Severity of brain injury correlates to an extent with the period of post traumatic amnesia (PTA) but brain injury results in complex and subtle changes in brain function over prolonged periods.

86. Assessment of impairment and the degree of disability require skilled assessment and they may be out of proportion to the apparent severity of injury. Evidence of structural brain injury may be found in cases in which injury and symptoms appear mild. Data on severity of injury are difficult to obtain and interpret and differences in the criteria for the minimum degree of severity required for classification as a head injury or for hospital admission make comparative studies of head injury rates problematic.

87. In assessing need for services data are required on not only the epidemiology of injury and other causes, but also on the epidemiology of subsequent activity limitation (Van Balen 1996). Whilst some information is available on the degree, duration and severity of disability after TBI much less is known about the consequences of the other causes.

Traumatic brain injury (TBI)

88. In Britain the death rate from head injury is low and has been falling since 1968 and was estimated to be only seven per 100,000 in 1994 (Jennet 1998). Male rates are approximately twice those in females and the peak age is from 15 to 30.

89. Admission rates to hospital are a less reliable indicator of head injury incidence or severity because of varying admission policies. In Britain the admission rate varies between 200 and 400 per 100,000 per year in different regions. In most areas 80% of these admissions are classified as mild and only 5 to 10% as severe. The British Society of Rehabilitation Medicine (1998) has estimated that in an average health authority population (250,000) there will be between 250 and 375 disabled survivors of head injury.

90. The aetiology of injuries in genera and head injury in particular has been the subject of intensive research world-wide over the past thirty years. The importance of injury as a threat to health is now well recognised in the public health community. Unintentional injuries are a major cause of death and ill-health particularly in children.

91. Alcohol, violence and road traffic, home and work related injuries are important factors and there is good evidence that preventive interventions can be cost-effective. Previous reports from SNAP have detailed much of the evidence for this.

92. The admission numbers to hospitals in Scotland for the period 1991-7 are shown in Table 7. This data is not linked and episodes may be repeated admissions of one individual. They give some indication of workload and time trends and illustrate the difficulties of determining numbers using coded episodes.

Table 7. Discharges from acute general hospitals in Scotland with any mention of a diagnosis of Acquired Brain Injury, 1991-1997p (including unspecified brain injury)

All Ages	Scotland
1991	16408
1992	16397
1993	15906
1994	17884
1995	17951
1996	17167
1997[p]	16702

Source: SMR 1/01: ICD Coding used: ICD9 854 ICD10 S09.9

93. A study of adults admitted to five hospitals in Glasgow over a one-year period (Thornhill 1998) identified 3005 head-injured adults admitted to one of five Glasgow general hospitals. This covered a population of 909,598 a rate of approximately 330 per 100,000 per year and equivalent to 17,000 per year for the whole of Scotland. Of these, 90% had head injuries rated as mild and 10% were referred to the regional neurosurgical unit. Peak incidence was in young adult males, a third resulted from assault and two-thirds involved alcohol.

94. This study found 150 adults per 100,000 population per year suffering disability at one year following head injury, several times higher than former estimates. Of even 'mildly' injured patients only 45% made a good recovery and 63% of moderate and 85% of severe injuries were disabled at one year.

95. There is little data available on trends in ABI in the UK. Falls in the prevalence of TBI, both major and minor, (from 113 to 79 per 100,000) were seen between 1982 and 1986 in Holland (Van Balen 1996). A quarter of these cases was under 15 years old and a third were aged between 14 and 30.

Disability

96. In the three to seven years following injury in this same series, 67% of those suffering major TBI suffered long term situational, cognitive and behavioural disabilities. Rates of disability after head injury require careful interpretation because of the problems of assessing severity of injury and of subsequent impairment. A household survey in Scotland (Bryden 1989) found a prevalence rate of 100 per 100,000 in the population for 'considerable disability' compared with an incidence rate of two per 100,000 of newly disabled survivors.

Stroke and other forms of ABI

97. In 1998 in Scotland there were 19,847 hospital discharges with a diagnosis of cerebrovascular disease. In 1992 the number of new cases of stroke in Scotland was 15,250 and the total number of people alive with a history of stroke was 53748 (SNAP 1994). Around half of those who survive stroke have significant disabilities and the incidence is strongly linked to age with about 28% of strokes occurring in people under the age of 65 years. (HAS 1996)

98. If half of the approximately 50,000 people alive in Scotland after stroke are disabled and a quarter of these is under 65 years, the number of disabled people under 65 is approximately 6,250 in a population of 5 million – suggesting about 125/100,000 of the population.

Prevalence of Disability after ABI

99. Many will have complex sensory and cognitive problems requiring expert care and rehabilitation but detailed information on this is not available. There are no accurate figures for the other categories of ABI.

100. Combing the estimates for TBI and stroke gives an estimate of 275 (150 +125)/100,000 population. It is reasonable to round this figure to 300/100,000 to take account of other forms of ABI.

EARLY ONSET DEMENTIA

101. Although some writers make a distinction between senile dementia and pre-senile dementia, most no longer do so because the causes of dementia are distinct from the ageing process and the symptoms tend to be the same, whatever the age of onset.

102. Dementia is associated with old age and has over a hundred 'known' causes. The occurrence and mortality of senile dementia of the Alzheimer type (SDAT) has been extensively studied whilst the epidemiology of dementia in those under 65 (EOD) is less well documented.

Incidence and prevalence

103. Alzheimers Disease is the commonest cause of dementia and this is also the case for those in whom the condition starts before the age of 65 (PSDAT). Another important group numerically is those cases due to cerebro-vascular disease or multi-infarcts (VaD).

104. McGonigal (1993) studied the Scottish population between 1974 and 1988 and identified, in this 14-year period, a total of 6581 cases of dementia. Of these a quarter were aged 40 to 64 at onset of dementia (EOD). Of the latter group a

further quarter were 'probable' PSDAT and a fifth were VaD. This study did not estimate point prevalence but calculated incidence of dementia for the 14-year period at between 23 and 41 per 100,000-population aged 40-64 (depending on the definition of dementia used).

105. Harvey attempted to identify every case of dementia in those under the age of 65 in two London boroughs. An overall dementia prevalence rate of 67 cases per 100,000 in the 30-64 year age group was calculated. The prevalence rates per 100, 000 population for specific dementias were:

22	Alzheimer's Disease
11	Vascular Dementia
9	Frontotemporal Dementia
8	Alcohol Related Dementia

106. Hospital contact is a poor indicator for this condition but the SMR 1 figures are given here for comparison. The increase in hospital discharges with a diagnosis of EOD in 1996 and 1997 is notable. It may reflect increased awareness of the condition, a change in clinical practice or possibly a change in coding or counting practice. This requires further investigation.

107. It should be remembered that combinations of these conditions occur. There is overlap between them both in diagnostic terms and with assessment and service development. In particular overlap with mental health services may be significant since there is increasing development of community mental health teams and dementia teams. The interface between all the services needs to be built into joint planning and commissioning processes.

Table 8. Discharges from acute general hospitals in Scotland with any mention of a diagnosis of Early Onset Dementia, 1991-1997

Early Onset Dementia	Scotland
All Ages	
1991	283
1992	306
1993	280
1994	308
1995	369
1996	631
1997[p]	886

Source: SMR 1/01

108. Newens (1993) estimated the point prevalence of dementia in people aged between 45 and 64 in the Northern Region of England in 1986 to be 34.6 per 100,000 in this age range (SDAT) with an additional 23.0 per 100,000 dementias

due to other causes (including VaD). The annual incidence of SDAT was 7.2 per 100,000 in this age range.

109. Woodburn has pointed out that different epidemiological studies give very different figures. Hofman's estimates based on twelve studies conducted in eight European countries are useful as an indication of the likely overall prevalence of dementia in Scotland and also the prevalence estimates for Health Board or local authority areas. (Hofman 1991)

110. The wide variations in estimates of dementia prevalence are mainly due to varying case-definitions in the studies cited and to different approaches to case ascertainment.

111. The earlier SNAP Report on Dementia (1997) uses the Hofman prevalence figures and provides a figure of just over 5,000 for the number of people in Scotland with early onset dementia. This is consistent with the Newens data.

112. Women appear to be more at risk than men for SDAT with a F:M ratio of around 1.7, the reverse of the situation for VaD. The reasons for this are unknown. A small proportion of people under 65 with EOD may be below the age of 45 a figure of 10% has been suggested. (HAS 1996).

Survival

113. The five-year survival of prevalent cases of EOD in the Northern Region study was 42% from the census date and the median length of survival was 4.3 years. This represents a reduction in life expectancy of 30% compared with a population matched for age and sex.

Geographical variation.

114. Table 9 summarises the numbers of cases of these three conditions in the population. These are estimates and should be used with caution bearing in mind the limitations of the data drawn attention to it in the text.

Table 9. Estimated prevalence: HD, ABI and EOD per 100,000 population in Scotland

Condition	Prevalence/100,000	Numbers in Scotland
Huntington's Disease	12	600
Acquired Brain Injury		
• Traumatic	150	
• Stroke	125	13750
Early Onset Dementia		
• Alzheimer type	35	3000
• Other causes	23	

CHAPTER 4

POLICY CONTEXT

115. This report is being written at a time of considerable change in the policies that drive health and welfare provision in Scotland. These policies are intended to modernise the welfare state and, in doing so, have the potential to have a significant impact on the provision of services to meet the needs of young adults with the conditions dealt with in this report.

116. The foundation for community care services for young adults with brain disorders is the NHS and Community Care Act (1990). This legislation gave local authorities lead responsibility for community care planning and recognises that the NHS, housing authorities, voluntary organisations and the private sector are partners in the provision of services. Local authorities were given additional funding that had previously been provided through the social security system for some of the costs of residential and nursing home care. The intention was that, following assessment of a person's needs, they would receive services in the community e.g. day care, home support and only residential or nursing home care if that was what was needed.

117. The Act was implemented in 1993 but, although its objectives included the promotion of domiciliary, day and respite services to enable people to live in their home whenever feasible and sensible, progress has been slower than intended. A Scottish Affairs Committee Inquiry into community care concluded:

> *"No one expected community care to be a cheap option and the funding has proved inadequate to meet the Government's original aims as set out in the 1990 Act".*

118. Community care plans produced by local authorities have given limited attention to client groups that are numerically small. Sometimes younger people with dementia have been included with older people, and people with Huntington's disease are often not even mentioned. Some, but not all, community care plans include people with brain injury as a distinct client group. In part this is because the needs of young adults with brain disorder do not fit easily into categories of mental and long-term physical illnesses and consequently some common needs (e.g. respite care) are not being adequately addressed. Also, the client groupings that are the subject of this report are not routinely used in community care plans.

119. Since the recent change in government a number of initiatives designed to modernise community care and address the problems facing it have been produced. 'Modernising Community Care: An Action Plan' was published in October 1998. This concentrated on key areas where improvements are expected to be achievable in the short term through:

- Better and quicker decision making
- Helping more people get care at home
- Developing more locality based working through new local partnerships
- Setting standards of care
- Ensuring that agencies achieve Best Value
- Considering if the current powers for joint working are adequate

120. Local authorities are required to include a chapter on Acquired Brain Injury in their community care plans. Not all have done so. By consulting about the powers for joint working the Action Plan adopts a cautious approach to the pooling of budgets and joint commissioning which it also advocated. This probably reflects the impracticality of more radical proposals until the new Primary Care Trusts have had time to settle down after their establishment in April 1999.

121. NHS MEL (1996) 22 'NHS Responsibility for Continuing Health Care' is relevant to these client groups because their needs cross the boundaries of Health and Social Care and they are often the subject of debate about whose funding should meet their needs

122. The White Paper 'Aiming for Excellence' (1999) sets out Government proposals for modernising social work services in Scotland. Although its scope is much wider than community care, it will have a major impact on community services for young adults with brain disorders. The key points of the paper are:

- lead responsibility for community care will remain with local authorities
- creation of an Independent Scottish Commission for the Regulation of Care
- creation of a Scottish Social Services Council that will maintain registers of specified groups of staff, regulate professional and vocational education and publish codes of conduct and practice for all staff.

123. The Government's final response to the Report of the Royal Commission on Long Term Care is not yet known. It is primarily about the long-term care needs of older people and as such is a report of considerable potential significance. It proposes, amongst other things, that personal care services should be free at the point of delivery after assessment and that the Government should consider how the provision of care according to need would relate to Independent Living Fund provision for the personal care needs of younger disabled people. This could lead to young adults with brain disorders being given greater opportunities to purchase (and choose for themselves) the services they need rather than being told which services they will receive.

124. The Community Care (Direct Payments) Act may also offer new opportunities for purchase of community care services. The legislation allows local authorities to make direct payments to people assessed as being able to manage their direct payment, with assistance if necessary. However, he or she remains accountable for how the direct payment is spent and no guidance is given about the level of assistance that is acceptable. Local authorities are understandably wary about extending the availability of direct payments to people who will need a great deal

of assistance. This will be a relevant debate for the client groups that are the subject of this report but direct payments may offer opportunities to some people that would not otherwise be available.

125. The final significant policy development for community care services is 'Caring about Carers: A National Strategy for Carers' (1999). This sets out three strategic elements:

- A new charter on what people can expect from care services
- Support and recognition i.e. involve carers in planning and providing services
- Carers' rights to have their own health needs met, new powers for local authorities to provide services for carers as well as those being cared for and respite care

126. Many young adults with brain disorder either permanently or temporarily become incapable of taking decisions for themselves. The current law on mental incapacity is archaic, fragmented and can leave people at risk of abuse. Following an extensive review of the law by the Scottish Law Commission (1995) the Government has announced its intention to include an Incapable Adults (Scotland) Bill in the first legislative programme of the Scottish Parliament. When enacted, these reforms will:

- Increase protection from abuse
- Promote independence
- Respect the view of carers
- Provide ways to give carers powers to make decisions for whom they care

127. Arrangements governing the provision of services within the Health sector are also undergoing a period of transformation driven by a number of recent policy initiatives. These are designed to operate within the wider framework of what is termed Social Inclusion. This aims to provide an overarching policy framework within which co-ordination of the various strategies and implementation plans of statutory, voluntary and independent agencies can act to improve health and personal outcomes for people.

128. The key publications which reflect these initiatives and which provide the statutory guidance and responsibilities to agencies are:

REPORT	PUBLISHED	SOURCE
A Framework for Mental Health Services in Scotland	September 1997	Scottish Office
Designed to Care	December 1997	Government White Paper
Priorities and Planning Guidance for the NHS in Scotland 1999-2004	September 1998	Scottish Office
Towards a Healthier Scotland	February 1999	Government White Paper
Social Exclusion in Scotland	January 1998	Government Green Paper
Social Inclusion Strategy	February 1999	Scottish Social Inclusion Network

129. 'Framework for Mental Health Services in Scotland' was a major initiative designed to maintain and develop effective Mental Health strategies through close multi-agency working. It sets out the government's expectations for the development of services over the ensuing six years:

- Generating consensus over the key issues in achieving transition to local comprehensive mental health services
- Providing a template against which those responsible for mental health services can, *in consultation with the people who receive these services and who care for them,* agree priorities for action that are related to outcomes and to clinical and cost-effectiveness
- Establishing a yardstick by which the Scottish Office can assess local strategies and action plans and monitor progress
- Stressing the importance of involving those who receive mental health services in their own care and in the planning of services. Advocacy has a key role in enabling people to make informed choices about and to remain in control of their own care.
- The SODoH, SWSG and Development departments have worked together and are committed to maintaining this approach in promoting its implementation. In monitoring progress they will expect evidence of greater co-operative working by the NHS, local authorities and other agencies.

130. The Mental Health Framework includes people with dementia. The Framework is being used extensively to implement strategy and is underpinned by the Mental Health Development Fund. The framework therefore offers an opportunity to highlight the needs of those with dementia and other mental health needs who have an associated condition.

131. **Designed to Care** sets out the process for achieving the Government's vision of a modernised and responsive health service. Key principles and issues are:

- Co-operation should replace competition
- Services should be "patient" or "people" centred
- Health improvement, prevention of disease and inequalities in health outcomes should have a high priority
- Health Improvement and Implementation Plans are to be the key mechanisms
- The gap between Social and Health care responsibilities should be narrowed
- Primary Care is to be at the forefront of these changes

132. **The Priorities and Planning Guidance for the NHS in Scotland (1999-2003)** sets out guidance to Health Boards in planning and developing services over the next 3 years. It emphasises Government strategic aims of improving health; tackling inequalities in health care and outcomes; developing Primary and Community Care and reshaping hospital services

133. **Towards a Healthier Scotland** is the policy statement that has been developed from "Working Together for a Healthier Scotland". It seeks implementation through a wide-ranging programme of action.

134. Health Boards and local authorities will need ideally to work together to produce Community and Health Improvement Plans which have synergy. In addition, innovative health improvement and prevention pilots will be funded from a variety of funding streams e.g. Healthy Living Centres.

135. **The Social Inclusion Strategy** represents the broad policy framework for a range of initiatives expected to prevent social exclusion - individuals and communities being excluded from mainstream social benefits and participation. The Scottish Social Inclusion Network was established in 1998 and has representation from Government, Statutory and Voluntary agencies and Community Groups. Action plans are being developed and it is in this context that we should consider how the care needs of young adults with brain disorder can be met more effectively

CHAPTER 5

HEALTH CARE NEEDS: REQUIRED PATHWAYS OF CARE, REHABILITATION AND INTEGRATION

136. Various models have been used by professionals and agencies to meet and co-ordinate the health and social care needs of individuals and their families, including

- Care Programme Approach (CPA)
- Case Management
- Care Management
- Care Pathways

Each of these models has different elements and the approaches are not mutually exclusive. Each aims to meet the needs of the individuals and their families through multi-disciplinary, inter-agency working, co-ordinated care and effective communication.

Care Programme Approach (CPA)

137. The aim of the CPA is to ensure that individuals with severe and enduring mental illness and personality disorders (including dementia) who also have complex health and social needs, receive ongoing care and supervision. There is a requirement placed on Local Authorities and Health Boards to ensure that there are appropriate management arrangements for the planning, co-ordination and implementation of the CPA. (SWSG 16/960)

Integrated Care Pathways

138. Integrated care pathways are structured multi-disciplinary care plans which detail essential steps in the care of patients with a specific clinical problem. They have been proposed as a way of encouraging the translation of national guidelines into local protocols and their subsequent application to clinical practice. They are also a means of improving systematic collection of clinical data for audit and of promoting change in practice. (Campbell 1998)

Care Management

139. The NHS and Community Care Act 1990 introduced sections 12A and 13A into the Social Work (Scotland) Act 1968. These sections place duties on local authorities to carry out assessments of need and provide community care services. Care management is a term used by social work services to describe the process of tailoring services to individual need. Care Managers have key responsibilities associated with this process

Case Management

140. The term "care management" is now used in preference to the term "case management" as it is the care and not the person which is being managed. (DOH 1991)

HUNTINGTON'S DISEASE

Needs of Individuals with HD

141. Adequate social surveys of the views of people with HD are rare, as are studies of the medical needs and provision of care to this patient group. Comments from families to the voluntary family group highlight poor service provision and inadequate co-ordination of such services plus inappropriate resources for long term and respite care

142. There are five main areas to address in the provision of care for these families.

The at risk group

143. The time of discovery of risk is traumatic and often complicated by inaccurate and frightening information. Commonly there is no good source of information available and considerable problems arise as a result.

144. Young people can react to their risk by dropping out of school, drug or alcohol abuse, or by leaving the family home, or all three. Individuals at risk of inheriting HD require comprehensive counselling and access to a specialist who can offer medical information, and referral for clinical genetics advice as required. There is a lack of counselling services available to assist this group, with the exception of the limited resources of the Scottish Huntington's Advisory Services. Many families are unaware of the existence of this group, unless the physician involved with the family has informed them.

145. At risk individuals should have ready access to Clinical Genetics services throughout Scotland where they can receive advice about presymptomatic predictive testing, and undergo such testing if they choose. In addition, there should be access to information about antenatal testing. Clinical Genetics centres exist in Grampian (Grampian University Hospitals Trust at Aberdeen), Highland (provided by Aberdeen at Raigmore General Hospital), Angus (Ninewells Hospital), Glasgow (Glasgow Genetics Services at Duncan Guthrie Institute, Yorkhill Hospital) and Lothian (Western General Hospital). These centres follow a protocol designed by the Huntington's Disease Research Group of the World Federation of Neurology, and which has the approval of the International HD Group (the family organisation). It allows for counselling and support, and sets minimum standards of care for this patient group.

The Pre-symptomatic and Newly Diagnosed

146. Pre-symptomatic Individuals who have been shown to have the gene should be offered regular contact in a clinical environment to monitor for onset of disease. Such monitoring should be by clinical neuropsychological assessment and neurological examination. Access to clinical genetics should be given for discussion about antenatal tests, as well as predictive tests for other family members.

147. Early diagnosis, assessment and referral to appropriate services are regularly voiced wishes of carers and families living with HD. There is evidence of inconsistency of approach and information offered at this stage, with the exception of one dedicated Huntington's Management Clinic offered by the department of Medical Genetics in Aberdeen.

148. Diagnosis should be confirmed by molecular investigation after the possibility of HD has been raised with the family. Carers or partners should be offered an appointment so that information can be given about HD. Clinical genetics advice should be offered to at risk individuals within the family, although it is important to avoid persuading individuals to undergo such tests. The family ought to be made aware of the SHA.

149. Patients have a mixed pattern of neurological and psychiatric features, and misdiagnosis, or late diagnosis is still very common. The average age at diagnosis is in the late thirties or early forties but this can vary widely. Most people are still working at the time of diagnosis, but may be unable to continue in their usual occupation. The decision to divulge the diagnosis to employers, especially since this may result in them losing their job, is not an easy one.

150. Many clinicians, including neurologists, psychiatrists, and general physicians may be involved in making the diagnosis, but consequent regular follow up is rare. Access to localised expertise is rarely available and individuals are therefore deprived of advice and guidance that they require.

The Carer/Partner

151. These individuals often have two roles. They are usually the parent of an at-risk individual as well as the partner of an affected person. The diagnosis is often made at the time of difficult marital relationships, as the personality changes associated with the disease in the early or even prodromal stages of the disease can place considerable strain on the marriage and family unit.

152. Families and individuals living with HD often highlight the problem of obtaining accurate information on Huntington's Disease and on coping strategies, and there are few resources designed for carers in these circumstances. The only literature available within the public domain are the information leaflets and booklets funded and provided by the Scottish Huntington's Association

153. In addition the carer as parent has to decide whether to tell the children about their 50% risk of developing the disease these children have seen developing in their parent. In some cases the carer will not tell, and this causes increasing strains within the family. On occasion a well-meaning friend or relative gives the information to the child with consequent difficulties for the family unit.

154. The carer is often a parent to an individual at risk of HD. Such people require information about predictive testing as well as considerable support as they contemplate their future

155. As the disease progresses the need for respite becomes more desperate, and crisis management often results as the carer reaches the limit of tolerance. There are no designated units for the care of HD in Scotland, and consequently such emergency care may take place in psychiatric units or geriatric units where the skills of the staff are deemed suitable for such patients. This type of care is inappropriate for the patient, and distressing for the family and carer.

156. In order to maintain an affected person within the community, it is important that the carer is fully informed and supported throughout the process of managing the affected individual. Regular respite is vital and support within the home environment should also be considered. The SHA play an important role in the provision of carers support groups and the production of leaflets with information on various aspects of HD.

157. Living with an affected individual causes significant family tensions and many families break up as a result. Counselling for the families should ideally be provided, with community psychiatric involvement where necessary.

Symptomatic Individuals

158. Neuropsychological assessment should take place on an annual or biennial basis together with treatment of depression, speech and language and swallowing assessment.

159. Dietary needs become important and eating becomes increasingly hazardous. Up to three times the average calorie intake can be required, yet eating can be painfully slow, and aspiration a constant risk.

160. Assessment of the home environment should take place as needs dictate. HD is a progressive disorder: needs fluctuate and change with time. Financial advice ought to be made available so that appropriate benefits can be claimed. There is a huge social and financial impact on the person and their family when the diagnosis is made and many social and practical problems result. They often become social outcasts as the choreic movements develop.

161. Mobility becomes more difficult as the disease progresses, with wheelchair provision or carer support necessary when walking. Often psychological difficulties may result in the affected individual rarely leaving the home, and the

carer can be similarly confined as the affected person is too vulnerable to be left alone.

162. The movement disorder can be disabling and require medication. There is a trend to overprescribe for HD, with resulting depression, apathy and sedation. Depression is common, but frequently not recognised.

Respite/Residential Care

163. Plans to use respite care should be discussed. If this is introduced early in the disease process, then the patient will more readily accept the need for such intermittent care, and the family be enabled to cope better with the caring process. Ideally, this unit would be prepared to give palliative care to the patient.

164. End-stage care is frequently provided within an institution, as the physical and emotional aspects of the disease become intolerable for the family. There is evidence that often the staff are not aware of the familial nature of the disease, or are misinformed about it. Nursing staff who care for affected individuals frequently report that the family do not visit. However it is especially difficult if the carer is someone at risk himself or herself, who has to witness what may well be their own future.

165. Flowchart for Huntington's Disease

Managed Care Pathways

Establish Local Clinical Leadership

(either) Neurologist
Geneticist
Neuro-psychiatrist
Psychiatrist

Diagnosis

Genetics → Pre and post Test counselling → Faulty gene not identified

↓

Faulty gene identified

↓

Clinical assessment

↓

Symptomatic/Asymptomatic

↓

Refer to H.D. Management Clinic (early referral advised)
(Usually acute Hospital Based Care)

Neuro Psychological Assessment / Continued assessment
Speech and Language Therapy assessment / Continued assessment
Psychiatric assessment if necessary
(Outreach Services from above)

↓

Primary Health Care / Community Care
(multi/interdisciplinary/interagency working)

GP
Care Programme Approach
Expert Care Co-ordinators
Huntington's Advisory Service (Scottish Huntington's Association)
Dietetic Services
Physiotherapy (Rehabilitation Teams)
Occupational Therapy
Community Psychiatric Services
Specialist social workers
Housing
Benefits/employment advice
Appropriate Respite / long term care/palliative care
Advocacy
Carers Support

Specialist Services	Memory Clinic / Cognitive dysfunction clinic
Other specialist information.	Advice about driving

N.B. Evidence indicates that the most dynamic force for change is the existence of a clinical team with a special remit for Huntington's Disease. Training is also centrally important to the improvement of services for people with Huntington's Disease and their families.

An illustrative case study for an individual with HD is provided in Appendix 4.

ACQUIRED BRAIN INJURY

166. There is a no one common pattern of recovery in acquired brain injury. However, taking severe head injury (TBI) as an example, an outline of required pathways of care can be developed and some examples of this are presented here. Throughout the process care should be focused on the family rather than simply on the injured person.

The need for specially designated services

167. Those who survive ABI present with a complex mixture of physical, cognitive, emotional and behavioural disorders. Medical services have traditionally been divided according to specialties dealing with different groups of symptoms and signs. Thus people with disorders resulting in physical problems are seen by physicians and surgeons often in general hospitals; those with mental illness (disturbed emotion or behaviour) by psychiatrists in psychiatric hospitals; and those with cognitive difficulties by specialists in learning facilities.

168. This has meant that people who have a mixture of physical, mental and cognitive problems do not fit neatly within the system and so all too often have received poor levels of care. These issues remain relevant after the person leaves hospital with regard to accessing health services but also in relation to attending day facilities run by the social work department or voluntary sector.

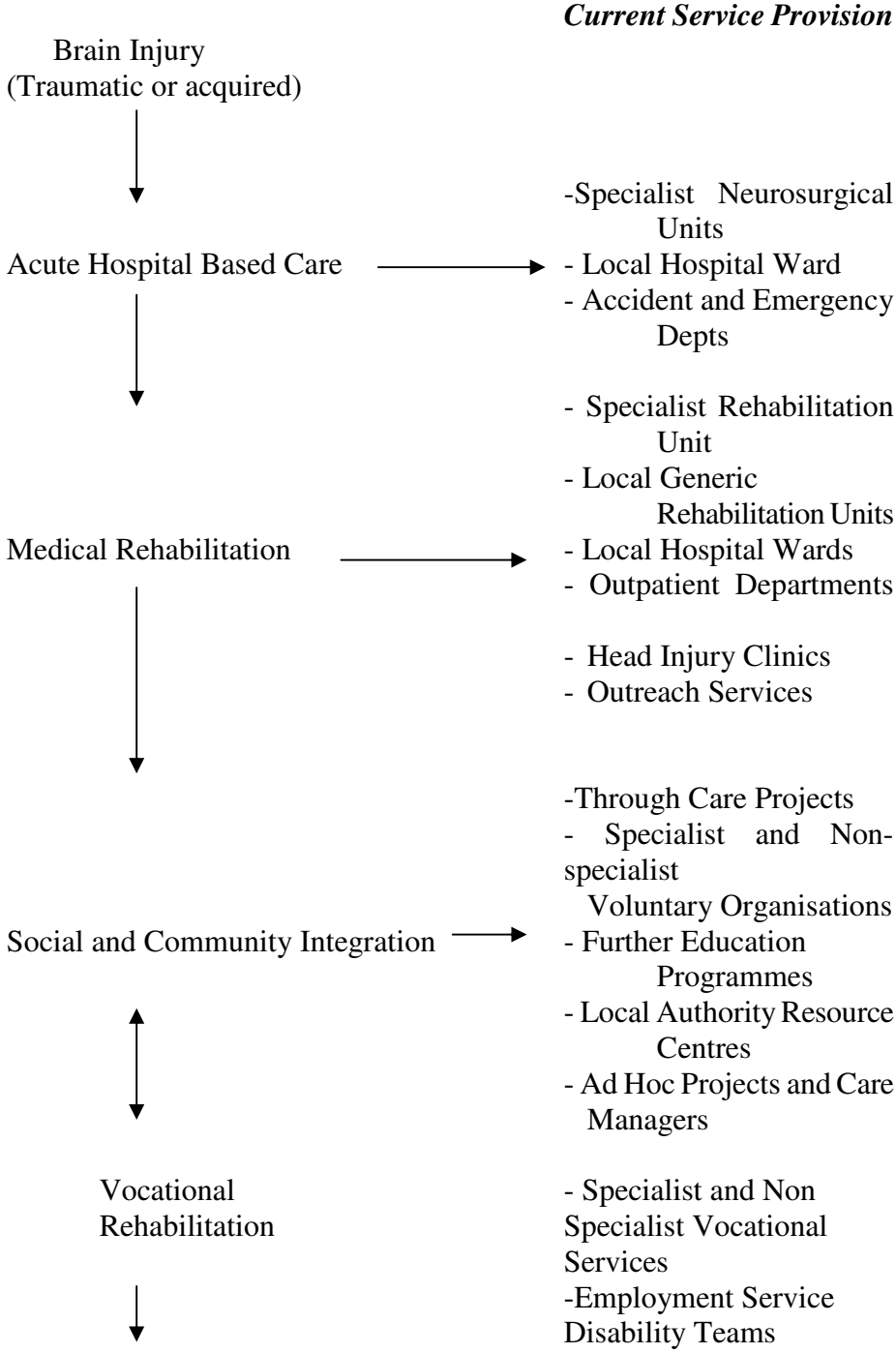
169. The need for designated services designed to address the particular needs of the ABI group has been recognised for many years but only in the last decade or so has some such provision been made (Helios Guidelines for Good Practice 1996). It remains patchy and poorly co-ordinated.

The consequences for families

170. ABI impacts on the family or closest friends of the individual affected often to a dramatic extent. Indeed the person who suffers the brain damage may lack insight into their circumstances and exhibit no distress or suffering. In contrast the parent, spouse or child of that individual may be faced with supporting someone who bears little resemblance in terms of behaviour, function and even appearance to their loved one and they are only too well aware of what is involved.

Acquired Brain Injury

Flow Chart: pathways of care in ABI



Outcomes

- Social/Community integration
- Independent/supported living/long term care
- Family/Carer Support
- Open or supported employment

An illustrative case study of ABI is provided in Appendix 4

171. The nature of care and management will be determined by the individual and depend upon their specific needs. They may require transfer to a specialised brain injury rehabilitation unit or at least assessment and management by a multi-disciplinary team comprising specialist medical, nursing, remedial therapy, psychology and social work staff. In addition to continuing the efforts to prevent avoidable complications, the team aims to assist the individual to make the most of their recovery, adapting to persisting disabilities and plan their return home and reintegration in the community.

172. The great majority of brain injured people will return to live in the community, usually in their own home. Some may spend a relatively short time in hospital while others can be hospitalised for weeks or months. In all cases discharge should be planned – preparing the person for the community and organising the community for the person. Communication and co-ordination between the hospital rehabilitation team and community agencies is essential with the injured person and their family fully involved in the process.

173. A range of different models of community based transitional living facilities should be considered where:

- Personal, cognitive and/or behavioural difficulties prevent early discharge from hospital-based care to family or carer
- Family breakdown or insecure housing impedes the rehabilitation process
- Community rehabilitation is needed to follow on from specialist rehabilitation

174. Independent living activities can then be undertaken in a safe and normalised environment before moving people on to other independent living options. In some instances, people with severe behavioural difficulties will require specialist rehabilitation. In relatively few cases, the combination of a high level of medical and nursing care needs and family circumstances may require long term placement in a residential setting.

175. Following hospital discharge, patients and family often face the greatest challenges. The process of rebuilding lives may change life styles and roles. Thus if the injured person is a husband and father support may be needed to help him resume these roles; he may have to adjust to no longer being able to drive, follow previous leisure activities or return to previous employment. A range of specialist advice may be required from family counsellors, vocational rehabilitationists, and voluntary agency workers from organisations such as HITS and Headway.

176. The specific aim of community-based, social and vocational rehabilitation is to enable the person to live as independently as possible and to be re-integrated back into their community, work and society in general. It is well established that the residual deficits of a cognitive and personal nature have to be carefully and specifically addressed if community and vocational integration are to work. Early

intervention is required to help the person adopt a range of strategies to compensate for residual loss of function. Family and friends often fade away as the realisation of personality change hits home. Family and individual counselling with accurate and honest information will aid healthy adjustment and minimise the likelihood of personal relationship failure.

177. Research supports the view that without a focussed, multi-skilled and specialist approach to the idiosyncratic mix of cognitive, emotional and/or psychological deficits common with brain injury, return to work rates are very poor. For a proportion of patients return to work may be unrealistic but, by building on learned cognitive, emotional and social strategies, it is possible to improve community re-integration by supporting people into leisure, voluntary or further educational opportunities. Services should be developed to address pre-vocational issues and vocational services should adopt a "job coaching" or "place and train" model with individualised support for the person, employers and co-workers over varying periods of time (Brookes 1987)(Kreutzer 1990)

178. The family and carers have a crucial role to play in the rehabilitation of the individual. They require support, and crucially, involvement throughout all stages, but are particularly important once the person has left hospital. With the right level of advice and support, family members and partners or carers can help maintain routine, prevent over-dependency and encourage social and community integration. There are particular problems in managing children and adolescents with ABI and no special services for these age groups currently exist.

179. The overall strategy of the rehabilitation process with regard to brain injury is to recognise that a continuum of care, rehabilitation and integration is necessary. The aim is not cure, but to minimise impairment and then to help both the individual and significant others to make healthy adjustment. This should lead onto developing compensatory strategies and the relearning of a range of social and personal skills which will aid integration. Services should be flexible and provide the opportunity for individuals to access appropriate support, as needs change over the long term.

180. Although this example has been that of a severe head injury, it applies to other forms of severe injury. However, even when the brain injury has been ostensibly mild it is essential to detect deficits that, if unrecognised, may cause the individual problems in getting back to a full life. This could be achieved by screening all brain-injured individuals at discharge or shortly thereafter and/or by ensuring easy access to advice and support for all cases when problems are encountered.

EARLY ONSET DEMENTIA

181. Individual pathways of care for people with early onset dementia will depend on:

- how people make contact with services
- the stage of the illness when diagnosed
- family and other social circumstances
- severity of problems caused by symptoms
- the presence of other illnesses or disabilities.

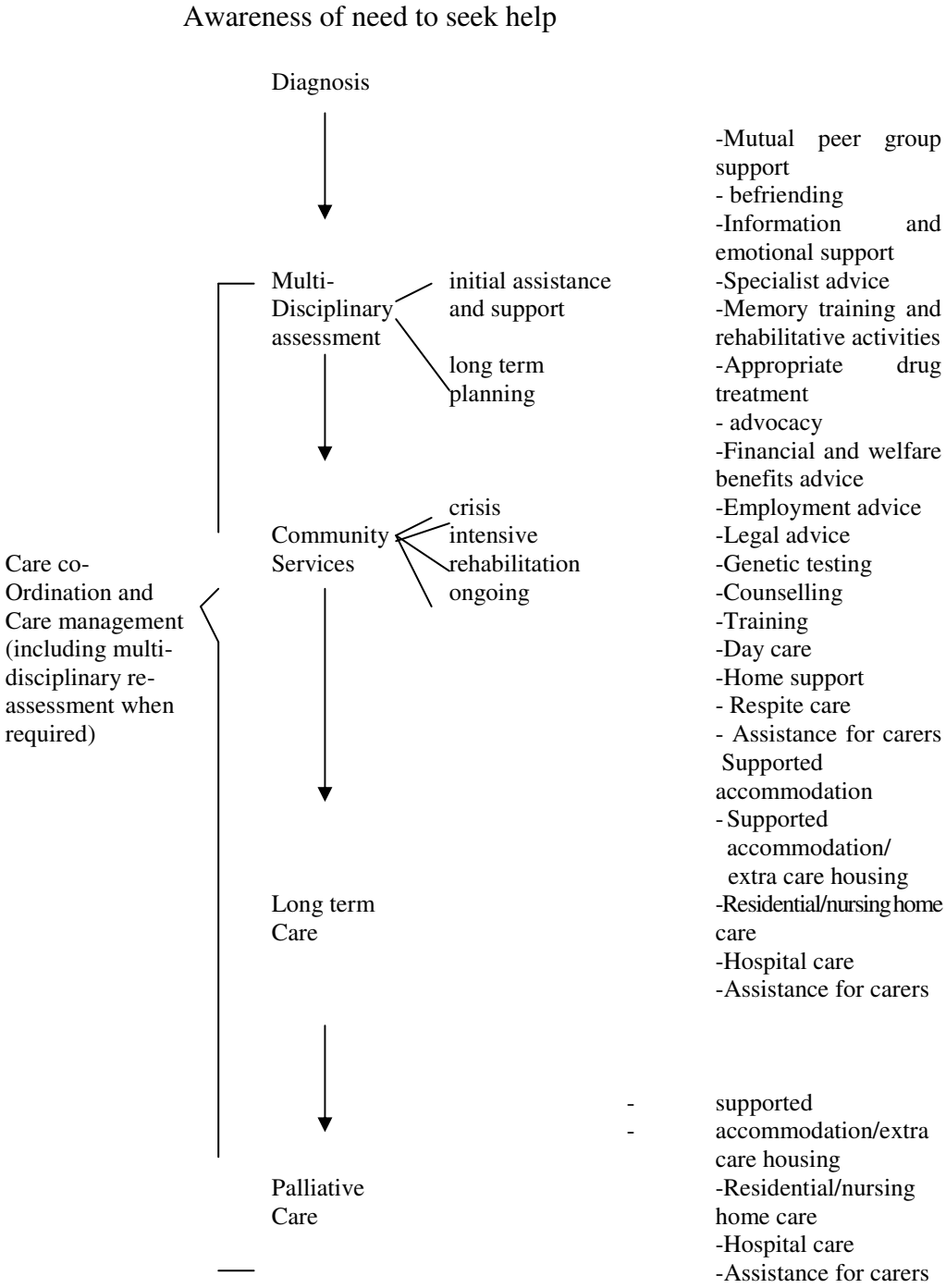
182. The illness may last for up to 12/15 years and the services required will change as the illness progresses. Hence it is important that people with dementia and their carers have access to sensitive and responsive services.

183. The first hurdle is a diagnosis. This may be delayed if the person with dementia is unwilling to seek help from their GP or is reluctant to discuss their growing concerns about emerging difficulties. Services such as housing and social work may not recognise the significance of early symptoms. It is these factors that led Cox (1999) to recommend that "sensitising the community and mainstream services to the needs of people with dementia of all ages" is a critical issue if a more positive system of care for people with early onset dementia is to be designed.

184. Once into the system it is important to ensure that services are continuous rather than disjointed. Multi-disciplinary assessment should be available throughout the course of the illness and not just at the diagnosis stage. The current division of responsibilities between health and social work is unhelpful in this respect unless there has been agreement about lead agency responsibility for dementia care or a detailed protocol has been drawn up allocating responsibilities and ensuring that there is effective inter-agency communication.

185. The key tool for ensuring that the person with dementia is properly supported is an up-to-date individual care programme.

Flow Chart: Pathways of Care in EOD



186. A case study of an individual with EOD is provided in Appendix 4

187. These services need to be sufficiently specialised to understand the needs of people with early onset dementia and skilled enough to respond appropriately.

The relatively small number of people with early onset dementia means that only the larger local authorities will have sufficient numbers to justify specialist projects. This points to the need for specialist projects to operate across local authority boundaries and use outreach workers to ensure that services are accessible to all people with early onset dementia. It is also desirable to be able to provide specialised services within community settings and mainstream services where this can be achieved. This is particularly important for people with early onset dementia because it helps them feel that services are designed with their needs in mind and not just provided for older people with dementia because it would be more convenient.

188. The following local authorities have been estimated to have more than 150 people with dementia under the age of 65 in their areas:

Aberdeen City	Glasgow City
Aberdeenshire	Highland
Dumfries and Galloway	North Lanarkshire
Dundee City	Renfrewshire
Edinburgh City	South Lanarkshire
Fife	

Summary

189. The preceding chapter has outlined the care needs of people with these conditions.

190. Check lists are provided in the appendix for health or social services considering the coverage of care services in their area.

CHAPTER 6

CURRENT PROVISION

191. The concept and development of rehabilitation has evolved over time in response to a range of factors. Historically, the term was used for physical methods of treatment applied to restore function and fitness after disease or injury e.g. patients with rheumatological problems or to assist the war wounded to return to active war service.

192. Rehabilitation has been extended to an increasing number of specialities e.g. cardiology, respiratory, neurology and medical services for older people amongst others. Such developments have occurred in response to clinical enthusiasm as opposed to clear strategic planning.

193. Other factors include demographic changes, policy agendas of both the NHS and Social Work Services and increasing professional recognition of the importance of rehabilitation in improving quality of life. Also, there has been increasing emphasis in the NHS on ensuring efficient use of scarce health service resources in a context of reduced length of stay in acute hospital settings and greater emphasis on community-focussed health care.

194. The concepts of care managers and care management tailoring services to individual needs on a needs-led basis rather than a service-led system of fitting people into services was promoted as an opportunity for developing rehabilitation within the context of community and within long term care.

195. Whilst there has been increasing recognition that rehabilitation should form part of mainstream clinical services and an integral element of both acute and long term medical conditions, the locus of control and physical location of rehabilitation emphasises a medical model of rehabilitation. Proponents of a social model of disability argue that disability is "socially created" and there is a need to adopt a different approach to rehabilitation which focuses on reducing environmental barriers to independent living and enables individuals to take control of the rehabilitation process.

196. In practice, there has been increasing recognition of both the clinical and social component of rehabilitation with a specific focus on restoration of physical, psychological, social, environmental, educational and occupational capabilities. Rehabilitation is a function of many of the services previously described and is not the sole prerogative of rehabilitation services that are explicitly named as such.

197. Deficiencies in availability and distribution of rehabilitation facilities in Scotland have been acknowledged, as is the need to extend and develop dedicated rehabilitation services nationally. (Rehabilitation Services in Scotland).

198. A review of effective practice in rehabilitation identifies that the clinical effectiveness and research base for rehabilitation is ill understood and is compounded by the "uncertainty about the role and place of rehabilitation in modern health care delivery. This may exacerbated by the low public appeal for a 'low-tech' type of health care". (Effective Practice in Rehabilitation – the evidence of systematic reviews).

199. The need for further research and service evaluation as part of a quality improvement approach should be a means of developing and monitoring the practice of rehabilitation

HUNTINGTON'S DISEASE

Specialist Services

200. There are currently no specialist residential or day units in Scotland for people with Huntington's Disease where assessment, counselling and appropriate management can take place.

201. There is only one dedicated management clinic for people with Huntington's Disease in Scotland, and this exists in Grampian region. There is close liaison with a clinical neuropsychologist, speech and language therapist and a Huntington's Advisor from the Scottish Huntington's Association, as well as dieticians, occupational therapists and social work care managers. Attendance at this outpatient clinic allows monitoring for onset of disease, offers support for the family, and updates in current research.

202. In Glasgow there is a pilot service based on the above model, operated by a Senior Registrar in Psychiatry in conjunction with the Clinical Genetics Services. This is not a dedicated clinic and the long term future beyond the placement term of the Senior Registrar is unknown.

203. The Sue Ryder Nursing home in the Scottish Borders has developed some expertise in managing end-stage Huntington's Disease. The rural geographical location is not always acceptable to families. In addition, as it is the only such unit in Scotland, there is limited availability.

204. The disease affects all age groups, and is not unique to middle age. The provision of care is haphazard, and until recently many more individuals were cared for inappropriately in psychiatric and geriatric hospital units.

Advisory Services

205. The Scottish Huntington's Association is developing a network of Specialist Huntington's Advisory services to provide information, support and expertise for individuals and families living with Huntington's Disease in Scotland. At present these services operate an open referral system and provide one Advisor to cover a wide geographical area acting in an Advisory capacity to families affected and

professional caregivers. The current service provides an Advisor in the following areas:

- Ayrshire (Commenced April'99)
- Angus
- Fife
- Glasgow
- Grampian
- Highlands (Commenced Feb'99)
- Lothian

206. The Scottish Huntington's Association is regularly contacted by professional staff and carers asking for advice regarding facilities for assessment, review, day care, respite, long term care and terminal care for people with Huntington's Disease. This problem is compounded by the lack of appropriate research into the health and social impact of living with HD and by the lack of accurate epidemiological evidence on prevalence and incidence of HD in Scotland.

ACQUIRED BRAIN INJURY

Introduction

207. There is a range of services funded in a number of ways and provided by Health Trust, Social Work Services, the Voluntary Sector and the Independent Sector. The geographic inequity of these services is indicative of the lack of strategic planning and co-ordination. The absence of service standards is highlighted by the variety of services with different aims and objectives

208. **Acute**

There are four neurosurgical units in Scotland:

UNIT	LOCATION	DESIGNATION
Southern General Hospital	Glasgow	Health Trust
Western General Hospital	Edinburgh	Health Trust
Aberdeen Royal Infirmary	Aberdeen	Health Trust
Ninewells Hospital	Dundee	Health Trust

209. **Post Acute Rehabilitation**

There is a range of post acute rehabilitation facilities at national and local levels providing specialist brain injury rehabilitation and generic rehabilitation:

National - Specialist		
Scottish Brain Injury Rehabilitation Unit	Astley Ainslie Hospital, Edinburgh	Health Trust
Scotcare Brain Injury Rehabilitation Unit	Murdostoun Castle, Bonkle, Wishaw	Independent
Robert Fergusson Unit	Royal Edinburgh Hospital, Edinburgh	Health Trust
Local-Specialist		
Brain Injury Rehabilitation Services	Dundee	Health Trust
Local-Generic		
Physical Disability Rehabilitation Unit	Southern General Hospital, Glasgow	Health Trust
Rehabilitation Services	Woodend Hospital, Aberdeen	Health Trust
Sir George Sharp Unit	Cameron Hospital, Fife	Health Trust
Rehabilitation Services	Raigmore Hospital, Inverness	Health Trust
Douglas Grant Unit	Ayrshire Central Hospital, Irvine	Health Trust

210. **Through Care*, Outreach Nurse Services and Head Injury Clinics**

UNIT	LOCATION	PROVIDER SECTOR
Through Care	Glasgow Royal Infirmary, Glasgow	Social Work
Through Care	Southern General Hospital, Glasgow	Social Work
Through Care	Proposed West Lothian Ability Centre	Social Work (new development for 1999)
Outreach Nurse, Scottish Brain Injury Rehabilitation Unit	Astley Ainslie Hospital, Edinburgh	Health Trust
Head Injury Clinic	Glasgow Royal Infirmary, Glasgow	Health Trust
Head Injury Clinic	Royal Victoria Hospital. Dundee	Health Trust

*Through Care is based on the concept of providing a service link between hospital and community. Projects vary in character but usually provide elements of assessment, care or case management and co-ordination of resources

211. **Community Based Rehabilitation**

Information and Support Services Service	Location	Provider Sector
Head Injuries Trust for Scotland	Glasgow, Grangemouth, Dumbarton, Galashiels, Broughty Ferry	Voluntary Sector
Headway	Ayr, Dumfries, Dundee, Edinburgh, Fife, Perth, Glasgow, Grangemouth, Gourock, Inverness, Airdrie, Motherwell	Voluntary Sector

Social & Community Integration Services		
Head Injuries Trust for Scotland	Glasgow, Dumbarton, Galashiels, Grangemouth	Voluntary Sector
Headway	Ayr, Edinburgh, Dumfries	Voluntary Sector
Dirrans Centre	Kilwinning	Social Work
Brain Injury Project	Motherwell	Social Work
Brain Injury Services	Dunfermline	Social Work
Connections	Edinburgh	Voluntary Sector
Rehab Scotland	Glasgow	Voluntary Sector
Pre Vocational and Vocational Services		
Rehab Scotland	Glasgow, Kirkcaldy, Aberdeen	Voluntary Sector
Physical Disability Rehabilitation Teams		
Forth Valley Area Rehabilitation Service	Forth Valley Health Board Area	Health Trust
Glasgow North Physical Rehabilitation Team	Greater Glasgow Health Board Area – North of River Clyde	Health Trust
Glasgow South Physical Rehabilitation Team	Greater Glasgow Health Board Area – South of River Clyde	Health Trust

212. **Others**

Designation	Location	Provider Sector
Acquired Brain Injury Worker	Angus	Social Work
Care Manager	Dundee	Social Work
Imprint	Edinburgh	Further Education
Neurorehabilitation Team	Inverclyde	Health Trust and Social Work
Head Injury Officer	East Renfrew	Social Work

EARLY ONSET DEMENTIA

213. There are few services in Scotland that are targeted on the needs of the approximately 3-5000 people with early onset dementia. Consequently these people are fitted into a mixture of neurology, psychiatry, old-age psychiatry and general services for older people as far as the health service is concerned and mental health and older people's services provided by local authorities.

214. The following services specific to EOD are known:

LOCATION	FACILITIES
Marshall House, Edinburgh NHS Trust	Long term nursing home and respite care (men only)
The Early Onset Dementia Project, Viewforth Resource Centre, Kirkcaldy	Day care and some outreach support, Fife Social Work
Dementia Initiative Scheme (Falkirk, Stirling and Clackmannanshire) Joint Social Work and Forth Valley Health Board project	• 1 to 1 home support scheme • Stirling and Clackmannanshire Early Onset Carers' Group (joint initiative with Alzheimer Scotland)
Glasgow Younger Person's Project, Alzheimer Scotland	Home support, day care, counselling and befriending
Lothian Early Onset Support Service, Alzheimer Scotland	Home support, outreach support and befriending

215. In addition there are general projects which have part of their work specifically aimed at younger people with dementia: These include:

LOCATION	FACILITIES
Irvine Project, Alzheimer Scotland	Day care and home support
Kilmarnock Project, Alzheimer Scotland	Day care, home support and carers' support group
Kincardine and Deeside Project, Alzheimer Scotland	Young Person's Group
Dementia Initiative Scheme, (Falkirk, Stirling and Clackmannanshire) Joint Social Work and Forth Valley Board Project	Mutual support Group Counselling Home from Home Day Care Time to Share (family placement scheme)

CHAPTER 7

KEY ISSUES/GAPS IN SERVICES/ROUTES TO IMPROVEMENT

HUNTINGTON'S DISEASE

Key Issues

216. Mis-diagnosis or late diagnosis is still very common. The carer or partner of people with these conditions has major problems in obtaining accurate information on Huntington's Disease and coping strategies. Families need better access to the SHA at an early stage. This requires a greater awareness amongst medical staff of the appropriate routes of referral.

217. With no designated units for the care of HD in Scotland emergency care commonly takes place in unsuitable psychiatric units or geriatric units. This type of care is inappropriate for the patient, and distressing for the family and carer.

218. Specialist management is needed for dietary problems, depression and other common psychiatric problems that are frequently not recognised. Involvement of employers and counselling about employment is needed but can only be available in specialist units.

219. Symptomatic individuals rarely have access to expert neuro-psychological assessment, speech and language assessment or swallowing assessment.

220. Respite care needs are being poorly met at present. Inefficient and unsatisfactory crisis management is often the result.

221. End stage care is frequently provided within an institution. A specialist unit can best provide specialist advice on the management of this phase.

Gaps in Service

222. There is a lack of:

- specialised, co-ordinated care for affected individuals from diagnosis to continuing care.
- specialised support and access to information for the families.
- specialised units to provide day care, respite and terminal care for affected HD patients.
- specific information about HD and its burden upon health and social services, such that the problem is underestimated and under resourced.

Routes to Improvement

223. Service strategy and planning should ideally be based on a comprehensive assessment of need in each region of Scotland. This should include prevalence figures based on inpatient, outpatient and community data, as it is clear that present statistics from ISD, which are based on inpatient data, provide a very incomplete picture of HD within Scotland.

224. Service planning should involve commissioning strategies with Health and Social Services as well as the voluntary sector to ensure productive partnerships between these groups.

225. Clear Pathways of Care for diagnosis, continuing management and terminal care should be implemented. These pathways should enable access to specialist care and information as well as support and advice as required.

226. There should be provision of accurate and accessible literature at the primary care level.

227. The development of suitable training, supervision and support for family carers and care staff would greatly aid the provision of adequate care for the complex needs of this patient group.

ACQUIRED BRAIN INJURY

Key Issues

228. Notwithstanding the National Specialist Rehabilitation Units, there is a lack of access to locally based, specialist multi-disciplinary rehabilitation services. Such services could be led by a Consultant in Rehabilitation, Neuropsychology or Neuropsychiatry.

229. Even where services are well represented, there is a lack of strategic co-ordination. Joint Health and Social Work strategies should be designed to ensure individuals are properly tracked, can have needs identified at the appropriate phase of rehabilitation and can be linked to required services.

230. The currently accepted prevalence rates of people with significant problems arising from mild, moderate and severe injury underestimate the true picture.

231. There is a need to have comprehensive, easily understood and accessible information for both patients and carers. Equally, staff within specialist units as well as other staff who will interface with service users need good information and appropriate training.

232. There is a noticeable lack of services specific to children and young people. Particularly important at the so called ' development stage' such services would greatly aid the process of transition into adult care.

233. At each stage of the rehabilitation process, the family/partner requires accessible information and active involvement

Gaps in Service

234. Current services are, in the main, lacking in co-ordination and are unevenly distributed throughout the country. Noticeable gaps occur in the North and North West of Scotland. There is a lack of provision in rural areas generally.

235. There is a need to evaluate the efficacy of Transitional Living Facilities, as there is currently a complete lack of this type of service provision.

Routes to Improvement

236. The development of better Joint Health and Social Work strategic planning and funding mechanisms.

237. Systems to help bridge the boundaries between the differing phases of rehabilitation and to co-ordinate and case manage needs at a local level are required. The Throughcare Project Model in Glasgow may offer one solution. This model adopts the notion of multi-disciplinary assessments, early intervention and the building of strong links with community integration and vocational services.

238. In some parts of the country, alcohol is known to be a significant casual factor in TBI, sometimes in a majority of cases. Alcohol and other substance misuse are also often seen, after injury as a serious impediment to the rehabilitation process. Current services, for alcohol problems on the one hand and TBI on the other, do not usually address this linkage. Therefore, services tend to exist in parallel thus preventing effective reciprocal working. There is a need therefore for models of service provision that could overcome this problem.

239. Because of the historical and ad hoc way in which services have grown, often with a wide range of funding sources and with little strategic co-ordination, the result has been variable quality and effectiveness. There is therefore a need to explore how best to develop good quality standards across the range of rehabilitation phases.

240. The development of community based Transitional Living Facilities could be helpful in a number of ways especially by enabling people to move to a range of independent and/or supported living options.

241. There is a need to develop well-resourced prevention strategies. These could be monitored and reviewed via Health Boards' Health Improvement Plans and Trust Implementation Plans.

242. There is a need to strengthen the so called 'mixed economy of care 'by promoting and developing partnerships between statutory, voluntary and independent sectors

in keeping with recent policy statements such as "Modernising Community Care".

EARLY ONSET DEMENTIA

243. Current provision falls far short of the pathways of care that have been proposed for people with dementia. Perhaps the most important issues for people with EOD is the need for planners, commissioners and providers of services to recognise their distinctive requirements. Without this recognition and understanding it is unlikely that services will be either adequate or responsive to their needs.

244. More specifically people with early onset dementia require early diagnosis and continuity of care or 'joined up' services. Early diagnosis is needed because this is the entry point, the beginning of the care pathway. Well co-ordinated services are needed because of changing needs as the illness progresses.

Gaps in Services

245. The first essential is a diagnosis. This needs to be linked to immediate support and effective access to a wide range of information. Younger people with dementia and their carers need supported diagnosis which requires the provision of ongoing help for the person with dementia and their family to come to terms with the diagnosis and to access information, assessment, rehabilitative services, counselling, peer support and benefits advice.

246. There is a lack of continuity in service provision that can occur throughout the illness. The long term and progressive nature of the illness points to the need for the appointment of a key worker who will get to know the family and develop a relationship of trust so that individualised services can be provided.

247. There is a notable lack of specialist services for people with early onset dementia and a lack of understanding by generic services of their distinctive needs.

248. The final gap is the narrow range of alternatives to residential and nursing homes for the later stages of the illness. There is an urgent need for a broader range of housing and accommodation options, with support.

Routes to improvement

249. People with early onset dementia and their carers need to become involved in the planning of services in general and of their own services in particular. This is an essential requirement if services are to be sensitive to their needs. Service planners and commissioners should consult service users and potential service users.

250. The development of protocols that set out agreed understandings about the roles of GPs and hospital consultants will help improve communication between doctors. Access to diagnostic services that people will not see as stigmatising,

such as Memory Clinics, are one helpful development. The other will be to increase public awareness about the early symptoms of the illness and encourage discussion with their GPs.

251. Joint planning of services between health, social welfare and housing or even better joint commissioning of services and pooling of budgets is the ideal needed. This could ensure that there is continuity of care along the care pathways that people with early onset dementia are likely to travel from their diagnosis to their need for palliative care.

252. If joint work between health, social welfare and housing results in the creation of the care pathways, there will still be a need for a guide. This is the role of care co-ordination and care management. Sometimes this is linked to the idea of a key worker being allocated to each person with dementia.

Quality Issues

253. High quality services are established on the foundation of appropriately trained staff, including the awareness and understanding of what good quality dementia care is for those staff who come in contact with people with dementia as well as for those who work regularly with them. High quality services will also need well-designed physical environments.

General

254. Greater involvement of patients and carers in planning and developing services would be a powerful way of shaping user-friendly systems of care. Also patients and carers need help in articulating their needs and help with the choices and decisions needed to move them through the 'care-maze'. Advocacy is one way in which this can be achieved

255. Advocacy workers can enable patients and carers to consider their choices for care, firstly for immediate care, but also for long-term care, recording their preferences for care as they become increasingly affected by the illness so that their wishes can be considered when longer-term decisions are being made.

256. This service may be provided by the specialist voluntary agencies or by more generic advocacy workers. In the latter case close liaison with the specialist voluntary providers would be the ideal. This would provide useful two-way learning and service development opportunities.

CHAPTER 8

SUMMARY

Size of the problem

257. In Scotland for every 100,000 population there are approximately 10 to 12 individuals with Huntington's disease; 300 with acquired brain injury and 60 with Early Onset Dementia.

258. In addition to this is a large number of carers and family members who take on much of the burden of care usually over long periods of time.

Nature of the problem

259. These conditions result in complex health and social needs and their impact is out of proportion to the numbers of cases presenting. This is because the conditions result in a complex and challenging mixture of motor, sensory and cognitive disabilities. These result in loss of independence and income, difficulties in communication and often in breakdown in family and social relations at a time when these supports are most needed.

260. The disabilities are often subtle and require skilled professional help and well-managed support from the statutory and voluntary sectors. Moreover, needs change over time and services need to be flexible to anticipate and respond to this.

261. It is not surprising that there are areas where need is not being adequately met and current services appear poorly adapted to address this situation or that the early period when diagnosis is made commonly gives way to a failure to provide and maintain patient-friendly, co-ordinated care in the longer term.

262. Individuals and families affected by these conditions tend not to fit neatly into the prevailing medically based models of care; as a result there is generally no strong ownership of the problems or a desire to lead or co-ordinate services from those with influence over service development. Professionals probably underestimate what can be done to alleviate suffering and improve quality of life for these patients and there is a lack of insight into the scale of the problem and the need for specialist assessment and care.

263. In addition, the kind of long-term problems of recovery and adaptation seem to remain stubbornly invisible to the caring services; they lack the appeal of high profile conditions such as coronary heart disease and cancer. The costs of caring are, however, borne by patients, families and carers and there are important issues of equity involved.

264. Patients and carers have great difficulty in negotiating what is, for them, a complex maze of options for a number of reasons. They may not be sufficiently articulate; they may lack the vital information they need and they may not have a clear idea of their rights or of what represents an acceptable standard of care.

Professionals may communicate poorly with patients and carers and with one another and across the many agencies providing care.

265. For these reasons the role of the specialist voluntary agencies is especially important both in advising statutory agencies on service needs and service gaps and in direct provision of services; information; respite; advice and advocacy.

266. Advocacy is one way round this dilemma for patients and one which is being increasingly explored and developed as a way of empowering patients and carers and giving them a voice.

Key problems

267. Patients and carers identify a large number of areas in which they wish to see improved information and services. These include:

- information and emotional support
- specialist advice, counselling and advocacy
- memory training
- rehabilitation
- appropriate drug treatments
- carer training and education
- day care, home support and respite care
- supported accommodation and residential/nursing home care
- palliative care.

268. This is potentially an overwhelming list of needs but advice from those in the voluntary agencies representing these patient groups and evidence from published work suggests there are a number of key problems that can be addressed. These are:

Diagnosis

269. Early access to early expert multi-disciplinary assessment is regarded as a key priority for all these patient groups. This requires awareness amongst health and social care staff about the needs of people with these conditions and the existence and location of services.

Counselling and information

270. Patients and carers set a high priority on improving the availability and quality of this. Advice on services, the implications of the illness and the availability of support is needed early on and failure to get this results commonly in frustration and resentment.

Patient and carer information

271. Patients and carers often feel this is not getting to them when they need it. Also we usually do not know who the patients are out there in the community who need care. One approach to this, which has been applied in other fields, is the use of patient registers. These could allow information to be targeted to those who need it; would facilitate the process of consultation on service development and would be a considerable aid to planning services. There are, however, problems in placing individuals with genetic, progressive disabling conditions on registers and these might outweigh possible benefits. More work needs done on this and it should be investigated as a way of improving the targeting of information and services to patients and carers.

Co-ordination and continuity of care

272. This is needed both over time and between services. Poor inter-agency collaboration is common and there is a great deal of anecdotal evidence of wasted time and effort as patients attempt to find their way round the system. As mentioned above, advocacy provided by trained and experienced personnel is an important way of reducing this problem. It is currently provided by the specialist voluntary agencies. This and other ways of providing advocacy need to be supported and developed.

273. There are examples of good well co-ordinated services but these tend to occur where an individual has developed and championed a service - the challenge is to find ways of improving care for all patients even when such product champions are not present.

274. Advice for patients and carers is needed in a number of ways. They should know what services are available locally and further afield. They should have access to high quality written advice on diagnosis, treatment and rehabilitation. They should be given guidance on the standards of care they should expect and how to access these services. This is needed to create a more informed and able patient/carer group.

275. Professionals also need better access to information on diagnosis and care and on facilities available locally, regionally and nationally. This is particularly pressing because individual professionals may each encounter only a few cases so expertise is difficult to acquire.

276. Information technology and particularly the NHS Net needs to be exploited to improve the information needs of patients, carers and professionals.

277. There is wide agreement that a Care Pathway approach is needed to improve this situation. There would be considerable advantage in having work on this co-ordinated at a national level in some way. The Scottish Intercollegiate Guidelines Network (SIGN) has successfully used this approach to clinical guidelines. It has the potential to reduce duplication of work and result in high quality recommendations being made available for adoption to local and regional use.

278. Implementation of this will require care co-ordinators, liaison nurses or specialist consultants. Lack of accountability is currently a major problem as no one currently has clear responsibility for improving the care of this group.

279. Whoever takes the lead needs to manage the issue of inter-agency collaboration sensitively - particularly involving the voluntary and self-help agencies which are able to provide care in a way which is less stigmatising and more acceptable to users and carers (HAS Report 1996). Joint budgets covering health and social care should be a way forward and this is already being attempted in a number of areas.

Expert clinical care

280. This is available where there are specialist teams but many parts of the country do not have access to these. This could be provided in a number of ways. For example in the case of Huntington's disease by the increased use of local expert advisers. In the case of all three conditions the existing nucleus of expert teams in the major centres could act as a resource regionally providing advice and training. This would require some formalisation but probably only small amounts of additional resource.

281. A gap here is the absence of clinical guidelines on the management of the wider care of these cases. The Scottish Intercollegiate Guidelines Network (SIGN) should consider this. One difficulty is that these cases do not fit neatly into a medical model and this may be a challenge for SIGN to address.

Mental health services

282. These patients and their carers have a high risk of developing mental health problems and an important issue is access to appropriate services for treatable mental illness in patients or carers arising as part of the developing illness process. These treatable conditions may be missed in the complex of presenting symptoms and signs.

Rehabilitation

283. Multi-disciplinary team-based rehabilitation is the ideal for all patient groups and is essential if the needs of patients are to be met. However it requires strong leadership and support. It is particularly relevant to ABI as without it an individual patient's chances of returning to normal life are greatly reduced. However it is also essential in HD and EOD to preserve function and independence. It does not have to occur in a hospital setting.

Respite care and appropriate long-term placement.

284. The former appears to be difficult to obtain and the latter is often in inappropriate surroundings e.g. in facilities designed for older people. The extent of the problem is unknown and needs to be determined by special surveys of current facilities.

Lack of data

285. We are hampered by a lack of data on the true numbers of cases, on where and how appropriately they are located and on whether they are in contact with effective services. This information is not obtainable from routinely collected data and special studies are needed.

286. A certain amount of data is available from the voluntary agencies but it is not comprehensive. This inhibits effective planning and adds to the underestimation of the size and nature of the problem. Properly resourced surveys of current provision and/ or a series of visits to existing facilities are required to address this.

Prevention

287. Interventions known to be effective in preventing traumatic brain injury are available. (Cochrane1998) In view of the prolonged requirements for care resulting from head injury particularly in younger people (the highest risk group) this is amongst the most cost-effective primary prevention available.

288. This should focus policy makers on the need to implement effective preventive measures. This has been the subject of SNAP reports on Injury and Accident prevention. Similarly we know that effective management of risk factors reduces the incidence of vascular and haemorrhage brain injury.

CHAPTER 9

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APPENDICES

Appendix 1

HUNTINGTON'S DISEASE

Table 1: Age of diagnosis of Huntington's Disease within Grampian region

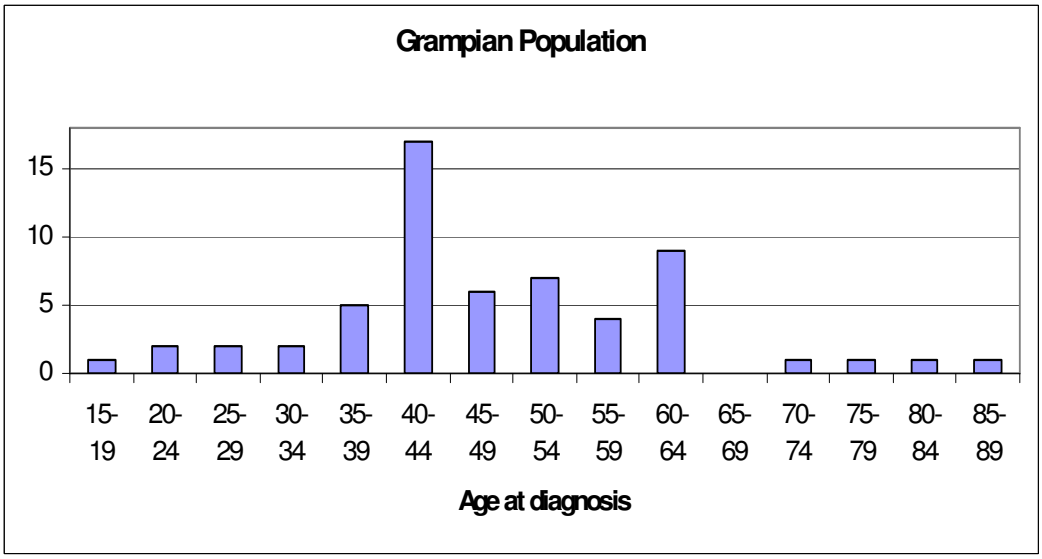
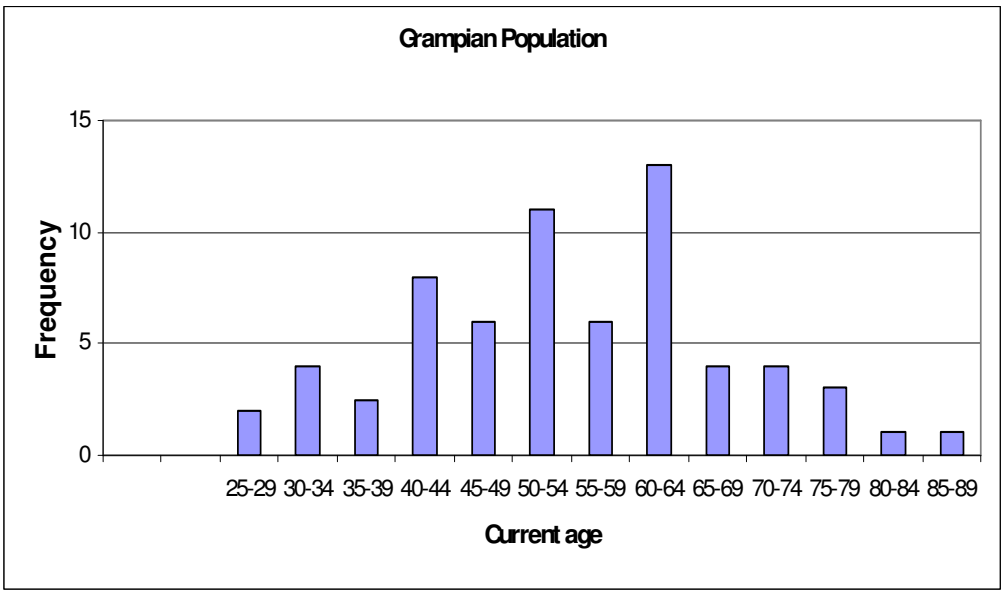


Table 2: Analysis of people diagnosed with Huntington's Disease within Grampian area



Appendix 2

CHECKLIST

A summary of key issues for the commissioners of services

Commissioning Services for People with HD

Some key considerations include:

- Know the nature, natural history, impacts and consequences (family, work and leisure) of disability in people who have HD
- Know where patients with HD are likely to be and how many of them there are within the district
- Integrate genetic counselling and neuropsychiatric services
- Anticipate long-term needs and plan service responses from the time when the diagnosis is made
- Integrate the roles and work of the statutory and non-statutory services

Source: HAS 1996

Commissioning Services for People with ABI

Some key considerations include:

- Know the nature, natural history, impacts and consequences (family, work and leisure) of disability in people who are brain injured
- Know where patients with ABI are likely to be and how many of them there are within the district
- Be aware that, because of their other injuries, patients with TBI may be dealt with in orthopaedic or general surgical wards where their brain injuries and their longer-term consequences may be missed or given insufficient attention
- Have a clear statement of what is wanted from each service provider.
- Integrate the roles and work of the statutory (health, social, housing and education services) and non-statutory services

Source: HAS 1996

Commissioning services for people with EOD

Some key considerations include:

- Know the nature, natural history, impacts and consequences of early onset dementia
- Know where patients with early onset dementia are likely to be and how many of them there are within the district
- Integrate the roles and work of the statutory and non-statutory services
- Ensure that effective multi-disciplinary assessment facilities are available and clearly identified
- Ensure that the assessment services communicate clearly with the services that provide continuing care
- Be clear about who is providing mental health services for this client group and for their carers
- Anticipate future care and nursing needs and plan service responses to meet them once the diagnosis is established

Source: HAS 1996

Appendix 3

Case Study/Huntington's Disease

This case study is an example from the SHA's Advisory files
(Personal details have been changed to protect identity)

Peter is a 34-year-old gentleman with Huntington's disease. Peter was married but is now divorced. He has three children, a son Paul who is now seventeen years old, a son Kevin who is twelve and a daughter Amy who is eleven. Peter has no contact with his oldest son (from a previous relationship) nor does his oldest son know of his father's diagnosis nor that he (Paul) is at 50% risk. Both of Peter's other children are aware that their dad has health problems but are unaware of the actual diagnosis or the implications this has for them. Peter wishes to tell his children but his ex-wife does not.

Peter is also part of a large family. He remembers his father as a hard and violent man who spent the latter years of his life in and out of psychiatric care. His father had Huntington's disease and died when he was 49years as a result of suicide.

Peter's mum is alive and remarried. Peter has three sisters all of whom carry the faulty gene for Huntington's Disease. His oldest sister Mary (38yrs) has Huntington's disease, is unemployed and lives with her second husband; she is at present trying for a family. She has one son with whom she has no contact. His sister Catherine (36yrs) has not been diagnosed as symptomatic but lives in constant fear of symptoms and suffers from long periods of depression. She is married and has one daughter (14yrs) whom she has not informed about Huntington's Disease. She is at present finding it difficult to sustain employment. His younger sister (31yrs) is married and has decided not to have children. She is very aware of the consequences of Huntington's disease, practices as a general practitioner and is at present asymptomatic.

Peter referred himself to the Huntington's advisory service approximately two years ago. He was living alone in B&B and had little contact with his family. On referral and following assessment several problems were identified. Peter was living in a very small room at the top of a flight of stairs. The private washing facilities, to which he had access, were in a terrible state of disrepair. The wash hand basin was cracked and the sink blocked. The kitchens had been closed down by environmental health a week prior to the advisor's visit. Peter had few clothes and was living on the minimum State Benefit. Peter physically had an obvious movement disorder with a marked unsteady gait. He had problems with swallowing and reported frequent episodes of aspiration. His speech was unclear at times. He had obvious signs of cognitive dysfunction reporting problems with his memory and concentration. Peter had accessed psychiatric services in the past but had no recollection of involvement with any other agency. Peter was abusing drugs and alcohol and was involved with the criminal justice team from social services. Peter had lost contact with many of his family and friends, links which he desperately wished to re-establish.

Immediate (same day) referrals were made by the advisory service to housing, psychology, speech & language therapy and social work. The housing department responded immediately, considered the 'high risk' environment, and re-housed the client

the following day in temporary supported accommodation, which had been especially adapted for someone with a disability. It was not possible to get Peter referred for expert clinical advice.

The clinical psychologist carried out a base-line cognitive assessment, and provided crucial information primarily to the client but then to the multidisciplinary team. Strategies for coping with Peter's changing behaviour were introduced and Peter was in turn much more able to cope with everyday living. Support and assessment was ongoing.

The speech and language therapist also completed baseline assessments and worked with Peter through a therapeutic programme. Peter did have difficulties with retention of information and learning new skills but overall the therapy was health promoting. The dietetic services were then approached via the GP and Peter was provided with written and continuing advice on consistency of diet and examples of menus and calorific need. Peter was initially underweight but is now able to sustain normal weight.

Work with the Criminal Justice Team and courts, particularly provision of accurate information about Huntington's Disease, resulted in appropriate support and understanding rather than a prison sentence.

Peter is now financially much more secure and receives relevant benefits for someone with a disability. He also has free access to public transport and is able to visit his family much more readily.

Peter re-established links with all of his family. They were given support as requested, primarily from the Huntington's advisory service and the clinical psychologist. Some family members are very supportive of Peter and others find supporting him much more difficult. Peter has become quite disinhibited and is often unaware of the social consequence of his actions. He has also experienced a degree of emotional blunting and egocentrism. This behaviour for many of his family and friends is difficult to understand and often misinterpreted as selfishness.

Peter is much happier within himself. He no longer takes drugs and his alcohol consumption has reduced. He feels the greatest difference over the past year has been an increase in confidence. He has not required any input from the criminal justice team for some considerable time. He continues to get support from a multidisciplinary team. His greatest achievement over the past year was to go on a walking holiday with a group of friends. His greatest disappointment was to be excluded from a sponsored parachute jump because he has a brain disease called Huntington's.

Case Study/Acquired Brain Injury

Michael is a 43-year-old male who sustained a traumatic brain injury as the result of a fall in a public house in December 1995.

Michael's family originates from Scotland, but lived in England until he was 7 years old. When Michael was 20 his mother died following a brain haemorrhage. His father, who was a heavy drinker, with a history of violence and aggression towards his wife, died in 1983. Michael has an older brother and younger sister, but has little contact with them.

Prior to his injury, Michael had a number of jobs, and was also unemployed for almost 8 years. After his father died he moved to England and lived for a number of years in a long-term relationship with a girlfriend. He had been a heavy drinker from the age of 18, and by 1985 was having real problems as a result of his alcohol misuse. Consequently he was admitted to a day unit for alcohol treatment in 1988.

Michael's long-term relationship ended in 1994 and he returned to Scotland to seek work and to re-establish family contact. Unfortunately he had little success, and his alcohol use increased again. In late 1995 he sustained his injury following a fall, suffering an acute right frontal subdural haematoma, intracerebral haematoma, and with some frontal contusion.

Following admission to Hospital via the Accident and Emergency Department, he underwent neurosurgery involving a right frontal lobectomy. In the immediate recovery period there was evidence of significant memory impairment, as well as impairment to the frontal executive function, including verbal fluency and problem solving. Over the next 12 months his cognitive functions steadily improved, but rehabilitation was impeded by a general lack of motivation. He was not physically impaired or disabled by his injury.

After several months in a rehabilitation ward, Michael became both verbally and physically abusive to staff and other patients. He was then transferred to a long stay ward where it was felt he would receive closer supervision. His behaviour continued to be a problem for staff, particularly at night and especially with females. He was sexually disinhibited and also began to bully other patients. At this time he was prescribed tranquillisers and was also started on anti-convulsive therapy.

In February 1997, 14 months after his injury, Michael was referred to the Scotcare Brain Injury Rehabilitation Unit for a period of assessment. He continued to exhibit aggressive and irrational behaviour, especially towards females. He was seen to have impaired memory and verbal and language skills, poor concentration, and a lack of insight regarding his challenging behaviour. A long-term placement in a supported environment was recommended, and after 6 months rehabilitation he was transferred to a long-term chronic sick unit.

Once in the unit Michael's behaviour deteriorated rapidly. He refused to accept authority from females and began to exhibit openly sexual and physical aggression to staff and other residents. As a result he was referred to the Robert Ferguson Unit in Edinburgh for specialist treatment and modification to his challenging behaviour. He was admitted in September 1998.

Michael responded well to the specialist and structured approach and, by the end of 1998, consideration was given to referring him to specialist community and vocational integration. This was thought appropriate as he was more co-operative and motivated, although his cognitive, language and problem solving impairments remained. Michael also continued to exhibit some inappropriate sexual and verbal behaviour, but this was much more easily controlled by both staff and by Michael himself.

Michael was described at this time as being usually pleasant, having a sense of humour and being motivated to live in the community with the hope of sustaining some form of supported employment. The discharge plan for Michael envisaged ordinary living, but in a supportive environment, perhaps with transitional living in the medium term. He had shown increasing motivation to undertake social, leisure, and vocational activities, and would benefit from community based services in these areas. He was referred to Rehab Scotland's Glasgow based Brain Injury Services, but this has been difficult to progress due to a lack of supported and/or transitional living in the Glasgow area where Michael would like to settle and live.

Issues highlighted:

- After severe injury, with significant cognitive and executive impairment, Michael has gone through phases of recovery, but has required specialist help to address his challenging behaviours.
- Michael will require high levels of structured support, at least for the foreseeable future.
- Michael has begun to show motivation and a desire to integrate back into community life and work, but requires some transitional help to achieve this goal.

Case Study/Early Onset Dementia

John lost his job after making a series of serious mistakes. At the time he insisted that there was nothing wrong and that all he needed was a less stressful job. As the months went on he didn't find another job and his family became increasingly worried about his moody and strange behaviour. Planting garden plants one day and digging them up the next. At first the GP said there was nothing to worry about and prescribed some pills. A few months later the problems were getting worse and John was sent to see a neurologist.

The consultant gave John a full medical examination and carried out a battery of tests. Following a scan John was diagnosed as having multi-infarct dementia and sent back to his GP. The GP told John's wife that there was nothing he could do but to come back if there were any changes.

Meanwhile John deteriorated quite rapidly and one day became lost while going to collect his daily newspaper. The police brought him home late at night. The family went back to the doctor who then suggested that they contact the social work department. While waiting for an assessment the family became worried. John's teenage daughter became extremely angry because she said she wouldn't bring her friends around any more because of her father's strange behaviour. A friend suggested they should contact the Dementia Helpline (0808 808 3000) The Helpline sent the family information about dementia and encouraged them to get in touch with a local dementia project. When they did this the project arranged for the social work assessment to be completed and offered John a place in the day centre one day a week in order to give his family a break. The day centre was for older people with dementia but there were no local alternatives and there were lots of young helpers. For some months this helped the family but John's wife increasingly felt that she couldn't leave him alone when she went out shopping. This time it was social work that came to the rescue when they arranged for a home carer to visit two afternoons a week.

Meanwhile the dementia project had been advising John's family how to apply for various welfare benefits they hadn't claimed. They also put her in touch with a solicitor to see if it would still be possible to arrange a power of attorney to enable John's wife to look after his financial affairs. Unfortunately it was too late and for the rest of his life his family had to negotiate a series of legal obstacles in order to manage the family finances.

John continued to deteriorate and it became increasingly difficult for the family to cope. The social work department was not able to offer additional home care and the day centre was full for the days when John didn't attend. It was agreed that John needed respite care but the only available place was in the psychogeriatric ward of a nearby hospital. John became very angry when he found himself in a hospital with "old" people.

Eventually even the occasional respite care breaks were not enough and it was decided to look for permanent residential or nursing home care. Most residential homes didn't want John because he had dementia and they felt that as a younger person his challenging behaviour would be disruptive. Some of the nursing homes were not keen to accept him but eventually one was found. Unfortunately the cost was more than the local authority

were prepared to pay and the family had to find an additional £50 per week. This was difficult because they were already dependent on income support, John's wife having given up her job to care for him. However John's brother was able to pay for this.
Some years later John died and his family were exhausted. Everyone had tried his or her best but it could have been better.

1. John's employers could have had an occupational health assessment before dismissing him, this could have resulted in an early diagnosis, and possibly him being able to remain at work for longer.
2. The GP could have referred John for diagnosis earlier and provided more active support.
3. When given the diagnosis the consultant should also have put John and his family in touch with the social work department, the local Dementia Project and given them the number of the Dementia Helpline.
4. Earlier diagnosis would have made it possible for John to receive practical support including counselling and to give his wife power of attorney and to sort out his affairs while he was still capable of doing so.
5. There should have been a specialist service for younger people with dementia instead of having to use existing services, which were already fully extended, and unable to offer the extra help when required.
6. Respite care should have been in more suitable surroundings.
7. There should have been a better choice of residential and nursing homes - all of which should be able to manage residents whose behaviour might be disruptive and those who have dementia.
8. There should be better ways of financing residential and nursing home care. If John's brother hadn't been able to pay the top-up he would have had to go into a nursing home much further from his family home.