

## 15. CERTAIN NEUROLOGICAL DISORDERS

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## 15.2 Multiple Sclerosis

### 15.2.1 Introduction

- (i) Multiple sclerosis (MS) is a chronic, progressive, degenerative disease, characterized in the early stages by remissions and relapses in its clinical course leading in some, though not all cases, to persisting and progressive disablement. Destruction of areas of myelin (a substance surrounding nerve fibres which enables nerve impulses to travel along the fibre) throughout the brain and spinal cord is followed by scar formation [known as a "plaque"]. Nerve fibres eventually degenerate, leading to failure of transmission of nerve impulses. The optic nerves (the nerves to the eyes) and spinal cord in the neck region are most frequently affected.
- (ii) Multiple sclerosis affects 30-80 people per 100,000 of population. The incidence of the disease rises throughout the second decade of life and is at its highest in the thirties. Slightly more women than men are affected. The disease rarely begins in people aged over 65.

### **15.2.2 Care Needs and Mobility Considerations**

- (i)** The principal manifestations of MS are weakness of one or more limbs, spasticity (muscle rigidity or pronounced stiffness), muscle fatigue, unsteadiness of gait, and difficulties with speech. Tremor sufficient to interfere with the use of the upper limbs may develop. Loss of sensation may occur. Difficulty in bladder control is common, the person having to rush to reach the toilet in time and often being incontinent.
- (ii)** As the disease progresses walking may become increasingly difficult either because of weakness and spasticity or because of unsteadiness leading to falls. This may be compounded by impaired vision and by weakness or tremor in the upper limbs which make the use of walking aids difficult or impossible. When walking, the severity of the muscle fatigue may result in the person's having to stop and rest frequently.
- (iii)** Short-term memory defects, impairment of speech, and mood swings are features of the more advanced stages of the condition.
- (iv)** The person may ultimately become chair or bed-bound. If this stage is reached there may be need for moving them frequently to prevent the occurrence of pressure sores.

### **15.2.3 Duration of Need**

The average survival of patients from the time of diagnosis is 30 years. The average age at onset is 32 years, and so there is probably a slight reduction in overall life expectancy. However, the course and effects of MS vary widely from one person to another and are unpredictable. Spontaneous remissions of varying length are common particularly in the early years of the disease, and in some instances appear to be life-long. Very rarely, multiple sclerosis takes a very rapid downhill course. However, the most common pattern is of frequently recurring relapses leading to chronic disability, dependency and complications. This process may take 20 years or more. Although the needs may change dramatically in the early years, if after two years there is a persistence or increase in needs, it is unlikely that there will be further improvements.

### **15.2.4 Further Evidence**

A medical report may be helpful where the needs over any period of time are not clear because of the variable nature and progress of the disease.

## **15.3 Parkinson's Disease and Parkinsonism**

### **15.3.1 Introduction**

- (i)** Parkinson's disease (Parkinsonism) is a complex clinical syndrome associated with decreased concentration of a naturally occurring chemical, called dopamine, in certain parts of the brain. It is characterised by tremor,

rigidity of muscles and difficulty in initiating movement.

- (ii) Parkinson's disease affects 100 people per 100,000 of the population in the United Kingdom. Men and women are affected equally. Symptoms usually begin after the age of 50 years, but it can also affect younger people. After the age of 50, the incidence rises rapidly with increasing age. There are probably between 60,000 & 80,000 people suffering from Parkinson's disease in the United Kingdom at any time.

### **15.3.2 General**

- (i) The earliest manifestation is a general, barely perceptible, impairment of movement. This leads to loss of facial expression, less frequent gestures, slowing of gait and a gradual increase in the time taken to perform any task including dressing and eating. Tremor may appear, initially in the hands, and muscular rigidity develops. As the disease progresses these symptoms become more marked. Mobility is particularly affected. The gait becomes short stepped and shuffling. Involuntary, progressive acceleration of steps (festinating gait) may occur making it difficult to stop and predisposing to falls.
- (ii) Alternatively the person may become rooted to the spot and unable to move. All tasks become more difficult especially those requiring fine movements of the fingers (tying shoe laces, doing up buttons, handling cutlery). Speech becomes slurred, weak and may be difficult to hear. At night the person may have difficulty turning in bed and so be unable to sleep because of pain and stiffness in the limbs. Mental function is not affected in the early stages, but in the late stages a proportion of sufferers become demented.
- (iii) The management of Parkinsonism has been revolutionized by the introduction of the drug L-Dopa, and other drugs, which increase the level of dopamine in the brain. Drug treatment reduces symptoms in 80% of cases; people who untreated would have required considerable attention are enabled to remain independent for some years. However, as the disease progresses treatment may lose its effect, symptoms increase and independence is lost.

### **15.3.3 Care Needs**

- (i) Attention needs arise principally from muscle rigidity, paucity and slowness of movement and tremor. The amount of attention and/or supervision required by day will depend upon the severity of symptoms and the stage the disease has reached in the individual case. In people first diagnosed as suffering from Parkinson's disease who have just embarked upon an appropriate drug regimen, the beneficial response is usually achieved quickly, resulting in a dramatic improvement in the disablement and its associated needs. If after a year significant improvement has not occurred

the situation is unlikely to change.

- (ii) Those already in long-term receipt of drugs who still have substantial needs because of persisting muscle rigidity, tremor or slowness of movement, are unlikely to show any further beneficial response. People in this category will probably progressively deteriorate and have increasing needs throughout the remainder of their lives.
- (iii) Independence is likely to be maintained by most sufferers in the early stages of the disease. As the disease progresses slowness of movement may make it impossible to undertake household tasks. Later assistance may be required with dressing and undressing and cutting up food. In some cases the person continues these tasks unaided but takes an inordinate length of time. Feeding, including chewing and swallowing, may also become very slow. In these circumstances assistance is a reasonable requirement. In the later stages of the disease the need for attention during the day will normally increase. The likelihood of falls occurring, and the danger they pose, is an important factor in determining whether there is a need for supervision.
- (iv) A requirement for attention at night is unlikely to be encountered until the later stages of the disease. Needs for attention at night arise in those who have difficulty in changing position in bed and in getting out of bed. The ability to use a portable urinal will also be impaired in those whose upper limb function is markedly affected by rigidity and tremor.

#### **15.3.4 Mobility Considerations**

The disturbances of gait and mobility described in paragraph 15.3.2 will be important considerations in the assessment of walking ability.

### **15.4 MOTOR NEURONE DISEASE**

#### **15.4.1 Introduction**

- (i) This is the name given to a group of related diseases affecting the motor nerve cells (which control movement) in the brain and spinal cord. In these conditions the motor nerve cells degenerate and the muscles which they supply become wasted and lose their ability to contract fully. The cause of the disease is unknown. The onset occurs most commonly between 40 and 60 years of age, although it can affect younger people.
- (ii) The way in which the condition first presents depends on which groups of motor nerve cells are affected. Weakness of the limbs is often the initial presentation with loss of fine movements of the hands and difficulty in walking. In addition to gradual paralysis of the limbs, difficulties with speaking, swallowing and breathing develop (bulbar palsy).

- (iii) The disease is progressive and its relentless nature means that disabilities may develop rapidly over a relatively short time scale. Overall the length of the illness can be between three to five years. Diagnosis may however be delayed and the average life expectancy after diagnosis is fourteen months. For many sufferers it is a rapidly progressive disease with a fatal outcome. Those cases presenting early with bulbar palsy may deteriorate quickly and have an especially poor prognosis as a result of respiratory difficulties.
- (iv) About 10 to 15 percent of people may have survival times lasting up to five years. These tend to be those who develop the condition at a younger age, where the limbs are predominantly affected initially and who have some of the rarer types of the group of motor neurone related diseases.

#### **15.4.2 Needs and Mobility Considerations**

- (i) The need for attention with personal care and bodily functions will depend on the muscles involved and the extent of the weakness. Involvement of the upper and lower limbs will lead to the need for assistance in feeding, dressing, washing, rising from a chair and with the toilet. Difficulty may be experienced in turning at night in bed. The debilitating effects of fatigue, difficulty in swallowing saliva, weight loss, anxiety and depression may exacerbate the other physical difficulties increasing the need for care as the disease progresses.
- (ii) Unsteadiness and a propensity to fall will affect mobility. As weakness of the lower limbs worsens the ability to walk is severely restricted and use of a wheelchair becomes necessary.
- (iii) Ten percent of sufferers may develop a mild form of dementia. It may be necessary to consider the need for some supervision, although it is likely that a need for attention with bodily functions will be more predominant.
- (iv) When difficulties with swallowing, speech and breathing develop (bulbar palsy) a small feeding tube inserted directly into the stomach through the abdominal wall (percutaneous endoscopic gastrostomy) will enable liquid feeds to be given to maintain nutrition. Help in communicating may be needed as the ability to speak deteriorates. In an increasing number of people assisted ventilation is used to aid breathing.

#### **15.4.3 Further Evidence**

- (i) A hospital factual report from a neurologist or specialist nurse attached to the hospital clinic will usually confirm the diagnosis including the type of disease and indicate the extent of disability at that time.

Occupational therapists may also have made assessments of disability with recommendations for aids and equipment. Physiotherapists and speech therapists advise sufferers on means of alleviating disability as the disease progresses.

- (ii)** In the latter stages of the disease people may be cared for predominantly by general practitioners, palliative care nurses and hospice doctors. These can be a valuable source of medical evidence that the disease is in an advanced stage and that life expectancy is reasonably expected to be less than six months. Referring factual medical reports to Medical Services both at this stage, and earlier in the process, can help in making an accurate assessment of mobility/care needs. This is of importance in determining the prognosis of a condition in which the severity of the disabling effects may evolve quickly.
- (iii)** An EMP report would not be the first choice of medical evidence in most cases. It is likely to be of greatest use in the small number of cases who have a longer life expectancy, and when disability is less severe earlier in the disease.

## **15.5 Peripheral Neuropathy**

- 15.5.1** The term peripheral neuropathy refers to damage to a nerve or nerves outside the brain and spinal cord. There are a wide variety of causes leading to variable degrees of loss of power and sensation in the area(s) of the body supplied by the damaged nerve(s).
- 15.5.2** Peripheral neuropathies can be divided into two broad categories depending on the distribution of involvement. The first group comprises damage to single peripheral nerves. Their effects alone are not likely to give rise to significant care or mobility needs. Secondly, there may be a diffuse and symmetrical disturbance of function due to the involvement of several nerves which can be called polyneuropathy (poly= many). In general, this results from causes that act diffusely, such as metabolic disturbances (ie. diabetes mellitus), toxins (poisons), deficiencies of certain vitamins etc. There are also hereditary polyneuropathies such as peroneal muscular atrophy. Because their effects are more widespread, polyneuropathies can cause significantly more disability and so the question of care and mobility needs is more likely to arise.
- 15.5.3** There are two polyneuropathies seen commonly. These are Guillain-Barre syndrome and the peripheral neuropathy that complicates diabetes mellitus. These are dealt with here.

## 15.6

### **Guillain-Barre Syndrome (Acute Idiopathic Inflammatory Polyneuritis)**

#### **15.6.1 Introduction**

- (i)** This is a rare condition causing inflammation of peripheral nerves leading to weakness of muscles and sensory loss. The disease can affect people of any age from early childhood onwards but the incidence is highest between the ages of 50 to 74. The condition develops over a few days or weeks and may be preceded, in about sixty percent of cases, by an infection such as an upper respiratory tract infection or gastroenteritis. It has also been associated with early HIV infection .
- (ii)** The distribution and severity of the weakness and sensory impairment is variable and gives rise to differing types of disability. Involvement of motor nerves is more predominant than sensory impairment, which tends to affect the sense of touch, appreciation of vibration and position sense in the limbs. The illness often begins with numbness and tingling in both feet, which is followed by muscle weakness in the lower limbs. The arms and face may become progressively affected, and in severe cases there is paralysis and difficulty in breathing. Such cases may need assisted ventilation in an intensive care setting.
- (iii)** Seventy to eighty percent of cases make a complete or almost complete recovery over several weeks or months with little residual disability. Children and younger people tend to recover more quickly. Those in whom recovery is not full often have residual weakness of the extremities, for example, of the hand or foot.

#### **15.6.2. Care Needs and Mobility Considerations**

- (i)** At the height of the illness when the person is most disabled, they are likely to be treated in hospital. Those with paralysis of the limbs and/or respiratory difficulties necessitating mechanical ventilation will have been admitted to intensive care for a period of time. Severe disease in this context can also affect function of the gastrointestinal tract, bladder and the blood pressure. Some people may require intravenous or tube feeding for a period of time. Physiotherapy plays a pivotal role in treatment as function returns to the limbs and other parts of the body affected by weakness or sensory loss. The use of splints will help to maintain function in the hands and feet as the condition improves. The sensory symptoms do not usually cause persistent impairment.
- (ii)** Depending on the time scale of recovery and the degree of residual disability there will be a wide variation in the amount of care required. Some individuals may need assistance with tasks such as washing, dressing, feeding as a result of decreased manual dexterity. Some people may need help to rise from a chair due to weakness of the trunk

muscles and lower limbs.

- (iii) The amount of difficulty in walking varies from person to person. Initial difficulty improves over several weeks or months after which there is usually little or no residual problem. Those who do not recover completely may be left with some difficulty in moving the foot or lower leg fully, a condition known as foot drop. The disease does not affect higher functions of the brain and it is unlikely that there will be a need for supervision.

### **15.6.3 Duration of Need**

Most cases recover quickly, within a few weeks or months, and there is little significant disability beyond six months after the acute illness. However in some cases recovery can take six to eighteen months or longer. People who were paralysed and needed assisted ventilation are likely to take longer to recover and may have residual disability. In a small number of cases, around 10% of the total, severe impairment persists and little or no improvement can be expected after two years. Some individuals (5 to 9 % of cases) may experience one or more recurrences.

### **15.6.4 Further Evidence**

Most people with Guillain Barre Syndrome will have been seen in hospital, and the more severe cases will have been admitted. It should be possible to obtain a report from a neurologist or specialist nurse attached to the neurological clinic. A specialist in rehabilitation medicine, a physiotherapist or an occupational therapist may also be able to provide information about the diagnosis, treatment and extent of recovery. General practitioners may be useful in providing reports about individuals who have not been seen at the hospital for some time; similarly an EMP report can give helpful information when the disability is stable or there is any doubt about the exact diagnosis.

## **15.7 Peripheral Neuropathy of Diabetes Mellitus**

### **15.7.1 Introduction**

- (i) About 15% of people with diabetes develop a significant degree of peripheral neuropathy. This is usually a symmetrical sensory polyneuropathy which can have important consequences since the loss of sensation leads to injury and the diabetes causes problems with healing. Commonly, the sensory neuropathy is mild, giving rise to numbness and tingling in the toes and feet and, less commonly, in the fingers. This does not normally give rise to significant problems. More rarely a severe sensory

neuropathy develops which is associated with the loss of pain sense. Damage to the joints of the toes and the ankles can occur, leading to degeneration of these joints. Minor injuries go unnoticed and because of poor healing small cuts can develop into large ulcers which become infected. In many cases the situation is complicated by peripheral vascular disease and often the only successful treatment is amputation of the foot or even the leg.

### **15.7.2 Care Needs and Mobility Considerations**

With the mild degree of sensory impairment usually seen there should be no significant increase in care and mobility needs on account of the neuropathy. With the more severe form there will be obvious problems with mobility and, depending on the age and capabilities of the individual, there may also be care needs. If there are significant needs these are likely to be permanent.

### **15.7.3 Further Evidence**

The degree of disability caused by the neuropathy should be fairly clear. If it is not, a factual report from the doctor taking care of the person (GP or hospital doctor) should provide the additional information needed.

## **15.8 Huntington's Disease**

### **15.8.1 Introduction**

- (i)** Huntington's disease is an inherited condition, occurring in 1 per 20,000 people (ie. about 3,000 in the United Kingdom). Children of an affected individual have a 50% chance of also developing the disease. Its onset is usually in middle life, between the ages of 30 and 50, although it can present as progressive dystonia (rigidity) in the teens. It is a progressive disease leading inevitably to severe disability and death. Death occurs on average 14 years after the onset.
  
- (ii)** The condition is due to a slowly progressive destruction of the cells of the brain, and is characterised by the gradual onset of uncontrollable movements (chorea) of either an abrupt jerking, or slow writhing nature; together with progressive loss of mental function. The initial symptoms are frequently of a change of personality and behaviour, but chorea may be the first sign. As the disease progresses dementia [See Chapter 21] becomes more obvious and the chorea more severe. The person has increasing difficulty with walking, with use of the hands and with speech. Gait is impaired by uncontrollable lurching and staggering. Attempts to pick up a pen or cup of tea set off wild, uncontrolled lunges. At this stage insight is lost, as is awareness of dangers, and behavioural problems increase. In the final stages many people develop increasing muscle rigidity and loss of

movement. This leaves the person totally helpless and bed-ridden.

### **15.8.2 Care Needs and Mobility Considerations**

- (i)** These will depend on the stage of the disease reached and the particular features shown by the affected individual. In the very early stages of Huntington's disease when the chorea is mild and insight is retained there may be few problems with day-to-day activities or with walking. As the chorea worsens, walking becomes more and more difficult as do many everyday activities such as dressing, eating, drinking and managing at the toilet. If dementia is a significant feature then there will be a serious risk of danger and a need for supervision.
- (ii)** Disability is relentlessly progressive, and in the final stages when the person is emaciated and bed-ridden a considerable amount of attention will be needed both by day and by night.

### **15.8.3 Further Evidence**

In most instances the care and mobility needs should be clear. Factual reports may help in those early cases where, either the diagnosis is not clear, or where there may be a significant suicide risk. In these early stages when insight is retained, with an awareness of what may be in store, depression is not uncommon and suicide is a risk. In the first instance a hospital factual report should be obtained, but if the person has not attended hospital, the GP may well be able to confirm the presence of the disease in previous generations of the family.

## **15.9 Myasthenia Gravis**

### **15.9.1 Introduction**

- (i)** Myasthenia gravis is characterised by rapid and extreme tiredness of the muscles. Thus, whilst muscle power may initially be strong, it rapidly fades with sustained effort. The disease usually develops in early adult life, but may start in childhood or even late in life. It is twice as common in women as in men.
- (ii)** The muscles of the face, throat and neck are almost invariably the first and most seriously affected. The condition commonly presents with drooping of the eyelids and double vision. Typically, symptoms first appear in the evening when the person is tired, and disappear after a night's sleep. There may be difficulty swallowing and chewing, complaints that worsen during the course of a meal. Speech may also be affected.
- (iii)** The muscles around the shoulders are also frequently involved and it is not uncommon for the muscles of the hips and thighs to be affected. The

muscles of respiration may also be affected leading to the need for assistance with breathing.

- (iv) Although the disease is generally progressive, there tend to be remissions and relapses. In some cases remission is complete and long lasting, and these people will not have significant care and mobility problems. In all but the later more severe stages of the disease, treatment is highly effective and may result in there being neither care needs nor problems with walking.

### **15.9.2 Care Needs and Mobility Considerations**

- (i) These will depend on the severity of the disease, the muscle groups affected and response to treatment. In mild cases, particularly those affecting the muscles of the face and throat alone there may be no need for any help of significance. It may be necessary to adapt the lifestyle somewhat to cope with the increased fatiguability at the end of the day but otherwise it should be possible to lead a normal life.
- (ii) Those whose shoulder muscles are affected may need help lifting heavy objects. They may later need help at the beginning and end of the day with dressing, washing and bathing. In severe cases they may be able to do very little with their arms and need a considerable amount of help both by day and at night.
- (iii) Those whose hip and thigh muscles are affected will have difficulty getting up from a chair or bed, and mobility will be reduced. In most instances the upper limbs will also be affected adding to the difficulty.
- (iv) If the muscles of respiration are affected the person may have difficulty breathing, in some instances to a degree sufficient to warrant mechanical assistance. In this situation the care and mobility needs will be considerable.
- (v) Insofar as myasthenia gravis does not lead to intellectual impairment, supervisory needs are not normally a feature of the disease. Only in its advanced stages is there likely to be a tendency to fall, by which time attention needs will predominate.

### **15.9.3 Duration of Need**

Most remissions occur in the first five years of the disease. If at any time disability is found to be severe and unresponsive to drug treatment, particularly if the respiratory muscles are involved, there is unlikely to be significant improvement in the future.

### **15.9.4 Further Evidence**

In most cases the level of disability and its duration will be clear. If it is not,

a report from the GP or hospital should provide the necessary additional information. Since response to specific treatment with certain drugs is usually good, factual information should be sought on how well the person is managed with appropriate treatments.

## **15.10 Meniere's Disease**

### **15.10.1 Introduction**

- (i)** This is a disease of the inner ear characterised by attacks of vertigo (dizziness), nausea and vomiting, associated with tinnitus (ringing in the ear) and increasing deafness. It can occur at any age but usually starts in the 4th or 5th decades.
- (ii)** Although attacks may be extremely incapacitating at the time, it is uncommon for them to last for more than a few hours at a time or to occur frequently for more than a few weeks at a time. The overall care and mobility needs will usually be slight.
- (iii)** The first symptoms usually noticed are deafness and tinnitus; but there may be vague unsteadiness, hearing difficulties, slight nausea or a feeling of pressure in the head. These normally affect one ear only but may progress to involve both. The attacks of vertigo may begin abruptly but are normally preceded by an intensification of the tinnitus. This or other warnings usually gives the sufferer time to sit or lie down so falls are unusual. There is usually no precipitating event for the attacks and they may occur during sleep.
- (iv)** The attacks usually last between 15 minutes and 2 hours during which time it is unusual for there to be any need for attention or supervision. The attacks can be frightening so reassurance may be necessary, but the person is unlikely to be in any danger. There may be long periods of freedom from attacks and in those cases that do progress the attacks may cease or eventually diminish with increasing deafness. Some persons may be left with some disturbance of balance.

### **15.10.2 Care Needs and Mobility Considerations**

Because the attacks usually occur in clusters with weeks or months in between, any care and mobility needs that may arise are intermittent and short-lived. At most there may be a need for help for a few weeks at a time. For the periods free from attacks there will be no attendance need. The person usually gets warning of an attack. The affected person would not usually have any mobility problems.

### **15.10.3 Further Evidence**

If there appear to be care and mobility needs in excess of those mentioned

above, a report should be obtained from the GP or any hospital the person is attending.

## **15.11 Migraine**

**15.11.1** This condition is characterised by periodic headaches, which are typically one-sided and are often associated with visual disturbances and vomiting. It is more common amongst women and tends to run in families. The condition usually starts after puberty and continues until late middle life when the attacks tend to lessen. Even for those who suffer severe attacks, their intermittent nature means that the overall needs for assistance are slight. There should be no need for assistance from another person during such an attack. Reassurance may be required but there should be no significant danger. The occurrence of migraine should not result in any mobility needs.

### **15.11.2 Further Evidence**

If there appear to be significant care or mobility needs a factual report should be obtained from the GP.

## **15.12 Poliomyelitis**

### **15.12.1 Introduction**

- (i)** Poliomyelitis is an infectious disease caused by the poliovirus. In the UK and most developed countries the acute infective form of the disease has been virtually eradicated by childhood immunisation programmes and better public health measures. The number of new cases is very small and likely to be linked to complications of vaccination.
- (ii)** In the past in the UK mainly children and young adults contracted the infection. Most cases (95%) were mild and complete recovery occurred within a few weeks. The poliovirus can however cause a severe, potentially fatal neurological illness in which there is profound weakness due to muscle paralysis. The virus attacks the motor nerve cells (motor neurones i.e. the nerves that control movement) in the brain and spinal cord. The muscles supplied by these nerve cells become paralysed. The paralysis may affect the muscles controlling breathing, and artificial ventilation in an intensive care unit is necessary to maintain life. One variant of the infection - bulbar poliomyelitis - may also affect the ability to speak and swallow as well as to breathe. In the paralytic form of the disease varying groups of muscle may be affected in an asymmetric pattern. Classically one lower limb, or part of the limb, might be severely affected while the opposite leg is unaffected. About half of those with paralysis recover completely, while the remainder have residual disabilities of a varying degree. Recovery from paralytic

poliomyelitis takes several months up to a year or more. There is no damage to sensory nerves in the infective illness.

- (iii) Individuals presenting with disabilities now are likely to be adults who contracted polio at a younger age. The last epidemic of polio in the UK occurred in the early 1950s and many of the survivors are now middle aged or older. Between 1985 and 1992 there were only 21 new cases of acute poliomyelitis in England and Wales.
- (iv) Since the disease is commonly contracted in childhood or adolescence before growth is complete, there is often asymmetric development of normal parts of the body compared to the paralysed parts. An affected lower limb may be considerably wasted and shorter than the normal leg causing difficulty in walking. There may be marked curvature of the spine due to abnormal growth patterns. The joints of affected limbs become stiff and unstable, and are liable to develop osteoarthritic changes. In some cases difficulties with breathing persist and there is a requirement for assisted breathing at night, and occasionally during the day. Some sufferers may continue to have problems with speaking and swallowing following the bulbar form of the infection.
- (v) It is well recognised that many polio survivors experience an increase in their degree of disability as the years progress. There is debate about the nature of the pathological processes that may lead to a decrease in function, and about the terminology used to describe them. The types of subsequent changes will be described in paragraphs (vi), (viii) and (ix). These distinctions however are of secondary importance in determining mobility/care needs in individual cases, where the resultant disabilities from previous polio require to be assessed on an individual basis in the light of the medical evidence.
- (vi) Disabilities arising some years after the original infection are described as being due to **late functional deterioration**. This term is used to describe the degenerative and secondary changes that may develop and increase impairment. Severe degenerative arthritis may occur in the joints of affected limbs and the spine, especially the cervical spine, leading to further restriction of movement. Pressure on individual nerves or on the spinal cord can lead to greater impairment of limb movement and the development of sensory symptoms. The degree of disability may be exacerbated by obesity and general physical deconditioning linked to poor mobility. There may be further loss of muscle bulk, coldness of limbs and a tendency to increasing feelings of fatigue and breathlessness. Difficulties in swallowing and speaking are also described. Individuals who need aid with respiration may have to use their ventilators for longer periods, or those who have not used ventilators for many years may have to start using them again for limited periods of the day or night.
- (vii) In addition other common medical conditions that develop in the middle years such as diabetes and heart disease may have additional disabling

effects.

- (viii)** The term **post polio syndrome** is used to describe the development of new neuromuscular symptoms, mainly increasing muscle weakness, in an individual whose neurological impairment has been stable over many years since the original attack of paralytic polio. It may occur fifteen to twenty years, or more, after the original illness; it does not appear to be directly related to the age of the person. There is an insidious onset of weakness and loss of muscle bulk accompanied by symptoms of pain and fatigability, usually in the muscles affected by the original infection. The condition may be due to further deterioration in the nerves (motor neurones) that supply the affected muscles.
- (ix)** The term **late effects of polio** can be used to encompass the features described above under **late functional deterioration** and **post polio syndrome**. Polio survivors experiencing deterioration in function from whatever cause can benefit from a specialist reassessment of their case. Disabilities can be ameliorated by provision of new aids, mechanical devices and adaptations. Treatment of osteoarthritis and other musculoskeletal problems including joint replacement may improve limb function. Physiotherapy and more extensive programmes of rehabilitation also help to maintain and improve function. It is thought that there are around 30,000 people in the UK who have survived the last epidemics of polio. Each case requires to be assessed individually for the overall degree of disability, taking into account the impairments of the original illness and the effects of subsequent deterioration whatever the cause. There may be much variation in the range of disability between individuals and the extent to which it has increased.

### **15.12.2 Care Needs and Mobility Considerations**

- (i)** Descriptions of care needs /mobility apply to all groups described above. Further information on paralysis and osteoarthritis can be found in other chapters.
- (ii)** As a result of impaired limb function, both upper and lower, individuals may need help with washing, dressing, rising from a chair, using the toilet, feeding and preparing food. In more severe degrees of paralysis help may be needed to turn in bed or maintain a suitable position.
- (iii)** Some people may be partially reliant on assisted ventilation during the day and at night. They may need assistance in using the equipment if they have impaired limb function.
- (iv)** The legs are more commonly affected in the original illness than the arms. As described above one leg predominantly may be paralysed and wasted leading to difficulties in walking, standing, bending and with balance. There will however be much variation from case to case. Some

people may walk well without aids; others manage with the aid of callipers or a walking stick. In cases of severe weakness a wheelchair will be used. With increasing disability the tendency to fall may become a problem.

### **15.12.3 Duration of Needs**

After the recovery phase of the original infective illness any residual muscle paralysis is permanent. Following rehabilitation the level of disability is likely to remain stable for many years. Subsequently care needs/mobility restrictions may increase as the disabilities evolve.

### **15.12.4 Further Evidence**

- (i)** In almost all cases evidence will be being sought about a person with long standing disabilities who had poliomyelitis many years ago. It may be difficult to obtain up to date medical information if the person is not attending a hospital clinic. General practitioners may have records of assessments following the original infection in addition to their personal recent knowledge of the patient. A report from an examining medical practitioner may be most helpful in assessing disability, especially where disabilities are multiple, and where factual reports are sparse in detail.
- (ii)** Some people may have maintained regular hospital follow up over the years, often under the care of orthopaedic surgeons. Hospital reports may also be available from neurologists, rehabilitation specialists and specialist nurses attached to hospital clinics. Physiotherapists and occupational therapists may also be a source of factual reports.

## **15.13 HYDROCEPHALUS**

### **15.13.1 Introduction**

- (i)** Hydrocephalus is an abnormal collection of fluid (cerebrospinal fluid) in the brain. The condition can lead to enlargement of the skull and compression of the brain tissues. The cerebrospinal fluid is produced within the brain; it circulates around the spinal cord and the brain ultimately being reabsorbed inside the skull. An excessive accumulation of the fluid occurs if the drainage pathways are blocked at any point; occasionally the condition can be due to an excess production of fluid.
- (ii)** The effect of an increasing accumulation of fluid is to interfere with the function of the brain. Initially this leads to tiredness, irritability, headache, drowsiness, associated seizures and ultimately to loss of consciousness. Untreated the condition causes progressive damage to and destruction of brain cells resulting in loss of motor function, diminished co-ordination, deterioration of learning skills, visual loss and other

neurological deficits.

- (iii) Hydrocephalus can be congenital or develop soon after birth, for example, due to a brain haemorrhage in a preterm baby. It may be associated with **spina bifida** or other developmental abnormalities of the spinal cord or the brain. Although it is often thought of as a condition of children many people survive to adulthood as a result of successful treatment. The condition can also arise in adults as well as in childhood, often as a complication of other diseases such as meningitis, cerebral haemorrhage or tumour. There is also a special form of arrested or compensated hydrocephalus that occurs in late adult life (normal pressure hydrocephalus).
- (iv) The main form of treatment is the surgical insertion of a shunt - a drainage tube that diverts the accumulated fluid around the obstruction. There are a number of different types of shunt which drain fluid from the cavities of the brain (the dilated ventricles), either to the heart or to the peritoneal cavity of the abdomen (the latter is the preferred option). Problems can arise with the shunt as a result of blockage, detachment or infection leading to a build up of pressure with its detrimental effects on the brain. In children further operations may be necessary to revise the shunt as the child grows. Problems with a shunt may present with headache, seizures, dizziness and drowsiness, and sometimes with more subtle changes such as behavioural problems or poor school performance.
- (v) People with hydrocephalus may also have seizures (epilepsy) and be prescribed anticonvulsant medication. Visual problems can occur including squint, involuntary eye movements (nystagmus), restricted visual fields and considerable visual loss. Urinary incontinence may also be found in association with coexisting neurological problems.

### **15.13.2 Care needs and Mobility Considerations**

- (i) If treatment of the hydrocephalus is successful, and there is little or no neurological damage or intellectual deficit, care needs may not be greater than that of a normal child. Significant care needs may be attributable to the coexisting neurological conditions such as spina bifida or associated learning disability (see related sections of handbook). In those children in whom the condition is not successfully or entirely relieved by insertion of a shunt some deterioration in neurological and intellectual function may occur. Their level of disability will need to be reviewed.
- (ii) Some children having recurrent problems with their shunts may be subject to episodes of drowsiness, which can lead to loss of consciousness. It is unlikely that this problem would be so precipitous that there was no warning of the decreasing level of consciousness, or that another person would have to be in attendance throughout the day

and night to prevent harm. Associated learning disabilities and/or visual impairment may lead to a requirement for supervision at home and when walking out of doors.

- (iii) Significant difficulty in walking is likely to be due to coexisting paralysis due to spina bifida rather than the hydrocephalus alone. Some adults with hydrocephalus do have problems with balance.

### **15.13.3 Duration of need**

If the condition is stabilised with treatment disability can be mild and will change little in the future. For those in whom raised pressure in the brain was not treated quickly enough long term damage can occur despite surgical treatment. Established neurological deficit and intellectual disability does not improve in these cases. The aim of any further surgical treatment is to prevent deterioration in the future; it will not lead to any lessening of care or supervisory needs. People with hydrocephalus are likely to be under long term hospital review. Each case needs to be assessed on its merits for the disabilities arising from the hydrocephalus and from the coexisting neurological conditions.

### **15.13.4 Further evidence**

Children and adults with hydrocephalus will be under the care of a hospital specialist, usually a neurologist and/or a neurosurgeon. A paediatrician with a special interest in neurology or developmental disorders may be the main specialist for some children. A hospital report is likely to be the most useful source of information, and sometimes could be provided by a specialist nurse attached to the clinic. An occupational therapist or physiotherapist may also be able to provide a report. The general practitioner may be able to provide a report for adults whose condition is stable, or who only attend hospital infrequently. An EMP report may be the best option in the latter group if no recent factual information is available or disability appears mild.

## **15.14 DYSTONIA**

### **15.14.1 Introduction**

- (i) The term dystonia is used to describe a group of illnesses in which abnormal and prolonged muscle contraction gives rise to uncontrollable, repetitive and abnormal movements or postures. The cause of the condition is not fully understood but it is thought to be due to an abnormality in part of the brain called the basal ganglia, an area of the brain that plays a role in controlling movement.
- (ii) Dystonia can be **primary** (or idiopathic), when no obvious cause can be

identified, or **secondary** when it occurs as part of other diseases such as cerebral palsy or stroke. The condition can arise as a side effect of some medications (neuroleptic drugs) used to treat mental health disorders such as schizophrenia.

- (iii) In practice it is useful to classify the condition by the part of the body which is affected. If only one area such as the eyes or neck is affected, it is referred to as **focal dystonia**. Involvement of a limb would be described as **segmental dystonia**, while widespread dystonia is called **generalised dystonia**.
- (iv) The most common types of dystonia are focal affecting the neck **spasmodic torticollis** causing involuntary twisting of the neck, **blepharospasm** uncontrollable blinking of the eyes and **writer's cramp** due to spasms of muscle contraction in the hand or forearm. In addition to the abnormal movements these conditions may cause pain. The focal forms tend to occur in adults and do not generally spread elsewhere in the body.
- (v) The more widespread forms of dystonia are rare and often start in childhood. Initially a lower limb is affected but the condition then spreads to include the trunk, neck and upper limbs.
- (vi) Dystonia can be treated with medication, which tends to alleviate the condition rather than gives complete relief. The focal types can be treated by injections of botulinum toxin, which causes a temporary weakness of the affected muscles. In some cases this treatment produces good relief but it needs to be repeated every few months in a hospital clinic. In some people the condition may improve spontaneously to a degree.

#### **15.14.2 Care needs and Mobility Considerations**

- (i) In general it is unlikely that the focal types of dystonia, such as spasmodic torticollis, would cause sufficient impairment of limb function to give rise to significant care needs. Writer's cramp usually affects a single limb only and might cause some mild impairment of grip. Involvement of the lower limbs in adults is rare and restrictions in the ability to walk are unlikely.
- (ii) In the severe cases of generalised dystonia involving all four limbs and the trunk care needs may be extensive and the child would use a wheelchair. Some adults may have dystonia secondary to a condition such as a stroke. Care needs and mobility would require the assessment of the impairments arising from the underlying illness as well as the additive disabling effects of the dystonia.
- (iii) Some people with severe and persistent blepharospasm may complain that they are effectively unable to see, although the eyes themselves are

normal. The condition can vary, and people may report that at some times reading is difficult while at others it is easy. In most cases episodes of blepharospasm are likely to be brief, although recurrent. Some sufferers report that the condition is aggravated outdoors by the light or windy conditions. In addition blepharospasm sometimes disappears spontaneously. Any claim for increased supervision as a result of visual impairment would require careful consideration, probably with a specialist report.

- (iv) People with dystonia may develop symptoms of anxiety and depression due to the embarrassing appearance of the abnormal movements. Anxiety itself may worsen the dystonia. These psychological effects can lead to social withdrawal and may exacerbate the perceived level of care needs.

#### **15.14.3 Duration of needs**

When dystonia has been diagnosed recently, it is reasonable to assume that there may be a response to treatment. If the condition is established or generalised the impairment is likely to be long term.

#### **15.14.4 Further evidence**

Diagnosis and treatment of dystonia usually occurs in a hospital clinic and a report may be obtained from a neurologist or a specialist nurse attached to a neurology clinic. For those people not attending a hospital clinic the general practitioner should be able to provide a report. People with more extensive disability may have been assessed by an occupational therapist. An EMP report will be valuable if the severity of the condition is unclear from the available evidence, or there is no recent hospital involvement.