CHAPTER 1
Introduction

1.1 Economic impact of long-term neurological conditions

It is estimated that 10 million people in the United Kingdom live with some form of neurological condition that impacts on their everyday lives (Department of Health [DoH], 2005). Neurological conditions account for one in five emergency hospital admissions, one in eight general practice consultations and a high proportion of severe and progressive disability in the population (Association of British Neurologists, 2003). As many as 350,000 people in the United Kingdom need help with activities of daily living because of a neurological condition and 850,000 people care for someone with a neurological condition (DoH, 2005). Due to their devastating impact and their generally progressive nature, neurological conditions are considered as long-term affecting individuals throughout their life span.

Occupational therapy is defined as ‘a client-centred health profession concerned with promoting health and well-being through occupation enabling people to participate in everyday life’ (World Federation of Occupational Therapists, 2011). Occupational therapy practice focuses on enabling individuals to modify and adapt elements of their roles, occupations or environments to support occupational participation in response to changes within their lives. Occupational therapists have a key role to play in supporting people living with a long-term neurological condition to manage a life of unpredictability and uncertainty. This requires a complex combination of knowledge and skills to address the physical, psychological, cognitive and emotional needs of people together with a broad range of assessments and interventions.
1.2 Definition of long-term neurological conditions

The DoH (2005) describes ‘long-term neurological conditions’ as

a range of conditions affecting the brain or spinal cord which occur through a variety of mechanisms which include the following:

- Sudden onset conditions (e.g. acquired brain injury of any cause, stroke and spinal cord injury)
- Intermittent conditions (e.g. epilepsy)
- Progressive conditions (e.g. multiple sclerosis (MS), motor neurone disease (MND), Parkinson’s and other degenerative disorders)
- Stable conditions with/without age-related degeneration (e.g. polio or cerebral palsy).

This book specifically focuses on the following progressive neurological conditions:

- Huntington’s disease (HD)
- Motor neurone disease (MND)
- Multiple sclerosis (MS)
- Parkinson’s.

Whilst there is an abundance of literature relating to each of these medical conditions the primary aim of this book is to place this knowledge and understanding within the context of occupational therapy practice. In order to fully understand the holistic needs of their clients occupational therapists are required to develop knowledge of the underlying pathology of each of these neurological conditions. However this understanding from a medical perspective should not be assumed to represent a medical model of care with an emphasis on symptomatic management. Throughout this book the focus is on delivering person-centred models of practice which support the complexity of the needs of people with neurological conditions from an occupational perspective.

1.3 International Classification of Functioning, Disability and Health

The International Classification of Functioning, Disability and Health (ICF) offers a conceptual basis for the definition and measurement of health and disability (World Health Organisation [WHO], 2002). Developed within a biopsychosocial model, ICF views disability and functioning as outcomes of interactions between health conditions (diseases, disorders and injuries) and contextual factors, as shown in Figure 1.1. Amongst contextual factors are external environmental factors (e.g., social attitudes, architectural characteristics, legal and social structures) and internal factors which include gender, age, coping styles, social background,
past and current experience, character and other factors that influence how disability is experienced by the individual (WHO, 2002).

Within this framework ICF defines three levels of human functioning: functioning at the level of body or body part (impairment), the whole person (activity limitations) and the whole person in a social context (participation restrictions). The formal definitions of these components of ICF are provided in Box 1.1 (WHO, 2002).

The remainder of this chapter presents each of the four neurological conditions in relation to body functions, body structures and impairments, highlighting the differences and similarities of each condition. Subsequent chapters explore the wider implications for activity and participation.
1.4 Huntington’s disease

HD is a rare disease, affecting an estimated 7–10 people per 100,000 or somewhere in the region of between 4200 and 6000 people in the United Kingdom (Quarrell, 2008). The onset of the disease is insidious and the age of onset depends on a number of different factors. Most people develop the condition between the ages of 30 and 50 years, but the disease can appear in all age groups (Nance et al., 2013). The HD gene is dominant, which means that each child of a parent with HD has a 50% chance of inheriting the disease and is said to be ‘at-risk’. Males and females have the same risk of inheriting the disease. HD occurs in all races (Nance et al., 2013).

There is currently no cure or treatment which can halt, slow or reverse the progression of the disease (Nance et al., 2013) and people with HD tend to die, on average, between 15 and 16 years after the onset of symptoms (Quarrell, 2008). People don’t die from HD itself, but they die from complications such as choking, heart failure, and infection or aspiration pneumonia (Nance et al., 2013).

1.4.1 Body functions
HD is a hereditary neurodegenerative genetic disorder caused by an expansion of a repeating CAG triplet series in the huntingtin gene on chromosome 4, which results in a protein with an abnormally long polyglutamine sequence (Nance et al., 2013).

1.4.2 Body structures
HD causes cells in the brain to die, specifically the caudate and the putamen and, as the disease progresses, the cerebral cortex. These organic changes lead to cognitive, motor and psychiatric changes that have a devastating impact on the individual. As the brain cells die, a person with HD becomes less able to control their movements, recall events, make decisions and control their emotions (Nance et al., 2013).

1.4.3 Stages of HD
Early stage
Symptoms may include minor involuntary movements, subtle loss of coordination, difficulty thinking through complex problems, depression, irritability, or disinhibition (Nance et al., 2013). Early symptoms of the disease often include subtle cognitive changes including the following:

• Difficulty organising routine matters or coping effectively with new situations
• Difficulty recalling information may make them appear forgetful
• Work activities may become more time-consuming
• Decision-making and attention to details may be impaired
• Irritability
Slight physical changes may also develop at this stage. There can be involuntary movements which may initially consist of ‘nervous’ activity, fidgeting, a twitching of the hands or feet, or excessive restlessness. Individuals may also notice a little awkwardness, changes in handwriting or difficulty with daily tasks such as driving (Nance et al., 2013).

Middle stage
Chorea may be prominent, and people with HD have increasing difficulty with voluntary motor tasks. There may be issues with swallowing, balance, falls and weight loss. Problem solving becomes more difficult due to difficulties sequencing, organising or prioritising information (Nance et al., 2013).

The initial physical symptoms will gradually develop into more obvious involuntary movements such as jerking and twitching of the head, neck and arms and legs. These movements may interfere with walking, speaking and swallowing. People at this stage of HD often stagger when they walk and their speech may become slurred. They may have increasing difficulty working or managing a household, but they can still deal with most activities of daily living (Nance et al., 2013).

Late stage
Chorea may be severe, or be replaced by rigidity, dystonia and bradykinesia. Although they are unable to speak in the end stages, it is important to note that people with HD retain a level of comprehension (Nance et al., 2013). People in these stages of HD can no longer manage the activities of daily living and usually require professional nursing care. Difficulties with swallowing and weight loss are common (Nance et al., 2013).

1.4.4 Impairments

- **Chorea**
  More than 90% of people with HD have chorea. It is characterised by ‘involuntary movements which are often sudden, irregular and purposeless or semi-purposeful. The movements are often more prominent in the extremities early in the disease, but progress to include facial grimacing, eyelid elevation, neck, shoulder, trunk, and leg movements as the disease progresses’ (Nance et al., 2013).

- **Dystonia**
  Characterised by ‘a repetitive, abnormal pattern of muscle contraction which is frequently associated with a twisting quality’ (Nance et al., 2013).

- **Bradykinesia**
  ‘Slowness of movement can include loss of facial expressivity, absence of arm swing, rapid alternating movements and gait slowness’ (Nance et al., 2013).

- **Tics**
  ‘are sudden brief, intermittent movements, gestures or vocalisations which can occur with HD. Respiratory and vocal tics can produce sniffs, grunts, moans or coughs’ (Nance et al., 2013).
• Loss of motor control
  ◦ Progressive loss of voluntary motor control
  ◦ Clumsy, awkward movement
  ◦ Akinetic
  ◦ Rigidity
  ◦ Hyper reflexia
  ◦ Extensor plantar reflexes (Nance et al., 2013)
• Gait impairment and falls
  ◦ Slower wide-based gait
  ◦ Trunk dystonia
  ◦ Chorea
  ◦ Displaced centre of gravity (Nance et al., 2013)
• Communication and swallowing
  ◦ Dysarthria
  ◦ Changes in speech rhythm
  ◦ Voice changes, that is soft spoken or explosive
  ◦ Complete loss of speech often occurs
  ◦ Difficulties with speech initiation
  ◦ Word-finding difficulties
  ◦ Impaired breathing (Nance et al., 2013)
  ◦ Dysphagia
  ◦ Aspiration
• Bowel and bladder dysfunction
• Weight Loss
• Cognitive impairment
  ◦ Attentional deficits
  ◦ Speed of processing
  ◦ Memory
  ◦ Visuospatial abilities
  ◦ Executive function
  ◦ Planning
  ◦ Lack of insight
  ◦ Behavioural regulation
  ◦ Lack of initiation
  ◦ Perseveration
  ◦ Impulse control (Huntington’s Disease Association, 2012)
• Emotional and behavioural changes
  ◦ Depression
  ◦ Apathy
  ◦ Irritability
  ◦ Disinhibition
  ◦ Jocularity
Obsessive compulsive disorder

- Impaired judgement
- Mania
- Agitation
- Delirium
- Sexual disorders including loss of libido or making inappropriate sexual demands (Huntington’s Disease Association, 2012)

1.4.5 Diagnosing HD

Genetic testing in HD can serve two purposes: as a diagnostic tool and as a predictive test to identify level of risk. Genetic testing involves the examination of an individual’s DNA, which is obtained from a blood sample. DNA molecules consist of four bases, known as A (adenine), T (thymine), G (guanine) and C (cytosine). The gene that causes HD is called the HD gene, and within it there is a region in which a sequence of the three bases (CAG) is repeated many times. For individuals with HD, the CAG sequence has increased (expanded) into a range that is abnormal. Testing is done in a specialised laboratory to determine the number of CAG repeats in both copies of the HD gene (Huntington’s Disease Association, 2012).

An HD gene expansion is passed on in families and children of a parent with this expansion have a 50% chance of developing the disease. Predictive testing is a process whereby an individual at risk of the disease can discover whether or not they have inherited the expanded HD gene, and will go on to develop HD. A ‘gene negative’ result is where the number of CAG repeats is 26 or less. The individual will not go on to develop the HD and their children will not be at increased risk either (Huntington’s Disease Association, 2012).

An intermediate result is a result where the number of CAG repeats is between 27 and 35. This means that the individual will not go on to develop HD but, in some cases, may pass on an expansion to their children because the CAG repeat can be unstable when passed from one generation to the next. This can mean that sometimes children will be at higher risk for developing HD (Huntington’s Disease Association, 2012).

A reduced penetrance result is one where the number of CAG repeats is between 36 and 39. An individual with a result in this range may not develop any symptoms of HD; however, this result also means that the next generation may be at risk of inheriting a larger expansion as it would also be unstable (Huntington’s Disease Association, 2012).

A full penetrance or ‘gene positive’ is a result where the number of CAG repeats is 40 or more. The individual with this result will always go on to develop HD at some point in the future. The result does not give information on the age of onset of symptoms (Huntington’s Disease Association, 2012).
Case study

Luke is 41 years old. Last year Luke underwent genetic testing following the death of his father to Huntington’s disease 8 years ago. Although Luke was aware of the genetic risk of HD he previously did not feel able to cope with genetic testing and opted to continue life without knowing the potential risk. However as he began to realise that some potential signs might be emerging within his everyday life, he felt it was now necessary for him to have a more definite prediction of what might lie ahead. It was confirmed that Luke had a full penetrance result confirming that he would go on to develop HD. Luke was devastated by the outcome as he now had two young children of his own. While Luke had been aware of his father’s condition he previously refused to attend for genetic counselling. Luke and his wife Amy aspired to have a normal family life and did not wish to acknowledge the potential risks when planning their family.

Luke works as a self-employed plumbing and heating engineer. Recently he had noticed some slight clumsiness or lack of co-ordination when working with his tools. This did not really impact on his ability to complete jobs, but he found he was becoming slightly weaker when unscrewing tight fastenings, greater difficulty when working within confined spaces and some difficulties with tasks requiring the simultaneous use of both hands. He was aware that it was taking him longer to complete jobs which placed him under significant pressure as in his trade time was money and this could have significant financial implications if he was unable to accept the same number of jobs. He also had many regular customers and had spent several years building a reputation as a reliable and dependable tradesman. Luke did not wish to let his customers down.

In addition to the practical aspects of his job Luke was also aware that he was finding it harder to plan his work schedule. He felt he was wasting valuable time as he was not planning his jobs in the same way to minimise travel time, and on occasions was significantly under-estimating how long it would take to complete a job. Previously Luke would have done this automatically, but now he was finding that he had to give this much more thought and was becoming angry and frustrated with himself because of this. There had also been some occasions when Luke had turned up for a job but had not brought the right tools or equipment. Again this had time and financial implications for him.

Luke also had responsibility for the administration aspects of his business including tax returns, book-keeping, preparation of customer invoices, and he was very concerned that he was making some mistakes with this. Luke had previously been a bit of a perfectionist, but now he was regularly making small mistakes and errors. Initially he attributed this to tiredness as he worked long hours, but he didn’t cope well with this change and got very frustrated with himself for making mistakes. He was starting to doubt his own ability and found himself taking more time to check his work to see that it was accurate.

Amy was aware of changes in Luke as he was forgetting things that she had told him during conversations. There were numerous occasions when Luke would say ‘You never told me that…’ when Amy was confident that a discussion had taken place. Luke had previously been very gentle and mild mannered and loved spending time with his family. More recently Luke had become irritable with Amy and the children. He seemed less tolerant of the children’s behaviour and seemed to be more reactive and angry with them all.

On occasions Luke had made rather unusual comments to people such as when waiting in the queue in the supermarket, or when trying to find a parking space. Amy was quite embarrassed by this and was concerned that Luke did not seem to understand why this might be inappropriate. Luke was very aware of twitching in his hands and feet and was
MND is a term used to describe a group of related diseases that attack the motor neurones (MND Association, 2012). It is a life-limiting condition that progressively impacts on the ability to perform daily functions. The average age at symptom onset is between 50 and 70 years, although it may occur earlier or later in life. Typically symptoms such as stumbling, foot drop, weakened grip, slurred speech, cramp, muscle wasting and fatigue occur in the early stages becoming progressively worse over a 2–5-year period. The current incidence is 2:100 000 per annum, rising with age and prevalence rates of 7:100 000, with a male:female ratio of 3:2 (MND Association, 2012).

**Amyotrophic lateral sclerosis**

Around 85% of people with MND will be diagnosed with amyotrophic lateral sclerosis (ALS). Early symptoms can include muscle weakness with spasticity in the upper or lower limbs which become progressively worse over a 2–5-year period. Where the initial onset is in the bulbar territory, survival tends to be shorter (2–3 years) (MND Association, 2012).

**Progressive bulbar palsy**

In this form of MND early bulbar signs can be relatively confined for several months before limb involvement becomes apparent. The overall survival rate for this form is 6 months to 4 years with a gender ratio of 1:3 male:female (MND Association, 2012).
Progressive muscular atrophy
Affecting less than 10% of people with MND progressive muscular atrophy (PMA) is characterised by a slowly progressive, proximal upper limb weakness which is usually symmetrical, and accompanied by visible fasciculations (MND Association, 2012).

Primary lateral sclerosis
This is the rarest form affecting approximately 5% of people with MND. It is characterised by spasticity and increased reflex response, as only upper motor neurone damage occurs. Often balance is affected and survival is notably longer (10–20 years) (MND Association, 2012).

1.5.1 Body functions
The motor neurone diseases are a group of progressive neurological disorders that destroy motor neurones, the cells that control essential voluntary muscle activity such as speaking, walking, breathing and swallowing.

1.5.2 Body structures
Motor neurones are nerve cells which carry an electrical transmission from the central nervous system to the muscles triggering a muscle to either contract or relax. The damage which occurs in MND leads to an interruption in the electrical transmission, resulting in a breakdown in the communications between the brain and the muscles. Impairment of the upper motor neurones leads to weakness and stiffness in the muscles, while lower motor neurone damage results in weak, floppy muscles and fasciculations (MND Association, 2013; Figure 1.2).

![Figure 1.2](image_url) Comparison of healthy and motor neurone affected by MND. (Source: MND Association, 2013, p. 7. Reproduced with permission of MND Association.)
1.5.3 Genetic Risk

**Sporadic MND**
While there is no known reason currently why some people may be at greater risk than others of developing MND genetic susceptibility and environmental factors may contribute to increased chance. For 90–95% of people diagnosed with MND, there is no family history of the disease (MND Association, 2012).

**Familial MND**
Of the remaining 5–10%, however, the disease is caused by genetic mutation. Currently there is no genetic test available that can confirm whether or not the disease is familial. Familial MND nearly always displays autosomal dominant inheritance, although penetrance is sometimes reduced, and the disease can appear to skip a generation. Age and site of onset can vary between cases within the same family (MND Association, 2012).

1.5.4 Impairments
- Muscle weakness
- Spasticity or stiffness in the limb muscles
- Slow and effortful movements
- Knee and ankle jerks
- Sudden muscle cramps
- Loss of voluntary movement
- Twitching and fasciculation
- Pain
- Dysarthria
- Dysphagia
- Respiratory insufficiency
- Acute dyspnoea
- Saliva and mucus problems
- Fatigue
- Constipation
- Cognitive changes including executive functions and memory problems and, in some cases, frontotemporal dementia
- Emotional and behavioural change including the following:
  - Emotional lability
  - Significant personality change
  - Disinhibition and impulsivity
  - Perseveration
  - Change in eating behaviour (sweet food preference)
  - Loss of emotional understanding (appear egocentric/selfish)
  - Withdrawn (apathy/failure to initiate)
  - Stereotyped/ritualistic behaviour
  - Behaviour change (MND Association, 2012)
1.5.5 Diagnosing MND

No specific diagnostic tests currently exist, but neurological investigations should normally include EMG, nerve conduction studies, blood tests and investigations that sometimes include MRI/CT scanning, lumbar puncture and muscle biopsy to exclude possibility of other neurological conditions. According to the MND Association (2013), MND can be extremely difficult to diagnose for several reasons:

- It is a comparatively rare disease.
- The early symptoms can be quite slight, such as clumsiness, mild weakness or slightly slurred speech, all of which may have been attributed to a variety of other causes.
- It can be some time before someone feels it necessary to see a GP.
- The disease affects each individual in different ways, not all symptoms may be experienced or appear in the same sequence.

Case study

Evie is 54 years old and lives with her husband Colin. They have three daughters and a son, three of whom live locally within the small village in which Evie grew up. Evie worked in the local school dinner hall and had recently become aware of some loss of function in her left hand. When she examined this a bit more closely, there was a noticeable loss of muscle bulk in her forearm which concerned her. Although seemingly unrelated at the time Evie was aware that at times she seemed to have difficulty forming words and her speech sounded slurred. One evening while Evie was sitting watching television, her husband drew her attention to the muscles in her forearm which were ‘flickering’ uncontrollably. Evie was aware of this but did not know why this was happening. While Evie did not connect any of the observations, she nevertheless made an appointment with her GP to allay her concerns.

When Evie attended the appointment with her GP, they discussed a number of factors. Evie then recognised that she had been experiencing muscle cramps in her legs but thought this was just due to her exercise regime. Evie remembered that she had tripped on a few occasions recently as her foot seemed to be tired, but this seemed to depend on which shoes she was wearing. Evie had never thought about her swallowing before, but when asked by her GP she realised that her family had made comments about her not taking time to digest her food properly. While at this stage none of this seemed to connect for Evie, there was sufficient evidence for a referral to the neurologist.

Evie attended the neurology appointment with her husband and eldest daughter. Following a thorough clinical examination and history Evie was referred for nerve conduction studies, blood tests, an EMG and an MRI scan. Evie was keen to understand what the neurologist was considering but he was giving very little away at this stage. Evie attended for all her appointments as arranged but in the meantime began to look for information on the Internet regarding the combination of symptoms which were becoming apparent. Evie’s aunt had died several years previously from motor neurone disease and Evie was beginning to wonder if there was any connection.

When Evie returned to the neurology clinic, she was given the diagnosis of motor neurone disease. Evie’s family were devastated as they had never anticipated this. Evie, being slightly more prepared for this possibility seemed more able to take on board the information which was being shared at this point. Evie was immediately referred to the MND Regional Care Officer and to the multi-disciplinary team. Evie had never heard of occupational therapy but was happy to meet the occupational therapist at the clinic.
1.6 Multiple sclerosis

MS is the most common disabling neurological condition in young adults, affecting around 100,000 people in the United Kingdom. It is most often diagnosed in people between 20 and 40 years of age and affects three times more females than males (MS Society, 2011).

The clinical course of MS may follow a variable pattern over time (Lublin and Reingold, 1996) and is usually characterised by episodic acute periods of worsening, described as attacks, exacerbations or relapse. Typically attacks arise subacutely over hours to days, then plateau and remit partially or fully over the course of weeks or months, either spontaneously or with intervention (Keegan and Noseworthy, 2002). MS is classified according to disease course with each presenting different clinical and epidemiological patterns. The most common classifications of MS include relapsing-remitting (RRMS), primary progressive (PPMS), secondary progressive (SPMS) and benign MS.
In the early stages following diagnosis people with MS typically present with a relapsing-remitting course in which clearly defined relapses are followed by full or partial recovery. Periods between disease relapses demonstrate a lack of disease progression. After an average of 15 years, the disease develops into a secondary progressive phase during which the course becomes either continuously progressive or progresses with occasional relapses, minor relapses and plateaus. PPMS is characterised by disease progression from onset with occasional plateaus and temporary minor improvements apparent (Lublin and Reingold, 1996).

1.6.1 Body functions
The function of the brain is to interpret sensations and initiate movements and other responses to those sensations. This activity depends upon a highly complex communication system of nerves running from the brain throughout the body via the spinal cord.

1.6.2 Body structures
Each nerve of this complex communication system can be compared to an electric cable. The inner part of the nerve, the axon, is made of conductive tissue and carries messages, or impulses, throughout the body. The axon is surrounded by a layer of fatty substance, the myelin sheath, which protects and insulates the nerve to prevent interference to the impulses passing along it.

Although the aetiology remains unknown, MS is thought to be an autoimmune disorder in which the immune system attacks the myelin sheath around the axons of the central nervous system, which results in plaques or lesions. Damage to the myelin interrupts and disturbs the transfer of information along the axon, with a subsequent failure to carry clear messages to various parts of the body. Axonal damage occurs in addition to demyelination and may be the cause of later permanent disability (Keegan and Noseworthy, 2002; MS Trust website, 2015) as shown in Figure 1.3.

![Healthy nerve cell and demyelination in MS. (Source: MS Trust website. Reproduced with permission of MS Trust.)](image-url)
1.6.3 Impairments
There are a number of symptoms or impairments associated with MS, none of which are unique to MS. There is no typical pattern of symptoms that applies to everyone and symptoms can vary in duration and intensity. It is unlikely that a person with MS will develop all of the symptoms listed below (MS Society, 2011):

- **Bladder dysfunction**
  - Urinary urgency
  - Urinary frequency

- **Bowel dysfunction**
  - Loss of sensation and neurological control
  - Faecal incontinence
  - Constipation

- **Sexual dysfunction**
  - Impotence
  - Decreased libido
  - Decreased sensation in the genital area
  - Decreased ability to maintain an erection
  - Absence of ejaculation
  - Decreased lubrication in the vaginal area (MS Trust, 2011)

- **Visual disturbances**
  - Optic neuritis
  - Diplopia (double vision)
  - Nystagmus

- **Fatigue**
  - Fatigability: a form of tiredness which occurs after prolonged activity and from which the person recovers after rest
  - Lassitude or a persistent feeling of exhaustion which is not related to rest and sleep

- **Pain**
  - Neuropathic or neurogenic pain, which is typically described as ‘burning, shooting, tingling, stabbing and/or hypersensitivity’ (MS Trust, 2011)
  - Nociceptive pain or joint pain

- **Spasticity**
  - Increased muscle tone due to muscle stretching, peripheral stimulation or infection
  - Involuntary muscle spasms

- **Cognitive dysfunction**
  - Memory
  - Attention
  - Processing speed
  - Visuospatial abilities
  - Executive functions including planning, problem solving, behavioural regulation, lack of insight, apathy and lack of motivation
• Tremor
  ◦ Postural tremor
  ◦ Kinetic tremor
  ◦ Intention tremor
• Muscle weakness
  ◦ Pattern of weakness usually more distal than proximal
  ◦ Generally greater weakness apparent in the upper limb extensor muscles and the lower limb flexors
  ◦ Footdrop impacting on mobility
• Balance
  ◦ Disturbance of the visual system due to double vision
  ◦ Impaired vestibular processing
  ◦ Damage to the cerebellum resulting in ineffective proprioceptive output
• Vertigo
  ◦ Transient experience or ‘feeling of dizziness where the world appears to be spinning’ (MS Society, 2013)
  ◦ Inner ear involvement due to nerve damage in the cerebellum or brainstem (MS Society, 2013)
• Altered sensation:
  ◦ Numbness (people often say they feel like they’ve had a local anaesthetic)
  ◦ Tingling or ‘electric shocks’
  ◦ Feeling extremities swollen/ feeling of ‘largeness’
  ◦ ‘Band-like’ sensation around chest or limb
  ◦ ‘Burning’
  ◦ Pins and needles
  ◦ Feelings like ants crawling under the skin
  ◦ Lack of awareness of temperature, both hot and cold
  ◦ Transient facial numbness (MS Society, 2011)
• Depression
  ◦ Low mood for more than 2 weeks
  ◦ Variation in mood throughout the day
  ◦ Negative thoughts about self
  ◦ Irrational or illogical thoughts
  ◦ Suicidal ideation
  ◦ Unable to experience pleasure and enjoyment (MS Trust, 2011)
• Emotional and behavioural changes
  ◦ Altered mood in both directions, including euphoria
  ◦ Mood swings
  ◦ Uncontrollable (and often inappropriate) laughing or crying
  ◦ Lack of insight
  ◦ Disinhibition
  ◦ Lack of initiative
  ◦ Withdrawal from usual activities (MS Society, 2011)
• Communication and swallowing
  ◦ Dysarthria or difficulty with speech production and intelligibility
  ◦ Dysphagia or difficulty in swallowing

1.6.4 Diagnosing MS
There is currently no single test or clinical feature which is exclusive to MS, and therefore other conditions must first be eliminated (MS Trust, 2011). The National Institute for Health and Care Excellence (NICE) guideline on the management of MS (NICE, 2014) requires that:

• Only a consultant neurologist should make the diagnosis of MS on the basis of established up-to-date criteria, such as the revised 2010 McDonald criteria (Polman et al., 2011) after:
  ◦ assessing that episodes are consistent with an inflammatory process
  ◦ excluding alternative diagnoses
  ◦ establishing that lesions have developed at different times and are in different anatomical locations for a diagnosis of relapsing-remitting MS
  ◦ establishing progressive neurological deterioration over 1 year or more for a diagnosis of primary progressive MS.

Case study

Brian is 42 years old and was diagnosed with relapsing-remitting multiple sclerosis 8 years ago. Brian went to his GP initially when he had several episodes of double vision, some ‘strange’ sensations in his right arm and because he was aware that his foot was dragging when he was walking. Brian ran his own business and worked long hours to make this successful. Brian was reluctant to visit his GP as he thought he was just tired from working excessive hours and that his wife was being over dramatic with her concerns.

Brian visited his GP who referred him for a lumbar puncture and an MRI scan. Brian felt this was very thorough, but somewhat unnecessary as he was still determined that his difficulties were due to over exertion at work. Brian attended for the investigations before being invited to attend a follow-up appointment to which he was invited to bring his wife along. Typically, Brian was running late for his appointment, trying to fit in that last minute work appointment.

During this appointment the neurologist advised Brian that there was evidence of plaques forming on his brain which were indicative of multiple sclerosis. He advised Brian that it was not possible to predict how this would progress but that he was presenting with a relapsing-remitting form of MS. Brian was not offered any additional support at this stage and was advised that he could continue to work and drive for the time being.

Brian was to arrange a follow up appointment with his GP and the MS specialist nurse.

Brian was naturally devastated at the outcome. However he did not fully understand the diagnosis and did not learn much more from his follow up appointment with his GP. Brian did not alter his lifestyle at all in response to this and continued to work long hours. Brian's wife was particularly concerned about him as she was aware that the happy-go-lucky man she married had become more irritable, always tired, reluctant to go out with friends, and all discussions about starting a family had ceased.
Within 2 months of the diagnosis Brian began to experience increasing difficulties with his walking, numbness in his right arm, and increasing difficulties reading documents at work. Brian felt even more tired than normal. Brian’s GP discussed the possibility of disease modifying therapies (DMTs), but Brian said that he did not feel that his symptoms were severe enough at this stage to start on this regime. Brian did not understand the purpose of DMTs and, therefore, was not able to make a fully informed decision about his ongoing care.

An appointment was arranged for Brian with the MS specialist nurse who was able to properly explain the purpose of DMTs once it was established that Brian was a suitable candidate for this intervention. The MS specialist nurse identified some mild cognitive deficits and was aware that Brian was having difficulty with some aspects of his work. Brian was referred to an occupational therapist at this stage.

By the time Brian attended the occupational therapy appointment he had made some fairly significant life decisions. He had decided to sell his business, had put his house on the market and both he and his wife had decided that they would not start a family. Brian was still driving and had not notified DVLA of his diagnosis. Brian presented with difficulties with impaired sensation, reduced mobility, diplopia, urinary urgency, fatigue, mild memory problems and difficulties with executive function. Brian feared for the future as he felt that everything that was important in his life had been slowly eroded and he found it hard to see where his future would lie.

Brian had never heard of occupational therapy before and had no idea why he had been referred. Initially he did not think that occupational therapy would be of any benefit to him until the occupational therapist helped Brian to truly understand what was going on in his life and why this was happening. Together they developed a programme of interventions which helped Brian to continue to engage in meaningful occupations and participate within his wider communities.

1.7 Parkinson’s

Parkinson’s is predominantly a progressive movement disorder affecting 1 in 500 people. The common age of presentation of symptoms is 65 years; however, Parkinson’s can occur in younger people as 20% of those newly diagnosed are under the age of 40 years. Statistically, men are slightly more likely to develop the condition than women (Parkinson’s Disease Society, 2007).

1.7.1 Body functions

Dopamine is a major neurotransmitter (a chemical messenger) produced in the basal ganglia which activates receptors in the motor cortex to produce coordinated, voluntary and semi-automatic motor skills and movement sequences (Parkinson’s Disease Society, 2007). Depletion or deficiency in dopamine can lead to delayed and uncoordinated movements (Parkinson’s Disease Society, 2007).

1.7.2 Body structures

Parkinson’s is characterised by slow or impoverished movements which impact on the ability to function normally within daily routines. In addition to the
disruption of smooth co-ordinated movement patterns dopamine depletion can also contribute to changes in speed of processing, problem solving, decision-making, visual perception, attention, mood and motivation (Parkinson’s Disease Society, 2007). While the cause of Parkinson’s remains uncertain like many long-term neurological conditions, environmental factors and genetic susceptibility are contributory factors. Approximately 5% of cases of Parkinson’s are directly inherited and are usually manifest as early-onset Parkinson’s (less than 40 years of age). To date, 10 genes associated with inherited Parkinson’s have been identified (Parkinson’s Disease Society, 2007).

1.7.3 Impairments

- Bradykinesia or slowness of movement
- R rigidity
- Balance
- Hypokinesia
  - Loss of facial expression
  - Loss of arm swing
- Tremor
  - Apparent at rest
  - Postural tremor
  - Intention tremor
  - Fine rhythmic movement of the thumb and index finger (pill-rolling)
- Freezing
  - Loss of initiation of movement
  - Leads to unpredictable ‘on’ and ‘off’ phases
- Pain
  - Neuropathic or ‘nerve’ pain
  - Nociceptive or musculoskeletal pain
- Restless legs syndrome (RLS)
  - Unpleasant sensations in the legs
  - Uncontrollable urge to move the legs
  - Involuntary jerking of the arms and legs
- Sleep problems
  - Excessive daytime sleepiness
  - Poor sleep regulation
  - Sudden onset of sleep
- Fatigue
  - Fatigability
  - Lassitude
- Bladder problems
  - Nocturia (night-time voids)
  - Urinary urgency
  - Urinary frequency
• Bowel problems
  ◦ Constipation
  ◦ Problems emptying the bowel due to weak abdominal straining and the anal sphincter not relaxing
• Visual problems
  ◦ Difficulty moving eyes
  ◦ Blurred vision
  ◦ Diplopia
  ◦ Dry eyes
  ◦ Involuntary opening and closing of eyelids
• Speech and communication
  ◦ Dysarthria
  ◦ Breathy, nasal or harsh voice
  ◦ Monotony with reduced loudness and pitch range
  ◦ Difficulties with speech initiation
  ◦ Variable rate of delivery of speech
  ◦ Stuttering speech patterns with frequent pauses
  ◦ Short rushes of speech
  ◦ Imprecise consonants
  ◦ Difficulty writing
  ◦ Problems with auditory comprehension
  ◦ Limited eye contact
  ◦ Lack of facial expression (Parkinson’s Disease Society, 2007)
• Swallowing
  ◦ Dysphagia
  ◦ Saliva overproduction leading to drooling
  ◦ Weight loss
  ◦ Fear of swallowing
  ◦ A ‘gurgly’ voice
  ◦ Coughing before, during or after swallowing
  ◦ Difficulty swallowing medications
  ◦ Reduced social contact
  ◦ Recurring chest infections
  ◦ Bronchopneumonia (Parkinson’s Disease Society, 2007)
• Cognitive impairment
  ◦ Executive function
  ◦ Planning
  ◦ Sequencing
  ◦ Working memory
• Dementia
  ◦ Parkinson’s with dementia (PDD) resulting in the following:
    ◦ Marked cognitive slowing
    ◦ Impairment of visuospatial abilities
    ◦ Memory problems
Introduction

Dementia with Lewy bodies (DLB) characterised by the following:
- Cognitive fluctuations (resembling a chronic confusional state)
- Visual hallucinations
- Mild Parkinsonism (Parkinson’s Disease Society, 2007)

Emotional and behavioural change
- Anxiety
- Depression
- Hallucinations

1.7.4 Diagnosing Parkinson’s
There is no definitive diagnostic test for Parkinson’s. Single-photon emission computed topography (SPECT) scanning should be considered for people with tremor where essential tremor cannot be clinically differentiated from Parkinsonism (NICE, 2006).

Case study

James is 58 years old and lives alone, following the death of his mother 4 years previously. James had never married and lived within the family home with his parents all his life. James is a quantity surveyor and worked all over the country depending on where his company placed him. James was a hardworking and loyal employee having worked with the same company for over 30 years. James has many friends and enjoys a very active life including swimming, cycling and running.

Two years ago James became aware of stiffness in his dominant arm. At times there would be a shakiness apparent, but James wasn’t aware of this happening at particular times or when doing specific tasks, other than that he was more aware of it when he was relaxing watching the television. James maintained a fairly regular routine, going to bed quite early in the evening so that he could get up early in the morning to exercise before he went into work at least an hour before his starting time. Gradually James felt that he wasn’t sleeping as well describing jerkiness in his legs during the night. This woke James up on occasions, but he seemed to settle again once he moved his legs.

During a routine medical assessment at work, James shared his concerns about the stiffness in his arm. James was advised to attend his GP who referred him to a Neurologist. Initially James was advised that he had an essential tremor. James however did not feel reassured by this diagnosis as this did not explain the stiffness in his arm. During a follow-up appointment with the neurologist, however, James was diagnosed with Parkinson’s. In addition to the stiffness becoming more apparent in James’ upper limb, he had become aware of changes in his posture, his walking was slower and stiffer, he was constipated, had dry eyes and was experiencing difficulties with writing and occasionally choking on his food.

James was referred to a Parkinson’s specialist and the multi-disciplinary team, including occupational therapy. The occupational therapist carried out a full assessment of James both in the clinic and within his own home. On assessment it became apparent that James was experiencing a range of difficulties including: standing from a low armchair; preparing and eating food; accessing his shower; loss of confidence in his driving ability; reduced stamina and tolerance in walking. James was climbing into bed on his ‘all fours’ but could...
not turn to get into a lying position once he was on the bed. He was lying awake at night worrying that he would need to get up to the toilet as he had difficulty getting back out of bed.

In addition to the practical difficulties James was also experiencing a general loss of confidence reflected in his withdrawal from social activities. He was now on long-term sickness leave from work and was negotiating a medical retirement package with his employer. James was no longer exercising as he found he was particularly stiff in the morning and needed some time before his medication kicked in, allowing him to move more freely. James had notified DVLA of his diagnosis but was no longer driving as his friends had indicated that they felt his driving was unsafe due to his slow decision-making and commented that he was leaving extra distance between other vehicles when he was driving. James felt he did not need to drive and was able to access sufficient support from family and friends when he needed to go anywhere.

James had commenced a medication regime but was experiencing fluctuating levels of ability in response to this. He described episodes of freezing, particularly when going through doorways. This made James feel anxious about his ability to cope, which at times made him feel quite sad. James worried about disease progression as he was a very private man and worried about having to accept support in the form of professional carers coming into his home. James had privately purchased a chair which had been expensive and which was not making it any easier for him to stand up. James was experiencing some difficulties negotiating with his employers due to their apparent lack of understanding about his reasons for being unfit for work.

1.8 Self-evaluation questions

1 What is the economic impact of long-term neurological conditions on health and social care systems?
2 What are the three levels of human functioning within the ICF framework?
3 What are the most common cognitive impairments in HD?
4 What are the most common classifications of MS?
5 What are the key differences between tremor associated with MS and a Parkinsonian tremor?
6 Which of the four neurological conditions – HD, MND, MS and PD – have a genetic inheritance?
7 What is the difference between fatigability and lassitude?
8 What are the main forms of MND?
9 What is the function of dopamine?
10 What are the main causes of depression for people with long-term neurological conditions?

References


