

4

PALLIATIVE CARE FOR PEOPLE WITH MULTIPLE SCLEROSIS

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LEARNING OUTCOMES

- Identify the common symptoms and different subtypes of multiple sclerosis (MS)
- Describe the experiences patients often encounter in living with the disease and its unique nature
- Consider the palliative care as it relates specifically to MS
- Describe the role of the nurse in supporting patients and families at the end of life

INTRODUCTION

The purpose of this chapter is to provide information on the pathophysiology of MS and some of the common symptoms and subtypes of the disease. It also looks at the various treatments used to control relapses and prevent symptoms such as muscle spasm through the use of prescribed medication, massage and the use of medicinal cannabis (sativex). Moreover, consideration is given to the unique nature of MS and how each individual with the disease has their own experiences. It does this through the use of case studies which examine the problematic nature of diagnosis and the importance of quality of life and living with the disease. The case studies are used as an anchor within the chapter to help focus the rest of the symptom management and patient experiences. Due to the complex nature of MS, patients may be referred to a neuro-rehabilitation team or Multiple Sclerosis

specialist practice nurse for symptom management. As the disease progresses, social care can be provided to assist with Activities of Daily Living (ADL). When the disease reaches the palliative phase, social care support often continues in conjunction with palliative services.

As the case study will illustrate, palliative care is focused on providing optimal care, not just for the patient but the family caregivers who are provided with various amounts of support from professional caregivers and organisations such as the MS Society and MS Trust. Finally, the chapter looks at palliative care end of life, highlighting a number of challenges for healthcare professionals.

BACKGROUND

Multiple sclerosis (MS) is a serious degenerative neurological condition that involves damage to the body's neurological system, specifically the myelin sheath covering the nerve (Alonso et al., 2007). The global prevalence of MS is increasing with an estimated 2.3 million people diagnosed in 2013, representing an increase of 0.2 million since 2008. In the UK it has been estimated that over 100,000 people are living with MS (Alonso et al., 2007). These figures are likely to underestimate the growing problem which is confounded by the high incidence of misdiagnosis, which is a very challenging and persistent problem that has significant consequences for patients and healthcare professionals (Solomon and Corboy, 2017). The condition often begins between the ages of 20 and 50 and is more common in women than men on a ratio of three females to every one male (3:1) (MS Society, 2017). The damage to the nerve cell involves a breakdown in the outer covering of the nerve, known as demyelination. Demyelination is the key pathophysiology of the disease, which can lead to numerous physical and psychological problems that impact on ADL, resulting from the neurological damage.

SYMPTOM RECOGNITION AND EFFECTS

MS is a long-term illness characterised by periods of remission and relapses of symptoms often referred to as attacks. These episodes can cause pain, lack of mobility, incoordination and a number of psychological issues such as anger and depression. One of the features of these relapses is their lack of predictability. Practitioners working with people with MS need to be aware not only of the disease progression but also the impact it has on the individual patient. In the palliative phase of the illness, the relapses may become more frequent, severe and debilitating. One of the consistent features of MS throughout the disease progression is fatigue, which is more than feeling tired and involves a range of psychological issues like frustration and anxiety which can be treated and the person supported.

BOX 4.1 MAIN SYMPTOMS OF MS

- Fatigue
- Muscle spasticity, weakness and ataxia (balance problems)
- Visual disturbances
- Speech difficulties (dysarthria)
- Swallowing problems (dysphagia)
- Cognitive impairment
- Depression
- Anxiety and frustration
- Unstable mood
- Pain, sensation (hypoesthesia, and tingling – paraesthesia)
- Bowel problems (incontinence, diarrhoea and constipation)
- Urinary frequency and/or retention

Box 4.1 provides an overview of the main symptoms associated with MS. MS leads to difficulties in coordination, problems with balance and mobility, eyesight, musculoskeletal problems, bladder difficulties, speech problems and tingling sensations in the peripheral limbs. There are also a wide range of other problems, sometimes referred to as invisible, that are associated with MS that become part of the more physical symptoms. These include fatigue, which is ever present throughout the condition and can be severe during attacks. Many people with MS report that they experience a combination of symptoms at the same time.

Some of the symptoms illustrated in Box 4.1, such as speech difficulties, may occur in the initial stages of MS as a result of dysarthria and thereafter not become troublesome to the patient as they adapt to the difficulty in mobilising the tongue in the mouth. Similarly, visual problems can be accommodated and, although initially worrying, can be less of a problem than symptoms such as muscle spasticity which can cause pain, discomfort and lack of mobility. Muscle spasticity can be a consistent feature and one that the chapter will discuss in more detail due to the problem in managing this distressing symptom. Other symptoms such as hypoesthesia and paraesthesia and bowel disturbances can result from the use of certain medications such as buscapan, used to control muscle spasm. The psychological effects of MS, such as anxiety, mood changes and cognitive impairment, affect some people more than others. Much depends on the individual's attitude, personality and level of tolerance. Moreover, the support provided by others and the care and attention they receive can make a significant difference to individual wellbeing. In Greater Manchester the MS Centre exists as an information provider and source of social support for people with MS. As a day facility it provides people with MS and their caregivers with a place where they can receive treatment and social support, very important in the management of symptoms. One of the universal issues about symptom management in MS is its variability. People with MS may not experience

the range and severity of some symptoms like visual disturbances. Most would agree that muscle spasticity is a prevalent feature and one that causes pain and anxiety. It can also become a distressing problem giving rise to other symptoms such as anxiety and severe frustration. Due to the unpredictable nature of MS, not everyone experiences all these symptoms and certainly not at one time.

DIFFERENT FORMS OF MS

MS takes several forms and has different subtypes with new symptoms either occurring in isolated attacks (relapsing forms) or building up over time (progressive forms) – see Box 4.2. A recurrence of the symptoms once they have remained dormant for several months or years is referred to as an attack. Between attacks, symptoms may disappear completely; however, permanent neurological problems often remain, especially as the disease advances. Despite the underlying cause being ambiguous, the pathology of the disease is focused on dysfunction of the immune system or failure of the myelin-producing cells. Proposed causes for this include genetics and environmental factors, with viral infection being a likely trigger. Another likely trigger is stress, which like other illnesses such as stroke can precede the onset of symptoms (Methley et al., 2014). MS is usually diagnosed based on the presenting signs and symptoms and the results of supporting medical tests.

BOX 4.2 SUBTYPES OF MS

1. Progressive-relapsing MS: Characterised by steady decline from onset with superimposed attacks
2. Secondary progressive MS: Characterised by initial relapsing-remitting MS that suddenly begins to decline without periods of remission
3. Primary progressive MS: Characterised by a steady increase in disability without attacks
4. Relapsing-remitting MS: Characterised by unpredictable attacks which may or may not have permanent decline followed by periods of remission (Methley et al., 2014).

DIAGNOSING MS

MS is commonly diagnosed between the ages of 20 and 40, although it can be diagnosed in children and in later life. A confirmed diagnosis, which matches the symptoms to MS, happens after the patient has had two isolated episodes or attacks. In terms of Dennis and his experiences, it is necessary to say that despite the signs of MS no two people have exactly the same symptoms (Solomon and Corboy, 2017).

CASE STUDY 4.1 DENNIS

Dennis was a 27-year-old who lived at home with his parents. His father (Eric) was diagnosed with MS in his 20s, around the same age Dennis began to have symptoms of MS. It seemed to start very innocently. He was aware that MS had a tendency to be familial and he had looked up what the symptoms were online. He put it out of his head, arguing with himself that it was not worth worrying about since he could not do anything about it anyway. He had an older brother who did not have MS, married with a baby girl. Dennis enjoyed his job working for a university as an IT technician and was very competent at his job. He had an active social life and was passionate about football, playing for a local Sunday league team. He enjoyed the games and the social life that existed in the pub after the matches.

It was just after his 28th birthday that he recalled feeling lethargic, but he put this down to the amount of drinking at his birthday party and the subsequent hangover. He started to experience blurred vision, although at first it was hard to say exactly what he was experiencing except that he did not feel as though he was seeing things properly. He also started to feel frustrated at work, becoming unable to sort out problems which he normally would have dealt with well. Perhaps most noticeably, he seemed to lose his football skills and suddenly felt as though he had two left feet. He thought he was just going through a low period and carried on as normal.

In the next few weeks, things started to deteriorate. His girlfriend (Tanya) noticed that he was not as quick to answer questions at the pub quiz. He put his general lack of motivation down to stress at work and the threat of redundancy. Had it not been for a conversation Tanya had with his mum when she told her he was skipping football practice, Dennis would have carried on for a long time without a diagnosis. His mum too had noticed his lack of interest and lethargy and commented that it was unlike him not to want to go out and play football. Dennis admitted that he felt a bit out of sorts and his mum wondered whether he should see his GP. Dennis also said he had begun to experience pins and needles in his arms when he used his laptop and once or twice felt giddy when he got up from the chair. His mum suspected the worst and was glad when Dennis agreed to make a GP appointment.

The GP asked Dennis a few questions about his symptoms. He also took a blood sample. After he disclosed that his dad had MS, the GP decided to refer him to a neurologist at the local hospital.

The neurologist explained to Dennis that he would need to have a series of tests to investigate his symptoms. These included a lumbar puncture (LP, or spinal tap) to check the fluid in his spinal column as well as a magnetic resonance imaging (MRI) scan of his brain. He also explained that to confirm diagnosis he would need a number of electrical tests (evoked potential) to see what damage may have occurred to his nerve pathways.

After a few weeks Dennis was asked to go back and see the neurologist who confirmed test results showing signs of early onset MS.

The investigations carried out to diagnose MS vary, although MRI scans rather than the more traditional and invasive lumbar puncture are often performed. MRI scans are a very effective way of demonstrating changes in the brain caused by multiple sclerosis. It can show the neurologist clear signs of inflammation in the deep parts of the brain and spinal cord that are indicative of MS. However,

the MRI brain scan test alone is not always conclusive since older people and those with high blood pressure and diabetes can also show inflammation in the brain which may imitate MS. It is necessary to do several investigations before confirming the diagnosis. It has also been known for a person with MS to have a normal MRI scan, as approximately 5 per cent of people with the condition do not have lesions in the brain; instead they may have lesions that do not show up on the MRI scan.

For a GP this is a very challenging situation, mainly because they may not encounter such patients very often. This makes it difficult to diagnose symptoms, especially as there is no single definitive test for MS. This accounts for the fact that many people remain undiagnosed for long periods.

DISEASE PROGRESSION AND TREATMENT

Disease progression and long-term outcomes are difficult to predict in MS. Positive outcomes are, moreover, more common in women, those who develop the disease early in life, those with a relapsing course and those who initially experienced few attacks (Pugliatti et al., 2002). Treatment attempts to improve function after an attack and prevent new attacks. There are 11 licensed treatments for MS, often referred to as disease-modifying therapies (DMTs). The drugs are largely available for people with relapsing, remitting MS and range from Tysabri, Lemtrada, oral therapies, to copaxone and the interferons. Large doses of steroids (methylprednisolone) are often used to treat relapses in order to reduce inflammation and are given intravenously in hospital or taken orally. Medications used to treat MS, while modestly effective, can have side effects and be poorly tolerated. Drug regimens such as interferon require a high level of compliance in order to achieve optimal efficiency (Freidel et al., 2015). In their study of the effects of interferon and nursing care, Freidel et al. found that in order to gain full benefit from disease-modifying therapies such as interferon β -1b, patients with MS needed to adhere to treatment in the long term. Their findings indicated that patients who rated nursing care as effective and valuable were more adherent to the drug regime. In other words, patients found it useful when nurses focused their attention on the individual problems identified by the patient. They also found that nursing care was valuable because they could trust the judgements of the nurse when providing information about the course of the disease. Despite the lack of therapeutic change in their condition following drug treatment, patients benefited from effective care that included information giving, telephone follow-up and close contact with nurses.

MEDICINAL CANNABIS AND MS

More recently, many people with MS are looking to complementary and alternative medicine (CAM) for answers to symptom control issues, despite a lack of

sound evidence. One area of pain management that is becoming increasingly popular for people with MS is the use of medicinal cannabis for symptom control, specifically muscle spasticity. Specifically the use of cannabis for people with MS is symptomatic as MS is a neurological condition that gives rise to muscular pain and discomfort. Medicinal cannabis (sativex) can help to control the distress caused by muscle spasm and pain. It is available on the NHS although not in all parts of the country. Some GPs will prescribe it despite the cost and the lack of endorsement from NICE. It is available as a spray. The MS Society has published data on the rhetoric and reality of using cannabis on their website (www.mssociety.org.uk). Despite claims that cannabis is a natural drug its main ingredient is tetrahydrocannabinol (THC). THC is ‘psychoactive’, and can alter thinking and create hallucinations. People with MS who may have a family history of mental health problems (such as schizophrenia or bipolar disorder) are advised not to use cannabis as it can make the mental health symptoms worse. The other active ingredients in cannabis are cannabinoids (CBD), which have anti-inflammatory and antispasmodic properties. Here are some of the comments from patients with MS who have used cannabis.

NICKY’S STORY

“Muscle spasms and stiffness cause my legs to clamp together, my arms to go rigid, and my body to fling itself backwards. Around four years ago I was able to try medicinal cannabis (sativex). It transformed my life. My neurologist and my GP submitted three requests for me to get sativex but each was unsuccessful. I’m now paying for sativex myself. It costs £500 a month. It’s horrible for my kids to see me crying in pain. So I think the sacrifices have been worth it.”

STEVE’S STORY

“Spasticity causes excruciating cramps and rigidity in my legs. Sativex hugely improved my quality of life. But I have to pay for it myself. Over the past four years, I’ve only managed to buy seven months’ worth. Cannabis isn’t right for me but for those it helps, it should be made legal.”

The clear message from the MS Society is that since cannabis is illegal there can be no full guidance about doses or quality; therefore, it is not possible to be confident that smoking cannabis is entirely safe and effective. This is why the MS Society is asking the UK government to review the current laws and make cannabis available for medicinal use in the treatment of muscular pain and spasms where other treatments have failed.

AUTOLOGOUS HAEMATOPOIETIC STEM CELL TRANSPLANTATION (HSCT)

Another area of palliative care treatment and symptom control for MS is autologous haematopoietic stem cell transplantation (HSCT), which involves the use of high doses of cancer chemotherapy drugs to wipe out harmful cells in the immune system. The patient's own stem cells are used to 'regrow' their immune system so that it no longer attacks myelin, thus reducing disease progression and preventing inflammation in the brain and spinal cord. The process involves taking a number of drugs to stimulate release of stem cells from the bone marrow into the blood stream. These cells are collected and kept frozen. The patient is then required to take chemotherapy drugs which either completely eliminate (myeloablative or high intensity chemotherapy) or partially eliminate (non-myeloablative or low-intensity chemotherapy) the bone marrow and immune system (Nash et al., 2017). Stem cell transplants carry risks and although all measures are taken to reduce these risks, clinical trials since 2001 have still had treatment-related death rates of one or two people in every hundred (Muraro et al., 2017). Current treatment methods for people with MS have favoured lower-intensity chemotherapy, which carries a lower risk of complications and death. This method results in the replenishment of the patient's immune system using his/her own stem cells. These cells then develop into the different types of cells found in the blood, including some cells which are part of the immune system. Recent research into the long-term outcomes of HSCT (Muraro et al., 2017; Nash et al., 2017) have reported on levels of disability in people with MS five years after receiving stem cell transplants. It is perhaps not surprising that HSCT take-up is quite low (Nash et al., 2017), especially when it is taken into account that the treatment involves enduring some distressing side effects from the drug.

PALLIATIVE CARE FOR PEOPLE WITH MS

Palliative care for people with MS is often not provided until the disease reaches the end of life stage. People with MS have symptom flare-ups and periods of remission when they feel independent and symptom free. This makes it difficult to provide consistent palliative care services, particularly when patients place great value on their ability to remain independent. However, when patients reach the palliative phase in their illness, palliative care can play a role in sustaining independence, rehabilitation and ensuring quality of life. This can occur through inpatient physiotherapy and occupational therapy (OT), both of which play a significant role in enabling people with MS to remain independent and develop self-management strategies to help optimise ADL functioning. Moreover, people with MS benefit from interventions made by occupational therapists who provide palliative care services, such as the provision of adaptations to the home. The OT services are focused on keeping the patient independent by using a range of equipment such as ramps for wheelchair access and grab rails to ease access to rooms and toilet

facilities. In the palliative phase, the OT can facilitate the fitting of ceiling track hoists to aid moving and handling in and out of bed. Equipment is also provided to assist with a range of enhancements to ADL such as eating, drinking, washing, dressing and elimination needs.

One of the key features of the palliative care offered to people with MS is the focus on survivorship and wellbeing. The key message from groups like the MS Society internationally seems to be focused on the development of physical and psychological wellness through effective health promotion. Palliative care is focused on quality of life, and for many people with MS feeling good about themselves physically and psychologically is closely related to overall wellbeing (Wollin et al., 2006). Wellbeing is promoted in a variety of ways including health education and advice on issues such as nutrition, although at present, the role of nutrition is unclear, and MS therapy is not associated with any particular diet (Mowry et al., 2017). There is also help available on how to get support and treatment from professionals as well as help and information about medication and how to manage drug side effects as well as alternatives to conventional drugs. The following case study illustrates some of the key points about palliative care for people with MS.

CASE STUDY 4.2 SARAH

Sarah was a 66-year-old woman who lived in North Wales, diagnosed with MS at the age of 22. Sarah and her husband Eric had been married for 42 years with no children. In the initial years, Sarah was told that she had the progressive degenerative type of MS, which followed a pattern of remission and relapses. She spent much of her early life with MS getting used to periods when she felt very tired, and despite treatments and various remedies the fatigue became a difficult burden to bear. She also had problems with muscular weakness and spasticity. Just after she retired from work as a civil servant, Sarah suddenly became very unwell and she felt as if she had the flu. Her initial symptoms included a tremendous feeling of fatigue. She adapted to these symptoms initially although they did progressively become more severe. Over a period of one year she gradually began to lose her balance and this was followed by a lack of mobility. Over a two-year period she gradually became unable to walk. Sarah had always been underweight and over the initial few weeks of feeling ill her weight dropped to under 9 stone. Her GP paid a visit and confirmed her worst fears that it was in fact a severe flare-up of her MS. She was at least used to these over the years but since entering retirement she lived in fear and uncertainty about her health. Over a relatively short period Sarah's condition had deteriorated. Fortunately, with Eric's help and with assistance and support from friends, district nurses, Macmillan and Marie Curie nurses, Sarah was cared for at home. The district nurses called in a specialist MS nurse (Jenny) to advise them on how to provide the best care and she visited Sarah to help with her nursing assessment. Jenny listened to Sarah and asked her a lot of questions about her symptoms and what she felt they could do for her. Jenny checked the prescribed pain relief and also spent time talking to Eric and asking him how much support he was receiving. Eric received help from the community nursing team and

Marie Curie nurses provided a night sitting service once a week to enable him to get a good night's sleep. Jenny also advised Sarah to attend an MS therapy centre in Greater Manchester for specialist treatment and support. Sarah was interested, but was put off by the travelling. Over the next six months Sarah's condition deteriorated. The next visit by the district nurse prompted Sarah to discuss advance care planning.

Sarah's stated preference was to spend her last days at home with Eric and her Labrador dog Sheba. Sarah's main physical problems were anorexia, incontinence, weight loss, the development of pressure sores, swallowing difficulties and pain related to muscle spasticity. Despite the rapid decline in her physical condition she remained relatively alert and focused mentally. Sarah received palliative care from a range of community staff, including the GP, community physiotherapist and occupational therapists, social worker, speech therapist and community nursing staff (including the continence advisor nurse). They all played a part in enabling her to remain at home with a reasonable quality of life. The community nurses arranged for an NHS profile bed (with a pressure-relieving mattress) to be provided that enabled her to sit up. Ceiling hoists were fitted due to mobility issues and to help with moving and handling. In Sarah's case, her weight loss was quite extensive and Eric found he could move her about the bed by himself using a slide sheet. A commode was made available at the bedside. This was used in conjunction with a soft plastic bedpan that Sarah sometimes preferred to use. She eventually had an indwelling urinary catheter inserted towards the end of her life. Sarah was able to sit out of bed which she appreciated – having the dog at her feet and looking out of the window onto the countryside around her which she loved. Sarah was provided with oxygen via a nasal cannula (24%) as her breathing became more laboured especially as a result of the chest infection she developed towards the end of her life. She developed a chest infection and refused antibiotics, as she was aware that she was near the end of life. Sarah died in the early hours of the morning three months after her initial flu symptoms. She remained mentally competent most of the time. Eric described it as a 'good death'.

Sarah's case study highlights a number of physical problems that occur throughout the course of the disease that remain very relevant at the end of life. One of these, which is susceptible to palliative care treatment, is spasticity of the muscles (Embrey, 2009; Edwards et al., 2008). It is one of the most distressing symptoms of MS and requires the use of a range of muscle relaxants such as buscapan and anti-spasmodics such as baclofen, and drugs such as diazepam to treat muscular spasms that occur at night. Sarah's GP prescribed a range of drugs that could be used as required that included baclofen, gabapentin steroids and a range of pain killers non-opiates such as tramadol and morphine (MST). NICE guidance (2014) on the use of antispasmodics recommends the use of baclofen or gabapentin as first-line drug treatments to treat spasticity in MS depending on contraindications and the person's comorbidities and preferences. Gabapentin is a drug used in the treatment of neuropathic pain in multiple sclerosis. It is specifically used to treat pain caused by the effects of MS, such as trigeminal neuralgia or abnormal sensations (dysaesthesia), such as burning or pins and needles. It is also used for the treatment of spasticity although one of its side effects is fatigue. NICE guidance (2014) suggests

that if the person with MS cannot tolerate one of these drugs, consider switching to other antispasmodic drugs such as tizanidine or dantrolene as a second-line option to treat spasticity. A third option is the use of benzodiazepines.

In some cases, patients can (although Sarah did not) have antispasmodic drugs intrathecally via a pump in order to ensure therapeutic levels are maintained. Sarah's GP, Bob, was aware of her MS and had prescribed antispasmodics for many years previously, he was aware of her wishes and ensured that she had a regular prescription.

NICE GUIDANCE ON SYMPTOM MANAGEMENT

- People with MS should have tried the drug at an optimal dose, or the maximum dose they can tolerate. Moreover, the drug should be discontinued if there is no benefit at the maximum tolerated dose.
- Once the optimal dose has been reached, the patient needs to have all their medication reviewed at least annually.
- Drugs used to control spasticity – i.e. baclofen or gabapentin – should be used as first-line drugs to treat spasticity in MS depending on contraindications and the person's comorbidities and preferences. If the person with MS cannot tolerate one of these drugs, consider switching to the other.
- Drugs such as tizanidine or dantrolene should be used as second-line options to treat spasticity in people with MS.
- Benzodiazepines should be considered as a third-line option to treat spasticity in MS. Prescribing physicians need to be aware of their potential benefit in treating nocturnal spasms.
- Should symptoms of spasticity not be managed with any of the stated pharmacological treatments, the patient should be referred to specialist spasticity services.

NICE guidance (2014) on the management of symptoms does not endorse the use of medicinal cannabis (sativex) because it is not cost effective. The guidance makes it clear that individual GPs can make their own decisions about its cost and effectiveness.

NON-PHARMACOLOGICAL FORMS OF TREATMENT

The control of muscle stiffness for Sarah was a key symptom and was focused on by the nurses who carried out active and passive leg exercises (as Eric did in their absence), physical movement was supported by the physiotherapist, a key contributor to the palliative care team (Holland et al., 2011). The overarching problem of muscle spasm and muscle pain is often a very common problem (Rosti-Otajarv and Hamalainen, 2011) and patients like Sarah use their own strategies to relieve stiffness such as heat pads, and warm baths so it's important to listen and act on the patient and carer's advice as healthcare professionals

(Malcolmson et al., 2008). Closely related to muscle management is insomnia as the reason for lack of sleep is often muscle spasm. The MS Society UK produces some very useful advice on how to manage insomnia that includes doing more during the day and basic exercises. Together Sarah and Eric ensured that they had a routine: a set time to wake up and to go to bed, regular meal times, lots of rest and some activity.

Management of continence was a key issue for Sarah as well as many patients with MS at the end of life. Finding and utilising appropriate continence aids can improve quality of life and the continence advisor was beneficial in helping to assess Sarah's individual needs. Initially Sarah was reluctant to have an indwelling urinary catheter because of the risk of infection in the long term. This was discussed during the session on advanced care planning and the nurses persuaded Sarah of the need to stay continent by having an indwelling urinary catheter inserted towards the end of her life in order to prevent the complications of pressure ulcers and to prevent loss of dignity. Consideration was given to having a suprapubic catheter surgically inserted to prevent infection, in order to avoid the problem of bypassing of urine around the catheter. Sarah did not wish to have that type of catheter and wanted minimal invasive medical intervention. A further physical challenge to optimising palliative care was maintaining skin integrity. Skin care and the prevention of decubitus ulcers in patients towards the end of life is a prime consideration in terms of quality of life. Physical challenges often centre on the prevention and management of the consequences of problems such as pressure ulcers that can add to disability.

This is an issue highlighted in the NICE guidelines. Infections (especially respiratory and urinary tract infection) are a constant threat to quality of life especially in the advanced stages of the disease (NICE, 2003). The guidelines highlight the need for attention to be given to hygiene (both person and caregiver, with a focus on hand washing when emptying catheters and if self-catheterisation is used (NICE, 2003). In Sarah's case, Eric was such a diligent and meticulous caregiver that options such as self-catheterisation were not required. Sarah used a Silastic indwelling urinary catheter with a short leg bag at night and a long bag during the day when able to sit out of bed. Infection is a major issue depending on the person's level of mobility; the more disabled people become, the greater the risk of infection. Many people with MS lose the ability to walk before death, 90 per cent are capable of independent walking at 10 years from onset, and 75 per cent at 15 years (Rossier and Wade, 2002).

Apart from the physical challenges, there are numerous psychosocial aspects to living with MS that nurses and healthcare professionals need to be aware of in order to provide optimal care (Patten and Metz, 2002). The psychological issues can and do begin with the diagnosis and the transition to living with MS, coping with the symptoms, the relapses and the treatment (Malcolmson et al., 2008). Depression and suicide are more common in people with MS than other neurological conditions (Royal College of Physicians, 2011). This is related to the roller

coaster type illness trajectory which accompanies MS, with some patients having no active symptoms for years yet knowing they may reoccur, living in a cognitive and emotional climate of uncertainty about remission and relapse times. Specialist nurses who work with people with MS express empathy, compassion and understanding of the disease but remain focused on keeping the patient in control as much as possible to retain autonomy (De Broe et al., 2001). Clearly, the focus of the nurse is on the patient, although palliative care practitioners will recognise the importance of supporting caregivers who may often feel despair and frustration towards the patient, especially when employment is affected and financial stability becomes an issue (McKeown et al., 2003). Patients with MS also express concern about the level of social care provided by statutory bodies as limited resources and cutbacks can mean more financial hardship for families and individuals (Chang et al., 2002).

PALLIATIVE CARE AT THE END OF LIFE FOR PEOPLE WITH MS

In relation to palliative care at the end of life, illness progression and cause of death are invariably related to the consequences of the disease, with infection being a major cause of mortality at the end of life (Tsang and Macdonell, 2011). The patient's prognosis is dependent on a number of factors, namely: gender, age, the subtype of the disease and the early onset of symptoms as well as the extent of the individual's disability (Ontaneda et al., 2017). One of the key features of MS, as illustrated by Sarah's case study, was the prolonged uncertainty that periods of remission can create. For many patients and their partners and caregivers, the experience of MS is shrouded in uncertainty. This can create a false sense of security and hope that the disease itself has burnt out. This may perhaps help to explain the irritation and severe frustration that is often reported by people with MS. The previous case study provides an illustration of one woman's experience of living with and dying from the progressive degenerative type of MS. There are several major nursing challenges to the provision of effective palliative care for people like Sarah at the end of life. Broadly, these challenges can be considered as physical, psychological and social (Freidel et al., 2015). One of the key symptoms experienced by many patients with MS is fatigue (Motl et al., 2017).

Fatigue may be described as an extreme form of tiredness in which the lethargy has a pervading effect on the individual's ability to carry out activities of daily living (ADL) and impacts on their emotional wellbeing. Motl et al. (2017) refer to the term 'wellness' when considering a range of symptoms associated with fatigue. One of the key physical challenges with Sarah was nutrition since her weight loss impacted on her risk of developing a pressure ulcer. The state of knowledge regarding diet and MS is limited and no single type of diet has demonstrated any proven

effects on the prevention of symptom development (Lorenz et al., 2008). It is important in the palliative phase of Sarah's illness to refer her to the dietician and speech therapist for advice on the best diet, means of hydration and ways to promote swallowing (Motl et al., 2017).

In terms of nutrition, a major challenge for people with advanced MS like Sarah was swallowing. Dysphagia represents a safety issue due to problems of choking and often requires food to be purified or made into a soft small bolus for easier digestion. One of the key contributors to the multidisciplinary palliative care team is the speech and language therapist (SLT). The speech therapist and dietician played a key role in advising the district nurses about her diet; little and often, small amounts of food to be cooked by Eric, food made easier to swallow by the use of a blender, crème fraîche, ice cream, bananas, fruit and her favourite dish lasagne, followed by chocolate. The speech therapist recommended that if, and when, it became impossible for Sarah to swallow, a nasogastric tube or percutaneous endoscopic gastrostomy (PEG), tube feeding, could be used. The latter involves inserting a tube into the stomach and suturing the tube into the abdominal wall. Neither of these were acceptable to Sarah and while quite invasive, PEG tubes are often preferred to a nasogastric tube that is visible and very uncomfortable. In her advance care planning, Sarah stipulated that she was against artificial forms of feeding on the basis that she lost her dignity. An even more invasive form of feeding is parenteral feeding, where patients are fed with special dietary fluid directly into the vein, although this form of feeding is often not preferred by patients unless absolutely necessary as it limits their mobility. In all cases of feeding it is important that nurses ensure that the patient has good oral hygiene and where possible is encouraged to feed themselves even if it is small amounts of fluid or sucking ice cubes to refresh the mouth. There is no substitute for eating food that has been selected and preferred by the patient.

Spirituality is an area not touched on in the case study, partly because it was not an issue that came up between Sarah, Eric and the palliative care team. Spiritual care is often discussed in relation to end of life, although few nurses would claim to have a thorough knowledge of what spiritual care involves in a secular society (Cobb et al., 2012). Should the patient state a particular religious orientation, they may be offered the services of a member of the hospital/hospice spiritual care team. For a more in-depth discussion of spiritual care see Chapter 9.

SUPPORT FOR CAREGIVERS

Central to effective palliative care at the end of life is emotional and practical support for caregivers (Costello, 2017). In the case study Eric, Sarah's husband, received a range of support from the community nurses, the local MS Society group, through online contact, local groups, as well as friends and ex-work colleagues.

CASE STUDY 4.3 ERIC'S STORY (CASE STUDY 4.2 CONTINUED)

I was not terribly shocked by the news that Sarah's MS had become much worse. In many ways I had been anticipating this event for many years. Strangely enough, the support we received from the local MS group made us aware that this time would come. That does not mean to say that it was not a distressing and worrying time. At first I busied myself with making changes to the house to accommodate Sarah's loss of independence. Like many people with MS this is a big issue. I moved the bed downstairs and the community nurses arranged for a commode which she hated but put up with because I made sure I looked after it. I was surprised by the suddenness of the change, even though we had talked about it in the local support group. Somehow, you can never be fully prepared. Looking back, I was glad of having lots to do. I had been retired two years and I had my wood turning and my workshop which I used to retire to when I wanted time alone. On reflection, I feel as if I was given a lot of support and it was always available to me if I needed it. I was made aware that a lot of couples split up after a few years of living with MS. I suppose I was lucky in that I knew what I was getting into with Sarah as she was diagnosed before we married. I understood MS and we had a good life and we helped each other, it was not one way traffic, although towards the end I was having to do a lot. Most of the support I received came from the local support group. People would phone me up and we would chat together with Sarah on speaker. I had a lot of discussions online which was helpful. The nurses were good but I knew their time was limited so I always tried to let them know I was OK and they should get on with their work. Our biggest challenge (or mine) was during the night when I could hear Sarah groaning with the pain and she needed turning because she was getting sore. I was worried about her getting pressure sores and made it my mission to prevent this, although she did have small ones on each ankle and redness on her hips and bottom. This caused Sheba, who slept by her bed, to start barking and we ended up having tea and talking for a while after. Afterwards I found it hard to get back to sleep and the next day I was physically and mentally tired and would fall asleep and nap. Sarah tried not to disturb me and let me rest even when she needed me, like the time she had an accident because she let me sleep during the day. We argued about this type of thing. Eventually we got night sitters to come in, which was a great help as I could go out, have a couple of beers, get her ready for bed and I could get a good night's sleep. We were lucky in that we had a good team of nurses and others like the physio who helped Sarah at the end to get her breathing sorted. Sarah was the star. She was her usual thoughtful, kind and considerate self and although we had a plan for the end, it was not easy and I heard her shed a few tears alone. We both shared a lot of truths about the end but made sure not to dwell on it. The last week, I did not sleep much. We were both tired, her chest was bad but the morphine helped a lot and I was happy to give her sips of water to moisten her lips and do all the things the nurses did. It gave me something to do and helped me to feel wanted. I look back and consider myself lucky to have had such a great wife and I feel slightly proud that I contributed to what I call a 'good death', if there is such a thing.

CONCLUSION

This chapter has described the key symptoms and varied treatments used to improve the quality of life for people who have MS. Moreover, the chapter has

focused on the provision of palliative care for people with MS at intermittent stages during the illness and at the end of life. It has been acknowledged that the provision of quality palliative care takes into account the unpredictable nature of the condition. In general, most people with MS and other non-cancer conditions prefer to die at home, as the case study of Sarah showed. It is clear, however, that for many reasons, such as the vulnerability of the caregiver, this is not always possible. The case studies illustrate the complexity of multiple sclerosis and its diagnosis, and the role of palliative care before the onset of advanced disease and in providing end of life care. Moreover, the chapter situated the importance of the role of palliative care in meeting the individual needs of the patient and providing emotional support for the caregiver. Palliative care can be provided as short-term care or can encompass more long-term provision to incorporate end of life care.

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