Condition Insight Report (CIR)

Motor Neurone Disease (MND)

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Motor Neurone Disease Association

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Overview

What is the condition usually called / any abbreviations used?

Motor Neurone Disease (MND)

Brief overview of the condition

Motor neurone disease (MND) is a fatal, rapidly progressing disease that attacks the motor neurones, or nerves, in the brain and spinal cord; messages stop reaching the muscles. There is no cure and no single diagnostic test. Diagnosis is based on features in the clinical history and examination, usually accompanied by electrophysiological tests, blood tests, lumbar puncture and muscle biopsy. The purpose of these tests is to exclude other neurological conditions.

People with MND appear to fall into four groups in terms of cognitive change:

- 50% are unaffected by cognitive change
- 35% experience mild cognitive change, with specific deficits in executive functions, language and/or social cognition
- 15% develop frontotemporal dementia (FTD), either at the same time or after diagnosis of MND
- 15% of people diagnosed with FTD go on to develop MND

Watch **'what is MND'** video **HERE**

Watch **'Understanding MND' HERE**

What is the generally preferred term for someone with this condition?

A person living with MND

Presenting Symptoms

This is a summary of the common symptoms and effects of MND:

- Pain and discomfort, not caused directly by MND instead indirect causes such as stiff joints
- Muscle cramps and spasms (fasciculations and/or spasticity)
- Muscle wasting and weakness
- Incontinence, not usually associated with MND, but may occur if mobility is restricted and getting to the toilet becomes more difficult
- Bowel problems, not usually caused directly by MND, but constipation may occur due to restricted mobility and/or changes to diet - diarrhoea can sometimes happen with a severely constipated bowel
- Speech and communication issues e.g. dysarthria
- Eating and drinking difficulties may become an issue if swallowing is affected and tube feeding will be considered
- Saliva and mucus may cause issues if problems occur with swallowing - saliva may pool in the mouth or it may become thick and sticky
- Coughing and a feeling of choking may occur as a result of food or saliva becoming lodged in the airway
- Respiratory muscle weakness affects most people with MND
- Recurrent chest infections
- Orthopnoea
- Fatigue
- Associated Mental health conditions such as depression and anxiety
- Cognitive changes may occur for some people living with MND, where difficulties with memory, learning, language and poor concentration may be experienced - this is commonly known as cognitive change
- Disturbed sleep
- Changes can occur in the frontal and temporal areas of the brain, which affect thinking, reasoning and behaviour
- A small number of people will develop frontotemporal dementia

Fluctuations

There are no fluctuations. Once functionality has been lost it's permanent and there is no 'getting better' or recovery. Function degenerates over time, but people do often have worse days generally following over exertion. Individuals cope much better with a routine. If they have to do something out of the ordinary this can cause a huge amount of anxiety in advance, a very challenging day, and exhaustion for days after.

There is no single set pattern to the degeneration; it can be uncertain with the length of time with prognosis being unpredictable.

Think about exploring things like:

- **How has the condition been over the past 12 months?** What changes have they seen, over what period of time did they experience any deterioration, what has this meant for their functional ability now compared to previous?
- **What level of any pain/fatigue?** Can they quantify this, do they use a scale and can they describe it? Can they share what a ‘typical’ day might be for them? What are their triggers? Whilst exertion is a main trigger what amount of exertion is enough to cause further limitation? How are triggers managed?
- **For spasms** How frequent, where, for how often and what does this impact?
- **For any shortness of breath** Any recurrent chest infections, if so how frequent? What is breathing like at rest and on exertion, what is their recovery period like?

Reliability

What specific areas should be covered to ensure a complete, reflective report?

SAFETY	TIMELY	ACCEPTABLE STANDARD	REPEATEDLY
Do they have any symptoms which could cause a safety consideration?	For any activities where restriction is reported how long does it take them to complete these activities? Has how long it takes them changed over time?	How have they adapted to completing tasks over time – is this different to what might be considered ‘normal’?	Are they able to repeat a task as often as required? Is this the same every day?
Due to the varied symptoms which may impact safety, this is a critical area which needs to be explored. Cover how any risks are mitigated and if there is a history of incidents within activities 1, 2, 3, 4, 5, 9, 11 and 12 to ensure you have addressed the needs of the individual claimant. Safety considerations can be a result from physical and psychological symptoms.	Freezing, pain, fatigue, stiffness, spasms can all affect time frames of how long it takes an individual to complete a task. This will vary for each individual. Remember that there are no specific time frames outlined for what is reasonable. **Ensure to ask:** • How long it takes to complete tasks such as 1,4,5,6,7 and 12. • Why they feel it takes them this long? • Where they report having any breaks in task, how long are these breaks and how frequent?	How they are adapting is important and needs in depth, targeted questioning. Exploring **<u>HOW</u>** they complete the task to draw out any compensation strategies which can then be explored further. E.g. using a sink to help get on/off the toilet. Remember that not everyone is comfortable talking about their restrictions. Be sensitive and explore both their physical and psychological symptom impact.	Due to the degeneration that occurs over time and the variation between days individuals can experience this needs to be explored for all relevant activities. You should try to explore how someone may be before, during and after activities where a restriction is present to determine fully whether the task is repeatable in the same way as often as required especially with elements like fatigue affecting their presentation.

Sensitivities

What areas might they find difficult to mention or perhaps understate the impact of?

Individuals may find all aspects of their condition difficult to discuss, but in particular:

- An MND diagnosis is always a diagnosis of a terminal condition
- All treatment is palliative
- Riluzole is the only licensed drug available that may extend life in MND and trials have shown it can extend survival by around three to six months if taken for 18 months

Watch people talk about their lived experience of MND:

Gemma

Vivienne

Lucy

Customer Care

How is it best to ask about any sensitive topics and what are the common courtesies?

In general

- Please talk with them directly, don’t ignore them
- Be sensitive due to the terminal diagnosis and be considerate in your questioning
- Sensitively explore further to ensure they are not overlooking something e.g. *“can you talk me through how you do that?”*
- Give them plenty of time to reply to questions
- Do not rush an answer or attempt to finish their sentences
- Make use of open questions to allow them to bring out the information they want to, however consider cognitive impairments and communication aids which may require you to ask more simple questions with closed responses
- Include any companion present in the assessment and recognise cognitive changes may be present

During face to face interactions

- Ask if they require assistance
- Do not expect them to walk long distances
- If given a drink, it should only be half full at most
- Ensure you are close enough to hear a response so you do not need to ask them to repeat comments unnecessarily however don't be afraid to ask them to repeat if their voice is indistinct, slurred or faint, as it may be impaired by MND

Functional Impact

A brief summary of the functional impact those living with this condition may experience

Activity 1: Preparing food

Pain, muscle wasting, stiff joints, spasms, fatigue. This can affect their ability to stand, prepare fresh ingredients and cook. Symptoms such as flail arm where they have no use of their arms can cause great difficulty and require them to need full support.

There may also be cognitive effects and associated mental health conditions from the condition which mean individuals need full support.

Remember in PIP…

Can someone prepare a meal reliably? Someone's physical ability to complete an activity is not the only area to be considered here. If there is evidence of cognitive deficit, are they safe cooking? Can they tell if food is cooked? Are they safe around knives and flames? Furthermore, if there is a lack of motivation, do they require prompting to cook? Are they cooking the majority of days? Do they require support or supervision? If there are issues with motor skills, can they safely cut food or hold pans? Do they require physical assistance?

Activity 2: Taking nutrition

Loss of the ability to speak and swallow food or drink. There is likely to be a period where softened or blended foods can help, but require specific preparation. Commonly people receive fluid, nutrition and medication through gastrostomy or nasogastric tube (nasogastric less common). Advice is often given to undergo gastrostomy surgery while respiratory function is still strong enough, even if tube feeding is not yet needed as it will almost certainly be necessary as the disease progresses. Others complain more of thickened mucus in the mouth and throat, which is difficult to swallow, or phlegm in the airways, which is difficult to cough up due to weakened respiratory muscles and an ineffective cough.

Remember in PIP…

You need to have established the individual’s ability to be nourished, either by cutting food into pieces, conveying it to the mouth and chewing and swallowing; or through the use of therapeutic sources. Are they at risk of choking? Do they require soft or blended food? If there is weight loss secondary to their condition, do they require prompting to eat regularly? Is this prompting throughout the duration of the meal?

Activity 3: Managing therapy and monitoring a health condition

Non-invasive ventilation (NIV) is a method of providing ventilator support to a person with MND using a close-fitting mask that may cover the nose and/or mouth. The ventilator detects when the person tries to take a breath and delivers an extra flow of air to increase the volume inhaled.

The MND Association “Just in Case” kit is a small box designed to hold medication that may be needed if a person with MND experiences a sudden change with symptoms such as breathlessness, coughing or choking.

A mechanical insufflation-exsufflation (MI:E) device is often used to support and enhance the ability of weakened inspiratory and expiratory muscles in order to improve the cough and aid secretion clearance and can help to prevent respiratory infections.

Remember in PIP...

If someone lacks insight, you must establish how they are managing their treatment. Do they require prompting or support to administer their medication? Do they have poor compliance? Furthermore, if they receive therapy in the home environment, do they require assistance with this? How long does this assistance take?

Functional Impact

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Activity 4: Washing and Bathing

Associated mental health conditions, affecting thinking, reasoning, changes in behaviour, changes in respiratory function and muscle wasting can hugely impact this activity where some may require prompting and others may need full assistance depending on the severity. Some lose trunk control and have poor balance and unable to support themselves to reach out of their base of support to complete the activity whereas others may physically still be able to do so but limited by fatigue.

Remember in PIP…

You must not only consider physical ability to complete the activity, but also recognition of the requirement to wash and motivation. Do they require prompting to wash regularly? In the absence of prompting, how often would they wash? If they have a physical restriction, how are they managing to wash their entire body? Can they physically get in and out of an unadapted bath? How long can they stand for? Do they require assistance to wash their body? If so, is this their entire body? How long would it take them to wash unassisted? Would this be acceptable when considering the extent of their symptoms?.

Activity 5: Managing toileting needs and incontinence

Bowel problems and incontinence can be present. Whilst pain, cramps, stiff joints and fatigue can all impact someone's ability to get on/off and manage their hygiene needs. Lack of mobility can also cause constipation. Changes to diet - e.g. blended foods or tube feeding, can also cause changes in bowel habits.

Remember in PIP…

You must explore not only someone’s ability to get on and off the toilet, but also to reliably clean themselves. This will be a sensitive area to explore, so should be probed delicately. Can they reliably clean themselves after going to the toilet? If they have aids, can they reliably use them or do they require assistance? If they report incontinence, can they self manage? Is this of both bladder and bowel?

Activity 6: Dressing and undressing

Associated mental health conditions, affecting thinking, reasoning, changes in behaviour, changes in respiratory function and muscle wasting can hugely impact this activity where some may require prompting and others may need full assistance depending on the severity. Some lose grip, movement in their arms, trunk control and have poor balance and unable to support themselves to reach out of their base of support to complete the activity. Others may physically still be able to do some tasks but initially are limited by fatigue. Many have to learn to adapt and then accept dependence as the condition progresses, which can lead to frustration and distress.

Remember in PIP…

How do they manage to dress themselves? Are they capable of selecting appropriate clothing? Do they require prompting to do this? If they can dress, how long does it take? Can they manage fastenings, buttons and laces? How do they feel after? Do they require assistance or use any aids to dress?

Functional Impact

A brief summary of the functional impact those living with this condition may experience

Activity 7: Communicating Verbally

80% of people with MND experience slurred, quiet or complete loss of speech because of spasticity and weakness of the tongue, lips, facial muscles, pharynx and larynx. The term AAC covers a wide range of techniques and communication aids that support or replace spoken and/or written communication. These may be used to communicate face-to-face or electronically. AAC is commonly used to refer to electronic communication aids and products (high-tech) but equally it includes simple, non-electronic (low-tech) options such as pen and paper, gesture, signing, symbols, communication charts, and letter, word or picture boards or books. Gesture and expression can also be affected, making communication of any kind more and more difficult over time. This is where there is a real potential of people with MND becoming 'locked in' and specialist help may be required to ensure their needs and wishes are considered. This may require a specialist advocate.

Remember in PIP...

The scope of the activity is to be able to both express **and** understand. It is not enough that someone can hear what another person is saying. Can they process it and respond appropriately and in a timely manner? How do they manage with communicating with others? Do they require support to do so?

Activity 8: Reading and understanding signs and symbols

With any cognitive changes, this can affect the ability to process and understand written information and in some cases support is required.

Remember in PIP…

Simply being able to read information is not adequate to manage this activity. The information someone is reading must be **understood**. What level of information can they understand? Although retention of information is not considered, it is clinically reasonable that someone will have difficulties processing, and this should absolutely be explored during the assessment.

Activity 9: Engaging with others face to face

Dysarthria can have a significant impact on emotional wellbeing because it can cause isolation, loss of intimacy, loss of identity, frustration, fear and anxiety, low self-esteem, a loss of control and increased sadness and depression. Can make it difficult for individuals to engage with others face to face.

Remember in PIP…

If there are cognitive changes reported, you must explore their ability to safely engage with others. Are they vulnerable? Are they at risk when engaging? Do they require support? If they have anxiety, who can they engage with regularly? How do they manage engaging with unfamiliar people? Do they require support from a specific individual to do so? **REMEMBER: Just because someone reports they can mix with others, this does not mean they are doing so safely.**

Functional Impact

A brief summary of the functional impact those living with this condition may experience

Activity 10: Budgeting

As any cognitive functions are affected or an individual experiences anxiety or depression this can also result in assistance being needed for this task.

Remember in PIP…

If someone has a cognitive deficit it may be that they are unable to prioritise finances. Could they reliably manage a household budget? Do they require support? How would they cope with an unexpected bill? Can they manage future financial planning? Also, could they understand change expected in a shop?

Lack of motivation and anxiety alongside impulsivity must also be considered here.

Activity 11: Planning and following a journey

Changes can occur in the frontal and temporal areas of the brain, which affect thinking, reasoning and behaviour. This may also make them impulsive, disinhibited, perseverative or compulsive in behaviour. Problems with executive functioning may lead to problems with:

- Setting and achieving goals
- Planning and problem solving
- Responding to new situations
- Shifting attention or dividing attention between different tasks
- Initiating and inhibiting responses

Remember in PIP…

You must explore safety in this activity. If someone is completing journeys, are they safe? Do they have road safety? Also could they maintain concentration when out of the home? Are they vulnerable? Could they manage a diversion or plan and follow a new route? How would they cope on public transport? What would they do if they got lost? Have there been any incidents when out of the home?

Activity 12: Moving Around

Muscle wasting and fatigue can cause falls, reduce distance that can be walked, increase timings and will affect the repeatability of ability to move around. Many use wheelchairs as their source of moving around, due to weakness it is likely these are attendant push or electric in most cases to support movement. Many will need to be fully supported with trunk control support and head rests.

Remember in PIP...

Firstly, establish when someone is walking, do they require an aid? If so, what aid? Was this prescribed or self purchased? How often are they having falls? How do they manage this? What is their gait?
How far can they walk and how long does this take? How does walking make them feel? Does pain/fatigue increase each time they walk? When they walk a second time, does pace reduce or distance reduce? If they experience fatigue, how do they feel later in the day? How many times could they repeat the distance they walk? How long does it take to recover from mobilising?
Lived examples are also extremely beneficial. How do they manage up and down the stairs? How do they manage with chores? How do they manage their shopping? How many aisles can they walk and how long does it take? If they go to their GP surgery or hospital, how far is it from the car? How long does it take them to get to the required department and how far is this?

Additional reading or other resources

EXTERNAL

- For further information see NHS Conditions and Treatments: https://www.nhs.uk/conditions/motor-neurone-disease/
- www.mndassociation.org/careinfo
- www.mndassociation.org/professionals/publications

INTERNAL

- Desktop Aid – CSE, Activity 12, Activity 6, Fatigue, Breathlessness
- **Motor Neurone Disease Video which is a presentation given by the MP for Bridgend and a representative from the Motor Neurone Disease Association (MNDA) in June 2018.**

Version control