YPI Information Pack

Here are some Bullet Points you might want to use to help make a presentation.
(If you use them all your presentation will take more than an hour, so be selective. As ALS is the commonest form of MND you might want to use only ALS information.)

MND is an umbrella term for several neurological conditions

- Amyotrophic Lateral Sclerosis **(ALS**) affects about 9/10 people with MND
    - Progressive Bulbar Palsy (**PBP**) also called Bulbar onset MND is a subtype of ALS
- Progressive Muscular Atrophy(**PMA**) affects about 9/100 people with MND
- Primary Lateral Sclerosis (**PLS**) affects less than 2/100 people with MND

These conditions are differentiated by which type of motor neurone is affected.

- If it affects mainly the motor nerves coming from the brain to the spinal cord (these are called Upper Motor Neurones) - that causes **Primary Lateral Sclerosis**.

- If it affects mainly the motor nerves coming from the spinal cord to the muscles (these are called Lower Motor Neurones) - that causes **Progressive Muscular Atrophy**

- When both kinds of motor nerve are affected - that causes **Amyotrophic Lateral Sclerosis. Progressive Bulbar Palsy** mainly affects the muscles of the face, throat and tongue.

Progressive Muscular Atrophy (PMA)

Progressive disease involving lower motor neurones causes

- Increasing weakness
- Fading reflexes
- Muscle wasting
- Life expectancy ~5+ years from 1st symptoms
-

Primary Lateral Sclerosis (PLS) is a progressive disease of mainly the upper motor neurones

- Increased muscle tone
- Brisk reflexes
- Spasticity/contractures
- Life expectancy ~ 20 + years from 1st symptoms

Amyotrophic Lateral Sclerosis (ALS) is a progressive disease involving both upper and lower motor neurones.

- Bulbar (speech and swallowing) onset ~25% of cases
- Limb or Trunk onset ~65% of cases

ALS has a mixture of upper and lower MN signs
Life expectancy from 1st symptoms - 6 months to 5 years for most people

Progressive Bulbar Palsy
Early symptoms may include slurring of speech or difficulty swallowing.
Life expectancy is between six months and three years from onset of symptoms.

MND is….

- incurable and life-shortening
- always progressive (only ever gets worse, there is no recovery)
- neither contagious nor infectious
- inherited in about 5 to 10% of cases (Familial ALS,)
- Has no family history in 90 to 95% of cases (Sporadic,) and
- Can strike in any decade of life,
- Mostly affects people older than 40, 10% under the age of 40 our youngest ever patient was 14 and died shortly before she was 17
- Average age at diagnosis in Scotland is ~67,
- Affects more men than women (3 men : 2 women)
- About 200 people diagnosed each year in Scotland,
- About 400 people have the condition in Scotland to date

Diagnosis
There is no single test that diagnoses MND, so other possible causes for the symptoms need to be excluded.
Tests to exclude other diseases, blood tests (Lyme Disease), neurophysiology, nerve conduction studies (speed of nerves electrical activity), electromyography (EMG records electrical activity of the skeletal muscles), MRI scans to rule out tumours or trapped nerves/slipped discs.

Scottish Statistics

Incidence ~ 4 cases diagnosed each year for every 100,000 people
Prevalence ~ just over 8 people living with the condition for every 100,000 people (What is Scotland's population? How many people live in your area, so how many would you expect to have MND?)

Presentation
- ~ 25% their first symptom is speech problems
- ~ 33% arm or hand problems
- ~ 33% leg or foot problems
- ~ 4% breathing problems

48% die within a year of diagnosis
<5% will live longer than 5 years after diagnosis
Average life expectancy after diagnosis ~18 months
Lifetime risk = 1 person in 350 will develop MND

MND - The Effects

Motor Neurones degenerate and die.
Gradual loss of muscle control, increasing weakness and paralysis.
Common problems include foot-drop, poor grip, head-drop, limited range of movement, breathing, speech problems.
Loss of body mass due to inactivity, malnutrition, ALS causes a higher metabolic rate.
Fasciculations (What are these?- explain)
Rate of progression, which muscles are affected and in which order varies between individuals.
Sensory neurones are unaffected so touch, sight, smell, taste and hearing remain intact.
Sexual function and bowel and bladder muscles are usually not affected.
Emotions and intellect are unaffected for most people most of the time (See later about FTD)

Common Problems

Muscle wastage–

- Causes lack of support for joints, lack of padding/ protection for bones – pain Wastage made worse by increased metabolic rate
- weakness leading to immobility - pain
- poor return circulation – cramps (What causes these?)
- poor lymphatic drainage – tissues retain fluid

Spasticity/Contractures due to muscle contractions

Eating drinking and swallowing - there can be decreased intake of food or fluids at any time
Loss of swallowing reflexes leads to choking – this can cause aspiration pneumonia, people might also suffer malnutrition or dehydration because they fear choking,
Some people can also have excessive saliva, (thin & runny or thick & tenacious) which is a problem if they can't swallow
Breathing problems can come from a weakened diaphragm this causes

- Respiratory infections
- Interrupted sleep due to low O2 (Initially normal levels in the blood while waking)

In turn this causes

- Difficult to waken
- Daytime sleepiness
- Confusion, difficulty concentrating, memory lapses
- Morning headache

Non-Invasive Positive Pressure Ventilation can help with breathing problems, but the mask has to be tight to the face and this can cause

- Pressure sores, which can lead to Ulceration, which can cause Sepsis/Septicaemia

Communication problems (affects up to about 75% overall)

- Dysarthria (i.e. slurring of words)
- Hyper-nasal voice
- Weak voice
- Anarthria (loss of voice)

Toileting

- Problems can come from reduced activity level, reduced fibre intake, reduced fluid intake or medication side effects. Inability to "push" with abdominal muscles can cause problems too.

Pain

- Bone pain, cramps, inability to change position

Anxiety (If you had this disease, what things would you be anxious about?)

Cognitive Change

Some of the genes that cause familial ALS can also cause changes to how people think and behave.
Up to 15% of people with ALS can also have Fronto-temporal dementia. Some people with FTD will go in to develop MND.
About 50% of people with ALS can also have changes to their behaviour or thinking.
Behavioural changes include apathy, impulsive behaviour, acting strangely.
Cognitive problems cause language problems such as finding the right words, problems planning and organizing tasks.
Emotional Lability is common - laughing or crying at inappropriate times eg laughing during a funeral.

End of Life

Sudden unpredictable causes at almost any point
17% die very suddenly probably due to Cardiac arrest.
Others die of:-
Respiratory infections
Respiratory insufficiency (not able to get rid of CO2, which causes the person to enter a coma.

Other sources of Information

YouTube has lots of videos about MND and ALS in particular. You might find some of these useful if you want to learn about the condition:-

www.youtube.com/watch?v=8AlUBSeo_Z8&list=RD8AlUBSeo_Z8#t=4 is a collection of 50 videos

www.youtube.com/watch?v=LwypwQxlXhQ from 3 minutes after the start

www.youtube.com/watch?v=StS4Rj_8ty8 American

www.youtube.com/watch?v=8AlUBSeo_Z8