

# What happens as MND progresses?

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# What Happens as MND Progresses?

- What changes have you encountered or what might you anticipate?



- [05 06 Case Study Motor Neurone Disease](#)
- The aim of this session is to help you identify the main clinical and practical issues that patients with motor neurone disease face, so that you can better support them and their families in whatever setting you are working.

# What can affect discussions and decision making?

- Communication impairment
- Cognitive impairment
- Fatigue
- Preference to avoid
  
- Our reluctance

# Physical changes

- Limbs
- Trunk
- Breathing
- Swallowing
- Speaking

# Cognitive changes

## **Some cognitive changes that MND can cause**

- Changes in personality
- Difficulty processing new information
- Restriction of verbal fluency
- Difficulty solving problems
- Difficulty doing two things at once

# Communication difficulties

MND can affect communication in a number of ways through bulbar muscle impairment. Conversations may take a considerable amount of time for even just a few sentences

- Increased muscle tone affects the vocal cords leading to a high-pitched voice.
- Slow tongue movements means vocalising words is slowed down and speech is hard to interpret.
- Weak facial muscles mean that expressions are more difficult to project and lip reading is less easy to understand
- The use of assisted communication devices to overcome these problems can also be limited, due to the loss of function or limited function in the patient's arms.



# @intheenditwasok

## Joe Hammond

Writing as I say goodbye  
to my body and those I



**The  
Guardian**

- [Head Hoovering](#)

## Questions and answers

- How did you choose my name?
- What advice do you have for dating and going through puberty?
- Do you have any favourite sayings/recipes?
- How do you handle anger?
- What is your favourite after-shave/perfume?
- Can you speak about your favourite books?
- What was my birth like?
- What advice can you give me to help me grieve for you?
- What do you wish for me?

# Dying from MND

*European Journal of Neurology* 2008, **15**: 1245–1251

doi:10.1111/j.1468-1331.2008.02307.x

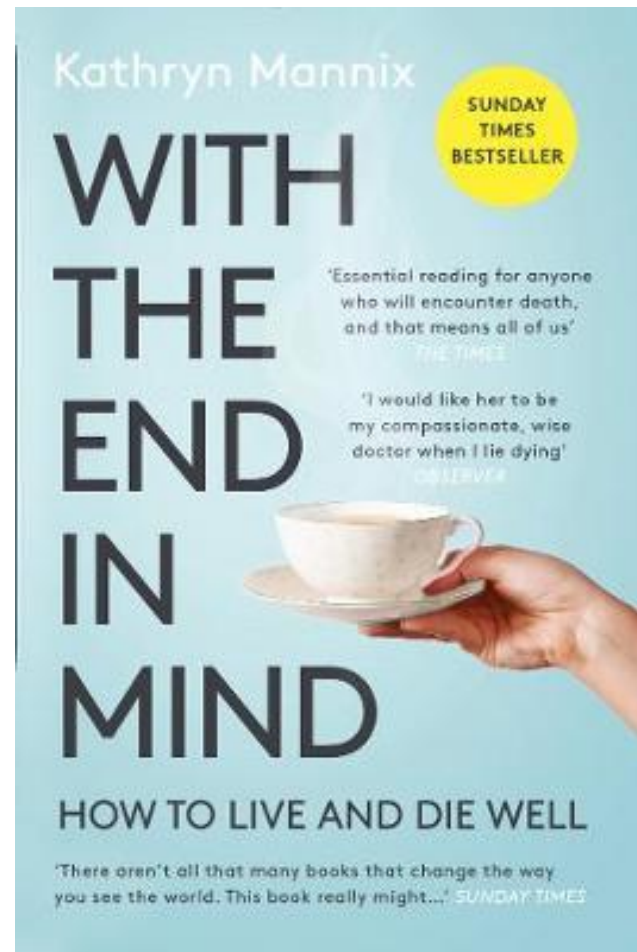
Causes of death amongst French patients with amyotrophic lateral sclerosis:  
a prospective study

- Sudden and unexpected
- Infection 14%
- Respiratory failure 58%
- PE 2%
- Foreign body 3%
- Other causes 10%
- Undertermined 13%
- 302 patients 2006-7
- 16 ALS centres
- 63% died in medical facility
- 33% NIV
- 3% TV
- 37% gastrostomy

# @drkathrynmannix

"I have come to appreciate that patience, compassion and skill must go hand in hand with honesty and realism.

When chances of survival are waning, the hope for dying well is hugely important."



This is **Amanda**. She is 31 years old and married to Peter. They have one son, Mark, aged 18 months.

In the summer, Amanda began to feel that her arms were getting heavy and felt that lifting Mark was becoming more difficult.

She noticed twitching in her muscles that became more pronounced over time. During this time Amanda also started complaining of cramp in both legs, especially at night.

By the onset of winter, Amanda was no longer able to lift her arms up and had no functional use of either hand. Following five months of tests, she was finally diagnosed with MND.



Amanda chooses to have a PEG fitted.

Initially, she only uses this for additional nutrition at night and still eats for pleasure during the day. However, over the coming months she begins to use the PEG more as she finds eating becoming more difficult and tiring.

During this time, Amanda also starts complaining of occasional episodes of **coughing** and **choking** when she is drinking. She tells the nurse that she's very frightened about these episodes and that she's terrified of choking to death.

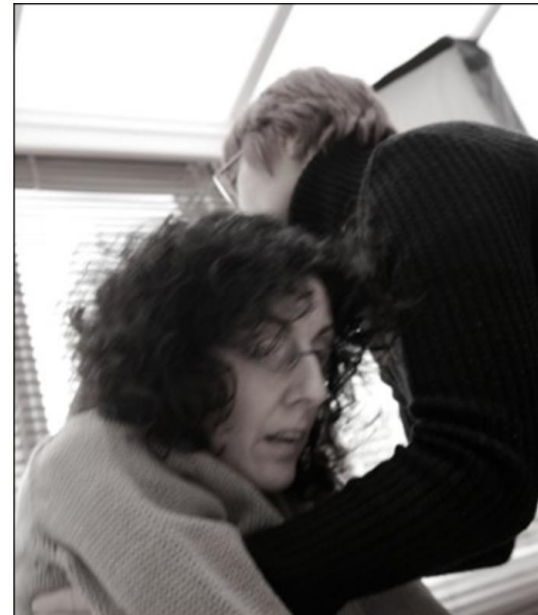
Is she right to be scared? Do you think people with MND choke to death?

Select one option from the answers below, then submit.

- A. Yes
- B. No

Submit

Image



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Is she right to be scared? Do you think people with MND choke to death?

Select one option from the answers below, then submit.

- A. Yes  ✓
- B. No

Submitted

Feedback

**Incorrect.**

Retry

**A. Correct.**

**B. Incorrect.**

The popular press often portray that choking is the way that people with MND die and this mostly is not true. The majority of people with MND die very peaceful deaths. There is, however, a risk of choking if someone can not co-ordinate their swallowing and very occasionally a patient has died from a blockage of the air passages.

This blockage would require a solid piece of food or other object of a size of >1 cm. Although inhaling (aspirating) liquids can cause enormous irritation to the airways it will not cause death by choking as air can still be taken in to the lungs. Aspiration of food and perhaps saliva can lead to infection and pneumonia.

Image

A few weeks later, Amanda complains of feeling breathless during the day. She reports that she has taken to sleeping in her armchair as it's easier than lying down.

Which of the following statements about breathlessness (dyspnoea) in MND are correct?

Select true or false for each of the following statements, then submit.

	True	False
A. It is a prominent symptom for many patients in the last few months of their life	<input type="radio"/>	<input type="radio"/>
B. The problem lies in the mechanics of breathing rather than a change in the physiology of the lungs	<input type="radio"/>	<input type="radio"/>
C. It occurs because the diaphragm is innervated by motor nerves, which are susceptible to weakness and paralysis	<input type="radio"/>	<input type="radio"/>
D. Breathing becomes less effective when lying flat because the diaphragm is too weak to move against the abdominal wall	<input type="radio"/>	<input type="radio"/>
E. Respiratory weakness is the cause of death in most people with MND	<input type="radio"/>	<input type="radio"/>

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Incorrect.

Retry

A. True

B. True.

C. True.

D. True.

E. True.

Dyspnoea should be a trigger for thinking about referral to specialist palliative care services if the patient is not already known to them.

Breathing becomes less effective when lying down to sleep, resulting in a rise in carbon dioxide (CO<sub>2</sub>) levels.

This causes a number of symptoms which you will look at later in the session.

Image



What symptoms related to her dyspnoea might Amanda complain of?

Select [here](#) to find out what Amanda says to the nurse about her symptoms.



What symptoms related to her dyspnoea might Amanda complain of?

**Amanda:**

“ I get a feeling of panic when I'm lying down, like I can't breathe properly.

I've started having these terrible nightmares, and sometimes I wake up desperately needing the toilet.

I get these bad headaches in the morning. Thankfully they usually go away about an hour after getting up.

I get so tired during the day. Sometimes I even fall asleep during a conversation. It's embarrassing! And I find it so hard to concentrate sometimes. It's difficult to take stuff in. It's like my mind has gone all fuzzy.

My appetite has definitely got worse. I don't feel so hungry these days.

”



Let's look at Amanda's symptoms in more detail.

Select each of the speech bubbles to find out more.

A feeling of panic when lying down	Terrible nightmares
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Finding it hard to concentrate	Decreased appetite
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Waking up needing to urinate	Extreme fatigue during the day
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Morning headaches
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# The last part of life and dying

It is now 12 months since Amanda received her diagnosis.

She is unable to move, her speech is unintelligible and she can only communicate via eye blink responses of 'yes' and 'no' to closed questions.

She can no longer eat or drink, and receives full enteral feeding via a PEG.

Over the last three weeks, Amanda has become more drowsy during the day - despite using non-invasive ventilation (NIV) - and she has been complaining of feeling breathless. She has become increasingly distressed.

Amanda has not felt able to discuss much about this stage of her illness other than saying she wants to remain at home where she can see and hear her son playing in the background.

