

DAVID PEACE and MND. STATE OF PLAY as of Saturday 13 March 2021

Brief background:

Early 2019 some strange feelings in the mouth, and some slurring in speech. Tests. July 2019 diagnosed with MND/ALS. More tests. October 2019 MND/ALS confirmed. It's terminal – no treatment, no cure. It will spread at its own pace and so far no one can stop it. For ease I'll use 'MND' from now on..

MND can start in many parts of the body, often manifesting itself when someone starts dropping things, or when a foot doesn't behave properly. In my case it started in the mouth and throat. I have MND 'bulbar onset'. Bulbar: "The brain stem (also known as the bulbar region) controls the muscles needed for swallowing, speaking, chewing, and other functions"

AS OF TODAY:

The mouth has mostly stopped functioning. The lips have no power and they can't close without strong jaw effort, which doesn't last. As a result they can't prevent anything that is in the mouth coming out, and the mouth is open all of the time. The tongue is completely paralysed: it can't direct food or liquid down the throat, so anything in the mouth might go the wrong way and could block the windpipe – potentially fatal. As the mouth is mostly open the tongue is completely dry. The muscles used for swallowing have 99% stopped working. Any attempt to swallow risks choking. With lips, tongue and other muscles permanently unemployed, I can't speak other than a few guttural sounds. Smiling is difficult. Due to the weakness of the muscles the face is more or less fixed in a type of grimace.

Saliva and mouth 'secretions': the most exhausting part of my daily life. Most people produce between 1 and 1.5 litres of saliva each day, and the tongue and other muscles direct the liquid round the mouth, lubricating every part, and then back, tidily, down the throat. Mine don't. The only way saliva can leave the mouth is out through my helpless lips. I carry towelling napkins everywhere to mop up the endless dribbling. It's continual work and it's truly infuriating. To try to reduce it I've had Botox injections into the salivary glands, I syringe a medicine into my stomach three times a day, and every three days I change a patch behind an ear. They probably do have an effect but the problem is still intense and it's very wearing. Despite the endless flow of liquid the tongue is often completely dry. Mucus gathers in the mouth, and with luck it can be removed manually. Mucus down in the throat is impossible to remove at the moment, and that is the most danger.

Eating and Drinking. In November, after two earlier operations each of which was abandoned in the process, a PEG tube was fitted into my stomach. Now, five times a day I syringe in combinations of two special nutritional liquids (1.32 litres total p.d.) and one litre of water, and something to thin the secretions, and a laxative, and the one medicine authorised in Britain for MND, which is believed to extend life for up to 18 months. The 'feeding' through the pipe doesn't hurt any more. It's tedious but it keeps me going. The NHS provides all my nutrition. At any one time there are over two hundred 400cl bottles of the liquids under the bed. To my surprise, having lost 15 kilos in the 15 months between diagnosis and PEG, I seem to have gained 3 kilos in the 3 months since the PEG.

The head. Whatever muscles keep one's head up have almost stopped working. When standing, the head drops down, chin on chest. It takes an effort to raise it. With effort I can keep it up for about a minute. If I'm standing or walking outside I wear an uncomfortable neck brace under the chin. Then I have to be careful in a different way. When you speak you normally keep your head still and move the jaw up and down. With the brace it's the opposite: the jaw is fixed. Any movement then jerks the whole head up and down, and it can be difficult to stop the teeth banging together.

Mobility: OK inside my flat, though when standing or walking the head is slumped. Outside I carry a walking stick, trying not to use it much, and if it's more than, say, five minutes I have to be accompanied and can then take a break in the wheelchair.

Walking is more like tottering. Needs care and concentration. I'm not sure if it's a balance thing or if the muscles are weakening. The walking stick is useful for stability when needed.

Left arm and hand: both are weakening. The left hand can reach to the top of my head but not much more. I need to concentrate on simple things like getting dressed and undressed, squeezing a toothpaste tube and nail trimming. With clippers in the right hand I can trim the nails on the left hand, but not vice versa: the left hand doesn't have enough strength to press the clippers.

Tiredness: all the time, I'm afraid. It doesn't seem to depend on sleep pattern. Always tired and often out of breath even when walking around the flat.

Sleeping: can be difficult because of the coughing but I generally get around five or six hours. I've found that sitting upright in bed, while not very comfortable, reduces the coughing, so I often spend the night in that position.

The brain is working more or less normally I think, but very short term memory is suffering. I'm forgetting many recent things, and that has happened quickly, too quickly for it to be just 'ageing'.

The 'pseudo-bulbar affect': note *affect*, not *effect*. A phrase, a look, a moment of sadness, certain pieces of music, reduce me to tears and I can't stop crying. Conversely, something humorous can get me laughing and I can't stop that either. Laughter can turn to tears, although that's happened to me only once. It's infuriating. In the past, tears were almost non-existent. Like many people I'd feel sad but it wouldn't lead to sobbing. Makes me angry but I just can't control it. And continual laughter, well beyond former limits, is maddening. Both are by-products of MND and other illnesses. It's one of the aspects of 'emotional lability'.

Emotions and Morale: I feel that morale is not too bad. Emotionally I adjusted to the diagnosis quite quickly; I've accepted it and have prepared for death (under my control I desperately hope). The things described above make life extremely difficult day by day, and there's serious discomfort especially in the mouth and in trying to communicate, but there isn't any actual pain. If there were pain I think morale would be much lower than it is.

The NHS and the Motor Neurone Disease Association (MNDA) have supplied me with two support bars in the shower; a bidet attachment to the toilet; a remote door opening mechanism to let people into the flat; a suction machine to remove mouth 'secretions'; a nebuliser machine to improve breathing; a wrist-worn security button to get emergency help; an oximeter to detect low blood pressure and oxygen levels; a walking stick; a neck brace; a wheelchair; all nutrition; medicines; specially supported cushions; and more. After a night in hospital next week they might give me a 'cough-assist' machine to strengthen the cough so that I can clear the mucus from the throat.

My local NHS team includes voice therapist, physiotherapist, occupational therapist, dentist, podiatrist and dietitian, all of whom visit. The GP practice and pharmacy are less than five minutes walk away, and during the pandemic the pharmacy has delivered everything to the door of my third floor flat. The MND specialists in the National Hospital for Neurology and Neurosurgery are regularly in touch. The company that supplies the nutritional liquid also provides a nurse to check on the PEG feeding and equipment maintenance.

Next. What comes next depends entirely on how the MND moves through the body, and how quickly. No one can tell. If it moves to the next set of muscles, that could be the lungs. Eventually they would need invasive ventilation – a permanent pipe into the throat – which I will refuse. We shall see.

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