NOTES ON MND/ALS, BULBAR VERSION
by David Peace
as of 18 April 2020

Time-line
In January 2019 I sensed two things: some words were being slurred, and I found that food was sticking to the roof of my mouth. On 17 April an ENT (ear, nose throat) specialist said it might be a neurological problem. More tests: muscles, electric response, MRI of the spine, lumbar puncture. On 4 July the diagnosis – motor neurone disease, starting in the bulbar area, i.e. the mouth and throat. The diagnosis was re-confirmed by another specialist at the end of October. Today is Saturday 18 April 2020.

My situation – whole body: I'm walking the parks and doing my own shopping; I'm starting to play the piano again after a few decades of neglect; I live alone in a flat almost 50 steps up from the street and am managing that without much difficulty; I tinker in my little in my roof garden; and I email and write a blog. 15 months on from my original concerns the main area of difficulty is still the mouth and throat. As expected they have considerably worsened.

If you know someone with MND it's likely that their illness started elsewhere, perhaps in a foot or leg or arm. If so what I will describe is probably what will happen when it finally spreads up to the bulbar area. If, like me, their MND started as a bulbar problem then this might be familiar.

The muscles

<u>The lips</u>
The lips are weak and I can't control them. Food, liquid and saliva can't be held in easily. Anything in the mouth can slide out without me knowing until it's too late. For that reason I have stopped eating out in restaurants, cafés, pubs and so on. When eating at home I wear an apron and have at hand a tea towel and a kitchen roll to catch anything. With care I can drink, but it's easy for thin liquids to trigger a cough or a bout of choking. Clearing the throat is vital but it can take a few minutes of very noisy, donkey-braying activity.

<u>The tongue</u> is weak and now has very limited movement. A year ago I could put it in either cheek but now I can't. Last year I had thought that food was sticking to the roof of my mouth, but in fact it was the tongue losing its power: one of its roles is to move food to the sides for it to be chewed, and it wasn't and isn't doing that. Chewing is vital because the throat is very sensitive: if anything solid touches it I choke, or I manage to cough it out. The danger is that it will go down the windpipe and into the lungs, resulting in pneumonia. All food must therefore be very well-chewed. This is not easy because the movement of the jaw is also impaired. I'm currently managing though.

Another problem is that the tongue normally clears most bits of food from the teeth. That doesn't happen anymore. After eating there's a lot wedged there so I'm always in the bathroom brushing and flossing.

Speaking suffers badly from the lack of movement of tongue and lips, and the interference of saliva.

Saliva
In the last few months the amount of saliva has doubled or tripled – very large quantities, and therefore embarrassing when it can't be held in. Dribbling and drooling, regrettably, and it can interfere badly with speaking. A handkerchief, cloth or paper towel should be at hand.

The nose
This is odd: I can't clear it properly. Blowing the nose doesn't work well - the muscles are weak and it sounds like air being released from a balloon!

The jaw
The jaw seems to be acting more slowly. I have to chew everything into tiny quantities but it takes a lot of effort and a lot more time than in the past.

The lungs
This is puzzling. I live near Hyde Park, Kensington Gardens and Regent's Park and can walk for a few miles there without feeling particularly tired. I get into a rhythm, walk at a steady semi-fast pace and, being 6'2"/1.88m tall, overtake many others. What does make me breathless? Filling or emptying the washing machine, or trying to talk as I walk. Very odd.

The voice
It's weak, gravelly, slow, slurred, and it takes a lot of effort to articulate anything. I will lose my voice at some point.

Possible Measures

Drug:
Britain has approved only one drug for MND/ALS. Riluzole is believed to extend life expectancy by 3 months, and possibly to slow down the rate of change in the bulbar area. It comes in tablet and liquid form. I tried the tablets but unchewed solids are very difficult to swallow so I'm now taking the liquid form – a 10ml dose in the morning and evening. A few weeks ago I told my consultant neurologist that I didn't see the point of doing this, in my case anyway, because, frankly, 3 months wouldn't make much difference to me. She said that the medics aren't sure whether it kicks in at the very end, i.e. when someone's totally paralysed, wheel-chair-bound and so on (which I don't intend to be), or whether it's earlier in the treatment, i.e. it could be slowing down the bulbar effects even now; so she recommended that I continue. Seemed reasonable, so I am taking it.

Voice:
My Westminster/NHS-support voice therapist arranged, through the MNDA, to give me free use of American software, ModelTalker, plus a set of high quality headphones. At the computer I had to articulate some 1800 sentences provided by the software, each attempt

being measured in terms of Speed, Volume and Clarity, so a number of sentences had to be repeated a few times until I met the standards. In the US they then turned it into a sort of file/database and returned it to me. Then I was given Predictable, software on the iPad or other machines, where I will be able to type what I want to say, and the machine will speak the words with 'my' voice. It's called 'voice banking'.

If any MND sufferer still has his or her normal voice, i.e. if the illness started somewhere else in the body and hasn't got to the bulbar area yet, please bank the voice before that happens. Regrettably I could do so only after the degradation started, so the electronic result is virtually as bad as my speaking voice now.

Eating:
Everything MUST be chewed or soft. Nothing should be allowed near the throat or windpipe that isn't chewed or soft. It's quite difficult, so (a) I choose food carefully, (b) if it's solid I cut it into tiny pieces, and (c) I generally eat with a teaspoon – for me that's the best practical way to ensure that the quantities are small.

With the teaspoon or occasionally the fork I try to place the food to the side of the mouth, i.e. touching the cheeks, so that the teeth can chew. If the quantity is too large it can escape to the centre of the mouth, which is very dangerous. It does happen though, so if I feel there is something in the centre of the mouth the awkward answer is either to throw the head back and sideways, trying with a half-paralysed tongue to move it to the side, or else to use a finger to do so. Occasionally both go wrong and food is swallowed anyway. Very uncomfortable and potentially dangerous.

If I'm cooking with my own ingredients, and knowing that solids, however small, can trigger a cough or a choke, I cut them up into small bits and then mix them with a thick liquid, Hollandaise sauce or a yoghurt or suchlike. The paste-like consistency is easier to control and swallow. Likewise if I ever have to take tablets they go in a spoon of yoghurt.

There are categories for people with dysphagia - swallowing difficulties. I'm currently Level 6, "Soft and Bite-Sized", but as things get worse I'll go down to Level 5 and then Level 4. An NHS summary of the levels is here. Fortunately I get deliveries of complete Level 6 frozen meals, which can then be simply microwaved or heated in the oven. The company I use is Wiltshire Farm Foods. They're not expensive, and the quality is good.

I do still buy from shops and supermarkets I but have to choose carefully.

Vitamins and other supplements are preferably in liquid form or spray (there's a vitamin D spray for example).

Liquids:
Liquids are as difficult as solids but in a different way: they're not controllable so they can run anywhere and trigger a cough or choking. The answer is to make them thicker whenever possible. The ideal consistency is like yoghurt, so if here's gravy in a meal I mix it

with the 'soft' food as far as possible. Weetabix with rather less milk than in the past turns it into an easier virtual paste. Soup can be OK, but given the atrophying of the lips it's easier to drink it from a mug than from a spoon. Water, and plenty of it, is essential of course, but it can be particularly risky. I'm persevering with water at the moment, and my voice therapist says she can provide a thickening powder when I need it. So far I've not given up the occasional, rather careful, glass of wine or beer!

The same coughing, choking, and speaking problem is caused by the excess of saliva. The neurologist suggested two non-drug remedies: sage tea and/or red grape juice. I tried the tea and still do, but so far it hasn't seemed to help. Haven't tried the juice. The medics also suggested I try an anti-travel-sickness remedy, a small patch that goes behind the ear (the version I have is called Scopoderm). It dries out the mouth but also the skin. The first patch seemed to cause the skin on my hands to crack, though that might have been also a result of regular hand-washing advised by HMG to avoid spreading or catching the coronavirus. The second patch now seems to be working – less saliva – so I'll continue with it. Interestingly one side effect, for me anyway, is that I sleep deeply, which is very welcome.

The nose:
There are various sprays and also a liquid 'irrigation' system. I use both, and they help.

Exercise:
Whatever one can do. For me walking is good, but two days ago I received the official NHS letter saying that I'm in the high-risk category, being over 70 and with an underlying illness, and therefore I must stay indoors for 12 weeks. That's difficult: I have quite a nice one-bedroom top floor flat with a roof garden, but keeping the lungs and other still-working muscles active will be a challenge. Fortunately I took delivery of an indoor exercise bike last week, so that should help. And/or perhaps I should go up and down the 50 steps 30 times a day!

Invasion:
Once the swallowing stops, or preferably well before it stops, the answer is a peg (percutaneous endoscopic gastrostomy) tube, permanently fitted into the stomach, for food, hydration and nutrition. It will need to be checked, cleaned, disinfected and so on, and special food/liquid gently pushed in. I don't want that.

When or before the lungs lose their power, non-invasive and then invasive ventilation will have to be provided – ultimately a permanent mask with tubes going in and out, a pipe down the throat, and the appropriate pump to keep air going in and Co2 coming out. I don't want that either.

Any MND sufferer whose illness started elsewhere (not bulbar) will probably have to have those interventions in time, as the paralysis creeps up the body. With me it started in that area, so if I do accept the interventions I might have many months or some years, incapacitated with the invasive equipment permanently in place, as the illness travels down the body at its own pace. That is not for me.

Finally, attitude:
I'm not sure if I'm lucky or not. Most of the muscles still work, so even with a restricted voice, and caution when swallowing, life is generally alright – I can do much more than those whose MND started in arms or legs and are already physically incapacitated. And I'll try to live without a voice. On the other hand it's the bulbar area that requires the invasive treatment early on, so there's not much to look forward to after that.

One other thing: if people don't know you they can conclude you're a simpleton: they hear a slow, goofy, slurring voice, sometimes hard to understand; and they see you struggling to speak, fighting saliva and dribbling; and you're forced to say only simple, short phrases because you know the muscles won't respond to anything more complicated. As a result there can be a level of patronising. Having read a couple of blogs by MND sufferers it's clear that the patronising upsets them greatly. They know that their mind, brain, memory, sense of humour and so on are all intact, but they're trapped and can't demonstrate that fact. My own friends and neighbours know me well and so would never patronise, but it can happen with others. One way around that, although slow and laborious, is to write letters or tap the keyboard to replicate your voice, or write emails, or blog and so on, just to prove that the grey cells are still active. That's one reason why I'm doing this.

That's probably more than enough for now. When there are significant changes I'll do a short update.

If you read this, thank you. If you know anyone whose life is affected by MND in any way, and if you think this might help to prepare them, then do pass it on. An alternative is to give them the whole blog address, www.deardavid.co.uk , so long as you make them aware that it's not dedicated solely to MND: there will be a lot of non-MND-related items. If you do pass it on, you might like to suggest they start on Day 1 – April 1st – and then track it through because there will be occasional references to the illness in other topics.

With good wishes, and in particular to those who are involved in the illness.

David
as of 18 April 2020.