

NOTES ON MND/ALS, BULBAR VERSION

by David Peace
as of 9 May 2020

This is an update on the 18 April Note. That Note is below this one.

Possibly these Notes will not be comfortable to read. Sorry. The reality is important; I want to leave a record of my experience, how it changes me physically, and how it affects me personally. The Notes are for those reading the blog so that they can understand a little of (a) what MND/ALS means for me and (b) what it means for others whose illness either started in the bulbar area or has reached there. The Notes are for others also: I might send them to the MNDA or to anyone I know who might be helping someone with MND/ALS. And they are for me as a reminder, to track it.

General: in the last three weeks the illness has 'progressed' as the doctors say - not greatly, but sufficient for me to gauge the pace of change. It's expected and it's inevitable, so although it makes life more difficult it's not a surprise: I'm ready for it.

So, from the top:

Head, mouth, throat:

Speech more difficult. Tongue, lips, uvula, throat, all more paralysed. Voice slower and more guttural, growling. Needs greater effort to articulate anything.

Saliva/liquid: the mouth is permanently full, which is very tiring. I gasp a lot and am continually trying to collect the liquid and swallow, but with muscles that don't work it's difficult. Losing control happens more often. The lips have no power and so it's inevitable: liquid continually escapes. I don't know it's happening until it's too late.

Lips: the best description: each lip has the strength of a strip of tissue paper. Food and liquid can't be held in the mouth. They fall out down my chin, down my shirt, onto my lap, or onto the floor. I have developed some ways of stopping that, but it still happens.

Tongue: more limited movement. I can't rely on it to move food to the sides to be chewed, so the danger is that food will go down the trachea/windpipe. Have developed other techniques. And after each meal the teeth are covered in food because the tongue doesn't clear it. Continual brushing and flossing.

Cheeks: both are losing power and are slightly concave (the return of the handsome DP cheekbones!). Result is that occasionally, when chewing, I bite the inside flesh – painful and could be dangerous but fortunately no blood so far. I can still smile but in time all power will be lost and there will be no more smiling – but the sense of humour will remain, naturally!

Jaw: it still moves and so I can still chew, which is essential.

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Lungs: still a mystery. For the first two hours each morning I'm gasping a lot just doing the simplest of things. The appetite is good: breakfast is some or all of Weetabix, eggs, toast, yoghurt with sliced fruit. I'm then exhausted and have to sit for half an hour taking deep breaths, after which things are generally better. Strangely though, if I go shopping or have a walk even before breakfast the lungs seem OK. Just walking seems to help. So perhaps I should get out of bed and then get out of the house!

Rest of body: two weeks ago I saw my hands shaking. Again this is inevitable, I thought, as the illness spreads. But I can't see the shaking any more. Perhaps it's sporadic at the moment.

Arms, legs, heart, brain: not too bad as far as I can tell.

Fumbling: yes. Dropping and knocking things occasionally; awkwardness in moving things; extra time trying to fit keys into locks; and sometimes stepping this way and that, unnecessarily, at home. My current interpretation is that this is not a brain or muscle problem. I think it's lack of concentration: subconsciously my mind is on the major illness and so I'm not paying enough attention to what I'm doing minute by minute. That might be the wrong explanation but it's my way of seeing it at the moment.

Weight: I've been 14 stone/88 kilos for many years, and that's not too heavy for someone of my build. A visit to a health clinic last November reduced it to about 13.5 stone/86 kilos. I'm eating well but I'm now just under 13 stone/81 kilos. Apparently that's not good news, although it's not affecting me as far as I know, and I feel good at this weight. Also I can at last fit into old trousers again!

Techniques:

Eating: as mentioned on 18 April I'm still ordering Level 6, 'Soft and Bite-Size' meals with tiny solids. Also am shopping for similar soft food such as fish pies. Vital to avoid things that are too liquid (no control where it will go) and too solid (too big to manoeuvre in the mouth, impossible to chew sufficiently, and dangerous to swallow unchecked or let it anywhere near the trachea).

Food is taken with a teaspoon, which restricts each quantity, and it's placed inside, against my right cheek so that I can chew it; but it's still hard to control. I now tilt my head back so that gravity keeps the food in, and I use my fingers to move the food if necessary.

I've bought long, washable bibs which go close round my neck and are long enough to cover my lap. When eating I wear one and also have a tea towel and a kitchen roll in case of coughing.

In a couple of weeks I will start to order Level 5 food: 'Minced Meals' which have no solids. Later I'll have to move to Level 4: 'Puréed'.

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Drinking: I've not found any way to prevent liquid running down my chin and down the glass, cup or mug. The lips won't control it. I've tried different shapes including enclosed mugs with specially shaped spouts, but it still happens. Straws are no good as the lips won't close round them. I'm still searching for something and will be having a video chat with Amanda my voice therapist next week. Again though, it's often a case of tilting the head back, cautiously.

Yesterday, thanks to Amanda, I took delivery of tubs of 'liquid thickener'. The ideal consistency is actually that of a smoothie because it's more controllable, but smoothies don't give the necessary amount of hydration. It seems that this thickening powder can go in water and other thin liquids to enable some control but also some hydration.

Saliva/Liquid. Doctor's advice: The simplest recommended aid was sage tea. I still drink it but it doesn't seem to help me. The next was a prescription for hyoscine patches, small round plasters that go behind the ear and feed various chemicals through the skin – they're designed for travel sickness but they also dry out excess saliva and the skin. Each patch lasts three days. They did help but they also had two side effects: they gave me very good nights' sleep but also extreme and overpowering tiredness during the day. I have stopped them. I'm now sleeping not so well but there's far less tiredness during the day; and the excess saliva has returned. The third thing prescribed by the doctor, and expected soon, are atropine eye drops, apparently also with a drying side effect.

Coughing, Choking: still happen when something goes wrong, even just saliva. I generally feel I'm in control and can clear it, but it does take a time, it's messy, and it's very noisy.

My Day

These days, early awake – around 6.30. Read in bed. 7.30 up. 90 minutes for breakfast, bathroom, dressing, because I'm fumbling, gasping, and slow. Rest for half an hour. Then anything: emails, blog, sitting in garden, walking outside, the exercise bike for a short time; perhaps short, difficult phone calls, slow cleaning of the flat, TV (but mainly news- I'm not a fan). Lunch. Similar afternoon activities. Dinner. Generally late bed, after 11.00.

The Future

My life is now in permanent lockdown in this sense: I cannot again go to restaurants, pubs, cafés, coffee shops, hotels or other people's homes (unless they can cope with the mess and awful sounds of my attempts). Social life will be extremely limited: at some point I will not be able to speak, and although I will learn to produce it through the iPad, effective conversation will be close to zero. The paralysis will spread, affecting my swallowing, lungs and other limbs. It might take a year, or a few years, to consume the whole body. As I have said before, that is not the life I want. I must take action well before that happens, because when the swallowing stops I will of course dehydrate and starve unless the pipe is fitted permanently into my stomach; and then when the lungs stop there will have to be permanent invasive ventilation, similar to that inflicted on extreme coronavirus patients but in my case, I repeat, permanent. So the future is bleak. Unlike almost every other illness there is no chance of this changing. I can see and feel it approaching. That is why, for my

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own sake, for my wellbeing, and for my enjoyment of the life I have left, I have made plans and I must action those plans, to avoid awful suffering, without hope, for perhaps years. I want my friends to understand that. I hope you do.

Attitude

Not bad. Not depressed. Trying hard to get the most enjoyment out of an increasingly restricted life. Fortunately I love my home and do not find it burdensome to spend long periods of time alone. Very anxious to keep some sort of control. I can't allow myself to be trapped by the illness, because if that happens I will never be able to escape and it could continue, worse and worse, for years. Important to stay positive and determined.

That's it for 9 May!

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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

The lips

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

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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.

The tongue 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

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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,

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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

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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

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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.