

Pick's Disease by Dr. Bob Fay

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I have Pick's Disease, alias Frontotemporal Dementia. Looking back I must have had it for about 10 years. The good news is that it is in my case very slowly progressive: the bad news is that I have it at all. Pick's is a sodder. It has changed me in subtle ways that outsiders find very difficult to understand, but are all too apparent to my wife and family, It has cut short my career as a General Practitioner; it has stopped me from driving; it has caused much grief and distress to my family. It has a name, but no known cause. It has a very unpredictable course, and it has no treatment. It's a sodder, it's a sodder, it's a sodder. I am not seeking sympathy, but I hope that by conveying the truth about this disease, from my point of view, from the inside, you may be enabled to understand it better. Dementia has no dignity, no compensations: it continues day by day, year by year, to take and take and take, until death comes as a relief to all.

The general public have no knowledge of unusual dementias whatever. That is understandable. Most people know of someone with Alzheimer's (AD), but unless that knowledge comes from close first hand experience, the knowledge is usually very superficial and straws of comfort are readily grasped: like the comforting belief that behind all the obvious confusion and muddle and failing physical health, the person inside is quite happy, and is unchanged in the inner person. Dementia brings anxiety, anger and grieving to the sufferer. It may be bravely born, but it is not fun; it is not an acceptable alternative lifestyle. Our experience has been that in fact some psychiatrists, geriatricians and neurologists are fairly ignorant of some of the rare young-onset dementias like Pick's, and if they have areas of ignorance in those conditions, then I suspect that most of the other health professionals who are involved with dementia sufferers, are similarly lacking in knowledge. That is quite understandable and I am not seeking to belittle, after all I was a GP only 8 years ago who was totally ignorant of any dementia that wasn't Alzheimer's.

Incidentally what a very unfriendly word "Dementia" is! Technically I must have been "dementing" for about 10 years. To me the term suggests INSANITY (and the dictionaries agree.) To be demented implies being frantic, overactive, out of one's mind. I haven't settled on a better term, but I usually either say I have Pick's disease or that I have a degenerative brain disease. Sometimes I call myself "an old Dementonian" but then people think I'm claiming to have had an elitist education! I think the general public gets misled by the term dementia. Ginty, my wife, has reminded me of an incident at a party where she was talking to one of these infuriating women who can not only manage her family but also run a successful business and apparently neither succumb to stress or a bad hair day. Inevitably she asked "and what do you do?" When Ginty was forced into saying she was a carer,

this lady replied " Oh, how awfully interesting. Who do you care for?" And when Ginty explained that she cared for the man over there who was her husband, and was driven to reveal that I had a frontotemporal dementia, the woman replied "well, how extraordinary! I've just been speaking to him and there doesn't seem to be anything the matter with him to me!" She had expected someone with dementia to be totally socially unpresentable, someone so far out of it as to need to be tidily put away out of sight. I do feel that somehow we need to find a less loaded term to cover these conditions which, medically speaking, involve dementia. It's so hard for people to realise that in the early stages one may have a very real handicap, but still seem normal.

I'm not alone in finding that the most difficult feature of my condition is that people just can't see it. A recent AS newsletter gives the story of a lady 2 years into a vascular dementia. "I've been laughed at when I've said I have dementia," she says. "People just don't believe me".

When I was first diagnosed 6 years ago we were told that there were perhaps 600 cases of Pick's in the country. I'm sure that number by now needs another nought on it. We know of 3 other cases within 10 miles of us in rural Somerset. I suspect that for every known case of Pick's there are several more undiagnosed cases. And it is terribly important to diagnose them because undiagnosed Pick's is lethal! It's a disaster waiting to happen. Someone with hidden Pick's is going to make an awful mess of things sooner or later. For a doctor with Pick's it means missing vital diagnoses or giving wrong drugs, or practicing "masterful inactivity" once too often; for a business man it means errors of judgement that may lead to financial ruin. We know of several cases where a once prosperous family has lost everything because the husband's judgement went down the pan, and no one would listen to the wife who just knew something was wrong. It is just so tragic to meet someone who once had a good income, a lovely house, and the prospect of a decent pension, but now literally wonders where the next meal is coming from, and finds that the pension has totally been swallowed up by the nursing home fees. And all because by the time Pick's was diagnosed the husband had blown his business. Also: listen to and believe the spouse or carer. Frequently the spouse gets blamed "Dick's all right, the trouble is his wife is depressed". We know of a case where that situation went on for months and months. It should go without saying that the carer must be seen alone - well it should go without saying, but where my first diagnosis went wrong was because the Consultant literally turned his back on Ginty in the consultation and gave her no opportunity to speak to him alone. At times I can be fairly plausible, but if he had listened to Ginty he would have got the truth. There is just no room here for being PC and thinking it's immoral to speak behind the patient's back. It is in the patient's best interest and that's all there is to it.

I'm going to give you a brief run-down of what the features of Pick's disease are, just in case you know as little about it as I did when I was a GP. The onset, unlike AD, is usually between 50 and 60. Men and women are said to be affected equally, though you wouldn't think so if you have anything to do with the support group.

Here the carers all seem to be the wives of the sufferers, with very few husbands seeking support. We think this just reflects the different way men go about seeking help, or rather, not seeking help. There is a family history quite commonly, though not in my case.

It's a disease which affects the personality and social behaviour. Memory is affected but it's not the salient feature that it is in AD. So, considering the personality changes the sufferer may become more outgoing, or conversely - withdrawn. He may lose the ability to empathise with others becoming a cold fish, and being selfish and unfeeling. Aggressive behaviour may develop. He becomes less flexible, and will be irritated by being contradicted. He may develop obsessive routines, or get over absorbed in one particular idea - what in my case I call "getting a bee in my bonnet". He may lose his inhibitions and become sexually inappropriate. In other ways he may become inappropriate, making tactless comments, joking at the wrong moment or being rude, and generally being an anxiety and embarrassment to his wife.

Failure at work is often an early symptom, and is caused by the loss of the ability to make good decisions; also by the inability to notice changing circumstances, or, if he has actually noticed something has changed - he fails to act on it. So the businessman notices his junior is spending the company's money wildly, but does nothing. The doctor notices a significant lump, but takes no action.

Failure at work is also caused by loss of concentration, or by loss of social skills. I can't see when I'm really irritating someone. Another disastrous feature is that the sufferer may become a spendthrift. And finally, memory lapses will inevitably affect one's work performance. An important feature of the disease is loss of driving skills. This seems to be brought about by a mixture of loss of judgment together with an inability to be aware of peripheral things when the attention is focused to the front. Speech problems are a feature, and in true Pick's disease the sufferer may end up mute. An odd feature sometimes seen is overeating-simply stuffing one's face, and boozing. Other oddities are being unable to see things that are under one's nose; giving an opposite answer to that intended "coffee or tea?" and I say "tea" when I mean "coffee". Lack of drive is common. Needing more sleep, and falling asleep frequently during the day may be a feature. Increased sensitivity to pain or temperature may also occur. My family complain that I have a "Pick's face". I frequently have a cold, hard look, when I am anxious, agitated, or just in a thoroughly bad mood, but sometimes this face in no way reflects how I'm feeling at all. I occasionally catch this face in the mirror and I can see why it upsets the family because it just isn't me at all.

So you can see that frontotemporal dementia is a totally different picture from AD, and that's where many people have difficulty with my Disease - they expect to find something like AD, but in the early stages its nothing like AD. The later stages of the disease become like those of AD, so I'm told... one of the features of my picture is an extraordinary detachment about my condition so that I can talk about it as

though it were happening to someone else. This can be a bit unnerving to the listener, or on the other hand may make them suspect I'm a fraud.

Incidentally the definitive diagnosis of Pick's can only be made at PM ...and I'm not ready for that yet, though I have agreed to leave my brain for research when I die. There's always a need for nice normal brains, so if you are willing to be a Blue Card carrier...

But on a lighter note --- have you heard the story of the lady whose husband had just died suddenly. She went to pick up the death certificate after the PM had been done. "What did he die of?" she asked. "I'm sorry to say your husband had a massive myocardial infarct, a really massive infarct," the doctor said. "That figures," she says. So she goes home and the family say "Mum, what did our Dad die of?" "The doctor says" she said, "The doctor says, he had a massive internal fart."

I shall now give you my story which will illustrate not only some of the features of Pick's but will also tell of the failings of some of those who cared for us. I reckon I've probably had it about 10 years, and was signed off as sick in October 1995. For about 2 years before that I had noticed that I was having difficulty recalling what had been said in a patient's previous consultation, and was compensating by making more detailed notes. Also I simply could not find things on my desk when they were under my nose. Ginty was becoming critical of the way I drove and I was increasingly being beeped at. I noticed that on days when I was tired I was tending to slur my speech. But much worse, I was coming very close to physical violence with one of my partners. Practice meetings were stormy, to say the least. And that was quite out of character for me: I used to be mostly benign. Also I was getting totally exhausted in a way that was unusual for me. A night call would shatter me for days. Patients were coming off my list and I could not understand why. I've never been good with names, but I began to realise I was having difficulty recognising patients faces. The crunch came when I went to visit a lady who had just had a baby. I had been giving her antenatal care. When she answered the door I saw a lady apparently in her early 40s and I didn't recognize her. Was this the mother or Granny? I went home and told Ginty about it and about my exhaustion and we decided to see my GP.

I said "David, I'm either suffering from stress or I've got Alzheimers". Talking it through he grasped that something was wrong, but couldn't really say what. So he said "Shall I sign you off for a few weeks and arrange for you to see someone?" I agreed. So he referred me to a neuropsychiatrist. Interestingly, when asked at this time "what do a cow and a table have in common?" I was completely floored and could not think of any answer. In the meantime I was feeling quite exhausted, not miserable, but physically tired and convinced that my brain was failing and I would not be returning to work. The neuropsychiatrist was very thorough. I had an MRI brain scan which he thought was slightly abnormal but others thought was within normal limits; a radioactive vascular brain scan which showed diminished blood flow in the fronto-temporal areas; and psychometric tests. I did appallingly on the

latter. There was a lot of emphasis on memory tests: a story full of facts would be read out and I was asked to recall as much as I could.

Well, I couldn't recall much at all. So at the end of it all he said " I'm very sorry, I can't be 100% sure but I think you have Alzheimer's and because you're only 57 I would expect the downhill course to be rapid. Within 2 years you probably won't be good for much". "Oh, and goodbye, because this is just a diagnostic unit, and oh yes, I wouldn't advise you to drive." When pressed by Ginty for some sort of follow up, he agreed to refer me to a Memory Clinic. Well, you can imagine how totally shattered Ginty was. I wasn't - it was just as though it was happening to someone else, though I was fairly fed up about the driving. And so we went to the Memory Clinic as much for some sort of support for Ginty as anything else. I'm not sure I ever really understood as a GP what giving support means, but just being there, keeping contact, and listening, is immensely helpful. Anyway our experience has been that even an hour of a CPN's time every 6 months can make all the difference between coping and folding. So at the Memory Clinic we had a consultation with the Consultant followed by psychometric tests. I scored very much better than the first time, and no one could understand it. A few months later I scored even better. What had happened was that I was learning how to do the tests and I could remember them from previous times. It should have been evident that because this just doesn't happen in Alzheimer's the diagnosis was wrong. Now the Consultant made the mistake that I told you about before: he turned his back on Ginty and paid no attention to anything she said. He, I am sure, was wondering if I had become bored with General Practice and was trying to play the system. He started to express these doubts, which nearly screwed up our benefits. Sometimes doctors seem to have no idea the power they wield in such situations concerning the patients' finances. And then he made another terrible mistake: I asked him about driving because he had intimated that he allowed some patients with early AD to continue driving. He then said "well the obvious way to settle this is to ask Ginty what she thinks, so would you mind leaving?" When I returned 5 minutes later he said "sorry old chap, it's not on." So he had forced Ginty to pull the rug from under my feet and I rather naturally felt betrayed. Which is another major point; don't involve the nearest and dearest in the decision about driving; there is another way. So we were in a real pickle; our consultant was pretty sure I hadn't got Alzheimer's and was putting doubt into Ginty's mind. She was starting to think she ought to get me to pull myself together and I was fed up that she had spilled the beans about my driving skills. So Ginty tackled the Consultant and said, "alright, if you don't think its Alzheimer's what do you think it is?" "Oh he's probably got some other dementia", he said. "Like what?" she said. He wouldn't be pinned down, "there are masses of other dementias" he said loftily. But he really should have suggested referring me to someone who might have got to grips with my disease.

It was time to turn to our network of medical friends and find a doctor who could understand my disease and make a sensible diagnosis.

Our enquiries led us fairly rapidly to a Consultant who specialises in unusual and young-onset dementias. He listened to the story and spent time with Ginty alone and was able to say at the end of the consultation that he was virtually sure I had a frontotemporal dementia. He admitted me for the full work up; Lumber Puncture, MRI scan, and psychometric tests by the hour. These provided only very slight backup evidence for his clinical diagnosis. In the 6 years that have elapsed since then there have been only very slight changes on the serial MRI scans and psychometric testing, but there has been a definite deterioration in the clinical picture. The trouble with these tests is - the operators find it very difficult to do anything other than give me the same tests each time and I can remember them and even practice in my mind the day before where I slipped up the last time. Isn't that just so childish? But then I haven't told you that childish behavior is common in Pick's! I was still smarting about not being allowed to drive. "After all, "I thought," how can a Doctor possibly make that decision when he hasn't seen me drive?" It was for me the very worst feature of the whole disease, and I thought it was unjust. So I brought it up with the Consultant. His approach was so compassionate. He said" yes of course, I quite understand, and we can settle it quite easily by asking the DVLA's test centre at Banstead to test you. But I'm quite sure they will find you're not safe." So off we went to Banstead and I had a full days test, firstly on simulators and then an hour and a half at the wheel on the roads. And of course I failed but I could see why I wasn't safe, not only were my reaction times slow but if something approached from the side I just didn't see it. When my attention is focused on one thing it takes an awful lot to make me aware of something else that is going on. For example if I'm in the garden digging, Ginty may call out to me and I won't hear. She will come closer and closer calling out loudly and waving and I just won't register until she's right by me. I must add that it's not only my driving skills that have deteriorated. I used to be quite a competent dinghy helmsman, but I realized 3 summers ago that I had lost that skill too.

I must give you an example at how not connecting what I have observed to what I then do can get me into trouble. I just love having bonfires but sadly our next door neighbour doesn't seem to appreciate the wonderful aroma of a good bonfire clinging to her washing on the line. So one day I lit up just as she was hanging up her washing. She was furious with me and I did the same thing the next day making no connection with her temper or the circumstances only the day before. The next thing we knew was that we had an official letter of complaint from the Council. When Ginty went round to make peace, she simply would not accept there was anything wrong with me as "I seemed to know her name and what the time of day was." Pick's can get one in a frightful pickle.

So it took us 2 years to get the right diagnosis. During those 2 years I had been trying to find out why my brain had gone wrong. I read a book by someone who said he had been diagnosed as having AD and he had "proved" that it was all due to aluminium in the brain. So I got obsessed with that for a while, and then I heard that mercury dental fillings were to blame and that I was being poisoned like the old felt hatters. I got very keen on that one and seriously considered having all my

stoppings changed. Then I got obsessed with folic acid and B12. The Consultant finally got me to see sense. That illustrates another feature of my disease: I get bees in my bonnet and worry them to death. The truth is that no one as yet knows what causes Pick's except that in some cases it is familial.

I'm now going to try to picture for you how Pick's has changed me, and how it has affected those around me. It's not easy because obviously I don't have total insight, but I have collaborated with Ginty She feels I am not the person she married and I recognise that. All I can say is that it's wonderful how, despite that, she goes on loving me, and caring for me, so that we make the best out of it that is possible. There are many changes in me and yet to the casual observer I appear to be much the same. One person in our church even said that she now found me easier to get on with. This is because I have lost some of my inhibitions and find it easier to talk to people, including complete strangers. But others may find me rude and too blunt. I find too that I can often laugh inappropriately, and call out in public meetings.

Finding oneself inside dementia is like being suddenly transported from one's familiar house and village to a distant unknown island. Before one can settle down and feel at home on the island one has to explore it and find out what sort of a new country one is in. There are no maps, no guides. So it is in dementia. The skills one had and were proud of become changed, warped and devalued. The personality I had has changed subtly so neither I nor Ginty know exactly where we are. I relate to people differently. I fail to interpret the nuances of their expressions and body language so I can easily cause offence without realising it. My ability to make valid decisions has flown out the window. My memory is unreliable. And so on. During the first four years or so we had to painfully explore our new boundaries. I had to give up all sorts of things and become reconciled to that, and Ginty had to learn some new skills and assume new roles. Previously we had always done as much together as we could. Now we had to learn painfully that it was better if we spent quite a lot of time doing different things so Ginty could get breathing space. Before I took up medicine I trained as a Chartered Accountant and so throughout our married life until Pick's came on the scene, I looked after the bill paying and the money in general. It became apparent about 7 years ago that I was no longer as accurate with figures as I used to be and since we thought then that I was going to go down the pan rapidly, we decided that Ginty should take over that role. And the maddening thing was that despite not being a natural with figures, she has improved on my system and reconciles the bank account nearly always first time, which I never did!

In conversations I quite often switch off, and this is something the children find particularly distressing. During a family occasion I'm not able to participate in the way I used to because I easily lose the thread, or get the wrong end of the stick. I get easily irritated by noises. If there's a background radio on with someone talking on it I will find it so irritating that even if I am in someone else's house I will have to turn it off. I've already described getting bees in my bonnet, so you can

begin to see that living with me, or as Ginty so kindly puts it - living with Pick's - is living on a knife edge of wondering what's going to set me off next. In the first 4 years there was a lot of tension between us with my frustrations coming out in anger and aggression. If I'm contradicted I have the greatest difficulty containing myself, and can suddenly get very angry. I've had to learn that the best thing to do is to leave the room and cool off. Making decisions can be a difficult area. I often find I just can't choose between 2 courses of action and just have to say to Ginty "I'll have to leave that one to you". In ordinary day-to-day things it can be quite comical. I can be stood in the drive intending to go into the garden. So I start towards the larger gate and then think "no, that one's a bit stiff", so I turn towards the smaller gate and then think "but if I went through the other gate I could pick up a watering can". So I start again towards that one, and then realise that because I've already got something in both hands I can't manage the latch without putting something down and so head back to the other one. Its one way of passing the time I suppose. But if one of the children want some advice about how to handle a situation, perhaps a major career decision, I can find it impossible to give useful advice.

My memory obviously is affected, but this is nothing like the salient feature of the disease that it is in AD. At times there seems to be a complete block so I can't recall what I was thinking of literally 30 seconds ago. It's as if my computer screen has just suddenly gone blank, and whatever key I press I can't get things to move on. My word processor produces a little message that says "memory full". Well with me it feels like just the opposite. If I really try to register a conversation as I'm listening to it I often can, but otherwise I can come off the phone and be unable to recall much at all.

My speech has been affected as I mentioned earlier. In the early days I was very aware that it became slurred easily, that I would slightly mispronounce words, and have difficulty finding the word I wanted. I wondered early on if one could fight the disease by practicing the mental maneuvers that seemed difficult. So for about 6 weeks or so I read out loud for half an hour a day, and it did seem to improve my fluency. I also try to do a simple crossword every day. Inertia can be a real problem. I can see all sorts of little tasks that need to be done, but the inclination is rather to sit and read the newspaper endlessly. But when I do get started I'm fine, and that's one area where I need Ginty so much. At great sacrifice to herself she doesn't go out to work, but is around to be my helper and stimulator. I'm sure that is very largely why we have managed to keep our heads above water. Being able to laugh together is very important. Even more importantly - we were lucky to have had a very happy marriage before this all started: its hard to go on being loving towards your husband who has Pick's when you have always thought he was a bastard in the first place! And we are also so fortunate to have a rock-sure faith in God and Jesus, and to share that with our children. We often say to each other that we can't think how we could cope without the comfort God gives us. People often dismiss the Christian religion as being "just a crutch." It IS a crutch, a mighty amazing crutch! And whoever thinks he or she is so self sufficient that they never need

support in their lives fool themselves. We all need help in our lives at times, and for some of us its all the time. Its such a male delusion, that we are in control of our lives! The poet W.E.Henley wrote "I am the master of my fate: I am the captain of my soul," Bollocks! Pick's is a disease which obviously affects far more people than the sufferer. The true sufferer, as you may be beginning to see, is not the patient, who is relatively indifferent to it, but is the carer. Ginty and the children are the real sufferers, and this is what we hear again and again from other Pick's families. Of course it is true also of Alzheimer's. There is not much the doctors can do for me, but an awful lot that can be done for Ginty.

One of the best things about finding ourselves under the Consultant Neurologist's was that we then had access to the Pick's Disease support group. When Ginty was at the end of her tether or simply wanted reliable information about some aspect of the disease she could ring up and be helped. However despite this the strain of it all affected her health, and about 5 years ago she needed hospital admission for colitis. For support locally Ginty was offered 2 appointments a year with a CPN. Amazingly that tiny amount of input made a big difference to her, so much so that when it was suddenly withdrawn she had a relapse of her colitis. It was withdrawn without any reference to her as it was deemed "that she no longer needed it!" Eventually, after letters of complaint from our GP and Ginty to the Chief Executive of the local health trust, we had an apology, access to a CPN on a much more frequent basis, and I was allowed to have a day a week in the carpentry class of the local psychiatric team's Day Hospital. This not only gives me something creative to do but, more importantly, gives Ginty a day when she can feel free of Pick's. It has made the hugest difference to Ginty's stress levels and ultimately to my stability.

I find winter is the most difficult for me, and I can get a bit low. Ginty has been most resourceful at finding things to engage my interest. She saw an article about hooking rugs 4 years ago, and arranged for us to go on a day's course to learn about it. It's not very difficult and I had soon got the hang of it and have made some lovely rugs. However the right sort of wool amazingly could only come from America, so my ever resourceful wife thought that with the aid of a local evening class we might be able to spin our own. It wasn't quite that easy however as it is virtually impossible to spin a yarn of uniform thickness. But as a result of that we've taken up weaving as well and we both find it utterly absorbing. We both believe that a most important part of managing ongoing Pick's is keeping the patient absorbed in interesting pastimes and activities. I've mentioned carpentry, rug-making and weaving, but if you are wondering if we ever take any exercise I must add that last year Ginty got us cycling. The 2 of us did a sponsored ride across the breadth of Ireland and raised J5,500 for the AS. We had to choose the route carefully as I'm not safe in complicated traffic. I need Ginty's help crossing the road as I get too impetuous and start to cross with only a very hurried assessment of the safety.

I have already mentioned this, but I want to emphasise it again; some part of the week needs to be guaranteed Pick's free for the carer. Before we had this built into

the system our set-up was precarious. Now that we have this, Ginty is able to keep her head above water and as a result I'm less tetchy and less likely to "lose it". I am very ready to admit that I am dependent on Ginty. I fully realise that I could not live on my own, and if something were to happen to her I'm not sure what I would do. I would need to be in some sort of a protected environment. You see, it's easier if you have Alzheimer's. Ginty's father died of it and her mother is now in a Care Home for it. Our experience was that when we needed help with their care, support was immediately available and there was a choice of services as appropriate. But for the young-onset, less understood dementia, it's a different story. We are constantly aware that what we need doesn't fit into any health care box. I'm not geriatric, I don't have Alzheimer's, and I don't fit into the right mould for the local Mental Health Team, and it seems one is rather a nuisance. Whatever we now have in place has been the result of much kicking and screaming.

Our friends' response to my disease has been disappointing. The doctors are particularly clueless at understanding it. I have one old friend, a surgeon, who came to see us 4 or 5 years ago. Our son had just been staying with him and had done his best to explain the disease, but despite that he asked me if I was doing any locums and was I driving again? As if one just got over Pick's! And my relations have only just got it that to ring me up and ask for a medical opinion is not really very clever. The most infuriating comment I hear when I try to describe my defects, is "oh yes, I do that" "oh yes, I do that!" Well yes, your memory may have lapses, you may have moments of indecision, you may get bees in your bonnet, you may be unable to find your pen when its under you nose, but you don't do all those things all the time, to the extent that someone with Pick's does-- or at least I HOPE not.

So, I hope that has given you a picture of Pick's From the Inside Out.

Dr Bob Fay