



INFORMATION BOOKLET

**Personal case studies of
Frontotemporal dementia (FTD)**

Rare Dementia Support runs specialist support group services for individuals living with, or affected by, a rare dementia diagnosis. Our vision is for all individuals with or at risk of one of these forms of dementia to have access to information, support and contact with others affected by similar conditions.

Rare Dementia Support is a fund held by The National Brain Appeal (registered charity number: 290173).

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Foreword

Rare Dementia Support (RDS) runs specialist support group services for individuals living with, or affected by several rare dementia diagnoses. Our vision is for all individuals with or at risk of frontotemporal dementia (FTD) to have access to information, support and contact with others affected by similar conditions.

Rare Dementia Support seeks to:

- Provide access to disease specific information
- Facilitate contact between people embracing similar diagnoses
- To represent the views of people affected by rare dementia diagnoses across a range of strategy and policy influencing platforms

This booklet brings together information that we hope will be of use to people embracing a range of FTD diagnoses. There are many symptoms described in this booklet which fall under the spectrum of an FTD diagnosis. Not everyone will experience every symptom listed in this booklet.

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Personal case studies of behavioural variant frontotemporal dementia (bvFTD)

Ken by Mary Dawber

My husband, Ken was diagnosed with frontotemporal dementia at the age of 65, although previous signs of personality and behavioural changes began to present approximately 7- 8 years ago. Ken was a quiet, unassuming man, gentle, caring; he was my best friend and companion as well as my husband. He was an active and participating member of the local Baptist church. He loved to freely and generously use his gardening and physical skills to help anyone who found these tasks difficult. He drove a tractor for Manchester Corporation Ground Maintenance Dept. for 38 years, was described as a conscientious, reliable employee with only minimal sickness time in his whole career. He loved the outdoor life i.e. walking in the remote Highlands and Hebrides and bird watching. We had been married for 20 years (I had known him for 25), and he was devoted to his stepdaughter, then aged 10, and treated her with love and care. Around the age of 54 - 56 years, with hindsight, very subtle signs began to become noticeable. He gradually became unable to handle responsibilities at work. His writing deteriorated and he was struggling to write and spell words he had previously had no difficulty with. He would be told to drive to a certain place, set out, then be unable to recall where he had been told to go and became unable to handle any paperwork. With hindsight, he tried to cover up these failings at first - it was only after talking to his manager when he had to give up work that the full picture became apparent to me. He appeared to be irritable, but unable to explain any reasons; he experienced mood swings unrelated to external events, demonstrated outbursts of irrational rage and became very fearful and anxious for no apparent reason. He appeared to demonstrate a gradual failure of logical and rational thinking and developed fixed, rigid, irrational views, making reasonable, logical discussions impossible. Now, unless it is a matter of safety, we agree that black is white as it is fruitless to make him see reason. He was becoming disinhibited; tactless remarks with inappropriate sexual overtures were being made to my daughter, her friends, middle-aged ladies and elderly spinsters. He was becoming verbally and physically aggressive both at work and at home. His appetite for sweet foods was uncontrollable - he would eat a whole cake and then say he had only had one piece. He lacked insight into the changes of aggression and disinhibition and when he began to behave in a bizarre manner I decided something more than mid-life crisis was wrong. I remember with horror, whilst I was driving the car with him as a passenger on the middle lane of the M6, he opened the car door and attempted to get out of the moving car. He could not understand why I was so shaken up and attempting to keep him inside the car. In May 97 when I knew he had struggled at work that day, but not the full extent, I experienced the beginnings of more bizarre behaviour. He lay on the floor, kicking the furniture and me; logical conversation was impossible. The GP visited and asked him to go to the Psychiatric Hospital, which Ken agreed to do. He was admitted to a psychiatric ward for about a week and discharged being told his

problems were due to tensions and difficulties within the marriage. This statement was made without obtaining a relevant history of changes from his pre-morbid personality. Had the psychiatrist enquired what tensions and difficulties we might have been experiencing, he would have been told that two years previously I had been diagnosed with cancer and my husband's reaction was one of fear that he might not cope with being widowed. With hindsight, I now can see that this inappropriate emotional response to my diagnosis was an important sign. Ken attended anger management classes but had no insight into the reasons why he became angry - he was given antidepressant medication with no improvement. He remained off work, sick, continued to deteriorate in the sense of being unable to function, regressing into very childlike dependant behaviour, becoming easily distracted with poor concentration, and loss of ability to think with reason and logic. The GP asked if we could pay privately to see a different psychiatrist, which I agreed to do as it was becoming increasingly apparent that something was really wrong. Many times I quietly observed and listened to him, coming to the conclusion that this was certainly not manipulative behaviour. Ken definitely had no insight into his childlike, irrational state and he was genuinely struggling with procedures that required more than one set of actions. This second psychiatrist queried an organic process and Ken was referred for a SPECT scan. His behavioural problems of disinhibition and lack of insight were documented. Ken then had to finish work at the age of 58 years. He saw the Company Medical Officer, who on seeing Ken who was just pacing up and down the corridor, giving no logical answers to questioning, asked me how on earth was I coping with this situation? Ken has never driven again since May 1997 - fortunately he has never requested to do so. He becomes very agitated as a passenger - the only roads he copes with are single-track roads in the Highlands. Transport is a big problem as he becomes too agitated to travel in a small car; he could not cope with a busy railway station or sit still on a long rail journey. He enjoys the hour long air journey to Aberdeen or Inverness, but although his disinhibited, socially inappropriate behaviour can be a problem, I carry a Rare Dementia Support card which states 'this person has a brain disease. There may be problems with speech, behaviour and confusion. Your help and patience would be appreciated.' He insisted on reaching across me from his window seat to poke the charming airhostess on her bottom requesting more sandwiches etc. A large lady got on the plane and he promptly told her that she was far too fat to be taking just one seat! After the initial diagnosis in 1998 we had an excellent psychiatrist and Community Psychiatric Nurse who supported me. Ken went to a Day Centre for 3 - 4 days weekly which he loved - he enjoyed painting, knitting and telling his stereotyped jokes. We had to change Consultants and I kept trying to convey how irrationally aggressive, both verbally and physically, Ken had become, how his level of functioning was deteriorating and how he was losing social skills and awareness and behaving in a disinhibited manner. All my attempts at conveying these observations were dismissed. I was accused of exaggerating symptoms for financial gain. I repeatedly stated I was tolerating abusive behaviour and violent outbursts, but I was told "All you want to do is stick him in a Nursing Home". I was very upset at this slur on my character. We made a formal complaint, which led to Ken being discharged

completely from all services, with no assessments or history being taken or believed. We were told all his behavioural problems stemmed from tensions within the marriage! A specialist, higher academic opinion was obtained where the diagnosis of frontotemporal dementia was given, and confirmed at subsequent follow-up appointments with objective clinical findings i.e. dysexecutive function on neuropsychological testing, frontal and parietal atrophy shown on MRI scan, comparison being made between the MRI scan of 1998 and 2003. My husband's behaviour continues to slowly but steadily deteriorate. He is completely dependent in a childlike way, approaches strangers with odd and disinhibited remarks, wears odd combinations of clothes, undresses in public places and is rapidly losing social awareness and graces. I have been asked "Is he drunk?" He presents with a fatuous sense of humour, but fails to recognise the inappropriateness of his remarks i.e. on entering the GP's room he loudly announced "Two of your staff are pregnant out here - is this anything to do with you?" Someone once asked me if I coped by laughing with him. I replied "Yes" but if he's asking an elderly spinster for the 10th time if she's enjoyed her night in bed with the milkman, postman etc. people don't always find it funny". He has lost his judgement and reasoning ability - he is unable to weigh up the pros and cons of a situation and make a rational decision. He has fixed ideas and attempts at common sense and reasoning are impossible. He has set routes which he walks and will not consider any changes. He has developed obsessive ideas i.e. everything has to be in exactly the same place, a strict timetable has to be adhered to even if it means you are an hour early for appointments .Wherever there are displays of leaflets, he picks up 2 copies of every one, whether we already have them at home or not. Anything he wants goes into his pocket regardless of whether it is appropriate to take it without asking. He has stereotyped phrases and responses - it is like playing a tape over and over again. He is losing his ability to understand both spoken and written word - he is asking me what fairly simple words mean. He had no previous difficulty with these words. Sometimes he gets the syllables the wrong way round. He quickly becomes aggressive if faced with an uncertain situation, or it is necessary to restrict or challenge him to maintain his or other people's safety. He appears to have no judgement of danger i.e. he stands within inches of the edge of a high, steep cliff with his telescope and wonders why you are suggesting he stands a bit farther back. He has lost his natural empathy; he responds with inappropriate emotions to situations i.e. laughs at distressing situations, is oblivious to others' pain, tiredness or distress. When challenged by professionals he responds aggressively. This is becoming more noticeable now; at first the aggressiveness was more noticeable to the family, but now it is demonstrated in all situations. He attends the Day Centre where within the confines of a structured routine, with activities to distract him, his aggressiveness is not demonstrated so freely, or is quickly diverted by professional staff. Disinhibited behaviour is tolerated in a mental health setting to a greater degree than in general society or is quickly defused by distraction. He enjoys the activities, apparently getting up and pacing around, going from room to room as he desires but benefits from the security of a structured day routine. He can be apathetic, at times unable to maintain concentration, but then begins pacing. The problem with some people

with dementia is that they will not eat, but our problem is that he will not stop eating. I have given up trying to keep food from him, and although my food bills are enormous, I find it an enormous strain to try to contain him. Caring is so tiring, especially when in the early stages, not only was I disbelieved, but also I was accused of making up symptoms for financial gain. Social life is virtually nil. I tell people they have to be broad minded and unshockable to walk into our house, and there are very few people who are able to cope. I cannot leave him for very long as he forgets where I am, and I have to frequently ring him to reassure him. Broken nights contribute to tiredness – he awakes, hearing noises outside and insists the cat is brought in to lie on top of him. I live in fear of his unpredictable behaviour, wondering if I will be able to cope much longer and what the next stage will bring. There are still ongoing issues at local level - the refusal to accept the diagnosis of the tertiary centre is still ongoing and brings its own problems. Denial of diagnosis results in substandard care and lack of support and services, and is not in the patient's best interest, causing additional stress for the patient and carer. The slur on one's character brings additional stresses.

Bob by Ginty Faye

My husband was a GP and had to retire early eight years ago aged fifty-seven. The first sign that there was something wrong was the fact that he could not recognise people he had regular contact with as patients. Also he was finding that he could not "process" the answers to questions that he asked patients. He described it as feeling as if his brain was a computer screen that had "gone blank". I was very worried about his driving which had become very erratic and careless, one could even say dangerous. He also seemed to get excessively tired - little wonder when you think how hard he was having to battle to keep his brain going to perform the daily tasks of a GP let alone normal daily life. Bob was first diagnosed as having possible early Alzheimer's disease, but as his performance on testing actually improved over time the diagnosis was withdrawn. We were left in a no-man's land of no diagnosis. No interest was taken by the specialist in trying to find out what was wrong. One of our most worrying times was the financial implications of withdrawing the initial diagnosis followed by a lack of commitment to finding the true diagnosis. All our financial provision and government benefits were in total jeopardy and the worrying harassment continued for years. Once branded as a possible fraud you never shake off the label. However, having made our own private arrangements with the help of our very concerned GP we finally saw a consultant at a specialist centre where the diagnosis of FTD/Pick's was made. Our situation was at last taken very seriously. It was a great help to be reassured that Bob had a genuine illness and that we were not imagining it. We have over the years discovered that Bob's type of the illness is extremely slow in its progression. He is still, eight years on, able to put on a very good show socially so that it is not readily apparent what major limitations he has. This has proved to be a huge difficulty with many people saying "Oh he looks fine to me" and "doesn't retirement suit him?" etc. On the other hand when he loses his cool which is very much a feature of the disease and not a feature of his character before, others view him as being a rather difficult, nasty, rude person. They cannot relate it to the disease. The social implications of FTD/Pick's disease are wide ranging. We have had great difficulty with neighbours not understanding despite being told of the situation. We have had complaints to the local council about timing and frequency of bonfires and we then had threatening letters from the council. With FTD you don't connect any problem with the neighbours hanging up her washing and you in clear view choosing that moment to light your bonfire. When I was at particularly low ebb I was confiding in a close friend and their response was to try and help us by giving Bob some counselling (similar to marriage guidance I think). And I really thought they understood the disease. Some people have actually said that Bob looks better now than he used to - I wonder if this may be because he has a full time carer who is constantly trying to be two jumps ahead whilst walking on eggshells and find fulfilling things for him to do as well as keeping the environment around him as stable as possible! There is the ever present problem of how to pass the time. We live four miles from our nearest town and not being able to drive means that Bob is very dependent on me for getting about. We go to evening classes together to learn weaving and that is a huge source of enjoyment for Bob at home too. He does gardening in the summer,

we have a very "Pick's garden" as a keen gardening friend once says to me. Six foot plants in the front of the border and six inch plants at the back struggling for light. As soon as something is beginning to be established it is rooted up and moved somewhere else. Fortunately the only thing I care about is that Bob spends hours of time and pleasure in the garden. Reading was a huge help but unfortunately novels have to have simple plots with few characters now - library books are frequently put aside after a few chapters these days. One of the hardest things is the affect that the disease has on our children. We have four children who were 12, 14, 17 and 18 when Bob went off sick. So teenage years and now young adulthood have encompassed friends, boyfriends, wives etc. having to become acquainted with, and having the ability to cope with sudden outbursts, the inappropriate comments or the misunderstood threads of conversation. I must admit that young people never fail to amaze me with their willingness to try to understand - it puts our long term medical friends, in particular, to utter shame. Bob also has a tendency to withdraw emotionally when there are too many threads to the conversation. This gives the impression of a lack of interest which when you have previously been a father who has taken 100% interest can be very distressing. One of our children in particular was very badly affected and at the age of sixteen was diagnosed with anorexia. This was thought by the eating specialist to be a direct result of the changes taking place in her father. It was heart-breaking when it was advised that she could be helped by becoming a weekly boarder in her school which was just down the road. During the eight years we have had to fight for support. Life has been a real battle and everything we have in place is only there because I have fought for it. This includes regular contact with a CPN or a carer support worker, a social worker (only as recently as nine months ago did I realise we should have one). Two days per week are spent in the woodwork department or the mental health day centre and now after a recent crisis Bob has two separate weeks respite per year. There is absolutely no provision for this in our area so we have to travel to the other end of the country and the funding for it is the latest fight that I am in the middle of now. During these eight years alongside this, mother developed AD; she needed huge support while trying to live independently 4 miles away and is now in full time care. My physically disabled, unmarried brother's condition deteriorated before he finally died tragically 4 years ago. A huge amount of time had to be spent winding up his affairs. Naturally of course there is also the continuing hurly burly of 4 teenagers growing into adulthood. The stresses are manifold and you need to access every possible support available (and kick and scream until somebody hears you!)

Pick's from the inside out by Bob Faye

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 succumbs 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 recognise 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 Alzheimer's". 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 frontotemporal 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 it's 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 behaviour 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 realised 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 is 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. It's one way of passing the time I suppose. But if one of the children wants 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 has difficulty finding the word I wanted. I wondered early on if one could fight the disease by practicing the mental manoeuvres 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: it's 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 it's all the time. It's 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 £5,500 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 it's under your 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.

Talk for The Journal of Dementia Care' International Conference 2003 on behalf of the Alzheimer's Society.

Personal case studies of primary progressive aphasia (PPA)

Semantic dementia

John by Liz Pooley

When I first met John he was combining a career as a professional trumpet player with teaching music; he travelled thousands of miles a year, loved his work, had a wicked sense of humour and was a seasoned raconteur of funny stories. We spent many happy years teaching alongside each other in a busy department, were content with our lot and had every hope and expectation that we would remain that way until retirement.

With hindsight, the symptoms started appearing after we'd been married about 6 years and some 6 years before diagnosis, but it was very easy to apply the "my husband never listens to me" label and make excuses, and besides, it was difficult to actually put one's finger on anything in particular.

Over time he became rather distant and less co-operative, appearing to pay little attention to those closest to him, and yet he still liked to be the centre of attention at times; the jokes and funny stories stopped. He hung on to routines; still loved going to work, but at the same time became less aware of others and their needs. It is difficult to put a time scale on these things but I remember one day suddenly realising that I hadn't seen him laugh for a very long time.

Others were obviously noticing things too: he seemed to need to talk a lot but it wasn't always relevant to the matter in hand; the symptoms, though vague in nature, were very public because of his job, and because we worked closely I witnessed his sometimes strange behaviour, and the outbursts of wordiness. It started to become obvious that his sense of empathy and ability to weigh up a situation were failing and he would get extremely anxious when he thought he was getting things wrong and he didn't know why. Life was not good but still it was impossible to put one's finger on anything. The atmosphere at the school deteriorated and he started to feel a failure. Other people began looking for reasons and there was a consensus that it must be depression; he was even threatened with the sack if he didn't go for counselling. In fact he seemed to enjoy the counselling and was signed off from that with a clean sheet. We seemed to be lurching from one little crisis to the next and more and more friends and colleagues were urging visits to the GP: we seemed to be on a roller coaster that only went downwards and never up.

Anti-depressants were, of course, useless. Once started, the whole diagnosis period seemed to go on and on; countless visits to different doctors and hospitals. I had a secret hope that someone in the medical world would come up with a quick fix of magic remedy but at the same time had an underlying feeling that I had to come to terms with a major long term deterioration. The diagnosis itself was a huge relief for me: having a label for the condition and being able to explain to others that John had not been deliberately difficult and intentionally awkward and embarrassing all this time was a turning point. He did not

comprehend the reasons for giving up work, nor for having his driving licence taken away and 18 months on, still misses these acutely.

I have to say that throughout all of this, I had spent hours and hours sitting at the computer, “googling” desperately. When I happened on the Pick’s Disease Support Group (now Rare Dementia Support) website and read about other people’s experiences, one account in particular struck me: the parallels I saw were remarkable and I was convinced, afraid and comforted all at the same time. Personally I cannot thank the writers of those accounts enough for what they did for me – I think it was then I realised that life actually would go on but it was time to “think outside the box”.

Words are John’s biggest problem at the moment: he writes himself little “aides- memoire” – not jobs to do (as in my own constant lists!) but people’s names and common words that he suddenly seems to have trouble spelling. He is losing the ability to listen, to understand and absorb instructions and to answer the simplest questions.

Sometimes I have to “translate” for other people because of his strange mix up of words: dogs for doves, tractors for trains, flies for fish, snow for swans, even people with the same initials like calling a person Tony Blair because his name began with T and B. The rhythm and pattern of words and names is really important.

Fortunately, John is physically very fit; he loves seeing people and getting out and about and has a need to be busy and likes to help people. He gets very anxious when things are not as they should be and very upset when he thinks he’s wrong. He wears himself out with his compulsive behaviour: weeding, painting, picking up dead leaves, picking the leaves off the trees in the autumn so that they don’t fall off and make the garden look untidy. Its very difficult to get him to sit still and relax when we’re at home because he feels he has to be doing something and justifying not going out to work. He doesn’t enjoy television much because he can’t seem to follow a story line but will watch nature programmes and programmes with lots of outdoor scenery.

And yet, throughout all this, he continued to (and still does) play his trumpet exceedingly well. He practises seriously every day, performs with a group and occasionally does solos. The orchestral work has dried up since he officially finished work – it’s not the playing that is difficult for him but following instructions during rehearsals.

Now, seven and a half years on from the original symptoms, John’s speech is mostly fluent but with a little stammering at times when he’s trying to think of correct words; he speaks loudly, unaware of the volume, but there is not much substance to his speech. He has many phrases that he repeats frequently such as “I’m sorry” and is inclined to ask the same questions many times over. It is difficult finding activities that suit him and ways of focusing his amazing energy but he has recently started working as a volunteer at a local charity and this provides a tremendous opportunity for him to meet new people and get out and about and to retrieve a bit of this independence that he so misses.

Progressive non-fluent aphasia

Bob by Val Bywater

The man I married 43 years ago was courteous, considerate, kind and gentle. We met at College on a Youth Hostelling weekend and Bob became my best friend and companion as well as a loving husband and father to David and Kay. He was a quiet, capable, intelligent man, very practical and able to solve complex problems.

Bob was just 52 when signs of his illness began to show 13 years ago. He enjoyed his work and was self-employed as an Electronic Engineer and Computer Consultant in Research and Development. He began to work even longer hours to compensate for slowing down. He was concerned about losing his memory and became increasingly withdrawn; he was treated for stress and depression. Bob looked normal and was still able to do complex calculations and use logic. However, anything speech based had become a problem and our son gradually took over answered the phone and dealing with clients. With hindsight, I think the initial difficulty was being unable to find the correct noun to express his meaning. No-one would accept Bob's concern that he was losing his memory, since outwardly he still appeared to be a successful, confident and capable businessman. He always had a fascination with language and was able to hide the problems he had with finding the right word for some time.

When Bob was diagnosed with early-onset dementia at the National Hospital for Neurology and Neurosurgery a year later, we had no understanding of what it entailed. I found it very difficult to accept that my fit, active and loving husband could not be cured. Bob ran his last London marathon in under 4 hours a few months after diagnosis and was upset when major headaches meant he had to give up his main hobby. He became very agitated and also developed tinnitus, insisting on continual silence at home. These symptoms all disappeared after a few years.

In the early years, routine was important and everything had to be done to time. Bob worried unnecessarily about spending money and turned off every light and power switch that he could. He was concerned that his family were long-lived and continually asked me to have him 'put to sleep' like the dog. At this time he also rejected and blamed me for his illness. I found this hard to cope with and attended a few counselling sessions provided by Age Concern, which helped me to realise it was not surprising that I was struggling to cope! Luckily, Bob never understood the extent of his future deterioration.

We had previously arranged Power of Attorney and gradually I took over the practical and organisational tasks that we had shared. When we were out together I found myself thinking and answering for both of us, so that Bob's problems were less evident. At this time it was also great to meet other members of the Pick's Disease Support group at varying stages of their caring role.

After several years, Bob became less aware of what was happening to him and started to smile again. He has never been aggressive. Bob had by then finished work and wanted to be out walking, or cycling all the time. He spent long hours listening to simple music through headphones. Driving was also important, since he felt it was the last useful thing he could do. After a special test he held a one year licence and still drove well, since the spatial side of his brain appeared to be unaffected. Bob eventually accepted the decision that he should stop surprisingly well. We had many enjoyable self-catering holidays at home and abroad. Gradually, the use and understanding of words deteriorated further until finally Bob became mute, but still made odd noises!

We made the house and garden secure, so that at least I knew where Bob was; he was still physically very fit and could move very fast. Bob also tried an Alzheimer's Day Centre, but his attempts to kiss everyone were not appreciated. After escaping and getting into a strange lady's car, he was asked to stay away! For several years, with the help of good friends, Support Workers and Crossroads, we managed to give Bob a range of activities outside the house each day. One friend took him out regularly in the car with a picnic lunch to country shows, antique fairs and car boot sales. Support Workers took him for drives and country walks and I took him shopping and also dealt with all his personal care. We both enjoyed attending a weekly day centre together, where people had a range of disabilities.

As problems arose Support Workers helped me to cope with them. Issues we dealt with included Bob's lack of inhibition, his obsession with cars, the making of inappropriate loud noises, choking on food stuffed into his mouth and even running away up the road. One evening Bob did not return from his regular two-hour walk alone. He walked all night and was found about 25 miles away at 5.30 am, as he attempted to gain entry to a stranger's house. After this he was always accompanied, since he was by then unable to ask for help if he took a wrong turn.

During these this time I also looked after my mother, with early Alzheimer's disease, so wherever we went, Bob came too! After seven years, our Key Worker from the Community Mental Health team suggested we might be eligible for NHS Continuing Care funding at home. Luckily, I understood about assessment criteria and Bob's application was successful. The live-in Care Worker deals with all personal care and shares with me the organisation of appropriate day-to-day activities, whilst I look after Bob at night. I could not have managed without the companionship, friendship and support of the young people who have been our live-in Care Workers.

Since Bob's diagnosis, 11 years ago, life has sometimes been difficult, but I am so glad that we persevered with the fight for good disability equipment which enables us to provide Bob with quality care at home. Without realising it we have moved from 'caring' to 'nursing' and visiting Health Professionals are impressed by the high quality of nursing care that Bob receives. Information and support are available locally, and I have attended several excellent practical skills training days and information sessions organised by Carer's Support

and Action for Carers, as well as researching on the internet. I enjoy many practical hobbies at home and join Active Life hydro fit and aqua aerobics each week. We are very fortunate that the Consultant visits Bob at home and we now have a range of Healthcare Specialists supporting us.

One of the hardest things has been watching the cruel progression of this illness, until Bob has become totally incapable, requiring constant care. 18 months ago Bob stopped recognising people and then lost his balance and became unable to support his weight. His mood is now unpredictable; he is often noisy and demands a great deal of attention when not asleep. He has no communication, little understanding and is incontinent. Food is prepared so that it is easy to swallow, since tongue action is impaired. Bob still recognises and enjoys all the food and drink he is fed, although he has lost a lot of weight. He has poor balance, his arms, hands and legs are rigid and twisted, he is hoisted everywhere and sleeps downstairs. However, his heart and lungs still seem to be sound and Bob only occasionally has a mild infection.

Bob's eyes still follow me as I move around the room and he sometimes watches a large TV screen. He enjoys company and still seems to take comfort from my touch and voice. He also likes to be cuddled and kissed, when he gives a deep sigh and relaxes. Although Bob is physically with me, he doesn't smile now and I sometimes feel as if we have already lost him. Many times during his illness I have been close to despair and afraid of being able to cope. I don't know what life will bring in the next few years, but I am sure that it is better for us both to stay together at home. In spite of the problems and lack of sleep, Bob certainly benefits from consistent, loving one-to-one care. I am sure that I am much calmer and less stressed whilst still in control of our lives.

Logopenic aphasia

Phil by Janet Gregory

My husband, Phil, was 62 years of age and winding down towards retirement when I began to notice he was sometimes struggling to find a word - and not necessarily a complicated word, maybe something as simple as 'belt'. My daughter also noticed this, during her telephone calls home. At first Phil was able to substitute another word, but this became increasingly difficult for him. Unbeknown to me, he mentioned his difficulties to the GP, who thought he might have had a stroke, but tests were negative, and he was referred to a neurologist. At this stage I never accompanied Phil to medical appointments, so I am afraid it was a long time before I became aware that there could be a serious problem.

Meanwhile, Phil was diagnosed with myeloma, and underwent chemotherapy and a stem cell transplant. During this time, his communication difficulties were put on the back burner for a while. Eventually however, I went with him too see the neurologist and we were given a diagnosis of PPA, but no information about the condition, though a referral to speech therapy was suggested.

Phil has never shown much insight into, or curiosity about, PPA, and I am still not sure he understands the diagnosis. It was left to me to find out more via Google (scary), and to make contact with Jill Walton, to whom I was most grateful for her sympathetic ear and offer of information and support. Phil enrolled in the research programme, and enjoys his visits to London. The researchers have all been very friendly and supportive, always ready to answer questions.

We live a long way from London, and have no local support groups, but I pressed for a referral to memory clinic, and made contact with the Alzheimer's Society, who have provided a Befriender. We also attend a Singing for the Brain group, which Phil enjoys for the social contact. He can sing along to a song he knows if the words are provided, even though he would not now be able to read those words, without the tune. He volunteers for a local charity for a couple of hours a week, refurbishing tools, and a couple of old friends take him out from time to time. Phil's speech is now limited, he likes to chat but it is hard for anyone other than myself to understand what he is talking about, and his understanding is limited. Recent tests showed he retains understanding of single words and simple phrases, but no longer sentences. The speech therapist suggests writing down short phrases to be sure he understands, or showing him a photograph.

It is now more coming up to five years since we began to notice difficulties. Phil still enjoys a good quality of life; he enjoys socialising and still recognises friends and family, though he cannot remember names. His short term memory is poor, but he remembers things which happened in his childhood very clearly. He is still able to wash and dress himself, but cannot

remember how to use the computer (he used to be an IT specialist), his mobile phone or camera, or how to do any of the DIY jobs he used to love. He used to enjoy number puzzles such as Sudoku, but can no longer manage these. He sometimes does a simple crossword, and likes drawing and colouring, but all these activities have to be suggested to him, as he cannot think of things to do for himself. Phil has been taking donepezil for some time, and is now also prescribed Memantine, on the recommendation of the consultant at the memory clinic.

For me, life can be very lonely. We can no longer have long conversations as we used to do, Phil cannot really contribute to decision making, and of course I have to take charge of all practical matters. He has just decided to give up driving, rather than face another test. I miss him cooking for me, he used to really enjoy preparing special meals, but can no longer follow a recipe. I sometimes talk him through cooking a simple dish, and he is always pleased to prepare vegetables. It would not be safe for him to cook on his own now, and there have been a couple of near disasters when he tried to make toast or a cup of coffee while I was out of the house. I sometimes find it quite tiring, having to plan Phil's activities, remember and accompany him to all his appointments, administer his medication, and get routine jobs done. Phil likes to be busy so I often ask him to vacuum the carpets or do some dusting, but of course I feel guilty that perhaps I should be looking for something more constructive for him to do.

Last summer Phil did realise that we need to downsize as we can no longer manage the garden, and he agreed with my suggestion that we leave our village and move to a larger town, with better services, and a bit nearer to our daughter and her family. The house move has not proved easy to arrange and I have found it particularly stressful, but I am grateful that Phil does not seem upset at leaving familiar surroundings. He still seems able to orientate himself in new places, and likes getting to know new people. I think I am much more nervous about the move than he is, but I am sure it is the right thing to do, for the long term. His social worker has suggested that Phil would enjoy attending the Day Centre, and such services are much easier to access in a more urban area. We will also be able to get to see our little grandson more easily. Phil really enjoys our weekly visits, playing, talking and even 'reading' to the little one; he is a very proud grandfather.

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RARE DEMENTIA SUPPORT

Rare Dementia Support (RDS) runs specialist support group services for individuals living with, or affected by, a rare dementia diagnosis. Our vision is for all individuals with or at risk of one of these forms of dementia to have access to information, support and contact with others affected by similar conditions.

Rare Dementia Support seeks to:

- Provide access to disease specific information
- Facilitate contact between people embracing similar diagnoses
- To represent the views of people affected by rare dementia diagnoses across a range of strategy and policy influencing platforms

For more information about our services, please contact:

Rare Dementia Support

Dementia Research Centre | UCL Institute of Neurology

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