My dad and progressive supranuclear palsy (PSP)

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Dad aged 60 years was a powerhouse—energetic, handsome, tall and powerful. I thought he would live forever. He and mum were mountaineers, opening a treacherous climb in the Drakensberg, South Africa, which bears their name ([figure 1](#)), fleeing apartheid and starting a new life in Australia. He was a pioneering academic in his field after a terrible childhood. An exceptional marriage, three children, lots of adventures.

Then at 64 a colleague noticed that he looked a little stiff, and he was diagnosed with Parkinson's disease.

He kept falling—a dramatic plunge head first into a swimming pool at a posh party in his finest clothes, stumbles and trips on a bushwalk, multiple falls backwards from ladders. This was made all the more worrying as he was on warfarin for pulmonary emboli. His neck was really rigid and he didn't move his eyes.

Figure 1 Peter Angus-Leppan in the foothills of the Drakensberg during a 2-week climbing trip in 1961 opening up new rock climbs on Champagne Castle and other peaks in Lesotho (then Basutoland). Photo by Pam Angus-Leppan (personal collection of Angus-Leppan) (item 1961-03-01).

I realised this was progressive supranuclear palsy (PSP). I didn't want to tell him or the family; it seemed too awful to bear. Maybe my diagnosis was wrong. Maybe I should wait a bit. I didn't know how we could cope with this terrible disease.

He kept going. There were some twists in the tale. The early misdiagnosis helped him—he did have some response to levodopa and he may not have been offered this if he had been correctly diagnosed. Mum was incredible—against her bossy neurologist daughter's advice she kept him doing things. He went on buses to his volunteering—working for a charity promoting zero population growth (we chuckled at him having a daughter with four then five children) and he walked along the beachfront every day. He fell lots, bloodied his knees and sometimes his nose, but he kept going. Mum made him dress and wash himself every day even if it took an hour or more.

He didn't complain. Once, long after he had a percutaneous endoscopic gastrostomy (PEG) tube, and fluids caused him violent coughing, he said to me with a smile that he would really like a cup of tea, but he knew he couldn't. The blunting of his previously fine frontal cortex was a blessing. He didn't look in horror at what he had lost, or at what lay ahead.

And when things got worse, and he had nothing by mouth, he bonded over the PEG feeds, which his 5 year-old grandson often did for him. Dad was loved by his carers, who took him on walks and joked with him. Mum never hovered over them, she left them to it. As soon as they arrived, mum was off, grabbing an hour to do her own thing, to recharge her enthusiasm and her drive.

Dad walked his younger daughter down the aisle to be married, just months before he died. He cuddled his youngest granddaughter days before he died.

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Table 1 Learning points

Key points	Comments	References
Raise the possibility of a Parkinsonian syndrome if in doubt	Initial features may be subtle or equivocal	Early descriptions and recent revisions emphasise the variability of clinical features ^{1 2}
Offer a trial of levodopa to people with PSP	Some people with PSP initially respond	Response rate estimates vary from 10% to 50%; the PSP-Parkinson's form has the highest levodopa response rate ^{2 3}
Dad seemed less distressed about his condition than us, viewed by me as 'blunting of his previously fine frontal cortex'	In fact, PSP is predominantly subcortical so there may be apathy ('acceptance') or frontal disinhibition Responses will be slower so give PSP patients time to express themselves	Cognitive dysfunction occurs in most people with PSP Executive function is the most affected involving subcortical/frontal connections ^{4 5}
Medical relatives may seem angry or interfering and adopt a healthcare role, without having insight	This is often due to fear and powerlessness Guide us to stop trying to be our relative's doctor or nurse	There is general information for relatives of people who are terminally ill and PSP associations worldwide that offer support ^{6 7}
Excellent palliative care is crucial	Compassionate end-of-life care has a major impact Resist cut-backs that threaten progress in palliative care	Worldwide, only 14% of people who need palliative care receive it ⁸
Mum was sent a letter about dad's histology results confirming PSP	This could be distressing for some families, particularly if the results are not as expected	The UK Brain Bank information leaflet states that relatives will be sent a letter with results Best practice should include an offer for relatives to meet with the treating clinician to discuss the results ⁹

PSP, progressive supranuclear palsy.

The final weeks were awful. There was an unremitting cough, which nothing seemed to settle. In the hospice I fought to hold on to him, until my little brother told me to stop trying to be dad's doctor, and just be his daughter. I cried tears of guilt and anger. Why did I live so far away, why hadn't I done more, maybe there's another treatment, maybe if he gets through this hurdle, he will have another few good months. But he didn't, and there wasn't another treatment, and in the end I thanked my brother.

We were all there when dad died and he was peaceful. His body was just a body, I couldn't get over that the life had left it.

Lots of people came to dad's cremation. And there was even humour, when the words of dad's favourite song, 'Bobby McGee' played at slightly the wrong time. His coffin slid through the final doors to 'Busted flat in Batin Rouge, headin' for the train'.

Dad donated his brain to research and the diagnosis of PSP was confirmed. Mum received a copy of the histology by mail—she wanted to see the report but it would probably have been better for this to be delivered in person (table 1). Fortunately it did not faze her. Mum cut down dad's trousers and wore them so she felt close to him.

And 9 months later dad's youngest grandson was born. We wished they could have met.

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