

Clinical tales in neurology: a vegetative existence

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Forty-eight-year-old JM was admitted to the hospital for minor abdominal surgery. He underwent a general medical and cardiological check-up and a thorough pre-anaesthetic check-up, all of which showed nothing abnormal. On the day of the operation, his wife went to church, lit candles and prayed for him. JM was wheeled into the operation theatre at 9.30 a.m. He thought he would be back in his room in a couple of hours. He was keen to have the operation done, as he planned a holiday with his family in his village.

The senior surgeon had a hectic schedule on that day and asked one of his assistants to do the operation. The assistant in turn passed JM on to a junior colleague. The same thing happened with the anaesthetist. A postgraduate student in anaesthesia finally got the case. JM was eventually taken up for surgery at 2.30 in the afternoon. The surgeon was making the abdominal incision when he stopped midway. "Hey, what's happening? Why is this guy blue? Check his heart," he told the anaesthetist.

By the time the senior anaesthetist who was supervising the postgraduate student rushed in, JM had suffered a cardiac arrest. It took a few minutes before he was resuscitated. A ventilator breathed for him.

Suddenly JM threw a fit. An urgent neurology consultation was asked for. The doctor on call from neurology saw him 10 minutes later.

Cassandra's curse

The young neurologist saw a deeply unconscious patient who jerked his limbs at random. He gave JM an intravenous anti-epileptic. He hoped everything would be all right soon, but nothing became right for JM. Forever. Despite the anti-epileptic, he continued to get limb jerks and two more generalised seizures. He did not become conscious even after four hours. The frightened young neurologist called his senior.

The senior neurologist saw JM without delay. JM was still deeply unconscious but his seizures were controlled by intravenous phenytoin. There was no response whatsoever when he was called or pinched. On opening his closed eyelids, the doctor found the pupils were large but shrank well on throwing a bright light. Turning his neck to both sides produced weak movements of the eyes. Further investigations like CT or MRI scans were not yet available in the city. The patient still had a chance to live, but the neurologist knew that chances of recovery were remote. He wrote his detailed findings on the case sheet and came out to meet the patient's family.

By then only a small team of anaesthetists were there, to monitor the ventilator. The chief anaesthetist had told the family that the neurologists would decide the rest of the

treatment. The senior neurologist knew that he should watch out for trouble. He looked at the frightened family and as gently as possible, he suggested that it was too early to predict anything. God willing, the patient would recover. JM's wife wanted to know when that would be. But the other relatives were very angry and demanded to know what had happened to JM and why. The senior neurologist's personal charisma helped pacify them to some extent.

The next day the family held special prayers for JM in their church. Their priest predicted that the patient would be all right by the evening. They came to the hospital with the priest and prayed for JM. Some relatives went to the administrators to complain about the 'anaesthetic accident'. They threatened the doctors. Everyone thought a legal battle was certain. The police came for an enquiry. The atmosphere was tense.

As luck would have it, at noon the patient made a move. He stretched out his limbs rigidly. Seeing some feeble respiratory movements, the anaesthetist tentatively unhooked the respirator. JM breathed spontaneously. He opened his eyes slowly. The priest proclaimed that JM was on his way to recovery. The anaesthetists were happy to accept the verdict. Tension was eased for the time being.

The neurologist kept his mouth shut but wrote that while it was too early to say anything definite, the outcome was likely to be poor. He told his anaesthetic colleague that the patient had sustained severe brain damage. The anaesthetist told him that he would be called again if needed. In fact, he was not called for the next couple of weeks.

The senior neurologist remembered the mythological story of Cassandra. The god Apollo gave her the gift of prophecy – and also a curse, that no one would believe her. The senior had encountered many such situations, and knew that time would prove him right. Two weeks, later he read a newspaper article about JM.

Unscrupulous hacks and sob stories

Suddenly JM became a cause celebre. The press took up the story but distorted the facts. They called the doctors callous and wrote sob stories about JM's family. One photographer captured the haunting look of a tiny girl sitting by JM's side. There were heart-wrenching pictures of JM with tubes hanging about him. A governmental enquiry was ordered.

JM was unconscious but the doctors felt that he was improving as once in a way he opened his eyes on his own. Their hopes dimmed as the days passed, and six weeks later they knew that they were in a soup. The neurologist was called in again. He reiterated his diagnosis — extensive brain damage due to sudden cessation of breathing and heart during anaesthesia. When cornered, he gave the problem a name — 'persistent vegetative state'.

The patient didn't hear anything, didn't open his eyes and didn't change the position of his repose. He was riddled with plastic tubes – in his nose through which he was fed,

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into his urethra to drain his urine, and into his veins to give him fluids. When his eyelids were **lifted**, his unseeing eyes roved from side to side. If liquids were poured into his mouth, he would choke. He stretched his limbs tightly on being pinched. No other movements could be induced. Unaware of all the clamour in the newspapers about him, he lay there, not for days or weeks, but for more than six years.

Six years to die

A year later, the administrative enquiry wrote off JM's case as an unfortunate anaesthetic accident. He never woke up from his lethal sleep. As belated compensation, he was allotted a special room, free of charge, in the hospital. That room became home for JM, his wife and two children. He had to be fed, cleaned, bathed, shaved and nursed by his frail wife. Sometimes her two children helped her. They had to nurse him like an ever-sleeping baby. The doctors dutifully saw him every day, prescribed many drugs, which his wife found beyond her means to buy. The family went bankrupt. JM's wife went from pillar to post. Everyone sympathised with her. But sympathy did not buy her food or medicines.

Then two sincere journalists took up his case with enhanced vigour. The details of the anaesthetic accident were probed once more. It was becoming clear that something in that episode was being hidden. To stop further probing, the only way out was to give JM's wife a job in the office where he had worked. All news becomes stale after a few days, and dies a natural death.

After a few months, the urinary catheter was removed and condom drainage was installed instead. The feeding Ryles tube was taken out, but JM could eat nothing without it, so it was reinserted. To everyone's surprise, he survived repeated pneumonias, urinary infections and diarrhoeas. At the end of a year, everyone, including his wife and children, was tired and hoped that one of the **intercurrent** infections would kill him.

Six years is too long a time for anyone to die. JM dried out like dead wood. His limbs developed contractures and became flexed. His nails grew and cut into his flesh. Like a macerated foetus he lay curled and quiet for years. One night he just passed away, no one knew how. There was no autopsy, no long funeral service, he was buried unceremoniously in his church. Everyone was in a hurry to forget him.

Persistent vegetative state

Jennett and Plum coined the term 'Persistent Vegetative State' (PVS) in 1972 (1). Others had already described almost all its clinical phenomena. Jennett and Plum named it properly. From the closed-eyed comatose state, patients recover to some extent. Their unseeing eyes are open but they do not respond to the usual stimuli. They yawn and sleep but are incapable of anything else. Jennett and Plum didn't use the term 'permanent' vegetative state, as some did recover in the course of time (2).

In medicine, once a disease is named, others tend to use the name indiscriminately. In fact there is a distinct possibility of errors in making the diagnosis of PVS (3,4). Errors inevitably bring on media attention. If one such

patient awakens, it becomes front-page news.

With the increased use of cardiopulmonary resuscitative measures, the number of patients with PVS increased all over the world. Unexpected problems cropped up when they continued to live. It necessitated a detailed study of all the problems connected with PVS.

Living corpse

The Multi-Society Taskforce on PVS (5) laid down criteria for diagnosing PVS. Patients exhibit no evidence of awareness of themselves or their environment; they are incapable of interacting with others; they have no responses to seeing, hearing, touch or pain. They use no words, no language; their sleep-wake cycle is okay; they survive if they are looked after, because their vegetative functions work; they have some preserved reflex functions like blinking, swallowing, breathing, gurgling and even some limb movements, but these are not made consciously.

Consciousness is a tricky word. We would like to use it 'scientifically' but then we really do not know everything about it. It has two components (6) — 'wakefulness' and 'awareness of the self and surroundings'. The brain stem takes care of the first, and the second is handled by the cerebral hemispheres. In PVS, the brain stem functions to a great extent. But the cerebral functions are lost.

The Task Force found that nearly a third of head-injured people in PVS die. Seven per cent recover well. Fifteen per cent persist in PVS **till their death**. The rest come out with varying amount deficits. But if the cause is something other than trauma, 85 per cent die.

The average life expectancy for people in PVS is two to five years. Recovery after a year is unlikely though it has been reported even after 30 months. There are some indomitable people who live longer than 10 years with PVS. The longer they live, the more the problems.

In the arms of others

The legal pyrotechnics about PVS started in 1975 when 17-year-old Karen Ann Quinlan collapsed after taking a gin and tonic with diazepam. She stopped breathing twice, was resuscitated but in a few days became a PVS patient.

It soon became clear that Karen would not come out a vegetative state. The Quinlans' priest in the Roman Catholic Church agreed with Karen's father that the Church did not consider it 'immoral' to allow Karen to die by unplugging the respirator. But her neurologist, Dr. Morse, thought otherwise and refused to oblige because it would kill her.

Her parents took the issue up to the New Jersey Supreme Court. The Court sided with the Quinlans but ironically the hospital by then had weaned Karen off the respirator. She lived for eight long years as a PVS patient (7).

If Karen's parents asked to terminate her treatment, Helga Wangle's relatives refused the same suggestion from the hospital. Nancy Cruzan's fate was worse. This 32-year-old became a PVS patient after she was thrown from a car. The contractures of her body were so terrible that her fingernails cut into her wrists. Her family wanted the tube feeding to be withdrawn so that she could die. Though the trial court

agreed with the family's request, the public hue and cry made the Missouri Supreme Court reverse the lower court's verdict. It wrote: "The State's interest is not only the quality of life; instead the State's interest is in life; that interest is unqualified." (8)

Have you made your 'living will'?

Most doctors have confronted such problems, **creating** issues far beyond their clinical training. Few know of the legal, ethical and medical guidelines regarding them. (10)

The first is quite simple: non-maleficence. '*Primum non nocere*' ('First, do no harm'). The physician's duty is not to inflict evil harm on any patient. Of course none of us would do harm knowingly, but this can be caused at times by negligence and ignorance.

The second is 'beneficence'. It is to promote good and prevent harm to the patient. Whatever doctors do should be beneficial to the patient.

The third is the trickiest. Patient autonomy is (and should be) the first aspect of patient management. It is the individual's right to be self-governing. His will is ultimate in the choice of his treatment. But if the patient cannot express his opinion when the doctor needs it, the situation becomes full of twists and turns.

Anyone can execute a 'living will' regarding the type of treatment he should get if he becomes unable to express himself. This could be a 'Do Not Resuscitate' order (DNR). This means that if he falls down unconscious, he need not be resuscitated. Or it could be advance directives as to the extent to which doctors should strive to keep him alive. The 'living will' can provide clear statements of the patient's preferences regarding the choice of treatment.

The legality of the 'living will' is accepted by most states in the US. But there are some problems. A 'living will' executed when someone is healthy need not necessarily reflect the desire of the same person when he actually confronts death. Additionally, the physician's responsibility to the patient under his care need not necessarily tally with the patient's advance directive.

How long to prolong?

Sooner or later, a stage is reached in the treatment of a PVS patient when most people concerned recognise the futility of continuing treatment. But for a small minority, as Langfitt put it, 'enough is never enough' (9). They want the treatment to be continued till the last heartbeat.

At times, decisions on treatment must be made by relatives. Though this is the norm in our country, it is illegal and irresponsible unless authorised by the patient himself, or if the patient is a child or mentally incompetent. Most western countries have strict rules on surrogate decisions. The gold standard is the patient's best interests, weighing the anticipated burdens versus the benefits accrued by the treatment.

James Bernat is a champion of human rights in the medical profession. In an exhaustive monograph on ethical issues in neurology (10) he has addressed various problems related to practice guidelines for terminating medical treatment in

PVS. He stresses the importance of establishing the correct diagnosis and prognosis; identifying the patient's and family's preferences; choosing an appropriate level of treatment; seeking the opinion of the hospital ethics committee, and, if needed, seeking the view of the court.

Stopping treatment includes feeding. There are legal judgements in the USA that hydration and nutrition are indistinguishable from other modalities of therapy and can be withheld once the decision to terminate the treatment is taken (10).

I wonder how many Indian physicians have faced the terrible choice to stop treatment in PVS. Here more often than not, Thanatos comes in the nature of infections and gently takes the patient away.

Have we considered the emotional effects on the nursing staff? They feed these patients, look after them, get acquainted with their families. Nenner has given a touching account of one such patient (11). When the respirator was turned off and feeding stopped, the patient continued to live for nine days. The family stayed away. When a nurse placed a damp gauze pad to his lips, he hungrily sucked at it as if demanding in a voiceless manner to be fed. Gradually when merciful death snatched his life, the nursing staff felt that they were part of his dying.

When concerned people express their pain and anguish, at least listen to them.

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