

# Clinical review

## The patient's journey: Living with locked-in syndrome

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This is the story of Nick, who has lived with locked-in syndrome since 2000. His narrative is interspersed with information on his condition and a commentary on the clinical and ethical issues that arise in locked-in syndrome.

### Nick's story, in his own words

This is a story of my experiences since a terrible accident. I started the story in hospital and then added to it over the years. I had my accident on the rugby field on 29 July 2000 about 2 00 pm, just before the ball was thrown into a line-out. It just felt like a simple case of concussion (everything went blurry). I staggered to the sideline, the coach asked me "What's wrong"? He said I told him I just felt sick and to put me back on the field in 10 minutes. Then I collapsed and was rushed to hospital (unconscious) in an ambulance with the staff struggling to keep me alive.

After three days, doctors thought I was all right and were going to send me home. Then it started: I nearly collapsed again, taking a shower (I became extremely dizzy and lost my balance). For days, the specialists didn't know what was wrong with me. My girlfriend at the time went mad at the specialists to do something.

### The diagnosis

After six days of going in and out of seizures, after what seemed like all the tests known to man, they said I had had several strokes of the brain stem and then one major one, which left me with the extremely rare condition known as locked-in syndrome (box 1), not able to do anything.

### Start of the journey

Words can't describe the situation I have been left in—but this is as close as I can get it: an extremely horrific experience that I wouldn't wish on my worst enemy. The incredibly immense frustration levels at times have eased slightly over the years because of physical and health gains I have made.

If dying is as painless and peaceful as just drifting off to sleep, then there's plenty of really very frustrating times that at a particular point I wished I wasn't here any more.

Still, with mind and memory at 100% (sometimes I wonder if it's a good thing or not), external feeling 100%, internal feeling about 30%, although just after the accident I had no feeling. All my senses are normal, if not enhanced (sight and hearing). I'm just left



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trapped inside this body. All my muscles wouldn't work. Basically I couldn't talk, which went first; then I couldn't walk, eat, or excrete.

### A slow journey: the early days of recovery

Through a tremendous amount of hard work and perseverance I have gained ever so slowly since my accident and still continue to make gains now (they seem huge gains to others, but they seem far too slow for me—I've got to live through it all), with the much appreciated help and support of others and a copious amount of extremely hard work and sometimes pain.

It felt like I was in a really bad nightmare constantly for about the first three months. I could only just hear (I couldn't even open my eyes or breathe by myself); without them even knowing that I still could hear, the doctors and specialists in front of me said to my mum that I would die. They even asked my mum if she wanted them to turn the life support machine off after a few days.

### Box 1: The map: locked-in syndrome and clinical uncertainty

Locked-in syndrome (also known as *coma vigilante*) poses problems for clinicians, who just do not understand that their patient is a silent and unresponsive witness to everything that is happening. It is more often relatives than medical staff who realise the patient's predicament (usually by noticing intuitively that the patient is awake and registering what is going on). Nick's mother and his girlfriend pleaded with the medical staff to realise that he was aware of what was happening, and when the clinicians appreciated that the diagnosis was locked-in syndrome, the climate of care changed. A patient in locked-in syndrome cannot interact with us because he or she has lost the ability to control his or her body (except, in most cases, the ability to move the eyes up and down) but, importantly, the subliminal cues that intuitively alert us to the presence of another person are all that is needed for the suspicion to form and then the diagnosis to be confirmed by imaging and bedside interaction. Locked-in syndrome is caused either by a lesion in the brainstem (usually vascular) or by extensive demyelination, denying the brain its peripheral connections.

#### What locked-in syndrome is not

Nick, aged 23 at the time, was plunged into locked-in syndrome by a dissection of the vertebral arteries during a rugby game. The syndrome is quite different from other forms of coma or the persistent vegetative state, although they may be confused at the bedside. The confusion affects ethics as well as clinical assessment, but the ethical implications of the two conditions are quite different.

#### Locked-in syndrome and persistent vegetative state

In a persistent vegetative state (as distinct from locked-in syndrome) the higher levels of the brain have been devastated, usually by a combination of shearing stress injury to neurones and global cerebral anoxia. These selectively damage the higher brain—the cortical system and its ramified connections, so that there are no longer enough “megabytes” to do the work required for conscious experience and cognition. Both comprise mental activity on data gathered from the environment (in the philosophical literature—intentional functions) and they make use of the extensive processing capacity of the neocortex. Consciousness therefore rests on the coordinated and holistic functioning of widespread and dynamically interconnected cerebral functions. Nick's story makes it evident that he was from the beginning interacting with his environment in as many ways as his impairments allowed despite the fact that only a few people realised it. He recognised objects and people, conceptualised experiences, and experienced a range of feelings. These states do not survive in persistent vegetative state because there is insufficient (and insufficiently integrated) brain function to support them so that a patient in this state no longer has typical persistent vegetative state and is a consequence of the loss of the neural capacity required for that intelligent adaptation to the environment that forms the basis of human mental life. To put it simply, someone in a persistent vegetative state is not conscious because he or she cannot engage in the many acts of being conscious of the things of which Nick was conscious.

to wear an eye patch sometimes. For about four months I couldn't use a call bell. So if something was wrong or I was in pain or I needed something, I had absolutely no way of attracting anyone's attention.

Early on after my coma the muscles in my body were incredibly tight. I couldn't even rest the left side of my face on a pillow, even while I was lying on my left side. An itch is completely unbearable and incredibly frustrating because I can't scratch it. Now it's slightly better because I can move my hand a bit, but it's still incredibly frustrating. I had to wear a mouth guard constantly for about the first nine months, because my muscles were so tight that my teeth would grind. They still do sometimes.

I thought of suicide often (mainly when alone in bed at night). I never had enough courage then to go through with it. Even if I wanted to do it now I couldn't, it's physically impossible. I just have to deal with it the best I can. I guess this situation I was left in is as close as I could get to it. Before Hanmer [Hanmer Springs is the location of a rehabilitation unit in New Zealand] I used to think “What is this life for?” Sometimes now I definitely think just that. It's too difficult and extremely frustrating for me, most people just don't know how to communicate with me.

Even my case manager at the Accident Compensation Commission at the time said in the same room that I was in (my eyes were still closed), that “even if I did live, I wouldn't want to anyway.” One specialist told me to get used to the wheelchair, because I'll be in one for the rest of my life. What do they really know? They only know what they read in textbooks (box 2).

### Box 2: A “locked-in” witness to ethical decisions

In Nick's case, a clear diagnosis was made based on the anatomy of the injury and careful attention to the subtle signs of consciousness that he showed, but notice the tendency to marginalise or “absent” Nick as an intelligent participant despite the fact that he is an intelligent witness to these conversations about life and death. We can imagine the sheer awfulness of hearing others discuss turning off one's life support.

#### Locked-in syndrome, mind, and consciousness

The lesion in locked-in syndrome causes a loss of control of all the bodily functions that make a person accessible to others; in fact, once the care team realised what was going on, they then developed and used a series of interactive systems (such as eye movements) to open up lines of communication. However, despite this realisation, he heard things said about his prognosis and his life that paid little regard to what he might have been thinking.

It is worth dwelling for a moment on a philosophical claim by Wittgenstein: “Consciousness is as clear in his face and behaviour as it is in myself.” This remark is apparently disproved both by Nick's testimony and our common intuition that our conscious life is private and internal. Wittgenstein argues that the meaning of any term is a function of its use in a sufficient range of typical or paradigmatic cases, so we might infer that where the normal conditions do not hold, our normal ability to recognise what we are seeing may be impaired. Therefore, if we normally judge that something or someone is conscious on the basis of the myriad subtle cues we pick up in everyday interaction with an intact human being, then certain kinds of neurological damage will make it hard to make that judgment. Locked-in syndrome is exactly such a situation, where the fact that the patient is conscious is unrecognisable to everybody except the person concerned (and perhaps those particularly “attuned” to him or her). Nevertheless he or she uses all the perceptual and cognitive skills that normally reveal what goes on around us.

### Travelling alone

After my coma, it took a long time before I could even open only one eye. For a couple of months, when I could open both my eyes, I would see double and had

**Box 3: Misjudging patients with locked-in syndrome**

In the locked-in case, the mind of the patient is fully functioning but the integrated functioning of the psychosomatic whole is disturbed: connecting lines are cut, even though the internal hard drive is more or less intact. Therefore, as in other cases, such as profound cerebral palsy, the state of the person may be completely misjudged because, although it is subtly manifest, it may not be readily manifest to us. This is the implication of the fact that people who are attuned to the patient or on the alert for the condition can discern what is happening. But the thinking that affects our acute assessments also, and quite evidently, blinds us to the ongoing work of “self remaking” that someone like Nick is doing.

When we understand the problem, we can see exactly how it comes about that the person is still interacting with the world around him. But it is all one way traffic in that, however things strike him, he cannot show it. His showing and our ability to recognise his “presence” to us depends, in locked-in syndrome, on a myriad of subtle manifestations evident in eye and face and bodily habitus, even though voluntary activity and obvious responsiveness is what we normally rely on (especially in clinical life). Therefore in locked-in syndrome we can, and do, make mistakes about what is happening to the patient. We must make special efforts to reach past the locked-in syndrome barrier and allow the patient to connect with us. Locked-in syndrome is the kind of state that defeats our capacities to know how it is with another person, and when we are given an insight into his or her situation we can readily identify with Nick’s frustration at being unable to convey how it is with him.

Notice that a similar strategy defeats any attempt to argue for the abiding worry that can affect our attitudes to patients in a persistent vegetative state, where we worry about an enduring presence incommunicable to “us outside.” But in a patient in such a state, the capacity to be conscious has been destroyed so that it is almost the exact reverse of locked-in syndrome. The cognitive or experiential core of human life is gone in persistent vegetative state whereas in locked-in syndrome this core is intact, as Nick so eloquently tells us.

**Losing the path: frustration and loneliness**

Even a very simple task like getting my teeth cleaned is a mission because I can’t voluntarily open my mouth for a long period of time, and I can’t hold my breath. My natural reaction is to bite down hard on the toothbrush if anything other than teeth are touched with the brush. I also have to swallow all the toothpaste because I can’t voluntarily spit it out either. Not being able to swallow on cue can become immensely frustrating.

When you’re like this (despite having 24 hour care) it’s an incredibly lonely existence at times. It’s amazing how much time I have to think about things now since the accident. There’s heaps of thoughts that I don’t bother even expressing.

I talk by using a transparent Perspex board (about A2 size) with the letters of the alphabet spaced out on it (identically on both sides). The person holds it up between our eyes (about 800 mm apart). I spell out each letter of my sentence using my eyes (similar to a typewriter), with the other person guessing each letter I’m staring at, until I’ve spelt out a whole sentence—extremely laborious! It’s also very difficult (almost impossible) to express yourself or be sarcastic. When the person doesn’t understand what I’m trying to spell, it rapidly becomes immensely frustrating for me, and the frustration very quickly turns into anger, mostly because the spell board is my only method of explaining it (box 3).

**The courage to be**

Some time in 2001 I had to meet with my neurologist again. He wasn’t at all positive (telling me bluntly), “Whatever gains you have made to date, they’re all the gains you’ll ever make.” After being home about four months, I had to see one specialist who, ages ago, said I wouldn’t get any better (he basically said I’d never move or talk), he asked me to show him what I could do and say now, so I did. He apologised and took back everything he had said. I gave him the finger, and the carer and I left.

I’ve always thought, fuck what they think and say—or I would have been dead at the start. Most specialists and doctors with whom I’ve dealt since my accident have been so extremely negative.

**Travelling companions**

My family, friends, and carers have been so great to me in my recovery. Heaps of people have helped me in some way or other. Another friend has taken me to most of my old rugby team’s games in 2002, 2003, and 2004. All past support was immensely appreciated. I don’t think I could’ve made it this far without the support of my friends, carers, and family. I have met a lot of

**Box 4: Ethical implications**

We have touched on one set of ethical issues to do with the life of somebody who has survived locked-in syndrome and there are many others that take us far beyond the discussion so far. One might concur with Nick when he says that his life in the state to which he has been reduced is not worth living and that at times he just wants to be left to die but also be overwhelmed by the courage and determination he shows by going on living. In fact his determination to go on living is congruent with the majority response of patients in this condition. A person such as Nick has been tragically struck down and needs all the help he or she can get to rediscover something worth living for. After that has been done, it is his or her decision what then happens.

**Locked-in syndrome and persistent vegetative state** It seems that we can, if we listen to a story such as Nick’s, find a real life situation against which to assess our ethical thinking about persistent vegetative state and other brain injured states. In fact, the ethical implications of locked-in syndrome and persistent vegetative state are completely different.

In a persistent vegetative state, the experience of a person as a person (“a conscious living thing that hath reason and reflection,” to quote English philosopher John Locke) is ended and can never be restored because of a massive loss of neural circuitry. Thus the story of the person’s life, as a story experienced “from the inside,” has ended.

A patient in a persistent vegetative state cannot take part in life or in any decisions that are made about him or her. We are therefore left only with an advanced declaration (if one exists) or an objective standard of substantial benefit in making an end of life decision such a case. But Nick presents us with a different challenge. He requires only that we care enough to rebuild the tools of communication (through interactive technology and massive personal commitment) so that he can begin to live again among us, albeit with severe disabilities.

### Additional educational resources for patients and professionals

Locke J (ed. P. Niddich). *An essay concerning human understanding*. Oxford: Clarendon, 1689, 1975.

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Auckland Area Health Board v Attorney-General [1993] NZLR 235.

Gillett G. *Bioethics in the clinic*. Baltimore: Johns Hopkins University Press 2004. (Especially chapter 10.)

Multi-society task force on PVS medical aspects of the persistent vegetative state. *New Engl J Med* 1994;1499:1503.

Gillett G McMillan J. *Consciousness and intentionality*. Amsterdam: John Benjamins. 2001.

Gillett G. Wittgenstein's startling claim: consciousness and the persistent vegetative state. In: Elliot C, ed. *Slow cures and bad philosophers*. Durham: Duke University Press, 2001:70-88.

Wittgenstein L. *Zettel*. Oxford: Blackwell, 1967:221.

people since my accident. Some have become friends; some have become great friends.

I only live for hope of recovery now. I can't live like this for about another 40 years or so and will not (box 4).

### Travelling at varying speeds

From having to put in a gigantic effort to make just a sound, I can now say heaps of words, count, pronounce about four carers' names relatively clearly, sometimes stringing some words together when lying down and relaxed.

To be really honest, I would erratically and uncontrollably without warning literally "shit myself." Believe me, when you're 30 it's totally degrading. And nowhere more so than in the public gym, in front of people. It definitely changes my mood extremely quickly when it happens, as you can imagine. The specialists even wanted to operate and give me a colostomy bag... stuff that.

Some people say how determined and stubborn I am, but to cope with this and to have hope of full recovery you have to be. I could just moan constantly and deteriorate by staying in bed, but I want the old Nick back again, really badly.

It is definitely a crazy, mixed-up world. I'm just glad to still be alive—most of the time anyway. I accepted the fact that the accident happened, long ago. Shit definitely happens; I just have to make the most of each day in my journey towards recovery.

Most of the time (when living like this) frustration levels are pushed to the max and eventually I explode. Sometimes I wish I had died in the ambulance on the way to hospital. It would've been a lot less frustrating, for me anyway.

### Change is optional—but the journey does not end here

I feel extremely sorry for anyone with this syndrome who is scared of taking risks. Most things I do entail some form of risk—even something simple, such as eating. As it is in everyone's life—change is optional. I can choose to stay bedridden (which I once was), wither away, and eventually die. Don't will for death: it will come to you. Just sometimes sooner than expected.

### A little understanding

When I was a house officer I was bleeped to a ward to watch a man pass the last few moments of his life. He was "not for resuscitation," but I was called because his relatives were so distressed. On arrival, I remember feeling overwhelmed and helpless. I became rather angry at the nursing staff for the inappropriate call. Afterwards I told my colleagues about how it really wasn't my job to be there for the relatives. "What could I possibly have offered?" I naively said. It was the combination of avoiding a difficult and awkward situation and plain stupidity that led me to believe that I shouldn't be dealing with such circumstances.

Recently this memory surfaced when I lost my cousin. He was like a brother to me and was the first person close to me to have died. It was doubly tragic because he committed suicide. The incident changed my life. I journeyed through countless emotions, from guilt through anger to sadness.

The responses I received from people were also very varied. It is no doubt a difficult time, watching someone you care about cope with bereavement. However, perhaps naively again, I had hoped that doctors would be able to deal with it better than most. It saddened me to find that this wasn't the case. I was surprised how many doctors would rather talk about their exams or their rotas than offer their condolences. I was equally surprised how many perceive grief as being contained and never encroaching on your work. Anyone who has experienced grief knows the contrary to be true. Maybe I am being harsh; I know that words

cannot bring back my cousin, and nothing anyone says will alter the grieving process. All I wanted was an appreciation that it was a difficult time for me.

We experience the death of patients in hospital all the time, and it is considered part and parcel of the job until it affects our personal lives. I wonder if it is the doctors who think that it isn't their "job to cope with the relatives" of a dead patient who find it so difficult to know how to behave. My advice to any junior doctors out there who may be as naive as I was is that we are all someone's relative. Those of you fortunate enough not to have experienced loss should realise that losing a relative is one of the most difficult tests that we face. What we all can offer is some understanding and support, no matter what time and no matter who it is.

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We welcome articles up to 600 words on topics such as *A memorable patient, A paper that changed my practice, My most unfortunate mistake*, or any other piece conveying instruction, pathos, or humour. Please submit the article on <http://submit.bmj.com> Permission is needed from the patient or a relative if an identifiable patient is referred to. We also welcome contributions for "Endpieces," consisting of quotations of up to 80 words (but most are considerably shorter) from any source, ancient or modern, which have appealed to the reader.