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ARTICLE
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Locked in Syndrome, PVS and Ethics at the End of Life

A Neurodiversity Theme Article

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Nick Chisholm

NICK CHISHOLM

I had my accident on the rugby field on July 29, 2000 about 2.00 p.m. during a simple line-out, even before the ball was thrown in. It just felt like another 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, eventually blacked out and then was rushed to hospital unconscious in an ambulance with them struggling drastically to keep me alive.

After three days of being in there, they thought I was alright and were going to send me home. Then it started happening. First I nearly collapsed again taking a shower (I became extremely dizzy and lost a lot of balance). For days the specialists didn't know what was wrong with me. My girlfriend at the time, who had rushed down from Wanaka when she had heard it had happened (she and her mother were absolutely awesome throughout my time living in hospitals, considering the situation) went mad at the specialists to do something.

After six days of going in and out of seizures, finally after what seemed like all the tests known to man, they said I had suffered several brain stem strokes then one massive major one, which altogether had left me diagnosed with the extremely rare and unknown condition only known to a few as "locked in syndrome".

Locked in Syndrome (LiS) – the Phenomenon[i]

LiS (also known as *coma vigilante*) poses problems for clinicians who often do not understand that their patient is a silent and unresponsive witness to everything that is happening to them[ii]. In fact it is usually relatives rather than medical staff who realise the patient's predicament (they tend to notice that the patient is registering what is going on). In Nick's case, his mother and his girlfriend pleaded with medical staff to see that he was aware of what was happening and when that realization finally dawned, the climate of care changed. A patient in LiS cannot interact with us because he has lost the ability to control his body (except, in most cases, the ability to move the eyes up and down in the orbit) but 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.

LiS is caused in one of two ways:

(1) by a lesion in the brainstem (usually vascular); or

(2) by extensive demyelination denying the brain its peripheral connections.

Nick's LiS was caused by as vascular occlusion of the basilar artery due to a propagated blood clot from a vertebral artery dissection after the artery had been damaged in its course through the neck.

Diagnosing Locked in Syndrome

In Nick's case, the diagnosis of LiS was delayed as it often is and, in fact, publicizing his case has made us aware that many patients are only belatedly realized to be in this state. In some cases the diagnosis has not been made and they have died. That mistake can

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be avoided but it is a condition where the clinicians must actively think about pathophysiology and functional anatomy so that they ask the critical questions:

(a) Where is the lesion?; and

(b) What is the lesion?

The former is, these days, revealed by imaging techniques (although careful clinical assessment can tell one where and how to look). The latter is revealed by what older neurologists called a "badness time curve" for spontaneous medical conditions. The curve may be that of cerebro-vascular event (sudden loss of function), infection (relatively acute decline), tumour (subacute decline), or degeneration (slower decline perhaps with remissions and relapses) and it allows one to interpret the changes seen in imaging.

But, in the case of LiS we must also listen to those who know the patient because, as Nick's story shows, they may notice subtle indicators that hold the key to the diagnosis and may be missed by busy health care professionals. Once the diagnosis is made, the ethical and epistemological predicament is clear: how can we establish communication with this person who is somebody (a being-in-the-world-among-us) but who cannot convey it to the rest of us?

The means to communicate with the patient are limited and dependent upon a number of contingencies which commonly include the preservation of Eye movements of some kind due to the spared circuitry to some or all of the extra-ocular muscles (through cranial nerves III, IV, and VI). This has allowed Nick to establish communication using a transparent Perspex board and a trained therapist (prior to his regaining some of the movements of his upper limbs which allow him to use a joystick and computer).

A Contrast: LiS and Persistent Vegetative State (PVS)

> 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%. I have feeling throughout entire body, although just after accident I had no feeling. Despite some lack of internal feeling, all senses are normal, if not enhanced (e.g., sight and hearing). I'm just left trapped inside this body.
>
> At times it feels so surreal and still does sometimes to this day.
>
> To cut things short, considering I could only just hear (I couldn't even open my eyes or breath by myself), without them even knowing that I still could hear, the doctors and specialists in front of me said I would die to my mum.

Notice that Nick, in LiS recognizes that his cognition is "100%" whereas in PVS we have almost the opposite: the higher levels of the brain have been devastated, usually by a combination of shearing stress injury to neurones and global cerebral anoxia[iii]. 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 of conscious experience and cognition. Consciousness and cognition comprise mental activity on data gathered from the environment (in the philosophical literature - intentional functions[iv]) and depend on the extensive processing capacity of the neocortex[v]. Consciousness, we could say, arises from the coordinated and holistic functioning of widespread and dynamically inter-connected cerebral functions. Nick is, from the time he "comes to" interacts with his environment and gathers information from it in as many ways as his impairments allow despite the fact that only a few people realized it. He recognized objects and people, conceptualized experiences, and experienced a range of feelings. These states do not survive in PVS because there is insufficient (and insufficiently integrated) brain function to support them. The extensive neurological injury (a loss of the neural capacity required for the intelligent adaptation to the environment that forms the basis of human mental life) entails that a patient in PVS no longer has typical human experience (as is indicated by the severely attenuated EEG and Evoked Potential activity seen in PVS). The person in PVS is not *conscious (simpliciter)* because he or she cannot engage in the many acts of being *conscious* of things that Nick was capable of.

Locked in Syndrome and Being Somebody

Note that LiS is a state in which communication is lost but consciousness, thought and memory are intact so that "there is somebody in there" and that person is desperate to show that he or she is *somebody*, a being-in-the-world-with-others.

> It's too difficult and extremely frustrating for me, most people just don't know how to communicate with me.
>
> 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, there was absolutely no way of attracting anyone's attention.
>
> I talk by using a transparent perspex board (about the size of an A2 sheet of paper) with the letters of the alphabet spaced out on it (identically on both sides). The person holds it up between our eyes (standing about 800mm 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 starring at, until I've spelt out whole sentence. Extremely laborious! It's also very difficult (almost impossible) to express yourself or be sarcastic.

To be somebody is not just to be a body but it is to be there in the sense of being-in-the-world-with-others (a way of capturing the content of Heidegger's *Da-sein*)[vi]. As *somebody*, one has a name

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and an identity which is formed and reformed as one shapes oneself and is shaped in a cultural and interpersonal context by interacting with others who share that time and place (a place bounded only by possibilities of communication). This is an aspect of one's subjectivity as a human being – to be recognised, engaged with and taken seriously by those around one through entanglement in the world and the many conversations whereby one articulates and develops one's own abilities to respond to the world through whatever means and by using whatever is to hand in one's context or environment. Nick converses but what is fit equipment for him to use in communication is his Perspex board and that imposes significant limitations on who he can be to others in that he cannot be witty or sarcastic in the free-flowing way he would like to be because the rhythm of his equipment for entanglement does not allow that.

We tend to forget the way that rhythm and timing are part of personality and identity so that an individual for whom these are altered (as in muscular dystrophy or cerebral palsey) must find ways of being in conversation that allow them to "come through" to others and transcend their neurological impairments. For Nick the possibility of an easy two way flow in which his identity and agency can be manifest is "kludgy", bogged down, and made cumbersome in ways that frustrate his ability to be who he is and to develop his being through interaction and a permissive or non-intrusive mode of embodiment. His body is his being-in-the-world but there is both a continuity and a mis-match with the Nick whom he became through his lived narrative before his brain insult.

> I'm just a typical mind imprisoned in this body, I feel as though I'm encased in concrete that I'm constantly and painstakingly breaking through ever so slowly. Since the physical gains I've made over the years, I imagine it's become similar to trying to fight your way out of a 'Straightjacket'.

Notice that although we might be tempted to invoke Cartesian conceptualizations here, as suggested by "a typical mind imprisoned in this body" we are vividly shown how much the mind and spirit, in detail, is a matter of the lived body through which one takes up modes of being-in-the-world as a person with a distinct personality and style of relating to others. Nick's imprisonment is a matter of altered being not just the impairment of a separable add-on to who he is (in essence or in himself) even though the echo or trace of who he is does transcend his present subjective embodiment as a psychosomatic whole and always has done.

Ethics and Endings: Three Principles

> I feel extremely sorry for anyone with this syndrome that is scared of taking risks, most things I do involve some form of risk - even something simple like 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.

Nick takes risks and so for him there is a fundamental phenomenological difference in value between life and death. To what extent is this conscious and to what extent merely instinctive? The difference between consciousness and instinct and the valorization of conscious rational choice is, to some extent, a product of post-Cartesian existential thinking in which everything valued becomes a free choice made at the level of rational conscious valuation[vii]. In fact we live as subjective bodies – our embodiment is the condition of subjectivity. That is not to say that living as a human being is merely a matter of instinct because our being is to be the kind of beings for whom being is a question that can be asked. We live resolutely in the face of mortality and therefore we confront nothingness[viii]. Nick also reminds us that the possibility of ending it all for a person in LiS, is a matter of lived subjectivity and not a paternalistic judgment from the "high moral plane" of "normality". The subject who lives LiS can show us the world from his or her moral plane so all our elevated ethical deliberation is so much inauthentic "flummery and nonsense" conducted in abstraction and condemned to irrelevance because it does not inhabit the situation of the subject and situation is a crucial property of subjectivity. We can ask some standard questions but presuming to answer them on behalf of an-other who is radically other is a bridge too far.

When the subject is no more than a trace left displayed in a living inscribed body and is not a lived subjectivity the questions that can be asked touch on the following.

(i) *Benefit or prevention of harm:* Is this leading to an outcome which now or in the future the patient would consider worthwhile?

(ii) *Dignity:* Is living like this consistent with respecting the dignity of the person who this body has been?

(iii) *Consent or implicit consent:* If s/he could be asked would the patient want us to continue with the interventions maintaining life in this state?

In cases where the patient themselves has gone beyond the possibility of participating in clinical decisions, we can ask these things on their behalf. But, as Nick himself testifies, we may have to confess that it is impossible to know the end from the beginning and therefore difficult to answer these questions in an unconflicted way.

> 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 did happen, long ago. Shit does definitely happen, I just have to make the most of each day in my journey towards recovery.
>
> Some people think I should live a relatively normal life. Really ..... how do expect someone who can't speak or move limbs properly live a relatively normal life.
>
> 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.

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Our questions have an important application where the subject (or the capacity for subjectivity of a distinctly human type) has been destroyed as in PVS. In such a case the questions serve to reconnect us as decision-makers with the person at their epicentre.

We can ask about *Benefit or prevention of harm* by asking "What are the prospects here?" We can further that question through the concept of *substantial benefit* - an outcome which now or in the future the patient would regard as worthwhile[ix]. To that we might add a question about the RUB – "What is the Risk of producing an outcome that this person would regard as Unacceptable Badness?" Is it certain, or as near certainty as we can ever be that what we now have is just such an outcome in the person concerned?

Those questions have a direct bearing on the issue of *Consent* - "If per *impossibile* s/he could be asked, would this patient give you consent to do what you are doing?" And we follow that up with the further question, "Why should this patient, solely because he or she is unable to communicate, be treated in a way that s/he would not approve of?"

We can finally turn to the issue of *Dignity* by asking "Is this ending the right kind of ending for this patient's life?" This is, however, deeply problematic because we tend to have a much too stuffy and conventional view of dignity whereby the pretentious and posturing stances that are taken by public figures become our model rather than the truly admirable human dignity required to persist in the face of hardship and degradation.

> To be really honest I would erratically and uncontrollably without warning literary 'shit myself'!!! Believe me, when you're thirty it's TOTALLY DEGRADING! No place more so than the public gym, in front of people. It definitely changes my mood extremely quickly when it happens, as you could imagine.
>
> The specialists even wanted to operate and give me a colostomy bag.....stuff that!

Nick says that he does not want death to come, even to spare what we might think of as his dignity (even though he acutely feels its loss) but there are conditions where one could tellingly ask "Would this patient want to be remembered as the kind of person whose life ended this way?" or, in some cases, "Is what is happening consistent with the ethos of this family?" One might find that the most authentic answer one could give to these questions, in some cases, is to honour the person by stopping whatever intervention we are using to prolong the remnant of human life that remains of the subjective being who was once a being-in-the-world-with-us in a distinctively human way and took some pride in being the somebody they once were. In making this judgment we key in to a distinctly narrative framework for ethical questioning.

We should not, however, presume to answer them for a person when that person themselves is a being-in-the-midst-of-us who can answer them him or herself (like Nick). In that case our ethical responsibility is to reassure that person that we recognize her/him for who s/he is and want to find ways to add richness to his/her being amongst us.

A Human Life and the End of the Story?

A narrative question about the end of a human life is: "Has this person reached the end of his or her human story?" There are cases in which we feel, intuitively that that is so, a thought that aligns well with the idea of a lived subjectivity as the core of a human life story. We could list among those conditions (where the subjective story has ended) PVS from whatever cause in that the person concerned is unconscious, with severe diffuse neocortical damage. PVS has laid waste the information processing systems of a human being so that it is the loss of subjectivity and intentionality and therefore the loss of the consciousness which can interrogate the world and find meaning in it. The diurnal variation in EEG characteristics that some patients show do not indicate (even in the "waking" phase) any consistent responsiveness to events around them and there is usually a distressing absence of consistent reactions to relatives or acquaintances. The patient needs complete nursing care, medical nutrition and hydration, and has no prospect of recovery (that fact should be established by suitably expert clinicians after a thorough review of the etiology and pathological diagnosis. The best we can do in such a case, and in the light of the consent related questions above, is to respect the person by ensuring that his or her life does not end in a way they would hate it to. The appropriate end does, however, vary from culture to culture and even from family to family (which may raise questions of justice in a publicly funded health system)[x].

In contrast to PVS (and closely related conditions), a patient with Locked in Syndrome can be reconnected with the world as Nick clearly and emphatically shows. PVS patients (in sharp contrast with LiS) cannot reconnect to the world and, on most accounts (even those framed within ancient and deep religious traditions), a person no longer connected to this world should not be kept tied to a domain of mortal change and decay. Therefore it looks appropriate to acknowledge this fact and to allow (in the words of a Maori spiritual guide) "the spirit to go on its journey to the place where it now belongs". A secular ethicist may think of this as a realm populated only by the memories and stories of the living but a more religiously inclined observer might have a quite different way of capturing the reality that follows bodily death and is marked by ritual and myth[xi]. In either case, for a person whose connection with the world has run its course, we are justified in making a certain kind of end-of-life decision to respect that fact and the very fact that it is proper, on many accounts, to make that decision reflects the radical difference between such a case and LiS.

What, by contrast, are our ethical dues to the LiS patient?

The Duty of Care

At the end of any clinical journey we should be able to reflect on a duty of care properly discharged through the clinical team's engagement with the patient as the one most vitally concerned

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with what happens. To conduct oneself in the light of that truth and the partnership it grounds is to exhibit certain features in one's clinical management.

Attention to and *recognition* of the patient as the creator of and living being at the centre of the story of his or her own life. This attention and recognition can only happen through *conversation* in which one exhibits care for the person concerned. Nick needed that recognition and partnership as a being-in-the-world entrapped by his brain injury in ways that pose a constant challenge to his spirit and determination. Only through that engagement can he transcend the limits we constantly assign to his potential.

> Sometime 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'.
>
> They first Speech therapist told me because of the severity of my accident and the damage it caused, I would never be able to even eat again.

When things are done in the right way – which often is more by friends and those lower on the hierarchy of health care than ("exalted") specialists – the clinical experience is not a matter of determining limits which curb the potential of the patient but rather they embed a "being-with" properly enacted by those who have seen the problem before and others who are prepared to accompany the patient on a difficult and sometimes cruel journey. Recognising and witnessing to what a person is going through is deeply ethical in a way that is far more basic than any assessment of consequences or outcomes as it acknowledges a person's place among us as a moral community. Far too often the person who has suffered a physical or mental impairment of some kind (which go together in neurological disease) suffers the double injury of also losing their place as a member of the moral community or the kingdom of ends through paternalism, infantilisation, and the loss of dignity and respect that those attitudes imply.

For those whose subjectivity has been utterly destroyed or who are facing their own imminent mortality, being-with a person in the basic attitude of I-thou is the mark of a life juncture properly negotiated[xii].

What I Have Learnt from Nick.

I have learnt a great deal from Nick. I have learnt how much effort it takes when the subject who values his own being-in-the-world-with-others fights, in Luria's words, "with the courage of the damned to recover the use of his shattered brain" and remake something of his life[xiii]. I have learnt that every human being needs recognition for who he or she is and needs witnesses to their living who reflect the truth about that life (as it is lived). I have learnt that the human spirit is truly transcendent of the human body and its ills even though it is still a fragile thing and is touched in its being by the ills that flesh is heir to. I have learnt that who one is as a human being is an embodied subjectivity whose embodiment is inseparable from one's subjectivity even though one can also transcend many of the limitations of that embodiment. In those for whom the story has ended (as it most definitely is not for Nick) I have learnt that the clinician cannot go far wrong who gets alongside the patient (or their family) and has an open-ended conversation about what is happening and what should, in his or her opinion, be done, in the light of the thought that the outcome should be one that the patient would consider fitting (or worthwhile).

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Acknowledgements: *None*

Endnotes:

i) The indented passages in this article are verbatim transcriptions from Nick Chisholm's memoires as a Locked in Patient.

ii) Smith E Delargy M Locked in Syndrome BMJ 2005; 330:406-9

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

iv) Gillett G McMillan J *Consciousness and Intentionality* Amsterdam: John Benjamins 2001.

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

vi) Heidegger, M 1958 *Being and Time* (Tr J Stambaugh) New York (NY): SUNY Press, 1996.
Martin Heidegger

vii) I have discussed the difference between consciousness of the human type and that of other creatures in several places but the fullest discussion is in *Consciousness and intentionality* G.Gillett & J.McMillan (Amsterdam: John Benjamins, 2001).

viii) This is the way in which Jean Paul Sartre at least has made this ontic fact about us – that we in ourselves can question our own being and realise its contingency – vivid and memorable {Sartre JP, (1943 [1958]) *Being and nothingness* (Tr. HE Barnes) London: Methuen.}

ix) Campbell, A, Gillett G, and Jones, D *Medical Ethics* (4th Edition) Oxford: University Press, 2005.

x) I have discussed end-of-life issues at length in *Bioethics in the Clinic*

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(Baltimore: Johns Hopkins University Press, 2004)

xi) The term myth is used to indicate a structure of meaning that can be applied to an event and clothe it with deep significance for human beings and their understanding of themselves, their history, and their place in the scheme of things. It does not necessarily indicate a false belief or fantasy.

xii) Martin Buber, (1970) *I and thou* (Tr. W Kaufmann) Edinburgh: T & T Clark.

xiii) Luria AR, (1972) *The man with a shattered world* London: Penguin.

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