

Phenomenology of the Locked-In Syndrome: an Overview and Some Suggestions

Fernando Vidal 

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Abstract There is no systematic knowledge about how individuals with Locked-in Syndrome (LIS) experience their situation. A phenomenology of LIS, in the sense of a description of subjective experience as lived by the ill persons themselves, does not yet exist as an organized endeavor. The present article takes a step in that direction by reviewing various materials and making some suggestions. First-person narratives provide the most important sources, but very few have been discussed. LIS barely appears in bioethics and neuroethics. Research on Quality of Life (QOL) provides relevant information, one questionnaire study explores the sense of personal continuity in LIS patients, and LIS has been used as a test case of theories in “embodied cognition” and to explore issues in the phenomenology of illness and communication. A systematic phenomenology of LIS would draw on these different areas: while some deal directly with subjective experience, others throw light on its psychological, sociocultural and materials conditions. Such an undertaking can contribute to the improvement of care and QOL, and help inform philosophical questions, such as those concerning the properties that define persons, the conditions of their identity and continuity, or the dynamics of embodiment and intersubjectivity.

Keywords Amyotrophic lateral sclerosis (ALS) · Illness narratives · Locked-in syndrome (LIS) · Personhood · Phenomenology · Quality of life (QOL)

Introduction

This article discusses a framework and some steps that may be taken toward a phenomenology of the locked-in syndrome (LIS) – which does not yet exist as a systematic endeavor – by reviewing relevant materials and making some suggestions. *Phenomenology* is here understood as a description of subjective experience as lived by the ill persons themselves [29]. Though primarily descriptive, it can contribute to the improvement of care, and to informing philosophical questions, such as those concerning the properties and processes that define persons and the conditions of their identity and continuity. While phenomenology is about experience in the first-person perspective, individual experience is largely shaped by conditions whose study falls outside the purview of phenomenological analysis, and whose examination must therefore inform the task of interpreting the first-person perspective. After an overview of LIS as a medical condition, a brief discussion of LIS as an “illness” and a “limit situation,” and an outline of the challenges of communication, this article reviews four areas that offer materials directly relevant for a phenomenology of LIS: first-person narratives, philosophy, research into quality of life and personal identity, bioethics and neuroethics. It concludes with

F. Vidal (✉)
ICREA (Catalan Institution for Research and Advanced Studies),
Barcelona / Medical Anthropology Research Center, Universitat
Rovira i Virgili, Av. Catalunya 35, 43005 Tarragona, Catalonia,
Spain
e-mail: fernando.vidal@icrea.cat

suggestions for deliberately pursuing a phenomenology of the locked-in syndrome.

LIS and the Disorders of Consciousness

The locked-in syndrome (LIS), a disorder of profound paralysis with preserved cognition identified and named in 1966 [173], is a condition of extremely low prevalence.¹ Persons in LIS suffer from paralysis of the four limbs (quadriplegia) and cannot speak (anarthria), but have visual and auditory perception, consciousness, cognitive and emotional functions, and bodily sensations. They are “locked in” an almost entirely motionless body, and only exceptionally do they recover speech or movement. LIS presents three forms [9]. In *classic* LIS, blinking or vertical eye movement are preserved; *incomplete* LIS is similar, but presents additional voluntary movements that vary from case to case; in contrast, *total* or *complete* LIS is defined by full (including eye) immobility.

The chief cause of LIS is brainstem stroke – more precisely, a bilateral ventral pontine damage due to occlusion of the vertebralbasilar artery. Since there is no injury to the reticular formation or the cerebral hemispheres, consciousness is preserved. Damage to the corticospinal tracts makes speech and facial movement impossible, but since the supranuclear ocular motor pathways are spared, vertical eye movements and blinking are not affected (for overviews, see [21, 102, 126, 127, 197]). LIS can also arise in the advanced stage of a neurodegenerative disease such as spinal muscular atrophy or, more commonly, amyotrophic lateral sclerosis (ALS). Some individual cases reveal moderate and selective cognitive impairment in LIS of vascular origin (e.g. [150, 170, 187]), but others illustrate the prevailing outcome, where personality, cognition and decisional capacity are preserved (e.g. [3, 26, 27, 136, 193]).²

¹ There are no official figures. In Orphanet, the portal for rare diseases and orphan drugs, its prevalence is estimated at <1/1,000,000 (https://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=GBandExpert=2406; reviewed by Marie-Aurélié Bruno and Steven Laureys, last updated in December 2012). Using a functional (rather than etiological) definition of LIS, Dutch researchers calculated for the Netherlands a prevalence of 0.73 cases per 100,000 inhabitants [168], and in a survey of all Dutch long-term care organizations, prevalence of classic LIS was found to be 0.7/10,000 [109].

² In contrast, about 50% of individuals with ALS will exhibit cognitive deficits (e.g. [74, 169]). This is clearly relevant for a comparative phenomenology of LIS (vascular vs. neurodegenerative etiology).

Reaching a diagnosis of LIS requires regular patient assessment and a combination of methods (see overview by [114] and the recent case report by [207]). Since LIS may arise after a period of unconsciousness where cognitive function has been absent, chances of erroneous diagnosis of coma, vegetative state or akinetic mutism are high. LIS is the most common immediate target of research into brain-computer interfaces (BCI) for communication; increasingly user-centered, such research constitutes a “multidisciplinary challenge” well beyond strictly technical complexities [86, 106, 115, 119]. Specialists have proposed speaking of a “locked-in state” in order to emphasize that different etiologies can leave persons with a residual voluntary control over a few muscles, which allows them to communicate [116]. Needless to say, the care and rehabilitation of LIS patients requires very substantial human and technological resources (see overviews and case descriptions by [99, 156, 180]).

Mortality is high in the early stages of LIS of vascular origin (87% in the first 4 months), but stable patients have a considerable life expectancy: 83% live 10 years and 40% live 20 years after the cerebrovascular accident (CVA) that left them locked in [186]. An early review of over 130 cases described between 1959 and 1983 (supplemented by six first-hand cases) concluded that prognosis for survival and recovery was better for LIS of nonvascular origin (infections or tumors) than for LIS with a vascular etiology, but that early intensive rehabilitation benefited both groups and could lead to some degree of functional recovery [166]. This has been since confirmed [34, 90, 164, 189]. Patients who become locked-in as consequence of a stroke or traumatic injury sometimes evolve from classic to incomplete LIS; ALS patients, in contrast, are likely to move toward total LIS.

As Stanislas Dehaene ([51], 207-209) emphasizes, LIS is most definitely not one of the disorders of consciousness (DOC). It is, however, usually discussed together with them – not because it affects consciousness, but because it can be misdiagnosed as DOC, or even as brain-death [85, 146], and because, as illustrated in Fig. 1, it can be compared to DOC along the dimensions of consciousness and motor and cognitive function (e.g. [13, 17, 20, 36]). DOC include coma, the vegetative state (VS) and the minimally conscious state (MCS). Clinically, consciousness is defined by wakefulness and awareness. The former is assessed by eye opening and sleep-wake cycles, the latter by oriented behavior to external stimulation, such as command

following. At one end, comatose patients lack both awareness and sleep-wake cycles, and respond to stimulation only with primitive reflexes. At the other end, healthy and LIS subjects have both awareness and wakefulness. Patients in VS display wakefulness, but their visual, motor and auditory functions are restricted to reflexes, which is why VS has been renamed “unresponsive wakefulness syndrome” (UWS; [125]). Persons in MCS [73] may display visual pursuit or follow simple commands, recognize an object, or respond to single words or short phrases; some can communicate by modulating brain activity as recorded via fMRI [11].

As displayed in Fig. 1, LIS is like the normal state from the cognitive standpoint, but close to VS as regards motor abilities. (A “functional” LIS can also develop as recovery phase of VS; [69].) These differences between DOC and LIS account for the distinctive ethical challenges they raise. According to prevailing bioethical norms, obligations toward patients mostly depend on their mental life. The moral significance of correct diagnoses with respect to unresponsive persons derives from the fact that consciousness remains the “most critical moral constitutional standard for human personhood”

[49] and that, therefore, “our ethical considerations are specifically directed at conscious beings” ([161], 11-12; also [61]). The history of medical ethics is largely the history of concern for patient autonomy [63].³ Informed consent protocols expect patients to assume responsibility over their life through autonomous self-determination, and that requires consciousness and self-consciousness. Such a situation accounts for the high stakes involved in discriminating VS and MCS, and speaks in favor of focusing, not on the metaphysics of personhood or consciousness, but on biological distinctions and clinical assessments.⁴ A metaphysical position is nonetheless embedded in the possibilities that follow from a diagnosis: for it is the link between consciousness and personhood that explains why there is never any doubt that individuals in LIS are fully persons.

A philosophically subtler perspective going beyond the definition of consciousness as wakefulness with awareness recognizes locked-in persons as having not only phenomenal consciousness (i.e., a qualitative feel for their experiences, an awareness of themselves and their surroundings), as may be the case of some individuals diagnosed as VS, but also access consciousness – the capacity to remember, to conceive of oneself as persisting in time, to plan, to introspect and to decide. Whether or not one agrees with the idea

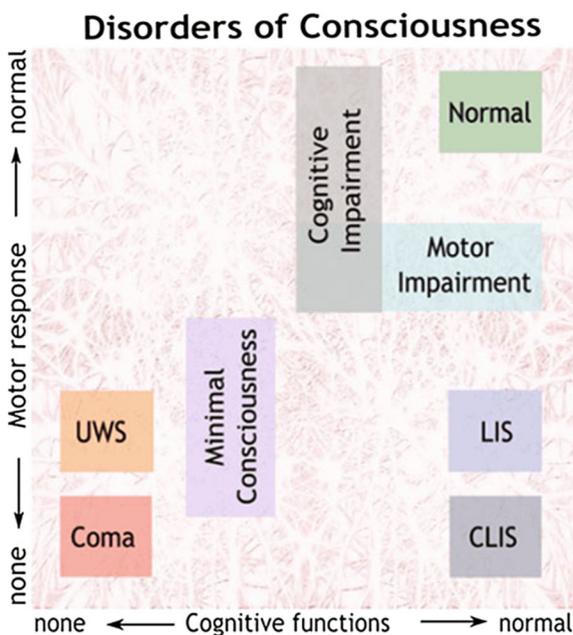


Fig. 1 Motor responses and cognitive functions in Coma, Unresponsive Wakefulness State (UWS), Minimally Conscious State (MCS), Locked-in Syndrome (LIS), and Complete Locked-in Syndrome (CLIS). From Guger et al. ([81], 106). Permission obtained through Copyright Clearance Center, Inc.

³ The notion of autonomy appears several times in this article. It has slightly different meanings depending on the framework being discussed, but it is always presented as a value, and as a constitutive feature of personhood whose core is the individual’s self-conscious ability to freely govern himself or herself. In neuroethics and biomedical ethics, it refers mainly to the capacity to choose or renounce treatment with full assumption of individual responsibility. Phenomenological approaches imply that the quality of being a person includes, but is irreducible to that kind of autonomy. The “enactivist” and constructivist interpretation of LIS argues that autonomy results from intersubjective relations. Beyond circumstances of extreme dependency such as LIS, the relational viewpoint seems particularly relevant to end-of-life decisions in intensive care, where it may be considered that the loss of decision-making capacity does not put an end to autonomy and personhood (e.g. [222]). These various usages are generally consistent with the first general principle of the United Nations Convention on the Rights of Persons with Disabilities, also mentioned below, which reads: “Respect for inherent dignity, individual autonomy including the freedom to make one’s own choices, and independence of persons.” The stress on individual autonomy as a crucial feature of personhood is a modern development of the Western tradition, which can be contrasted to the situation in other cultures (we here mention Japan). For an overview of the concept, see Christman [41].

⁴ See Fins and Schiff [67] for a recent statement of this position. Neuroimaging and electroencephalography have been used for assessing awareness by seeking to prompt and register the voluntary modulation of brain activity, and electroencephalographic methods have been employed for establishing neurophysiological measures that may provide indications about the future improvement of consciousness in patients diagnosed as vegetative [16, 195].

that a full right to life requires *both* phenomenal *and* access consciousness [128], the difference between LIS and DOC is in this respect crystal clear. And while it may be said that in persons with Alzheimer's Disease and dementia, the *accident* (the disease as nonessential property of being) has become their *essence* ([137], 193-195), no such transformation happens to persons in LIS, who maintain self-awareness and a strong sense of personal continuity.

LIS as Illness and Limit Situation

The clinical features of LIS open and sustain the possibility of elaborating a phenomenology – and even call for it. Indeed, for locked-in individuals, their medical condition represents a new manner of self-conscious existence and a novel experience of being in the world. Two distinctions contribute to frame these circumstances. First, the distinction between *disease* and *illness*, which has been present in theoretical writings on medicine since the 1950s [89]. The former is defined by the biological processes and pathological entities described and named in a particular biomedical context; the latter is the subjective experience of suffering persons, which is colored by their life history as well as by values, knowledge, categories and vocabularies of their socio-cultural milieu. A phenomenology of LIS implies a focus on *illness*.

The second distinction, found in Jean-Paul Sartre and Maurice Merleau-Ponty among others, is that between the *lived body* we are (*corps propre* or *corps phénoménal*) and the anatomical and physiological body we have. *Being* and *having* here indicate viewpoints, ways of considering the body as something we experience or as an objectified thing. German phenomenology, on which the French draw, speaks of *Leib* and *Körper*, *Leibsein* and *Körperhaben*. A phenomenology of LIS implies a focus on the *lived body*, which is the body involved in *illness*. There are examples of such undertaking for other conditions leading to paralysis – e.g. the first-person narratives by S. Kay Toombs [213] and Albert Robillard [181], who suffered respectively from multiple sclerosis and ALS; Robert Murphy, who suffered from a spinal cord tumor, calls his memoir “a social history of a paralytic illness” ([148], 3), but, like Toombs' and Robillard's, it is also a deep and insightful contribution to phenomenology.

We can think of LIS as illness in terms of what the psychiatrist and philosopher Karl Jaspers [94] called “limit situations.” These are unavoidable events, such as death, suffering and being subjected to chance, which

we usually forget or conceal to ourselves. Circumstances may push us to confront them and become aware of the human condition they materialize. While LIS is obviously not a universal, inescapable predicament, it poses the conceptual and empirical question of the relationship between the self, the body, others, and the world in the framework of an extreme existential situation. Like Jaspers' *Grenzsituationen*, it magnifies those questions and brings into focus crucial factors at play in exploring them. Thus, Havi Carel ([31], 346) speaks of illness as a “limit case of embodied experience” that, by virtue of its distancing effect, can become a “philosophical tool for the study of normally tacit aspects of human existence.” Indeed, as Murphy ([148], 5) put it, the disabled represent “humanity reduced to its bare essentials;” they are “not a breed apart but a metaphor of the human condition.”

The Challenge of Communication

I think I could tolerate a lot happening to my body, but if I lost the ability to communicate permanently, I just might be thinking Game Over.

ALS patient Kate, quoted in Caron and Light ([32], 688)

The major challenges in LIS reside in the conditions and possibility of communication, which is an essential element in locked-in persons' quality of life (QOL), emotional state and sense of existence. Not without good reasons do literature, TV and film emphasize them when representing LIS [46, 82].⁵ Beyond its practical usefulness, communication gives LIS patients full membership in the community of human persons. Testimonies agree on this, but the most daunting is surely Julia Tavalaro's *Look Up For Yes*:

⁵ Kondziella [112] claims that Roald Dahl's 1959 story “William and Mary” describes complete LIS and predicts recent successes in detecting covert awareness. However, the story (which I have often used in my teaching) is about William's brain kept alive in a vat, with one of his eyes connected to it; thus, even though William is told that he will be able to communicate thanks to an encephalograph-like device, the lack of a full body marks a considerable subjective, intersubjective and philosophical difference between William's vital circumstances and those of a locked-in person. We meet here the well-known limits of thought experiments [226].

“Can you close your eyes, Mrs. Tavalaro?”

With these words, I’m shocked back into reality. This is no dream. I’m actually being spoken *to*. I close my eyes. I open them and see Arlene’s face. “Can you blink twice?”

I do it.

Silence fills the space between us. Her face shows shock and grief and happiness at once. In the previous six years, no one had thought to ask me these simple questions.

“Okay, Mrs. Tavalaro. I’d like you to respond with eye movements. Can you move your eyes up, like this?” She rolls her eyes toward her forehead.

I watch her do this. Then, with a quick movement of my eyes I feel my mind rise from the ocean depths of pain. For the first time in six years, I feel whole. ([209], 121)

Tavalaro (1935–2003) entered the locked-in state, as a consequence of a stroke, in 1966. She was considered in VS and often maltreated; her mother and sister claimed that she was conscious, but were not heeded. The confirmation came only in 1973 thanks to the speech therapist Arlene Kraat. Over time, Julia was able to communicate with a switch-based scan and use head movement to drive a power wheelchair. She later participated in writing workshops, began to compose poetry, and wrote her memoir in collaboration with the poet Richard Tayson. (In addition to [100, 143], see [188], a testimony of Julia’s occupational therapist from 1973 to 1981.) Beyond medical misfortune, circumstances make Julia’s story exceptionally heartrending.⁶ But all other

⁶ Tavalaro’s story is in this respect analogous to Martin Pistorius’ (born 1975). Aged 12, Pistorius fell into a vegetative-like state of unidentified origin, began regaining consciousness about 4 years later, and was fully conscious at around age 19. It took about six more years before a caregiver noticed and convinced his family to have him systematically assessed for cognitive capacity and communicative potential [171, 172]. Andrea Ostrum (1941–1994), who suffered traumatic brain injury in 1985, underwent a different yet related kind of ordeal. She was discovered to be locked-in after several months, when a court required evaluation before her feeding tube could be removed. She thus lived – but only to enter “a nightmare that was worse.” She writes: “. . . although I had injured my brainstem. . . I did not conform to the other distinguishing textbook characteristics [of traumatic brain injury]. . . in my thoughts and my core identity, I was my old self. . . Now, because I was trapped in a badly injured body and because the medical books said it was not possible, I no longer existed. For 3 years, I was addressed by every new person as if I weren’t in my right mind. . . .” ([160], 97). In a less angry testimony, she observed that many people regard the brain-damaged “as a species apart,” and defended their right to “be treated as individuals, not textbook cases. After all ‘if you prick us do we not bleed?’” ([159], 624).

autobiographical narratives document daunting interactional difficulties, give communication a central role and describe it as the locked-in individual’s major existential challenge.

Persons in classic and incomplete LIS can communicate via the voluntary control of blinking or vertical eye movement to signal *yes* or *no* in answer to questions, as well as to select letters and compose words and sentences. The most widespread system, known as partner-assisted scanning, uses an alphabet spell board: the conversation partner points to letters or calls them out, the patient chooses one, and the process is repeated as needed. Spell boards are generally transparent; some include words and phrases, and the order of letters usually corresponds to their frequency in the language of conversation. Most, like the widespread AEIOU alphabet board [197, 230], imply holding it so as to enable the patient to look at it. Kopsky et al. [113] have recently tested a grid that is easier to memorize and must not be physically held; this facilitates eye contact and enables a “more natural” communication.

Persons in incomplete LIS can benefit from other communication methods (overview in [71]). No matter how small, movements of the head, finger, shoulder or mouth are essential in the choice of rehabilitation strategies [178], and can be exploited to move a cursor on a screen, surf the Internet, write email or generate speech via a voice synthesizer [132]. Other systems, such as eye-tracking, rely on the ability to control eye muscles [98], and can be supplemented by implanted BCIs thanks to which the patient can make “brain clicks” by trying to move a hand [215]. After losing his speech in 1985, physicist Stephen Hawking (1942–2018), who suffered from a slow-progressing form of ALS, became the most famous incompletely locked-in person relying on technology-based tools for augmentative and alternative communication (AAC). Though seldom associated with LIS, he was a prime example of the function of AAC systems in the quality of life and social integration of LIS patients.⁷ It is in this connection noteworthy that locked-in persons’ relatives do not locate their troubles in difficulties of communication or the patient’s physical

⁷ Hawking is on the Wikipedia “List of people with locked-in syndrome,” and the “continuity of his brilliant productivity” is said to confirm that locked-in persons “can be productive members of society” ([124], 497). Media coverage, however, does not usually associate Hawking with LIS, and neither does a major study on how he became an icon of the lonely genius and the disembodied mind [144].

limitations, but rather in administrative procedures, inadequate facilities, insufficient medical and paramedical staff, and a lack of emotional support [133].

The greatest hope for completely locked-in persons resides in the development of BCI (see [79] for a general introduction; for LIS, see [37, 81, 116, 117, 149, 151]). They will be able to participate in decision-making “by proclamation rather than proxy” ([88], 1853). In the most recent advance reported at the time of writing these lines, four ALS patients (two in permanent complete LIS and two entering the condition) learned to respond to personal questions with known answers as well as to open dichotomous questions by using frontocentral oxygenation changes measured with functional near infrared spectroscopy ([39]; questioned by [201]). Niels Birbaumer, a BCI pioneer, described the result, in which the voluntary modulation of brain activity is used to communicate, as “the first sign that completely locked-in syndrome may be abolished forever, because with all of these patients, we can now ask them the most critical questions in life” [190].

Illness Narratives

Yet what do we know about how persons with LIS envisage such critical questions and connect them to the extreme existential situation in which they find themselves? Rather little. QOL research and other work afford valuable information. A richer picture emerges from journalism (sometimes with interviews) devoted to persons with classic LIS (e.g. [6, 7, 54, 68, 97, 100, 143, 184, 224]), as well as from a recent collection of conversations [165]. The most absorbing and detailed materials are locked-in individuals’ autobiographical narratives – but only an extremely small number have been discussed.

The favorite has been *The Diving Bell and the Butterfly* [8], by Jean-Dominique Bauby (1952-1997). At the time of suffering a CVA in 1995, Bauby was editor of the French fashion magazine *Elle*; before his death, he founded the French *Association du Locked-in Syndrome* (www.alis-asso.fr). Bauby was able to blink with the left eye; he dictated his memoir, one letter at a time, after composing and memorizing his text day after day before engaging with his conversation partner. Bauby died two days after the publication of his book, which

became a great success.⁸ Jean-Jacques Beineix shot with him the documentary *Assigné à résidence* [10], and a decade later Julian Schnabel released the prize-winning film *The Diving Bell and the Butterfly* [192], which won international awards and was included in the BBC list of the best one-hundred movies of the twenty-first century.⁹

The Diving Bell is indeed an outstanding book of great literary quality. There are, however, about thirty first-person LIS narratives in Western European languages (a few titles in Italian, Spanish, English and French can be added to the list of 23 by Bruno, Laureys and Demertzi ([20], 206)); some are articles [40, 159, 160]. This corpus has never been approached as a totality, and only three books have been the object of analysis and commentary: Bauby’s, Tavalaro’s, and Philippe and Stéphane Vigand’s *Only the Eyes Say Yes* [220]. Bauby’s memoir (like Vigand’s, originally in French) has by far attracted the most attention; Vigand and Tavalaro have been discussed only in a comparison of the three books. Almost everything thus remains to be done to integrate LIS memoirs into the field of “illness narratives” and use them to inform a phenomenology of the syndrome.

Since the 1980s, the analysis of illness narratives has been incorporated into medical anthropology, medical education, and clinical practice to explore the social, relational and psychological dimensions of illness, raise empathic awareness of patients’ and caregivers’ experience, further communication, and help in healing and improving care and quality of life; closely connected to the emergence of medical humanities and the “narrative turn” in the human sciences, narrative approaches have also become prominent in biomedical ethics (e.g. [145]), and “narrative medicine” has become an institutionalized field offering degrees to professionals with a broad

⁸ Over 300,000 copies had been sold by January 1998 (<http://www.lepoint.fr/actualites-litterature/2007-01-25/majuscules/1038/0/88082#>); a number of Internet sources (e.g. <http://www.critiqueslibres.com/fr.php/vcrit/3864>) report sales of over a million copies and translations into 23 languages.

⁹ <http://www.bbc.com/culture/story/20160819-the-21st-centurys-100-greatest-films>. In my (admittedly limited) experience, many people have heard of LIS only through Schnabel’s film. They often relate it to Alejandro Amenábar’s *Mar adentro* (*The Sea Inside*, 2004), which is based on the life of Ramón Sampedro (1943-1998). For decades, Sampedro, who was not locked-in but remained quadriplegic in 1968 due to a diving accident, unsuccessfully fought for the right to commit assisted suicide in Spanish and European courts.

variety of backgrounds (see [35] for a concise discussion).

Illness narratives can be fictional or non-fictional, and be written not only by patients, but also by doctors and caregivers; “judging by present practice, any text in which illness plays a conspicuous part can count as an illness narrative” ([217], 388; also [92]). The emphasis is naturally placed on *illness*, i.e. the lived, subjective experience of a biological process, rather than on *disease*. It is with this distinction in mind that medical anthropologist Arthur Kleinman [108] coined *illness narrative* in the groundbreaking book that used it as a title. Although lived experience encompasses much that does not enter a story, narrativizing it may help suffering persons reconstruct their identity and their subjective and intersubjective worlds ([77], ch. 6). Many accounts are “narratives of triumph” [47]; the better ones, however, avoid the trap of “positive thinking,” and can be seen as contributions to the philosophical tradition of doing ethics on the basis of lived examples [103].

Insofar as illness narratives concern subjective experience, they have intrinsic phenomenological value. It is therefore not surprising that, so far, the most direct contributions to the phenomenology of LIS are close readings of locked-in person’s testimonies – even if their aim is to explore *by way of LIS* themes in philosophical anthropology, philosophy of mind or biomedical ethics. Perhaps precisely because a phenomenology of LIS is not their main objective, they do not connect to relevant knowledge gained by other means. In any case, the fact that we meet narrators and protagonists of a story rather than flesh-and-blood people defines the reach and limitations of narrative analysis.

Phenomenological Readings

Richard Zaner focuses on the narrator of *The Diving Bell and the Butterfly* alongside Joe Bonham, the fictional protagonist of Dalton Trumbo’s anti-war masterpiece *Johnny Got His Gun* [214]. A young soldier in World War I, Joe loses his arms, legs and face, including eyes, ears, teeth and tongue in an explosion. His mind, however, functions perfectly. Kept alive by means of a tracheostomy, he communicates by banging his head on his pillow in Morse code. Zaner treats Joe as the exact equivalent of Bauby. This implies ignoring differences between author and narrator, or “real” and fictional character, but highlights that both stories are written in the first person from within a locked-in state. Bauby and

Joe are therefore unlike two literary characters who have been “diagnosed” with LIS [82], Monsieur Noirtier de Villefort in Alexandre Dumas’ *The Count of Montecristo* (1844–45) and Madame Raquin in Émile Zola’s *Thérèse Raquin* (1867), who are mainly depicted from the outside by an omniscient narrator.

Zaner underlines the “unbridgeable distances” that LIS creates between the locked-in person and others when the body ceases to be the “communicating vehicle” we take for granted ([231], 190, 191). Yet because those persons are nonetheless embodied, their condition raises such questions as, “What are the limits beyond which a human body can no longer embody a human life?” and “What is the minimal wherewithal for embodiment?” (ib., 195). In spite of their plight, Bauby and Joe remain communicative beings who desire recognition as such, and thus call attention to the ontological phenomenon Zaner places “at the heart of human life,” namely “the being of Self” as “being-with-another-Self in the most concrete, immediate and intimate ways of the flesh, where touch seems the most dense and significant” (ib., 204, 205).

Zaner’s article illustrates how, in skillful hands, illness narratives illuminate general issues in philosophical anthropology. In turn, by highlighting issues of embodiment and communication, the philosophical gaze can guide approaches to the real-life experience of LIS. It also leads to a reflection on narrative itself. Precisely because encountering real persons is so different from reading about them, illness narratives allow readers “to know subjectively” what many of those who meet concrete patients cannot perceive or understand. Readers, Zaner notes, “know better,” and may feel more empathy than those who materially interacted with Joe and Bauby: “Perhaps, too, we attain the truth of the matter only by stories” (ib., 200).

In her article on *The Diving Bell and the Butterfly*, published while she was Zaner’s student, Denise Dudzinski argues, “Bauby describes two different selves shaped by two different bodies, one before and one after the stroke” ([58], 34). She detects in the narrative two phenomenologically relevant aspects of physical illness: “the alienation that occurs when one no longer recognizes one’s body as one’s own,” and “the creativity and freedom that persists through this changing identity” (ib., 35). Bauby, Dudzinski observes, conveys “the feeling that his paralytic body is not *he*,” the disconnection between self-identity and embodiment, and the sense that “his body is no longer useful to him” (ib., 36, 42).

There occurs in the locked-in person a breakdown of what the French philosopher Gabriel Marcel (1889–1973) called the *noyau senti*, whereby “I am implicated in my body and my body is implicated in me” (ib., 43). Bauby tries to recapture this “felt kernel” by way of memory, imagination and narrative.

Dudzinski’s educational use of *The Diving Bell* involves new phenomenologically relevant observations about communication, autonomy and vulnerability. She notes, for example, how Bauby is “buoyed and empowered by his relationships” and how correspondence with friends and outings with family become for him “precious liberties” ([59], 242). LIS manifests “the irony of autonomy,” which rests on paying vulnerability its due and “on others’ willingness to listen and connect with us” ([60], 603). Bauby, Dudzinski (ib., 605) concludes, thus reminds us “that what we owe patients is not respect for autonomy *per se*, but respect for persons.”

In 2009, Yumiko Kawaguchi, then president of the Japanese ALS Association, published in Japanese *Maybe She Just Wants to Breathe: An Everyday Life of an ALS Patient and Her Family*. The patient was her mother: diagnosed with ALS in 1995, she was placed under artificial respiration at home in 1996, entered the total locked-in state in 1999, and died in 2007. In a crucial episode, Kawaguchi describes how her mother’s body became expressive after she (the mother) said her two last words with her eyes: the capillaries then became eloquent, and sweat told “the real feelings.” Such bodily-mediated communication is the focus of a subtle reading by the University of Osaka philosopher Yasuhiko Murakami [147].¹⁰

Framing his analysis within the *Körper-Leib* distinction, Murakami observes that in complete paralysis, the sense of corporality associated with *Leib* tends to disappear, and the totally locked-in person connects with the world through the caregiver’s body. At this point, a “dialogue of the body” without ordinary communication emerges. Different kinds of sweat, the temperature and coloring of the skin, or changes in blood pressure sustain this unusual discourse that is not truly intersubjective (since there is no intention to transmit contents and no thoughts are shared), but resides in the perception of the material body: “the care of the body itself

becomes communication.” Kawaguchi reports that the “world of silence” she had feared when her mother entered total LIS did not materialize, and that the “dialogue with the body” continued until the end. Murakami interprets this process as an example of how life is “phenomenologically revealed” in the experience of the other. Like Zaner or Dudzinski, he does not pursue a specific phenomenology of LIS. He nonetheless demonstrates the philosophical and anthropological significance of first-person testimony, and illuminates phenomena involved in patient care when communication via movement or BCI is lacking.

Narrating the Unspeakable

In 2001, a group of authors, including a historian of medicine, two scholars trained in literary narrative theory, and two using narrative analysis in qualitative research in the health and social sciences, published a set of readings of *The Diving Bell and the Butterfly*. “Narrating the Unspeakable,” as the collection was titled, is the most sustained piece of scholarship focused on the analysis of a LIS memoir. Although its main goal was to study narrative form, it calls attention to issues of phenomenological relevance and, as all the other materials discussed here, gives salience to communication.

Valerie Raoul underlines Bauby’s “self-performance in the text” ([177], 186). Made up of twenty-nine fragments including descriptions, reflections, memories, dreams, complaints and fantasies, *The Diving Bell* embodies the leitmotiv of “biographical disruption” [24], while at the same time constructing a character who in fundamental ways remains the humorous and seductive “former Bauby” ([177], 188).¹¹ James Overboe in turn argues that Bauby sees his life “as a continuation of his existence as a desiring person” who communicates “an experience that is usually considered incommunicable,” and thus prevents other “from perceiving him as a nonhuman kept alive by medical technology” (ib., 190). Confronted with LIS, people may assume that if there is “no vehicle of communication,” then “no one is there” (ib., 192). Bauby retorts with an entire book that validates his ontological status and the value of his lived experience. Similarly, Connie Canam remarks that what distinguishes the individuals who try to decipher Bauby’s attempts to

¹⁰ I thank Dr. Kawaguchi for providing me with an English translation of two chapters from her book [101]. Murakami translates the title differently, as *Le corps qui ne meurt pas – Vivre la vie quotidienne du SLA (The Body That Does Not Die: Living Day to Day with ALS)*.

¹¹ Williams [227] updates the notion of “biographical disruption,” emphasizing that it is only one of the ways in which chronic illness can become integrated into a person’s life.

communicate from those who do not is whether they relate to him as a person. The latter “deny him a voice and depersonalize him as a ‘case;’ they unwittingly send him the message that they see no person in his body” (ib., 194–195). Communication and relationality safeguard the body, but above all personhood itself.

Carla Paterson next remarks that Bauby barely uses the current vocabulary of neurological impairment, and hardly mentions the methods and machines used to manage severe paralysis (ib., 197). Instead, he draws attention to traces of the history of the rehabilitation facility where he lives, and brings them together with evocations of his own past; he thereby escapes a present moment whose most unbearable feature is the “inability to communicate with his fellow inhabitants” (ib., 199). Finally, Gloria Onyeoziri notes that the irony of Bauby’s narrative is that the diving bell of his inert body supports the flight of the butterfly: “His one eye can still see, yet it is the eyelid that covers it that enables him to communicate his observations and gives him a powerful ironic voice” (ib., 203). In conclusion, *The Diving Bell and the Butterfly* draws attention “to the potential for people with LIS to communicate and to their need to be recognized as people by health professionals and others” (ib., 206). Such recognition can take different forms, and, ultimately, values rather than disciplines decide the weight readings choose to give to “advocacy, therapy, aesthetic effect, or commercial success” (ib., 205). This reflection concerns the methodology of studying illness narratives; but it also applies to phenomenology as an intellectual endeavor.

Questioned Consciousness

Henry C. Stewart’s unpublished dissertation on “questioned consciousness” in Bauby, Tavalaro and Vigand is the only study so far to undertake a comparative reading of LIS first-person narratives.¹² “Questioned consciousness” refers to the fact that the narrators were considered for some time as lacking consciousness – for as long as 6 years in Tavalaro’s case.¹³ Stewart hopes that

¹² I thank Dr. Stewart for kindly sending me a copy of his dissertation.

¹³ Born in 1958, Philippe Vigand entered the locked-in state after suffering a stroke in 1990. He lives at home and communicates via computer or a blinking code. At the time of the CVA, he and his wife Stéphane had two small daughters; a son was born in 1992. His memoir *Only the eyes say yes: A love story (Putain de silence)*, 1997, was published the same year as Bauby’s and Tavalaro’s; one half was written by him, the other by Stéphane. Philippe Vigand has since then published three other books. The lives of Bauby and Tavalaro have been sketched above.

an engagement with illness narratives will improve diagnosis and care, and has used Bauby in his teaching [205]. His purpose, however, is to identify commonalities across the chosen memoirs, and bring narratives of questioned consciousness into the literary “canon of disability” ([206], 34).¹⁴ Although he mentions the significance of writing methods, he does not explain how they influence the final products and stays focused on contents.

The main common literary device Stewart identifies is humor (e.g. [206], 29). The narrative episodes of comic relief might be related to the patients’ “surprising degree of optimism” (ib., 17). (It is perhaps not surprising to find optimism in three persons who look ahead in their lives and choose to write about themselves. However, they are a minority. Most patients do not write memoirs, and do not even respond to surveys. As we shall see later, this bias implies a major challenge for the study of LIS and chronic illness in general.)

Other common themes include dreams and memories, dependency, self-perception and the perception of others, hope, a motivation to proceed with one’s existence, emotions (anger, rage and frustration, but also appreciation, love and gratitude), mistrust of the medical establishment, interactions with medical personnel, and life in institutional settings. Again, however, the most salient motifs are communication, personal identity, and the body.

Communication always represents the most urgent need. The “terrifying realization” (ib., 17) the locked-in face when they emerge from coma is that they are unable to communicate. Such inability and the resulting isolation is the most immediate source of frustration, anxiety and desperation; surmounting it becomes their major motivation, and its difficulties, their main challenge. The three authors “enthusiastically discuss the crucial first moment of communication with others,” and “seem extraordinarily grateful to both the medical staff and the family members who enable them to communicate” (ib., 28). Communication is essential not only for pragmatic, but also for existential reasons. Bauby, Tavalaro and Vigand “want more than to declare their humanity; they also want to announce that they are

¹⁴ The analysis of commonalities occupies the entire chapter 2 of the dissertation. Stewart does not discuss the relationship between fiction and non-fiction, but plays down the difference, for example when he writes that, of various characters, “Bauby and Bonham are the most amusing” (2012, 31).

the same humans they were prior to their injuries” (ib., 19). No matter how impaired and unrecognizable, they proclaim that “the person remains” (ib., 22).

The narratives suggest the constitutive significance of embodiment and relationality for personhood and personal identity. As Stewart (ib., 26) aptly remarks, the mind may be the seat of the sense of self and the source of the locked-in persons’ communicative drive, but their bodies is what we first encounter in their writings. The narrators long to transcend a body that remains sentient; they all report pain and situations (Bauby, for example, on stuck eyelashes or a fly on the nose) that become tormenting moments. The bodies with which they no longer identify are nonetheless theirs; poignant narrative episodes relate occasions in which they discover them directly in a mirror, or indirectly, in other people’s gaze. In short, Stewart’s reading underlines locked-in persons’ sense of personal continuity, as well as the role of relationality and embodiment for their experience of the self and the meaning they attribute to life. This resonates with work done from other points of view.

Philosophical Engagement with LIS

We mentioned already the central role of consciousness in defining personhood and determining ethical obligations toward patients, and how that implies that individuals diagnosed with LIS are persons. Nevertheless, as LIS narratives demonstrate, from the point of view of the experience and empirical conditions of personhood, LIS patients are not mere minds imprisoned in paralyzed bodies. The concrete circumstances of locked-in embodiment are for them a source of suffering, but also confirm their ontological and moral status. Personhood in their case is enabled through relationships with human agents and non-human apparatuses. Does that real-life situation inform a philosophical understanding of personhood? The agony of incommunicability and the pain of not being able to touch their loved ones is a recurrent theme in LIS testimonies. Is relationality then constitutive or incidental to selfhood? Communication may be “crucial for personhood because it allows a sense of agency” [154]. But does that mean that there is

no personhood without communication or intersubjectivity? And what about embodiment? What does it mean that locked-in persons tend to report a sense of continuity in their identity in spite of massive bodily changes? Does it substantiate the widespread belief that “we are our brains” [219]?

Philosophical engagement with LIS around such questions has been limited. The most sustained attempt is due to Miriam Kyselo, who defends the perspective known as *4E-cognition*, with *E* standing for *enacted*, *extended*, *embodied* and *embedded*. For Dudzinski, Bauby’s case demonstrated that an altered bodily condition transforms a person’s identity, and splits it into two eras – before and after the stroke. Kyselo [121], in contrast, argues that Bauby’s self-alienation results mainly from “an altered dynamical relation to other people.” It is not because the individual is embodied that a disturbance of the body is a disturbance of the self. Rather, the self “is *indirectly* affected by impaired embodiment, in that the disability impacts the individual’s capacities to engage with the social environment;” thus, the “experienced alienation of the body” results from an altered social interaction process (ib., 56, 57). It also follows that autonomy relies “on communicative (re)actions of the social environment” ([122], 589).

Kyselo’s goal is not to understand LIS itself, but to test by way of LIS different approaches in embodied cognitive science [123]. Nonetheless, the analysis that leads her to attribute a central role to intersubjectivity and the social environment directs our attention to the interactive, relational and communicative mechanisms involved in locked-in individuals’ experience. LIS, Kyselo [121] concludes, can be considered a “social injury” because, rather than affecting the self directly, it “impacts on bodily and socially mediated action, which are both necessary for the maintenance of the self.” Similarly, the constructivist interpretation of LIS emphasizes the connection between patients and their human and non-human environments, and argues that autonomy, as a constitutive property of personhood, is an “effect” of fragile relations among persons and between persons and machines ([56]; see also [62] and related arguments in [83, 152], and [223]).

With regard to LIS specifically, the debate over these interpretations still has to take place. Whether, how or to

what extent LIS questions standard accounts of personhood or selfhood is an open question. Overall, however, there are two main ways of articulating philosophy and an existential situation such as LIS. On the one hand, as in the enactivist and constructivist interpretations just sketched, LIS can be used to illustrate or test philosophical theories. On the other hand, LIS as a real-life situation can inform philosophical inquiry. For instance, the survey on bodily changes and personal identity that is summarized below contrasts patients' self-report "from the wheelchair" to philosophers' perspective "from the armchair" ([153], 432). These two stances need not be opposed to each other: philosophical analysis may profit from empirical knowledge about LIS, and the survey questionnaire is partly framed by philosophical concerns. One can give more or less weight to philosophical theory or to social-science methodologies, while making them interact and considering them as complementary investigative strategies.

Personal Identity and Quality of Life

There is a considerable gap between patients' perceptions of their lives, and notions held by healthy people and medical professionals.¹⁵ Similarly to what happens with respect to DOC, which are judged "worse than death" ([80]; see also [104, 105, 131]), there is a tendency to think that life in LIS is not worth living and to underestimate how locked-in people rate their circumstances. While healthy controls assert that they would not want to live in a locked-in state [153], a high proportion of the LIS patients who respond to surveys report subjective well-being and a meaningful quality of life, and as a population display low rates of depression (though higher than in controls according to some studies), suicidal thoughts, euthanasia requests, and do-not-resuscitate orders ([18, 19, 48, 53, 55, 124, 134, 167, 185, 186]; see also [228], on an individual case). The contrast between healthy people's judgment and

¹⁵ "Quality of life" is by definition based on self-assessment. The World Health Organization defines it "as an individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns," and adds, "It is a broad ranging concept affected in a complex way by the person's physical health, psychological state, personal beliefs, social relationships and their relationship to salient features of their environment." <http://www.who.int/healthinfo/survey/whoqol-qualityoflife/en/> Last accessed on 4 October 2018.

patients' self-assessed QOL has been named the "disability paradox" [2]. However, as Limburg, Pols and Limburg [129] point out, the paradox is only apparent: while medical staff tends to assimilate QOL to functionality and healthy people cannot imagine living with such a disorder, patients tend to focus on their actual possibilities and their assessment changes with time (those who have spent longer in LIS report better QOL than recently diagnosed individuals); in short, QOL has many dimensions, and is, as they put it, "a changeable and dynamic concept."

Disability paradoxes embody different interests and viewpoints, or even contrasting understandings of subjective experience and the meaning of words. For example, the reputed neuroscientist Antonio Damasio [50] claims that a "remarkable feature" of LIS is that locked-in persons "do not experience the anguish and turmoil that their horrifying situation would *lead observers to expect*," they have "a considerable range of feelings," but "do not report the terror that *one imagines would arise* in their horrible circumstances," and "do not seem to have *anything like the acute fear experienced by so many perfectly healthy and mobile individuals* inside a magnetic resonance scanner, not to mention a crowded elevator" (my emphasis). Damasio's "surprising finding" about an alleged lack of anguish, terror and fear is abundantly contradicted by locked-in patients' first-person narratives. Indeed, as Birbaumer [15] notes, "subjective emotional experience is obviously not changed by complete paralysis." In addition to being empirically unfounded, Damasio's observations about fear in LIS combine typically "disability paradox" expectations with a singularly limited conception of fear.

In addition to what they reveal about locked-in persons' QOL, surveys and narratives suggest that mind as pure cognitive function is for them of little value without a sense of being supported by the environment, a certain measure of perceived control, and especially the capacity and opportunities to communicate and relate to others [33, 196, 198, 229].¹⁶ QOL surveys throw valuable indirect light onto the phenomenology of LIS, but there is practically no research specifically devoted to it – as there is, based on testimonies and interviews, with persons who suffer other neurological impairments of

¹⁶ These factors, however, do not suffice to sustain the desire to live; an example is the well-publicized case of Tony Nicklinson, an ALS locked-in patient who let himself die after losing his legal battle in favor of assisted suicide [22].

movement and sensation (e.g. [44, 45]). Views about “lost” and “shattered” selves betray the assumption that these patients are dispossessed of their sense of personal identity. The patients themselves, however, report feeling loss and discontinuity as regards the public dimension of the self, but interior continuity [72, 142].

The only study that directly concerns these issues in LIS was carried out by Marie-Christine Nizzi and members of the Coma Science Group at the University of Liège. Their goal was to investigate how the massive bodily changes involved in LIS are integrated in individuals’ personal identity. They used a fifteen-item closed-ended questionnaire organized as a four-point Likert scale. The questionnaire had three parts: *The accident and my identity*; *My body and me*; *My life and me*. Each included five statements, with which subjects had to *totally agree*, *agree*, *disagree* or *totally disagree*. Most respondents felt essentially the same as before entering LIS, reporting a continuous experienced identity when they accepted their bodily changes, and a discontinuous one “when they reject the new body” ([153], 435). The paralyzed body, the authors conclude, “remains a strong component of patients’ experienced identity,” and “patients can adjust to objective changes perceived as meaningful” (ib., 431). A control group of healthy medical professionals well acquainted with LIS was asked to predict the patients’ responses; in line with what happens in other domains of disability, they failed.

The study by Nizzi et al., so far the only one of its kind, nuances on an empirical basis the narrative and philosophical analyses: entering LIS constitutes a fateful biographical disruption, but it alters the relationship to embodiment and personal identity in different ways in different individuals. Details about how and why body and being remain connected or dissociated in patients’ self-perception, or how LIS is experienced as an individual and as a social injury remain to be investigated.

LIS in Neuroethics and Biomedical Ethics

Insofar as locked-in individuals are fully *persons*, LIS does not give rise to the ethical dilemmas and procedural challenges that are customary in connection with VS and MCS. Contrary to VS and MCS patients, persons in classic and incomplete

LIS can understand explanations, report on their quality of life, give their opinion, express their choices, assert their wishes, and give or refuse consent – all that, provided they are equipped with communication-enabling tools. Only by communicating can they exert the autonomy that the United Nations Convention on the Rights of Persons with Disabilities considers constitutive of personhood. That is why, in their discussion of how locked-in persons are “locked out,” Johansson, Soekadar and Clausen ([95], 563) rightly note, “The moral responsibility toward persons in LIS consists in enabling a functional way to communicate.” This is a very substantial ethical and technical undertaking – one that Joseph Fins [64] has richly documented in case-studies where communication emerges as crucial in connection with the role of families, the recognition of personhood, societal integration, and the possibilities and limits of receiving care and financial support. That undertaking, however, does not imply rethinking the concept of personhood the UN Convention exemplifies. Such a situation is reflected in the place of LIS in neuroethics and bioethics overall.¹⁷

LIS sometimes appears only as a “confounding neurological state” in the context of diagnosing DOC ([175], 144, 149). Going further, Walter Glannon explains that whereas “patients in a permanent VS no longer exist as persons” and MCS patients lack the capacity “to adequately consider the reasons for or against life-sustaining treatment,” locked-in individuals do not suffer such incapacitating “cognitive impairment” ([75], 172, 157, 167, 170). They may vary in how they judge their situation and make different choices about their life, but there is no doubt that they judge and choose with the autonomy attributed to persons. While classic and incomplete LIS pose no problem in such a perspective, totally locked-in individuals may go unrecognized as persons. Arguably the chief leitmotif of writings on LIS, communicative capacities are not

¹⁷ LIS surfaces more marginally in objections the whole-brain definition of death, which emphasizes the role of the brain in maintaining the integrated functioning of the organism as a whole (the cessation of which defines death). It is noted that some integrative functions do not require the whole brain, and that the brains of LIS patients “appear to have no more integrative effects on their bodies than those of brain-dead individuals, and yet locked-in patients are undoubtedly alive” ([210], 531). These two objections have not led to the abandonment of the whole-brain criterion, but moved it away from justification on the basis of the brain’s integrative function.

merely practical tools for establishing patients' preferences and assessing their quality of life, but crucial for actualizing, corporealizing and ascertaining personhood. As Albert Robillard [181] demonstrated, refusing to use a patient's alphabet board amounts to denying his individuality.

LIS is also mentioned, as usual, in standard presentations of DOC [36, 66, 162, 225], or introduced as precisely *not* being DOC [65, 76]. Informed consent in LIS is also discussed, and it is considered "reasonable" for surrogates of totally locked-in persons to authorize interventions, such as brain implants, that may restore communicative ability, "even if they involve significant risks" ([107], 131; see also [87]). The standard position is that "it is hard to raise ethical objections" to attempts "to provide a locked-in patient with a means to communicate with the outside world" ([70], 196). The situations involved are technically challenging, but they assume, rather than lead to pondering, their status as persons.

A presentation of methods for detecting levels of consciousness in DOC recalls that LIS patients report a positive subjective QOL and considers them "a nice control population" ([52], 41). A more substantial discussion of the ethical implications of BCI emphasizes that advance directives are usually prepared long before the medical interventions they rule out might be needed [199]. They may therefore embody a self-applied paradox of disability, and contradict the wishes the patient would have at the time the illness appears and develops. That is why, at least within the German LIS Association, most respondents declare themselves happy not to have written advance directives.¹⁸

Such a situation parallels the fact that informed consent represents a dominant theme in discussions of the ethical aspects of BCI [23]. Yet it is not unique to total LIS, and raises challenges akin to trying to detect covert awareness in individuals who might have been misdiagnosed as VS [1, 38, 182, 183]. The above-mentioned possibility of communicating by a voluntary modulation of brain activity has inspired calls for neuroimaging to be made available to all VS patients [25, 93]. Such a demand would also apply to persons with total LIS, whose condition has inspired speculation

about what people with covert awareness might respond to questions concerning their wishes and QOL [78]. The ultimate goal is to ascertain a person's wishes about life and death *after*, not before she becomes a patient.

In this context, LIS and DOC raise complementary problems; as philosopher Alva Noë [155] put it, "Whereas with locked-in syndrome we are challenged to believe that behind the masklike wall of a face there is a lively intellect at work, with persistent vegetative state we struggle to take seriously the thought that there is an absence of feeling and subjectivity behind what moves us as an expressive face." Comparisons are philosophically and ethically illuminating, but amalgamating LIS and DOC may lead to misunderstandings. Thus, in a survey of German neurologists concerning diagnostic knowledge, attitudes toward limiting life-sustaining treatment and the care of DOC and LIS patients, the median QOL of the locked-in person was assessed as equivalent to that of VS patients, and rated lower than that of MCS patients – about whose QOL actually nothing is known; moreover, in spite of what studies show about LIS patients' QOL and preferences, the survey revealed similar attitudes toward limiting life-sustaining treatment for the VS and the LIS groups [120]. Thus, although establishing a prognosis of functional outcomes and determining the patient's wishes about treatment were, reasonably enough, seen as the most ethically challenging issues in all conditions, the imagined proximity of VS and LIS entails a grievous risk for locked-in persons.

To my knowledge, the most comprehensive discussion of the bioethical dimensions of LIS is to be found in two articles by James L. Bernat. In 1990, Bernat dealt with LIS through the examination of a case in which he was involved as treating physician and ethics consultant. As he explained at the time, ethical problems with LIS patients result from the unique features of the syndrome: communication is extremely difficult; since pain sensation remains intact, they can suffer intensely; they can hardly manipulate their environment effectively; and they can be misdiagnosed as lacking consciousness. Because their cognitive capacities are intact, they must be treated according to "the same standards governing decision making by any competent patient" ([12], 93). If the consent process is adequately conducted and the patient is not clinically depressed, a consistent wish to die and refuse medical treatment should be considered "a rational

¹⁸ See the documents available under <http://www.locked-in-syndrom.org/patientenverfuegungen.htm>, as well as the article "Patientenverfügung aus Sicht der Betroffenen." www.locked-in-syndrom.org/presse-not-1-2009.pdf. Last accessed on 4 October 2018.

decision;” hence the United States court decisions of 1987 allowing two persons with LIS to decline life-sustaining measures, which confirmed the “overriding importance of the rights of competent patients to consent or refuse proposed treatments” (ib., 95, 97).

Almost three decades later, much more is known about QOL, and there has been great progress in AAC systems; yet the ethical considerations remain basically the same. QOL and technology go hand in hand, since, as Bernat [14] notes, the inability to communicate adequately is the chief source of suffering in LIS patients, and communicative capacity has an enormous positive impact on their attitude and wellbeing. Prudence is of course imperative: only repeated and detailed conversations using systems that go beyond binary responses will allow physicians to be sure of a patient’s choice, and doctors should be well informed about patient satisfaction and QOL data to avoid negative biases toward life in LIS. As Tola-Arribas [212] points out, ethical dilemmas arise before therapeutic action is undertaken, and without such precautions, any decision runs the risk of being contrary to the patient’s wishes.

With respect to treatment and end-of-life decisions, LIS (regardless of etiology) is only the most profound of the several conditions that can lead to states of impaired communication while sparing cognition [141]. The ethical imperative always consists of trying to establish communication, ascertaining decision-making capacity, informing patients of their condition, prognosis and clinical course, treatment options, and right to choose whether to pursue or withhold treatment. As some cases concretely illustrate, in the light of existing studies about locked-in persons’ preferences, and given that most individuals who enter LIS due to a brainstem stroke are relatively young and cognitively intact, it is advisable not to try to reach decisions about palliation, conservative medical management or end-of-life during the acute period [4].

As for end of life, only 19% of the respondents to a 1995-96 survey of 93 German physicians working in intensive care units found active euthanasia defensible in cases of LIS, while 99% declared passive euthanasia permissible [211]. The debate remains open. The legal possibility of intentionally terminating life at the request of an adult patient has been advocated as a means of

respecting autonomy and individuality [110, 111], and rejected as potentially leading to the premature death of persons who might be depressed and not yet adapted to their new circumstances [15, 118]. The fact that locked-in respondents to surveys “report being happy with their present lives” is indeed “an important message that should be taken into account in the debate about end-of-life” ([5], 131). In his substantial discussion of “the neuroethics of being locked-in,” Birbaumer [15] suggests that the ethical problems concerning end of life in LIS may resolve themselves in the light of empirical knowledge. By the very nature of ethical debates, this seems unlikely. Each person has a different view of what would be a “living hell” – and, it has been argued, each should be given the right to end it [191].

Toward a Phenomenology of LIS: Some Suggestions

A systematic phenomenology of LIS would draw on the areas sketched here. While some methods deal directly with locked-in persons’ subjective experience, others throw light on its psychological, sociocultural and materials conditions. It will therefore be most productive to maintain a methodological and interpretive pluralism, and to use and compare approaches and results in an eclectic and pragmatic manner.

Illness Narratives

Locked-in individuals’ first-person narratives emerge as a major source, to be analyzed singly, as a corpus, and in intercultural comparison. Using them for a phenomenological apprehension of LIS will incorporate them into the broader field of illness narratives. Insofar as they will be treated as a corpus, they should be handled according to the principles of narrative analysis [179], in steps such as those described by Råheim and Håland [176] – gradually identifying themes and subthemes, comparing forms and contents, defining broader categories, and elaborating a view about the authors’ experience. The approach, though inductive, will be informed by questions ranging from care to philosophy, as well as by existing readings of narratives by persons suffering from other chronic and neurological conditions (e.g. [44, 91, 200]).

It will be important not to lose sight of who speaks in illness narratives. In a first approximation, we hear the voice of those who sign the texts and designate

themselves as *I*. However, even a first-person protagonist-narrator is a character in a story, and therefore different from the author. These distinctions underline the limits of illness narratives as a source for phenomenology. Zaner [231] is right to note that an encounter with Bauby or Trumbo's Joe Bonham "is decisively different from reading about them." Hence the need to combine narrative analysis with other empirical tools: questionnaires, interviews, observation. The locked-in individuals who publish first-person narratives are a minority within a minority; their experience cannot be simply generalized. Moreover, as we saw, different readers reach different conclusions.

Beyond Surveys

The corpus of published narratives can be supplemented by encouraging first-person storytelling with the help of such tools as the McGill Illness Narrative Interview,¹⁹ or with open-ended questions such as the ones we are using in our ongoing research project [218]. The unique investigation led by Marie-Christine Nizzi with French-speaking patients could be carried out in a variety of national and linguistic contexts. Moreover – and this to my knowledge has not been done – one should distinguish and compare groups according to etiology. The experience of persons who find themselves in LIS suddenly must differ significantly from the experience of those who enter the locked-in state gradually. Robillard [181], for instance, explains how the lip-signing system he devised with his wife changed with the gradual loss of facial musculature control and the ability to mouth recognizable words. Other examples could be given. In general, the sense of biographical disruption is unlikely to be same; beyond individual differences, the etiology conditions illness trajectories [43]. For stroke patients, the accident is the "zero hour" ([28], 13), followed by the realization of finding oneself in an irreversible situation. For ALS patients, in contrast, the diagnosis can be reached after months of medical exams, and in some cases, that means patients are prepared and do not sink in despair ([135], 15). Moreover, the nature of a condition that advances more or less quickly, but inexorably, makes a crucial difference. As the historian Tony Judt (1948–2010) put it in his beautiful memoir, "In contrast to almost every other serious or deadly disease, one is

thus left free to contemplate at leisure and in minimal discomfort the catastrophic progress of one's own deterioration. In effect, ALS constitutes progressive imprisonment without parole" [96].²⁰

In addition to narratives, seeking the phenomenological "density" that surveys lack demands qualitative methodologies of the ethnographic kind, involving conversations with patients and caregivers, as well as in situ observations of interpersonal dynamics. This might also help elucidate the self-selection bias that has brought about the perception of a "happy majority" [19]. Response rates vary (14% in [194], 32% in [153], 43% in [132], 54% in [167]). The "majority," mostly, is therefore neither happy nor miserable, but silent. Even if such bias is typical of survey research when respondents decide for themselves if they want to participate [158, 204], it would be important to find out why most locked-in persons do not respond, and what they think and feel.

Intercultural Comparison

Opportunities for intercultural comparison are limited: they require distinct but comparable contexts, and an accessible population of locked-in persons. Japan emerges as the best choice. First, there is in Japanese a large number of published LIS and ALS patients' narratives [208]. The existence of this corpus is connected with the social and material conditions of support for patients – which should also enter the comparison. Second, there are well-organized institutions through which LIS patients can be contacted. Third, the level of development of the health system and the medical and care professions, as well as of such fields as biomedical ethics and medical anthropology and sociology, is commensurate to that of Western Europe and North America. Comparisons would thus take place among comparable contexts.

At the same time, Japan offers significant cultural differences of potential relevance for a phenomenology of LIS. It is fair to conjecture that, with regard to subjectivity, corporality and the self, "Western" self-presentations, and real-life situations themselves, are partly structured by a stress on individual autonomy and such dichotomies as body-mind or self-other.

¹⁹ <https://www.mcgill.ca/tcpsych/research/cmhr/mini>. Last accessed on 30 July 2018.

²⁰ Quoted from the e-book text, therefore without page number. The quotation comes from the beginning of ch. 2, "Night," originally published in the *New York Review of Books* of 14 January 2010.

Japanese ones, in contrast, may reflect distinct features, such as an emphasis on the solidarity of wholes and parts (including individual, family and society), an understanding of mind (*kokoro*) as a unity of the affective, the sensitive and the rational, a perception of the body as a whole (*mi*), or the sense of a mind-body totality (*shinshin ichinyo*). (See on these concepts [84, 138], and [163] on how they can contribute to rethinking the sociology of the body.) The existence of a Japanese tradition of illness narratives (*tōbyōki* or “account of a struggle with disease;” [139, 221]) and the Japanese debate on brain death [130] suggest the weight of those cultural dimensions, which are likely to manifest themselves in narratives, attitudes, and intersubjective dynamics.

Philosophy and Biomedical Ethics

The limited attention that biomedical ethics devotes to LIS corresponds to the primacy of consciousness and autonomy as criteria for personhood and for defining obligations toward patients. Over three decades ago, in what is perhaps the first discussion of the bioethics of LIS, the *Hastings Center Report* published two opinions about Mr. B, a locked-in man who lacked a living will and had provided no clear basis for substituted judgment [203]. Mr. B could use a yes/no blinking code, but his capacity to respond was unstable, and he proved incapable of answering questions with an emotional content. Grant E. Steffen, a practicing internist, argued that under the circumstances, and given Mr. B’s prognosis and quality of life, life-sustaining therapy should not be provided. In contrast, Cory Franklin, director of an intensive care unit, held that the difficulty of ascertaining Mr. B’s wishes was no ground to withhold support. Although an opinion as categorical as Steffen’s has been rendered less acceptable by better knowledge of patients’ QOL and advances in care and AAC technologies, the questions and conceptual and practical challenges raised by LIS remain. Indeed, precisely because of those advances, they will increasingly require reflection and decision-making not only at the individual, but also at the societal level.

Judgments about personhood imply views about the connection of the mental, the bodily, the relational and the social. Which functions are necessary and sufficient for someone to be a person? When and under which circumstances does an organism begin and cease to be a person? To what an extent is personhood an inherently

individual quality and to what extent does it depend on relationality and community? These questions sound abstract, but how they are investigated and solved turns them into objects of public debate, contributes to shape medical practices and public health policy, to steer legislation, and to direct research in biomedicine and the human sciences. The existential situation of LIS could help rethink them.

We noticed that whereas self-consciousness and autonomy have been considered sufficient and necessary for a normative definition of personhood, communication and relationality seem constitutive of its empirical accomplishment. From Antiquity to the present, the philosophy of personhood and personal identity has comprised three main interrelated issues: *essence* (the necessary and sufficient attributes that define the quality of person), *individuation* (that which differentiates one person from another), and *re-identification* (the properties that make me the same person over time in spite of change). The metaphysics of personhood has given more or less weight to physical and psychological criteria, and the human sciences have argued for a more constitutive role for intersubjectivity and technological systems. Locked-in persons’ experience invites us to explore these issues by turning the usual viewpoint around – asking what LIS can do for theories rather than what theories can do for LIS. In such a framework, LIS would have to be examined together with conditions, such as DOC and dementias, which seem to problematize personhood more directly.

The law has already offered a limited but significant arena of debate. In 2000 and 2006, two Spanish locked-in persons who had been deprived of their civil rights, specifically the right to vote, reclaimed them in court [57]. Rights were given back to the patient who had recovered mobility of a finger and become able to communicate via computer; they were refused to the other, who could communicate only by blinking and depended on a human partner. Only the system made up of a human and a machine was trusted to express faithfully and reliably the subject’s genuine and autonomous will. A philosopher such as Martha Nussbaum [157] would certainly disagree with the unfavorable decision, since she defends the right of all disabled persons, not matter how deep their intellectual deficits, to participate through a surrogate in a nation’s political acts. The experience and the “transhuman” circumstances of LIS may help reconceptualize criteria of civil

personhood in this complex and controversial domain of disability and justice.²¹

The contrast between dependency on a machine or a human highlights the social, economic and material conditions of life in LIS. Subjectivity and first-person experience, including the very possibility of narrativizing them, are inseparable from levels of public and private support, from the structure of a health system, from access to quality of care and to communication devices, and from attitudes to illness and disability, which significantly impact clinical realities at their most concrete level (for example the use of tracheostomy ventilation in ALS, which is more frequent in Japan than in Western countries; [42, 174, 216]).

Phenomenology as a Resource

This article has suggested that phenomenology can be a resource in various practical and theoretical domains, from patient care to the philosophical theory of personhood, with effects that range from sensitizing us to ill persons' attitudes and perceptions to giving empirical consistency to abstract notions and discussions. But phenomenology, as philosopher Havi Carel [30] has proposed, can also be a resource for patients, a tool for linking the existential and the medical dimensions of their condition by delving into the lived experience of illness and its transformative effect on one's being in the world. With regard to LIS, in this connection too almost everything remains to be done.²²

²¹ In 2016, California's Senate Bill 589 amended the state's election code to expand the number of citizens with disabilities who can retain or regain voting rights. At first sight, LIS should be in this framework less problematic than conditions involving cognitive disabilities; that, however, has not been the case. David Rector, a locked-in person, was initially denied voting rights, but was able to recover them in time for the 2016 election. Yet his success is by no means the norm, and the battle against the disenfranchisement of people with disabilities continues. Rector has become the lead individual in the class action suit filed with the US Department of Justice by the Spectrum Institute, a disability rights organization, for California's failure to restore voting rights in a timely manner. On Rector's case and more generally, see [140] and [202].

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