Aim to investigate Locked-In Syndrome as portrayed in the film "The Diving Bell and the Butterfly", discussing: the Accuracy of the Film, and the effects of the syndrome.

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It is hard to envision ourselves being trapped in our body and enduring an extreme deprivation of independence. However this was the exact fate that Jean-Dominique Bauby suffered in 1995 after a brainstem stroke. With movement limited to his left eyelid, but consciousness totally intact, Bauby was diagnosed with Locked-in Syndrome (LiS). Despite this, he was determined to communicate with the rest of the world, and soon became the author of “The Diving Bell and the Butterfly” by painstakingly choosing each letter through eye blinks. This best-selling book became a feature film in 2007 and won awards at various ceremonies (Cannes Film Festival, Golden Globes, BAFTAs and Cesar Awards), as well as being nominated for four Academy Awards. Since the original French release "Le Scaphandre et le Papillon", it has become globally accessible in various languages including English.

The "Diving Bell and the Butterfly" has become the basis of our topic of interest as it explores the psychological consequences of LiS and allows us to investigate the neuroscientific and physiological outcomes of such a rare, yet fascinating case. Prior to the release of the movie, there was little public knowledge about LiS and the unpredictability of its onset. Nevertheless, due to such media exposure, there have been many support networks established, funds raised and heightened social pressure to find effective treatments and improvements to the quality of life of patients.

1. Neuroscientific Context:

1.1 Background

LiS is a rare neurological disorder which severely affects an individual's physical and psychological state of well-being. It often results in a complete loss of control in voluntary muscle movements (quadraplegia), face paralysis, respiratory difficulties and anarthria; an inability to verbally communicate (Smith & Delargy, 2005). Unlike vegetative (grossly impaired consciousness whereby an individual is incapable of voluntary or purposeful acts) or brain-dead patients, LiS patients are still cognitively and consciously aware of their environment, thus are seen as "trapped" in their own bodies. This syndrome most commonly occurs as a consequence of trauma or vascular injury to the brainstem, such as strokes and brain tumors. Less common causes include overdosing, hypoglycemia or damage to the nerve cells caused by multiple sclerosis (Patterson & Grabois, 1986; Laureys, Pellis, Van Eekhout, Ghorbel, Schnakers, Perrin, Berre, Faymonville, Pantke, Damas, Lamy, Moonen & Goldman, 2005).

There are three main classifications; 'Classic' which presents anarthria, quadraplegia and an intact consciousness, with eye muscle control; 'Incomplete' which is similar to classic, however is characterised by some motor ability like weak arm or leg movements; and 'Total' LiS which results in complete paralysis, anarthria and intact consciousness. LiS can also be classified as either Chronic or Temporary; however this page will mainly focus on Chronic LiS (Laureys et.al., 2005).
Although there is no current cure for LiS, a patient's quality of life is of utmost importance. Extensive rehabilitation is required for patients to regain independence, improve social interactions and assist in psychological well-being. Therefore scientists have developed innovative ways of communicating including eye-tracking and fMRI technologies. Several case studies have been investigated such as Jean-Dominique Bauby and Tony Nicklinson, which have given rise to ethical debates regarding euthanasia (painless putting someone to death with their consent) and the quality of life.

1.2 Brain Areas

LiS (also known as “ventral pontine syndrome”) represents a severe insult to the ventral pons; a region of the brainstem containing neurons that contribute to autonomic regulation (respiration, swallowing, bladder control) and sensory processing (taste, sensation). Moreover, the pons controls voluntary movement by acting as a relay station for signals travelling between the forebrain and cerebellum.

Specific areas of the pons are involved in the control of different functions. Crossing the ventral pons are the corticospinal, corticofacial and corticobulbar fibres which signal voluntary motor supply to the limbs, face and medullary motor nuclei respectively. Disruption of these pathways results in the classic immobility exhibited in LiS patients. Moreover, lateral gaze is controlled by the paramedian pontine reticular formation (PPRF), whereas vertical gaze and upper eyelid movement depends on signals from the superior colliculi of the mid-brain, and is therefore preserved during LiS. Furthermore, disruption to the pneumotaxic centre of the pons in LiS patients results in decreased respiratory rate, and increase depth of each breath, while cutaneous sensation is preserved due to the spinothalamic pathway being more laterally placed.

The midbrain and reticular formation are brain areas required for cognition and consciousness. They are above the level of the pons and hence are not disrupted in LiS. PET scanning has
revealed that the brains of LiS patients have significantly higher metabolic levels compared to patients in the vegetative state, but equal levels compared to age-matched healthy controls. Furthermore, LiS patients exhibit normal EEG activity, compared to slowed activity in the vegetative state (Laureys et.al., 2005). These combined results highlight the fact that, unlike in the vegetative state, the intellectual capacity and consciousness of an LiS patient remains perfectly intact.

1.3. Causes

Figure 2: 2 types of stroke and their effect

LiS can be classified into vascular and nonvascular groups, nevertheless, it is important to note that some cases are due to an overlap of the two. Vascular etiologies are most common, particularly associated with circulatory system diseases, hemorrhages and strokes caused by blockage of the basilar artery which supplies oxygenated blood to the pons (Patterson & Grabois, 1986). This leads to short-term inadequate blood supply to the area (transient ischemia), causing cell death due to lack of oxygen (infarction) and rapid loss of function. Therefore, disruption of blood flow to the brain stem and associated structures leads to gross physical impairment.

There are various nonvascular factors, the most common include traumas to the brainstem, prolonged hypoglycemia; insufficient supply of glucose to the brain due to its low content in blood (Chow & Heller, 2012); central pontine myelinolysis; neurological disease due to severe damage on myelin sheath (Hurley, Filley & Taber, 2011); tumors and infectious agents, such as measles, encephalitis, swine flu and toxins (tetrodotoxin).

The primary cause of death for LiS patients are the extension of the lesion, and respiratory and cardiac complications. Thus, patients with LiS due to nonvascular etiology have a greater rate of rapid functional recovery and a higher survival rate than patients with LiS due to vascular factors. (Patterson & Grabois, 1986).
1.4 Psychological Aspects

Diseases with partial or total physical restriction such as LiS have, as consensus, the idea that patients present negative psychological effects as well as a low degree quality of life. However, studies containing self-assessment questionnaires which patients answer using eye movement technologies, show that the majority of patients with LiS evaluate their quality of life similar to those presented by healthy participants (Bruno, Bernheim, Ledoux, Pellas, Demertz & Laureys,
The amplitudes of scores were highly correlated with positive and supportive contexts such as the environment surrounding the patient and social interactions with friends and family. Also, the treatment given by the medical team is extremely important for the development of the patient's motivation. Once that is achieved, suitable communication methods are investigated to enable the patient to express their feelings and needs (Sledz, Oddy & Beaumont, 2007).

Contrary to results found in patients with cancer or non-terminal chronic disease, the majority of LiS patients do not show signs of depression, suggesting that the development of this psychological state has no relation with physical restrictions (Bruno et al., 2011; see Figure 3). Studies have shown that quality of life is almost always related to the social environment than physical interaction and autonomy (Lule et al., 2009). According to Doble et al., (2003) LiS patients may present an estimated life of up to 20 years or more, compared to chronic disease patients whom have short or ambiguous life expectancy. Perhaps this is one of the reasons why patients with chronic diseases tend to have depression symptoms on a larger scale than those with LiS (Doble et. al., 2003). Furthermore, chronic illness patients may have their self-esteem weakened and a sense of hope in the future diminished (Hendler, 1984).

In fact, examples like Jean-Dominique Bauby, whom wrote his memoirs, and Julia Tavalaro, whom became a poet some years after being diagnosed with LiS, have shown that patients are able to further develop their intellectual abilities through composing and reading poetry and literature.

Figure 3: Self-Assessed Well-being in LiS scores derived from Bruno et.al. 2011 - LiS patients’ socio-demographic, disease characteristics, quality of life and end-of-life data in unhappy (ACSA ratings <0) versus happy (ACSA $0) respondents. Particularly note the depression ratings

1.5 Treatment and Technology

Treatment:

Because individual cases of LiS vary so greatly, there is no existing standardised treatment.

Full (or even partial) recovery of motor control is very rare to impossible for chronic LiS-sufferers but most patients achieve vertical eye movements and eye-blinking abilities (Smith & Delargy, 2005). However rehabilitation is crucial for learning coping skills and long term management of the disorder.

Therefore a holistic rehabilitation care plan requires physical, occupational, recreational and speech therapies. Various specialists from centres such as the Rehabilitation Institute of Chicago (RIC) endorse a home-based care plan. This involves teaching skills to primary caregivers and patients such as monitoring medication, basic communication, finding new leisurely interests and other essential skills. The main goal of rehabilitation is to improve patient quality of life by maximising their existing abilities, increasing independence and problem solving for daily challenges. Thus it is essential for LiS patients to engage in rehabilitation to
develop positive physical, mental and social capacities for everyday life (Harvey et al., 2011).

**Technology:**

Figure 4: Perspex board with the alphabet used to communicate using eye movements. These can be organised in a variety of ways ranging from alphabetical order or frequency of use order. Communication is an important part of rehabilitation as patients regain a degree of independence (Andrews, 1999). Those that have only eye-blink capacities (Classic LiS) require more sophisticated communicative technologies. Some patients such as Tony Nicklinson utilised a colour-coordinated perspex board to slowly spell out words and sentences to a primary carer. Advanced eye-movement tracking technology such as Eye-gaze Response Interface Computer Aid (ERICA) uses a camera and infra-red light to detect the position of gaze on a computer screen so the patient can “type” with their gaze (Harvey et al. 2011). These systems have been the most reliable for the majority of cases, however varying individual needs present application difficulties and decrease processing speed. This means that conversation often lags and can become tedious (Dobkin, 2007).

Figure 4.1: This eye-gaze system allows this patient to communicate using only his eye movements. A video camera focuses on the pupil of the user's eye, tracking its movement. To “press” a key on the screen, the user looks at the key for a specified time, prompting the computer to take the appropriate action.

Currently, research into Brain-Computer Interface (BCI) systems is promising in the field of artificial communication. These systems will operate with electrodes planted on the scalp or through use of a wireless micro-chip that sends electric brain signals to a wheelchair-mounted computer (Klose, 2007). This computer then interprets the received patterns of brain activity to produce speech or even possibly movement of a robotic arm (Dobkin, 2007). The challenge for
BCI systems is to recognise the huge variations in individual brain activity and to operate in real-time to produce normal conversation.

With all communicative technologies, financial, safety and reliability concerns arise. Although technology can lessen a patient’s impairment, rehabilitation primarily relies on individual cases of LiS, the level of care and technology patients can afford and how motivated they are to continue rehabilitation (Dobkin, 2007).

2. Critical Analysis

‘The Diving Bell and the Butterfly’ is a French film based on the life of Jean-Dominique Bauby and his memoirs after the onset of LiS. This film had huge international acclaim, being released in numerous countries and appealed to movie go-ers worldwide.

However the neuroscientific accuracy of the entire film is debatable as it is a dramatised portrayal of LiS. One major downfall of this media item is that it is quite specifically a subjective description of LiS. As this is a film and not a documentary, it focuses on the protagonist’s internal thoughts to describe the experience of having LiS. Although this is a rare insight into patient’s suffering, it is highly dramatised for the “Hollywood effect” in several ways. One of these include pulling on the audiences’ heart-strings by emphasising the thoughts and feelings of characters in reactions to LiS rather than presenting LiS symptoms as they were diagnosed in the film. Furthermore the film did not adequately explain the medical causes of LiS in scientific language as the dialogues had to be understandable to the general public.

On the other hand, the quality of information in the ‘Diving Bell and the Butterfly’ is very detailed in highlighting the difficulties faced by LiS patients in everyday life and the extensive rehabilitation required to overcome these challenges. One good example was the inability of the protagonist to swallow food and how he had to undertake massive amounts of rehabilitation to just perform this very basic daily function. Another particularly striking scene demonstrated the frustration, fear and helplessness accompanied by the inability to communicate when doctors sewed the patient’s right eye shut to prevent infection, against his (unvoiced) wishes.

Therefore the film is in accordance with the current literature in the observable effects of LiS, the quality of life patients face and their personal cognition. But it does not have a strong neuroscientific foundation. As this film was based on the memoirs of Jean-Dominique Bauby
during the 1990s, there is inadequate information about the technological advances made for communication. So in this respect it does not agree with the current scientific literature on eye-tracking computer systems (Dobkin, 2007) or the increased life expectancy of LiS patients since 1985 (Laureys, 2005). However the film achieves its goal to increase awareness of LiS and overall, it is a visually striking and compelling film.

3. Ethics and Case Studies:

3.1 Ethics

Ethical arguments for LiS include delay of diagnosis and misdiagnosis, and bed-side attitudes. It can take months for the doctor to diagnose the patient with LiS after its onset which causes severe stress for the apprehensive patient, family and friends. Furthermore, as LiS is a rare condition, misdiagnosis can lead to incorrect rehabilitation and life expectancy information. A combination of these can lead to insensitive bed-side attitudes if the doctor/family/friends believe the patient is vegetative (Wilson, B. A. et al., 2001).

Another really important ethical point is concerned with life-support treatment and euthanasia. Studies revealed that the majority of LiS patients do not wish to die and favour life-saving treatment if required (Laureys, 2005). However, there are some cases in which the patient desires death. This presents an issue as euthanasia is illegal in nearly every country, often viewed as an act of murder or failure of a doctor's legal obligation to minimise suffering. Therefore, the patients must fight the judicial system to have their wishes achieved, as in the case of Tony Nickson.

3.2 Case Studies

Tony Nicklinson:

Tony Nicklinson (58, 1954-2012) had been a victim of LiS since a stroke in 2005, which left him paralysed from neck down and taken away the ability to speak.

Tony’s life consisted of being washed, shaved, dressed and exercised by carers, as well as staying in his chair in front of a specialised computer that is adapted to detect eye movements, and communicating using a perspex board to spell out words, which are ever so slow. Tony stated that he was frustrated as he was reminded everyday of the independence and privacy he no longer possessed, and described his life as a “misery created by the accumulation of lots of
things, which are minor in themselves but taken together, ruin what’s left of my life” (Miller, 2012).

In June 2012, Tony and his family took the case to the court in order to gain the right to determine when, where and how a person dies and enable a third party to lawfully take the life if the subject was physically unable to do so.

Tony wished that he had a brain damage so that active and functioning thinking became impossible. As this implied that he would rather be in a vegetative state if death is not an option, it is a question whether people who strongly desire to end their life should be forced to live.

Tony passed away on 22 August 2012, from pneumonia - refusing food and fluid intake, after losing the court case on 16 August.

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Jean-Dominique Bauby:

Jean-Dominique Bauby (45, 1952-1997) is perhaps one of the most famous patients whom suffered from LiS. He is well known for the publication of his memoirs ‘The Diving Bell and the Butterfly’ which he wrote using an elaborate eye-blinking system. On the 5th of December, 1995 the once Editor in Chief of ‘Elle’ magazine in France suffered from an abrupt stroke in his brain stem, consequently resulting in LiS. Jean-Dominique passed away on March 9, 1997 due to a heart failure, only 2 days after completing his memoirs (Kat, 2011).

The best known record of his experiences are derived from his memoirs which were made into a movie in 2007. In his account, Jean-Dominique describes his psychological experiences and how LiS affected his day-to-day life. Such insight into his life can be helpful to medical practitioners and other carers of patients with LiS to improve their quality of life.

Jean-Dominique’s case, although tragic, did not dampen his will to live. He created his own association named the ‘Association du Locked-in Syndrome’ (ALIS) which supports other patients with LiS and their families with acquiring the necessary technological assistance, raise awareness of the cause and help improve the quality of life of such patients.
Appendix

The search strategy for this project was a team effort that came about through group discussion. We met one Friday after the lab where someone mentioned reading something about Locked-in Syndrome on the UNSW Psychology Facebook page. Then another group member suggested 'The Diving Bell and The Butterfly' as our media item and once we had all seen the movie trailer, we decided it was a good topic to investigate. From then, all group members have worked independently on their assigned section and we collaborated our ideas/findings during fortnightly group meetings. We have also made Wiki posts, emailed each other resources and made suggestions/clarifications for the project.

Feedback from fellow NEUR2201 classmates, in general, suggested that we needed to work on the Psychological Aspects and the Treatment/Technology section. Therefore, the group members in charge of those specific sections edited their parts independently: Luciana added more detail to the 'Psychological Aspects' and Brina shortened the 'Treatments & Technology'. Referencing was fixed throughout as suggested by review comments and the NMES (neuromuscular electrical stimulation) therapy was deleted due to lack of specifically named LiS cases where NMES was used (patient had only been called 'X') and its rareness nowadays as a treatment, as found by Brina. Suzanna added a clear definition of LiS at the beginning of the 'Introduction' and labelled a figure that had not been labelled which reviewers had found.
One reviewer said "some sentences ran too long" so we attempted to shorten sentences in general, however the reviewer did not specify WHERE those sentences were, so all group members fixed syntax, spelling and word length as appropriate throughout the Wiki page.

Some reviewers requested additional clips of the movie as they thought the trailer was not engaging/detailed enough. However, only the trailer of the film can be uploaded (as opposed to the whole movie) and we believed that this media item was concise enough to present our topic in an interesting way. We also felt that there were enough references to the full movie and Bauby's autobiographical book for viewers to find it themselves online/DVD and in a bookstore. That said, we added another clip from the film in the critical analysis section to increase involvement with the topic and the media item chosen.

At the final group meeting on Friday (Week 9), the whole group reviewed all the comments made by reviewers and created a list of the changes we have made in response to them for this 'Appendix'. The group discussed the nature of the comments and agreed that we did not require major theoretical changes, but mostly minor grammatical errors, which we addressed as best as possible.

Finally, the comment from one reviewer (z3218566) was given after the "Review Comments" due date, therefore according to the assessment instructions, we discounted these suggestions.

Overall, we worked very effectively as a team and did the best we could for this assessment. Please refer to the 'Progress and Administration' hyperlink (bottom of references) for more information.

References:


