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Challenges when diagnosing locked-in syndrome following TBI: The Story of U.P. a Clinical Puzzle

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Abstract

Background and aims: *Locked-in syndrome (LIS) is a rare neurological disorder; patients with LIS are awake, conscious with normal or nearly normal cognitive functioning. They cannot produce speech, facial or limb movements with complete paralysis of all voluntary muscles except for those controlling eye movements. LIS is associated with lesions of the brain stem and the pons, with 60% of people having sustained a stroke. LIS following traumatic brain injury (TBI) is rare; brain stem lesion plus cortical damage makes diagnosis of LIS challenging.*

Method: *We describe U.P, a 42 year old man who sustained a TBI. A CT scan showed traumatic subarachnoid haemorrhage with a pre-pontine bleed. Awake, conscious, presenting with right sided paralysis and severe left sided paresis;U.P could produce voluntary horizontal eye movement, tracking people and stimuli of interest. Vertical eye movements emerged later, reading simple instructions and following commands.*

Results: *U.P could use eyebrow movements for "yes" and a slight head shake for "no". He could read some written instructions; non-verbal responses were inconsistent and sometimes unreliable.*

Discussion: *Based on assessments from speech and language therapy and neuropsychology with U.P, we discuss LIS plus additional cognitive problems and the difficulties with diagnosing LIS following TBI.*

Introduction

The term "locked-in syndrome" (LIS) was first used by (Plum & Posner 1) [1]. Patients diagnosed with this condition had quadriplegia, lower cranial nerve paralysis, and mutism with preservation of consciousness, vertical gaze, and upper eyelid movement. The American Congress of Rehabilitation Medicine noted that LIS has five characteristics: namely (i) sustained eye opening (ii) preserved basic cognitive abilities, (iii) aphonia or severe hypophonia (loss of voice), (iv) quadriplegia or quadriparesis, and (v) a vertical or lateral eye movement or blinking of the upper eyelid as the primary means of communication [2]. Some people, of course, have problems with eye gaze and eye movements. Pierrot-Deseilligny, Milea and Muri say that a neuroscientist can tell much about the organisation of the brain through eye gaze abnormalities [3]. They also remind us that eye movements are controlled by ocular motor nuclei in the brain stem. The lesion causing LIS is also in the brain stem.

LIS is caused by damage to the pons, a part of the brainstem that contains nerve fibers that relay information to other areas of the brain. Most LIS patients have sustained a stroke in the basilar artery or suffered a pontine haemorrhage [1]. Schnakers et al, say that at least 60 per cent of LIS patients have sustained a basilar artery or pontine haemorrhage [4]. In addition to stroke, there are other causes of LIS. Smart et al include, among other things, TBI, tumour, encephalitis and toxins [5]. Although prognosis is generally poor and death can occur through pneumonia or thrombosis, with the right care people with LIS can live for many years and a few make a good or even a complete recovery [6].

People with LIS have normal or nearly normal cognitive functioning [7]. Duffy says that in LIS the individual is conscious and sufficiently intact cognitively to be able to communicate with eye movements [8]. So he seems to be implying that the ability to communicate is sufficient with regard to cognition. With some people it is easy to diagnose and assess LIS. Wilson et al, for example, report the case of Tracey who became locked-in after a fall in the gym which caused a tear in her brain stem [9]. Wilson et al tell the story of Paul who was diagnosed with LIS after a brain stem stroke [6]. Both these patients were able to communicate easily (albeit slowly) with eye movements. There are, however, those who have brain stem lesions plus cortical damage or those who have difficulty with eye movements such as ophthalmoplegia making it difficult for them to control their eye muscles. When patients like this are referred for assessment it can be difficult to determine whether or not they are truly locked-in with additional problems or not. We report the case of one man who challenged our ability to make such a diagnosis.

Case Study

At the age of 39 years, U.P. sustained a TBI in a road traffic accident. This resulted in a sub arachnoid haemorrhage, a prepontine bleed, a skull base fracture plus numerous other fractures. As well as a tracheostomy, a ventriculoperintoneal (VP) shunt was inserted after U.P developed hydrocephalus. He was 40 years old when first seen by the speech and language therapist and the neuropsychologist. During the various assessments, U.P. was always awake and appeared to be alert. He had some rhythmic mouth movements but did not verbalise. His left eye was partially stitched closed because he was unable to blink that eye. His right eye stared intently at people. Because he had nystagmus, it was not always easy to know if he really was looking at something or not [6]. Wilson et al report the neuropsychological assessments carried out with this man. They note that U.P was definitely awake and alert, was responsive to sound and appeared to look at objects when requested. However, it was impossible to establish a Yes/No response in order to confirm whether or not he had LIS. He was asked first to blink for "Yes", then to raise his eyes for "Yes" and finally, to look at "Yes" and "No" cards (first when these were side by side and then when they were placed one above the other in case of scanning difficulties). With each attempt, U.P. just looked at the tester. On the basis of the results at that time, U.P was believed to be emerging from the minimally conscious state (MCS). There was lack of certainty both about this diagnosis and whether the scores were reliable. It was not absolutely clear if U.P was really looking from one object to another or whether his nystagmus was making his eyes jump. The assessment, therefore, was repeated a few weeks later. The repeated assessment showed no changes in his behaviour. It was still unclear if he was really looking at a named object or whether his nystagmus made it appear as if he were looking. He did not try to communicate; and a Yes/No response could not be established. At this time, the previous diagnosis was confirmed.

The assessors, however, were always worried that they were under estimating U.P's level of functioning. This was because he differed from other people at the top end of, or who had just emerged from, the MCS. Because U.P stared intently with his good eye and he was always so alert, this led to the belief that he may have a LIS. Then his wife discovered he could read! Of course, someone who can read is well beyond the MCS. U.P was much better at the written word than the spoken word. As this is unusual in patients with naming and language disorders (which it was thought U.P. might have), this raised questions about his hearing. Inretrospect, his ability to hear should have been one of the first things to check for, but clinical experience suggests that relatively few people are deafened after a brain injury while many have problems with visual acuity and visual perception.

U.P. was then referred for an audiology examination and the report said that he had severe to profound sensori-neural hearing loss, worse on the left side; he also had a glue ear on the left. Once communication was established, via the written word, U.P. himself denied problems with his hearing. It is unclear why the audiology report said he had a hearing loss while U.P. himself felt he did not. It is possible that another reason he was better at the written than the spoken word, was due to something else entirely such as delayed processing orapraxia.

Once it was realised that U.P. could read, he was provided with an ipad on which questions were typed for him in large letters. When next assessed, his ipad was used. The purpose of the assessment was explained to U.P by writing this out in simple language. The questions for him were then typed out one at a time. At first, he was asked to blink again, but he did not respond. Yes/No cards were used to no effect. It was then determined that U.P could raise both eyebrows with prominent upward movement for "yes" and could indicate "no" by an almost imperceptible shake of his head. Three observation sessions were required to ensure correct interpretation of these very slight movements.

Finally, it was possible to assess him with the Putney Auditory Comprehension Screening Test [10]. He passed four of the five practice items and all of the biographical items but made some errors on the other questions (for example he responded incorrectly to the questions "is cyanide poisonous?" and "was Picasso a train driver?"). Although U.P's score of 49/60 was well above chance, he certainly had some cognitive problems. This was confirmed by another assessment, the Cognitive Assessment by Visual Election [11]. The CAVE assesses people who have emerged from a disorder of consciousness but are too impaired for traditional tests. There are six CAVE categories (objects, numbers, words, letters, pictures and colours). In each category twenty stimuli are presented, two at a time and the person being tested is asked to look at (or indicate), the correct one of each pair. Thus, a comb and a car may be presented and the person asked to look at the "CAR". Half the correct stimuli are on the right and half on the left. U.P scored 7/10 on objects; 7/10 on numbers; 8/10 onwords, 10/10 on letters; 10/10 on pictures and 10/10 on colours. In each case the instruction was written out on his ipad (for example, "Please look at CAR". The Test for Reception of Grammar (TROG-2) is a standardised assessment used with children through to young adults, assessing their auditory comprehension skills [12]. The participant is required to match the sentence heard to a choice of four target pictures. This assessment was appropriate to use with U.P. as an informal measure of his comprehension skills of a variety of grammatical constructs. The format of the TROG-2 was altered to meet his access needs. Each sentence was written on an iPad, with each of the four pictures presented on the corners of an e-tran frame. The written sentence was given first, presented for about five seconds, after which the e-tran frame was shown. The speech and language therapist and U.P were able to see one another through the central gap of the e-tran frame, with U.P's significant level of physical limitations he was allowed thirty seconds to look at all of the images and was then guided in clockwise around the e-tran frame, by the therapist pointing to each of the four pictures.U.P would follow the therapist's finger pointing at each picture, he would indicate "no" via head shake or "yes" via prominent eyebrow raise or head nod. Over five sessions U.P completed 14 out of 20 blocks of different types of grammatical constructs; achieving 100% correct responses with 4/14 of these blocks. The results showed U.P understood "two elements" (practise item), "reversible in and on", "three elements" and "not only X but also Y" constructs without difficulty. The results of the remaining 10 blocks were as follows:

- 75% accuracy with 2 blocks:"reversible SVO" and "reversible above and below" constructs
- 50% accuracy with 4 blocks: "two elements", "negative", "relative clause and subject" and "comparative/absolute" constructs
- 25% accuracy with 3 blocks: "four elements", "reversible passive" and "pronoun gender/number" constructs

• 0% with 1 block: “zero anaphor”

In subsequent speech and language therapy (SALT) sessions U.P demonstrated the ability to read upside down, he was already responding to written single words and phrase level instructions appropriately even before the therapist had finished typing them out on the iPad. U.P would often initiate social interaction with his family and staff by lifting his left hand from the wrist or smiling with right sided weakness whilst sustaining eye contact, for “hello” and “good bye”. During dysphagia assessments of ore-motor functioning U.P would attempt initiating movement of the muscles of his face and mouth, following written instruction. On several observations U.P looked down towards his mouth in an attempt to “help” move his tongue on instruction; however his lips would grope in weak and asymmetric patterns withmostly unsuccessful attempts of tongue movement. The impact of the traumatic brain injury left U.P with the reduced ability to manage copious secretions effectively; he was able to trigger involuntary swallows these would occur with infrequency. In SALT sessions he was able to trigger voluntary swallows when following the written instruction “swallow” on the iPad. The carryover and generalised is limited and U.P continues to require on-going support to manage the volume of secretions produced.

Additional cognitive deficits are, perhaps, unsurprising as U.P had several skull fractures sustained in the road traffic accident as well as his pre-pontine bleed. Thus, as well as pontine damage, U.P. also had cortical damage.

Discussion

This case study illustrates the problem of accurately diagnosing LIS in someone with problems controlling their eye muscles and with cognitive deficits. Although, as said earlier, people with the LIS have normal or nearly normal cognitive functioning it is possible that the necessary lesion for LIS, a pontine lesion, can co-occur with cortical damage and consequent cognitive deficits. In particular the administration of the TROG-2 assessment took up to five sessions yet remained uncompleted with fatigue and slow processing speed affecting U.P’s on-going participation despite being medically stable.U.P appeared to have ophthalmoplegia making it difficult for him to control his eye muscles. This, in turn, meant it was hard to establish communication with him. He could open his right eye but his left eye was more or less fully closed. A neurologist might have been able to say more about U.P’s eyes but in a clinical situation, without recourse to sophisticated equipment, finding the best way (or indeed any way) to converse is problematic.

One of the American Congress of Rehabilitation Medicine’s criteria for LIS is communication by eye movement; U.P was unable to do this. He could just about raise his right eyebrow for “Yes”, but could not voluntarily control his blinking, raise his eyes up or down or swallow. We believe that he may have partial LIS with additional cognitive problems. The American Congress of Rehabilitation Medicine states that basic cognitive abilities are intact with LIS patients but this begs the question “what are basic cognitive abilities?” Is it the ability to understand? Does it include memory, naming and attentional problems and so forth? We saw above, that Duffy (2000) seems to be implying the ability to communicate is sufficient with regard to determining cognition. U.P. can communicate, if not by eye movements, then by marginal movements of his head.

The point to make here is that it is not always easy to determine LIS and it is possible for some patients, like U.P to have LIS plus other cognitive problems.

Conclusion

The case of U.P illustrates the difficulty clinicians have in reliably determining whether or not someone has LIS when, in addition to a brain stem lesion, they are unable to control their eye muscles and have cortical damage with consequent cognitive impairments.

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