'Locked-in' but not 'locked-out'
A case report

A. S. DE GRAAF, MERYL D. RYBNIKAR

Summary

The application of modern electronic apparatus in patients with the 'locked-in' syndrome can significantly improve communication.


The term 'locked-in' syndrome was introduced by Plum and Posner in 1966 and since then has been generally accepted as a clinical concept. It describes a selectively de-afferented state of lower cranial and spinal cord neurons while leaving consciousness preserved. In most cases the only means of communication is by vertical eye movements and blinking.

Although most patients die after a cerebrovascular accident causing this syndrome, partial recovery with years of survival has been reported. As mental functions are fully preserved it is of paramount importance to establish some form of communication as soon as possible.

Although many papers on the 'locked-in' syndrome have appeared, scant attention has been paid to its management. Electronic computerized devices can significantly contribute to the patient's well-being. Attention is drawn to this aspect of management.

Case report

A 25-year-old man was transferred from a South Atlantic island weather-station to the intensive care unit of Tygerberg Hospital.

A detailed history was not available but it was learnt that his initial symptoms had appeared about 8 days before admission. He had developed a headache and slurred speech, and had later become anarthric. This was followed by quadriplegia, urinary incontinence, and inability to swallow. His body weight had increased and only able to react to questions by blinking his eyes. His head and eyes were responsive to very light pressure. Head position was carefully monitored to prevent movement. The adapted computer enabled the patient to state his problems and needs effectively.

On arrival he was mute, on only able to react to questions by blinking his eyes. His consciousness was somewhat decreased, pupils were equal and reactive, fundus normal, eye movements full and the corneal reflexes present and equal. Frowning was seen when emotionally upset or in pain, and vocalization was a groan when crying. No voluntary movement below the upper face could be elicited. Sensation appeared to be largely intact. There was a flaccid paraplegia and urinary incontinence. Except for bronchopneumonia, further physical examination was normal; the blood pressure was 130/90 mmHg. A tracheostomy was performed because of swallowing difficulties.

Nutrition was provided by nasogastric tube, a urinary catheter was inserted and intensive physiotherapy and occupational therapy started.

Although sleep was not recorded polygraphically it appeared to be normal.

Excerpt for heavy cigarette smoking and a strong family history of heart attacks and varicose veins, the patient's previous medical history was unremarkable.

Results of the following laboratory tests were normal: full blood count, erythrocyte sedimentation rate, serum electrolytes, hepatic and renal function tests, lipogram, CSF analysis, and serological tests. Serum electrophoresis showed an acute-phase reaction, probably due to the bronchopneumonia. Blood coagulation studies were compatible with an acute stress reaction. ECG, echocardiogram, EEG, brainstem auditory evoked potentials and somatosensory evoked potentials were within normal limits. Computed tomography revealed an area of decreased density centrally located in the pons. Vertebral angiography disclosed a complete occlusion of the proximal basilar artery. The vertebral arteries were patent.

During the following weeks spasticity, with both increased and pathological reflexes including an increased jaw jerk, developed. About a fortnight after the initial symptoms he could make lateral movements with his head and lift it slightly from the pillow.

Reliability could only involve the senses, information processing, voluntary eye and head movements. Based on these facts, an intensive therapeutic programme was developed.

'Interface', a new association recently formed in Cape Town with the aim of enhancing the quality of life for the mentally or physically handicapped by adapting computers, was contacted.

A BBC computer and disc drive was lent to the patient, specific software written and an interface made, the minimum number of movements required to control the computer being two.

An occupational therapist designed a harness to facilitate activation of the microswitches by the mandible. The switches needed to be static as they responded to very light pressure. The harness allowed lateral head movements, while eye contact with the monitor was not lost, thus minimizing eye muscle strain (Fig. 1). The initial program (the alphabet) needed re-evaluation within a week, because it was too slow and therefore frustrating.

A record program was written, using the patient's ideas. Morse code was considered too slow and was not attempted. Other systems were not available, e.g. the long-range optical pointer.

The approach to the patient was very important since he did not yet fully realize the extent of his physical disability and the consequences. The idea of communicating through a computer was discussed over a few days and the need for adaptation explained.

The adapted computer enabled the patient to state his problems as they occurred and he was therefore able to obtain the appropriate help and therapy without delay, and to start working through his emotional reaction to his physical state towards self-acceptance.

Thought content improved from 'They [the nurses] are nice' to 'Please do not talk when I am working on the computer' to precise instructions about how to change the software program.

One of the initial problems was hypertension which built up to flexor spasms towards evening. This was alleviated through expression of emotion, the ability to indicate needs and neurodevelopmental techniques.

While using the computer, the patient had a tendency to flex his head and therefore increase flexor tone in the arms. The head position was carefully monitored to prevent this.

Within 3 weeks, endurance improved from 15 minutes to 40 minutes twice a day. The pace of the treatment was set by the patient. Previously there was a passive acceptance of all therapy and medical care. Now there was the stimulation of being able to express control, self-expression and communication, which gave a more positive approach towards future planning. The ability to help others through trying out equipment and systems gave self-
Because of the gradual onset and lack of evidence for a source of embolism, this is in agreement with the report of Castaigne et al.\(^1\) on basilar occlusions, 94.4% of which resulted from atherosclerotic thrombosis.

After basilar artery occlusion the development of a stable collateral circulation is crucial. This is largely dependent on the anatomy and patency of the vascular system and its anomalies.\(^{1,2,15}\)

In order to overcome ischaemia of the posterior circulation during the first critical weeks, treatment in the supine position has been recommended, specifically for subjects with fluctuating signs.\(^{14,15}\)

Opinions on the use of anticoagulants and drugs which decrease platelet aggregation are still divided. During the first few weeks elementary questions can only be answered by eye blinking and eye movements. Morse code signalling in later stages has been successfully applied in isolated cases.\(^{11}\) In less critically placed lesions, eye and perhaps head movements provide additional possibilities. Among the factors influencing the final therapeutic achievements are the patient’s personality, intellect, mood and interests.

Sensitivity to the patient’s needs is required from the medical and paramedical staff and the family.

Modern electronic technology can provide an ever-increasing range of specially adapted devices.

### Addendum

Since submission of this article practical improvements in the alphabet were introduced; technical procedures facilitating its reading are in progress and the patient is starting and driving his wheelchair by electronic devices connected to his chin.

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### References

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