The Case of Locked-in Syndrome in Pregnancy

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HPI

• 29-year-old female with no known past medical history presented to the emergency department with seizure activity and posturing. Diagnostic studies revealed bilateral pons and left cerebellar infarcts. Also found to have occlusion of right vertebral and basilar artery and an intrauterine pregnancy at 6 weeks gestation.
Initial Hospital Admission

- Seizure in ED and was placed on AED.
- Established communication with up and down eye movements, yes and no respectively.
- Patient, with assistance of mother, elected for chemical abortion.
- Stroke alert called for loss of horizontal eye movements and MRI showing progression of pontine stroke.
- Tracheostomy and PEG placement
- Transferred to skilled nursing facility
2\textsuperscript{nd} Hospital Admission

- The patient was again admitted to acute care for continued vaginal bleeding.
- Work up revealed severe anemia and leukocytosis.
- Pt received blood transfusion, infectious workup was negative but thought to be a complication of previous chemical abortion.
- Dilation and Curettage performed revealing retained products of conception.
- Transferred to acute rehab facility
History

- PMH: none
- PSH: none
- Family Hx: unremarkable
- Social Hx/Prior level of function: Lives alone in an apartment. Previously independent w/ ambulation, functional transfers, and ADLs.
- Drug Hx: Marijuana, questionable methamphetamines
Physical Exam

- Pulm: Tracheostomy, lungs with rhonchi no rales
- Abdominal: Percutaneous endoscopic gastrostomy (PEG)
- Neuro: CN globally weak with vertical eye movement (superior and inferior). Weakly raises right lip and able to open mouth only. Flaccid paralysis of all extremities. Muscle contractions noted in various muscle with cough but not under voluntary control. Sensation to light touch intact.
What is Locked-In Syndrome (LIS)?

- A neurological disorder due to damage of the brainstem, typically of the ventral pons and caudal midbrain, resulting in quadriplegia, bulbar palsy, and whole body sensory loss.
  - Intact functions may include eye movements, blinking, hearing and cognitive function. Most likely due to an ischemic cause, such as basilar artery occlusion.
- Classified into 3 subtypes: classic, incomplete, & total
LIS Subtypes (Bauer 1979)

- Total: total immobility + inability (not unwillingness) to communicate + full consciousness (think no voluntary movements & closed eyes)
- Classic: quadriplegia + preserved vertical eye movements + preserved consciousness
- Incomplete: classic + additional voluntary movements (varies case by case) other than preserved vertical eye movement

*In all subtypes, conscious mental activity was show on EEG (Kotchoubey 2013)*
### Causes and mechanisms of locked-in syndrome

<table>
<thead>
<tr>
<th>Cause</th>
<th>Mechanism</th>
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<tbody>
<tr>
<td>Ischaemic</td>
<td>Basilar artery occlusion, hypotensive or hypoxic events</td>
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<tr>
<td>Haemorrhage</td>
<td>Haemorrhage originating within or infiltrating into the pons</td>
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<td>Traumatic</td>
<td>Direct brain stem contusion or vertebrobasilar axis dissection</td>
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<tr>
<td>Tumour</td>
<td>Primary or secondary infiltration of the ventral pons</td>
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<tr>
<td>Metabolic</td>
<td>Central pontine myelinolysis</td>
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<tr>
<td>Demyelination</td>
<td>Multiple sclerosis affecting the ventral pons</td>
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<tr>
<td>Infectious</td>
<td>Abscess infiltrating the ventral pons, brain stem encephalitis</td>
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Functionality

- Many symptoms are due to disruption of neuronal tracts that pass through ventral brainstem

- Functions lost:
  - Paralysis of voluntary muscles of limbs/torso
  - Bulbar palsy: anarthria/dysarthria & dysphagia
  - Difficulty with voluntary breathing, apnea, ataxia, hyperpnea
  - Dizziness
  - Vertigo

- Functions Retained
  - Retains: consciousness, language comprehension, orientation
  - Retention of hearing
  - Vertical eye movements and blinking ability
Rehab Progress

- Problems:
  - UTI s/p Foley – were unable to have suprapubic tube placed
  - Dysphagia - Tube feeds progressed to bolus
  - Pulm: s/p trach, no major complications
  - Skin: Stage I pressure ulcer improved with diligent wound care
  - Contracture prevention: PRAFOs, wrist splints, and regular ROM
Therapy Progress

- PT: Dependent (12/08) → improved head/neck control, UE/LE strength and motor control, visual tracking & sitting balance (01/20). BM-Dependent, T-MaxA, Power wheelchair mobility-ModA

- OT: Dependent (12/08) → Dependent (01/20)

- ST: Severe oral and pharyngeal dysphagia (12/08), unable to assess communication deficits 2/2 Locked-In Syndrome; NPO w/ tube feeds → non-verbal expression – ModA, Motor speech-ModA, Voice-MaxA, Swallowing-ModA; NPO w/ tube feeds 01/19)

- Goal:
  - Mobility: Power wheelchair, supervision
  - Transfers/ADLs: Max
  - 24/7 supervision
Prognosis and Recovery

- Early literature have demonstrated mortality as great as 60% (n = 139) (Patterson 1986). A recent study has shown with aggressive rehab within 1mo of acuity, mortality was only 14% at 5 years (n = 14) (Casanova 2003).

Classification of recovery of motor function

- No recovery—No return of motor function, total dependence for all activities of daily living.
- Minimal recovery—Minimal motor return, total dependence for all activities of daily living.
- Moderate recovery—Moderate motor return, independence in some but not all activities of daily living.
- Full recovery—Independence in all activities of daily living but some minimal neurological deficit.
- No neurological deficit—No reported residual deficits.

*4 pts were excluded due to insufficient data.

Figure 3. Reported mortality per time interval for 79 deaths following the onset of “locked-in syndrome.” The number of deaths per etiology is also shown for each time interval.
Prognosis and Recovery Cont.

- Specifically for LIS 2/2 basilar a. occlusion, McCusker 1982 described 4 patients who were made significant functional gains over a period of several months, with rehabilitation.
  - Pts were able to regain some ADLs such as: bowel and bladder continence, eating by mouth, and functional speech w/ residual dysarthria.

- Life longevity: Doble 2003 determined long LIS (n = 29, with 48% with cerebrovascular
  - 5 year survival: 83%
  - 10 year survival: 83%
  - 20 year survival: 40%
Prognosis and Recovery Cont.

- Casanova 2003
  - Demonstrated meaningful recovery with intensive rehab within 1 month of onset demonstrating \((n = 14)\)
  - Partial or full independence in activities of daily living within three to six months of onset was achieved by three patients (21 percent)
  - Complete swallowing ability recovered in six (43 percent)
  - Verbal communication recovered despite dysarthria and dysphonia in four (28 percent)
  - Ability to use a device by hand, finger, or head movement was achieved in six (43 percent)
  - Tracheostomy removal was achieved by six patients (43 percent)
Prognosis and Recovery Cont.

- Hocker 2015
- A case series presented with 14 pts with classic/total LIS seen 3-6 months following neurorehab demonstrated (% pts):
  - Motor recovery 21%
  - Return of swallowing 42%
  - Verbal communication 28%
  - Bowel control 35%
  - Weaning off vent 50%

- Conclusion: Patients improvement help transition from classical/total to incomplete.
Prognosis & Recovery (Hocker 2015)
Quality of Life

• Despite being in a immobile state, chronic LIS patients typically self-reported meaningful quality of life and infrequent demand for euthanasia (Laureys 2015)

• Several studies demonstrated a proportion of their patients having depression and contemplating suicide, however, all wanted life-sustaining treatments, none had DNR directives, and quality of life scales were not significantly different from controls. (Doble 2003, Anderson 1993, Rousseau 2013)
References


