Remarkable recovery in an infant presenting with extensive perinatal cervical cord injury

Israr Ul Haq,¹ A K Gururaj²

¹Department of Paediatric Neurology, Latifa Hospital, Dubai, United Arab Emirates
²Department of Paediatric Neurology, Latifa Hospital, Dubai, United Arab Emirates

Correspondence to
 Dr Israr Ul Haq,
 israrulhaq87@gmail.com

SUMMARY

Cervical-cord damage is a complication of a difficult delivery, and results in spinal shock with flaccidity progressing to spastic paralysis. Conventionally, outlook for such patients is extremely poor and most will recover only slightly from quadriplegia and autonomic dysfunction. Here, we report a case in which the extent of damage considerably contrasted with the outcome and recovery. A full-term baby girl born by difficult vaginal delivery displayed bilateral flaccid paralysis of the lower limbs with absent spontaneous movements, weakness of both upper limbs, hyporeflexia in all limbs and axial hypotonia. MRI of cervicothoracic spine exhibited raised signal intensity in the dorsal aspects of C7 to T1 signifying myelopathy. MRI at 4 months revealed a near-total transection of the cervical cord. However, at 6 months, the child could move all lower limbs independently with a marked increase in power. There was no spasticity, wasting or incontinence. Reflexes had also returned.

BACKGROUND

Birth injury to the spinal cord is quite rare. This case presents us with a valuable example of an extremely unexpected but favourable outcome of severe injury of the cervico-thoracic spine. Near-total cervical cord transection causes numerous neurological deficits in the body, including respiratory insufficiency, motor weakness in the limbs and even quadriplegia; however, in this patient, contrary to the prognostic outlook, within a few months, significant improvement had taken place. This is a rare case of an extremely devastating injury with an astonishingly favourable prognosis.

CASE PRESENTATION

A baby girl was born at 39 weeks and 6 days gestation, via normal vaginal delivery to healthy non-consanguineous parents, with a birth weight of 3.52 kg. The mother was 40 years of age and it was her second child. The pregnancy was uneventful but it was a difficult delivery as the baby had face presentation and shoulder dystocia. As a result, hyper-extension of the neck occurred during labour. APGAR score at 1 and 5 mins was 5 and 8, respectively. Soon after birth, the baby was noted to have decreased movements. She was transferred to the Neonatal Intensive Care Unit at a tertiary referral hospital on suspicion of severe neck injury.

When the baby arrived in the neonatal intensive care unit (NICU), her neck was stabilised. On examination, she was found to have hyporeflexia in all four limbs, weakness in her upper limbs and bilateral lower limb flaccidity with grade 1 power. Axial hypotonia was also noted. On chest,

abdominal and cardiovascular examination, findings were unremarkable. Her vitals were within the normal range and there was no respiratory distress.

INVESTIGATIONS

In the NICU, an MRI of the cervico-dorsal spine was conducted. It showed raised signal intensity at dorsal aspect of C7 to T1 with associated severe atrophy, cystic change and fibrotic bands.

MRI of the brain was normal. A CT-scan of the cervicodorsal spine showed no fractures in the vertebrae. A full blood count was carried out and it was normal. Blood culture showed no growth.

At 4 months, a follow-up MRI of the cervico-dorsal spine was again conducted. This time, there was evidence of a near-total spinal cord transection between C7 and T1 (figure 1).

DIFFERENTIAL DIAGNOSIS

Examination findings of bilateral flaccid paralysis in the lower limbs, axial hypotonia, weakness of the upper limbs and hyporeflexia of all limbs can direct the physician to a number of diagnoses. Cerebral palsy was a very likely possibility in view of the difficult birth which may have led to hypoxic damage of the brain, but a normal MRI of the brain put that suspicion to rest. This investigation also ruled out any intracranial haemorrhage or brain malignancy that could have been presenting with such symptoms.

A CT scan of the spine was performed but could not detect any fractures or vertebral dislocations.

A negative blood culture and maternal history showed that a neonatal infection could be disregarded too. There was no fever or any other positive clinical finding either for such a cause.

Subsequently, we performed an MRI in order to rule out non-traumatic spinal cord pathology such as a tumorous mass as this could have been compressing a part of the spinal cord and causing these types of symptoms. Syringomyelia could also be diagnosed by this investigation. However, it neither showed a mass nor any syrinx. However, it showed raised signal intensity along the dorsal aspects of C7 and T1. Cystic change and fibrotic bands could be seen. Taking into consideration the history of difficult labour, a diagnosis of cervical cord injury due to hyperextension of the neck fit soundly.

TREATMENT

She received three doses of intravenous steroids for 2 days in the NICU. This was reduced to twice daily for the next 2 days and gradually tapered off after that by day 6.

Physiotherapy in the form of strength training and passive range of motion exercises was started for all limbs for an indefinite period.

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Figure 1 MRI cervico-dorsal spine showing transection of spinal cord in the region of C7-T1.

At the age of 9 months she started taking L-Carnitine daily.

OUTCOME AND FOLLOW-UP

She started showing improvement in her lower limb power by day 8 after birth. By the time of discharge, on day 14, her lower limbs had grade 1 power. There was no complaint of urinary retention or incontinence. She seemed vitally stable with no respiratory insufficiency or bradycardia and had started on-demand breastfeeding while still in the NICU, hence was discharged without any medications, though the mother was advised to give the baby regular physiotherapy.

At her 4-month follow-up, when clinically examined, the child's condition had improved, even though a second MRI showed a degree of cervical cord transection. No weakness could be found in her upper limbs. Her lower limbs had improved too, as movements equating to grade 2 power were observed. Continuation of physiotherapy was advised.

On her second follow-up at 6 months of age, the infant had improved significantly and could now independently move both lower limbs with a power of Grade 3 when she was put in a supine position. She could manipulate objects with her upper limbs and pass them across the midline.

Now at 10 months of age, the baby was re-examined in the clinic. She was found to be active and alert, with good movement of the upper limbs and a firmer grasp of objects in the hand. She could sit with support for a short while. She was able to lift her head up completely when lying in a prone position. Lower limbs could be moved up off the bed from the supine position with the same grade 3 power. Knee reflexes were elicitable and brisk. However, ankle reflexes were still weak. There was no wasting or contractures in the limbs and neither was there complaint of urinary incontinence or retention. Vitals were normal. No flushing of the skin or dryness was seen and

the nails were fine. Overall, she seemed healthy and was steadily improving. The only complaint narrated by the mother was that the baby did not perspire on the lower body and that she had a long-running tactile fever, which comes and goes every few days. Initially, it used to reach 39°C but now does not go beyond 38°C.

DISCUSSION

Perinatal trauma to the spinal cord is decreasing in incidence and has become quite rare, in part due to better prenatal surveillance and improved obstetrical practices. However, it is still observed in difficult deliveries when there is an abnormal presentation of the fetus, as traction on the neck, whether it is longitudinal or lateral, can cause varying amounts of damage to the spinal cord.

Different areas of the cord can be affected but the most common is the cervico-thoracic junction.¹ It could be an incomplete spinal cord injury in which there is partial loss of function, or complete, in which all function below the level of insult is lost.

In the case of lower cervical injury, upper and lower limb, as well as bladder function, is lost, to a varying degree depending on whether the injury is total, near-total or partial. In near-total transection, the symptoms include some degree of quadriplegia. In addition, the body may not be able to regulate heart rate, blood pressure, sweating and body temperature.

Diagnosis is typically made on the basis of a positive history and is confirmed and localised by MRI.² CT can also be used.

Treatment for cervical cord damage consists of administering high-dose corticosteroids, which exert their maximum beneficial effect if started within 6 hours of injury.³ Therefore, early recognition and treatment is crucial. Further treatment consists of physiotherapy and occupational therapy, consisting of range of motion exercises and strength training in order to strengthen the weakened musculature.⁴ This increase in muscular activity helps to raise the chances of recovery.⁵ Acetyl-L-Carnitine is an amino acid that works by preserving the vitality of the mitochondria inside the muscle cells and providing them with an alternative biofuel, thereby increasing recovery in spinal cord injury.⁶

The death rate for perinatal cervical cord injury is high and the ones that survive often have a grim prognosis with a future of being confined to a wheelchair. However, some patients recover as shown in one study where patients with incomplete quadriplegia were able to walk 1 year after injury, though some required crutches or braces. There was recovery in muscles of the upper and lower body.⁷

The latest research suggests that the central nervous system is not as inherently static as was once thought.⁸ It is actually capable of recovering quite significantly after injury by regeneration.⁹ Studies show that regeneration of neurons is possible provided there is correct and timely management and it is imperative to provide input and stimulation to make them recover.¹⁰ Therefore, the future looks promising as we discover more about the degree of plasticity the central nervous system can undergo, and which factors influence it.

Learning points

- ▶ Early intervention with steroids and physiotherapy can achieve a favourable outcome in near-total cervical cord transection, as quick management is essential to limit damage and allow improvement in neurological function.
- ▶ L-Carnitine greatly improves the function of muscles in spinal cord injury.
- ▶ Rapid radiological investigations such as MRI and CT are vital when a baby presents with weakness and hyporeflexia of the limbs, as well as axial hypotonia. This speeds up diagnosis and allows localisation of the lesion.
- ▶ Careful interpretation of MRI images of such birth injuries of the spinal cord is mandatory as significant oedema may obscure the transection and lead to a misdiagnosis. Therefore, a repeat MRI should be carried out in a few weeks after steroid therapy once the oedema has decreased, to give a clearer extent of the lesion which may not have been visible initially.
- ▶ We may possibly be looking at a case of regeneration of spinal cord neurons. This may have occurred in this case as the injury was in a neonate and, at this age, there are increased chances of complete recovery by regeneration due to plasticity of the spinal cord. This should be investigated more as it holds promising news for babies that suffer perinatal neurological trauma.

Competing interests None.

Patient consent Obtained.

REFERENCES

- 1 Franken E. Spinal cord injury in the newborn infant. *Pediatr Radiol* 1975;3:101–4.
- 2 Parashari UC, Khanduri S, Bhadury S, *et al*. Diagnostic and prognostic role of MRI in spinal trauma, its comparison and correlation with clinical profile and neurological outcome, according to ASIA impairment scale. *J Craniovertebr Junction Spine* 2011;2:17–26.
- 3 Bracken MB. Steroids for acute spinal cord injury. *Cochrane Database Syst Rev* 2012;18:CD001046.
- 4 Phillips AA, Cote AT, Warburton DE. A systematic review of exercise as a therapeutic intervention to improve arterial function in persons living with spinal cord injury. *Spinal Cord* 2011;49:702–14.
- 5 Frood R. The use of treadmill training to recover locomotor ability in patients with spinal cord injury. *Oxford J* 2010;4:108–17.
- 6 Patel SP, Sullivan PG, Lyttle TS, *et al*. Acetyl-L-carnitine ameliorates mitochondrial dysfunction following contusion spinal cord injury. *J Neurochem* 2010;114:291–301.
- 7 Waters RL, Adkins RH, Yakura JS, *et al*. Motor and sensory recovery following incomplete tetraplegia. *Arch Phys Med Rehabil* 1994;75:306–11.
- 8 Hagg T, Oudega M. Degenerative and spontaneous regenerative processes after spinal cord injury. *J Neurotrauma* 2006;23:264–80.
- 9 Bregman BS. Regeneration in the spinal cord. *Curr Opin Neurobiol* 1998;8:800–7.
- 10 Dietz V. Neuronal plasticity after a human spinal cord injury: positive and negative effects. *Exp Neurol* 2012;235:110–15.

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