Introduction

- Neonatal hypotonia, or the 'floppy' infant is a common reason for referral to paediatric neurology and usually prompts urgent investigation for a number of underlying aetiologies (usually of a hypoxic-ischaemic, genetic, metabolic or neuromuscular nature).
- Meticulous clinical assessment coupled with findings from blood, cerebrospinal fluid and radiological sources are usually sufficient to make a specific diagnosis.
- However in a small number of cases persistent hypotonia and peripheral weakness persists despite extensive investigation and relatively little in the way of pertinent clinical findings from history and examination.
- While rare, spinal cord injury sustained during the process of delivery may mimic causes of neonatal hypotonia, often with a trivial birth history and subtle findings radiologically.
- We present a rare case of neonatal cervical cord injury which was only diagnosed at six months of age.

Case

- A term male infant was born following emergency caesarian section for fetal tachycardia, breech presentation and meconium stained liquor.
- He needed brief resuscitation at birth but was well with APGAR 9 at 10 minutes. At one hour of age he was intubated and ventilated for severe hypoxia and respiratory distress. He was then extubated onto non-invasive ventilation after 24-48 hours. He remained on CPAP until day 16 of life and was able to be discharged home in air, however had persistent difficulties with feeding requiring the assistance of an NG tube.
- From birth he was noted to be hypotonic with extremely limited antigravity movements in all four limbs. However was alert and fixed and followed well.
- Initial baseline microarray, Prader-Willi screen, SMN1 and congenital myotonic dystrophy screen were normal. MRI brain in the early neonatal period was reported as normal.
- In the first 4-6 months of life, he was readmitted to hospital with frequent episodes of urosepsis. Additionally he developed an oxygen requirement at home, as well as evolving hypertonus affecting all four limbs.
- He Was admitted to paediatric critical care at approximately 6 months of life with metapneumovirus respiratory tract infection, where his underlying diagnosis was revisited.
- Urgent repeat MRI brain and spine revealed a C1 injury (comprised of T1 hypointensity and T2 hyperintensity on STIR sequencing (see Fig1). On review of previous imaging from birth, this injury was present.
- At 3 years of age he has four-limb hypertonia with clonus and spasticity, particularly • affecting his lower limbs and requiring regular gabapentin for symptom control.
- He continues to require nocturnal respiratory support with BiPAP.

NEONATAL SPINAL CORD INJURY: NOT TO BE MISSED

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Fig 1; Sagittal T1 weighted MRI brain showing C1 arch hypointensity in keeping with perinatal injury (white arrow), on initial scan in the early neonatal period.

Discussion

- While rare, neonatal spinal cord injury is well described in literature and believed to affect approximately 1 in 29,000 livebirths.
- While one might expect association with a significantly difficult In neonates with significant hypotonia in the absence of persistent delivery and extraction, numerous cases are described involving encephalopathy, spinal cord injury should be considered in the differential relatively minor, or even no difficulty during the birth process (both diagnosis. normal vaginal delivery and caesarian section).
- Early MRI brain AND spine are crucial in making the diagnosis can clinch the The precise mechanism of injury is not completely understood, diagnosis, avoid unnecessary investigations and also help to tailor appropriate however factors such as fetal macrosomia, shoulder dystocia, neck supportive management. hyperextension and rotation (especially during breech vaginal References delivery) and difficult instrumental/vacuum delivery are perceived risk factors.
- While the severity of the clinical sequelae will vary significantly depending on the nature and extent of the injury, neonates are often extremely unwell at birth, requiring periods of ventilation secondary to apnoea, with diminished spontaneous movements of the limbs, areflexia and poor response to tactile and painful stimulation. Therefore, it is not uncommon for infants to undergo therapeutic hypothermia in the context of suspected HIE.





- However, encephalopathy rarely persists in isolated spinal cord injury, and as such, babies may be misdiagnosed with a neuromuscular disorder such as spinal muscular atrophy type 1.
- Biochemical, genetic and radiological investigations of the brain and spine will not be in keeping with the aforementioned disorders.
- Radiological assessment of the brain AND spine is therefore of utmost importance in the investigation of persistent neonatal hypotonia without encephalopathy.
- MRI is the recommended modality for assessment of suspected spinal cord injury.
- Findings include focal cord swelling/enlargement on saggital T1, oedema on sagittal T2 weighted imaging (increased signal in the context of swelling), contusion suggested by a rim of mixed hyperintensity on T2 and hypointesity on T1, haemorrhage and even cord transection.
- Radiological changes may be very subtle however (especially early in the presentation) and false negatives are commonly reported. It is therefore prudent to repeat a radiological assessment of the brain and spine with MRI at a later stage if there is ongoing diagnostic uncertaintly.
- In this case, repeat evaluation confirmed the presence of a C1 injury which was retrospectively detected on re-evaluation of neonatal imaging.
- Clinical findings of symmetrical pyramidal weakness, hypertonia and bowel/bladder dysfunction were important clues to making the correct underlying diagnosis.
- The treatment options for neonates with spinal cord injuries are limited with a small and mixed evidence base. Reports concerning early use of systemic corticosteroid therapy and or therapeutic hypothermia suggest evidence of benefit, but the extent of this remains unclear given the broad spectrum of severity of injury and further data are needed.

Conclusion

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- 3) N.S. Mahmood, R. Kadavigere, K.R. Avinash et-al. Magnetic resonance imaging in acute cervical spinal cord injury: a correlative study on spinal cord changes and 1 month motor recovery. Spinal Cord. 2008;46 (12): 791-7. doi:10.1038/sc.2008.55