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Case Report

Neurophysiologic monitoring during cervical traction in a pediatric patient with severe cognitive disability and atlantoaxial instability

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ABSTRACT

Background: Surgical management of atlantoaxial instability (AAI) in pediatric patients with Down syndrome is associated with high neurological morbidity. Moreover, Down syndrome cognitive impairment coupled to AAI removes traditional verbal communication to relay evolving symptoms and aid in neurologic examination. It is not clear whether surgical adjuncts can alter clinical outcomes in this vulnerable population.

Case Description: Herein, we report the case of a 6-year-old patient with significant developmental delay and severe AAI that was successfully managed by stabilization with guidance of neurophysiologic investigations in the perioperative phase.

Conclusion: Perioperative neurophysiologic monitoring is safe, useful, and reliable in pediatric patients with trisomy 21 undergoing cervical traction and occipitocervical instrumented fusion for AAI.

Keywords: Atlantoaxial instability, C1-C2 instability, Cognitive impairment, Neurophysiologic monitoring, Trisomy 21

INTRODUCTION

Atlantoaxial instability (AAI) is a rare and potentially fatal disturbance of the normal occipital-cervical anatomy that affects some populations disproportionately.[10,14,18] Approximately 14–22% of patients having trisomy 21/Down syndrome develop AAI in their lifetime, while some case series report a prevalence of up to 60%.[3] This is secondary to ligament laxity and odontoid dysplasia.[2] If left untreated, this pathology can cause permanent neurologic deficits or sagittal deformity which leads to significant and premature mortality and morbidity.[4] At present, there is a lack of consensus to the best approach to diagnose, characterize, and treat this condition.[9] When surgical management is pursued, the role of perioperative neurophysiologic monitoring (NPM) also remains unclear.[13] We report herein a pediatric patient with AAI and trisomy 21, with significant cognitive impairment, in which operative management was sought and successfully accomplished with the use of atlantoaxial reduction and stabilization with the aid of NPM.

Patient's legal guardian provided written informed consent to publish this case and images herein contained. This study was exempt from ethical approval according to the Institutional Review Board of Geisinger Medical Center (IRB# 2020-0412).

CASE REPORT

History

A 6-year-old recently adopted boy from Asia was diagnosed with Down syndrome at birth. Due to progressive weakness in all limbs and ultimately arrest of ambulation, a neurological evaluation was sought. After initial assessment and imaging acquisition by our neurology colleagues, the patient was ultimately referred to our office. On initial interview, the patient's parents produced an historical video of relatively normal ambulation and occasional jumping at 2.5 years of age. At 3.5 years of age, the patient sustained a brief viral-like syndrome at which point his ambulation gradually ceased. His parents reported a substantial lack of strength to hold his neck or trunk and seemed to have no meaningful movement on his left side. He had no bladder control, but preserved bowel control. To compound his profound intellectual disability, his language barrier made neurological examination and interview quite challenging.

Examination

Physical examination revealed typical stigmata of Down syndrome. Significant cognitive impairment was noted without ability to verbally communicate despite translation services available. He was unable to walk independently; crawled on his hands and knees, pulled himself to standing, and was able to roll over to get into a sitting position. On neurologic examination, he had diffuse hyperreflexia and bilateral clonus, which was sustained on the right. He was noted to have spastic, 3/5 strength throughout, except 1/5 in the left upper extremity. Preferential hyperextension of his neck was also appreciated. On assessment of his ambulation, he was able to stand up with maximal assistance and inversion of his feet, but unable to walk.

A cervical spine MRI demonstrated a dystopic os odontoideum associated with significant atlantoaxial subluxation. Evidence of encephalomalacia at the craniocervical junction was also noted secondary to acute angulation of the cord [Figure 1]. Short-tau inversion recovery signal changes were noted at C1-C2 (not shown). MR of the brain was unremarkable. Given the clear clinical evidence of progressive quadriparesis with concern for further strength loss in his upper extremities, operative stabilization of his craniocervical segment was offered. We decided to immobilize the patient with a rigid cervical collar as a bridge until definite fixation.

Operation

Plan for staged surgical correction was selected. First, the decision was to proceed with closed reduction with traction to reduce instability and gradually correct his spinal alignment. His profound intellectual disability and language barrier, however, posed unique challenges since traction usually demands patient participation and real-time neurological examination. To circumvent this problem and minimize risk of permanent neurological insult, we opted for general endotracheal anesthesia before installation of Gardner-Wells tongs under NPM. Somatosensory-evoked potential (SSEP) and transcranial electric motor-evoked potentials (TceMEP) were employed. Mean arterial pressure (MAP) was kept >75 (baseline approximately 70) to ensure adequate cord perfusion. Pretraction SSEP signals were unreliable and thus we employed TceMEP responses throughout the case, as they were present and monitorable at baseline. A ≥80% decrease in amplitude was used as warning criteria to indicate cord dysfunction. In addition, a mean baseline stimulation threshold for the motor evoked potentials of 485 + 85 V (range 387–600 V) was used. Gardner-Wells tongs were then placed. Neuromonitoring detected decreased MEP signals even with the smallest changes in position [Figure 2a]. We then placed 3 pounds of weight and positioned his head in a more extended position with return to baseline neuromonitoring [Figure 2b]. Extended time was required, as at least an extra hour was required to get his head in an appropriate position that allowed for adequate spinal cord perfusion. Fluoroscopy demonstrated significant reduction of the atlantoaxial displacement with some distraction. TceMEP signals were at baseline at the end of procedure. At this juncture, given the lability of TceMEP and lack of reliable examination and patient cooperation, we decided to defer extubation and emergence from anesthesia until after definitive occipitocervical fixation and fusion. Consequently, he was admitted to the pediatric intensive care unit (PICU) for close MAP maintenance and frequent NPM. For this, neuromuscular blockade was reversed, and TceMEP signals assessed every 2–4 h at bedside. No electrophysiological changes were detected during this part of management.

Approximately 48 h from his original procedure, he returned to the operating room for occipitocervical fusion as planned. His previous Gardner-Wells tongs were removed and a Mayfield 3-head pin holder was placed. He was turned in the prone position. NPM was stable and fluoroscopy demonstrated adequate positioning. Incision was performed in the midline from the inion down to C3 and subperiosteal dissection was carried out bilaterally down the laminae of C1 down to C3 as well as the suboccipital region. The joint capsules at C2-C3 were maintained intact. Bilateral C2 nerve roots were sacrificed proximal to the dorsal root ganglion to allow for C1 screw placement. Instrumented arthrodesis was accomplished

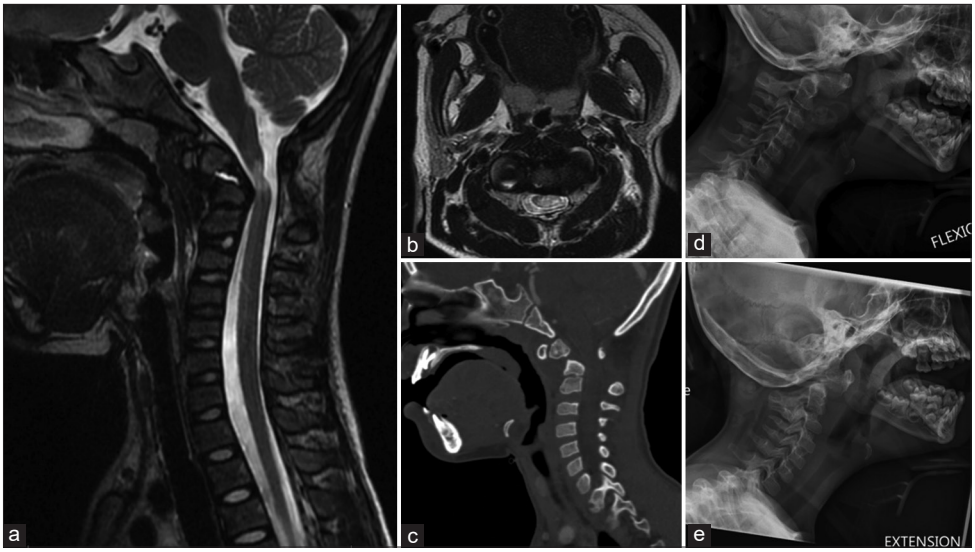

Figure 1: Imaging findings consistent with atlantoaxial subluxation and instability. (a) T2-weighted sagittal view of the cervical spine demonstrating sequelae of cord compression just below the level of the foramen magnum. The predental space is markedly increased measuring approximately 1.5 cm. Definite stenosis and cord compression at this level likely attributed to supine position during image acquisition. (b) The cord is flattened and of abnormal signal characteristic at C2 level, where there appears to be an anteriorly displaced os odontoideum. (c) Sagittal bone window of computerized tomography redemonstrating atlantoaxial subluxation and dystopic os odontoideum. Dynamic changes appreciated on plain radiographs in (d) flexion and (e) extension.

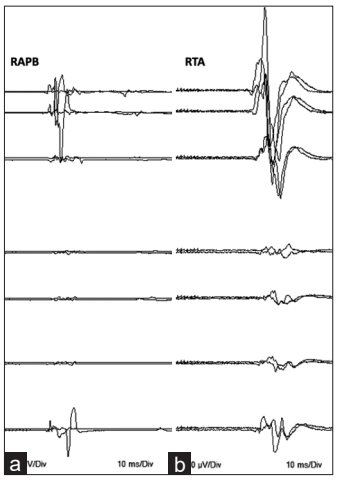

Figure 2: Transcranial motor-evoked potential recordings during closed cervical traction. Decreased amplitude in the right hand and leg noted during application of Gardner-Wells tongs and extension of neck (a) with subsequent return to baseline in the right lower extremity after initial weight placement and correction of cervical alignment (b). RAPB: Right abductor pollicis brevis, RTA: Right tibialis anterior.

as described previously.[1] Briefly, C2 pars screws were placed bilaterally at this point under fluoroscopic guidance. C1 lateral mass screws were subsequently placed, which were both left proud to be in-line with the rod. At this juncture, TceMEP demonstrated a decrease in the right hand [Figure 3a], thought to be secondary to excessive atlas mobility during screw placement despite the use of countertraction. The decision was then made to proceed with laminectomy of C1, at which point significant improvement was noted in TceMEP signals [Figure 3b]. An occipital plate with bicortical screws was used.

Decortication and autologous bone grafting with demineralized bone matrix were performed followed by rod placement. Importantly, further atlanto-occipital reduction was induced during final construct arrangement. Definitive arthrodesis was subsequently achieved by placing an iliac crest allograft between occiput and axis and securing with wires connected to the rods. At this point, electrophysiologic monitoring confirmed stability and our team proceeded to close the incision. A rigid cervical collar was placed and he subsequently transferred to the PICU for further management.

Postoperative course

The patient's immediate postoperative course was uneventful. In brief, he was kept mechanically ventilated under mild sedation. The MAP was carefully titrated to >70 mmHg for

the first 48 h after surgery. Ultimately, he was extubated on postoperative day 3 and a trial of oral intake was successful. His neurological examination remained at baseline with purposeful, antigravity, and symmetric movement in all extremities. He was discharged home on postoperative day 5 with occupational and physical services in place. During his wound check visit, the patient's parents reported good recovery aside from expected postoperative pain which subsided without further pharmacologic treatment. The patient tolerated cervical collar use. In addition, his parents reported potential motor improvements demonstrated by his attempts to stand up rather than crawl. Physical and occupational therapy continued.

Follow-up at 3 months after surgery revealed our patient displayed significant motor improvement with increased strength of his left upper extremity. Importantly, parents reported some ambulation with the aid of a walker. Follow-up at 2 years after surgery, his motor strength continued to improve. He demonstrated full strength in his right upper and lower extremities. Muscle tone was just slightly elevated on the left and normal on the right. No clonus on examination was noted with only trace hyperreflexia on the left lower extremity. He was able to open and close his left hand freely, bring it to his mouth and perform simple tasks of fine motor movements. He was able to dress and undress himself without any difficulty. Ambulation required only minimal assistance. Bladder incontinency improved considerably with reports of almost full volitional control of enuresis. His surgical incision healed without incidents. Interval imaging studies were also obtained demonstrating no instrumentation failure and adequate osseous fusion [Figure 4].

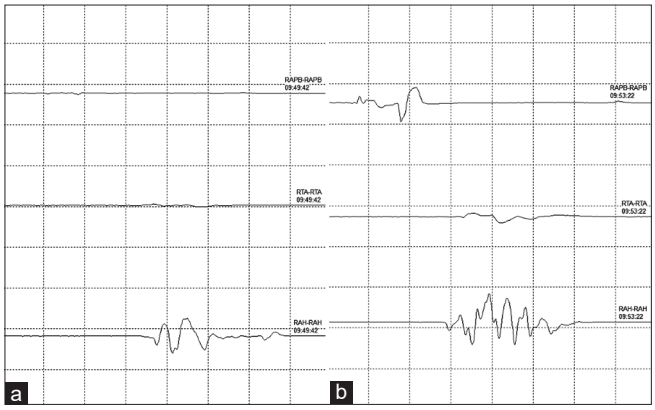

Figure 3: Transcranial motor-evoked potential recordings during occipitocervical instrumented fusion. Following C1 lateral mass placement, a decrease in amplitude was detected in the right hand thought to be secondary to excessive atlas mobility (a). Baseline signal acquisition occurred after rapid C1 laminectomy (b). RAPB: Right abductor pollicis brevis, RTA: Right tibialis anterior, RAH: Right abductor hallucis.

DISCUSSION

Here, we report the use of NPM as adjunct in the operative management of a pediatric patient with AAI and profound cognitive impairment. Our patient was a 6-year-old boy with Down syndrome who presented with significant atlantoaxial subluxation and worsening, progressive quadriparesis. As noted above, we sought to halt further progression by stabilizing his craniocervical junction. Specifically, closed traction before posterior occipitocervical instrumented arthrodesis was the selected approach. Importantly, the use of NPM reduced the potential for neurological complications and allowed us to circumvent our patient's language and cognitive deficits.

The role of intraoperative NPM in adult patients undergoing spinal operations has been established.[5] In pediatric patients, however, the utility of this surgical adjunct has not been clearly characterized.[15] This age group is as susceptible to

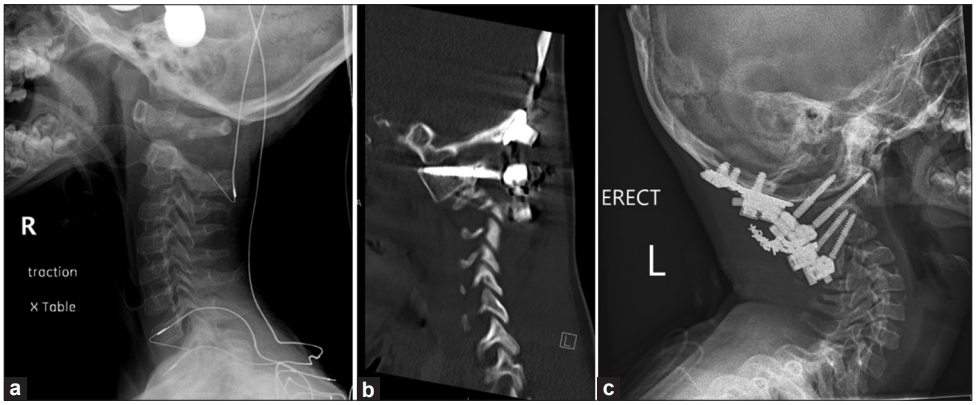

Figure 4: Intra and postoperative images demonstrating adequate reduction of atlantoaxial subluxation and occipitocervical fixation. Plain lateral radiograph of the cervical spine during traction with 3 pounds of weight; significant atlantoaxial reduction with some degree of distraction noted (a). Computerized tomography (b) and radiographic (c) studies performed 1 year after staged occipitocervical reduction and fixation with evidence of osseous fusion and no failure of instrumentation.

cord injury during spine surgery as adults[19] and could benefit as much but a more robust body of evidence is needed. It has been reported that multimodal intraoperative NPM is a safe and reliable tool in complex pediatric spine procedures,[15] especially during intramedullary pathologies.[7,11]

Since NPM is sensitive to the anesthesia technique,[16] neurological comorbidities, and extent of tract myelination, some groups have studied its use in children with underlying neuromuscular disorders[6] and trisomy 21/Down syndrome.[12] Intraoperative NPM can be safely utilized with standard anesthesia technique and stimulation parameters in patients with Down syndrome although the information it provides should be carefully interpreted and adequately applied.[12]

As previously mentioned, patients with Down syndrome have a higher prevalence of AAI. When clinical deterioration is detected, surgical management is often recommended. The rate of surgical complications in this patient population nonetheless remains high[17] and attempts to minimize risks should be pursued.

In our case, we first decided to promote an extension vector with closed cervical traction to correct alignment and reduce AAI. Given our patient's cognitive impairment and expected lack of cooperation, we relied on NPM to avoid potential complications. To the best of our knowledge, ours is the first report of the use of NPM in a pediatric patient with Down syndrome during cervical traction. While the SSEP responses were absent and thus not utilized, TceMEP consistently and reliably informed us the effects of maneuvers on cord integrity. This finding is in agreement with prior studies that have shown that SSEPs are less sensitive than MEPs to detect clinically significant cord injury,[8,12] although preexisting dorsal column dysfunction cannot be excluded in our patient.

The addition of perioperative NPM may have ultimately decreased the risk of neurological complications and could be considered as a valuable operative adjunct in this patient population not only during spinal surgery but also during cervical traction.

CONCLUSION

Perioperative NPM was useful and reliable in this pediatric patient with trisomy 21 undergoing cervical traction and occipitocervical instrumented fusion for AAI. This diagnostic modality can be considered in selected cases where there is a significant concern for neurological insult during preoperative closed traction as a bridge for definitive fixation.

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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