An infantile alantoaxial dislocation with patent foramen ovale managed with titanium cabling and allogenic bone grafts

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Abstract

Atlantoaxial dislocation is a disorder that is characterized with loss of stability of the atlas and axis (C1-C2) with consequential loss of usual articulation. Although this condition is very common, no one has reported a case as young as our patients. We present a 7-month infant with bilateral paralysis of the lower limbs for four (4) months with no history of trauma. Computer tomographic (CT-scan) imaging revealed alantoaxial dislocation with severe cervical spinal cord compression. The odontoid process is displaced outwardly with no bone destruction. Doppler echocardiogram done revealed patent foramen ovale. Thorough physical examination as well as radiological evaluation revealed no feather malformations. Electrophysiological studies reveal normal compound muscle action potentials (CMAP) and sensory nerve action potentials (SNAPs) in all the limbs. Electromyography (EMG) also revealed normal nerves in the limbs and the trunk. We attained a stable fusion and anatomical reduction using a posterior titanium wire and an iliac bone graft harvested from his mother. This is the youngest patient reported in literature. Infantile alantoaxial dislocation should be managed at early stage to prevent long-term neurologic disorders.

Introduction

Atlantoaxial dislocation is a disorder that is characterized with loss of stability of the atlas and axis (C1-C2) with consequential loss of usual articulation.[1-3] Functionally, the osseous articulations together with their enforcing ligaments essentially oppose forces in all directions of movement around the neck.[1,4] Destabilize of the atlas and axis which usually constitutes a junction may arise as a result of diverse disease situations such as traumatic, inflammatory, idiopathic, or congenital abnormalities.[1,2,4-8] This disorder can be substantially severe advancing disease with local pain, myelopathy or even death if the atlantoaxial instability is significate with no surgical intervention.[1,3,8,9] Furthermore, subluxations may be asymptomatic or present with inexplicable neuronal signs and symptoms.[1,4] Radiologically the distance between the anterior arch of the atlas and the odontoid is used in the diagnosis of atlantoaxial dislocation. This distance should not be greater than 3 mm in adults and 4.5 mm in children.[1-3,6-9]

In the management of patients with atlantoaxial dislocation, C1-C2 motion segment is precisely problematic.[7,9,10] Many surgeons advocated an initial anterior decompression preceded with stable posterior craniocervical junction fixation and ventral neuronal compression.[3,10] Also, to further decrease deformity, risk of potential neural damage as well as pain relief, posterior occipitocervical or atlantoaxial arthrodesis is recommended in patients with moderate to severe instability and in some instances asymptomatic patients.[1,6,10] Furthermore, rigid stabilization, maintenance of alignment, minimal postoperative immobilization, and enhanced fusion rates are the key advantages of internal fixation.[10] We present a case of congenital alantoaxial dislocation with patent foramen ovale managed with titanium cabling and allogenic bone grafts. To the best of our knowledge this is the youngest patient reported in literature and the first case with simultaneous occurrence of these two congenital disorders.

Ethics approval and consent to participate

The ethical committee of the hospital full approved our case study. The child's parents were informed about our intension to involve him in a case study and they agreed to partake in the study.

Consent for publication

The child's parents were dually informed about our intention to publish his case and they fully concerted to the use of these documents. The hospital also concerted to the use of this information for publication.

Case Report

We present a case of a 7-month infant with bilateral paralysis of the lower limbs for four (4) months. He was apparently doing well after birth until four months prior to presentation when the parents noticed that he could not move his neck freely and this progressed bilateral paralysis and severe irritability. He however had frequent upper repertory tract infection but no coughs. He did not have convulsions, feeding difficulties, painful micturition or passing stool. His parents denied history of any form of trauma during and after birth. Delivery was via spontaneous vagina delivery with no further instrumental assistance. His immunization was completed with his age.

On examination, we saw an infant who was warm to touch with a temperature of 38.8 degree Celsius and his neck in a cock robin posture. His head circumference was normal according to age with no abnormalities. Anterior and posterior fontanels were palpable but not bulging. Abdominal and chest examinations was unremarkable. Examination of the spine revealed tenderness at the cervical spine but no abnormities on thoracic and lumber spine. He however was not able to move his limbs. Further neurological examination did not yield

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much. The muscle bulk on all the limbs was normal with normal reflexes. Digital rectal examination revealed a normal spinster tone.

Laboratory investigations were normal but a Doppler echocardiogram done revealed patent foramen ovale. Computer tomographic (CT-scan) imaging revealed alantoaxial dislocation with severe cervical spinal cord compression. The odontoid process is displaced outwardly with no bone destruction. There is narrowing of the occipitocervical junction with a gross enlargement of distal cord (Figure 1A-D). 3D images also revealed a dislocation at the atlantoaxial joint (Figure 2). Electrophysiological studies reveal normal compound muscle action potentials (CMAP) and sensory nerve action potentials (SNAPs) in all the limbs. Electromyography (EMG) also revealed of normal nerves in the limbs and the truck. Thorough physical examination as well as radiological evaluation revealed no feather malformations. Based on the finding above, a working diagnosis of congenital atlantoaxial dislocation and patent foramen ovale with upper respiratory infection was made. After education and counselling of the patients, surgery was scheduled.

On the surgical table, we did not see any obvious abnormalities on skin around the neck (Figure 3A). After general anesthesia, the leads of the intraoperative neuromonitoring (IONM) which comprise of somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs) were inserted into the appropriate muscles to monitor the never on the upper limbs as well as low cranial nerves. We found atlantoaxial joint dislocation outwardly and frontal as well as dislocation of the odontoid process backwardly. There was narrowing of the occipitocervical junction with a gross enlargement of distal cord. We attained a stable fusion and anatomical reduction using a posterior titanium wire and an iliac bone graft harvested from the patient's mother (Figure 3B-D). Postoperative MRI (Figure 4A and B) and 3D imaging (Figure 4C and D) revealed a reduced atlantoaxial dislocation as well as titanium cabling and allogenic bone grafts in position. He was discharged from our ward ten (10) years after operation and scheduled cardiology outpatient follow-up arranged because of the patent foramen ovale. We also supported him with halo jacket for three months and he regained full motor function. Four (4) years follow-up revealed spontaneous closer of the patent foramen ovale and full motor function.

Discussion

Atlantoaxial dislocation is a disorder that is characterized with loss of stability of the atlas and axis (C1-C2) with consequential loss of usual articulation.[1-3] Functionally, the osseous articulations together with their enforcing ligaments essentially oppose forces in all directions of movement around the neck.[1,4] Although this disorder can present at any age, it has not been seen in infants in literature. It is often seen in adolescents and occasional children.[1,3,9] To the best of our knowledge this is the youngest patient reported in literature and the first case with simultaneous occurrence of these two congenital disorders. The mechanism of injury is still a matter of debate since no author has clearly

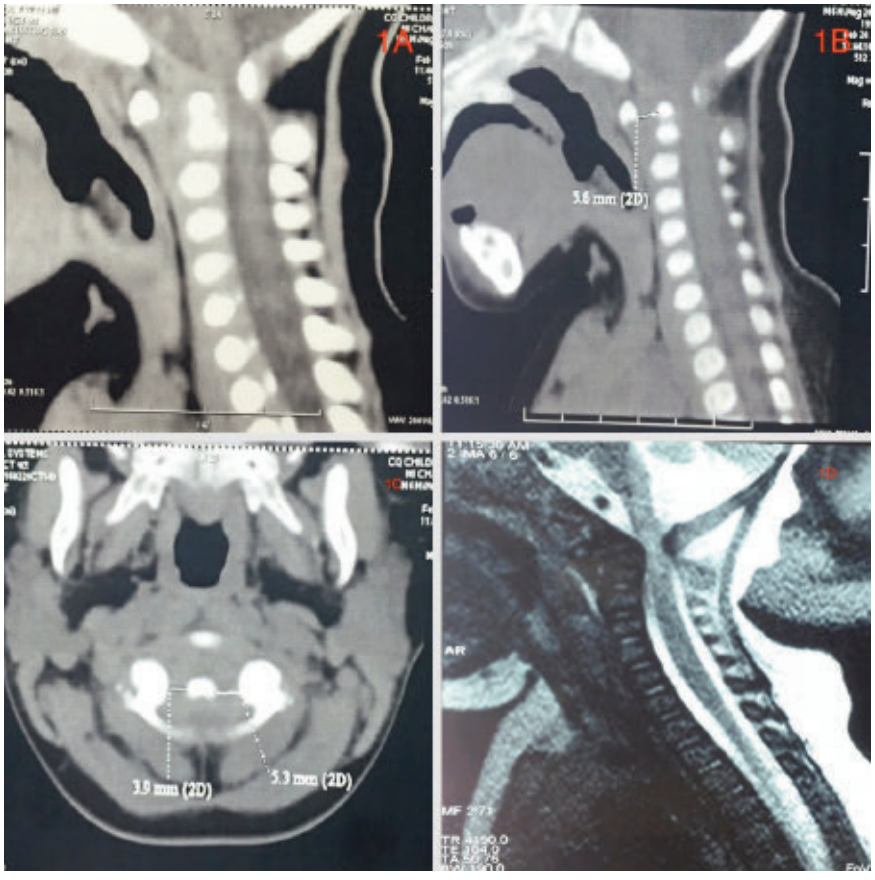

Figure 1. A-D) Are MRI showing the atlantoaxial dislocation with severe cervical spinal cord compression.

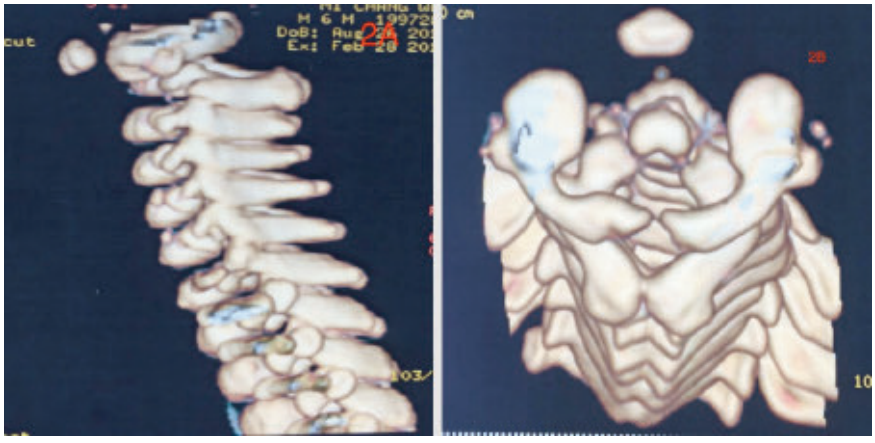

Figure 2. A-B) Are 3D images showing the atlantoaxial dislocation.

explained the pathological process involved in literature.[1] Many authors are of the view that the cause of atlantoaxial dislocation is generally multifactorial and can be broadly categorized into traumatic, congenital, or inflammatory etiologies.[1-8]

Traumatic atlantoaxial dislocation occurs when a force dislodges the neck from its original subluxation state leading to destruction of the transverse ligament.[1,11] Occasionally, there is concomitant interruption of the alar and apical ligaments as a result of injury of the transverse ligament.[1,11] The atlas may lose articulation with the dens, and the anterior atlantal arch may transmute totally superiorly and posteriorly with substantial destruction to the ligaments during these traumatic dislocations.[11-13] A number of congenital disorders have been linked to the development of craniocervical area distortions which predispose affected individuals to atlantoaxial dislocation.[1,14] Down syndrome (trisomy 21) is one of the congenital chromosomal syndrome is well known to be associated with atlantoaxial dislocation.[1,15] Hypermobility and instability as a result of ligamentous laxity and osseous deformities are the predisposing ramification of atlantoaxial dislocation occurring as result of downs syndrome in about 15-20% of cases.[1,16-18] Another congenital syndrome associated with atlantoaxial dislocation is spondyloepiphyseal dysplasia which comprises of numerous conditions and usually marked with anomalous development of the spinal vertebrae and epiphysis.[1] Short-trunk dwarfism, short proximal and middle limbs with however normal-sized hands and feet are cardinal signs seen in patients with spondyloepiphyseal dysplasia.[1,15] Furthermore, skeletal dysplasias are also a heterogeneous group of syndromes that are marked with anomalous cartilage and bone formation, growth as well as remodeling. Some of the conjoint spinal hitches that are associated with skeletal dysplasias are craniocervical junction anomalies, atlantoaxial dislocation, and kyphoscoliotic abnormalities.[1,15] Goldenhar syndrome is another heterogeneous condition marked with spinal anomalies, hemifacial microsomia, and epibulbar dermoid appendages. Hypoplasia of the dens with atlantoaxial instability has been seen in children with this syndrome.[1,15] The inflammatory condition that is commonly associated with atlantoaxial dislocation is chronic rheumatoid arthritis which mostly seen adult's patients. The incidence rates chronic rheumatoid arthritis advancing to atlantoaxial dislocation is about 23-86% and the atlantoaxial joint rheumatoid arthritis is usually the origin.[1,19-21]

Embryonically, the first and second cervical vertebrae have a distinct formation. The atlas or C1 is characterized with three primary ossification sites during it formation and these are, the anterior arch and the two neural arches which enclose the anterior arch and join as life advances to form the posterior arch.[4] The neural arches emerge during the 7th week of fetal life but the anterior arch ossifies at birth and seen as an ossification complex during the first year in about 20% of children. In many instances *nonfusion* the anterior arch with the neural arches may be mistaken for a fracture before age seven.[4,22,23] Furthermore, during the 3 year of life the neural arches fuse posteriorly but sporadically, the anterior ossification center of C1 is not formed and the neural arches try to fuse anteriorly leading to a fusion anomaly. This anomaly can be distinguished from a fracture because it displays sclerotic margins.[4]

The axis (C2), has the most multifarious and distinct formation pattern among all the vertebrae.[4] It comprises of four ossification centers when the child is born, usually one for each neural arch, one for the body, and one for the odontoid process. The odontoid process often develops in utero during the 7th fetal month from two separate ossification centers that fuse in the midline.[4] Furthermore, between 3 and 6 (years) as the child advances with age, a secondary ossification center appears at the apex of the odontoid process (os terminale) and fuses by age 12 (years).[4] It is however important to note that the body of C2 fuses with the odontoid process between age three and six. This fusion line also referred to as subdental synchondrosis, or the remnant of the cartilaginous synchondrosis is usually visible up to age 11 (years) and may be mistaken for a fracture. The neural arches fuse posteriorly between the ages of 2 and 3 (years) and with the body of the odontoid process between 3 and 6 years of age.[4,22,23]

The manifestations of atlantoaxial dislocation may vary from slight axial neck pain to very severe debilitating illness and finally death. Nearly 50% of cases are seen

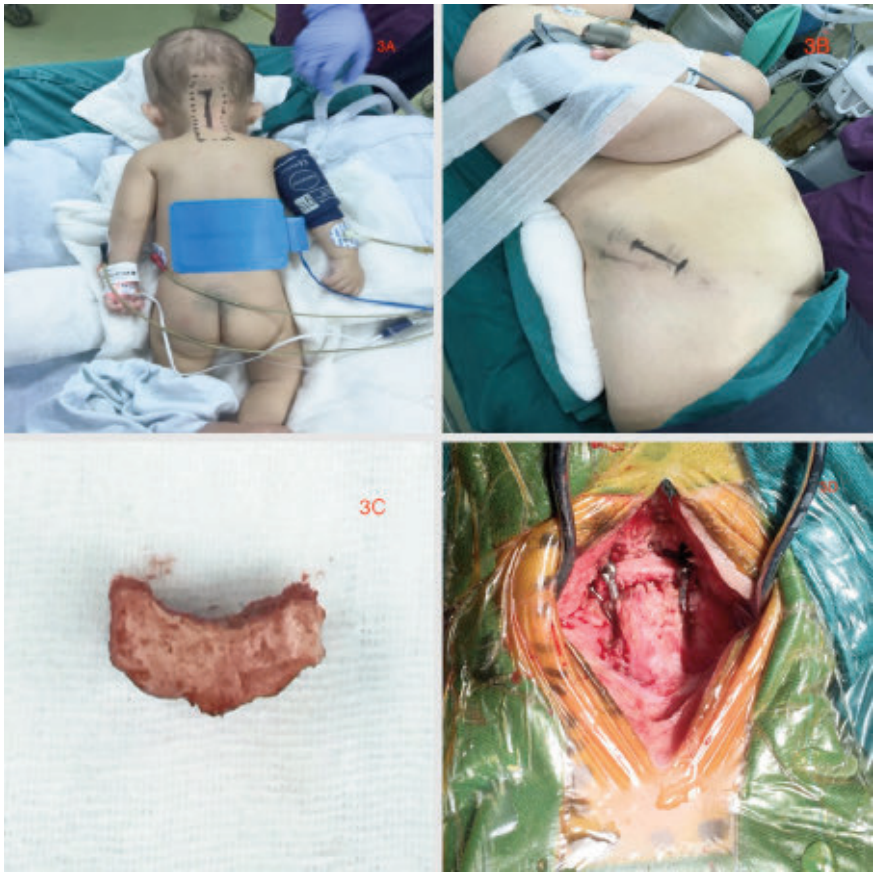

Figure 3. A) Showing child on the surgical table with a marking of the incision site; B) showing the patient's mother prepped for iliac bone graft harvest; C) showing the harvested bone graft with a slight curvature in the middle; and D) showing intraoperative C1-C2 fusion with the bone graft fixed by titanium wires.

with neck pain and/or neck movement restriction while 70% with weakness and/or numbness, and 90% with pyramidal signs.[1,24-26] Low sphincter tone and lower cranial nerve dysfunction have also been reported as the signs and symptoms of this disorder.[1] Furthermore, severe clinical manifestations such as myelopathy, respiratory distress or respiratory failure, vertebral artery dissection, neurologic compromise, and rarely quadriplegia or death if left untreated have also been reported.[1,27-31] Atlantoaxial dislocation should be suspected in a child who get irritated or reluctant turning their head when signs and symptoms do not match with the diagnosis of torticollis.[1,32] The clinical manifestation of atlantoaxial dislocation in adults has generally been due to rheumatoid arthritis, with varying symptoms such as instability and neurologic compromise. Torticollis, atlantoaxial rotary fixation, and odontoid fractures without atlantoaxial dislocation should usually be ruled out during the evaluation of patients with atlantoaxial dislocation.[1]

The distance between the anterior arch of the atlas and the odontoid is the most crucial radiological feature used to diagnose atlantoaxial dislocation. The presentation is very suggestive when this distance is greater than 3 mm in adults and 4.5 mm in children.[1-3,6-9] Furthermore, the distance between the postero-inferior rim of the anterior arch of atlas and the residual segment of the odontoid or the anterior superior edge of the body of axis should be suggestive in cases with hypoplastic odontoid.[9] Plain radiographs of the craniovertebral junction usually showing translateral views in neutral position, flexion and extension are advantageous in arriving at preliminary conclusion of atlantoaxial dislocation.[9] This usually serve as a reference for the successive ongoing evaluation of the degree of disruption and the alteration in curvature on the application of a traction; and, can be associated with the postoperative plain radiographs in deciding the appropriateness of posterior stabilization and bony union.[9]

Multiplanar evaluation with inflexion and extension using intrathecal contrast CT scan with axial, sagittal, coronal views as well as three-dimensional reconstructions is usually recommended for a three-dimensional evaluation of the bony configuration of the craniovertebral junction.[9] This is beneficial during the evaluating because it's able to determine the degree of cervicomedullary compression, presence of Chiari malformation as well as appropriate delineation of bony anomalies.[9] Furthermore, a preoperative MRI aids in the evaluation of soft tissue abnormalities such as syringomyelia and the degree of cervicomedullary compression as well as cord vicissitudes. However, an MRI at follow-up is recommended in situation in which titanium implants are used but not possible in cases where metallic wires or rods used as implants for posterior stabilization.[9] This makes our treatment option more appropriate because we can follow-up our case with MRI.[9]

Greenberg is one of the early clinicians who first proposed a classification system for atlantoaxial dislocation. He subcategorized this dislocation into reducible and irreducible and proposed management approach to this subcategorise.[1,33] He precisely specified that the management of irreducible atlantoaxial dislocation should be focused on immediate decompression as well as stabilization.[1,33] Fielding and Hawkins later come out with a new classification system which focuses on the direction of dislocation that is anterior, posterior, lateral, and rotational but their classification has setbacks.[1,34] Wang *et al.* classification system is most recent and novel because it gives a distinctive diagnosis and management decorum for patients with atlantoaxial dislocation.[1,35] The management algorithm proposed by Wang *et al.* which comprises of preoperative assessment using dynamic radiographs, reconstructive CT, and skeletal traction test. This classification therefore categorizes atlantoaxial dislocation into four types which includes, instability as type I, reducible dislocation as type II, irre-

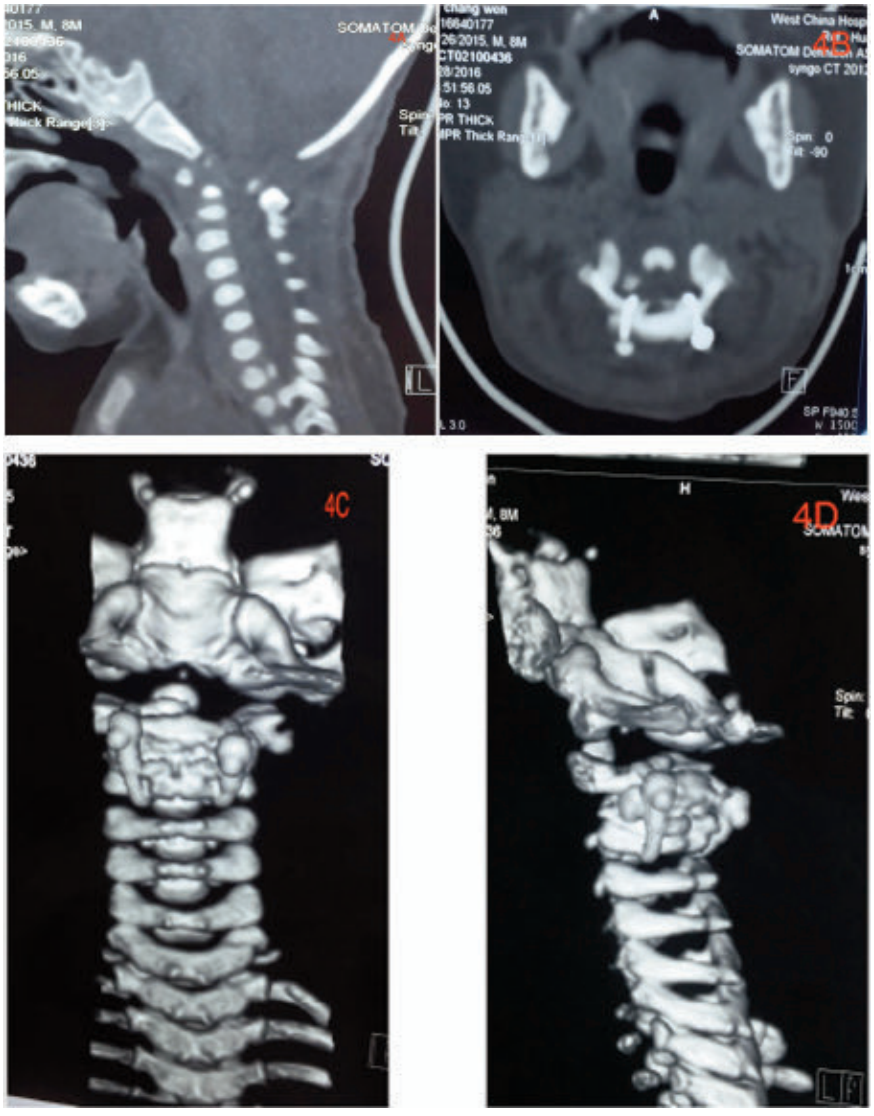

Figure 4. A-B) Are postoperative MRI showing good anatomical reduction; while C-D) are 3D postoperative images also showing good anatomical reduction.

ducible dislocation as type III, and bony dislocations as type IV.[1,35]

The general purpose of managing atlantoaxial dislocation is to rectify the sagittal structure of the upper cervical spine and the stabilization that is close to normal anatomical configuration.[1,8,36] Up to data, there is still no homogeneously recognized technique or management approaches to atlantoaxial dislocation. Therefore, we still have colossally anecdotal views on indications of nonoperative *versus* operative management options as well as which operative techniques are most suitable.[1,8,18,37-41]

Nonoperative management of atlantoaxial dislocation comprises of the application of a cervical halter traction in the supine position and active range-of-motion exercises for 24 to 48 hours first, preceded with ambulatory orthotic immobilization with active range-of-motion maneuvers until free movement returns.[1,42] Nonoperative treatment modality is recommended for children presenting acutely with evidence of transverse ligament disorder with no neurologic injury, diagnosed within 3 weeks.[1,43] This treatment option is also advocated for preventative treatment and screening as well. Preventative treatment and screening options is usually advocated in patients with Down syndrome.[1]

As part of their routine evaluation, serial cervical radiographs are done between 3 and 5 years of age and have their cervical canal width measured.[1,44] The ideal treatment modality for a subset of patients with Down syndrome with associated asymptomatic atlantoaxial dislocation is still a matter of debate.[1,45] Furthermore, cervical flexion-extension films every 6 months without contact sports is advocated in patients with Goldenhar syndrome with an atlantodental interval less than 6 mm. However, in any child with instability greater than 6 mm, surgical treatment is advocated to decrease the likelihood of disastrous spinal cord impingement. Moreover, patients with Goldenhar syndrome may necessitate extensive radiographic evaluation and planning before surgery due to the excessive incidence of cervical malformations.[1,46]

Some authors advice that attempts should be made at conversion of irreducible atlantoaxial dislocation to reducible via Traction.[1,8,34,41] Diverse intraoperative or preoperative modalities should be exploited to achieve reduction via traction when the dislocation is not effortlessly reducible by neck flexion or extension.[1] Traction stretches and slackens muscles across the dislocation, thus allowing resolving of structures into normal anatomical position. Furthermore, some authors advocate curarization of back and neck muscles to relieve tension before applying skeletal traction under general anesthesia. This allows for a rapid reduction before fixation in the same surgical operation.[1,8,18,41] The perfect reduction technique of curarization last about 10 minutes with an initial traction weight of 7-8% of total body weight and progressively adjusted to a maximum of about 7 kg. However, reduction ought to be monitored with serial lateral radiographs.[1,8,37,47]

Anterior surgical approach is advised in patients with irreducible dislocation with progressive cord compression leading to profound neurologic deficit after traction.[1,8,35] We advocate that intraoperative electrophysiological studies such as compound muscle action potentials (CMAP) and sensory nerve action potentials (SNAPs) be monitored. Electromyography (EMG) should also be applied to monitor nerves in the limbs and the trunk.[48] Intraoperatively, some authors advocate that irreducible atlantoaxial dislocation be decompressed prior to fixation although it is proven that neurologic status can improve with only effective fusion. Transoral odontoidectomy is now, the most recognized treatment modality for irreducible atlantoaxial dislocation although other anteriorly approaches like transoral anterior release and transoral atlantoaxial reduction plate (TARP), have also proven to be effective.[1,8,49] Anterior transarticular screw fixation another anterior approach used to reduce fixed atlantoaxial dislocation. This approach offers a securer and extra suitable milieu with high fusion rates and negligible hitches for difficult cases like in patients with osteolysis in infectious or tumorous disorders, aberrant vertebral artery, and/or narrow pars interarticularis.[1,8,50] However, safe screw dimension, insertion, and trajectory are the major short comings with the approach.[1]

The TARP is advantageous because it avoids the usual two-step operation. This technique utilizes a reduction plate that efficiently aid in anterior decompression, reduction, and fusion for anterior irreducible atlantoaxial dislocation in one operation.[1,8,51-53] In this technique, incision of the posterior pharynx and exposure of C1 and C2 are totally analogous to the other transoral approaches. The TARP is inserted at anterior portion of C1 while the reduction screw is then implanted into the C2 body. One notch of the TARP system secures the TARP at the midline and the other secures the protruding reduction screw, initiating interference between C1 and C2 resulting shifting of C1 superiorly with the reduction screw as a base. The joint is then fused with bone graft.[1,8,51,52] In the transoral odontoidectomy however, the apical and alar ligaments are separated with a curette followed by amputation of the dens from top down.[1,8,49] Furthermore, odontoidectomy endoscopically through a transnasal, transoral, or retropharyngeal approach is the now the treatment option adapted by most surgeons. The endoscopic transoral odontoidectomy system gains access vie an incision in the midline pharyngeal wall, circumventing any palatal piercing or use of self-retaining retractors. This approach has so far gained recognition because it improves neurologic status after surgery.[1,8,54] However, cerebrospinal fluid leakage, inadequate decompression, infection, abscess formation, vertebral artery injury, and spinal cord injury have been re-counted with transoral decompression.[1,8,40,49,55,56]

C1–C2 transarticular screw fixation, C1 lateral mass screw–to–C2 pedicle screw fixation, and C1 lateral mass screw–to–C2 laminar screw fixation are the most recognized posterior surgical approaches used in reducing atlantoaxial dislocation.[1,8] Posterior approaches are either use alone or in combination with anterior transoral decompression to manage certain kinds of irreducible atlantoaxial dislocation.[1,8] Gallie or Brooks technique of wiring and bone grafting to fuse and stabilize the atlantoaxial joint is original surgical managements for atlantoaxial dislocation.[1,8,57] This technique is rare used because posterior wiring involves external immobilization (bracing) of the head and neck postoperatively and lengthier stay in the hospital with a nonunion rate of about 30%.[1,8,58-61]

Magerl in 1986 described a much more better technique which involves C1–C2 transarticular screw fixation with posterior wiring. This technique was widely accepted as the gold standard treatment of atlantoaxial dislocation because of its superior biomechanical strength and high fusion rates as compared to posterior wiring.[1,8,61] Furthermore, the fusion rate of this technique is as high as 100%.[1,8,62-64] This technique is effective because it offers instant and direct fixation at the atlantoaxial joint. Intraoperatively, a midline incision is made to expose the posterior arch of C1 and lamina of C2 followed by the screw which positioned anteriorly through the C2 pars, across the C1–C2 joint, and into the anterior aspect of the lateral mass of the atlas.[1,8,62,63]

In 1995, McGuire and Harkey presented a modification of Magerl technique to curtail tissue dissection in which a negligible midline incision is accompanied with two bilateral 1-cmcaudal percutaneous incisions of about 2 cm lateral to the T1 spinous process.[1,8,62,65,66] Furthermore, another modified Magerl technique that utilizes

transarticular screw fixation with morselized autograft without posterior wiring has been described.[1,39] This technique has less biomechanical strength as compare to the Magerl technique. Additionally, patients well-maintained a high fusion rate and the risk of neurologic hitches as well as slackening of cables associated with posterior wiring.[1,39] The C1 lateral mass screw and C2 pedicle screw fixation (C1LC2PSF) utilizes a C1 lateral mass screw technique in combination with a C2 pedicle screw fixation connected by rods to stabilize the atlantoaxial joint, though certain hospitals favor plates connecting the fixation points.[1,60,67,68] The technique allows for extension to the occiput or subaxially if necessary, and screw positioning is more accurate.[1,69,70] Moreover, it permits intraoperative reduction after screw fixation.[1,60]

Yeom and colleagues compared the risk of vertebral artery injury in C1–C2 transarticular screw *versus* C2 pedicle screw and discovered that the two methods did not vary in risk of vertebral artery injury so they suggested the use of pedicle screw over transarticular screw in cases of high riding vertebral artery.[1,8,32] Moreover, the C1 lateral mass screw and C2 laminar screw fixation (C1L-C2LSF) method was explained in 2004 and it involves the use of a crossed screw method across the C2 lamina. This technique curtails the risk of vertebral artery injury as compared to transarticular screw fixation and C1L-C2PSF. It also permits direct visualization of the lamina to guide screw positioning intraoperatively.[1,8,11,26,45]

Conclusions

This is the youngest patient reported in literature. Atlantoaxial dislocation should be suspected in an infant who gets irritable when the caregivers try to move his or her neck. The cause of atlantoaxial dislocation is generally multifactorial and can be broadly categorized into separate traumatic, congenital, or inflammatory etiologies. The gold standard diagnostic modality for atlantoaxial dislocation is radiology. Infantile alantoaxial dislocation should be managed at early stage to prevent long-term neurologic disorders. Although so many surgical techniques have been described in literature, no one surgical approach has been singled out as best. We are therefore of the view that surgery approaches should be the surgeon's choice but must have limited complications.

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