Atlantoaxial Rotatory Subluxation
A Review for the Pediatric Emergency Physician

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Abstract: Pediatric emergency physicians must have a high clinical suspicion for atlantoaxial rotatory subluxation (AARS), particularly when a child presents with neck pain and an abnormal head posture without the ability to return to a neutral position. As shown in the neurosurgical literature, timely diagnosis and swift initiation of treatment have a greater chance of treatment success for the patient. However, timely treatment is complicated because torticollis can result from a variety of maladies, including: congenital abnormalities involving the C1-C2 joint or the surrounding supporting muscles and ligaments, central nervous system abnormalities, obstetric palsies from brachial plexus injuries, clavicle fractures, head and neck surgery, and infection. The treating pediatrician must discern the etiology of the underlying problem to determine both timing and treatment paradigms, which vary widely between these illnesses. We present a comprehensive review of AARS that is intended for pediatric emergency physicians. Management of AARS can vary widely bases on factors, such as duration of symptoms, as well as the patient's history. The goal of this review is to streamline the management paradigms and provide an inclusive review for pediatric emergency first responders.

Key Words: atlantoaxial subluxation, rotary subluxation, cervical spine trauma, axis, atlas, torticollis, trauma, AARS

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Pediatric emergency physicians may encounter atlantoaxial rotatory subluxation (AARS) in certain clinical circumstances. Although the majority of children with wry neck will have self-limited muscular torticollis, there is a selective subset of patient that may have AARS. Children predisposed to AARS may have Down syndromes or may have incurred severe trauma. Moreover, this should also be suspected in children presenting with neck pain and an abnormal head posture without the ability to return to a neutral position. As shown in the neurosurgical literature, timely diagnosis and swift initiation of treatment have a greater chance of treatment success for the patient.[1–4] However, timely treatment is complicated as torticollis can result from a variety of maladies, including: congenital abnormalities involving the C1-C2 joint or the surrounding supporting muscles and ligaments, central nervous system abnormalities, obstetric palsies from brachial plexus injuries, clavicle fractures, head and neck surgery, and infection.[5,6] The treating pediatrician must discern the etiology of the underlying problem to determine both timing and treatment paradigms, which vary widely between these illnesses.

Torticollis stemming from rotatory fixation of the atlantoaxial joint can occur spontaneously, due to trauma, infection, congenital abnormalities of the craniovertebral junction, or even from arthritidies.[3,7–11] The incidence of pediatric cervical spine trauma ranges between 1% and 4% of pediatric trauma admissions; however, the patterns of the cervical spine injury differ by patient age group.[1,12–16] Younger patients (0-9 years) are more likely to have injuries to the craniovertebral junction or spinal cord, and have a higher incidence of ligamentous injuries as compared with bony fractures.[12,17] This is due to the unique anatomic features of the pediatric spine that make it far more flexible than the adult spine and does not begin to mature until 9 years of age.[1,12]

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ETIOLOGY OF ATLANTOAXIAL ROTATORY SUBLUXATION

Pediatric Patients Are Anatomically Predisposed to AARS

The pediatric spine and the craniovertebral junction, in particular, are susceptible to dislocations and subluxations after relatively minor trauma or infection. Atlantoaxial rotatory subluxation is more common in younger children (<13 years) due to several unique and biomechanical features of the pediatric craniovertebral junction.[3,8,10] The ligaments and joints of the pediatric spine are more lax and can expand considerably tolerating trauma without tearing. The shallowness and horizontal orientation of the facet joints allow greater mobility, whereas the disproportionately larger head of the child can lead to more forceful and exaggerated movements causing greater stress to the upper cervical spine and predisposing the children to AARS.[1] Furthermore, the angle between the odontoid and the facets is steeper in younger children, making them more prone to subluxation. A cadaver study by Kawabe et al compared the anatomy of the infantile atlantoaxial joint to adult specimens of older than 65 years to help better explain the pathophysiology why AARS is more common in the younger pediatric population and found that infants have synovial folds attached to the capsule of the atlanto-occipital and atlantoaxial joints which can become trapped in the joint space with during extreme rotation, causing the subluxation and fixation which can help explain why children are more prone to AARS than adults.[1,3,18]

Surgery and Upper Respiratory Infections Are Associated With Torticollis

Torticollis associated with recent upper respiratory infection (URI) or recent head and neck surgery is well documented in the literature. The vast majority of children presenting with wry neck in this setting will have muscular torticollis. Respiratory illnesses are one of the most common reasons for pediatric emergency department visits for children under the age of 17 years in the United States, accounting for approximately 1 million visits per year.[19,20] Aëtius of Amida (circa 550 AD) was the first to associate the inflammation caused by a retropharyngeal abscess in the cervical spine with cervical subluxation.[5] The etiology of recent URI or surgery resulting in muscular torticollis is not well understood. It has been postulated that recent infection or surgery causes a local lymphadenitis which in turn produces cervical muscle spasm or a

hyperemia to the ligaments of the craniovertebral junction.[5] In children, who already have an inherent ligamentous laxity, this can cause muscular torticollis. Although muscular torticollis is seen in the overwhelming majority of children with wry neck, in select cases, AARS may be considered in the differential diagnosis. For example, recent URI or head and neck surgery causes periligamentous inflammation that causes distention and laxity of the craniovertebral joints and ligaments and can subsequently results in AARS.[3,5,10] Venous connections between pharynx, retropharyngeal space, periodontoidal area, perivertebral area, and suboccipital sinus are believed to provide a conduit for hematogenous spread of infection or inflammatory cytokines.[3,21–24] Moreover, this constellation of symptoms seen in URI that progress beyond muscular torticollis and may rarely develop Grisel syndrome.[21,23] Children with Grisel syndrome have a subluxation of the atlantoaxial joint, as a result of inflammation from adjacent tissue.[5,10,23]

Congenital Anomalies Predispose Patients to AARS

Congenital anomalies of the C1-C2 joint can also result in AARS. It has been reported that up to 5% of fetus have vertebral anomalies and approximately 1 in 40,000 to 42,000 children born worldwide will have a cervical spine anomaly.[17,25,26] Occipitalization of C1, either complete or incomplete, can tax the altantoaxial joint and result in AARS and other development anomalies affecting C1, C2, dens, atlantoaxial joint, and the craniovertebral junction. Development syndromes also associated with AARS include: Down syndrome, achondroplasia, spondyloepiphyseal dysplasia, Larsen syndrome, Klippel-Feil syndrome, and Morquio syndrome. Patients with these conditions are at especially high risk for AARS.[17,27]

BIOMECHANICS OF ATLANTOAXIAL ROTATORY SUBLUXATION

The C1-C2 joint is the most active joint in the body, moving on average about 600 times per hour.[28] The cervical spine can rotate up to 90 degrees with approximately 60% of the rotation accounted from the atlantoaxial joint, assisted by the horizontal orientation of the C1-C2 facet joints that allow increased rotation without compromising bony stability.[3,11,29] The ligamentous configuration of the atlantoaxial joint are optimized to stabilize this high degree of rotational movement. The transverse ligament keeps the odontoid opposed to the anterior arch of C1 and prevents excessive anterior motion of the atlas on the axis during movement. The paired alar ligaments which run from the tip of the odontoid process to the right and left occipital condyles also prevent excess anterior subluxation and function with the thickened C1-C2 capsular ligaments to resist excessive rotational movement.[3,29–31]

Physiologic rotation of C1 on C2 quoted in the literature between 25 and 53 degrees. In 1995, Pang and Li found that C1 moves independently until about 20 degrees of rotation, at which point C2 rotates in the same direction as C1 due to "yolking" of C2 by C1, with C1 rotating faster. At 65 degrees, both C1 and C2 rotate at the same speed with a fixed maximum separation angle of 45 degrees.[18,32] According to a review article by Roche et al, hypermobility of C1 on C2 occurs when atlantoaxial rotation is in excess of 56 degrees or a right-left difference greater than 8 degrees; rotation of C1 on C2 greater than 64 degrees will cause bifacetal dislocation. Hypomobility of C1 on C2 at maximal rotation is diagnosed when the angle is less than 28 degrees.[29] It is also important to remember that during rotation, the spinal canal at the C1-C2 level narrows because the movement of ipsilateral lateral mass swings out into the spinal canal causing a physiologic narrowing as C1 rotates on C2 (Fig. 1). Of great significance, the vertebral artery is intimately associated with the atlantoaxial joint and endures some physiologic stenosis particularly with excessive rotation coupled with anterior displacement. In extreme cases, AARS can cause brainstem and cerebellar infarct and even death.[3,33]

CLINICAL PRESENTATION OF AARS

Children with AARS present with persistent torticollis and a decreased neck range of motion, in a condition described as a "wry" neck. The head will be tilted to 1 side, slightly flexed, whereas the chin will be rotated to the contralateral side. This is known as the "cock-robin" position, because it looks similar to that of a robin listening for a worm.[1,3,33] A child with AARS is able to turn their head to exaggerate the deformity and may be able to rotate in the contralateral direction to partially reduce the deformity to the neutral position. Children with AARS most commonly present after a trauma, which is often minor, or after a recent URI or head and neck surgery.

Atlantoaxial rotatory subluxation usually presents without neurologic deficits; however, it can be associated with cervical myelopathy in cases with significant compression of the upper cervical spinal cord. Atlantoaxial rotatory subluxation can also present with an occipital neuralgia ipsilateral to the side of chin rotation. This happens because the greater occipital nerve can be stretched and irritated from the subluxation during the nerve's long course as it ascends from the dorsal ramus of C2, hooking around the posterior arch of atlas and enters the atlantoaxial joint space.[1–3]

The most accepted classification scheme for pediatric AARS was developed by Fielding and Hawkins[7] in 1977 in a study of 17 patients with AARS, and categorizes patients into 4 based on imaging findings on plain radiographs and cineradiography. Type

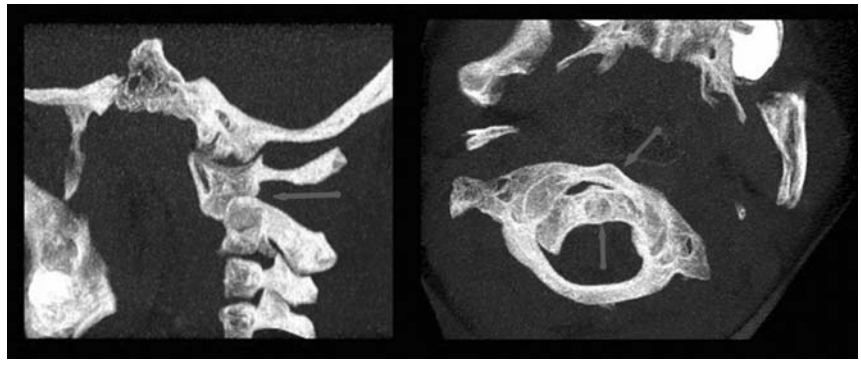

FIGURE 1. Sagittal and axial CT scans showing atlantoaxial rotatory subluxation of C1 on C2 with narrowing of the spinal canal.

I is a pure rotatory fixation without anterior subluxation and is the most common type of AARS. In type I AARS, the odontoid process acts as the pivot point. Because there is no anterior displacement, the atlantoaxial ligamentous structures are intact (Fig. 2A). In type II AARS, the second most common category of AARS, there is ligamentous disruption of the transverse ligament. Instead of the odontoid process, one of the articular masses acts as the pivot point, and there is 3 to 5 mm of anterior displacement of the anterior arch of C1 in relation to the odontoid (Fig. 2B). Fielding and Hawkins[7] type III AARS is very similar to type II, but with greater than 5 mm of anterior displacement of the arch of C1 and associated disruption of the transverse and alar ligaments (Fig. 2C). Type IV is the rarest type of AARS and is rotatory fixation with posterior displacement (Fig. 2D). This is associated with concomitant fractures or congenital anomalies of the odontoid process.[1,3,7]

More recently, Pang and Li proposed an alternate classification scheme of AARS based on the rotational dynamics of C1 on C2.[7] In their type I injuries, the most severe category, C1 and C2, are locked regardless of any type of counter corrective rotation. Type II injures show a reduction in the C1 on C2 angle with forced reduction, but C1 cannot be made to cross over C2. In type III injuries, C1 can be made to crossover C2 with forced rotation far to the opposite side of head rotation. Type IV is a type of AARS injury, referred as the "diagnostic grey zone" where the rotational dynamics of C1 and C2 lie between type III injury and normal.[1,32] Such classification schemes are important because the prognosis of success of treatment is based on severity of the injury and the timing of treatment.[1–4]

DIAGNOSING AARS

Distinguishing torticollis caused by AARS from muscular torticollis is difficult. There are 3 clinical signs to help differentiate AARS from muscular torticollis: (1) Sudek et al described there will be a palpable deviation of C2 spinous process in the same direction as head rotation in patients with AARS; (2) the ipsilateral sternocleidomastoid muscle (SCM) may spasm in an attempt to reduce the deformity because the normal action of the SCM is to rotate the head contralateral to the contraction, whereas in muscular torticollis, the opposite SCM is contracted, potentially causing the deformity; and (3) the inability to counter-rotate the head past midline.[3,4,34] A fourth finding on physical examination described in other studies is a bulge in the back wall of the pharynx representing anterior displacement of the arch of C1. In longstanding cases of AARS, the child's skull and face may be flattened ipsilateral to the side of the head tilt.[3]

Diagnosis is challenging, especially in the pediatric patient. Based on the patient's history, the pediatrician must have a high index of suspicion and confirm the diagnosis radiographically, which can be difficult. According to a recent review by Tat et al,[12] currently, there exists no definitive clinical criteria

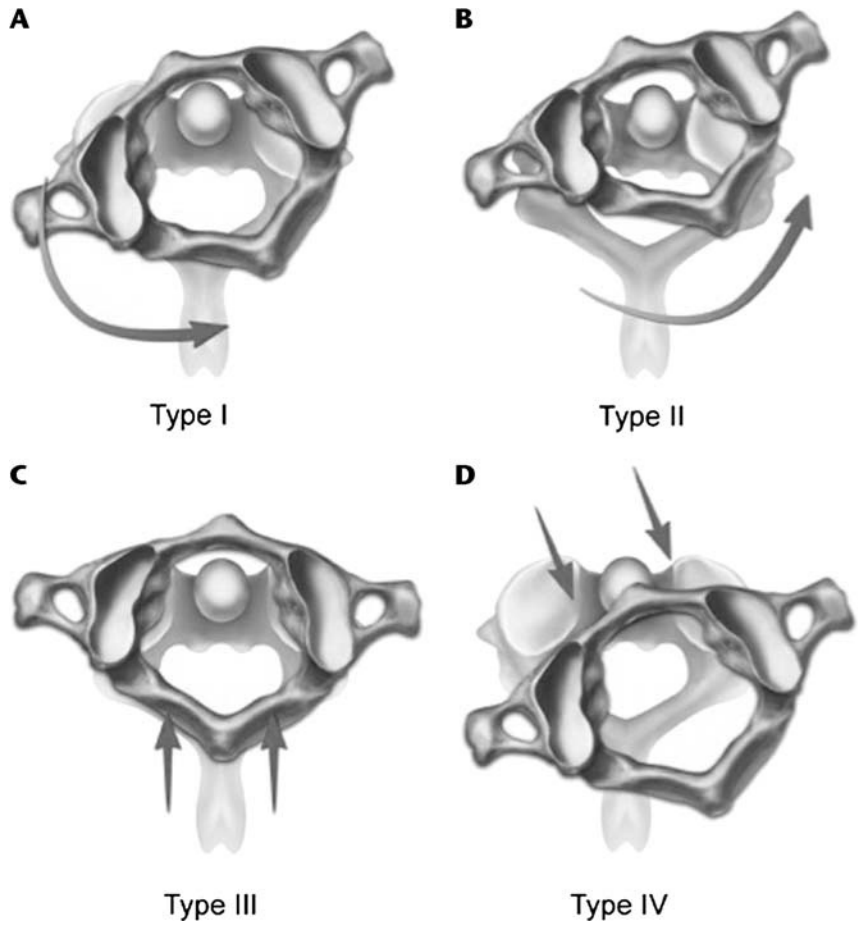

FIGURE 2. A, Fielding-Hawkins Type I injuries: C1 / C2 are locked without anterior displacement. B, Fielding-Hawkins Type II: C1 is rotated over the facet of C2 with 3-5 mm of anterior translation. C, Fielding-Hawkins Type III injuries: C1 crossing over C2 with more than 5 mm of anterior translation. D, Fielding-Hawkins Type IV: Rotatory fixation of C1 over C2 with posterior displacement.

specific to the pediatric population for diagnosis with AARS, and most providers use a modified version of the National Emergency X-Radiography Utilization Study (NEXUS) criteria. If possible, pediatric providers should be judicious in what imaging studies they order and minimize the amount of unnecessary radiation exposure to their young patients. Most authors recommend some sort of dynamic imaging study to assess the fixed rotation of the atlantoaxial region, because plain static radiographs in patients with AARS can be difficult to interpret. Open-mouth odontoid view can show asymmetry of the lateral masses in relation to the odontoid, but these are difficult to obtain in children. Lateral radiographs are difficult to interpret because of head tilt caused by torticollis, but depending on the degree of rotation, 1 lateral mass may be seen anterior to the odontoid and the left and right sides of the posterior arch of C1 may fail to superimpose. The clinician must also remember that these radiographic characteristics can be seen in children without AARS who voluntarily rotate their head.

Thus, dynamic tests are key in establishing the fixed deformity because that is what distinguishes AARS from muscular torticollis. Several authors recommend using cineradiography, including Fielding, which can show that the deformity of C1 on C2 is fixed throughout a range of motion as well as to detect any associated anterior subluxation on flexion views, but again, these are difficult to perform and interpret and impart a large radiation dose to the patient.[3,7]

Because of the difficulty in obtaining and interpreting plain radiographs including static and dynamic views, computed tomography (CT) imaging has become the essential in diagnosing AARS due to its ease and availability. Fine-cut, high-resolution CT imaging through the craniovertebral junction can show in axial view the rotated position of C1 on C2 and any associated anterior or posterior displacement. The AARS can be further studied on sagittal and coronal reformats, and 3-dimensional reconstructions can be made to assist patient evaluation. Furthermore, CT can reveal subtle fractures undetectable on plain films in trauma patients.[3] Dynamic CT imaging of the craniovertebral junction was first described by Rinaldi et al[4,35] in 1979, and consisted of 3 separate fine cut CT imaging studies to evaluate the fixed rotational deformity of C1 on C2. The first series is of the patient in the neutral position, whereas the next 2 series are of the patient turning their head as much as they can in either direction. In normal atlantoaxial rotation and muscular torticollis, the angle between C1 and C2 reduces to 0 or may even cross over C2 when the head is rotated to the opposite side, whereas in AARS, the angle between C1 and C2 fails to reduce and will never cross over C2.[4] In pediatric patients, where AARS is suspected, a focused CT scan of the occiput through C3 can be obtained, thus increasing the sensitivity of diagnosis and reducing the radiation exposure.[12,36,37]

Magnetic resonance imaging (MRI) can be helpful in diagnosing AARS and may be used as a complementary adjunct to CT. Magnetic resonance imaging resolution of bony structures is poor compared with CT, but MRI has superior soft tissue resolution. Thus, in cases where there is diagnostic doubt based on CT, MRI can be used to evaluate the integrity of the atlantoaxial ligaments. If there is neurologic deficit or suspected central nervous system origin of the torticollis, MRI can help evaluate spinal cord compression and/or pathology in the posterior fossa and upper cervical region. Furthermore, for patients with Grisel syndrome, MRI may be helpful in assessing extent of underlying craniovertebral infection.[3] However, in children with special needs (such a Down syndrome), acquiring an MRI may prove to be a challenge. These children may require sedation and even anesthesia. If AARS is suspected, there should be every attempt to acquire diagnostic imaging as soon as possible. Moreover, delays in diagnosis of RAS could decrease the success of reduction in traction. Patients with longstanding RAS are more likely to require surgical correction with fusion.[38]

MANAGEMENT OF AARS

The majority of children presenting to the pediatric emergency department with torticollis and neck pain will not have AARS due to its overall low incidence. Atraumatic cases of pediatric torticollis can be appropriately managed conservatively with soft collar and anti-inflammatories. Although children may not be very compliant with the soft collar in instance of muscular torticollis. In this circumstance, the soft collar does not confer actual stability to the spinal column, and the muscular torticollis maybe managed with anti-inflammatories alone. If AARS is suspected based on history and physical, it should be diagnosed quickly with the aid of dynamic CT imaging and promptly treated as early diagnosis and treatment portends a better outcome with nonsurgical closed reduction (Fig. 3).[2–4,11,39]

Atlantoaxial rotatory subluxation is initially managed medically; however, surgery is indicated with an unstable deformity, progressive neurologic symptoms, or significant structural deformity.[3,4] Subach et al[4] found that patients with a fixed deformity and pain for greater than 3 weeks had a significantly high rate of persistent or recurrent deformity after attempted reduction. Children who present with symptoms within the first 3 weeks can be managed with a trial of closed manual reduction followed

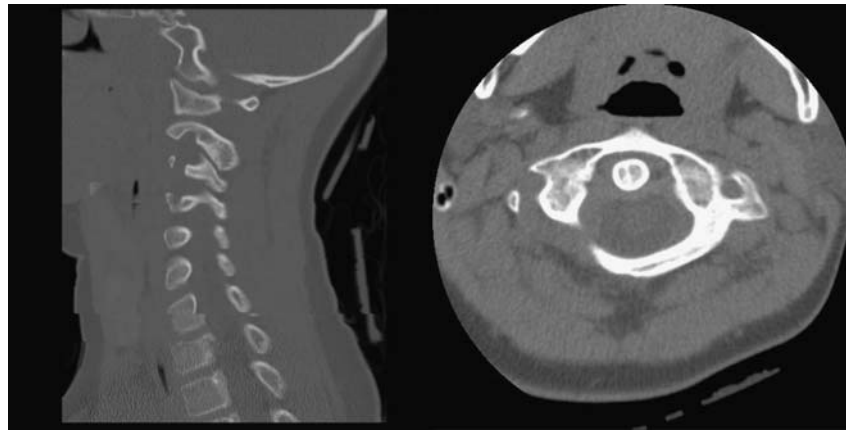

FIGURE 3. Sagittal and axial CT scans with return of normal alignment and rotation after treatment with conservative therapy.

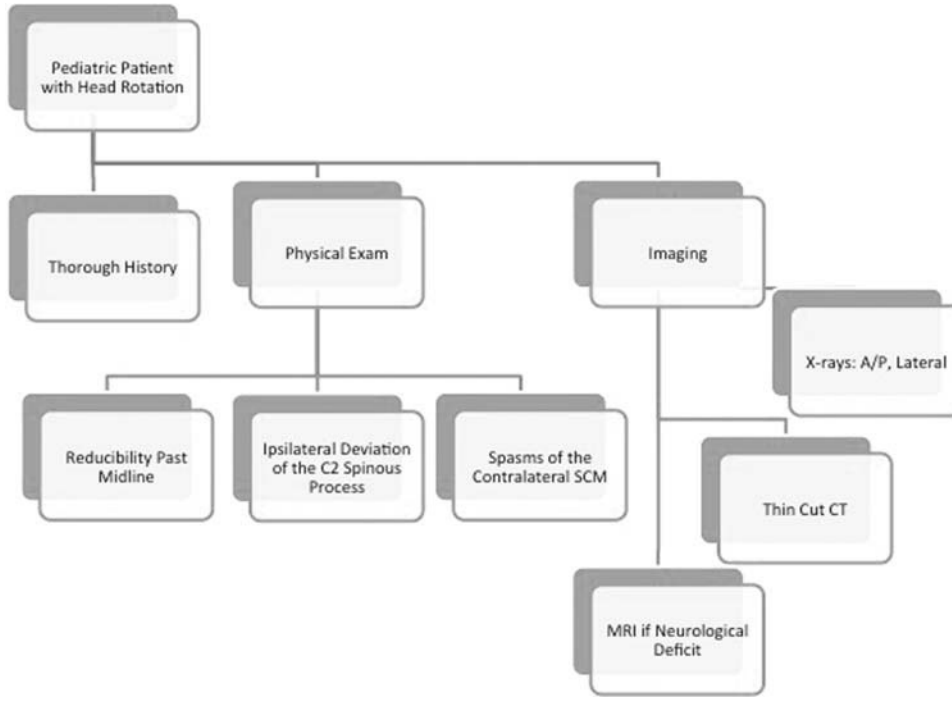

FIGURE 4. The general work-up when a pediatric patient presents with head rotation to the emergency department.

by cervical immobilization, anti-inflammatory agents, and close outpatient follow-up (Fig. 4). If recent upper respiratory tract infection is suspected as a precipitating cause of the torticollis, the URI should be evaluated and medically managed.

Patients that present later than 3 weeks after symptoms begin or that failed 2 weeks of conservative management, more aggressive treatment includes several days of cervical traction in combination with muscle relaxants and analgesics followed by 6 weeks of cervical mobilization in a hard cervical orthosis or brace.[4] Some authors recommend immobilization with halo vest, whereas others recommend a course of cervical traction with muscle relaxant and analgesia as a first line therapy, particularly with a mild degree of AARS (Fig. 5).[1,2] However, halo vests are associated with significant complication rates in children due to their thinner scalps and skulls; thus, it is our opinion that such measures should be reserved for very severe cases or if recurrence is likely (Fig. 6).[40,41] In a study by Fielding and Hawkins,[7,33,42] the recurrence of AARS was higher in patients with greater than 3 months of symptoms. Although these patients may initially reduce with a halo, they are more likely to have a recurrence of symptoms. In cases of recurrence or failed reduction after 6 weeks, cervical traction is again tried, and patients are immobilized for a longer period, usually 3 months.[4] If patients fail to reduce or recur a second time, operative fusion is suggested because there are low success rates of nonoperative treatment

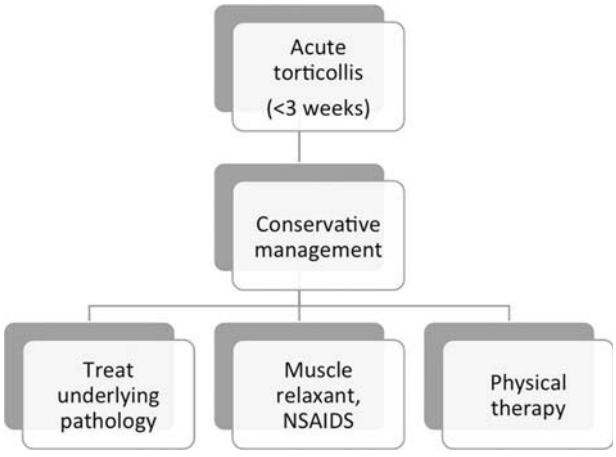

FIGURE 5. The management strategy for acute (less than 3 weeks) of head rotation.

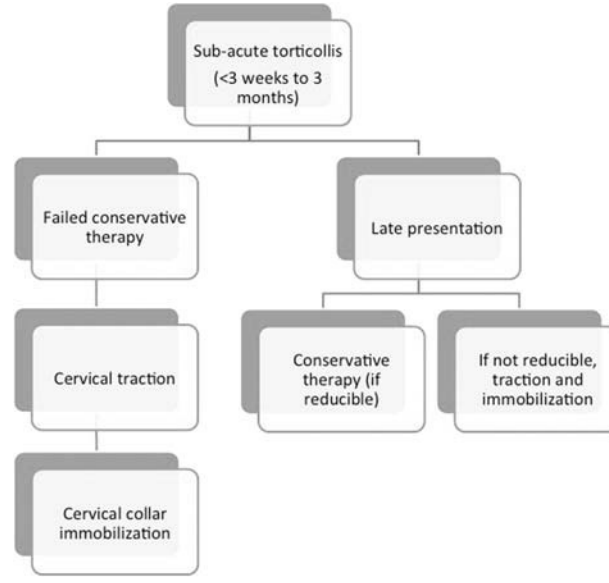

FIGURE 6. The management strategy for subacute (3 weeks to less than 3 months) of head rotation after a trial of conservative therapy.

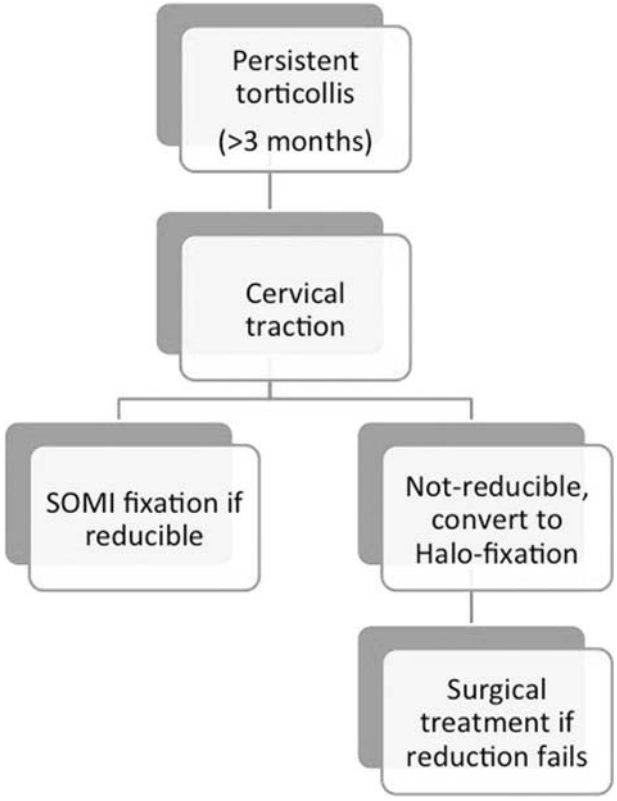

FIGURE 7. The management strategy for chronic (greater than 3 months) of head rotation after failed conservative therapy.

in such patients.[2,4] Furthermore, patients with coexisting conditions, such as rheumatoid arthritis, Down syndrome, were more likely to require fusion surgery because of their predisposition of atlantoaxial instability in concert with AARS (Fig. 7).[2,4,42]

CONCLUSIONS

The pediatric emergency physician should have a high suspicion for AARS for children who present to the pediatric emergency department with a "wry" neck with their head in the "cock-robin" position. Treatment is usually initially nonsurgical, and early diagnosis and treatment is associated with a greater chance of resolution with medical management in patients with less than 3 weeks of acute symptoms. There is no clear consensus on management between 3 and 12 weeks, surgical intervention maybe influenced by recurrent symptoms after reduction. Patients with symptoms greater than 3 months often need surgical correction.[42] Certain clues in the patient's history including and recent trauma or recent upper respiratory illness or head and neck surgery put AARS higher on the differential diagnosis. Patients with suspected AARS require aggressive investigation that includes evaluation by dynamic CT to assess the atlantoaxial joint and see if treatment needs to be initiated.

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