Atlantoaxial Rotatory Fixation in Children. A Clinical Case Study and Review of the Literature.

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Abstract

Background Data: Atlantoaxial rotatory fixation is a relatively uncommon condition. It remains a poorly understood entity despite many reports in the literature. There is no consensus about the best imaging modality for the diagnosis and the best therapeutic modality the patients benefit from.

Purpose: This study was conducted to determine the efficacy of imaging modality and treatment measures and to determine risk factors for recurrence in consideration of the cause, management, and outcome.

Study Design: This is a retrospective clinical case study and a literature review.

Patients and Methods: Our hospital records were reviewed between January 1995 and December 2011. We were able to track 12 children with atlantoaxial rotatory fixation treated by posterior fixation. The mean age was 8.6+2.4 years. Seven were boys and 5 were girls. Sports injury was reported in 4 cases, falls in 3 cases, unknown injury in 3 cases, and road traffic accident in 2 cases. All children had neck pain with torticollis and decreased cervical motion. The mean duration of symptoms before diagnosis was 21.3 days. All children were neurologically intact. All underwent plain cervical radiographs and computed tomography to document atlantoaxial rotation. All children were submitted to closed reduction with cervical traction followed by posterior surgical fixation and fusion after failure of conservative treatment.

Results: All procedures went uneventfully with no added neurologic deficits and a stable repair over at least 12 months follow-up period. Easiness of reduction was affected by the duration of symptoms prior to traction. All had radiographic documentation of stability and fusion. One girl had recurrent rotation two years after modified Gallie fixation after minor head trauma. She was repaired using Harms' technique.

Conclusion: The optimal therapy of atlantoaxial rotatory fixation entails early diagnosis with computed tomography. Reduction with cervical skull tongue traction followed by posterior fixation and fusion accomplished reduction and was curative to all patients. There was no correlation between recurrence and age, sex of the child or the cause of rotation. (2012ESJ011)

Keywords: Rotatory, Atlantoaxial, Subluxation, Fixation, Trauma, Children.

Introduction

Atlantoaxial rotatory fixation is a relatively uncommon condition. It remains a poorly understood entity despite many reports in the literature. The term AARF was coined first by Fielding and Hawkins in 1977[10]. In adults the condition is almost due to trauma[3,20], whereas in children it may results from mild injuries, infections or congenital laxity of the transverse ligaments, head and neck operation, Down syndrome and Morquio's syndrome[6,17,32].

The majority of patients presents with neck pain and torticollis without neurological deficit. Also the majority of patients respond well to non-surgical treatment. However the condition may recur or persist and this mandate surgical treatment. Generally there is no consensus about the best imaging modality for the diagnosis as well as the best therapeutic options the patients benefit from[17,18,27,28,30]. Different methods can be used in C1-C2 fusion augmented with sublaminar wiring[2,9,12] laminar clamps[15,33], transarticular screw[23] C1 lateral mass and C2 pedicle screw either screw plate[13] or polyaxial screw[16] or C2 laminar screw[35]. Although the safety and efficacy of different techniques are encouraging, nevertheless an acceptable rate of morbidity was reported depending on the technique itself[32].

This retrospective study was conducted to determine the efficacy of imaging modality and treatment measures and to determine risk factors for recurrence. Also the causes, management measures, and outcome were analyzed.

Patients and Methods

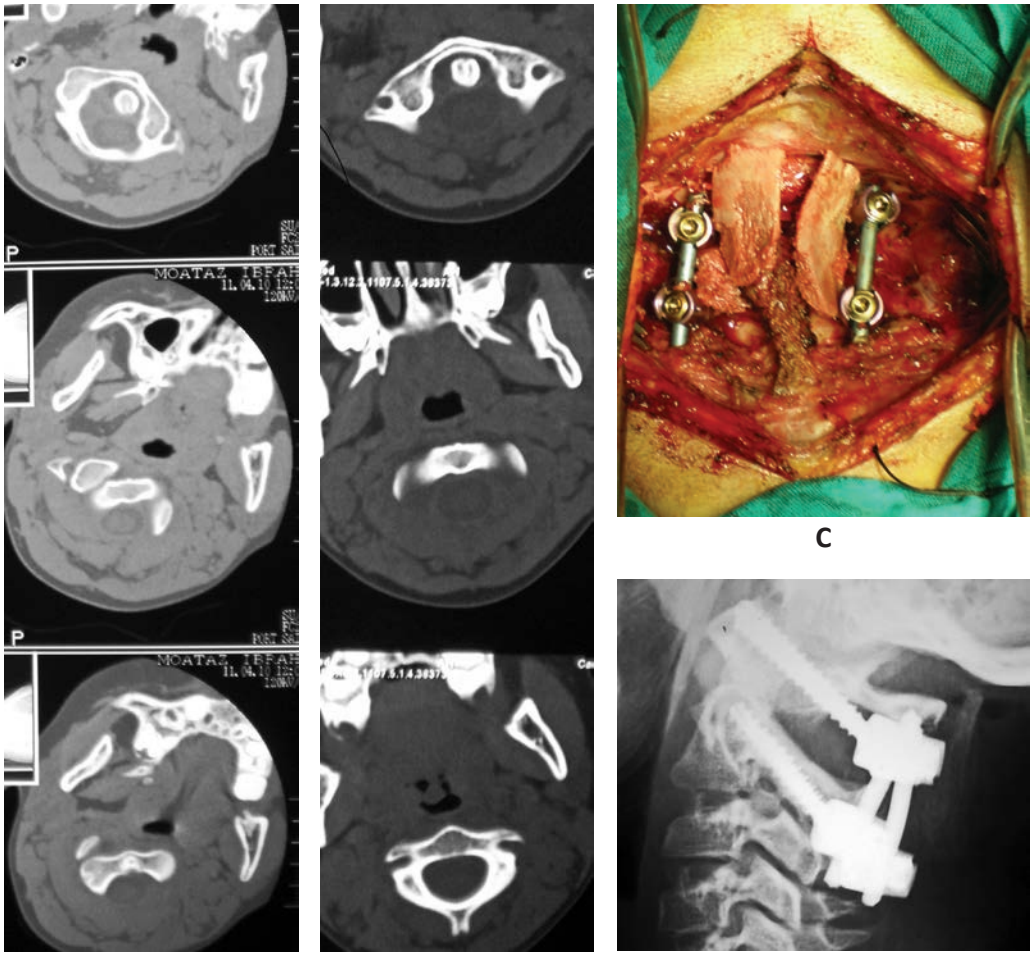
Our hospital records were reviewed between January 1995 and December 2011. A total of 53 patients were reported and managed for traumatic atlantoaxial rotatory subluxation. After exclusion of adults and conservatively managed patients we were able to track 12 children with atlantoaxial rotatory fixation treated by posterior fixation and fusion. The relevant data are summarized in table 1. The mean age was 8.6+2.4 years with a range of 4 to 12 years. Seven were boys and 5 were girls. Sports injury was reported in 4 cases, minor trauma/falls in 3, road traffic accident in 2, and unknown injury in 3 cases. All children presented with neck pain, torticollis and decreased range of cervical motion. Rotation was anti clock wise (left side) in 9 cases and clock wise (right side) in 3 cases. The mean duration of symptoms before diagnosis was 21.3 days and ranged from 7 to 42 days. All children were neurologically intact. None of them had associated systemic injuries. Three children reported upper respiratory tract infection in the last 2 weeks prior to presentation.

All underwent cervical plain radiographs anterior-posterior and lateral views, and cervical Computed Tomography to document atlantoaxial rotation. Only 5 children had 3D-Computed Tomography. Nine children had Magnetic Resonance Imaging study. Plain radiographs could not give the final diagnosis in all cases as well as MRI. The final diagnosis was retrieved from CT-study in all cases. Eight children were classified according to Fielding and Hawkinss' classification as type-II, two as type-I, two as type-III, and none were type-IV. We were not able to report any deformity of the axis lateral mass in our 5 cases with 3D-CT. None had associated cervical spine boney injuries.

According to our medical records, all children were initially submitted to successful closed reduction with cervical traction followed by the application of hard neck collar. Skull tong traction of 5-9 kg was used in a period varied from 2 to 5 days with CT follow-up in order to document full reduction of the rotation. They were discharged and followed regularly at the outpatient clinic. Then they reported recurrence of torticollis again during follow-up, where recurrent AARF were proved radiologically again. So it was decided to offer them posterior surgical fixation and fusion. It was observed that there was no correlation between recurrence of rotation and the cause of rotation, age, or sex of the child. Also easiness of reduction was affected by the duration of symptoms prior to traction. Early presented cases were easily reduced than later presented cases. A total of 3 cases were offered modified Gallie fusion[9], 3 were offered atlantoaxial transarticular screw fixation[23], and 6 were offered Harms' atlas lateral mass and axis pedicle polyaxial screw posterior fixation[16] (Figure 1).

All were fused using autologous chipped iliac crest bone grafting. All surgical procedures went uneventful. Mean hospital stay was 5 days, with a range of 4-12 days including preoperative traction time. Children were discharged in hard collar for 8 weeks and were followed-up at the outpatient

clinic. Children were assessed at follow-up both clinically and plain radiographicaly at 1, 3 months then at 6 months intervals. Follow-up ranged from 12 to 48 months with average of 21.7510.7± months (Table 1).



A B C D

Figure 1. Case number 7, 8 years boy with sports injury. A; Axial CT showing type-I clockwise recurrent AARF. B; Axial CT showing reduction of the AARF. C; Operative image showing screws and rod in situ with iliac crest grafting. D; Postoperative plain radiograph showing adequate screw purchase.

Table 1. Data of 12 children reported in study.

No.	Age in years /Sex	Cause	Type	Operation	Follow-up/months
1	9/M	Sport	2	Trans-articular	36
2	7/M	unknown	3	Trans-articular	24
3	9/F	unknown	2	Trans-articular	12
4	8/F	Minor fall	2	Modified Gallie > Harms'	48
5	6/M	RTA	2	Modified Gallie	18
6	7/F	Minor fall	2	Modified Gallie	24
7	8/M	Sport	1	Harms'	15
8	10/M	Minor fall	2	Harms'	24
9	4/M	unknown	1	Harms'	18
10	12/F	Sport	2	Harms'	15
11	11/F	Sport	3	Harms'	15
12	12/M	RTA	2	Harms'	12

Results

At last clinical follow-up all children were pain free with normal position of head and neck without any apparent deformity. All children had insignificant limitation in the range of their neck of motion. All were neurologically intact and has no limitation of their ordinary life activities. Plain radiographic study of our cases showed that metal constructs were stable in all patients but one (N=11) with documented sound boney fusion in 83% of cases (N=10). Post-operative CT-scan showed adequate purchase of all screws through both C1 lateral mass and pedicle of C2 vertebrae. The bi-cortical screw purchase of C1 lateral mass in cases managed with Harm's fixation has been documented in postoperative CT-scan in all cases (N=6). Post-operative CT-scan had shown reduction of the rotation as well as the anterior atlantodental interval (ADI) space in all patients. There was no reported morbidity either related to the traction, operative procedure or the donor site. There was no correlation between failure of conservative treatment and age, sex of the child or the cause of rotation.

An 8 years old girl (child number 4, Table 1) had recurrent painful torticollis after minor trauma two years after modified Gallie's fixation. She had recurrence of clock wise rotation this time instead of anti-clock wise rotation at the first injury. Here CT-scan documented C1-2 rotation, graft resorption and wire break. After skeletal traction and reduction of her rotation she was repaired using Harms' technique. She had a smooth operative and a postoperative follow-up for 2 years.

Discussion

Rotatory subluxation of the atlantoaxial joint was described for the first time by Corner[4] in 1907. Persistent subluxation causing torticollis was termed as atlantoaxial rotatory fixation (AARF) by Wortzman and Dewar[34] in 1968, followed by Fielding and Hawkins[10] in 1977. AARF is relatively uncommon condition that requires a high suspicion index for diagnosis. Simple X-ray or CT is used for the diagnosis of atlantoaxial rotatory subluxation. Correct diagnosis with X-ray is usually difficult because the patient has severely bended head and stiff neck in many cases. Mandibular and occipital bone can be overlapped and makes it difficult for diagnosis. Although CT scan minimizes these problems, 3D CT provides accurate diagnostic image of atlantoaxial relation in these cases[20,24]. This condition responds well for conservative treatment in the majority of cases. Conservative treatment is often unsuccessful in patients with a chronic irreducible or recurrent unstable AARF[7,8,17,22,32], that requiring surgical treatments[7,10,17,27]. In this conditions, the majority of surgeons favor surgical treatments[8,10,17,22,27].

In this report we operated on 12 out of total 53 patients with AARF after failure of conservative treatment during the last 15 years. In this study, the mean age was 8.6+2.4 years, and 7 were boys and 5 were girls. Most reported cases were due to minor traumatic events including sports injury in 4 cases, falls in 3 cases, unknown injury in 3 cases, and road traffic accident in 2 cases. Our epidemiological data matches most reports in the literatures[10,11,30,32]. We reported no associated injuries in our study. This can be explained by the minor trauma as a causative factor. In adult, trauma is the major reported cause of AARF[20]. In their study, Kim et al., reported a high incidence of associated head and cervical injuries and all their patients but one were males. All patients presented with neck pain, torticollis, and were neurologically intact. These also were reported by different authors[20,30,32].

In this study all patients had examined by X-Ray, CT scan, and MRI. However, CT was the key stone for the diagnosis of AARF. Although CT scan facilitated the diagnosis of this condition, still normal atlantoaxial rotation makes a challenge. Functional/ dynamic CT has been recommended by many authors for diagnosis of a fixed AARF before the decision of surgery is taken and to exclude normal atlantoaxial rotation[10,19,21,27,32]. Normal atlantoaxial rotation has to be excluded in order to avoid miss diagnosis of AARF. Mönckeberg et al.,in their study concluded that there is a risk of overdiagnosis and overtreatment (C1–C2 arthrodesis) when evaluating upper cervical spine rotational problems. The concept of both rotatory fixation and subluxation should be revised, and quantifying the rotational angle and contact surface loss between C1–C2 can be very useful[26].

The pathogenesis of irreducible or recurrent AARF remains unknown[17]. The results of our study demonstrated that the rotation was easily corrected in cases encountered shortly after the initation of symptoms. This was observed by different

authors[7,27,30,32]. In our study all cases were corrected with tong skull traction with 5-9 Kg during a period of 2-5 days. For irreducible cases, Crockard and Rogers[5] used the extreme lateral approach to break the atlantoaxial lateral mass fusion bilaterally. They managed their cases with the transarticular screw fixation technique. Govender and Kumar[14] used the transoral route to release atlantoaxial fxation, then they posteriorly fixed the reduced segment. These two studies reported more than 3 months symptoms duration prior to management. Different causes for irreducibility have been postulated including; ligamentocapsular contractures[10], the adherent and inflamed synovial fringes, fibrous interposition in the joint[5,14,29] or C1–2 osseous union.

Case number 4 in our study (Table 1) had recurrence of rotation and failure of 2 years old atlantoaxial modified Gallie. This has not been reported in the literature before but by in our previous study[1]. Short term recurrence with failure of conservative therapy is common. Recurrences were thought to be caused by overstretching of stabilizing ligaments, overdistension and subsequent laxity of the joint capsule[25], or excessive rotatory movement following damage to the articular surfaces of the C1–2 joint[7,30].

Various operative procedures have been advocated for atlantoaxial fixation[32]. Currently we are using the Harms' procedure for treating recurrent AARF. We reported no neurological or vascular injury. Stability and fusion were reported in all 6 cases of Harms' procedure. Our results as well as others[31] suggest the safety and efficacy of this procedure in recurrent AARF.

Conclusion

The optimal therapy of atlantoaxial rotatory fixation entails early diagnosis and computed tomography study. Easiness of reduction was affected by the duration of symptoms prior to traction. There was no correlation between recurrence and the cause of rotation, age, or sex of the child. Reduction with cervical skull tongue traction followed by posterior fixation and fusion accomplished reduction and was curative all patients of recurrent AARF.

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الملخص العربي

تثبيت تدويرية الفهقي لدى الأطفال. دراسة حالة سريرية ومراجعة الأبحاث

بيانات أساسية: إن تثبيت تدويرية الفهقي هو حاله شائعه نسبيا ولكن تبقى غير مفهومه على الرغم من العديد من التقارير في الأبحاث. ليس هناك توافق في الآراء حول أفضل طريقة أشعة تشخيصية وأفضل طريقة علاجية للمرضى.

الغرض: أجريت هذه الدراسة لتحديد مدى فعالية طريقة التشخيص وطرق العلاج وتحديد عوامل الخطر. كما تم تحليل الأسباب والعلاج.

تصميم الدراسة: دراسه بأثر رجعي لحالات سريرية ومراجعة الأبحاث.

الطرق: تم استعراض سجلات المستشفيات لدينا ما بين يناير ١٩٩٥ وديسمبر ٢٠١١. كنا قادرين على تتبع ١٢ طفلا مع تثبيت تدويرية الفهقي تم علاجهم بالتثبيت الخلفي. كان متوسط العمر ٨,٦ سنوات. سبعة أولاد و٥ من الفتيات. وأفادت التقارير أن الإصابة الرياضية تواجدت في ٤ حالات، السقوط في ٣ حالات، غير محدد في إصابة ٣ حالات، وحادث مرور في ٢ من الحالات. كان جميع الأطفال يعانون من آلام الرقبة وتقلصات وصعوبة في حركة العنق. وكان متوسط مدة الأعراض قبل التشخيص ٢١,٣ يوما. وكانت جميع الأطفال سليمة عصبيا. وخضعت جميع صور الأشعة العنقية العادية والأشعة المقطعية للمراجعه. وتم تثبيت العنق مع عمل جذب باستعمال أوزان يليها تثبيت جراحي خلفي بعد فشل العلاج المحافظ. وتم متابعة الأطفال على مدى ١٢ شهرا على الأقل في فترة المتابعة.

النتائج: كل الأطفال كانت نتائجهم مقبوله من حيث عدم وجود الم وبرغم وجود بعض الحد من حركة العنق ملحوظ ولكنها بدون الم. كان هناك فتاة عاودها تقلص الرقبة بعد فشل التثبيت بعد سنتين ناتج عن صدمه بعد تثبيت جالى وتم إصلاح الأضرار باستخدام تقنية هارمس.

الخلاصة: إن العلاج الأمثل لتثبيت تدويرية الفهقي يستلزم التشخيص المبكر مع التصوير المقطعي. والحد من حركة العنق مع الجمجمة تليها التثبيت الخلفي لكل المرضى. لم يكن هناك علاقة بين ارتجاع المرض والعمر والجنس للطفل.