

Recurrent Atlantoaxial Rotatory Fixation after failed posterior fusion: A Case Report & Review of the Literature.

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Background Data: AARF is a well-known disease entity as well as its recurrent form. However, recurrent AARF after posterior fusion was not reported before. This is the first report of a case of revisited recurrent AARF 2 years after posterior fusion.

Purpose: To describe the clinical and radiographic features and management of revisited recurrent AARF.

Study Design: Case report.

Methods: A type II AARF as described by Fielding recurred shortly after external orthosis in an 8 years old girl. She was subjected to traction until reduction and modified Sonntag fusion. The condition recurred after 2 years. She was put into traction for reduction. She was subjected for Harms' posterior fusion.

Results: Traction followed by C1 lateral mass and C2 pedicle screw fixation and fusion was successful in the management of a recurrent AARF.

Conclusion: Recurrent AARF can occur even after failed posterior fusion. Harms' posterior fusion is a sound procedure for the management of recurrent AARF. (2012ESJ015)

Key Words: Atlantoaxial rotatory subluxation, torticollis, Atlantoaxial rotatory fixation, recurrent AARF.

Introduction

Atlantoaxial rotatory fixation (AARF) refers to a pathological process involving C1-C2 joints causing irreducible disarticulation of the facets and transitional subluxation. The term AARF was coined first by Fielding and Hawkins in 1977⁸. In adults the condition is almost due to trauma, whereas in children it may result from infections or congenital laxity of the transverse ligaments^{3,16}.

Patients with AARF present with torticollis (cock-robin deformity of the neck) where the head is tilted to one side and rotated to the contra-lateral side with slight neck flexion. The condition is reduced by cervical traction and muscle relaxant in majority of cases. The child is then placed in rigid cervical collar or halo-vest as a definitive management in majority of cases^{16,17,25,29}.

Recurrent dislocation of C1-C2 denotes instability and termed chronic dislocation or chronic fixation and requires reduction and fusion. Failure of closed reduction or recurrent rotation mandate surgical treatment^{27,29}. Different methods can be used in C1-C2 fusion augmented with sublaminar wiring^{1,6,10} laminar clamps^{13,32}, transarticular screw²¹, C1 lateral mass and C2 pedicle screw either screw plate¹¹ or polyaxial screw¹⁴ or C2 laminar screw³³. Although the safety and efficacy of different techniques are encouraging, nevertheless an acceptable rate of morbidity was reported depending on the technique itself. As the standard management of AARF is conservative, failure of surgery was hardly reported. To the best of our knowledge we report the first case of recurrent AARF after failed posterior fusion.

Case report:

An eight-year old girl sustained violent neck twist

by her 2-years older cousin. This resulted in sudden neck pain and torticollis. We saw the girl at day 12 after trauma. There was no history of upper respiratory tract infection or family history of collagen deficiency or metabolic illness. On examination the patient was neurologically intact. Plain X-ray and CT-scan were performed and atlantoaxial rotatory fixation type-II fielding was diagnosed⁸(figure 1).

She was submitted for skull tong traction starting with 4 kg and adding one and half kg per day. The subluxation was reduced with 7kg traction and a post traction CT-scan was obtained (figure 2). We put the child in a Philadelphia neck collar and she was sent home for followed-up. She came back in a month with recurrent torticollis. She was submitted for traction again until reduction was documented.

The child was taken to the OR and a modified Sonntag posterior atlantoaxial fusion was performed⁶. The operative and post-operative course went uneventful. Post-operative X-ray was good and the graft was in position with good C1-C2 alignment (figure 3).

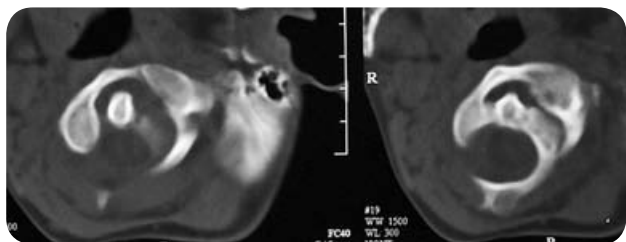


Figure 1. CT scan of a 7 years old female patient with Atlanto axial rotatory subluxation.



Figure 3. Post-operative X Ray lateral view following posterior C1-C2 fusion by modified Sonntag fusion.

The girl was discharged on day 4 after surgery in a hard collar. She was followed-up for a month then the patient's follow-up was discontinued as we lost any connection with her.

After two years she came back with same neck pain and torticollis. She reported her illness after minor neck trauma when she fell down her bed while playing with her sister one week before presentation. This time she was also neurologically intact with limited neck movement. New plain X-ray and CT-scan showed recurrent AARF with complete absorption of iliac graft and broken wire (figure 4).

She was submitted for gradual skull tong skeletal traction again. Here subluxation was reduced after 9 kg traction as shown on post-traction CT-scan. She was taken again to the OR and a C1 lateral mass C2 pedicle polyaxial screw posterior fixation was performed¹⁴. Operative and post-operative course went uneventful and post-operative X-ray and CT scan was OK (figure 5, 6 respectively). She was discharged in a hard neck collar. So far we are up to 18 months follow-up now and everything is fine.

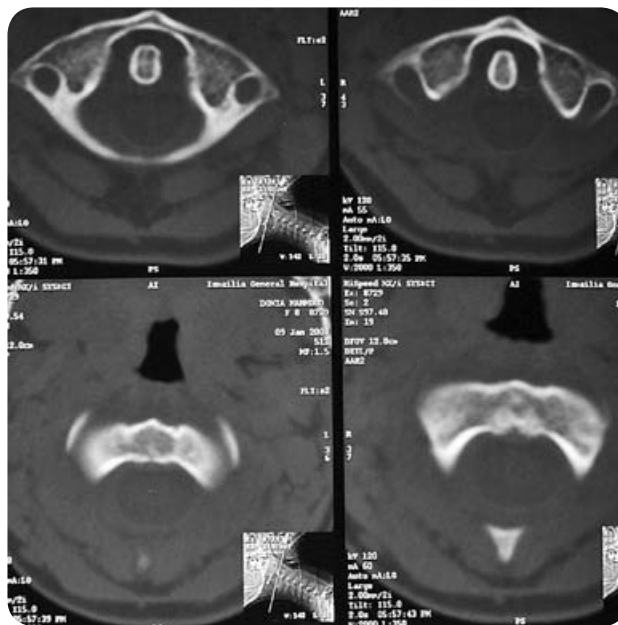


Figure 2. CT scan of same patient after reduction using skull tong traction with 7 Kg weight.

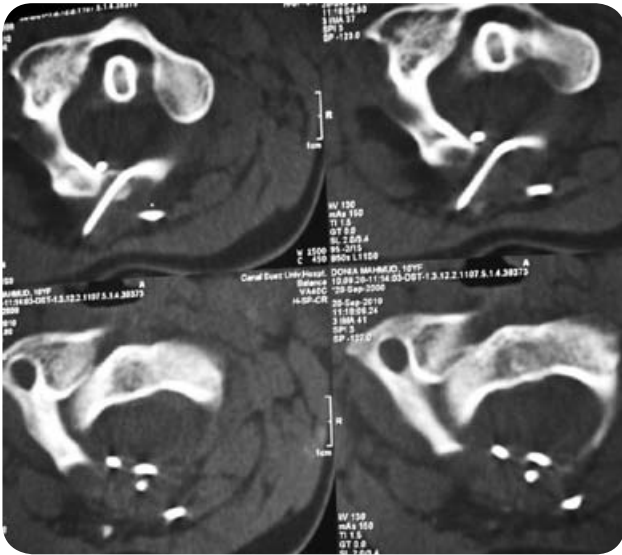


Figure 4. CT scan of the same patient following second trauma showing recurrent subluxation of C1-C2 joint.



Figure 5. X Ray cervical spine lateral view showing C1-C2 fixation using C1 lateral mass screw and C2 transpedicle screw



Figure 6. CT scan of C1-C2 vertebra showing C1 lateral mass screw and C2 transpedicular screw

Discussion

Atlantoaxial rotatory subluxation or fixation is more common in children in comparison to other ages. The mechanism that causes rotation is not fully understood. It may be due to muscle spasm^{9,31}, increased laxity of ligaments or joint capsules caused by pharynx infection, surgical treatment of the pharynx, or trauma^{7,9,31}. Also some conditions that enhance ligamentous laxity such as: Down syndrome, Morquio Syndrome and Marfan syndrome, correlate with a higher incidence of rotatory subluxation²².

A high suspicious index is important in diagnosing AARF among all painful torticollis. With plain radiography alone the diagnosis is not always possible because it is difficult to position the patient properly for imaging, and meanwhile, it is difficult to interpret the radiographs^{8,18,29}. Computed tomography is more advantageous and has been extensively described as the method of choice^{7-9,24}. While many authors^{8,18,26} has recommended the use of dynamic CT, recently others^{16,17} has recommended 3D CT scanning for proper diagnosis.

The rotation can be managed in acute cases successfully conservatively with traction and external orthoses^{8,18,25,31}. However when diagnosis is delayed or in neglected cases, intra- and

extraarticular lesions may eventually prevent closed reduction of the dislocation^{4,31} or cause recurrences even after successful closed reduction^{5,20,31}.

Surgery may be indicated for patients with recurrent AARF^{4,8,19,28} or when the rotation is irreducible. Ligamentocapsular contractures⁸, adherent and inflamed synovial fringes, fibrous interposition in the joint^{2,28}, or C1–2 osseous union are the most known causes for irreducibility. Short period of external orthosis with inadequate healing, persistent of primary pathology of rotation such as infection or collagen disease, overstretching of the stabilizing ligaments, distension and subsequent laxity of the joint capsule²³ or damage to the articular surfaces of the C1–2 joint^{5,29} are the most acceptable explanation for recurrent AARF. However still the pathogenesis of rotation and its recurrence is not clearly understood^{4,5,9,12,20}.

Recurrent subluxation after reduction and external fixation has been reported by different authors^{16,17}. Based on the assumption that it can be due to defective ligamentous healing by early mobilization of cervical spine or C2 facet deformity observed¹⁶, application of rigid cervical collar or halovest should be continued for enough time to allow full C2 facet remodeling to prevent further subluxation¹⁶.

It has been generally accepted to manage recurrent subluxation following failure of conservative treatment with posterior fusion²⁷. Several C1-C2 posterior fusion techniques have reported for the treatment of AARF with varying success rate^{13,30}. The non-rigid constrained construct included Gallie, Brook, interlaminar clamps, Sonntag^{1,6,10,15}, where the rigid constrained constructs included the transarticular screw fixation, Goel, Harms, translaminar screw^{11,14,21,33}. The second group of techniques was more biomechanically advantageous than the first group^{13,30}.

Our case was offered the Sonntag modified technique⁶ in the first go which failed and the condition recurred again after 2 years. To the best of our knowledge this has not been reported before in the English literatures. Although long period of external orthosis has been recommended to prevent recurrence of AARF^{16,17}, a 2 years of internal fixation did not prevent recurrence when fusion failed in our patients. This is very odd incidence in the natural history of pediatric trauma generally

and AARF specifically. The C1-C2 wiring techniques have generally resulted in suboptimal outcome in comparison to other procedures^{13,30} and our initial management is one of these cases with poor outcome. We can assume that the iliac graft has been resorbed with time and the segment got loose and with repetitive movement the wire ruptured. For some unclear reason the AARF did not heal and recurred again after this long period. A number of speculations can be postulated. It can be due to recurrent pharyngeal infection, damaged C1-C2 facets, or inherited ligamentous and capsular laxity. Poor neck musculature of such age augmented by the collar use may be another speculation. The high physical activity of the children with frequent neck trauma makes the need for a sounder fixation and fusion mechanisms an important issue.

Post-operative neck immobilization is recommended for at least three months following surgery in order to guarantee sound fusion especially in non-constrained C1-C2 fixation. However, this is not mandatory with constrained constructs. Close clinical and radiographic follow-up guided by efficient C2 facet remodeling is important to prevent recurrent dislocations following final treatment.

Conclusion

AARF in children should be treated conservatively with external orthosis following reduction. Recurrent cases should be treated surgically by posterior fusion. Posterior wiring stand-alone fusion techniques are inadequate and require prolonged external immobilization. AARF can recur after failure of posterior fusion. C1 lateral mass and C2 polyaxial pedicle screw fixation and fusion with iliac graft is a successful technique for AARF management.

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الملخص العربي

التثبيت المتكرر للخلع الجزئي الدائري للمفصل الأطلسي- المحوري بالفقرات العنقية بعد فشل محاولة اللحام الخلفي للفقرات (تقرير عن حالة ومراجعته بحثية) المقدمة: يعتبر الخلع الجزئي الدائري للمفصل الأطلسي- المحوري بالفقرات العنقية من الأمراض المعروفة والمدونة بالأبحاث المختلفة ولكن لا توجد تسجيل لحالات قد حدث لها تكرار الخلع بعد اجراء تثبيت ولحام خلفي للفقرتين. وتعتبر هذه الحالة من اولى الحالات التى حدث خلع جزئى متكرر بعد مرور عامين من اجراء جراحه تثبيت من الخلف. الهدف : شرح الأعراض الإكلينيكية المصاحبة للخلع الجزئى المتكرر مع شرح طرق التشخيص عن طريق الأشعات وطريقة العلاج المقترحة. نوع البحث: تسجيل لحاله فتاة تبلغ من العمر ثمانية اعوام وتعانى من خلع جزئى متكرر بالمفصل الأطلسي- المحوري بالفقرات العنقية بعد مرور سنتان من اجراء جراحه لها لتثبيت الفقرات العنقية من الخلف الوسائل: تم وضع المريضة التى حضرت مصابه بخلع جزئى من الدرجة الثانية حسب مقياس فيلدينج فى شدة عنقية لاسترجاع الخلع من عمل جراحة تثبيت للفقرات العنقية الاولى والثانية من الخلف عن طريق سلك معدنى مع عمل لحام عظمى بطريقه سونتاج المعدلة وبعد مرور عامين حدث خلع جزئى متكرر بالمفصل الأطلسي- المحوري مما استدعى اجراء جراحه لتثبيت الفقرات من الخلف باستخدام مسامير متعدده المحاور بين الفقرتين العنقية الاولى والثانية بطريقه هارمس بعد استرجاع الخلع النتائج: استرجاع الخلع عن طريق الشدة العنقية مع عمل تثبيت للفقرات العنقية الاولى والثانية عن طريق مسامير متعددة المحاور بطريقه هارمس هى افضل طريقه لعلاج الخلع الجزئى المتكرر للمفصل الاطلسي- المحوري بالفقرات العنقية الاستنتاجات: يمكن ان يتكرر حدوث خلع جزئى للمفصل الاطلسي- المحوري حتى بعد اجراء جراحه للتثبيت من الخلف عن طريق السلك المعدنى وفى هذه الحالة يكون التثبيت الخلفى بطريقه هارمس هى افضل الأساليب لعمل لحام عظمى جيد بين الفقرات.