Eur Spine J
DOI 10.1007/s00586-014-3648-5

GRAND ROUNDS

The surgical management of atlanto-axial subluxation in juvenile rheumatoid arthritis

Khalid M. I. Salem · Ajay Radhakrishnan · Eyal Behrbalk · B. M. Boszczyk

Received: 24 July 2014 / Revised: 27 October 2014 / Accepted: 29 October 2014

Abstract

Juvenile idiopathic arthritis (JIA) is a chronic condition affecting patients <16 years of age and can be associated with substantial morbidity. Atlanto-axial subluxation (AAS) is a known complication of JIA and can result in pain, reduced neck motion and neurological compromise. In this paper, we present the case of a 10-year old suffering with JIA and significant AAS; we discuss the management options and present the approach and outcome of treatment for this case.

Keywords Atlanto-axial subluxation · Juvenile idiopathic arthritis · Posterior arthrodesis

K. M. I. Salem · A. Radhakrishnan · E. Behrbalk · B. M. Boszczyk (✉)
Centre for Spinal Studies and Surgery, Queen's Medical Centre Campus, Nottingham University Hospital NHS Trust, Nottingham NG7 2UH, UK
e-mail: bronek.boszczyk@nuh.nhs.uk

K. M. I. Salem
e-mail: kmsalem@gmail.com

Case presentation

A 10-year-old male patient presented to the spinal clinic with a 9-month history of a painless torticollis after bending his head whilst taking a bath. He had no associated loss of manual dexterity or gait disturbance. Examination revealed a painless restricted neck range of motion and his neurological examination at presentation was normal in the upper and lower limbs with no abnormal upper motor neuron signs. His past medical history included a mild birth-related cerebral palsy and he suffered with juvenile idiopathic arthritis (JIA) for which he was treated with methyl prednisolone and subcutaneous methotrexate. Following initial X-rays of the cervical spine (including flexion/extension views), the diagnosis of atlanto-axial subluxation (AAS) was confirmed and he was admitted to the hospital for temporary reduction and stabilisation of his cervical spine using halo traction (5 pounds). Further imaging was organised and a definitive posterior stabilisation was performed 3 days post-admission (Fig. 1).

Historical review of the condition, epidemiology, diagnosis, pathology, differential diagnosis

Juvenile idiopathic arthritis is a term recently adopted to describe all forms of arthritis affecting children under the age of 16 and lasting for more than 6 weeks with a prevalence of 16–150 per 100,000. The term replaces what was previously known as Juvenile Rheumatoid Arthritis (JRA) or Juvenile Chronic Arthritis (JCA) [1, 2]. The most recent classification for the condition was proposed by the International League of Associations for Rheumatology (ILAR) dividing the presentation into 7 district subgroups including Systemic arthritis (4–17 %), Oligoarthritis (27–56 %),

Published online: 27 November 2014

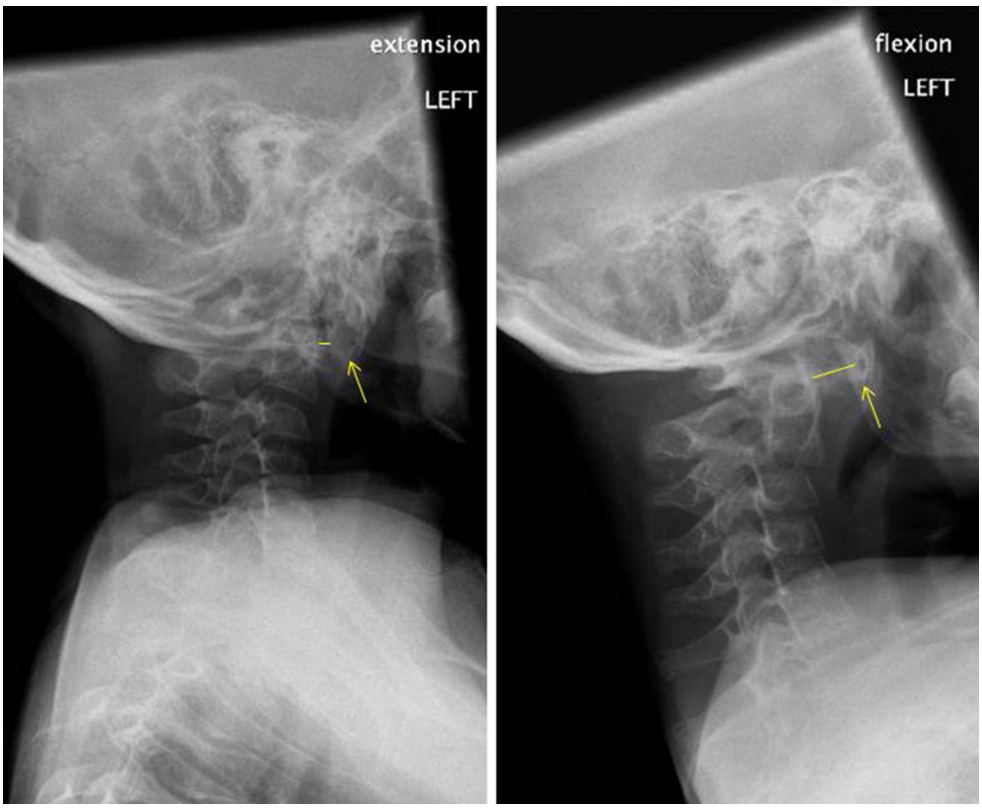

Fig. 1 Flexion/extension lateral cervical spine X-rays confirming significant atlanto-axial subluxation (AADI in extension = 2.3 mm, AADI in flexion = 11.4 mm)

rheumatoid factor–positive polyarthritis (2–7 %), rheumatoid factor–negative polyarthritis (11–28 %), enthesitis-related arthritis (3–11 %), psoriatic arthritis (2–11 %), and undifferentiated arthritis (11–21 %) [3] (Fig. 2).

The involvement of the cervical spine in the disease process has been expressed through a wide range from 25 to 80 % reflecting the variation in the duration of patient follow-up between studies and the sensitivity of the diagnostic modality (radiographs/MRI) used in establishing a diagnosis [4–6]. Pathologically, the cervical spine is affected by the same chronic systematic auto-immune inflammatory process affecting the rest of the body; a humoral auto-immune response is initiated by an exposure to an environmental trigger in the genetically prone individual. Following an exposure to a trigger, macrophages release cytokines resulting in the activation of T- and B-Cells, the formation of auto-antibodies (i.e., rheumatoid factor) and synovial infiltration with activated macrophages, lymphocytes, and fibroblasts (Pannus). The process results in enzyme-mediated and neovascularisation-mediated destruction of adjacent soft tissue, cartilage, and bone [5]. When the cervical spine is involved, the axial cervical spine appears to have a higher predilection to the condition. This is because the articulation at occiput/C1 and C1/C2 is purely synovial, additionally, the articulation orientation in the axial plane means that there is no bony barrier to subluxation [5]. In a paediatric setting, the situation is accentuated by the higher torque and sheer forces across this region making it more susceptible to instability. Other anatomical consideration in the paediatric spine include weak neck musculature, larger head–body mass ratio, ligamentous laxity, and a higher neck movement fulcrum (at C2/3) put this group of patients at a higher risk of instability [7] (Fig. 3).

Clinically, patients suffering with JIA and cervical involvement can present with neck pain and stiffness (classically cranio-cervical) and rarely torticollis. C2 neural compression can also result in occipital headaches, ear pain, or facial pain depending on the contribution of the C2 root to the greater auricular nerve, greater occipital nerve, and the nucleus of the spinal trigeminal tract. Cord compression can result in cervical myelopathy. The clinical findings can be used to categorise patients according to the Ranawat classification (Table 1) which is a useful guide for management [8] albeit such symptoms are less likely in paediatrics compared to adults [9] (Fig. 4).

Cervical radiological findings in JIA include apophyseal joint ankylosis (most common at C2/3), AAS, atlanto-axial impaction (AAI), vertebral body growth disturbances (occurs in 26 % of the cases), and/or subaxial subluxation

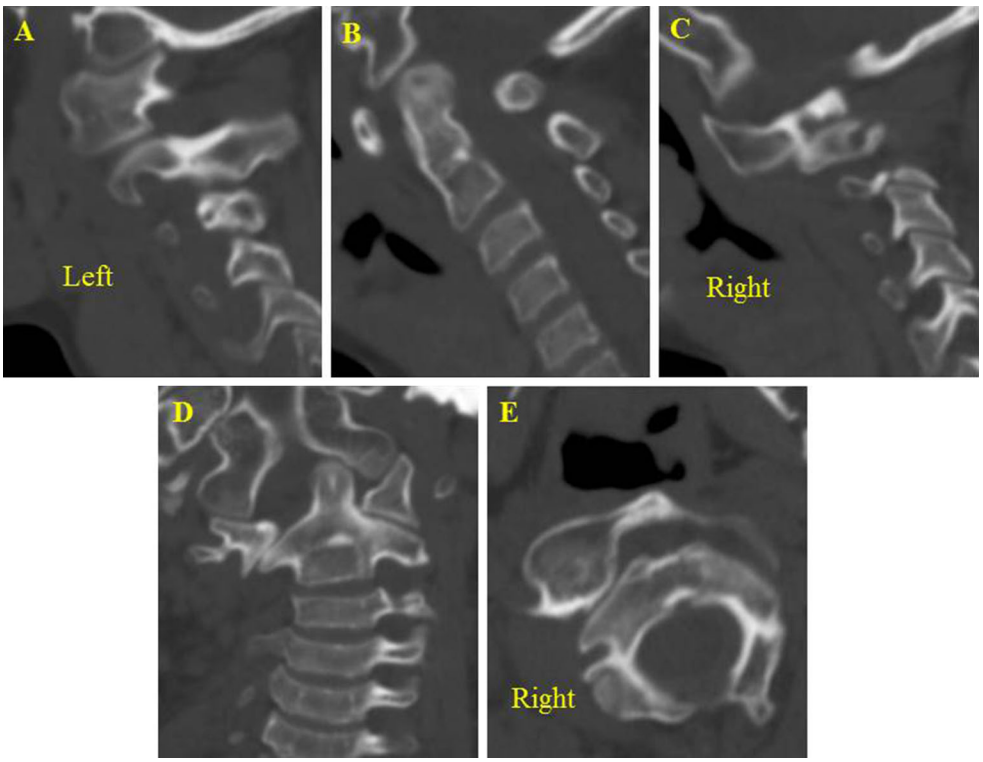

Fig. 2 Preoperative CT scan of the cervical spine (**a**, **b**, **c** are sagittal representations, **d** is a coronal cut and **e** axial cut) confirming the rotatory subluxation with a pivot over the left C2 lateral mass and no significant bony erosion

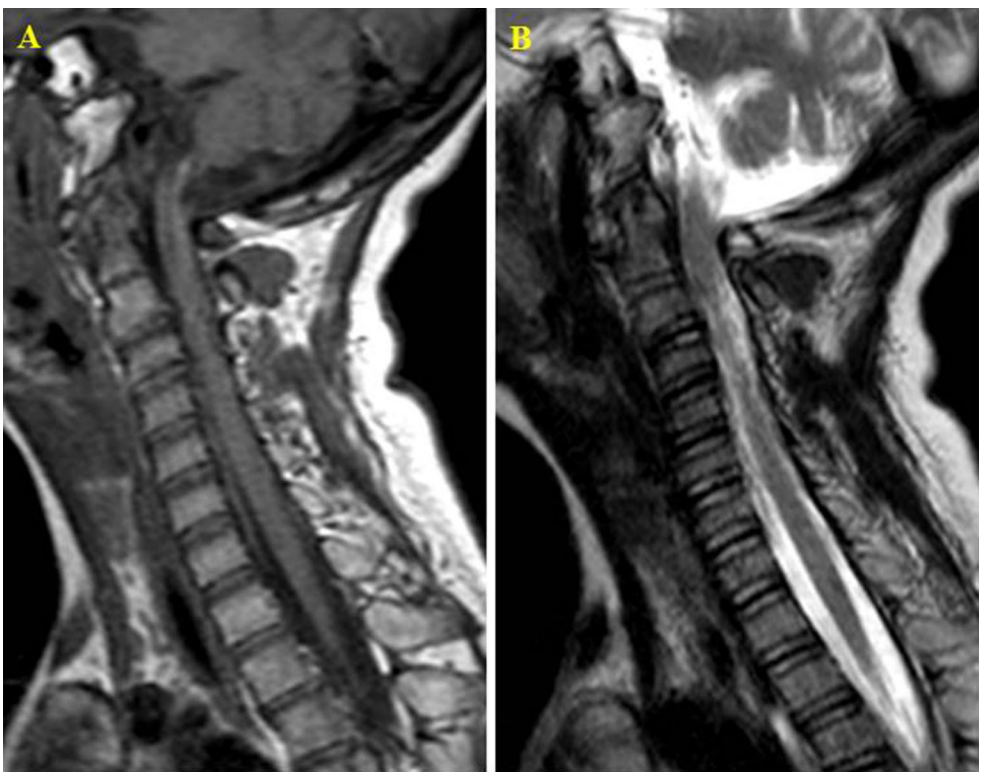

Fig. 3 Preoperative MRI of the cervical spine (**a** and **b** are T1 and T2 weighted images, respectively) showing no evidence of myelopathic changes or inflammatory lesions. Strong movement artefact affecting the quality of the images can be noted

Table 1 Ranawat classification

Class	Description
1	No neurological deficit
2	Subjective weakness, dysesthesia, and hyperreflexia
3A	Objective weakness and long tract signs, patient remains ambulatory
3B	Objective weakness and long tract signs, patient no longer ambulatory

(vertebral body displacement is >3 mm relative to the next lower vertebra when measured from the posterior line of the vertebral bodies) [10, 11].

Atlanto-axial subluxation prevalence in JIA is 33 % and diagnosed if the anterior atlanto-dental interval (AADI) was greater than 3 mm whilst an AADI >5 mm suggests instability was further classified by Fielding and Hawkins (Table 2) [12].

The absence of neck symptoms in JIA does not exclude radiological abnormality [9] and radiographic cervical spine assessment is, therefore, necessary in all JIA patients. Baseline cervical X-rays (AP and lateral) and dynamic images (flexion/extension views) are useful aids. Although a cervical spine MRI with intravenous gadolinium contrast has been advocated for all patients with JIA [13], we use it to evaluate symptomatic patients to help identify inflammation sites, osseous reaction, and erosions and to better determine the extent of canal narrowing and neural compromise [6].

Rationale for treatment and evidence-based literature

Long-term follow-up studies suggest that the course of JIA extends into adulthood with only 40 to 60 % of the patients in clinical remission at follow-up with predictors of poor outcome including severity and extent of arthritis at onset, symmetrical disease, early involvement of wrist and hip, early radiological findings, and a positive rheumatoid factor [14]. With this in mind, there has been a significant

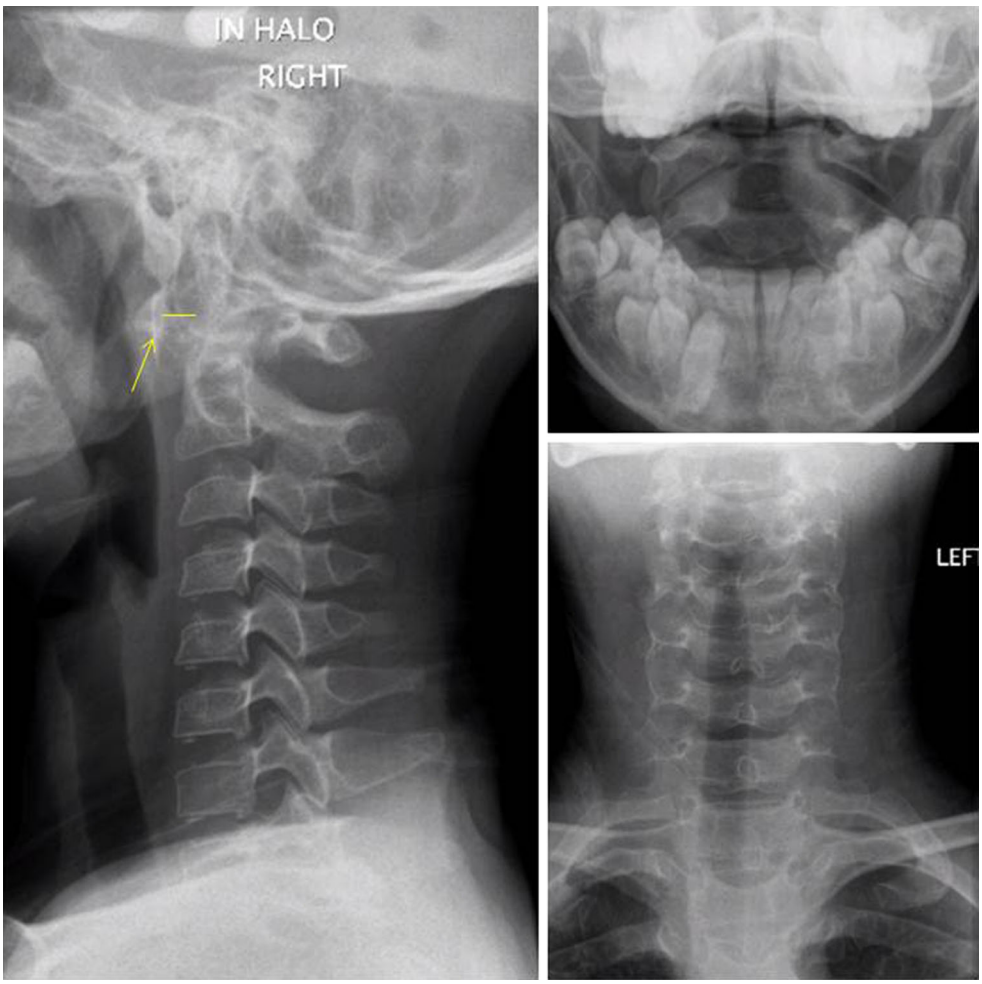

Fig. 4 Cervical spine X-rays in Halo traction. Note the C1/2 is better aligned but the AADI remains abnormally large at 5.6 mm

Table 2 Fielding and Hawkins classification of atlanto-axial subluxation

Type	Description	Anatomic consideration
I	Rotatory subluxation with no anterior displacement (AADI <3 mm)	Transverse ligament intact
II	Rotation about one intact facet with increase AADI 3–5 mm	Transverse ligament insufficient and unilateral capsular tear whilst the other acts as a pivot, alar ligaments intact
III	Bilateral anterior subluxation with AADI >5 mm	Failure of the alar ligament, C1 facet locks over C2
IV	Complete posterior displacement of the atlas	A posteriorly displaced type II dens fracture

improvement in the functional outcomes due to disease modification therapy [1, 15].

The aims of treatment in JIA cervical instability are to improve pain and protect neural tissue. Surgical indications for axial spine fusion include AADI >8 mm on flexion/extension view, or AADI = 5–8 mm with radiological evidence of cord impingement and in patients with a deteriorating neurological picture. Multiple C1/2 fusion methods have been described including posterior wiring (using Gallie's wiring on lay grafting, Brooks–Jenkins inter-laminar grafting), posterior inter-laminar clamps, Magerl's transarticular C1/2 screws and Goel's C1 lateral mass screw with C2 pars or pedicle screw fixation. Biomechanically, evidence suggests the Magerl's fixation to provide the highest stability across C1/2 when compared to wiring or inter-laminar clamps particularly in limiting rotation [16]. An anterior approach to the C1/2 fusion was also described either through a trans-oral approach using a plate or through an anterior cervical approach using transarticular screws [17].

Post-surgical complications after posterior fusion has been reported; these include pseudarthrosis, postoperative re-dislocation, neurological injury, and malalignment of the cervical spine (mainly with posterior wiring techniques and posterior inter-laminar clamping) [18].

Our case demonstrated significant C1/2 instability with significant malalignment despite the initial Halo stabilisation. During our surgical planning, our aim was to achieve a stiff, stable reduction to attain a solid fusion. Magerl's transarticular C1/2 screws and Goel's C1 lateral mass screw with C2 pars or pedicle screw fixation were options but anatomic considerations in a this paediatric patient (favourable C2 pedicle diameter, relatively small C1 lateral masses and the good quality of the cortical laminar bone) meant that using a hybrid fusion system combining posterior transarticular C1/2 fusion with a posterior C1 lamina clamp is likely to accomplish the intervention objectives whilst avoiding the instrumentation of small C1 lateral masses whilst providing the added rigidity from multipoint fixation compared to transarticular screws in isolation.

Procedure

The procedure was done under general anaesthesia, image intensifier guidance with the patient prone and the AAS was reduced before the Halo was held with a Doro clamp to maintain reduction. A small midline posterior incision to expose C1 and C2 was used in addition to two para-median stab incisions (approximately 1 cm off midline) positioned

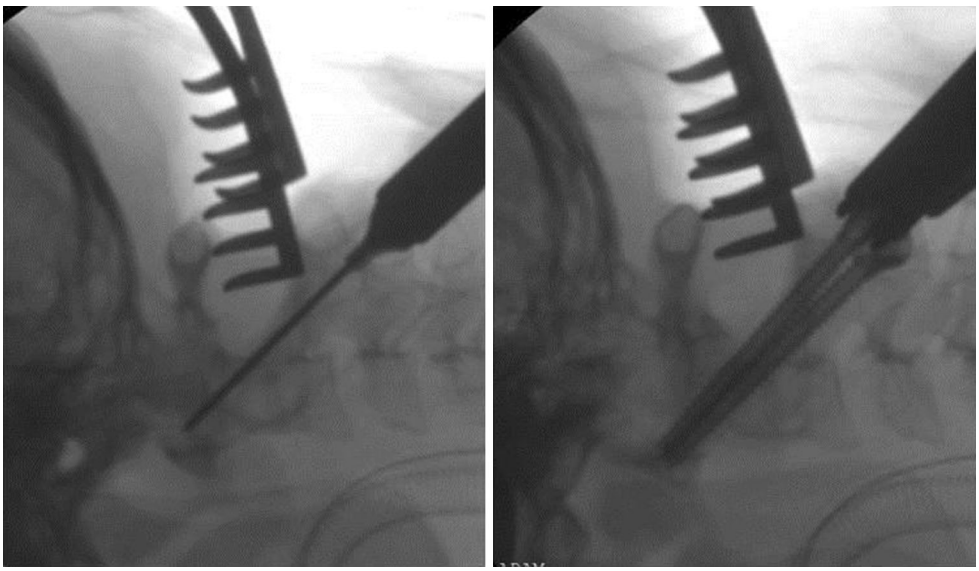

Fig. 5 Intraoperative K-wire guided insertion of transarticular C1/2 screws

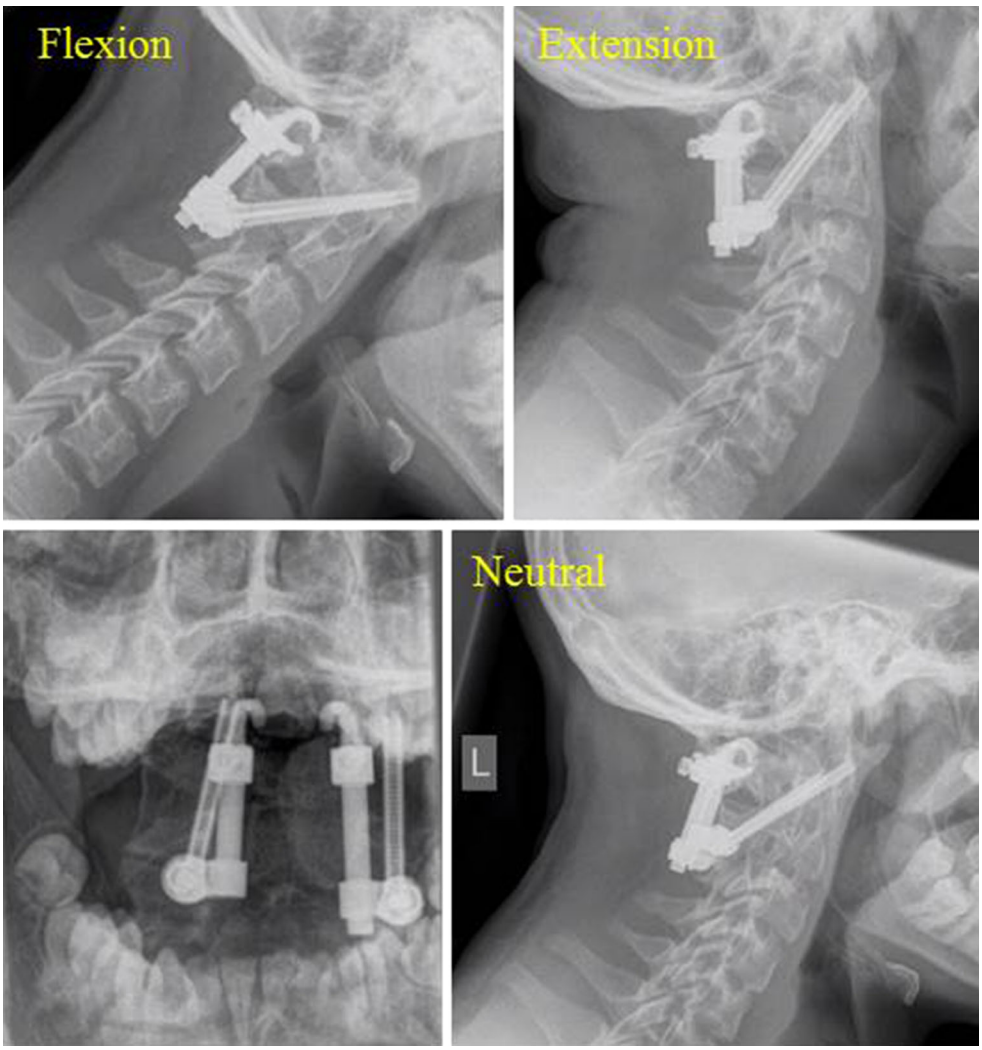

Fig. 6 Final follow-up cervical spine X-rays showing a stable C1/2 motion segment

with the aid of an image intensifier to facilitate the insertion of a guide wire from C2 to C1.

The entry point for the guide wire lies on the dorsal aspect of the axis at the junction of the lamina with the articular mass. The trajectory is guided by an image intensifier positioned for a lateral view and the wire is directed in a sagittal plane to avoid injury to the vertebral artery laterally and the contents of the spinal canal medially. A useful aid with the trajectory is the medial wall of the C2 pedicle which can be felt with a dissector to help avoid unintended wire conversion. The guide wires are directed down the C2 pars and pedicle complex and across the C1–C2 joint, aiming at the anterior tubercle of C1. The tip of the K-wire is advanced to a point 3–4 mm posterior to the anterior C1 tubercle (to avoid penetration of the wire into the retropharyngeal area). 3.5 × 40 mm cannulated screws were used. The screws were then connected to locked C1 posterior arch hooks (NEON^{TM} Cervical System).

Bone graft was then harvested from the iliac bone and put over the decorticated C1 and C2 laminae. Haemostasis achieved and the wound was closed over a gravity drain. Estimated blood loss was 500 ml (Fig. 5).

Outcome, follow-up

Postoperatively, the patient was monitored in a paediatric ward, had no neurological deficits and was mobilised 2 days following his surgery. He did not require the use of a cervical collar and his neck was mobilised as comfort allowed. He was discharged 6 days following his surgery after an uneventful stay in the hospital. The patient had satisfactory regular follow-ups as an outpatient and at his final review 6 years post-surgery, he remains asymptomatic with a no functional limitation relating to the cervical range of motion (Fig. 6).

Conflict of interest None.

References

1. Ravelli A, Martini A (2007) Juvenile idiopathic arthritis. Lancet 369(9563):767–778 Epub 2007/03/06
2. Singh S, Jat KR (2008) Juvenile idiopathic arthritis: an update. Indian J Rheumatol 3(3):110–119
3. Petty RE, Southwood TR, Manners P, Baum J, Glass DN, Goldenberg J et al (2004) International League of Associations for Rheumatology classification of juvenile idiopathic arthritis: second revision, Edmonton, 2001. J Rheumatol 31(2):390–392 Epub 2004/02/05
4. Fried JA, Athreya B, Gregg JR, Das M, Doughty R (1983) The cervical spine in juvenile rheumatoid arthritis. Clin Orthop Relat Res 179:102–106 Epub 1983/10/01
5. Reiter MF, Boden SD (1998) Inflammatory disorders of the cervical spine. Spine 23(24):2755–2766 Epub 1999/01/08
6. Hospach T, Maier J, Muller-Abt P, Patel A, Horneff G, von Kalle T (2014) Cervical spine involvement in patients with juvenile idiopathic arthritis—MRI follow-up study. Pediatr Rheumatol Online J12(1):9 Epub 2014/03/07
7. Samartzis D, Shen FH, Herman J, Mardjetko SM (2010) Atlantoaxial rotatory fixation in the setting of associated congenital malformations: a modified classification system. Spine 35(4): E119–E127 Epub 2010/02/18
8. Ranawat CS, O'Leary P, Pellicci P, Tsairis P, Marchisello P, Dorr L (1979) Cervical spine fusion in rheumatoid arthritis. J Bone Joint Surg Am 61(7):1003–1010 Epub 1979/10/01
9. Collins DN, Barnes CL, FitzRandolph RL (1991) Cervical spine instability in rheumatoid patients having total hip or knee arthroplasty. Clin Orthop Relat Res 272:127–135 Epub 1991/11/01
10. Elhai M, Wipff J, Bazeli R, Freire V, Feydy A, Drape JL et al (2013) Radiological cervical spine involvement in young adults with polyarticular juvenile idiopathic arthritis. Rheumatology (Oxford) 52(2):267–275 Epub 2012/04/20
11. Pennecot GF, Gouraud D, Hardy JR, Pouliquen JC (1984) Roentgenographical study of the stability of the cervical spine in children. J Pediatr Orthop 4(3):346–352 Epub 1984/05/01
12. Fielding JW, Hawkins RJ (1977) Atlanto-axial rotatory fixation. (Fixed rotatory subluxation of the atlanto-axial joint). J Bone Joint Surg Am 59(1):37–44 Epub 1977/01/01
13. Oren B, Oren H, Osma E, Cevik N (1996) Juvenile rheumatoid arthritis: cervical spine involvement and MRI in early diagnosis. Turk J Pediatr 38(2):189–194 Epub 1996/04/01
14. Ravelli A, Martini A (2003) Early predictors of outcome in juvenile idiopathic arthritis. Clin Exp Rheumatol 21(5 Suppl 31):S89–S93 Epub 2004/02/19
15. Oen K, Malleson PN, Cabral DA, Rosenberg AM, Petty RE, Cheang M (2002) Disease course and outcome of juvenile rheumatoid arthritis in a multicenter cohort. J Rheumatol 29(9):1989–1999 Epub 2002/09/18
16. Grob D, Crisco JJ 3rd, Panjabi MM, Wang P, Dvorak J (1992) Biomechanical evaluation of four different posterior atlantoaxial fixation techniques. Spine 17(5):480–490 Epub 1992/05/01
17. Sen MK, Steffen T, Beckman L, Tsantrizos A, Reindl R, Aebi M (2005) Atlantoaxial fusion using anterior transarticular screw fixation of C1–C2: technical innovation and biomechanical study. Eur Spine J 14(5):512–518 Epub 2005/01/26
18. Toyama Y, Matsumoto M, Chiba K, Asazuma T, Suzuki N, Fujimura Y et al (1994) Realignment of postoperative cervical kyphosis in children by vertebral remodeling. Spine 19(22): 2565–2570 Epub 1994/11/15