Atlanto-axial Chronic Rotational Subluxation

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Introduction. Atlanto-axial rotational subluxation, a relatively common condition in pediatric patients, is normally secondary to traumatic inflammatory processes and is favored by the great degree of cervical mobility in children. If untreated, the condition may become chronic and result in a fixed cervical deformity whose diagnosis and treatment may pose a significant challenge. Only a few series have been published, all of them containing a small number of cases and there is an absence of definite criteria for the diagnosis and treatment of this condition.
Clinical cases. Three cases are presented of atlanto-axial chronic rotational subluxation treated over a 15-year period. Ages at presentation were 9.7 and 16 year. In all patients, the same protocols, both diagnostic (x-rays, CT-scan and MRi in one case) and therapeutic (gradual halo traction until reduction was confirmed by a CT-scan and halo cast immobilization) were used. In one case full reduction was not achieved and, consequently, a C1-C2 posterior arthrodesis was performed.
Results. All three patients had a minimum follow-up of 2 years. Patients not subjected to arthrodesis showed a satisfactory clinical situation and normal cervical motion; the remaining patient had slight torticollis with somewhat limited cervical rotation. There have been no relapses.
Conclusions. Atlanto-axial chronic rotational subluxation is a condition with severe and painful repercussions for the patient and whose management poses serious challenges. In our series, all cases were treated following the same protocol: progressive cranial traction and halo cast immobilization. Complete reduction was achieved, while in the remaining one the reduction achieved was only partial, which made it necessary to perform a C1-C2 posterior arthrodesis. As the literature does not provide hard-and-fast treatment criteria, we suggest possible protocols for action.

Key words: *atlanto-axial rotational subluxation, rotational fixation, torticollis, halo traction.*

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Received: November 2005.
Accepted: June 2006.

Subluxación rotatoria crónica atlanto-axial

Introducción. La subluxación rotatoria atlantoaxial es relativamente frecuente en pacientes pediátricos, habitualmente secundaria a procesos inflamatorios o traumáticos, y favorecida por la gran movilidad cervical infantil. En pacientes no tratados puede cronificarse, produciendo una deformidad cervical fija de difícil diagnóstico y tratamiento. Hay escasas series publicadas, todas ellas con pocos casos, no existiendo criterios definidos para su diagnóstico y tratamiento.
Casos clínicos. Se presentan tres casos de subluxación rotatoria crónica atlanto-axial tratados en un período de 15 años. Las edades de presentación fueron 9,7 y 16 años. En todos se siguió el mismo protocolo diagnóstico (radiografías, tomografía axial computarizada, en un caso resonancia magnética nuclear y terapéutico: tracción progresiva con halo hasta la reducción comprobada por tomografía axial computarizada e inmovilización con halo-yeso; en 1 caso no se consiguió la reducción completa, por lo que se hizo una artrodesis posterior C1-C2.
Resultados. Los tres pacientes tienen un seguimiento mínimo de 2 años, con una buena situación clínica y movilidad cervical normal en los no artrodesados; el otro paciente presenta una ligera actitud en tortícolis con una leve limitación de la rotación cervical. No ha habido recidiva del cuadro.
Conclusiones. La subluxación rotatoria crónica atlanto-axial es una afección rara, que implica graves repercusiones funcionales y dolorosas para el paciente, siendo su manejo dificultoso; en esta serie, se trataron todos los casos siguiendo el mismo protocolo: tracción craneal progresiva e inmovilización con halo-yeso, obteniéndose una reducción completa en 2 casos e incompleta en uno, por lo que se efectuó artrodesis posterior C1-C2; en la literatura no existen criterios unificados de tratamiento, por lo que se proponen protocolos de actuación.

Palabras clave: *subluxación rotatoria atlanto-axial, fijación rotatoria, tortícolis, tracción halo.*

Atlanto-axial rotational subluxation, described by Bell in 1830, is one of the most frequent causes of torticollis in pediatric patients, and is normally secondary to different kinds of trauma, the upper-respiratory tract infection known as Grisel syndrome[1], head and neck surgery, inflammatory arthritis, etc. The denomination "rotational subluxation" has been questioned by various authors, due to the fact that rotational displacement takes place in children within their normal range of cervical mobility[2]; thus, several other names have been suggested to define this clinical entity: rotational dislocation, rotational deformity, rotational displacement and spontaneous hyperemic luxation[3]. The patient presents torticollis and his head is inclined laterally and with contralateral rotation and there is spasm of the ipsilateral sternocleidomastoid muscle in the attempt to overcome the torticollis. Radiological diagnosis poses a significant challenge and it is therefore necessary to resort to an axial CT-scan (CTS), which will render a clear image of the injury[4].

When the deformity persists, it may turn out to be irreducible, causing a condition of rotatory fixation, classified by Fielding into 4 types[5], assuming a true atlantoaxial chronic rotational subluxation (AACRS). The treatment that is generally applied is cranial or cervical traction, with progressive reduction followed by immobilization; however, there are no hard and fast guidelines for treatment just as there are no criteria to decide whether surgical stabilization is necessary or not[6,7]. Only a few series have been published and there is no agreement as to the correct criteria for diagnosis or treatment. Because of this, the aim of this work is to carry out a study of the cases we have treated and, by comparing it with the existing literature, propose a protocol for the treatment of AACRS.

CASE REPORTS

We present three cases of AACRS treated in our center between 1989 and 2003. The three patients had been referred from other centers and presented with a typical head deformity in lateral flexion and in contrarlateral rotation; observation with a three-dimensional CTS was carried out in the third case, and magnetic resonance imaging (MRi) in the first two, after which a diagnosis of AACRS was reached. They were classified using Fielding's categorization, effected according to the sagittal displacement of the atlas[5]: type I, no displacement; type II, anterior 3-5mm displacement; type III, over 5mm displacement; and type IV, posterior displacement. All the patients were treated, for a period of three weeks, with gradual halo traction in bed and wheelchair, reaching approximately one fourth of their body weight. In the same period, a control CTS was carried out and cervical mobility was tested clinically.

Case 1

We treated a 9-year-old male patient who presented torticollis associated to a fracture of the collar bone during a fall in the gym class. The patient was first treated in his home town with an 8-shaped bandage for the collar bone fracture, and with muscle relaxants and physiotherapy; 15 weeks later, he was sent to our center, due to the persistence of the symptoms (fig. 1). The X-ray images did not bear the necessary information, a fact which led to the performance of a CTS and an MRi, and the ensuing diagnosis of AACRS type I3, with a C1-C2 42° divergence (fig.2); gradual halo traction was initiated up to a weight of 7kg. at which it was stopped due to the appearance of dysarthria (17-day traction), with slightly reduced rotational mobility. The control CTS showed an 11° residual divergence, as a result of which and also with a view to preventing a relapse, a C1-C2 *in situ* arthrodesis was carried out with an iliac crest graft (fig. 3), maintaining traction during the operation and immobilizing the area with a halo cast. Once the traction was removed, the

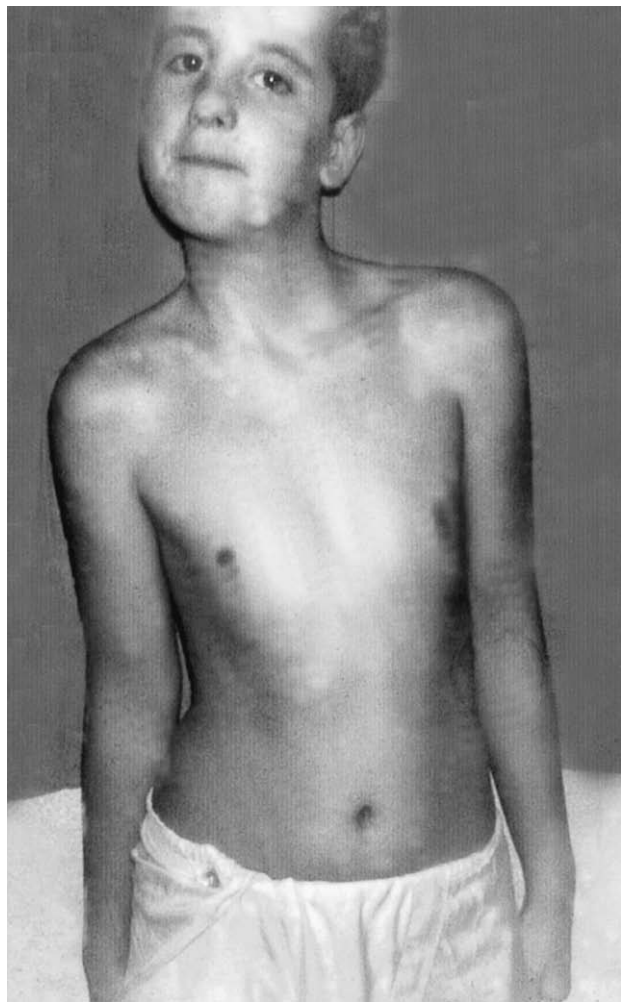

Figure 1. *Clinical view of the deformity of the first case.*

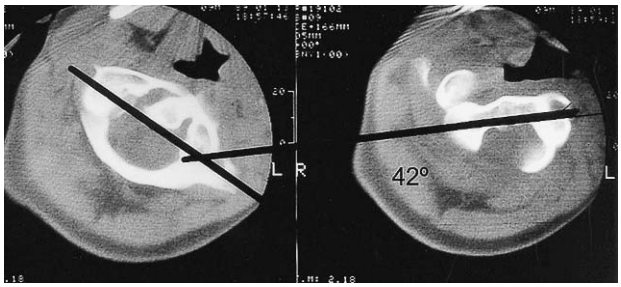

Figure 2. *Axial CT scan of patient n°1 showing the measurement method with a 42° atlanto-axial divergence.*

dysarthria remitted spontaneously. Fifteen years later, the patient presents with cervical mobility that is practically normal with a 10° limitation of rotation to the left.

Case 2

We treated a 7-year-old boy with a record of fixed torticollis that had evolved for 17 weeks and had originated in an infection in the upper respiratory tract. Before being sent to us, the patient had been treated with non-steroid anti-inflammatory drugs (NSAIDS), cervical orthosis of the Schanz type and elastic traction followed by cervical ortho-

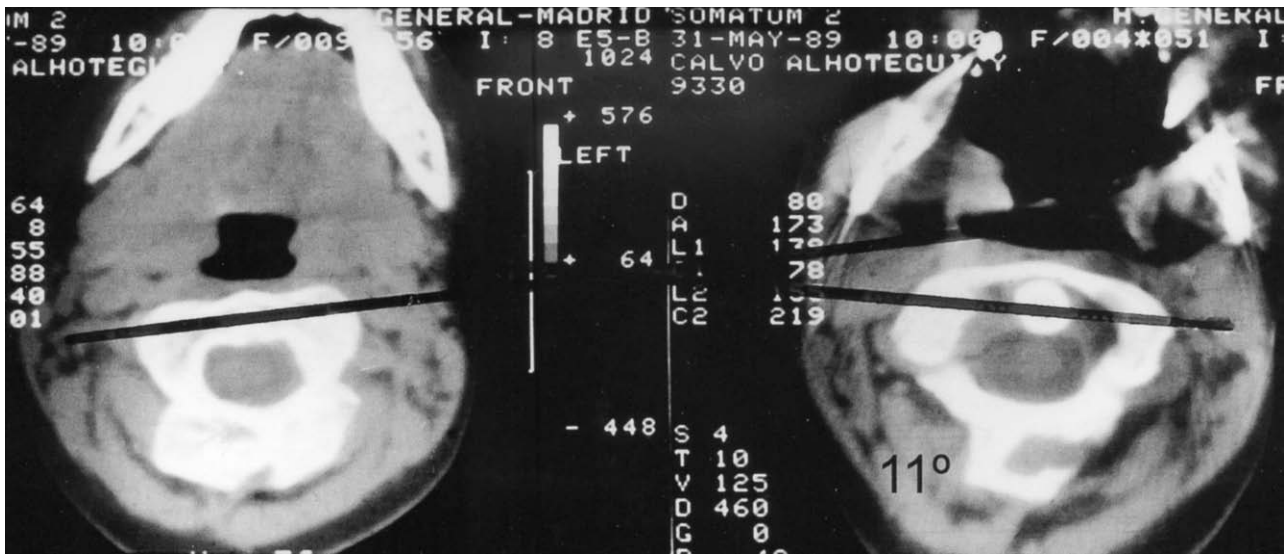

Figure 3. *Postsurgical CT scan of the first patient, where the satisfactory mass of arthrodesis and an 11° residual divergente are shown.*

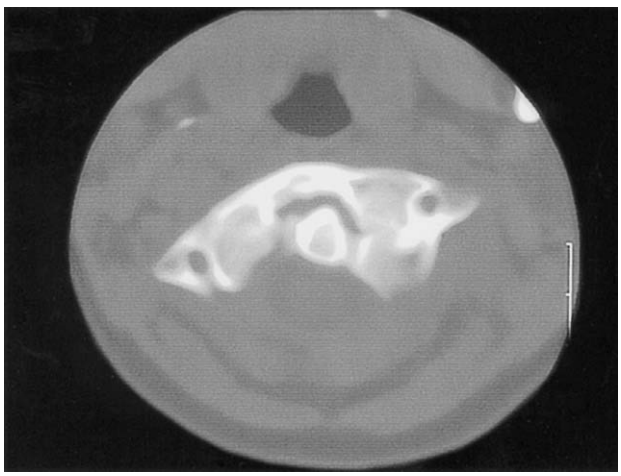

Figure 4. *Axial CT scan of the second patient showing the rotational deformity of the C1-C2 complex with an angle of 25°.*

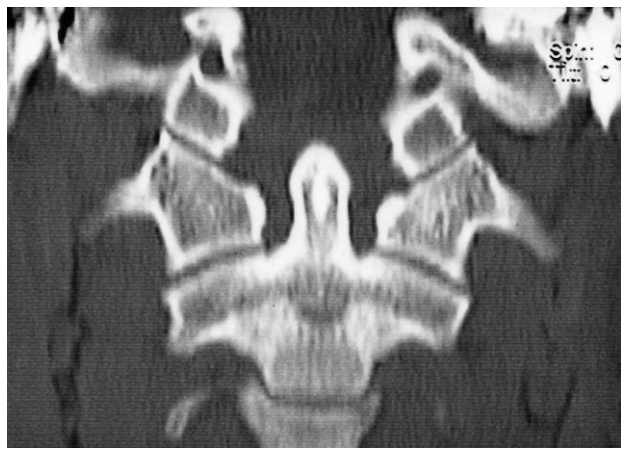

Figure 5. *Axial CT control scan reconstruction 3 weeks later, showing the symmetry between the odontoid process and the lateral masses of the atlas, with complete correction of rotation.*

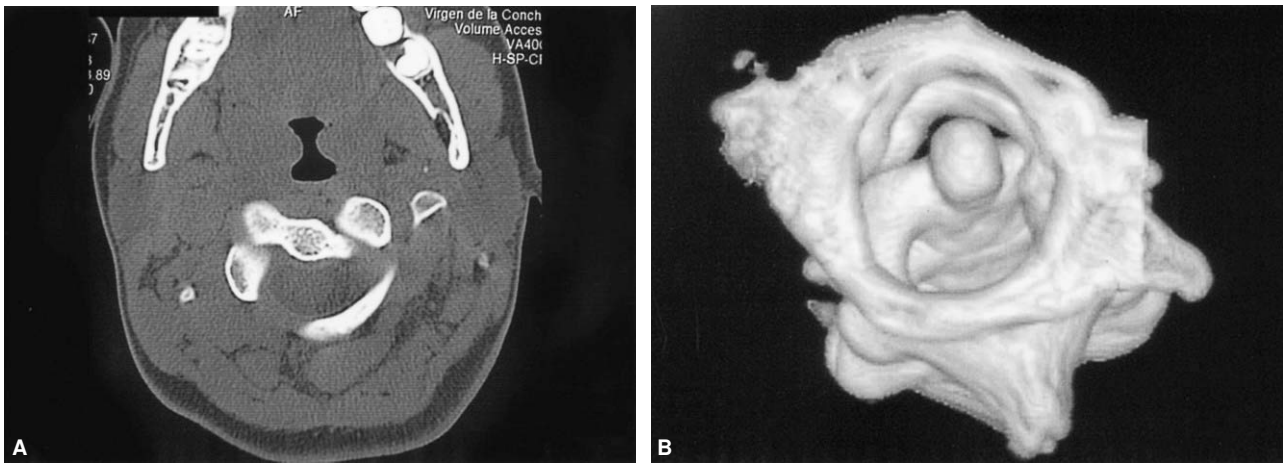

Figure 6. *A: Conventional axial CT-scan showing the 32° rotation between C1-C2 in patient N°3. B: three-dimensional reconstruction showing the typical deformity of rotation and C1-C2 impingement.*

sis, but there had been no positive results. The CTS led to a diagnosis of AACRS type I, with a 25° divergence (fig. 4), and treatment with traction was applied, up to 8kg. Three weeks later the patient presented symmetric rotations and the CTS (fig. 5) showed a complete reduction; consequently, the area was immobilized for 6 weeks with halo cast and treatment was subsequently continued, for another 6 weeks, with soft cervical orthosis. Eight years later, the functioning is normal and there are no sequels.

We treated a 16-year-old female patient with a record of fixed torticollis produced after thyroid surgery who was sent to our center after having showed no improvement on being treated with NSAIDS and a soft cervical orthosis. The diagnosis for AACRS Fielding type I, was reached through CT-scan (fig. 6A) with a three-dimensional reconstruction (fig. 6B) using the same protocol as with the other patients and reaching 14kg. Three weeks later mobility was symmetrical and the control CT-scan confirmed the complete reduction of the rotational divergence (fig. 7); accordingly, the area was immobilized with a halo cast for a period of 6 weeks and with a soft cervical orthosis for a further 6- week period. Two years later, the patient's function is completely normal, and there have been no instances of cervical pain or torticollis.

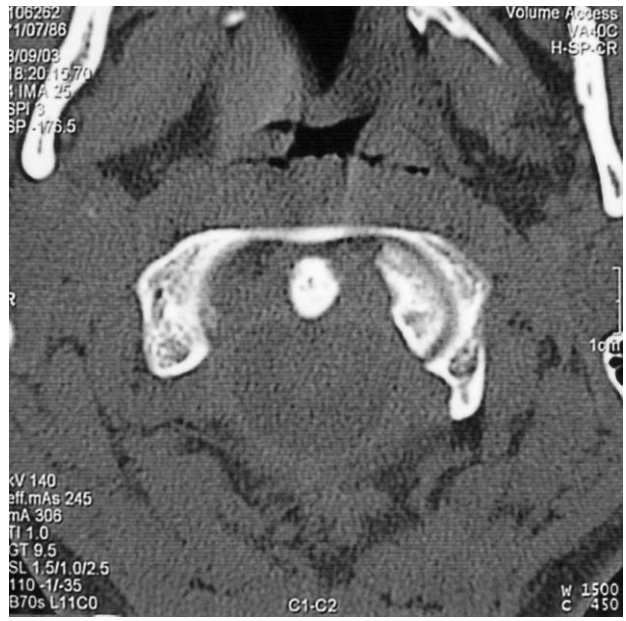

Figure 7. *CT- scan of the third patient, showing appropriate reduction after three weeks' traction treatment.*

DISCUSSION

AACRS is a typical pediatric condition due to the special anatomic features of the atlanto-axial joint in children. These features have been appropriately described by Kawabe et al[8]: the presence of richly vascularized synovial folds in the lateral joints, which disappear gradually with age, a major inclination and convexity of the C2 facet joints, and C1-C2 hypermobility[2]. The following is a possible sequence leading to the establishment of AACRS: synovial inflammation or joint capsule rupture, rotational displacement, muscle spasm fixing the rotational position, interposition of hypertrophic synovia or of ruptured capsule and, lastly, the establishment of irreducibility with local fibrosis and definitive muscular spasm[6,8].

Atlanto-axial rotational subluxation is one of the most frequent causes of torticollis in children and is normally secondary to different kinds of trauma[6], upper-tract breathing infection[1], head and neck surgery[9], inflammatory arthritis[10], etc. Also, several cases have been found to be associated to fracture of the collar bone[11]. On being examined clinically, the patient presents with a stiff neck with compensating trunk inclination, which progresses gradually to reach the final clinical state: head inclined to one side and

rotated contralaterally, with marked spasm of the ipsilateral sternocleidomastoid muscle on cephalic rotation (fig. 1) and in the attempt to correct the deformity, considerable cervical rigidity and rotational incapacity. When diagnosed in its initial stages, it usually responds to traditional treatment such as cervical orthosis associated to NSAIDS, traction followed by orthosis, physical therapy, etc[9]; in undiagnosed cases or in partially treated cases it may become chronic, constituting rigid AACRS, also known as atlanto-axial chronic rotational fixation[15,6,8].

Fielding classified AACRS into 4 types, according to the sagittal displacement of the atlas[5]. In type I the transverse ligament was intact and in the other three there could be potential rupture and instability. However, this classification was drawn up before the appearance of CT scanning, and was based on lateral x-rays and cineradiography. Waegeneers attempted a new classification based on CT-scanning, which is complex and difficult to manage–since it defines 5 types and 24 subtypes–and which questions the validity of type IV, pointing out that it does not seem to imply transverse ligament rupture and could therefore be included in type I.

Radiological diagnosis is difficult because deformity and pain make it difficult to obtain good x-ray views; thus, in the simple anteroposterior X-ray, the head is seen to overlap, and in the lateral X-ray there is a lack of parallelism between the skull and the spine; the transoral odontoid x-ray can be of great use, since it shows an asymmetry between the odontoid and the lateral masses of the atlas with possible facet joint impingement[8]. The use of dynamic CT-scans has been recommended, with rotation to the right and left of the head. However, under the condition of AACRS, movement is painful, due to which the patient will probably not cooperate and the diagnosis will be confusing[13]. CT-scans do not seem to be advisable in the cases of acute torticollis[14,15], but in persistent cases in which AACRS is suspected they are highly useful for confirming diagnosis, since they show the C1-C2 rotational divergence that can be quantified by measuring the angle between the lines that join the transverse processes of the atlas to those of the axis (fig. 2)[16]. The three-dimensional CT-scan shows the rotational deformity with the complete displacement of the facet joints, the asymmetric placement of the odontoid in the anterior arch of the atlas–which can in turn provide information about the integrity of the transverse ligament–and C24 spinal displacement. MRi is a good complementary method because it can provide information on the interposition of soft tissue as well as on possible damage to the transverse ligament, due to which it may help decide whether surgical stabilization is necessary or not. In our series, and on the presumption of an AACRS, a simple CT scan was used as diagnostic method (fig. 2 and 4A). In the third case, it was completed with a three-dimensional reconstruction (fig. 4B), which showed the torsion alterations in the atlanto-axial complex with clarity.

Due to the scantiness of published series, no agreement has been reached regarding treatment. Scapinelli[4], for instance, carried out a reduction with general anesthesia and immobilization with a Minerva type cast for three months; Phillips[13] made a proposal in which subluxation with more than a month's evolution is treated by means of traction, preferably halo and fixation in the cases in which there is no improvement or in which there is redisplacement. Loder and Hensinger[3] posit C1-C2 arthrodesis in cases with more than three months' evolution or in which there is transverse ligament rupture. Fielding[5] recommends traction followed by *in situ* arthrodesis for AACRS, even if the correction of the deformity is incomplete. Subach et al[9] suggest the use of traction followed by immobilization during 3 weeks, although they practiced open reduction with posterior arthrodesis in a case of AACRS; Park et al[18] put forward a case in which they performed skull traction for 6 weeks and immobilization for 6 months. Govender and Kumar[7] presented 7 cases of AACRS in which, on not obtaining reduction after skull traction for 11 days, they practiced posterior *in situ* arthrodesis before exeresis of fibrous tissue, interposed between the atlanto-axial and atlanto-odontoid joints, and effected transorally; Crockard et al[19] carried out open reduction laterally in two cases, and they even found bone healing. Cranial traction is widely accepted, being the technique used by the majority of authors[3,5-7,9,13,17], but there is no agreement regarding which type of traction is the best: soft or skeletal, halo or compass; neither is there agreement regarding the weight to be used, although Fielding arbitrarily suggests a 6.8kg traction limit for children[20], or the appropriate traction system to be used.

Subsequent treatment has not been appropriately determined either; accordingly, when reduction has been reached, some authors propose immobilization with acervical orthosis, whereas in cases of AACRS there seems to be a tendency towards arthrodesis[3,5,9,13].

In our case study we have used a determined strategy at all times: gradual halo traction of the Stagnara[21] type up to one fourth of body weight, following the principles used in the treatment of scoliosis[22] and maintaining traction for 3 weeks (since we believe that traction with halo provides greater control and is more comfortable than compass traction), which allows us to complete immobilization with a vest and cast. We also believe that three weeks is a satisfactory period of time for progressive reduction to take place, which is also favored by the patient's gradual weight gain. In the first case, traction had to be stopped due to the appearance of dysarthria, possibly caused by excessive traction on the hypoglossal nerve[23], which corrected itself spontaneously, once reduction had been confirmed through a CT-scan. In two cases treatment was continued with halo cast immobilization (which is notably more reliable than halo vest and is more easily tolerated by children), for three weeks and a soft cervical orthosis for another 6 weeks if there is no relapse into AACRS. As regards subsequent fix-

ation by means of arthrodesis–which, as we have stated above, is considered the treatment of choice by several authors–we considered it only in the cases of complete reduction, as suggested by Arlet et al[24]; the technique used, *in situ* arthrodesis with intraoperative traction maintained with autologous iliac crest grafting and halo cast immobilization, seems to be sufficient, since the tendency to spontaneous fusion after subperiostic dissection[25]–which renders the use of sublaminar wires or any other type of osteosynthesis unnecessary–is an important advantage to be considered together with the great osteogenic capacity of children and the high degree of tolerance to immobilization.

In conclusion, AACRS is an unusual condition, secondary to the presence of torticollis of any kind. Therefore the first step to be taken should be to prevent its establishment through appropriate treatment, such as cervical orthosis, soft traction, physical therapy, etc. In the cases in which it does appear, treatment should be immediate, for which we propose the following protocol: *1.* Halo traction for 3 weeks until, if necessary and possible, one fourth of the body weight is reached. *2.* If reduction is complete (confirmed by CT-scanning), immobilization with a halo vest or halo cast for 6 weeks and follow-up with another 6 weeks with a soft neck brace. *3.* If reduction is incomplete and there remains residual atlanto-axial divergence, *in situ* posterior C1-C2 arthrodesis should be used, followed by immobilization of the same kind as mentioned above.

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Conflict of interests: We, the authors, have not received any economic support to carry out this study. Nor have we signed any agreement with any commercial firm to receive benefits or fees. On the other hand, no commercial firm has provided nor will provide economic support to non-profit foundations, educational institutions or any of the other non-profit organizations that we are members of.