Accepted Manuscript

Title: Bilateral C1 laminar hooks combined with C2 pedicle screw fixation in the treatment of atlantoaxial subluxation after Grisel's syndrome

Author: Luis C. Morales, Fernando Alvarado, José A. Corredor, Andrés Rodríguez

PII: S1529-9430(16)30867-1
DOI: http://dx.doi.org/doi: 10.1016/j.spinee.2016.08.016
Reference: SPINEE 57132

To appear in: *The Spine Journal*

Received date: 12-8-2015
Revised date: 15-6-2016
Accepted date: 3-8-2016

Title: Bilateral C1 laminar hooks combined with C2 pedicle screw fixation in the treatment of atlantoaxial subluxation after Grisel´s syndrome

Concise title: Surgical treatment of Grisel´s Syndrome

Author names and affiliations

Luis C. Morales M.D. Orthopaedic and Spine Surgeon, Hospital Universitario Fundación Santa Fe de Bogotá, cirugía de Columna.

Postal Address: Carrera 7 No. 117-15. Bogotá D.C., Colombia

E-mail: luiscarlosmoralessaenz@gmail.com

Fernando Alvarado M.D. Orthopaedic and Spine Surgeon, Hospital Universitario Fundación Santa Fe de Bogotá, cirugía de Columna.

Postal Address: Carrera 7 No. 117-15. Bogotá D.C., Colombia

E-mail: falvaradogomez@gmail.com

Corresponding author:

José A. Corredor M.D., Orthopaedic and Spine Surgeon, Hospital Universitario Fundación Santa Fe de Bogotá, cirugía de Columna.

Address: Carrera 7 No. 117-15. Bogotá D.C., Colombia

Phone number: (+57) 3124747340 Fax: (+571) 603 0303

E-mail: joalcosa@yahoo.com

Andrés Rodríguez M.D. Orthopaedic and Spine Surgeon, Hospital Universitario Fundación Santa Fe de Bogotá, cirugía de Columna.

Postal Address: Carrera 7 No. 117-15. Bogotá D.C., Colombia

E-mail: andres.rodriguez@ama.com.co

Acknowledgments

The authors would like to thank Teryn Buna from University of British Columbia and Alexander Joeris MD and Melissa Wilhelmi from AO Clinical Investigation and Documentation for their contribution in preparation and copy-editing of the manuscript. Also, we thank Jimmy Peña and Miguel Farfan MD for the drawings in this manuscript.

Abstract

Background context. Many etiologies can lead to atlantoaxial subluxaion. In Grisel´s syndrome, this subluxation occurs spontaneously after inflammatory processes of the head and neck. Diagnosis is typically based on clinical history and a strong suspicion of this syndrome. Non-surgical treatment most often resolves the symptoms, however in some cases surgical treatment is necessary to repair the subluxation. Various surgical techniques and instrumentation systems have been used to treat atlantoaxial subluxation, although there is no consensus regarding the best treatment method for the pediatric population.

Purpose. To describe a case of atlantoaxial subluxation in a child with Grisel´s syndrome treated surgically with an alternative construct.

Study Design/Setting. Case report and literature review.

Methods. Our case study involves a five-year-old female with a six-month history of unresolved Fielding type II atlantoaxial subluxation due to Grisel´s syndrome. Despite conservative treatment, the patient´s symptoms continued to progress. After two failed closed reduction attempts, open reduction and C1-C2 fusion was performed with atlas laminar hook and axis pedicle polyaxial screws. A literature review of the surgical treatment of Grisel´s syndrome was also performed.

Results. After surgery, the patient exhibited full clinical and functional recovery with complete resolution of symptoms. At the 36-month follow-up examination, there was continual evidence of satisfactory reduction and fusion. No complications were observed. Upon completion of the literature review, eight Grisel´s syndrome cases were found to have been treated surgically with the minimum patient age being nine years.

Conclusions. Conservative management of Grisel´s syndrome is the most common and effective treatment; however a few surgical cases have been reported in the literature with good results. Satisfactory clinical results and fusion at 36 months post-surgery were seen in a pediatric patient with atlantoaxial subluxation and instability using atlas laminar hook and axis pedicle polyaxial screws.

Key words: Atlantoaxial subluxation; atlas laminar hook; axis pedicle polyaxial screws; C1 - C2 arthrodesis; Grisel´s syndrome; Goel–Harms arthrodesis.

Introduction

C1-C2 subluxation is an uncommon condition in clinical practice. If spontaneous subluxation of the atlantoaxial joint occurs secondary to infectious or inflammatory processes of the head and neck, it is known as Grisel´s syndrome (GS) and is most often seen in children. In 1830, Sir Charles described the first case of atlantoaxial subluxation and Pierre Grisel described three patients with pharyngitis and torticollis who also experienced dislocation of the C1 – C2 joint nearly a century later [1;2].

Conservative treatment of GS is the gold standard involving anti-inflammatory, antibiotics, and muscle relaxants therapies as well as bed rest, physiotherapy and immobilization with soft collars [3]. These methods typically offer relief of all symptom, and are considered as first line treatment prior to considering surgery [4]. The option to undergo surgical intervention is rare, and generally only taken when symptoms progress and initial treatments are no longer effective [5]. Occipitocervical fixation, C1-C2 wiring with bone grafting (Brooks-Jenkins fusion), transarticular atlantoaxial screws (Magerl technique) or C1 lateral mass screws and C2 transpedicular screw and rods (described by Goels and modified by Harms) [6-9] have been described to achieve intraoperative reduction and subsequent fixation to maintain correct alignment. Potential growth and the possibility of postoperative spinal deformity is a concern associated with cervical arthrodesis in children, especially those younger than ten years.

This case study describes a five-year-old girl with a six-month history of unresolved type II atlantoaxial subluxation classified according to Fielding-Hawkings [10]. Surgical treatment involving open reduction and C1-C2 fusion with atlas laminar hook and axis pedicle polyaxial screws was performed. Based on the final 36-month outcome of this case, we exhibit the outcome of the surgical treatment of atlantoaxial subluxation and instability secondary to inflammatory process (Grisel´s syndrome) in a child in which conservative treatment was unsuccessful.

Materials and Methods

Case presentation

A five-year-old female was admitted to the emergency room of our center with a 15-day history of neck pain, right-sided inclination and left-sided rotation of the head, cervical muscle spasm and a description of electric pain behind her right ear without any mechanism of injury (Figure 1). One week prior to admission, the patient had started micro-nebulization treatment for a broncho-obstructive syndrome. Physical examination revealed cervical adenopathy, pain on palpation of the left side of the neck, and restriction in all cervical movements. Radiographic investigations identified right lateral inclination of the head and rectification of the cervical lordosis (Figure 2). A CT scan revealed asymmetrical odontoid-lateral mass relationship (Figure 3). Based on these findings, initial treatment involved a Philadelphia soft collar and rest. At a two-week follow-up examination, the patient presented with acute sinusitis and further investigations identified persistent impairment of cervical range of motion. Treatment with cervical traction device (1 kg) was started and continued on a daily basis in combination with gentle soft tissue manipulation. Amoxicillin was prescribed for sinusitis treatment and physiotherapy. After 10 days following this inpatient treatment regimen the deformity persisted. Under general anesthesia, closed reduction under CT guidance was therefore attempted without success. A follow-up MRI revealed fluid in the C1-C2 joint. After discussion among all clinic spine surgeons, the diagnosis of Grisel´s syndrome was reached and further monitoring with home treatment including the use of soft collar, analgesia, and physical therapy was recommended. There was no evidence of improvement in symptoms by the five month clinical follow-up; further CT imaging confirmed progression of the dislocation (Figure 4), and the MRI revealed progression of C1-C2 subluxation and right rotation toward the C2 vertebral body, right facet C1–C2 dislocation, loss of relationship between the atlanto-odontoid joint and scoliosis. A new treatment intervention was proposed involving the cervical traction halo-traction system; this device was fitted under general anesthesia with the traction level starting at 1 kilogram (equivalent to 5% of the total weight) and progressively increasing to 20% of the total weight over 10 days (Figure 5). A second attempt of closed reduction under CT examination was done, using the transoral method according to Jeszenszky [11], achieving partial reduction. Further cervical traction with the halo traction device was undertaken for 8 days; however the reduction was not maintained. Consequently, surgical treatment for reduction was considered.

Surgical technique

The patient, stabilized with the halo-traction device, was placed in the prone position and neurophysiological monitoring (including somatosensory- and motor-evoked potential) was performed throughout the surgery. A posterior neutral approach was performed and C1-C2 was subperiosteally exposed. Upon further exposure of the posterior arch of C1 and the lamina of C2 evidence of right C2 facet indentation was apparent. Real-time fluoroscopy was used to place axis pedicle polyaxial screws 3.5 mm in diameter. Reduction was performed under C-arm fluoroscopy to verify alignment and positioning. Laminar hooks were placed on the posterior arch of the atlas, and connection of both constructs was made with posterior rods (size 3.5mm in diameter). The lamina of C1 and C2 were then decorticated posteriorly and a demineralized bone matrix allograft was applied. No intraoperative complications were experienced. Postoperative radiographs demonstrate proper alignment of cervical spine and placement of implant (Figure 6).

Postoperative course

After surgery, almost complete pain relief was achieved and the torticollis subsided. A rapid early postoperative recovery was observed and the patient was discharged five days post-surgery with upper cervical spine soft collar stabilization for 8 weeks. Thereafter, physical therapy was initiated, which included pain management techniques as well as exercises to regain cervical spine mobility. At the 36-month follow-up, C1-C2 spinal fusion was obtained based on radiographic evaluation without any spinal deformity (Figure 7), there were no further reports of neck pain, and gait abnormalities and urinary disturbances were nonexistent.

Discussion.

The case exhibit an effective and safe surgical construct that combines the C1 laminar hooks and C2 transpedicular screws united with connecting rods, for use in the pediatric population (Figure 8). This construct was proposed by Ni et al. as an alternative surgical technique to treat atlantoaxial instability secondary to odontoid fracture, os odontoideum or traumatic rupture of the transverse ligament [12]. A satisfying clinical result and fusion at the 36-month follow–up were achieved with this technique. The construct minimizes the risk of neurological or vascular injuries.

When a static CT scan is performed using the characteristic cock-robin posture of the head, there is a significant chance of misinterpreting the findings [13]. Static CT may facilitate the diagnosis of a subluxation or dislocation of C1 on C2, yet it does not demonstrate the dynamic relationship between this pair [14]. Dynamic relationship between C1 and C2 can be better seen with a Dynamic CT Scan (DCTS) [13,15]. Currently there is a lack of good quality studies in literature recommending its use to specifically evaluate rotatory subluxation. Alany et al, found that DCTS has poor reliability and reproducibility in the diagnosis of atlantoaxial subluxation in patients with acute torticollis. As a result, they suggest that routine use of DCTS is a costly choice in patients with acute torticollis as the deformity usually resolves by a simple cervical mobilization [16]. We consider DCTS doesn't change the treatment, moreover, increase costs and patient radiation exposure. In our case, sequential CT scan were performed, being obvious the progression of subluxation

There is no formal consensus advocating the optimal surgical treatment method, particularly since conservative treatment is most often effective. Analysis of the literature revealed only eight cases of GS treated with arthrodesis (Table 1) where the youngest patient was nine years old [8;17;18;19]. One case was treated with the Goel-Harms technique [8], one with the occipito-C3 technique and wires [17], five cases were treated with C1-C2 sub- and interlaminar wires with an interspinosus allograft [18], and the last one treated with C1-C2 transarticular screws and C1 lateral mass screws [19]. The union rate was 100% for the first two cases without any reported complications. For cases involving treatment using C1-C2 sub- and interlaminar wires and C1-C2 transarticular screws with C1 lateral mass screws, the union rate was not reported. For our knowledge this construct using atlas laminar hook and axis pedicle polyaxial screws has not previously described neither in pediatric population nor in the treatment of GS.

In the pediatric population, the Goel-Harms technique (bilateral insertion of polyaxial-head screws in the lateral mass of C1 and the pedicle of C2) has been an effective method for the management of atlantoaxial instability. Heuer et al. used this technique for six pediatric patients (age range 7.5 to 17 years) with instability secondary to os odontoideum [20]. All patients were reported with complete fusion and normal alignment after an average of 14 months follow-up. Only two patients experienced postoperative paresthesia in C2 dermatome, which resolved spontaneously within two weeks postoperative; there were no reports of vertebral artery injuries, infections or wound dehiscence.

Limitations of Goel-Harms construct are related to the technical difficulties of the surgery as well as the inherent risk associated with placing screws in the lateral masses of C-1. This is due to the presence of immature bone, small bone structures and extensive anatomical variability. In some patients, neurotomy of the spinal nerve must be performed to achieve suitable positioning of the screw in the lateral masses of C1 [12]. Based on these observations, Ni et al. propose arthrodesis using laminar hooks in C1 and transarticular screws in C2 connected with rods for pediatric patients. C1 laminar hooks avoid the neurological risk caused by sublaminar cables that are passed under the C1 arch as well as vascular injury arising from screws positioning within the C1 lateral masses[12]. A disadvantage of this construct is that the C1 level only has one fixation point with limited biomechanical stability; nevertheless the outcome for five patients was excellent, with fusion rates of 100% [12]. In line with this, we considered the use of sublaminar hooks in C1 as safe and effective for the management of atlantoaxial subluxation in pediatric patients.

In order to perform this technique, the C1 posterior arch is needed to bear fixation with hooks. Patients with complete or incomplete congenital absence of the C1 posterior arch (1.5- 5% of the population) [20] or those that require its surgical removal cannot be considered for this type of surgery. These patients would undergo the original Goel-Harms technique. Posterior arch hypoplasia is a relative contraindication that is more commonly found in some patients with atlantoaxial subluxation or instability [20]. The presence of a C2 short pedicle may also limit screw placement, although Sim et al. consider the possibility of using these screws in this anatomic variants; they reported stable fixation using long and short pedicle screws with a low occurrence of vascular or neurological injury [21].

This surgical technique may be applied to children with traumatic injuries, rheumatologic diseases, connective tissue disorders, Grisel´s syndrome and congenital malformations such as Chiari, Down syndrome and os odontoideum, in which C1-C2 fixation is required.

Conclusion.

Based on the outcome of the presented patient, we exhibit an effective, alternative and safe surgical construct using atlas laminar hooks, axis pedicle polyaxial screws, rows connectors and bone allograft in the pediatric population for the treatment of atlantoaxial subluxation and instability. A satisfying clinical result and fusion at the 36-month follow–up were achieved with this technique. As Gisel's Syndrome is a rare diagnosis in children, it might be interesting to evaluate the outcome of children with an atlantoaxial subluxation or instability secondary to traumatic injuries, rheumatologic diseases, connective tissue disorders and congenital malformations such as Chiari, down syndrome or os odontoideum, where C1-C2 fixation is required.

References

1. S. Osiro, K.J. Tiwari, P. Matusz. Grisel's syndrome: a comprehensive review with focus on pathogenesis, natural history, and current treatment options. Childs Nerv.Syst. 2012; 28: 821-5.

2. A. Harma , Y. Firat. Grisel syndrome: nontraumatic atlantoaxial rotatory subluxation. J.Craniofac.Surg. 2008; 19: 1119-21.

3. V. Salpietro, A. Polizzi, F. Granata, et al. Upper respiratory tract infection and torticollis in children: differential diagnosis of Grisel's syndrome. Clin.Neuroradiol. 2012; 22: 351-3.

4. B.R. Subach, M.R. McLaughlin, A.L. Albright et al. Current management of pediatric atlantoaxial rotatory subluxation. Spine (Phila Pa 1976.) 1998; 23: 2174-9.

5. V.J. Fernandez Cornejo, J.F. Martinez-Lage, C. Piqueras et al. Inflammatory atlanto-axial subluxation (Grisel's syndrome) in children: clinical diagnosis and management. Childs Nerv.Syst. 2003; 19: 342-7.

6. D.D Cavalcanti,A. Agrawal, U. Garcia-Gonzalez et al. Anterolateral C1-C2 transarticular fixation for atlantoaxial arthrodesis: landmarks, working area, and angles of approach. Neurosurgery 2010; 67: ons38-ons42.

7. J. Harms, R.P MelcheR. Posterior C1-C2 fusion with polyaxial screw and rod fixation. Spine (Phila Pa 1976.) 2001; 26: 2467-71.

8. R. Desai, C.B Stevenson, A.H. Crawford et al. C-1 lateral mass screw fixation in children with atlantoaxial instability: case series and technical report. J.Spinal Disord.Tech. 2010; 23: 474-9.

9. M.E. Jacobson, S.N. Khan, H.S. An. C1-C2 posterior fixation: indications, technique, and results. Orthop.Clin.North Am. 2012; 43: 11-8, vii.

10. C. Bocciolini, D. Dall'Olio, E. Cunsolo et al. Grisel's syndrome: a rare complication following adenoidectomy. Acta Otorhinolaryngol.Ital. 2005; 25: 245-9.

11. D. Jeszenszky, R. Willms, A. Steigleder et al. Closed reposition of C1/C2 rotatory subluxations, a new method. Presented at the XIV Annual Meeting of the Cervical Spine Research Society. Varese, Italy, September 17–19

12. B. Ni, Z. Zhu, F. Zhou, Q. Guo, J. Yang, J. Liu, F. Wang. Bilateral C1 laminar hooks combined with C2 pedicle screws fixation for treatment of C1-C2 instability not suitable for placement of transarticular screws. Eur Spine J. 2010 Aug;19(8):1378-82.

13. Been HD, Kerkhoffs GM, Maas M. Suspected atlantoaxial rotatory fixation-subluxation: the value of multidetector computed tomography scanning under general anesthesia. Spine (Phila Pa 1976). 2007 Mar 1;32(5):E163-7

14. Kowalski HM, Cohen WA, Cooper P, et al. Pitfalls in the CT diagnosis of atlanto-axial rotatory subluxation. AJR Am J Roentgenol 1987;149:595–600.

15. McGuire KJ, Silber J, Flynn JM, Levine M, Dormans JP. Torticollis in children: can dynamic computed tomography help determine severity and treatment. J Pediatr Orthop. 2002 Nov-Dec;22(6):766-70.

16. Alanay A, Hicazi A, Acaroglu E, Yazici M, Aksoy C, Cila A, Akalan N, Surat A. Reliability and necessity of dynamic computerized tomography in diagnosis of atlantoaxial rotatory subluxation. J Pediatr Orthop. 2002 Nov-Dec;22(6):763-5.

17. M. Yamazak, Y. Someya, M. Aramomi et al. Infection-related atlantoaxial subluxation (Grisel syndrome) in an adult with Down syndrome. Spine (Phila Pa 1976.) 2008; 33: E156-E160.

18. S.C. Lee, T.N. Lui, S.T. Lee. Atlantoaxial rotatory subluxation in skeletally immature patients. Br.J.Neurosurg. 2002; 16: 154-7.

19. E.A. Pereira, J. Hempenstall, P.M. Pretorius, A.A. Kamat, T.A. Cadoux-Hudson. La maladie de Grisel treated by combined C1-2 transarticular and C1 lateral mass screw fixation. Br J Neurosurg. 2010 Aug;24(4):499-501.

20. G.G Heuer, D.A. Hardesty, D.A Bhowmick et al. Treatment of pediatric atlantoaxial instability with traditional and modified Goel-Harms fusion constructs. Eur.Spine J. 2009; 18: 884-92.

21. H.B. Sim, J.W. Lee, J.T. Park et al. Biomechanical evaluations of various c1-c2 posterior fixation techniques. Spine (Phila Pa 1976.) 2011; 36: E401-E407.

Figure legend

Figure 1. Patient´ clinical image with right-sided inclination and left-sided rotation of the head.

Figure 2. a). Radiographic investigations identified right lateral inclination of the head and rectification of the cervical lordosis. b). Open-mouth radiograph showing asymmetry of the odontoid process in relation to the lateral masses of the atlas

Figure 3. Initial coronal and axial CT scan of C1-C2 revealed asymmetrical odontoid-lateral mass relationship (narrow)

Figure 4. a. Five-month coronal and axial CT scan of C1-C2 revealed progression of the subluxation. b. Axial and Coronal 3D-CTin atlanto-axial rotatory subluxation

Figure 5. Patient under cervical traction with the halo-traction system.

Figure 6. Postoperative open-mouth and lateral radiograph of cervical spine, demonstrate proper alignment of cervical spine and placement of atlas laminar hooks and axis pedicle screws.

Figure 7. Sagittal (a) coronal (b) and 3D – CT reconstruction (c) of cervical spine – 36 months after surgery, demonstrate fusion across C1-C2.

Figure 8. Alternative surgical construct combines C1 hooks and C2 pedicle screws united with connecting rods. a) Anteroposterior view b) lateral view c) superior and right rotated view.

Table 1. Literature review. Surgical treatment Grisel´s syndrome

AUTHOR	STUDY DESIGN AND YEAR	# PATIENTS	AGE	PROCEDURE	FOLLOW-UP	COMPLICATIONS	FUSION RATE
Lee	Case series (2002)	6 (5 treated with arthrodesis)	Mean: 9 years (range, 7 to 12)	C1 - C2 sublaminar wirings with graft	18 months	None	No reported
Yamazaki M	Case report (2008)	1	26 years.	Occipitocervical fixation and wires	3 years	None	100%
Desai R	Case series (2010)	1 case secondary inflammation process	9 years	C1 lateral mass screws and C2 transpedicular screw and rods	23 months	None	100%
Pereira	Case report (2010)	1	66 years	C1-C2 transarticular and C1 lateral mass screws with vertical connecting rods	12 months	None	No reported