

Wiring or Screwing at the Craniovertebral Junction in Childhood: Past and Present Personal Experience



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Abstract Background: Craniovertebral junction (CVJ) instrumentation and fusion in childhood are frequently performed with either sublaminar wires or screws in lateral masses, and both are considered quite safe procedures.

Methods: Our experience deals with 12 children: six (mean age 9.5 years) harbouring a congenital instability associated with Down's or Morquio's syndromes and primary os odontoideum; and six (mean age 11.5 years) with acquired iatrogenic instability due to transoral anterior decompression for different reasons (inferior clivectomy, anterior arch removal and odontoidectomy). All patients in the 'congenital group', except for one, had preoperative dynamic x-rays and underwent surgical correction by means of posterior wiring, fusion and an external orthosis. All patients in the 'iatrogenic group' had no preoperative dynamic x-rays and underwent a screwing technique with fusion and an external orthosis.

Results: The postoperative clinical picture had improved in all patients at the latest follow-up (observation range 63–202 months [mean 118.5 months]), with neuroradiological confirmation of satisfactory bony fusion and with neural decompression in all patients.

Conclusion: Although it requires a more accurate preoperative neuroradiological setting, the screwing technique takes less time and is characterized by less blood loss and less postoperative discomfort than the wiring technique. The latter features confirm the simplicity, safety (continuous fluoroscopic assistance is not necessary, and there is no risk of neurovascular injuries) and lower expense (neither complex hardware devices nor neuronavigation systems are required) of the screwing technique.

Keywords Craniocervical junction instability · Sublaminar wiring · Screwing technique · Mucopolysaccharidosis · Down's syndrome · Os odontoideum · Transoral approach

Introduction

Anterior transnasal or transoral decompression—which is used in treatment of irreducible neoplastic, dysembryogenic, inflammatory and chronic traumatic diseases of the anterior craniovertebral junction (CVJ)—has been reported in the literature for many years [1–4].

Surgical treatment of CVJ compression by the transoral or transnasal route is still strongly suggested in cases of irreducible dislocation. Surgical management of CVJ compression aims to achieve neural decompression and to stabilize the CVJ in order to relieve neurological manifestations arising from bulbospinal compression both at rest and during motion, secondary to CVJ instability [5]. *Functional decompression* is a concept in our therapeutic strategy, aiming to achieve neural decompression by performing simple reduction, instrumentation and fusion of the CVJ dislocation when it is reducible [6]. In cases in which accurate preoperative x-ray examinations demonstrate CVJ irreducibility and associated neural compression, the goal of surgery is to maintain anatomical alignment while preserving the motion of normal adjacent elements, with the aim of protecting the neural elements [7, 8]. In this paper we present an update of our personal experience of instrumentation and fusion in children, using titanium rods, sublaminar wires and screws [6].

Materials and Methods

From 1998 to 2018, 12 children were operated on in the Section of Paediatric Neurosurgery at Policlinic Gemelli, Catholic University School of Medicine, in Rome. Six female patients aged 6–14 years (mean age 9.5 years) were treated for os odontoideum (group 1). Five of these patients

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were affected by Down's syndrome, one had a metabolic disease (mucopolysaccharidosis type IV, i.e. Morquio's syndrome) and one had an isolated os odontoideum. A second group (group 2), consisting of six male patients aged 8–15 years (mean age 11.5 years), underwent transoral anterior decompression and staged posterior instrumentation and fusion with screws for different diseases. One of these patients had *impressio basilaris*, one had basilar invagination, two had os odontoideum, one had a C0–C1 developmental anomaly and one had a C2 fracture and dislocation. All patients underwent computed tomography (CT) scans and static and dynamic magnetic resonance imaging (MRI) of the brain and CVJ. Further preoperative static and dynamic x-rays were performed in patients in group 1, and CVJ instability was shown by atlantoaxial displacement greater than 4.5 mm in all but one patient (Table 1) [9]. One group 1 patient (patient #3) with a hyperintense signal at the medulla and at the bulbospinal junction had gait disturbances and dyspnoea, which led to an emergency tracheostomy [10]. In group 1, the CVJ shift was reducible in five patients and irreducible in one patient (patient #4). Preoperative fixation was accomplished by use of a hard collar.

Surgical Techniques: Posterior Instrumentation and Fusion

Group 1

Patients were placed in a prone position. Intraoperative traction and reduction of the C2 shift were obtained using a Mayfield headholder under fluoroscopic control. After preparation of the occiput and the cervical spine, occipitocervical instrumentation was carried out. Two burr holes into the occiput, 3 cm cranially to the rim of the foramen magnum, represented the proximal point for passing titanium wires. To facilitate the passage of the wires, notching (with Kerrison rongeurs) of the rim of the foramen magnum and of the cervical laminae to be fused, and removal of the atlanto-occipital membrane and ligamentum flavum, were carried out. In patients with C0–C1 assimilation (patients #1, #2 and #3), C1 laminectomy was performed. A wide-diameter (5 mm) non-threaded titanium rod was bent into a U shape, cut in a way that the ends extended a few millimetres beyond the fused segments to prevent them from slipping out during flexion and extension movements of the neck, and to adapt them to the bony contours of the CVJ. The assimilated and bifid posterior arch of the atlas was excised during posterior decompression of the posterior foramen magnum margin prior to passage of sublaminar wires. With use of the Sonntag method, Songer titanium wires were passed under the involved bone

segments and over the titanium rod and bone graft, being stretched up to approximately 10 pounds (Table 1).

Group 2

After transoral decompression a second staged procedure was performed. Under fluoroscopy, C0–C2–C3 screws 3.5 mm in diameter and 12 mm in length (Vertex System [Medtronic, Minneapolis, MN, USA]; Summit SI OCT Spinal Fixation System and Mountaineer OCT Spinal System [DePuy Synthes Spine, Warsaw, IN, USA]; and VuePoint OCT [NuVasive, San Diego, CA, USA]) were inserted in the C2 isthmus bilaterally in the centre of the lateral masses (taking care to spare the vertebral notch) and in the C3 lateral masses from medial to lateral and from caudal to cranial.

Both Groups

Bone fusion was performed by decortication of the occiput and the posterior arches of the cervical facet joints by a high-speed drill and curettes to facilitate bone fusion.

Autologous bone was harvested from the right posterior iliac crest, cut in a double-wing shape and fixed over the construct, using a silk suture along with antigen-free synthetic bone graft substitute fusion (beta-tricalcium phosphate [Vitoss Synthetic Cancellous Bone Void Filler; Stryker, Kalamazoo, MI, USA]). Moreover, cancellous bone was placed upon the levels to be fused when available after further posterior CVJ decompression.

Postoperative Care (Table 1)

Group 1

After completion of the surgical treatment, a halo or SOMI vest was utilized for 4 months in all patients except for patient #3, for whom it was necessary to prolong the application of the external orthosis.

Group 2

After completion of the surgical treatment, a halo or SOMI vest was utilized for no more than 3 months in all patients.

Bone fusion was evaluated on CVJ radiological studies and bone window CT scan examinations. Radiological and CT scans plus MRI and neurological examinations were performed 1 week after surgery, then every 4 months up to 1 year and finally at the last follow-up [11]. The Frankel scale and the Di Lorenzo disability grade were used to evaluate the neurological condition.

Table 36.1 Patients with congenital (1–6, Group 1) and iatrogenic (7–12, Group 2) instability

Pt initials Case N°.	Age (sex)	Primary disease	Radiology	Pre-op C1/C2 shift (X-rays)	Treatment	Post-op shift (X-rays)	Frankel scale and		
							Di	Lorenzo Grade changes	External orthosis (months)
CV 1	8 (F)	MPS (Morquio)	- C0-C1 assimilation - C1-C2 instability - C2-C3 partial fusion - Os odontoides reducible	>4.5 mm	- Suboccipital craniectomy and C1 laminectomy - C0-C3 reduction, instrumentation and fusion	No	E/E I/I	SOMI (4 months)	202
BS 2	6 (F)	Down syndrome	- C0-C1 assimilation - C1-C2 instability - Os odontoides, reducible	5 mm	- Suboccipital craniectomy and C1 laminectomy - C0-C3 reduction, instrumentation and fusion	No	E/E I/I	Halovest (1 month) SOMI (3 months)	175
TP 3	8 (F)	Down syndrome	- C0-C1 assimilation - C1-C2 instability - Os odontoides, reducible	10 mm	- Suboccipital craniectomy and C1 laminectomy - C0-C3 reduction, instrumentation and fusion	No	D/E III/I	SOMI (4 months) Halovest (4 months)	172
EAK 4	(7) (F)	Down syndrome	- C1 bifid - C1-C2 instability, - Os odontoides irreducible	>8 mm	- C0-C3 reduction, instrumentation and fusion;	No	E/E I/I	SOMI (4 months)	143
FI 5	14 (F)	Down syndrome	- C1-C2 instability - Os odontoides reducible	>4.5 mm	- C0-C3 reduction, instrumentation and fusion	No	E/E I/I	Halovest (4 months)	130
RRS 6	14 (F)	Down syndrome	- C1-C2 instability, - Os odontoides reducible	>4.5 mm	- C0-C3 reduction, instrumentation and fusion	No	E/E I/I	Halovest (4 months)	124
CL 7	15 (M)	Developmental anomaly C0-C1	- C0-C1 anterior compression - C1-C2 instability	>5 mm	1. Transoral decompression 2. C1 laminectomy, C0 double vertical screws, C2 pedicles and C3 lateral masses screws instrument.	No	D/E III/I	Soft collar (1 month)	74
EA 8	11 (M)	Impressio basilaris Os odontoides (Down s.)	C1-C2 anterior compression	Virtual, previously documented	Transoral C1 odontoidectomy and clivectomy in C0-C2-C3 screwing instrumentation and heterologous bone fusion (previously implanted)	No	D/E II/I	Soft collar (1 month)	70
RR 9	14 (M)	C2 fracture and dislocation	C2 fracture and C1-C2 dislocation with cervico medullary contusion	>7 mm	1. Transoral C1-C2 decompression 2. C0-C3 C5 screwing instrumentation and heterologous bone fusion	No	D/E II/I	Soft collar (1 month)	67
MP 10	8 (M)	Impressio basilaris	C0-C1-C2 compression	No	1. Transoral C1-C2 decompression 2. Staged C0-C2-C3 screwing instrumentation and fusion	No	D/E II/I	Halo + Philadelphia (2 months)	63
SS 11	9 (M)	Os odontoides	C2 compression	Virtual	1. Transoral C1-C2 decompression 2. C0-C2-C3 screwing instrumentation and fusion	No	D/E II/I	Halo + Philadelphia (2 months)	118
AE 12	12 (M)	Basilar invagination	C2 compression	Virtual	1. Transoral C0-C1-C2 decompression 2. C0-C2-C3 screwing instrumentation and fusion	No	D/E II/I	Halo + Philadelphia (3 months)	84

Results

The follow-up period ranged from 63 to 202 months (mean 118.5 months). All patients improved soon after surgery independently of the type of surgery they underwent, but an immediate clinical improvement in gait disturbance occurred in patient #3 in group 1, who had Down's syndrome; her Frankel grade changed from D to E and her Di Lorenzo grade changed from III to I. In this patient the improvement in respiratory dysfunction allowed closure of the tracheostomy 24 months after surgery. Nuchal pain disappeared in all of the children postoperatively.

No arterial injuries, bleeding, haematomas or systemic complications occurred. At the site of bone harvesting, no infections, cosmetic problems, pain or complaints were reported.

Blood loss ranged from 17 to 23 mL (mean 20 mL) in group 1 and from 11 to 19 mL (mean 15 mL) in group 2.

Concerning the duration of the posterior approach procedures, it ranged from 3.2 to 4.0 h for wiring and from 2.3 to 3 h for screwing.

Diagnostic imaging, immediately after surgery, showed restoration of bone alignment with decompression of the brainstem in all patients. Neuroradiological signs of bone fusion were already evident 4 months after surgery in all but two patients.

Bone Fusion

Group 1

In patient #3, failure of bone fusion occurred 9 months after surgery, as a consequence of a cerebrospinal fluid (CSF) fistula and wound infection; further bone grafting from the iliac crest was successful after resolution of the CSF leakage. A significant reduction in the cervicomedullary junction contusion was evident at late follow-up.

Group 2

In patient #10, the hardware was revised and the synthetic bone graft substitute was removed 2 weeks after the staged instrumentation and fusion procedure, because of infective dehiscence of the surgical wound. After 2 months of polyanthibiotic therapy (intravenous daptomycin 350 mg/day and oral rifampicin 600 mg/day), and 1 month after collar removal, a dynamic cervical MRI examination confirmed CVJ stability.

In both groups, despite the cranial fixation, no limitations in social life due to impaired head motion were observed in any patient.

Discussion

Sublaminar Instrumented Wiring Versus Lateral Mass Screw Implants

Wiring Technique

Sublaminar instrumented wiring remains an excellent and simple procedure for stabilizing the CVJ and upper cervical spine, resulting in a reasonably good mechanical outcome with a low incidence of complications [22]. The stiffness provided by the wiring, determined by the number of vertebrae enclosed by the instrumentation and augmented with external immobilization, is associated with bone fusion in 100% of cases [6, 22, 23]. This observation may help to overcome the early biomechanical drawbacks of sublaminar instrumented wiring with respect to lateral mass screw implants.

Screwing Technique

After early reports on small series of patients treated with this approach, several clinical studies reported that the results obtained with use of lateral mass screw implants were better than those obtained with sublaminar instrumented wiring [12–18]. However, complications reported at the very beginning of the experience (such as 30% of screws pulling out in the suboccipital area and a mortality rate of up to 9% after complex spine decompression and fixation) discouraged some paediatric spine surgeons from using lateral mass screw implants [14, 19]. Lateral mass screw implants in a paediatric population achieved bone fusion in 100% of cases, with a 10.4% complication rate, including vertebral artery injuries [7, 9, 20, 21].

Wiring Versus Screwing

More recently published experiences have seemed to report the same 100% incidence of fusion with both lateral mass screw implants and sublaminar instrumented wiring [6, 8, 14, 22, 23]. Despite a clear advantage of the screwing technique in terms of blood loss, surgery duration and postoperative immobilization, the infectious complication rate appears comparable.

Complications in Our Series

Wiring Technique

The difficulties encountered in patient #3, who had Down's syndrome, were ascribed to the patient's immunocompromised state (impaired monocyte and neutrophil chemotaxis, decreased phagocytosis and qualitative T-lymphocyte defi-

ciency), which may have predisposed her to respiratory infections and postoperative complications. In similar cases, the rate of bone fusion may also be lower than that in other patients, probably as a result of deficient collagen synthesis, which contributes to bone graft pseudoarthrosis [24]. In accordance with the literature, to prevent frequent neck movements in the postoperative period—especially in children with delayed mental milestones and those with spasticity—halo immobilization was instituted early and until there was objective evidence of bony fusion, which could take as long as 6 months [5, 25].

Screwing Technique

We can hypothesize that a long-lasting infection, which occurred in one of our patients, played a role in the ossification process implied in CVJ fusion, since the ossification occurred 33 months after the onset of the infection. Finally (and very surprisingly), in our case the postinfective bone fusion not only produced good fixation but also resulted in a kind of odontoid regeneration that has not been reported so far. In fact, although our group has recently published a description of ‘true’ odontoid process regeneration (along with clival regeneration and recurrence of Chiari malformation) after transoral decompression, in the present case we observed union of the remaining C2 and C3 bodies, strongly mimicking concomitant, rather complete axis and clival regeneration [26, 27].

Conclusion

The wiring technique is simple, safe (continuous fluoroscopic assistance is not necessary and there is no risk of neurovascular injuries) and less expensive than the screwing technique, as no complex hardware devices and no navigation systems are required.

The screwing technique requires a more accurate preoperative neuroradiological setting than the wiring technique but seems faster and is characterized by less blood loss and less postoperative discomfort.

Caution is needed to avoid postoperative complications (namely, a cerebrospinal fluid fistula) that might lead to secondary infections, bone graft pseudoarthrosis or external contamination.

Competing Interests The authors declare that they have no competing interests.

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