Cervical fixation in the pediatric patient: our experience

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Abstract The surgical management of cervical instability in children is a challenging issue. Although the indications for internal fixation are similar to those for adults, accurate pre-surgery study and sharp surgical techniques are necessary because of the size of such patients' anatomy, their peculiar tissue biology and the wide spectrum of diseases requiring cervical fusion. Our case study is made up of 31 patients, 15 male and 16 female, with an average age of 7 years and 6 months (2 years and 6 months to 18 years) who underwent cervical fusion for instability. Their physical condition presented various different pathologies ranging from congenital deformity, systemic skeletal disease, tumors, trauma, post-surgery instability. We performed occipito-cervical fusion in 11 cases, 5 of which involved stabilization at the cranium–vertebral junction. We used instrumentation in 13 cases (3 sublaminae wiring, 10 rigid adult instrumentation). We used rigid adult instrumentation in three patients under 10 years of age, treated by rod, occipital screws and sublaminae hook instrumentation in steel C0–C2 (9-year-old male, affected by os odontoideum in Down's syndrome; male of 7 years and 10 months, affected by os odontoideum in Down's syndrome; female of 4 years and 6 months with occipito-

cervical stenosis and C0–C2 instability in Hurler's syndrome). We operated on two patients under 3 years of age, using sublaminae wiring with bone precursors and allograft at level C0–C2 (one of these was a 30-month-old male with post-traumatic instability C0–C2, while the other was a 17-month-old male with C0–C2 instability in Larsen's syndrome). The average follow-up age was 7 years and 1 month (between 1 and 18 years). Cervical fusion was assessed by X-ray examinations at 4th and 12th weeks and at 6th and 12th months after surgery. Where implants could allow, RMN examination was performed at 1st month after surgery. In the other cases, in which implants do not allow RMN to be performed, CT scan and standard X-rays were carried out, and new X-rays were performed every other year. We experienced two cases of sublaminae wiring rupture without impairment of bone fusion. No patient suffered major complications (infection and osteomyelitis, rigid instrumentation mobilization, incomplete fusion with instability, neurologic impairment, insufficient cervical spine range of movement to cope with everyday life activities, cervical pain). Even though most authors still indicate that rigid instrumentation should be performed in cases over 10 years of age and sublaminae wiring in cases over 3 years of age, our findings demonstrate that this age limit can be lowered. We have treated children under 10 years of age by rigid adult instrumentation and under 36 months of age by wiring. The anatomic size of the patient is the most important factor in determining the use of instrument arthrodesis to treat pediatric cervical spine instability. Although not easy, it is possible and preferable in many cases to adapt fixation to child cervical spine even in very young patients.

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Introduction

Cervical fusion in pediatric patients is a difficult problem. While the indications for internal fixation in children are similar to those of adults, the data concerning techniques, complications and outcomes of spinal instrumentation are based on the findings and practice performed on adult patients. Diminutive osseous and ligamentous structures and anatomical variations associated with syndromic cervical abnormalities frequently complicate the approach and limit the use of internal fixation. Cervical arthrodesis in the children could limit growth potential and cause secondary deformity.

Poor healing potential in at-the-risk groups, such as patients with Down syndrome and os odontoideum or atlanto-axial instability in Larsen syndrome, makes fusion difficult [1, 2].

A wide spectrum of abnormalities is encountered in the pediatric patients which affects the craniocervical junction and the upper cervical spine. The initial surgical treatment of these problems is mainly by posterior route. The advances in neurodiagnostic imaging and in surgical instrumentation have been achieved through the improved understanding of the complex anatomy of the site, the dynamics of the diseases and the biomechanics of the cervical spine. This has opened new vistas and placed the entire circumference of the cervical spine and craniocervical junction within the spine surgeon's reach.

The indications to perform a cervical fusion in children comprise congenital abnormalities and acquired lesions [1, 3]. These can be divided into:

Morphologic abnormalities:

- absence of occipital condyles
- unilateral atlas assimilation
- absence of odontoid process
- dystrophic os odontoideum
- absence of posterior arch of C1
- cervical emispondylus

Congenital diseases:

- Morquio's syndrome
- Goldenhar's syndrome
- Conradi's syndrome
- Spondyloepiphyseal dysplasia
- Grisel's syndrome
- Larsen's syndrome
- Hurler's syndrome

Tumors

Post-infectious instability

Post-surgery instability

Trauma

These indications include acute conditions, where the pathology is at a critical stage and intervention is essential (fractures, dislocations, tumors), as well as progressive diseases, where the evolution of instability is unpredictable [2, 4–6].

All treatments aim to solve the pathology and obtain cervical spine stabilization. In tumors, the first goal of the treatment is the radical excision, with the spine stabilization. Cervical tumors are less frequent in pediatric patients than in the adult population.

Cervical arthrodesis must obtain spine stabilization in order to prevent and avoid neurologic lesions.

Materials and methods

We treated 31 patients, 15 males and 16 females, from 2 years and 8 months to 18 years of age (the average age was 7 years and 6 months).

All these patients were operated on by the same surgical team in the same institution, and arthrodesis was performed by the posterior route. All patients were placed in prone position with neck in flexion. No patient needed subsequent anterior route cervical procedures. In the cases in question, only two patients, affected by instability in Neurofibromatosis, were treated by combined anterior and posterior route surgery, following the original surgical planning. One patient, treated by simple arthrodesis for complex congenital malformation, presented partial loss of correction at the follow-up stage.

The pathology included:

- seven cases of Larsen's syndrome
- two cases of Neurofibromatosis
- four cases of dystrophic os odontoideum in Down's syndrome
- seven cases of spondyloepiphyseal dysplasia
- three cases of osteoblastoma at level 2 at C2 and 1 at C3
- one case of carcinoma at C3
- two cases of post-traumatic instability: C0–C2 and C5–C6
- one case of instability after laminectomy at C4
- one case of C0–C2 instability in Hurler's syndrome
- three cases of cervical fracture: one C3 and two C5 fracture

Eleven patients were treated by craniovertebral junction arthrodesis at these levels:

- five C0–C2 (two dystrophic os odontoideum in Down's syndrome, one post-traumatic instability C0–C2, one instability in Larsen's syndrome, one occipito-cervical stenosis and C0–C2 instability in Hurler's syndrome); two were instrumented

- three C0–C3 (one osteoblastoma C1, one spondyloepiphyseal dysplasia, one Larsen's syndrome)
- two C0–C4 (one instability after previous laminectomy, one cervical fracture C3), both instrumented
- one C0–C5, after C3 Carcinoma removal, instrumented

We performed instrumented arthrodesis in 13 cases, including 5 occipito-cervical instrumented fixations. In three patients, we performed contoured loop fixation. We used rigid adult instrumentation in the remaining ten cases, with rod, occipital screws and a laminar hook. In one case, we used a steel implant, in the others titanium instrumentation. These cases include:

Rigid adult instrumentation

- one occipito-cervical stenosis and C0–C2 instability in Hurler's syndrome
- two dystrophic os odontoideum in Down's syndrome C0–C2
- one instability in Larsen's syndrome C0–C2
- one post-traumatic instability C5–C6
- one instability after previous laminectomy C4
- one C0–C5, after carcinoma removal
- two C4–C6 in C5 fractures
- one C2–C4 in C3 fracture

Contoured loop fixation

- two post-traumatic instability C0–C2
- one C0–C2 instability in Larsen's syndrome

In the cases treated by instrumented fixation, we used rigid adult instrumentation in three patients under 10 years of age, treated by rod, occipital screws and sublaminar hook instrumentation in steel and such cases include:

- 9-year-old male, affected by os odontoideum in Down's syndrome, fixation C0–C2
- 7-year and 10-month-old male, affected by os odontoideum in Down's syndrome, fixation C0–C2
- 4-year and 6-month-old male with occipito-cervical stenosis and C0–C2 instability in Hurler's syndrome: in this case, we implanted occipito-cervical instrumentation C0–C4 (Fig. 1)

We operated on two patients under 36 months of age, using sublaminar wiring with bone precursors and allograft at C0–C2 level, whose pathologies include:

- 30-month-old male with post-traumatic instability C0–C2
- 17-month-old male with C0–C2 instability in Larsen's syndrome (Fig. 2)

We strongly recommend the use of instrumented arthrodesis in children where necessary even if most authors consider patients too young to sustain this procedure. In

literature, age limits are generally considered to be 10 years for rigid arthrodesis and 36 months for wiring.

In the other 18 cases, we performed simple arthrodesis using posterior iliac spine bone graft, in a few cases implemented by allograft.

In three cases under 5 years of age, we used bone precursor and allograft.

The average blood loss during procedure was 300 cc.

Four patients with tumors were treated by tumor removal and stabilized by posterior route. No tumor recurrence was observed during follow-up stage.

Before the procedure, all patients were examined by standard X-rays of cervical spine in antero-posterior, lateral and transverse projection. The cases treated by craniovertebral arthrodesis were studied before surgery by CT scan or 3D CT scan reconstruction of cervical spine and occipital region and RMN of medulla and brain.

The post-surgery treatment of these patients involved the cervical immobilization for an average of 12 weeks (10–24 weeks) in rigid Philadelphia/Schantz collar or in Minerva brace for cases treated by instrumentation; cervical immobilization for an average of 12 weeks (11–32 weeks) in a pro-Minerva cast followed by 8 weeks in Schantz collar was adopted for cases treated by simple arthrodesis. At the end of this period, the patient was free to move his/her neck if the imaging demonstrated that the fusion was successful.

The follow-up was 1–18 years (the average follow-up was 7 years and 1 month).

We examined patients in the 4th and 12th weeks and 6 months after surgery performing standard X-rays to study fusion. Where implanted instrumentation was possible, we performed a RMN examination 1 month after surgery. CT scan and standard X-rays were performed 1 year after surgery, and new X-rays every second year.

Results

All our patients, except one, healed without major complications. One patient, operated on for complex congenital malformation, treated by occipito-cervical arthrodesis without instrumentation, and adding a bone precursor, presented a subsequent loss of correction.

We consider the following as major complications:

- Infection and osteomyelitis
- Rigid instrumentation mobilization
- Incomplete fusion with instability
- Loss of correction
- Neurologic impairment
- Insufficient cervical spine range of movement to cope with everyday life activities
- Cervical pain

Fig. 1 **a–c** VI, 4-year and 6-month-old male, instability and stenosis in Hurler's syndrome. **d, e** Decompression, fixation arthrodesis by adult occipito-cervical rigid instrumentation C0–C4. **e, f** 11 months after intervention. **g, h** 18 months after intervention. **i** 30 months after surgery

Fig. 2 **a–c** BM, 17-month-old male C0–C2 instability in Larsen's syndrome. **d–f** Arthrodesis fixation by sublaminar wiring with bone precursors and allograft at level C0–C2. **g, h** 12 months after surgery,

wiring rupture. **j, k** Dynamic rx, no instability. Complete fusion, without symptoms. Wiring is not removed. **l, m** 3 years after intervention, **n–p** 4 years and 4 months after intervention

Fig. 2 continued

The X-ray examinations and RMN showed complete fusion 1 year after surgery.

At that time, patients no longer suffered pain or had neurologic symptoms. Cervical spine range of movements was wide enough for the patient to perform everyday life activities without impairment. Patients operated by occipito-cervical arthrodesis showed, of course, a limitation in the craniovertebral junction mobility, but this was sufficiently compensated by mobility in lower cervical levels.

Patients operated for dysplasia or upper cervical tumor removal, that at the presentation showed a considerable lateral imbalance of the spine, demonstrated a good biological compensation of deformity in the follow-up analysis. Cervical spine deviation was reduced to an acceptable esthetic level, while the range of the movement maintained a good functional level, without pain.

All craniovertebral junction arthrodesis cases resulted in complete fusion, except for one.

Two patients experienced sublamina wire rupture: one 12 months after surgery, the other 24 months after surgery. No arthrodesis failure was observed, and the patients did not have neurologic symptoms, pain, or cervical movements' impairment. The broken wiring was not removed [7].

Rigid adult instrumentation was used in ten cases, all of which demonstrated complete fusion 1 year after the procedure. No rupture or mobilization of instrumentation was observed during the follow-up stage and all ten patients had good cervical function without pain or neurologic impairment.

In one case, the patient, treated by occipito-cervical instrumentation, suffered occipital bursitis, which was treated by anti-inflammatory drugs until remission.

In most authors' opinion in literature, rigid adult instrumentation should be used in children from 10 years of age, while patients over the age of three can be treated by sublamina wiring [8–11]. In our series, however, we

have implanted instrumentation in significantly younger patients, without major complications and always obtaining stable bone fusion.

We used rigid adult instrumentation in three patients under 10 years of age, treated by rod, occipital screws and sublaminar hook instrumentation which include:

- 9-year-old male, affected by os odontoideum in Down's syndrome
- 7-year and 10-month-old male, affected by os odontoideum in Down's syndrome
- 4-year and 6-month-old male, affected instability and stenosis in Hurler's syndrome

We operated two patients under 36 months of age, using sublaminar wiring with bone precursors and allograft at level C0–C2 which include:

- 30-month-old male with post-traumatic instability C0–C2
- 17-month-old male with C0–C2 instability in Larsen's syndrome.

Our experience demonstrates that even in the case of a patient under previously established age limits instrumentation use is safe and effective, where the patient's anatomy is wide enough to be compatible with minimum dimension of implanted devices.

Instrumented arthrodesis is by far more stable than simple fusion, while in our follow-up we never had an implant failure.

Discussion

Surgical management of pediatric cervical spine instability is a challenging issue. A wide range of diseases could require a surgical arthrodesis in children's cervical spine: as the basic procedure is not very different from adult surgery, the size of the anatomy and the peculiar biology of pediatric spine require an accurate study of the patient and a sharp surgical technique.

While high osteointegration capability of a child's bone tissue allows for stable fusion after simple arthrodesis, many cervical instability cases are associated with conditions (i.e., Down's syndrome and os odontoideum) where poor healing potential makes fixation difficult.

Instrumentation can obtain more stable fixation, but greatly enhances the risk of complications; moreover, in literature its use is limited by the age of the patient and the size of his/her cervical spine. In most authors' opinion, rigid adult instrumentation can be used in children from 10 years of age while patients over 3 years of age can be treated by sublaminar wiring [8–11]. In our series, we used rigid adult instrumentation in three cases under 10 years of

age, and sublaminar wiring in two cases under 36 months of age. In one case, we stabilized by wiring at C0–C2 level in a 17-month-old boy affected by Larsen's syndrome. All these patients healed without major complications, bone fusion was completed within 1 year and cervical functions were completely satisfactory. Our results show that the limit in instrumentation use in a pediatric patient is determined by the accuracy of the surgical technique and by the anatomic size of the patient, and not by his/her age.

Peduncular screws are by far more stable than sublaminar wiring and hooks, but their use in the cervical spine is limited due to high risk of damaging vertebral artery, as minimum size screws in adult instrumentation are often still too big to avoid impingement in the vertebral canal, even in older children (over 10 years of age) [8, 12–14]. In younger children, the situation is even more demanding, as the size of vertebrae is minimal.

Moreover, the cartilaginous tissue in a child's cervical spine requires the screw insertion to be highly accurate, particularly in younger children. Any kind of mistake in placing screws leaves very little margin for correction, as secondary mobilization of implanted devices could lead to instrumentation failure, often requiring revision. The expertise and experience of the surgeon are the most important factors, as surgery in this area of the pediatric spine is difficult and the learning curve is rather long.

Surgical planning must be based on an accurate study of the anatomy, combining information from plain X-rays, CT scan and RMN. Angiography or angio-CT scan are useful in assessing the course and variation of vertebral artery, particularly in the case of severe vertebral deformity, where the vessels course could be altered. In the past 5 years in our institution, we have been systematically using 3D CT scan reconstruction of cervical spine to build a "real-life" bone anatomy model of the patients' cervical spine. This technique, combined with angiography or angio-CT, has a great potential in avoiding dangerous areas placing instrumentation [15].

Sublaminar wiring, although not as stable as rigid instrumentation, is much easier to place and maybe less invasive. Wiring has the advantage of being used in diminutive anatomy patients, where even the smallest adult instrumentation is too invasive. In our experience, two patients treated by sublaminar wiring experienced wire rupture 1 and 2 years after surgery. Fusion was complete in both cases and the rupture had no residual consequences [9–11].

Bone graft could pose others problems, as cervical arthrodesis requires bone stock addition to be completed. We have used posterior pelvis access to obtain autograft from posterior iliac spine, and, in recent cases, we used bone precursor with allograft. Both procedures were well

tolerated by children patients, and pelvis incision healed without any case of infection or residual pain.

In patients under 36 months of age, we have used bone promoter with local autograft. The results were very good, with complete fusion within 1 year from surgery [16].

We treated four cases of local aggressive tumors that required complete excision before stabilization. Tumor treatment aimed at completing the removal of neoplastic tissue as the first goal of surgery. Secondary instability must be treated by stabilization, and technique is not different from the other osseous diseases requiring similar surgery.

A major potential complication peculiar in pediatric cervical spine arthrodesis is the fusion proceeding to levels different from surgical program. This side effect is greatly determined by an invasive surgical technique, with the formation of subperiosteal hematomas and subsequent ossification, and it is more common in simple arthrodesis. This situation could cause impairment of cervical mobility over time.

It is not easy even for experienced surgeons to limit subperiosteal blood loss and avoid fusion spread over planned area. We have observed this effect in several patients, but in no case did the fusion determine a severe limitation in neck mobility, as free levels in the cervical spine were able to balance the impairment of movements in fusion levels.

Conclusion

Cervical fusions in pediatric patients often have the same problems of arthrodesis at other vertebral levels. Forecasts based on the potential growth of cervical spine are always difficult, and surgeons should take potential complications into consideration when choosing the most appropriate treatment to adopt.

The use of instrumentation and its choice is the main issue. While instrumented arthrodesis is more stable, it is commonly considered that children's cervical spine shows reduced tolerance for implants [15–17].

The absence of dedicated pediatric instrumentation means that it is necessary to use adult devices, which must be adapted to the child's cervical spine. Instrumented fusion of cervical spine with pedicle screws has several advantages compared with sublaminar wiring, such as stronger fixation and reduction of malalignment [15].

Even if many authors sustain the use of rigid instrumentation only in patients over 10 years of age and the use of sublaminar wiring, we have successfully operated three patients under 10 years using rigid fixation and two patients under 36 months using sublaminar wiring. All these cases healed with complete cervical function, without

residual instability or neurologic impairment. In our opinion, instrumentation should be performed in each patient whose anatomic size is compatible with minimum size of devices. In patients where cervical structures are sufficiently developed to tolerate screws and hooks, rigid instrumentation is preferable, as it grants a more stable fusion [18]. The risks of a more invasive implant are balanced out by potential results, since we never experienced major complications in the cases that were handled. In patients with cervical spine which is too small to cope with rigid instrumentation, stabilization can be obtained by sublaminar wiring. Even if wiring is less stable than rigid implant, it is less invasive, and still ensures a good fixation and a complete bone fusion. We achieved excellent results using bone precursors and allograft in arthrodesis in patients under 36 months.

While the surgeon's expertise and experience are the most important factors, a complete study of patient before surgery, the choice of the fusion level, an accurate and sharp surgical technique and prolonged and scheduled post-operation care are also paramount features in making such procedures successful.

Conflict of interest statement None of the authors has any potential conflict of interest.

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