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ORIGINAL PAPER

Instrumented arthrodesis for non-traumatic craniocervical instability in very young children

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Abstract

Purpose Occipitocervical instrumentation is infrequently required for stabilization of the axial and subaxial cervical spine in very young children. However, when it is necessary, unique surgical considerations arise in children when compared with similar procedures in adults.

Methods The authors reviewed literature describing fusion of the occipitocervical junction (OCJ) in toddlers and share their experience with eight cases of young children (age less than or equal to 4 years) receiving occiput to axial or subaxial spine instrumentation and fixation. Diagnoses and indications included severe or secondary Chiari malformation, skeletal dysplastic syndromes, Klippel-Feil syndrome, Pierre Robin syndrome, Gordon syndrome, hemivertebra and atlantal occipitalization, basilar impression, and iatrogenic causes.

Results All patients underwent occipital bone to cervical spine instrumentation and fixation at different levels. Constructs extended from the occiput to C2 and T1 utilizing various permutations of titanium rods, autologous rib autografts, Mersilene sutures, and combinations of autografts with bone matrix materials. All patients were placed in rigid cervical bracing or halo fixation postoperatively. No postoperative neurological deficits or intraoperative vascular injuries occurred.

Conclusion Instrumented arthrodesis can be a treatment option in very young children to address the non-traumatic craniocervical instability while reducing the need for prolonged external halo vest immobilization. Factors affecting fusion are addressed with respect to preoperative, intraoperative, and postoperative decision-making that may be unique to the toddler population.

Keywords Occipital bone · Cervical spine · Occipitocervical instability · Instrumentation · Occipitocervical fusion

Introduction

Occipitocervical junction (OCJ) instrumentation and fixation (OCF) are rarely indicated in the pediatric population; however, instability at the cervicomedullary junction may necessitate an OCF. Craniocervical instability (CCI) in this population mainly results from a congenital or traumatic pathology, i.e., mechanical instability from atlantoaxial rotatory subluxation, significant neck trauma, os odontoideum, infection, juvenile rheumatoid arthritis, craniofacial syndromes, mucopolysaccharidosis, spondyloepiphyseal dysplasia, "secondary" Chiari malformation syndromes (progressive atlantoaxial instability or unstable basilar invagination with crowded posterior cranial fossa), iatrogenic causes (previous suboccipital decompressions, anterior decompressive approaches, i.e., postendoscopic transnasal/transoral approaches to odontoid), or cervical decompression, and other less common entities [1–9, 34]. Although pediatric trauma accounts for only 1–4% of overall spinal trauma, and remains the common reason for occipitocervical instrumentation is not the main focus of this study. [8, 10, 11].

Typically, trauma localized to the axial spine or subaxial spine is more common in younger children due to unique

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anatomical differences that include disproportionately large head size compared with their cervical spine, ligamentous laxity, facet orientation, and less well developed uncinated processes. When the need arises for spinal fixation, the immature spinal anatomy of very young children is challenging. Fusion techniques need adequately address the instability, while remaining cognizant of the ongoing development growth plates of the vertebrae and ossification zones are often still evolving with cartilaginous interpositions [12]. Most of these young patients exhibit good immediate and late OCJ alignment with appropriate growth, but a small fraction of these children may suffer from diminished vertical growth across fused levels or a reduction in the height of the fusion mass. They may develop hyperlordosis due to asymmetric growth between anterior and posterior spinal column [13].

The surgical management of the OCJ instability in children is unique due to the characteristics of immature bone and ligamentous structures and biomechanical considerations needed to avert early treatment failures. Historically, fusion failures yielded by onlay techniques with laminar wiring and external immobilization resulted in poor arthrodesis. Gradually, these techniques were replaced by the development of occipitocervical internal fixation techniques. These strategies, adapted from adult populations, have improved pediatric fusion rates and have decreased the need for prolonged external arthrodesis. With the rarity of these cases, very few large series have rigorously examined the safety and long-term results of these methods in a very young pediatric population.

The purpose of this paper is to delineate an approach for toddlers with craniocervical instability and discuss our insight. As a framework for this discussion, we will report our experience with our eight cases and address lessons we have learned from these patients in relation to the sparse literature available.

Methods

We performed a PubMed literature search using the keywords "pediatric cervical spine fusion," "pediatric craniocervical," and "pediatric occipitocervical fusion." We reviewed all articles from peer-reviewed journals that reported an OCJ instrumentation in very young children (age less than or equal to 4 years). Fusion rates were assessed by radiological evaluation (AP and lateral cervical spine radiographs) with at least 12 months of follow-up and procedure-associated complications. Due to inherent variability in surgeons self-defining a successful fusion and the absence of CT imaging in certain studies, we included various definitions of presumed osseous fusion, including (1) fusion on AP and lateral radiographic views, (2) stability on flexion extension radiographs, and/or (3) CT scan-based evidence of a successful fusion. Exclusion criteria were (1) children older than 4 years of age at the time of surgery, (2) non-English published articles, (3) non-instrumented occipitocervical fusions, and (4) instrumentation and fixation below the cervical spine and first thoracic segments. All references were manually cross-referenced.

We retrospectively reviewed our patients at two pediatric medical centers (Weill Cornell Medicine, New York Presbyterian Hospital, New York, NY, and Shriners Hospitals for Children, Philadelphia, PA) between January 2008 and October 2017. Patients were included if they were 3 years of age or younger at the time of initial presentation. IRB approval was obtained from both institutions prior to the study. We report eight cases of occipitocervical junction instrumentation and fusion surgery in infants equal to and/or less than 4 years of age at the time of operation. We recorded demographic information, associated comorbidities, previous operations, radiographic imaging, clinical features, intraoperative details, follow-up, and complications. All patients underwent OCJ instrumentation and fixation procedures for their individualized indications, and variable constructs were utilized from the occiput to either axial or subaxial spine. The etiology of OCJ instability, procedure performed, segments/levels fused, construct material used, graft type, use of adjunct biologics, follow-up duration (12 to 108 months), fusion rate, and method of fusion were evaluated. We also reviewed constructs described in other published reports. The complexity of early childhood craniocervical pathology, defining the goals of surgery, and the challenges of stabilization in this unique subset of the pediatric population will all be addressed within the context of these patients.

Results

The combination of available literature and additional data from our own case series was used to help better define indications for surgery and report on the outcomes of occipitocervical instrumentation in a young pediatric population. Our series of eight very young children who have undergone occipitocervical fusion in the setting of congenital, iatrogenic, and/or pathological instability had a mean age of 22.28 ±12 months with a mean follow-up of 41.25 ± 29 months. The choice of construct was made with respect to the existing pathology and taking into consideration the future growth potential of each patient. All patients underwent rigid instrumentation with or without autologous rib grafts. Authors reported use of demineralized bone matrix (DBM) in only three cases (37.5%); however, no formal bone morphogenetic protein (BMP) was used. All of our cases documented good immediate and long-term craniocervical sagittal alignment (occiput to C2 and C2-7 angles). Only one patient (case 1) developed postoperative kyphosis/nonunion due to occipital screws pullout. One patient developed minor wound breakdown, which responded to a course of antibiotics in the outpatient setting. We did not

use a formal pediatric quality of life measure (PedsQL) to assess the overall outcome in our cases. However, parent-reported outcomes were utilized and equally considered as a valid measure to assess the overall outcome and patient satisfaction in this subset of the pediatric population.

Patients' demographic data, diagnoses, choice of construct, complications, and follow-up are summarized in Table 1. A more detailed description is provided in three of our unique cases.

Case 1

A 12-month-old male with Chiari malformation type II, L4 myelomeningocele, and hydrocephalus with a ventriculoperitoneal (VP) shunt and suboccipital craniectomy with C1 to C4 laminectomies presented with symptoms of respiratory distress. The patient had significant obstructive sleep apnea (OSA), stridor and poor neck tone, and iatrogenic kyphotic deformity of cervical spine. Radiological imaging confirmed ventral compression of the cervicomedullary junction with worsening of cervical spine syringomyelia and cervical spine kyphosis (Fig. 1a–b). We performed a suboccipital and C1–C5 revision of his decompression with occipitocervical instrumentation. Bilateral titanium rods (affixed with the occipital screws to the keel) were placed from occiput through T1 and sutured with 1-0 polyester sublaminar sutures (Mersilene [Ethicon, Somerville, NJ, USA]) of C6, C7, and T1. The right 10th and 11th ribs were harvested and sutured to the rods. The lateral gutters were augmented with an allograft of cortical cancellous bone chips and demineralized bone matrix (DBX) (DePuy Synthes Spine) (Fig. 2a–b). The patient was placed in hard cervical collar for 6 weeks after surgery to provide immobility and promote fusion.

Table 1 Demographics, diagnoses, operative details, and postoperative course

Case no.	Age (mos.)/ sex	Diagnoses	Type of construct	Complications	Postop follow-up (mos.)
1	12/M	Chiari malformation Type II + hydrocephalus + C2–C5 laminectomy (previous) with severe occipitocervical instability	Occiput to T1, occiput titanium rods and screw construct with sublaminar wires + T9 and 10 rib autografts, revision with Mersilene suture and cables at the occiput	Screw pullout (occipital bone)	42
2	36/M	Chiari malformation Type I with ventral brain stem compression + suboccipital craniectomy (previous)	Occiput to C3 arthrodesis, occiput plates with adjustable rod system to C2 pedicle and C3 lateral mass screws construct + decortication with right T9 rib autograft mixed with demineralized bone matrix (DBM)	None	24
3	24/M	Pierre Robin syndrome + Gordon syndrome + GDD [Global Developmental Delay] + torticollis + respiratory insufficiency and feeding difficulties	Two-stage procedure. Stage 1: Halo vest + sternomastoid muscle release. Stage 2: Occiput to C4 arthrodesis, occiput plate with hinged rod syndrome. Right C2 pedicle and left lateral mass screws, bilateral C4 lateral mass screws. Decortication autograft mixed with DBM.	None	48
4	27/M	Wolf-Hirschhorn syndrome with occipitocervical instability	Occiput sublaminar wires using Luque loop system with C2–5 lateral mass screws + autologous rib bone grafts	None	36
5	28/F	Klippel-Feil syndrome + torticollis with decreased range of left sided axial rotation	Initial halo vest placement followed by occiput to C3 arthrodesis, occiput plates with hinged rod system. C2 pedicle and right unilateral C3 lateral mass screw + left sublaminar wire construct. Decortication + left iliac crest autograft mixed with DBM	None	36
6	10/M	Left C1 hemivertebra and occipitalization + torticollis + infantile idiopathic scoliosis + tethered cord syndrome	Two-stage procedure. Stage 1: Halo vest + sternomastoid muscle release. Stage 2: occiput to C3 arthrodesis, occiput plate, left C2 pars, right C2 translaminar and bilateral C3 lateral mass screws, decortication with right T6–T9 rib autografts	None	12
7.	38/M	Chiari malformation type I + basilar impression + ventral brain stem compression	Two-stage procedure. Stage 1: halo vest placement. Stage 2: suboccipital decompression + C1 laminectomy followed by occiput to C2 arthrodesis, occiput connectors, C2 pars and C3 lateral mass screws with bilateral self-contoured rods, decortication with T7–8 rib autograft	None	108
8	8/M	Spondyloepiphyseal dysplasia with occipitocervical instability	Occiput to C2 arthrodesis, T9 to T11 rib autograft construct and sublaminar Mersilene sutures	Minor skin breakdown resolved with Minerva jacket	24

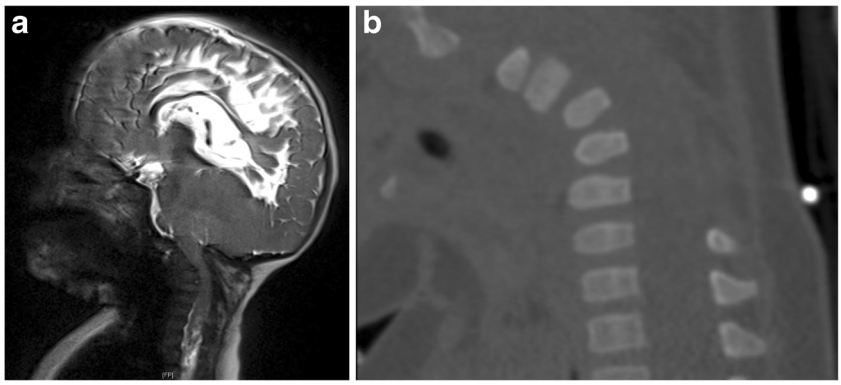

Fig. 1 **a** MRI of brain including cervical spine with postlaminectomy cervical kyphosis and intramedullary T2 hyperintensity suggestive of progression of syringomyelia from C5 to T2. **b** Cervical spine CT scan sagittal view with the progressive cervical spinal kyphosis and anterior subluxation of C4–C5

At his 6-month follow-up visit, scanty serous discharge was observed from occiput. CT scan with 3D reconstruction demonstrated fusion along rib autografts (Fig. 3a); however, there was occipital screw pullout with disengagement of the proximal end of the construct from the occiput **(**Fig. 3b). Suboccipital wound revision was performed up to C3, and rods were affixed to the occiput with cables utilizing four Mersilene sutures. The rostral extent of the rods were drilled and a small amount of bone cement [Hydroset® (Stryker, Kalamazoo, MI, USA)] was placed on top of the rods to provide a less irritating contour to facilitate wound healing (Fig. 3c–d). Postoperatively, he was placed in a halo vest brace and was discharged on postoperative day 5. On postoperative day 8, loosening of halo pins and patient discomfort directed us towards discontinuation of the halo and replacement with a Minerva vest. Follow-up cervical spine radiographs demonstrated a well aligned instrumentation with optimal bone fusion **(**Fig. 4**)**.

Case 2

A 36-month-old male presented to our clinic with progressive symptoms of swallowing and feeding difficulty, multiple aspirations, neurogenic bladder, and central sleep apnea secondary to ventral brainstem compression in the setting of an iatrogenic craniocervical instability. He had a gastrostomy tube (MIC-KEY) placed to meet all hydration/nutrition requirements. He had significant relief of his symptoms while wearing a hard collar. MRI scans of cervical spine demonstrated worsening of the clivo-axial angle (CXA) of 120° (Fig. 5a). Suboccipital epidural adhesiolysis revision surgery was performed with the widening of the lateral gutters of foramen magnum. Occiput to C3 instrumentation was performed with the bilateral adjustable rod system (MOUNTAINEER Spine System), DePuy Synthes Spine, Raynham, MA, USA) (Fig. 5b). Subsequently, cranioplasty was performed with a titanium mesh (4 × 2 cm) for strong attachment and to

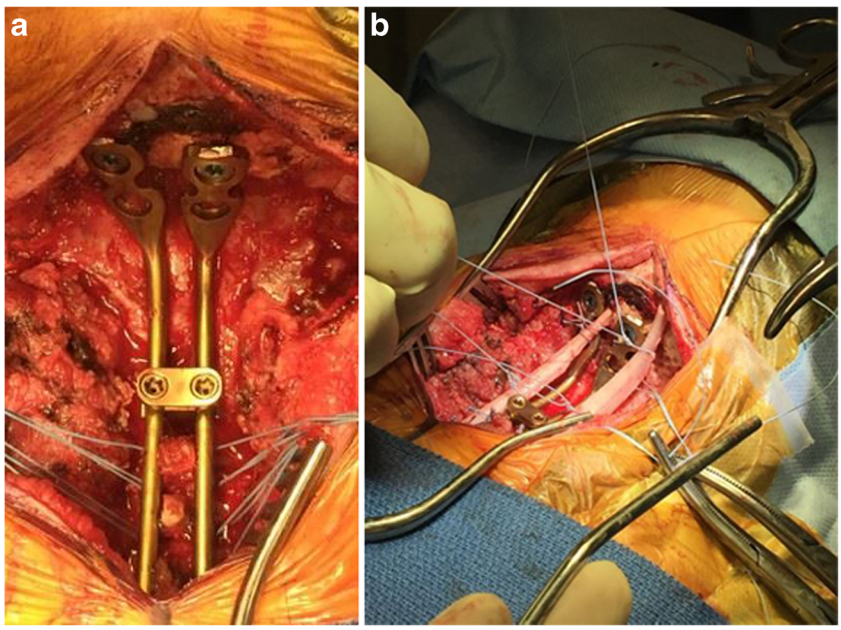

Fig. 2 **a**, **b** Intraoperative images with use of bilateral titanium rods (affixed with the occipital screws to the keel) placed from occiput down to T1 and sutured with 1-0 polyester sublaminar sutures (Mersilene) of C6, C7, and T1. Right-sided 10th and 11th ribs were harvested and sutured to the titanium rods

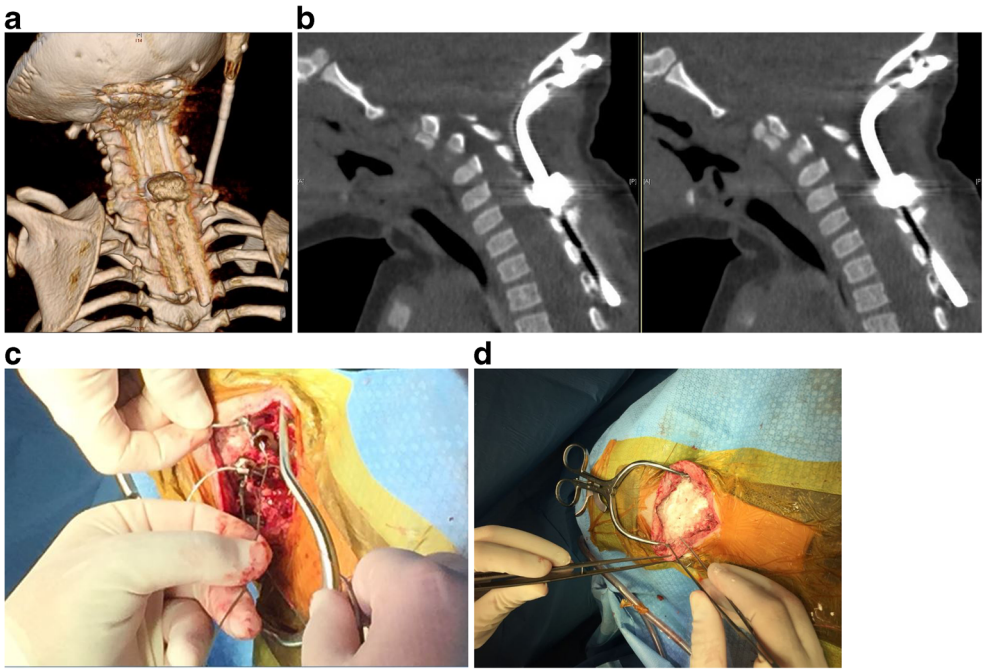

Fig. 3 **a** 3-D reconstruction CT scan of cervical spine with the fusion of bilateral rib autografts. **b** CT scan of cervical spine revealing bilateral occipital screws pull out. **c**, **d** Intraoperative images revealing placement of bilateral suboccipital wires around the top holes of the occipital plates followed by the use of HydroSet bone cement on top of the construct

avoid tethering of dura to the suboccipital muscles. The patient had significant improvement of his symptoms after surgery. Postoperatively, his CXA was improved to 135° (Fig. 5c). At 1 month follow-up visit, all symptoms improved, but there was a small area of superficial skin breakdown due to the collar which resolved with extra padding and skin care.

Case 6

A 10-month-old male presented with the symptoms and signs of global developmental delay (GDD) tethered cord syndrome, severe torticollis secondary to the left sided hemivertebra, and occipitalization of C1. On physical exam, he had left-sided positional plagiocephaly with significant head tilt and facial asymmetry with marked restriction of axial neck rotation to the left side. Neurological exam revealed early signs of neuraxial compression. Radiographic imaging showed cervical scoliosis as well as significant head tilt (Fig. 6a). CT scan of the cervical spine revealed occipitalization of C1 with hypoplasia of right C1 and C2 lateral masses (Fig. 6b). After several trials of a rigid collar, physical therapy without improvement, and progression of neuroaxial compression surgical intervention was undertaken with a planned two-stage procedure. The first stage was performed with release of the sternocleidomastoid muscle and the placement of a halo west. A week later, the second stage entailed occiput to C3 arthrodesis (occiput plate with left-sided C2 pars screw and a right-sided translaminar screw and bilateral C3 lateral mass screws (MOUNTAINEER Spine System, DePuy Synthes Spine, Raynham, MA, USA) with right-sided T6–T9 rib autografts. The patient was placed in the halo vest postoperatively. Good cervical alignment was attained after surgery. The halo vest was removed 1 month after surgery. During last follow-up (12 months after surgery), the patient was observed making good progress without any clinical or instrumentation-related issues (Fig 6c–d).

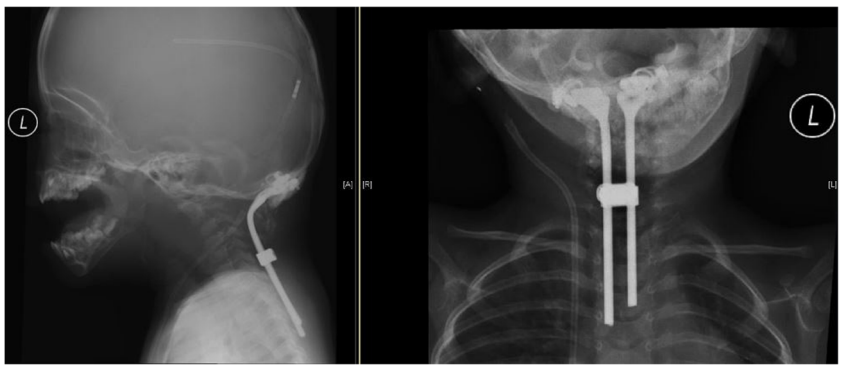

Fig. 4 Follow-up X-ray of cervical spine lateral view demonstrating the optimal craniocervical alignment with stable construct

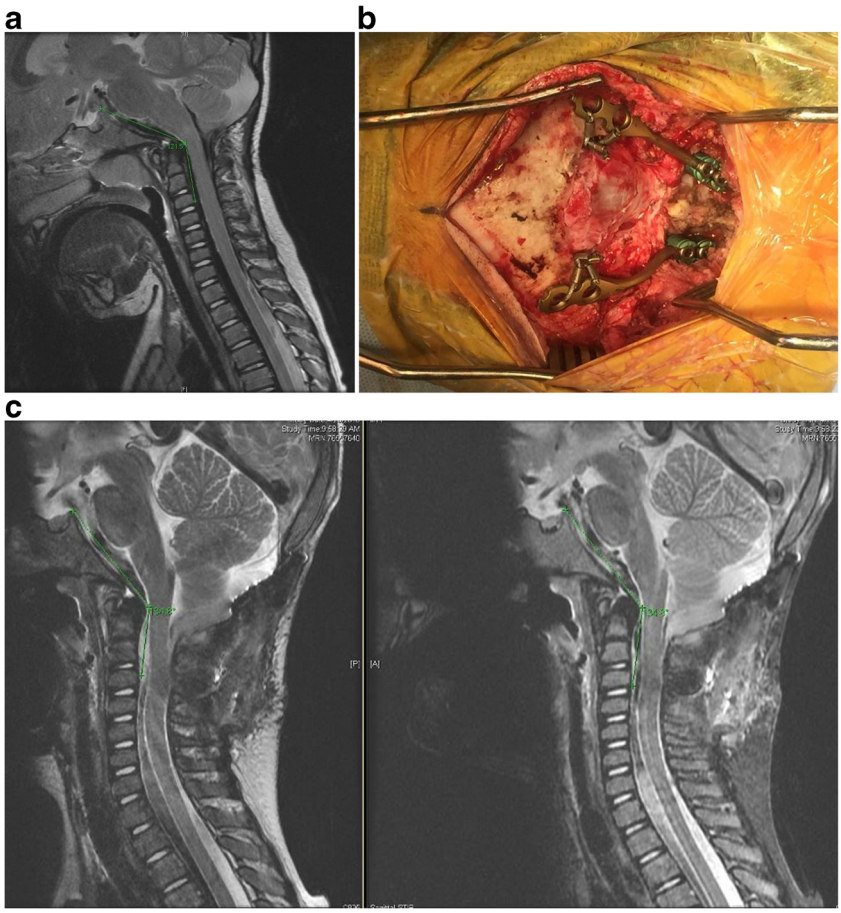

Fig. 5 **a** MRI of cervical spine with clivo-axial angle (CXA) of 120° with medullary kinking. **b** Postoperatively, the CXA improved to 135° with a CSF flow around the foramen magnum. **c** Revision suboccipital craniectomy revealing bilateral occipital rods construct which was affixed with cables tunneled through the occiput bone lateral to the craniectomy

Discussion

There are few indications for OCJ fusion in young children, but when radiographic suspicion of instability is paired with lower cranial nerve dysfunction and myelopathy or brainstem findings, it may be required. Inserting instrumentation into young children presents several technical challenges to pediatric spine surgeons. Most of these children have an underlying neuroanatomic dysfunction compounded by anatomic constraints due to size; translation of adult constructs to the pediatric spine is challenging, and the children manifest unique issues with respect to healing and arthrodesis. A successful fusion depends on meticulous surgical planning, selection of appropriate instrumentation, optimizing graft for fusion, and postoperative immobilization with the use of hard collar [14].

Nonoperative management with external orthosis

Initially, use of an external orthosis is considered for management of symptoms and signs of craniocervical instability. Among available options are hard collars, fixed cervicothoracic braces (CTSO) or jackets, or halo vests [15]. The hard collar is traditionally preferred as it both helps in the initial management and may aid in the diagnosis of CCI when this diagnosis may be uncertain. The other common indication for its use is to augment the stabilization and theoretically promote fusion after internal instrumentation surgery [16]. Postoperative use of an orthosis is necessary to promote optimal fusion in these young children. Noncompliance with collar wear and skin breakdown are two important considerations and should be part of routine preoperative discussion and predischarge teaching [15]. It is crucial to monitor the skin on a regular basis. Moreover, the use of soft padding or thermoplastic materials and custom-made braces improves compliance and helps to meet the goal of bracing without increasing morbidity from pressure points and skin breakdown [15].

Age, skeletal immaturity, and growth potential

Surgical instrumentation in a young child may raise concerns due to a patient's age and desirability of maintaining the growth of an immature spine. Moreover, delicate and small bony structures may not withstand rigid instrumentation due to excessive mobility, poor bone purchase at the time of instrumentation, and still developing ossification centers with cartilaginous interpositions [17]. It is thought

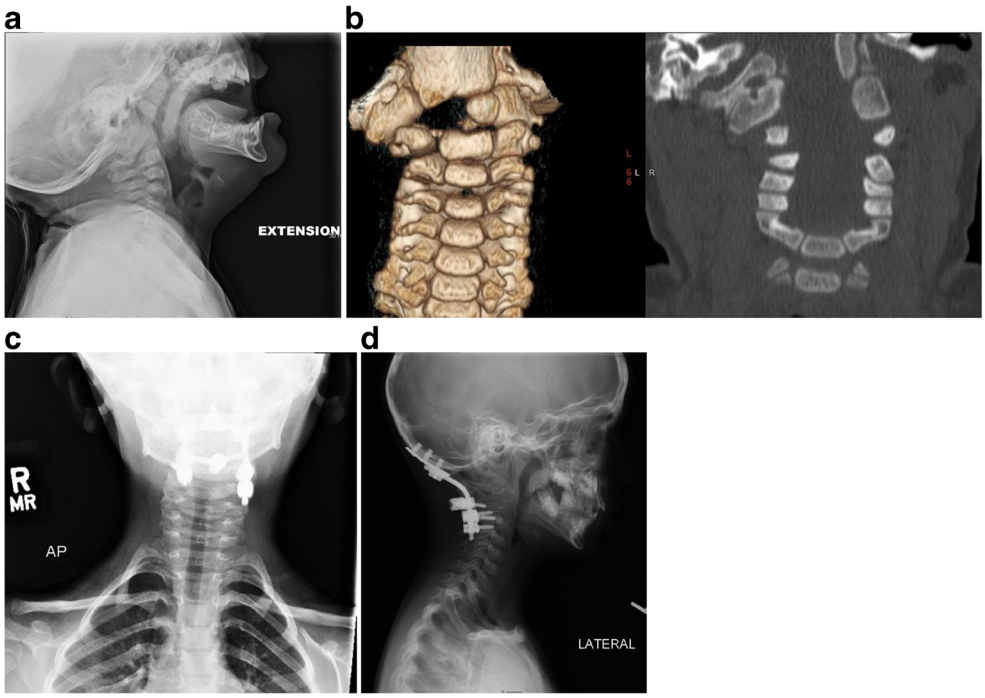

Fig.6 **a** X-rays of cervical spine lateral film revealing occipitalization of C1 with abnormal craniocervical alignment. **b** CT scan of cervical spine coronal view revealing hypoplasia of right lateral masses of C1 and C2 vertebra. **c**, **d** Cervical spine lateral and PA films revealing well aligned craniocervical junction with a stable hardware construct.

that the cervical spine matures at the age of 8 to 9 years with completed ossification and adult-like vertebral bodies and intervertebral discs [18]; occipitocervical fusion in earlier pediatric age groups has the potential to limit growth and cause secondary deformity [18]. Previously, accepted criteria for instrumenting the pediatric cervical spine, included signs of cervicomedullary kinking, significant spinal deformity, dynamic instability, and an age greater than 8 years [19]. Literature is sparse on instrumented fusion in toddlers managed by external orthosis or with minimal instrumentation [19, 20]. Generally, we favor deferring surgery using an external orthosis until patients are older and then select surgical intervention based on considerations as outlined below. We chose to include only children age 3 years or less in our study because the bony anatomy typically becomes much more favorable for instrumented fusion at around age 3 [17].

Selection of a particular construct for instrumentation in young children (age $\leq$ 3 years)

Proposing a particular construct to utilize in a very young child is based upon anatomic specifics, usually defined from preoperative CT scan, and may depend upon a surgeon's preference and familiarity with a given system. Within the cervical spine, the use of rods with sutures has been a favorable construct considering the delicate and immature bone of young children. The choice of absorbable versus nonabsorbable sublaminar sutures is a matter of personal preference. Ha et al. [19] used nonabsorbable sutures in severe distraction injury and reported no complications during their follow-up. The purported advantages of this technique include avoidance of growth disturbance, preservation of motion, and prevention of iatrogenic injury to the ossification zone and the smooth lamina, which may be caused by wires or screws [19]. Luque loops with wires have also been adopted by some surgeons. We have successfully used a similar construct in one of our patients (case 4) (Table 1). More recently, a number of screw fixation techniques have been described in the literature; however, osseous immaturity poses significant challenges. Odent et al. [21] warned about the significantly compromised screw purchase in toddlers. Benzel et al. [22] employed occiput to C2 constructs using autologous rib bone autograft and further augmented this with BMP-2 and Mersilene sutures used as "cross-connectors," similar to one of our techniques, to achieve an optimal fusion.

Hamoud et al. [20] recommended utilization of absorbable sutures with a rod construct to avoid foreign material and minimize the risk of deep infections. Moreover, fixation to the occiput demands a great attention due to a thinner keel. Utilizing occipital hooks for distraction and cervical sublaminar hook claws with C2 pedicle screws for immediate stability may provide better purchase in the immature occipital bone, avoiding complications from bicortical screws. However, these hooks may not be properly sized for children less than 3 years of age [23–25].

Instrumented fusion in toddlers has increased due to trauma and an increase in familiarity with instrumentation systems and techniques [12, 26, 27]. Screw-based constructs provide superior stabilization and biomechanical profiles when compared with sublaminar wires [25, 28, 29]. The literature is limited on occipitocervical fusion in very young children,

and most of the surgical techniques, outcomes, and complications are extrapolated from studies in either older children or adults [21]. Traditional onlay techniques with or without wiring have previously been successfully utilized [28, 29]. Poor control of stability in the rotational plane is a limitation to these methods; however, this can be overcome using screws or wires at the point of maximum load.

Excellent fusion rates have been reported in some case series while others have encountered a pseudarthrosis rate of 25–33% in children [29]. The documented 92.3% fusion rate in the study by Odent et al. [21] was concordant with other studies by Flint and Hockley [30] and Higo et al. [31], who reported exceptional fusion rates using a Hartshill (metal) rectangle and a Luque loop rod system, respectively. In a recent multicenter study with propensity matched cohorts of 28 children where half of the children were treated with non-rigid instrumentation (wires or cables) and the other half with screws or hooks and rods, a higher nonunion rate was observed in the non-rigid group ($p = 0.0057$), supporting the use of screws and rods with postoperative halo body jacket placement [32]. The available literature strongly supports the contention that rigid occipitocervical fusion aids not only in fusion but also promotes earlier recovery and faster rehabilitation in this subset of the pediatric population [13]. Choosing a construct with extension of instrumentation to C3 versus C2 might add a little more biomechanical strength or better load sharing to the construct. In one of our cases, the selection of a semi-rigid fixation was purely case based and based on the intraoperative findings of bone quality. Semi-rigid fixation is still a viable option considering the risk benefit ratio [35]. Moreover, authors have recommended case based/ intraoperative decision-making as a selection criteria for optimal occipitocervical instrumented fusion construct. Among selection of type of various available constructs, a midline keel fixation system is encouraged due to greater bone thickness and stronger bicortical fixation.

Choice of bone graft

The choice of bone graft is crucial once the instrumentation has been defined. Odent et al. [21] utilized autologous iliac crest; however, Ha et al. [19] suggested that it was acceptable to defer harvesting additional autologous bone grafts. Mueller et al. [33] did not report the use of any bone graft but advocated for 4 weeks in a halo followed by a semi-rigid cervical collar for 10 days. Mazur et al. [17] reported their results in large series with the use of autografts in the majority of cases (96.9%); biological adjuncts were used in 79.5% (DBM in78.7% and BMP in 5.5% of cases only). BMP was used in most of the revision operations. We used DBM in our cases where supplementation with the autograft was necessary [17]. However, BMP was not used even in the revision surgery (case 1). The formal use of BMP can be considered in lieu of revision surgeries if risk of poor fusion is high. However, the regular use of BMP in this age subgroup is still a matter of debate. Supplementation of these constructs with autografts is helpful to overcome skeletal immaturity, and for early bone healing. Authors have used rib autografts in some of the reported cases with good results. Alternatively, full thickness calvarium can be considered as a graft option.

Another non-traumatic consideration of OCJ fusion is in patients with irreducible atlantoaxial dislocations while, refuting the need for transnasal/transoral odontoid decompression surgery. Visocchi et al. [36] reported complete neuraxial decompression in three pediatric patients, (two with Down's syndrome, one rheumatoid arthritis) after an unsuccessful preoperative traction test. A combination of axial traction with light extension of neck on chest and flexion of the head followed by OCJ fixation documented good results.

Through literature review and analysis of our case series, we have tried to emphasize a number of factors. Strong preoperative planning, family counseling, and attention to the construct are crucial before undertaking these cases. A careful consideration of future growth potential and intended cervical alignment as well as very personalized adaptations for external bracing systems will help minimize immediate and late complications. Most of the published studies are short-term follow-up; therefore, limited information is available on postfusion adjacent segment disease or crankshaft phenomenon. Very close and long-term follow-up are necessary, more than in almost any other pediatric population due to risks of wound, instrumentation, growth related, and neurologic complications. Limitations in our study include relatively small sample size, short follow-up, and heterogeneity of surgical techniques employed across a diverse number of etiologies resulting in craniocervical and subaxial instability in very young children.

Conclusion

Posterior surgical instrumentation and fusion of the occipitocervical region and upper cervical spine in very young children is relatively rare, and treatment strategies can be challenging. Internal rigid fixation with autologous bone graft has yielded better fusion rates, reducing the need for prolonged external halo vest immobilization. Experience from our case series and a review of the literature support careful selection of personalized occipital plates, rods, screws and wire constructs depending on the underlying pathology, bone thickness, age, and degree of suspected instability. Thin or weak bone, revision surgeries with or without intraspinal pathologies, difficulty in compliance with immobilization, and skin integrity secondary to proud instrumentation on the occiput remain challenging issues. A properly fitted extended neck and chest brace and serial imaging with very frequent outpatient

follow-up visits are recommended. Fusion rates can be very high and neurologic outcomes are excellent; thus, pre- and perioperative management should be directed at minimizing and being prepared for the expected challenges during the healing process.

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Compliance with ethical standards

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