The development of rigid internal occipitocervical fixation has improved fusion rates in patients with occipitocervical instability and has largely eliminated the need for postoperative external fixation. Before the use of rigid internal fixation constructs, bone and wire constructs supported by rigid external orthoses were associated with significant halo-related morbidity and suboptimal fusion rates. In the adult population, current techniques for occipitocervical and atlantoaxial fixation have led to fusion rates approaching 100% without the use of postoperative halo fixation.
Rigid occipitocervical fusion with or without C-1 instrumentation

Authors have also reported excellent results using rigid internal fixation at the occipitocervical junction in children. Nevertheless, the safe placement of occipitocervical instrumentation in the pediatric population may be hindered by the size and developmental stage of the relevant anatomical structures as well as by the pathological conditions that require occipitocervical fusion, many of which are characterized by atypical regional anatomy. As such, multiple construct configurations are used to achieve rigid internal occipitocervical fixation, some of which do not include direct C-1 instrumentation.

Although some authors have advocated the use of multiple fixation points in multilevel segmental constructs to improve stability, Wolfla and colleagues have demonstrated in adult cadaveric specimens that the placement of C-1 lateral mass screws does not improve occipitocervical construct rigidity when compared with constructs without C-1 instrumentation. In the pediatric population, it is clear that there is risk associated with the placement of instrumentation in the upper cervical spine. For example, Jea and colleagues have described the placement of C-1 lateral mass screws in 4 young children, one of whom suffered VA injury. In the largest study of pediatric TAS placement, the rate of VA injury was slightly < 3% (2 of 67 patients). The purpose of the present study was to evaluate the fusion and perioperative complication rates in children who had undergone rigid internal occipitocervical instrumentation and fusion using a variety of constructs to determine if direct C-1 instrumentation was necessary.

Methods

We retrospectively reviewed the construct characteristics, fusion rates, and complications in 125 pediatric patients who had undergone occipitocervical or high cervical fusion at 9 different children’s hospitals. The study period ranged from January 1, 1995, through January 1, 2009. Inclusion criteria included occiput-C1 instability with or without C1–2 instability and without subaxial instability. Forty-eight patients either demonstrated subaxial instability or did not have atlantooccipital instability; these patients were excluded from the study, leaving 77 who underwent occiput-C2 instrumented fusion using rigid internal constructs. The details of each patient construct were evaluated, and each construct was placed into 1 of 3 groups based on the characteristics of the anchoring spinal instrumentation, as every patient construct included occipital screws. The groups were as follows: Group 1, C-2 instrumentation; Group 2, C-1 and C-2 instrumentation without TAS; and Group 3, any TASs (uni- or bilateral). There were no occipitocervical constructs with unilateral or C1-only spinal instrumentation. Iliac crest or rib autograft was used in almost all cases. Groups were compared for perioperative complications, rates of fusion, and methods of postoperative bracing. Long-term follow-up data were not assessed, as this study was designed to evaluate the safety of construct placement and initial fusion rates.

Results

The median age at surgery was 7.5 years (range 1.3–18.8 years). Indications for surgery included congenital anomalies (47 patients [61.0%]), trauma (22 patients [28.6%]), infection (1 patient [1.3%]), and unknown causes (7 patients [9.1%]). Among the 47 patients with congenital anomalies, 12 (25.5%) had Down syndrome and 2 each (4.3%) had Loeys-Dietz syndrome and Morquio syndrome. One patient (2.1%) had Hurler syndrome and 1 (2.1%) had multiple pterygium syndrome. The remaining 29 patients (61.7%) presented with unnamed congenital anomalies.

In all patients, preoperative occipitocervical anatomy was evaluated using thin-slice CT scans with sagittal and coronal plane reconstructions. Further preoperative imaging was undertaken at the discretion of the operating surgeon. Group 1 (C-2 instrumentation; Fig. 1) consisted of 16 patients (20.8%), Group 2 (C-1 and C-2 instrumentation without TASs; Fig. 2) included 22 patients (28.6%), and Group 3 (any TASs; Fig. 3) represented the remaining 39 patients (50.6%; Table 1). All C-2 instrumentation included a TAS, pars screw, or laminar screw. There were no constructs that included C-2 pedicle or lateral mass

![Fig. 1.](image-url)
screws, or wiring alone. Cables were used only to maintain the position of the autograft bone within the rigid construct. The median follow-up for all patients, including 2 who were lost to follow-up before radiographic evaluation of the fusion, was 14.2 months (range 1.0–56.2 months).

**Fusion Rates**

Overall, 75 patients underwent follow-up imaging, and radiographic fusion was demonstrated in all (100%) within 6 months of surgery. Fusion was evaluated using thin-slice CT scanning with sagittal and coronal plane reconstructions in all patients. Two patients (2.6%) from Group 2 were lost to follow-up prior to documented fusion (Table 2). Iliac crest or rib autograft was used in all but 4 cases, each from Group 2. In these 4 cases, morcelized cancellous allograft was used with bone morphogenetic protein–2 (Infuse, Medtronic); fusion was achieved in each of these cases. There were no instances of documented fusion failure. Groups 1 and 3 had fusion rates of 100%. Group 2 had a documented fusion rate of 90.9% (that is, 20 of 22 patients because of the 2 patients lost to follow-up). After documented fusion, patients were not followed up radiographically.

**Procedural Complications**

There were 7 perioperative complications in 6 patients, although none required construct revision. In Group 1 there were 2 complications (12.5%, wound hematoma and graft site infection), in Group 2 there were 3 complications in 2 patients (13.7%, VA injury, superficial wound infection, and reintubation), and in Group 3 there were 2 complications (5.1%, 2 VA injuries).

Three patients (3.9%) suffered VA injury, 2 during TAS placement and 1 during C-1 lateral mass screw placement. No VA injury resulted in neurological sequelae. There were no cases of VA injury in Group 1 (C-2 instrumentation only). The injuries during TAS placement occurred in a 3-year-old and a 12-year-old patient. In each case, the screw was left in place, and the patient was followed up with postoperative CT angiography, which demonstrated thrombosis of the VA to the junction of the vessel with the basilar artery. Both patients were treated with aspirin to minimize the risk of progressive thrombosis. The VA injury during C-1 lateral mass screw placement occurred after successful screw placement on the contralateral side in an 8-year-old boy. The VA was breached during pilot hole placement, and the screw was quickly placed to tamponade.
Rigid occipitocervical fusion with or without C-1 instrumentation

<table>
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<th>Table 1: Preoperative characteristics in 77 patients who underwent rigid internal fixation</th>
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*Pts = patients.

the injury. Postoperative CT angiography demonstrated thrombosis of the vessel without evidence of pseudoaneurysm formation. This patient was treated with aspirin.

**Postoperative Bracing**

Methods of postoperative bracing included a rigid cervical collar (63 patients [81.8%]), a halo (11 patients [14.3%]), and no bracing (3 patients [3.9%]). There were no significant differences in the rates of halo or hard collar use among the groups. Halo fixation was selected on an individual basis and was not related to the characteristics of the construct (Table 2).

**Discussion**

Data in this study demonstrate that rigid occipitocervical instrumentation can be safely placed in the pediatric population and that excellent fusion rates can be achieved. Moreover, C-1 instrumentation may not be required to achieve fusion in pediatric patients.

Experimental and clinical reports have demonstrated findings consistent with ours. A biomechanical study by Wolfla and colleagues has demonstrated in adult cadaveric models that occiput-C2 constructs including C-1 lateral mass screws did not impart additional rigidity when compared with those lacking C-1 screws, even the constructs with bilateral TASs, pars screws, and laminar screws. Although this study was performed in adult cadaveric specimens, the experimental findings are consistent with our clinical results in a pediatric population.

Clinically, excellent fusion rates have been achieved regardless of the construct design. For example, Gluf and Brockmeyer have reported a 100% fusion rate in 23 patients who underwent occiput-C2 instrumentation using TASs without halo fixation. Furthermore, Anderson and colleagues have described the successful application of flow diagrams to guide the choice of construct configuration in 25 children who could not safely undergo bilateral TAS placement. In that series, spinal instrumentation included C-1 lateral mass screws, C-2 pars screws, and C-2 laminar screws. Similarly, Schultz and colleagues have reported a 92.9% fusion rate in 14 children who underwent occipitocervical fusion with a variety of constructs. In smaller series, multiple construct configurations have also been successfully used.

A fundamental question remains, however, regarding how much instrumentation is required to achieve a solid fusion. This question is particularly germane in younger pediatric patients, whose anatomy often makes screw placement technically challenging and risky. Jea and colleagues, for example, have placed C-1 lateral mass screws in 4 children younger than 8 years of age. Although C-1 was instrumented in each of these patients, a VA injury occurred in 1 patient, highlighting the risk involved in placing C-1 lateral mass screws in children. Furthermore, the limited space and extreme angles at the craniocervical junction in young children often make it technically challenging if not impossible to place bilateral screws in both C-1 and C-2. Because the documented fusion rates in children with or without direct C-1 instrumentation in the present study were 100% (acknowledging that 2 patients were lost to follow-up prior to fusion), our data suggest that direct C-1 instrumentation may not be necessary in occiput-C2 constructs in children.

The primary strength of our study is that, to our knowledge, it represents the largest series of pediatric occipitocervical fusions using rigid instrumentation to date. Additionally, our results are likely to be generalizable, as the data were derived from multiple medical centers and include many different construct configurations. The retrospective study design, however, does limit the strength of the conclusions that can be drawn. Additionally, despite our having gathered data from 9 children’s hospitals over 14 years, the rarity of these cases has limited the number of patients we could study. This limitation prevented us from performing detailed statistical analyses of individual institutions or from mathematically demonstrating the absence of a difference between fusion rates in the study groups, although the data clearly imply there is no such difference. There were no patients with occiput-C1 constructs or unilateral-only instrumentation, making evaluation of these constructs impossible. Two patients were lost to follow-up, and we have not completed long-term follow-up in many patients, which would be important to the understanding of spinal growth in these children.

**Conclusions**

This retrospective study of 77 pediatric patients from 9 children’s hospitals demonstrates that occipitocervical fusion using rigid internal fixation can be reliably achieved using a variety of construct designs. Our data also suggest that direct C-1 lateral mass instrumentation may not be necessary in these constructs. Avoiding C-1 instrumentation may not only reduce hardware-related complications
such as vertebral artery injury, but also lower blood loss from paravertebral plexus dissection and reduce overall operative time. The large number of surgeons and construct configurations evaluated in this study implies that our findings, although retrospective in nature, are largely generalizable and should provide guidance regarding the amount of instrumentation required to safely achieve occipitocervical fusion in the pediatric population.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


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