Patient-reported outcomes of occipitocervical and atlantoaxial fusions in children

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OBJECTIVE There is limited literature on patient-reported outcomes (PROs) and health-related quality of life (HRQOL) outcomes in pediatric patients undergoing surgery for craniovertebral junction pathology. The aim of the present study was to assess surgical and quality of life outcomes in children who had undergone occipitocervical or atlantoaxial fusion.

METHODS The authors retrospectively reviewed the demographics, procedural data, and outcomes of 77 consecutive pediatric patients who underwent posterior occipitocervical or atlantoaxial fusion between 2008 and 2015 at Texas Children’s Hospital. Outcome measures (collected at last follow-up) included mortality, neurological improvement, complications, Scoliosis Research Society Outcomes Measure–22 (SRS-22) score, SF-36 score, Neck Disability Index (NDI), and Pediatric Quality of Life Inventory (PedsQL). Multivariate linear regression analysis was performed to identify factors affecting PROs and HRQOL scores at follow-up.

RESULTS The average age in this series was 10.6 ± 4.5 years. The median follow-up was 13.9 months (range 0.5–121.5 months). Sixty-three patients (81.8%) were treated with occipitocervical fusion, and 14 patients (18.1%) were treated with atlantoaxial fusion. The American Spinal Injury Association (ASIA) grade at discharge was unchanged in 73 patients (94.8%). The average PRO metrics at the time of last follow-up were as follows: SRS-22 score, 4.2 ± 0.8; NDI, 3.0 ± 2.6; the parent’s PedsQL (ParentPedsQL) score, 69.6 ± 22.7, and child’s PedsQL score, 75.5 ± 18.7. Multivariate linear regression analysis revealed that older age at surgery was significantly associated with lower SRS-22 scores at follow-up (B = −0.06, p = 0.03), and the presence of comorbidities was associated with poorer ParentPedsQL scores at follow-up (B = −19.68, p = 0.03).

CONCLUSIONS This study indicates that occipitocervical and atlantoaxial fusions in children preserve neurological function and are associated with acceptable PROs and ParentPedsQL scores, considering the serious nature and potential for morbidity in this patient population. However, longer follow-up and disease-specific scales are necessary to fully elucidate the impact of occipitocervical and atlantoaxial fusions on children.

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KEY WORDS surgical outcomes; patient-reported outcomes; health-related quality of life; occipitocervical fusion; atlantoaxial fusion; traumatic atlantooccipital dislocation; pediatric neurosurgery; spine

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ABBREVIATIONS ASIA = American Spinal Injury Association; ChildPedsQL = child-reported PedsQL; HRQOL = health-related quality of life; IONM = intraoperative neuro-monitoring; MCS = Mental Component Summary; NDI = Neck Disability Index; ParentPedsQL = parent-reported PedsQL; PCS = Physical Component Summary; PedsQL = Pediatric Quality of Life Inventory; PRO = patient-reported outcome; SRS-22 = Scoliosis Research Society Outcomes Measure–22.


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of PROs and health-related quality of life (HRQOL) outcomes for other pediatric conditions is increasing.10 To the best of our knowledge, the use of PRO and HRQOL tools has not been previously undertaken for pediatric patients undergoing surgery for craniovertebral junction pathology. The aim of the present study was to perform an assessment of surgical interventions in a series of children with craniovertebral junction pathology, with an emphasis on measuring PROs and HRQOL outcomes.

Methods

We reviewed all consecutive patients (age ≤ 18 years) who underwent occipitocervical or atlantoaxial fusion performed by the pediatric neurosurgery service at Texas Children’s Hospital from 2008 to 2015. This study received approval from the Baylor College of Medicine Institutional Review Board.

All patients underwent surgery via a standard posterior approach, and no patient underwent a combined anterior-posterior surgery. The surgical technique for occipitocervical fusion performed by our team has been described previously.11,12,23 In brief, a midline posterior approach with subperiosteal exposure of the occiput and required cervical levels was performed. Occipital screws, C-1 lateral mass, and C-2 pars or pedicle screws were placed under fluoroscopic guidance. For fusions extending to lower levels, the relevant anatomy was exposed, and lateral mass or laminar screws were placed in a standard fashion for the cervical levels.

Clinical Outcome Measures

Patient demographics, disease characteristics, treatment variables, readmissions or reoperations, and surgical morbidity and mortality were recorded for each case. Postoperative HRQOL outcomes were captured at the most recent clinic visit. PRO instruments included Scoliosis Research Society Outcomes Measure–22 (SRS-22), SF-36 Physical Component Summary (PCS), SF-36 Mental Component Summary (MCS), neck disability index (NDI), and Pediatric Quality of Life Inventory (PedsQL) parent-reported score (ParentPedsQL) and child-reported score (ChildPedsQL). All PROs were obtained with the help of clinical research personnel at the most recent clinic visit. The SRS-22, SF-36 PCS, SF-36 MCS, and NDI forms were given to patients and parents for self-administration. For the ChildPedsQL, age-specific forms (ages 5–7, 8–12, and 13–18 years) were completed by children. For children in the 5- to 7-years of age group, the instructions and items were read out loud to the child and his or her answers were recorded. For children in the age groups 8–12 years and 13–18 years, self-administration was encouraged. If cognitive impairments prevented the child from completing the form, data for the ChildPedsQL form were not recorded. Parents were given the ParentPedsQL forms to complete.

Statistical Analysis

Descriptive statistics (mean ± SD) were computed as necessary. Multivariate linear regression analysis was performed for the SRS-22, ParentPedsQL, and NDI scores to determine the effect of age, sex, diagnosis, preoperative American Spinal Injury Association (ASIA) grade, number of levels fused, intraoperative complications, as well as presence of comorbidities; p values were considered significant at < 0.05 (Student t-test). Analysis was done using SPSS Statistics (version 20, IBM Corp.).

Results

Patient Population

A total of 77 patients were included in our study. The mean age ± SD at the time of occipitocervical or atlantoaxial fusion was 10.6 ± 4.5 years. Forty-one patients (53.2%) were boys.

Operative Treatment

Sixty-three (81.8%) of the 77 patients were treated with occipitocervical fusion. All patients underwent spinal instrumentation. The average number of levels fused was 2.4 ± 0.9. Most procedures lasted 2 to 4 hours (n = 64, 83.1%). The mean estimated blood loss was 84.0 ± 156.7 ml. Intraoperative neuromonitoring (IONM) was used in the majority of cases (n = 72, 93.5%). The overall intraoperative complication rate was 14.3%. The most common intraoperative complications encountered in our series included durotomy (3 cases) and loss/change in IONM signals (3 cases) (Table 2).

Outcomes

Clinical follow-up was available for 57 patients at a median duration of 13.9 months (1.4–121.5 months), with a minimum 6-month follow-up for more than 80% of patients. Two patients (2.6%) died during postoperative follow-up. One patient sustained a cardiopulmonary arrest intraoperatively due to suspected primary cardiac event and died on postoperative Day 2. Another patient developed postoperative propofol infusion syndrome and died on postoperative Day 43.

Neurological stability with no change in ASIA grade at discharge was seen in 73 patients (94.8%). One patient who underwent a supplementary occipitocervical fusion after resection of clival chordoma had an improvement from ASIA Grade D to Grade E, and another patient with traumatic atlantooccipital dislocation improved from Grade B to Grade D at discharge. No patient had a worsened ASIA grade at discharge. During the postoperative period, the most common complications were CSF leak (4 cases), dysphagia (2 cases, including 1 patient who required a temporary gastrostomy tube), and pneumonia/sepsis (2 cases) (Table 2). The majority of patients (n = 65, 84.4%) were discharged home after surgery.

PROs and HRQOL Outcomes and Predictors

The mean PROs and HRQOL outcome scores are shown in Table 3. Multivariate linear regression analysis (Table 4) revealed that increased age at surgery was significantly associated with lower SRS-22 scores (i.e., worse outcome) at follow-up (B = −0.06, p = 0.03). Multivariate
linear regression analysis showed poorer ParentPedsQL scores (i.e., worse outcome) at follow-up in patients with associated comorbidities ($B = -19.68$, $p = 0.03$). No significant predictors for the NDI score at follow-up were noted using the same regression model. Number of levels fused, preoperative ASIA grade, intraoperative complications, and diagnosis were not associated with PROs or HRQOL outcome scores.

**Discussion**

The present study describes PROs and HRQOL outcome scores in a consecutive series of pediatric patients undergoing surgery for craniovertebral pathology. The majority of patients had stable neurological examination results postoperatively and showed favorable PROs and HRQOL outcome scores at follow-up. Age at surgery and the presence of comorbidities were found to be significant predictors of PROs and HRQOL outcome scores.

The mean PROs and HRQOL outcomes in our series were similar to the mean scores after other pediatric neurosurgical procedures. In our cohort, the average SRS-22 score (4.2) was comparable to the mean postoperative SRS-22 scores reported for patients with adolescent idiopathic
TABLE 3. PROs and HRQOL outcomes in pediatric patients undergoing occipitocervical and atlantoaxial fusion

<table>
<thead>
<tr>
<th>Outcome Tool</th>
<th>No. of Patients</th>
<th>Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>SRS-22</td>
<td>42</td>
<td>4.2 (0.8)</td>
</tr>
<tr>
<td>SF-36 PCS</td>
<td>13</td>
<td>49.6 (10.6)</td>
</tr>
<tr>
<td>SF-36 MCS</td>
<td>13</td>
<td>52.5 (12.7)</td>
</tr>
<tr>
<td>NDI</td>
<td>36</td>
<td>3.0 (2.6)</td>
</tr>
<tr>
<td>ParentPedsQL</td>
<td>36</td>
<td>69.6 (22.7)</td>
</tr>
<tr>
<td>ChildPedsQL</td>
<td>26</td>
<td>75.5 (18.7)</td>
</tr>
</tbody>
</table>

scoliosis. The average SF-36 PCS score in this study was similar to values in pediatric patients who underwent fusion for scoliosis, as well as in children undergoing surgery for lumbar disc disease. The average PedsQL score in the present study compared favorably with good outcomes obtained in children with adolescent idiopathic scoliosis and intramedullary spinal cord tumors. These results show that expected quality of life outcomes after craniovertebral junction surgery in children are equivalent to outcomes seen in other pediatric spine surgeries.

The pediatric spine literature has been sparse in analyzing effects of occipitocervical and atlantoaxial fusion on quality of life in children. To the best of our knowledge, this study is the first to examine patient-centered characterization of pain, disability, and quality of life after occipitocervical and atlantoaxial fusions in children. Since craniovertebral junction fusion leads to a decrease in the range of motion of the cervical spine, it is to be expected that quality of life scores would be low after surgery. The results of this study, however, indicate that although craniovertebral junction pathology is associated with morbidity, surgery is associated with acceptable quality of life scores at intermediate follow-up. Given the number of active years in a child after surgery, long-term HRQOL outcome scores are necessary to better evaluate the impact of occipitocervical and atlantoaxial fusions on quality of life over time.

Atlantoaxial and craniovertebral instabilities represent unique challenges in their diagnosis and management, especially in the pediatric population, where smaller vertebral body size, abnormal anatomy caused by craniovertebral anomalies, and an immature spine are important issues encountered by pediatric spine surgeons. Numerous authors have shown the feasibility of achieving occipitocervical and atlantoaxial fusions even in the youngest of patients with rigid spinal instrumentation. In our series, all patients had spinal instrumentation incorporated as part of their fusion. Our results for neurological outcomes, as well as surgical complication rates, are similar to previous studies. Furthermore, significantly improved prehospital care for cases of traumatic atlantoorcicipital dislocation, hospital care for patients with traumatic and nontraumatic spinal cord injuries, and advances in surgical technique for placement of spinal instrumentation in children have resulted in decreased morbidity and mortality for pediatric patients undergoing occipitocervical and atlantoaxial fusions.

Our study showed that older age is associated with lower SRS-22 scores at follow-up, and a similar trend has been observed in patients with idiopathic scoliosis. It has been shown that younger patients are more affected by the ceiling effects of the SRS-22 questionnaire. This may be attributable to an early diagnosis in young children, who experience less pain, less compromised function, and better mental scores than older patients. Additionally, it is likely that older children have a decreased ability to adapt to the decreased range of motion after fusion. The presence of comorbidities was associated with lower ParentPedsQL scores in our series, similar to HRQOL studies from other disease states. Comorbidities decreased HRQOL scores and affect generic HRQOL tools, such as the ParentPedsQL, more than disease-specific tools. Comorbidities contribute additional physical and psychological burden for parents and need to be accounted for in determining HRQOL outcomes in children. The identification and treatment of these comorbidities are important, and surgeons need to be a part of a comprehensive care program for these children.

The primary limitations of our study lie in the small sample size, attrition rate, and retrospective design. The lack of long-term follow-up PRO data is another limitation of this study. Since we tried to use a number of PRO instruments at follow-up, we had difficulty ensuring that patients or parents completed all the PROs, which led to missing data values. We did not have data on preopera-

TABLE 4. Multivariate linear regression analysis to determine factors associated with PROs and HRQOL outcomes at follow-up after occipitocervical and atlantoaxial fusion in children

<table>
<thead>
<tr>
<th>Variable</th>
<th>SRS-22</th>
<th>ParentPedsQL</th>
<th>NDI</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>B</td>
<td>p Value</td>
<td>B</td>
</tr>
<tr>
<td>Age at op</td>
<td>−0.06</td>
<td>0.03*</td>
<td>−0.19</td>
</tr>
<tr>
<td>Sex (F/M)</td>
<td>0.23</td>
<td>0.35</td>
<td>3.35</td>
</tr>
<tr>
<td>Diagnosis†</td>
<td>−0.02</td>
<td>0.87</td>
<td>−2.08</td>
</tr>
<tr>
<td>Comorbidities (no/yes)</td>
<td>−0.23</td>
<td>0.38</td>
<td>19.68</td>
</tr>
<tr>
<td>Preop ASIA grade</td>
<td>0.23</td>
<td>0.34</td>
<td>4.83</td>
</tr>
<tr>
<td>No. of levels fused</td>
<td>−0.02</td>
<td>0.89</td>
<td>−1.58</td>
</tr>
<tr>
<td>Intraop complications (no/yes)</td>
<td>−0.06</td>
<td>0.87</td>
<td>3.89</td>
</tr>
</tbody>
</table>

* p < 0.05.
† 1 = congenital, 2 = deformity, 3 = trauma, 4 = tumor.
tive scores for the PRO instruments and were unable to define the change in PRO scores after surgery. The ASIA grade has been previously used to define the neurological status in patients undergoing occipitocervical fusion. Although this scale is more appropriate for patients who have sustained a spinal cord injury, it does provide a standardized tool to evaluate neurological deficits for these patients. The instruments used in this study were not disease specific and possibly did not address the specific problems associated with occipitocervical fusion. While the SRS-22, SF-36, and NDI have been used in a few prior pediatric studies, these scales are not ideal for children. Additionally, missing data for the SF-36, NDI, and Child-PedsQL forms may have limited the ability to achieve statistically significant associations in our analysis. Currently, a concise, validated disease-specific HRQOL scale for craniovertebral junction disorders in children is lacking. The present study used the most widely used scales—SRS-22, NDI, SF-36, and the PedsQL—to describe the quality of life outcomes in this patient population; yet, there is a need to develop scales that are sensitive to the specific issues concerning these children. In this study, we have provided an early description of quality of life outcomes in this patient population and expect to carry this study forward by implementing a disease-specific scale to assess outcomes for children undergoing craniovertebral junction surgery.

Conclusions

Occipitocervical and atlantoaxial fusions for the treatment of craniovertebral junction disorders in children are relatively safe and effective procedures. They are associated with acceptable PROs and HRQOL scores, considering the serious nature and potential for morbidity in this patient population. However, longer follow-up and disease-specific scales are necessary to elucidate the full impact of occipitocervical and atlantoaxial fusions on children.

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

Author Contributions
Acquisition of data: Vedantam, Hansen, Briceño, Brayton. Analysis and interpretation of data: Jea, Vedantam. Drafting the article: Jea, Vedantam. Critically revising the article: Jea. Reviewed submitted version of manuscript: Jea. Study supervision: Jea.

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