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Rib Reconstruction of the Absent Mandibular Condyle in Children

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

Abstract

Objectives. To describe pediatric costochondral graft reconstruction of the absent mandibular condyle and to report the short-term and long-term outcomes and complications associated with performing this procedure in young children.

Study Design. Case series with a retrospective chart review.

Setting. Pediatric otolaryngology clinic and tertiary children’s hospital in a metropolitan area.

Subjects and Methods. All children treated for an absent mandibular condyle with a costochondral graft at Children’s Hospitals and Clinics of Minnesota were identified from 2002 through 2011, and a retrospective chart review was performed.

Results. Ten patients aged 3 to 11 years were identified. The most common diagnosis, in 8 of 10 patients, was oculo-auriculo-vertebral syndrome. Three of the patients had a tracheostomy, of which 1 was decannulated following condylar reconstruction. Functional improvement, defined as improved symmetry, chewing, or better oral opening, was observed in 8 of 10 patients. Five patients have required no further surgeries to date, with a mean follow-up time of 3.9 years. Severe overgrowth of the graft was noted in 1 case, and partial or complete resorption of the graft was also noted in 3 cases. Overgrowth occurred after 5.7 years, whereas resorption occurred after an average of 2.5 years.

Conclusions. Costochondral grafts are an excellent surgical treatment option for children with severe mandibular malformations. Short-term results show particular improvement in function and mandibular alignment. The mean follow-up time with no revision surgery was substantial and indicates that rib grafting is a good addition to the armamentarium of treatment for this patient population.

Keywords

pediatric, reconstruction, costochondral graft, mandibular defect, oculo-auriculo-vertebral syndrome, Goldenhar syndrome, Hemifacial microsomia

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Oculo-auriculo-vertebral spectrum (OAVS) is the second most common congenital craniofacial anomaly, with only cleft lip and palate occurring more frequently. Despite this, it is a relatively rare condition, affecting approximately 1:5600 live births.¹ Oculo-auriculo-vertebral spectrum is also referred to as hemifacial microsomia (HFM) and includes Goldenhar syndrome. These are a very diverse grouping of abnormalities thought to be caused by problems in the development of the first and second branchial arches. Specifically, these conditions are likely due to a vascular event involving the stapedial artery in early embryogenesis.² This can cause hypoplasia of the major facial bones and soft tissues, as well as malformation of the external and internal auricles and vertebral defects. One major challenge for children with OAVS can be breathing and feeding problems caused by facial asymmetry. These asymmetries can result from several disorders including cranial nerve VII paresis, maxillary deficiency, or the unilateral absence of a mandibular condyle.

Some authors have argued that it is important to diagnose and treat OAVS early, as it may be a progressive process that can lead to increased asymmetry and disability over time.³ More commonly, the primary purpose of reconstruction is to decrease disability and improve oral function and appearance. To ensure that pediatric patients are able to achieve as much function as possible, and to decrease the strain of caring for these patients on their families, it is necessary to create a functional mandibular condyle. Artificial prostheses are generally not appropriate for young patients, as their fixed dimensions cannot adapt to the maturing
skeleton. Historically, correction of the more severe deformities has been achieved with an autogenous costochondral bone graft (CCG). Despite this, there are few reports of this procedure in a young pediatric population. The objective of this article is to provide a review of the authors’ experience with this treatment and describe the outcomes experienced by these patients.

Methods
A retrospective review of all charts from patients seen in the Pediatric ENT Associates clinic and the Cleft and Craniofacial Clinic at Children’s Hospital of Minnesota from 2002 through 2011 was undertaken to identify all children treated with a CCG for an absent mandibular condyle. The Institutional Review Board of Children’s Hospital and Clinics of Minnesota granted approval for this study. A total of 10 children were identified. Demographic information, as well as data regarding history of breathing or eating difficulties, facial asymmetry, and operative notes were extracted from the patients’ charts and entered into a database. Outcome measures such as tracheotomy decannulation and functional improvement, as well as incidence data, such as graft resorption and overgrowth leading to increased asymmetry, were also added to the database. Functional improvement was defined as any mention of improved oral opening or asymmetry by the provider or subjective ease of food ingestion by the patient or their caregiver in the patient’s postoperative medical records. Patients were followed as long as possible, until they were lost to study or required other major reconstructive surgery.

Description of Operative Procedure
The patients underwent nasotracheal anesthesia. A suitable rib was palpated, outlined, and harvested. Then, a preauricular incision was made. Blunt dissection was performed in the direction of the expected course of the facial nerve with nerve integrity monitoring performed. Soft tissues were released posteriorly to identify the site of the primitive glenoid fossa, and it was evaluated for depth and lip. If it was not adequate, a round, diamond burr was used to hollow out and shape the fossa to accept the new condyle. Next, a small submandibular incision was created, and careful dissection to expose the mandible and preserve the marginal mandibular branch of the facial nerve was performed. The periosteum was incised along the inferior border of the mandible, and wide subperiosteal dissection was performed going in a superior direction until the edge of the malformed mandible was exposed on the superficial surface. A tunnel was then created through the soft tissue connecting the preauricular site and the submandibular exposure of native mandible. A Penrose drain was passed between the 2 sites (Figure 1). Alternatively, an intraoral approach was also used in some patients. In these cases, an incision was made in the mandibular buccal vestibule with electrocautery down to the mandibular bone with exposure of the native body and ramus. The incision was expanded via blunt dissection to the preauricular incision and preserved with a Penrose drain.

Finally, the rib graft was shaped to resemble a condyle of appropriate length for optimal realignment, with approximately 2 mm of shaped cartilage remaining at the superior end (Figure 2). The graft was trimmed, and the periosteum was removed on the medial surface so that a bur could be used to create a small depression for the native mandible. The graft was placed in the correct position, and lag screws with washers were used to secure the graft to the mandible (Figure 3). A small piece of zygomatic subperiosteum was insinuated between the cartilage cap and glenoid fossa and sutured to the lateral surface of the graft. Stability of the mandible and adequate range of motion was ensured before irrigation and closure.

Results
Descriptive features of the 10 patients included in this review are presented in Table 1. There were 3 male and 7 female patients. The average age at the time of surgery was 6.5 years (range, 3-11 years). The most common diagnosis,
in 8 of 10 patients, was OAVS. The other diagnoses were unspecified mandibular malformation and histiocytosis.

Three of the patients considered in this study had a tracheotomy prior to the CCG reconstruction. Of these, 1 was decannulated following surgery. Functional improvement, defined as improved symmetry, chewing, or better oral opening, was observed in 8 of 10 patients at short-term follow-up visits (Figures 4 and 5). Five patients have required no further surgeries to date, with a mean follow-up time of 3.9 years (range, 9 months to 8 years).

Five patients have required further surgery after the initial CCG reconstruction. One patient required mandibular distraction osteogenesis to improve continued micrognathia at 4 years postoperation. Severe overgrowth of the graft was noted in 1 case after 5.7 years. This same patient experienced improved oral opening and chewing at 1-year follow-up. Partial or complete resorption of the graft was also noted in 3 cases after an average of 2.5 years. Only 1 of these patients experienced no functional benefit from treatment, whereas the other 2 showed improved oral opening after surgery.

**Discussion**

Oculo-auriculo-vertebral spectrum (HFM) is a congenital condition exhibiting diverse phenotypic variation. One classification system devised by Pruzansky et al in 1969 further subdivides HFM into 3 subtypes: type I HFM is characterized by hypoplasia of the mandible, type II HFM indicates hypoplasia and malformation of the condyle or coronoid process, and type III HFM involves a complete or partial loss of the mandibular ramus, condyle, or glenoid fossa. Micrognathia leading to feeding and breathing difficulty in the newborn is a common symptom of HFM requiring treatment. The authors have previously commented on several different treatment options of varying complexity in proportion to the extent of dysgenesis. Ideally, simple prone positioning of an infant or a nasopharyngeal airway is adequate to ensure a patent airway in neonates suffering with this condition. However, in more severe type I and II malformations, a surgical intervention such as mandibular distraction osteogenesis (MDO) can be performed to accomplish airway patency without resorting to a tracheotomy. In type III HFM, the lack of a native condyle makes MDO impractical until a functional joint is created. In this most severe form of the syndrome, a new condyle is often constructed from autogenous bone. If necessary, additional reconstructive surgery such as MDO can be implemented later to resolve any continuing functional deficits. Definitive craniofacial reconstruction is not attempted until skeletal maturity, but early surgical intervention in children can prevent many of the morbidities associated with a tracheotomy.

In 1968, Ware and Taylor documented surgical treatment using autogenous rib grafts to replace missing mandibular condyles. This treatment methodology was not immediately accepted and can still be contentious to this day. Some authors argued that it was not necessary to reconstruct the

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**Table 1. Demographics and patient outcomes.**

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
<th>8</th>
<th>9</th>
<th>10</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at procedure, y</td>
<td>8.6</td>
<td>4.3</td>
<td>5.8</td>
<td>5.1</td>
<td>5.1</td>
<td>7</td>
<td>3.5</td>
<td>11.8</td>
<td>4.7</td>
<td>8.7</td>
</tr>
<tr>
<td>Gender</td>
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<td>Male</td>
<td>Female</td>
<td>Female</td>
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<td>Female</td>
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</tr>
<tr>
<td>R, L, bilateral</td>
<td>Left</td>
<td>Right</td>
<td>Left</td>
<td>Left</td>
<td>Left</td>
<td>Right</td>
<td>Right</td>
<td>Right</td>
<td>Left</td>
<td>Right</td>
</tr>
<tr>
<td>Diagnosis</td>
<td>OAVS</td>
<td>Histiocytosis</td>
<td>OAVS</td>
<td>OAVS</td>
<td>OAVS</td>
<td>OAVS</td>
<td>OAVS</td>
<td>OAVS</td>
<td>OAVS</td>
<td>OAVS</td>
</tr>
<tr>
<td>Tracheotomy decannulation</td>
<td>No</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>Yes</td>
<td>No</td>
<td>—</td>
</tr>
<tr>
<td>Functional improvement</td>
<td>No</td>
<td>Yes</td>
<td>—</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Time to overgrowth, y</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>5.7</td>
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<tr>
<td>Time to resorption, y</td>
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<td>—</td>
<td>4.5</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>1</td>
<td>—</td>
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<tr>
<td>Years since procedure</td>
<td>8.8</td>
<td>4.0</td>
<td>0.8</td>
<td>6.5</td>
<td>1.6</td>
<td>8.4</td>
<td>9.3</td>
<td>7.5</td>
<td>8.3</td>
<td>6.0</td>
</tr>
</tbody>
</table>

Abbreviation: OAVS, oculo-auriculo-vertebral spectrum.
temporomandibular joint after a condylectomy. They believed that the mandible would continue to grow without a condyle and that a pseudoarthrosis would be functional, as is seen in cancer resections in adults. However, in young children, the mandible plays a very important role in cranial skeletal development. Mature facial symmetry and proper tooth positioning are dependent on proper jaw alignment and force distribution. Today, many agree that early manipulation and correction of the mandible is crucial for symmetric growth and development of the facial skeleton during childhood.

Our results support the position that it is advantageous to act early to correct a malformed mandible with a missing condyle using a CCG. Functional improvement was observed for 80% of patients treated this way. One of the 2 patients who did not have documented functional improvement went on to have mandibular distraction osteogenesis less than a year later as part of an overall facial reconstruction plan. These patients had improvement of symptoms in the form of increased oral opening and facial symmetry. For some of the patients, this led to an increased ability to chew solid foods and in 1 case allowed decannulation of a tracheotomy.

There have been concerns with graft overgrowth and the predictability of the results since the early days of this procedure. In 1991, Ware and Brown published a case series of 10 pediatric patients in which 4 patients experienced substantial overgrowth. Guyuron and Lasa also experienced difficulty with growth predictability in their series of 8 patients, 4 of whom experienced overgrowth and 3 who did not grow at all. Much of this difficulty has been addressed by carefully trimming the amount of cartilage on the grafts. Perrott et al hypothesized that the size of the cartilage cap on the graft is directly related to its growth. Their series identified 26 patients who were treated with CCGs using 2 to 4 mm of cartilaginous end cap and had no clinically significant linear overgrowth. In our experience, there was only 1 CCG that experienced excessive linear overgrowth. This did not occur until almost 6 years after the surgery, when the patient was 17 years old. This overgrowth is not necessarily unexpected as hormonal changes through puberty can cause rapid bone growth. This is why, as mentioned above, it is common to delay definitive orthognathic reconstruction until skeletal maturity.

The young age of our patients at the time of surgery is likely to be one reason for our positive results. Other authors have also found that earlier treatment is likely to correspond with a better outcome. Ross reported an 80% success rate when CCG was performed in children 3 to 9 years old. However, his success dropped to only 50% when attempted on patients older than 14 years. Not all agree that earlier treatment results in improved outcomes, as noted by Padwa et al. They stated that older patients were more likely to have a positive result. This could simply be due to a difference in definition of success, as they were measuring postoperative occlusal cant as opposed to functional improvement. Also, the group labeled as unsuccessful had more significant deficits preoperatively and were only moderately younger than the successful group (mean of 5.8 vs 6.7 years).

One alternative to CCG for severe mandibular malformations is the use a vascularized free fibular flap. Foster et al published a large series in 1999 that showed good results with both nonvascularized and vascularized bone grafts. However, this was primarily an older adult population, and almost all of the vascularized grafts were used in patients treated for malignant disease. Guo et al also reported success with free flaps in a series of 16 pediatric patients. However, they commented that it is a very technically demanding surgery and should be used for only severe cases, such as those with defects larger than 6 cm, irradiated wound

Figure 4. Image showing patient 3 before the procedure.

Figure 5 Image showing patient 3 three months after the procedure.
beds, or failed previous bone grafts. Also, they mention that no growth was noted in any of the cases, which may necessitate additional future bone transfer. Another option that has been recently proposed is performing MDO in patients with no functional mandibular condyle and allowing a pseudoarthrosis to occur between the mandibular remnant and skull base. However, these patients were selected because of contraindications to rib grafting, and there are no long-term results to indicate this would be a preferable technique. It is possible that a pseudoarthrosis will not prove as durable and functional as a CCG.

Conclusion
Costochondral grafts are an excellent surgical treatment option for children with severe mandibular malformations. Short-term results show particular improvement in function and mandibular alignment. This can greatly improve the patients’ quality of life and opportunity for positive development in the formative years. Progressive improvement of facial symmetry and alignment until skeletal puberty can only enhance the final product of definitive orthognathic reconstruction. Mean follow-up time with no revision surgery was substantial, which reinforces the durability and effectiveness of this procedure. However, future study might focus on the results of patients once their craniofacial features have reached maturity.

A limitation of our study is that we attempted to quantify, through a retrospective review, subjective measures of improvement in patients’ lives. Our assertions could be bolstered by new research using prospectively collected data from patients, including objective measures of facial alignment and oral opening. Despite this, the authors feel that our data, along with much of the current literature, indicates that rib grafting is a good addition to the armamentarium of treatment for this patient population.

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Author Contributions
Derek Goerke, data acquisition and analysis, article drafting and revisions, and final approval; Daniel E. Sampson, manuscript conception, critical revisions, and final approval; Robert J. Tibesar, manuscript conception, critical revisions, and final approval; James D. Sidman, manuscript conception, design, critical revisions, and final approval.

Disclosures
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