Use of External Distractors and the Role of Imaging Prior to Mandibular Distraction in Infants With Isolated Pierre Robin Sequence and Stickler Syndrome

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IMPORTANCE Computed tomographic (CT) scans are often obtained before mandibular distraction osteogenesis in patients with isolated Pierre Robin sequence. There is concern regarding the risk of radiation exposure from CT in children.

OBJECTIVE To evaluate whether preoperative CT is necessary for adequate airway, feeding, and aesthetic outcomes following mandibular distraction with external distraction devices in infants with isolated Pierre Robin sequence.

DESIGN, SETTING, AND PARTICIPANTS In a retrospective review of medical records, infants who underwent mandibular distraction between January 1, 1998, and September 30, 2014, at 2 tertiary children’s hospitals were identified using procedure codes. Data analysis was conducted December 1, 2014, to March 31, 2015. Fifty-two patients fit the inclusion criteria of isolated Pierre Robin sequence or Stickler syndrome, of being younger than 9 months at the time of distraction, and of use of external distractors. Forty-two of these infants did not receive preoperative CT imaging.

EXPOSURE Mandibular distraction osteogenesis for isolated Pierre Robin sequence or Stickler syndrome.

MAIN OUTCOME MEASURES Number of infants who were able to avoid tracheostomy or achieve decannulation, who were able to avoid placement or achieve removal of a gastrostomy tube, and in whom there were no intraoperative complications, no open-bite deformity, no malocclusion, no asymmetry, and no postoperative complications.

RESULTS In comparison with the 10 infants who underwent preoperative CT, all 42 of the infants (100%) who did not receive preoperative CT imaging successfully avoided tracheostomy or achieved decannulation (P = .04) and 40 patients (95%) did not require placement of a gastrostomy tube or were able to undergo gastrostomy tube removal postoperatively (P < .001). There were no significant differences between the CT and non-CT groups in the other 5 outcome measures. Two patients (5%) required postoperative gastrostomy tube placement, 2 patients (5%) had minor intraoperative complications that might have been anticipated with CT, 2 patients (5%) demonstrated malocclusion, and 1 infant (2%) had asymmetry at the end of the distraction phase.

CONCLUSIONS AND RELEVANCE This series suggests that the absence of preoperative CT does not compromise functional or aesthetic outcomes in mandibular distraction with external distraction devices in infants with isolated Pierre Robin sequence or Stickler syndrome. This finding has implications for cost containment and reduction of radiation exposure to a vulnerable population.

LEVEL OF EVIDENCE 4.
Pierre Robin sequence (PRS) was first described in 1923 as a triad of micrognathia, glossoptosis, and airway compromise; it was later noted that a U-shaped cleft palate is also commonly present. Some controversy over the exact definition still exists, but the sequence is recognized to occur in approximately 1 of 8000 live births. Pierre Robin sequence can be an isolated event; however, up to 80% of patients with PRS have an associated syndrome, with the most common being Stickler and velocardiofacial syndromes. Infants with PRS may have symptoms of respiratory obstruction and feeding difficulty, with a wide range of severity. Most commonly, respiratory obstruction can be managed with prone positioning alone; however, some children develop severe symptoms and their condition can deteriorate rapidly. Tracheostomy, previously considered the conventional alternative, is also commonly present. Some controversy over the exact sequence can be an isolated event; however, up to 80% of patients with PRS have an associated syndrome, with the most common being Stickler and velocardiofacial syndromes. Infants with PRS may have symptoms of respiratory obstruction and feeding difficulty, with a wide range of severity. Most commonly, respiratory obstruction can be managed with prone positioning alone; however, some children develop severe symptoms and their condition can deteriorate rapidly. Tracheostomy, previously considered the conventional alternative, is also commonly present. Some controversy over the exact

In 1995, Codivilla introduced the idea of distraction osteogenesis, which was later refined and popularized by Ilizarov and Ledyav. In 1992, McCarthy et al described the use of craniofacial distraction osteogenesis to correct mandibular hypoplasia, and Moore et al later described the use of mandibular distraction osteogenesis (MDO) to alleviate upper airway obstruction due to glossoptosis. In 2001, Sidman et al reported their specific techniques for MDO in infants and young children; similar results were demonstrated by other authors. Since then, MDO has become an accepted method among otolaryngologists and craniofacial surgeons for alleviating upper airway obstruction, avoiding tracheostomy, and restoring feeding ability in infants with PRS.

Many institutions consider computed tomographic (CT) imaging to be a standard step for preoperative planning for MDO. Indications cited for preoperative CT include planning for osteotomies and vectors of distraction, locating the inferior alveolar nerve and inferior tooth buds, and assessing the temporomandibular joint. We do not routinely obtain preoperative CT scans for infants with isolated PRS (iPRS) or Stickler syndrome who undergo MDO. It is our practice to only use externally applied, multidirectional distraction devices in this patient population. We hypothesize that there is no significant difference in outcomes between patients who receive preoperative CT and those who do not when multivector external distractors are used in the iPRS and Stickler syndrome populations.

Results

Fifty-two infants with iPRS or Stickler syndrome underwent MDO with externally applied, multidirectional distraction devices during the study period. Mean age at MDO was 35 days (range, 6 days to 7 months). Follow-up ranged from 1 to 8 years. Forty-two patients did not receive preoperative imaging and 10 received preoperative CT with 3-dimensional reconstruction. Comparison of the 2 groups yielded statistically significant differences in results within 2 of 7 outcome measures for our definition of PRS: airway and feeding (Table 1). The infants with iPRS who did not undergo preoperative CT had higher success rates in both of these categories (Figure).

Airway Outcomes

Successful airway outcomes as defined by avoiding tracheostomy or achieving decannulation were significantly increased (P < .05) in the patients without preoperative imaging. Of the 42 patients who did not receive preoperative imaging, none (100%) required tracheostomy. Of the 10 patients who received imaging, 8 (80%) did not require tracheostomy, 1 (10%) was decannulated, and 1 (10%) was emergently intubated on postoperative day 9 and received a tracheostomy on postoperative day 14.

Feeding Outcomes

Successful feeding outcomes, as defined by avoiding gastrostomy tube placement or transitioning to oral intake, were significantly increased (P < .05) in the group without preoperative imaging. Of the 42 patients who did not receive imaging,

Methods

Approval was obtained from the Children’s Hospitals and Clinics of Minnesota and Tufts Medical Center institutional review boards for both study design and waiver of informed consent. Patient data were stored on a secure server at each institution and deidentified prior to analysis. Only deidentified data were shared between institutions. Eighty-nine infants who underwent MDO at Children’s Hospitals and Clinics of Minnesota from January 1, 1998, to September 30, 2014, were identified from a database using procedure codes, and 15 infants who underwent MDO at Tufts Medical Center from November 1, 2010, to September 30, 2014, were identified using a similar departmental database. Patients were eligible for the study if they underwent MDO during infancy with multidirectional external distraction devices and had a diagnosis of Stickler syndrome or iPRS as defined by cleft palate (any form), micrognathia, glossoptosis, and upper airway compromise. Patients were deemed ineligible for the study if they underwent MDO with submerged distraction devices, were older than 9 months, had an additional underlying syndrome, or did not fulfill the above criteria for our definition of PRS (ie, if they had micrognathia without cleft palate). Forty-three infants were identified as eligible for the study from Children’s Hospitals and Clinics of Minnesota and 9 patients were identified from Tufts Medical Center. A retrospective review of the medical records was performed on all 52 patients, including operative reports, hospital progress notes, and clinic follow-up appointments.

Statistical analysis was performed using a χ² test for independence in a 2 × 2 contingency table for each outcome measure with level of significance (α) equal to .05 and 1 df. Calculations for power and sample size were performed (Microsoft Excel, version 14.4.4; Microsoft Corp), with level of significance equal to .05, 1 df, standard upper limit of infinite sum, m = 40, and standard power (1 – β) equal to 0.8 or 80%. Data analysis was conducted from December 1, 2014, to March 31, 2015.

Table 1: Outcomes

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<thead>
<tr>
<th>Category</th>
<th>Preoperative CT</th>
<th>No Preoperative CT</th>
<th>P Value</th>
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<td>Successful outcomes (%)</td>
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<td>Airway outcomes</td>
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<td>Feeding outcomes</td>
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2 (5%) required postoperative gastrostomy tube placement: one inserted 8 days after distractor removal and the other on the same day of distractor removal.

Of the 10 patients who received preoperative imaging, only 5 (50%) did not require a gastrostomy tube or were able to transition to oral feedings after MDO. One patient had a gastrostomy tube inserted before surgery and was unable to transition to oral feeding, 1 patient had a gastrostomy tube inserted 15 days after distractor removal, and 3 patients underwent surgical gastrostomy prior to completion of the distraction phase.

**Intraoperative Complications**

There was no significant difference in the intraoperative complications between patients who received preoperative imaging vs those who did not. Of the 42 patients who did not receive imaging, 2 (5%) had minor intraoperative complications that might have been anticipated with preoperative CT. In one patient, a fracture between 2 posterior pins occurred and both pins were repositioned where there was better-quality bone stock. The other patient required a single-pin repositioning owing to poor stability of the pin in the mandible due to either poor bone stock or the presence of a tooth follicle. None of the 10 patients who received preoperative imaging had intraoperative complications.

**Postoperative Occlusion**

There was no significant difference in the rates of malocclusion, asymmetry, or open-bite deformity occurring immediately after the distraction phase in patients who received preoperative imaging vs those who did not. Of the 42 patients who did not receive imaging, 1 (2%) demonstrated malocclusion at the end of the distraction phase, described as a 2- to 3-mm class II occlusion at the time of device removal. One patient (2%) had asymmetry at the end of the distraction phase, described as a relative paucity of mandibular mass on one side without malocclusion in the context of underlying congenital muscular torticollis. Four other patients (10%) had later clinic follow-up notes that described malocclusion. One patient (2%) had a class II occlusion first noted at 11 months after distractor removal and then a slight class III pattern at 23 months. Another patient (2%) had a slight class III pattern noted at 6 months after distractor removal. Two other patients (5%) had class II occlusions noted several years postoperatively. In 2 patients (5%) there was asymmetry detected during the activation phase due to unilateral device failure. One of these patients required replacement of one device; both had resolution of asymmetry by the end of the activation phase.

Of the 10 patients who received preoperative imaging, only 1 individual (10%) demonstrated malocclusion at the end of the distraction phase. This patient had severe temporomandibular joint ankylosis after MDO; however, successful decannulation was achieved. There was no underlying syndrome identified during infancy, but this patient had a subsequent microarray analysis later in childhood and ultimately a 13:18 chromosomal translocation of unknown significance was identified. Since this translocation is not associated with any known
syndrome or craniofacial condition, we chose to include this patient in our analysis.

In all infants, dental follicles were noted in the region of the osteotomies. The inferior alveolar nerves were visualized in their expected location within the path of the osteotomy in all patients.

Postoperative Complications
There was no significant difference in the rates of postoperative complications in patients who received preoperative imaging vs those who did not. Of the 42 patients who did not receive imaging, 6 (14%) had postoperative complications, 2 of which may have been prevented or mitigated by obtaining preoperative CT. One patient (2%) had bilateral hardware loosening with symmetric mandibular regression; postdistraction malocclusion was not described in subsequent follow-up notes. The second patient (2%) had residual glossoptosis that resulted in prolonged postoperative intubation and pseudomonal tracheobronchitis requiring antibiotic treatment. This patient was successfully extubated 14 days after distractor removal. The other 4 patients (10%) had postoperative complications that likely would not have been prevented with preoperative imaging. Two patients (5%) developed postoperative infections, 1 involving a local pin site infection that was treated with antibiotics and the other with a methicillin-sensitive \textit{Staphylococcus aureus} infection of one distractor during the consolidation phase, which caused loosening and necessitated early removal. No problems with asymmetry or malocclusion resulted for either patient. One patient (2%) had slight unilateral marginal mandibular weakness noted after distractor removal and 1 patient (2%) had ongoing aspiration for which direct laryngoscopy and bronchoscopy could not identify an anatomic cause. There were no known instances of subjective sensory deficits in the distribution of the mental nerve in any of the participants. During the follow-up period (range, 1-8 years), normal sensation was documented in older children (>3 years) who were able to communicate lip and chin sensation on physical examination; however, objective data on detection of more subtle sensory deficits were never obtained.

Of the 10 patients who received preoperative CT, 2 (20%) developed postoperative complications. One patient (10%) had residual postoperative glossoptosis that did not allow decannulation. The other patient (10%), mentioned above, had severe bilateral temporomandibular joint ankylosis that resulted in gastrostomy-tube dependence.

Sample Size and Power Calculations
Sample size and power calculations were performed for each of the 7 outcome measures (Table 2). Only 1 of the outcome measures (successful avoidance or removal of gastrostomy tube) had a sufficient sample size to detect a standard statistical difference of 20% or less (power, 0.8). Excluding the category of open-bite deformity (which had 0% power because both CT and non-CT populations had a 0% rate of deformity), the remaining 5 outcome measures were powered from 7% to 54% (Table 2).

Discussion
The definition of PRS used in this study is the triad of micrognathia, glossoptosis, and cleft palate. We excluded “syndromic” PRS patients in our analysis because these individuals often have cognitive or neurologic abnormalities that complicate postoperative recovery and may influence functional outcomes. We also excluded patients with isolated micrognathia and no cleft palate. Patients with Stickler syndrome were included despite technically falling in the syndromic PRS category because these patients represent a significant subset of the PRS population and have largely unaffected neurologic systems and no additional temporomandibular joint abnormalities. Moreover, many children with Stickler syndrome are initially thought to have iPRS and the disorder is diagnosed later in childhood when additional syndromic features (myopia and flattened facies) become apparent and single gene testing is pursued.

Although some institutions may consider CT to be a standard step for preoperative planning of mandibular distraction osteogenesis, we do not routinely obtain preoperative imaging for patients with iPRS or Stickler syndrome. Our data suggest that in 5 of 7 outcome measures, there appeared to be no significant difference between patients who received preoperative imaging and those who did not (Table 1). However, the rates of long-term tooth loss or damage to secondary molars caused by the mandibulotomies were not specifically collected in this study. Prior outcomes studies examining similar surgical techniques report a long-term dental complication rate of approximately 20%, which may include malformed or absent second mandibular molars on either or both sides of the mandible.

In the areas that yielded statistically different results—upper airway and feeding outcomes—the patients without...
preoperative CT had higher percentages of success than those who received preoperative imaging (100% successful upper airway outcomes in non-CT patients vs 90% in CT patients, and 95% success with oral feeding in non-CT patients vs 50% in CT patients). Possible confounding factors contributing to this difference include increased patient complexity and less surgical experience with earlier MDO procedures. Of the patients at our institutions who received preoperative imaging, most cases occurred either prior to 2005 when MDO was an emerging procedure or there was clinical suspicion for additional anatomical abnormalities. The patients who underwent CT scanning may have been imaged because of a clinical instinct by the surgeon, regardless of an otherwise negative medical workup for the presence of a syndrome. The merits of this clinical intuition cannot be proven but may be supported by the fact that preoperative imaging seemed to correlate with an overall poorer functional outcome among the patients in this study.

Preoperative imaging may be helpful early in a surgeon’s experience with MDO to identify good bone stock for osteotomy and pin site planning. However, with practice and increased comfort with this procedure, imaging is not required to identify these landmarks because, by definition, there are no additional anatomic abnormalities in iPRS cases. Patients with Stickler syndrome have only mild dysmorphic features that spare the remainder of the mandible, and there is no need to assess for temporomandibular joint abnormalities in this population either. Our data suggest that there is no difference in intraoperative complications between patients who receive preoperative CT and those who do not (0% vs 5%, respectively). However, syndromic PRS patients may have additional temporomandibular joint or mandibular deformities that justify preoperative assessment with imaging, especially if the syndrome is poorly characterized.

Additional indications cited for preoperative imaging include identifying other key points of anatomy, such as the inferior alveolar nerve and inferior tooth buds. Although we did not specifically track inferior alveolar nerve complications in this study, we cite a previous report of zero identified subjective deficits with direct sensation testing in the mental nerve distribution in children old enough to test. We recognize that it is difficult to assess neurosensory deficits in the neonatal period; however, we have noticed anecdotally that even children with known inferior alveolar nerve transaction report normal sensation years later. Long-term outcomes of molar development are also poorly defined and of unclear clinical significance. Another previous report described an overall incidence of 16% tooth malformation, loss, or dentigerous cyst formation in patients with a mean follow-up interval of 5.6 years, most of which were discovered incidentally on routine dental radiographs. A review of 40 articles on MDO complications reported a rate of 22.5% intraoperative tooth injury, of which up to 71% resulted in migration of dental buds, molar bud perforation, dental root injuries, or dentigerous cysts within the first 1 to 2 years postoperatively. Further research on long-term dental outcomes is merited, but our current rates of long-term tooth morbidity appear to be comparable to those of other limited reports.

We only use multidirectional, external distraction devices in the neonatal population. These distractors can be easily adjusted as needed during daily progress checks throughout the inpatient postoperative recovery period and in follow-up clinic appointments. Therefore, CT is not required to plan precise distraction vectors when using external distractors. Our data show that there was no significant difference between the CT and non-CT group in terms of malocclusion (10% vs 2.4%), asymmetry (10% vs 2.4%), or open-bite deformity (0% in both groups) at the end of the distraction phase.

However, preoperative CT imaging may have increased benefit when a surgeon uses unidirectional, submerged distractors. Proponents of submerged distractors often cite reasons such as decreased rates of scarring compared with external distractors. However, because submerged distractors are unidirectional, they cannot be directly examined or adjusted postoperatively and therefore allow less room for error in planning and implementing the distraction vector. External distractors allow easy adjustment throughout the activation period and, as we have shown here, appear to allow the patient to forgo imaging radiation exposure without compromising alignment. The secondary procedure of hardware removal is simple with the use of external devices; pin removal takes minutes and can be done with or without sedation. There is no risk of bleeding, neuropraxia, or infection as compared with the secondary procedure required when using a buried distraction system.

Scarring is more substantial with external devices, but this often fades with age and proper incision care. In general, the rate of scar revision following neonatal mandibular distraction with external devices is low. Scott et al found that, although the rate of visible scarring approaches 16% with this technique, most parents choose to forgo scar revision.

In addition to the clinical costs of medical imaging, carcinogenesis has long topped the list of concerns regarding risks associated with ionizing radiation. It is well known that cancer risk increases with a lifetime accumulation of radiation exposure, and pediatric patients are believed to be at the greatest risk of developing a cancer related to medical imaging for several reasons. A child’s smaller size compared with an adult’s may result in a larger effective dose, and the rapidly dividing cells present in a growing child may be less able to efficiently repair mutations caused by radiation exposure compared with an adult. Children also have a longer lifespan during which the effect of radiation-induced mutations can manifest. These differences account for a sensitivity to imaging radiation more than 10 times greater than that of most adults. A recent systematic review estimated the risk of subsequent cancer associated with pediatric CT exposure. The estimated risk in the 10 years following CT exposure was 1 brain tumor per 10 000 children receiving a head CT when younger than 10 years. The data regarding maxillofacial- or otolaryngology-specific studies were limited but suggested a borderline significant increase in the risk of all combined cancers after facial CT (incidence rate ratio, 1.14; 95% CI, 1.01-1.28).
Conclusions

The risk of radiation exposure from CT exposure remains an active area of research, with initial data calling for decreasing unnecessary CT scans. This series suggests that preoperative CT imaging is not necessary to achieve consistently successful outcomes following MDO using multidirectional external distractors in infants with iPRS or Stickler syndrome. Although underpowered, these data show that, in 5 of 7 outcome measures there was no significant difference between patients who received preoperative imaging and those who did not. External, multi-vector distraction devices seem to allow avoidance of imaging without compromising postoperative alignment. This study has implications for cost containment and improved patient safety through reduction of medical imaging radiation exposure. A larger study would improve the quality of this comparison and we believe would strengthen our hypothesis.

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Study concept and design: Sidman, Sampson, Lander, Tibesar, Scott.
Acquisition, analysis, or interpretation of data: Mingo, Sidman, Scott.

Drafting of the manuscript: Mingo, Sidman, Tibesar, Scott.
Critical revision of the manuscript for important intellectual content: Mingo, Sidman, Sampson, Lander, Scott.

Statistical analysis: Mingo.

Administrative, technical, or material support: Sidman, Scott.

Study supervision: Sidman, Sampson, Lander, Tibesar, Scott.

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REFERENCES