Management of Type I and Type II laryngeal clefts: controversies and evidence

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Purpose of review
To summarize the pediatric Type I and Type II laryngeal cleft literature, paying special attention to recent trends, including evolution of surgical techniques, standardization of outcome assessments, and utilization of management algorithms.

Recent findings
There are a variety of options to choose from whenever considering Type I and Type II cleft repair, including endoscopic repair, transoral robotic surgery, and injection laryngoplasty. Conservative management including feeding therapy and treatment of comorbid medical conditions is recommended prior to repair. Validated outcome measures have arisen for swallow study interpretation and timing, as well as caregiver quality-of-life assessment. In addition, a series of medical algorithms have been proposed, which provide specific recommendations for diagnosis, treatment, and follow-up.

Summary
For clefts that fail conservative management, endoscopic repair has become the gold standard. In addition, injection laryngoplasty appears to provide both a diagnostic and therapeutic option in the management of these patients. Transoral robotic-assisted endoscopic repair appears well tolerated and feasible, although broader implementation of this technology remains limited. The development and refinement of best practice algorithms can help standardize management and improve decision-making. Furthermore, incorporating validated outcome measures, recorded and followed over time, will improve both patient care and research efforts moving forward.

Keywords
algorithm, laryngeal cleft, management, Type I, Type II

INTRODUCTION
Laryngeal clefts are abnormal posterior communications between the larynx, trachea, and esophagus. First described in 1792 by Richter, clefts are most commonly classified into four types as described by Benjamin and Inglis [1,2]. Type I is a supraglottic interarytenoid defect in which the cleft remains above the level of the vocal cords. Type II is a partial cricoid defect, extending below the level of the vocal cords (Fig. 1) [2]. Common presenting symptoms may include stridor, difficulty swallowing, choking with feeding, shortness of breath, chronic cough, recurrent lower respiratory infections, or failure to thrive [3,4]. Although high-grade clefts (i.e. Type III and Type IV) require operative intervention, the evaluation and treatment of low-grade clefts (i.e. Type I and Type II) remains controversial. This article reviews the pediatric Type I and Type II laryngeal cleft literature, paying special attention to recent trends, including evolution of surgical techniques, standardization of outcome assessments, and utilization of management algorithms.

SURGICAL TECHNIQUES
There are a variety of options to choose from whenever considering low-grade laryngeal cleft repair, including endoscopic repair, transoral robotic surgery, and injection laryngoplasty. However, most surgeons agree that a trial of conservative
Conservative management, which includes feeding therapy (i.e. adjustments in liquid viscosity, solid consistency, equipment, strategies, and positioning) and treatment of comorbid medical conditions (e.g. gastroesophageal reflux disease) is warranted prior to surgical intervention.

For Type I and Type II laryngeal clefts that fail conservative management, endoscopic repair is considered the gold standard.

The outcomes of injection laryngoplasty (no improvement, temporary improvement, and durable improvement) provide valuable information about the functional significance of the laryngeal cleft and offer a diagnostic and therapeutic tool in the management of patients with Type I laryngeal cleft.

Validated outcome measures have arisen for swallow study interpretation and timing, as well as caregiver quality-of-life assessment, and can offer consistency both within and between clinical practices and research studies moving forward.

The development and refinement of best practice algorithms can help standardize management and improve decisions made in the delivery of healthcare.

Endoscopic repair

For Type I and Type II laryngeal clefts that fail conservative management, endoscopic repair has become the gold standard [6–12]. The first endoscopic cases were performed with nasotracheal intubation, with the laryngoscope positioned to hold the endotracheal tube into the anterior commissure [12]. Since that time, anesthetic techniques have evolved and working closely with anesthesia, it is possible to perform ‘tubeless’ surgery [6]. In particular, the requirements for anesthesia during cleft repair include: immobility, analgesia, lack of airway reflexes, adequate ventilation and oxygenation, avoidance of apnea, and full view of the laryngeal inlet [13]. Induction is achieved by either inhalation of sevoflurane in oxygen and nitrous oxide or intravenous administration of propofol (1–2 mg/kg) to induce unconsciousness while maintaining spontaneous respiration. During the procedure, an infusion of propofol (300 mcg/kg/min) and remifentanil (0.05–1.0 mcg/kg/min) is used to maintain the anesthetic plane [13]. The vocal cords are also anesthetized with topical lidocaine (2 mg/kg). Medication

FIGURE 1. Benjamin and Inglis classification. Diagram showing four types of cleft defined by the increasing extent of communication between the larynx, trachea, and esophagus. Reproduced with permission from Benjamin and Inglis [2].
dosage is titrated as needed by the anesthetist. Finally, if the patient becomes apneic, either side-port ventilation or controlled, intermittent intubation may be performed to maintain oxygen saturation until respiration returns [13].

Initial endoscopic repairs utilized sharp dissection with the development of small mucosal flaps, facilitating a double-layer closure [12]. Over time, denuding of the interarytenoid mucosa has been performed with carbon dioxide laser, instead of laryngeal microscissors [14–15]. The carbon dioxide laser is commonly delivered by a micromanipulator, although in some cases access may be limited because of poor visualization [6,15]. In 2013, Watters et al. [15] provided the first description of the flexible carbon dioxide laser for cleft repair. The flexible nature of the fiber allows off-axis delivery of the laser, which can provide better application of energy to the interarytenoid region [15,16]. In addition to laser, needle-tip electrocautery can be used to provide demucosalization [17]. Regardless of the technique, once the mucosa is removed, attention is then directed at reapproximation. Though many authors continue to advocate for two-layer closure [18–20], similar success has been shown with single-layer closure [6,17] (Fig. 2).

Thus, endoscopic repair can be achieved with spontaneous ventilation without the need for endotracheal intubation. Demucosalization of the cleft can be performed with a variety of devices (e.g. laser, electrocautery). In addition, tissue reapproximation can be achieved with single-layer closure.

**Transoral robotic surgery**

The desire to move toward minimally invasive surgery has provided the impetus for development and application of robotic procedures [21]. In 2007, Rahbar et al. used four pediatric cadaver larynges in the laboratory setting to determine the best set-up for a robot-assisted airway operation. Following this, robot-assisted endoscopic repair was attempted in five patients with Type I or Type II laryngeal clefts [21]. They were able to perform the repair in two patients, with the remainder aborted because of limitations in transoral access. The authors acknowledged that smaller instrumentation and further advances in device technology would likely facilitate the incorporation of robotic equipment into the field of pediatric otolaryngology [21].

Since Rahbar et al.’s work, advances in robotic technology have allowed miniaturization of instrumentation. As a result, efforts to explore transoral robotic-assisted cleft repair have resurfaced [22]. Leonardis et al. reviewed the charts of five children with Type I laryngeal cleft who underwent repair with robotic assistance. Swallow testing performed 4 weeks postoperatively showed complete resolution of

![FIGURE 2](image-url). Endoscopic repair of Type I laryngeal cleft. Diagram showing cleft before repair (a), cleft after demucosalization (b), cleft mid-repair (c), and cleft after repair (d).
penetration and aspiration in all patients. Although the authors noted promising results, they recognized the need to perform further studies comparing robotic versus endoscopic repair to address the cost benefit of using robotic technology [22].

Certainly, robot-assisted surgery has advantages over traditional endoscopic approaches, including: improved optics with three-dimensional visualization, tremor filtration, and increasing freedom of instrument movement [21]. Pediatric transoral robotic surgery has been shown to be well tolerated and feasible for a variety of airway diseases, including laryngeal cleft repair [20–23,24]. As advancements in technology broaden robotic applications, further work will be necessary to define the optimal role and indications for robotic-assisted cleft repair.

Injection laryngoplasty

Congenital defects, including both anatomic malformations and coincident diseases are very common in patients with laryngeal cleft [6,9,11,18]. As a result of these potential confounding conditions, it can be difficult to determine whether a cleft is a significant contributor to a patient’s symptoms. In 2000, Kennedy et al. [25] presented a small case series in which patients with a Type I laryngeal cleft underwent Gelfoam injection of the interarytenoid space at the time of diagnosis. All eight patients demonstrated clinical improvement after injection. Half of the patients had return of their symptoms, which correlated well with the known length of Gelfoam persistence in situ (approximately 6 weeks) [25]. The remaining half did not have return of symptoms at the time of their last follow-up.

As a result, the outcomes of injection laryngoplasty (no improvement, temporary improvement, and durable improvement) provide valuable information about the functional significance of the laryngeal cleft and suggests the best management option moving forward [26]. Without improvement, the patient is less likely to benefit from formal repair. In the case of temporary benefit, especially if the time interval correlates with the expected resorption rate of the implanted material, the patient may benefit from repeat injection or formal repair. With durable improvement, additional procedures may be avoided, although close clinical follow-up is warranted [26]. In respect to those that achieve a durable effect, multiple explanations exist, including: stimulation of an inflammatory response with resultant scarring, development of compensatory swallowing strategies, or improvement in neurodevelopmental maturity [25,26,27].

Investigators have shown well tolerated and effective augmentation of the interarytenoid space using a variety of injection materials, including Gelfoam [25], sodium carboxymethylcellulose aqueous gel [10,26,27], and hyaluronic acid derivatives [28] (Fig. 3). Although use of these substances is off-label, injection laryngoplasty appears to provide both a diagnostic and therapeutic tool in the management of patients with Type I laryngeal cleft.

OUTCOME ASSESSMENTS

Whenever examining the original literature on endoscopic repair and injection laryngoplasty, the postoperative evaluation of success generally relied on a historical assessment of symptom control and, sometimes, a repeat swallow evaluation [12,25]. Fortunately, over the past few years, efforts to improve the quality and safety of patient care has enhanced the standardization of practices in the evaluation and management of laryngeal cleft.

Modular Barium Swallow or Videofluoroscopic Swallow Study interpretation

A Modular Barium Swallow (MBS) or Videofluoroscopic Swallow Study (VFSS) is the most widely

![FIGURE 3. Injection laryngoplasty of Type I laryngeal cleft. Diagram showing cleft before (a) and after (b) injection of sodium carboxy-methylcellulose aqueous gel.](image-url)
utilized tool for assessing swallowing dysfunction in children. Recently, Strychowsky et al. [29] reviewed the MBS studies and medical records of 175 patients who underwent laryngeal cleft repair. A speech language pathologist reviewed every MBS study and classified types of swallowing impairment as follows: oral phase impairment, swallow triggering impairment, and pharyngeal phase impairment (including penetration and aspiration). In addition, the Penetration–Aspiration Scale and Functional Oral Intake Scale (FIOS) was determined for each study [30,31]. The Penetration–Aspiration Scale is an 8-point validated assessment tool that relies on the classification of the depth to which material passes into the airway and whether or not it is expelled [30]. The FIOS is a 7-point validated assessment tool that documents the functional intake of food and liquids, particularly noting the degree of diet modification and compensation that is required [31]. Compensation refers to changes in equipment (e.g. nipples), strategies (e.g. pacing), or positioning (e.g. side-lying).

The authors showed that laryngeal cleft patients experience impairment across all phases of swallowing. Significant improvement in swallowing function was observed in all of the surgically managed groups, as well as in Type I patients who underwent conservative management [29]. Although, some individual patients continued to have swallowing dysfunction to varying degrees. Furthermore, they noted that patients may have a normal study, yet exhibit other clinical signs of swallowing impairment, as the MBS may not capture intermittent periods of dysfunction [29]. Thus, by taking a broad and standardized approach to assessment, the MBS study can help to guide feeding therapy, surgical decision-making, patient and family counseling, and recommendations for evaluation of comorbid conditions.

**Modified Barium Swallow or Videofluoroscopic Swallow Study timing**

Despite the fact that swallowing evaluations are an integral component to the management of laryngeal cleft patients, this must be balanced against the risks of repeated radiation exposure. The ALARA principal (‘as low as reasonably achievable’) is the underlying principle for the well tolerated use of radiological studies [32]. Hersh et al. [33] examined the charts of 78 children who had undergone management for a Type I laryngeal cleft. The mean number of VFSS each child received during the course of treatment was 3.24 studies (range 1–10). The authors calculated the mean effective radiation dose per pediatric VFSS to be 0.16 mSv (range 0.03–0.59 mSv), which was 9.4 times that of a standard pediatric chest X-ray (0.017 mSv). As a result, over the entire course of treatment, patients undergoing management of Type I cleft reached an exposure equivalent to 30.6 chest X-rays [33]. This study highlighted the need for critical evaluation of management pathways in order to reduce the number of VFSS both preoperatively and postoperatively.

Following along with their previous work, Wentland et al. [34] sought to decrease the number of postoperative VFSS by modifying the timing of assessment. In particular, patients were divided into groups, depending on whether or not they had certain comorbidities (i.e. cardiorespiratory disease, congenital syndromes, or neuromuscular disorders). VFSS was not performed until 12 weeks postoperatively for patients with comorbidities, delaying the previous 6-week time point. Additionally, children without comorbidities and with overt clinical signs of aspiration were evaluated with a clinical swallowing evaluation with close monitoring of diet advancement, sparing assessment at the 6-week time point. Whenever the authors compared their results to that of an earlier cohort of similar patients, they saw a decrease in the number of postoperative VFSS from 1.22 to 1.03 [34]. Continued efforts to evaluate the timing for postoperative swallowing evaluation, in order to reduce the risk of repeated radiation exposure, while still supporting diet advancement should be encouraged.

**Quality-of-life assessment**

The traditional outcome measures used to describe laryngeal cleft consist of clinical evaluation and MBS/VFSS before and after repair. Until recently, there was a lack of data looking into the effect of laryngeal cleft and aspiration on the quality of life of affected children and their families [17]. Health-related quality of life provides an assessment of how the individual’s well being may be affected over time by a disease. Within the pediatric population, this is generally assessed using questionnaires that include the family experience, in addition to that of the child. In 2014, a new caregiver instrument, the Feeding/Swallowing Impact Survey (FS-IS), was designed to describe the quality of life of caregivers of children with aspiration [35]. Recently, Fracchia et al. [17] sought to cross-validate this instrument within patients with Type I laryngeal cleft. The survey has 18 questions that evaluate daily activities, worry, and feeding difficulties using a 5-point scale, in which lower scores indicate a better quality of life. The authors noted evidence of convergent validity, as children with improved VFSS after cleft repair exhibited significantly improved FS-IS scores,
compared with those children without improvement on VFSS postrepair. In addition, discriminant validity was also proven, as the average FS-IS score in caregivers of children with improved VFSS after repair was significantly lower than those of children who had not yet undergone repair [17*]. Therefore, the FS-IS appears to be a valid metric for assessing the impact of aspiration on caregivers of children with laryngeal cleft.

**MANAGEMENT ALGORITHMS**

Medical algorithms based on best practice can help standardize diagnosis and treatment and improve decisions made in the delivery of healthcare. Chien et al. [9] were the first to propose a diagnostic and management algorithm that included MBS, functional endoscopic evaluation of swallow (FEES), and conservative therapy, prior to consideration of surgical repair. In 2011, Cohen et al. proposed an...
algorithm that included interarytenoid injection with absorbable material in patients who failed a trial of acid blockade and conservative measures. Repeat injection or formal closure was then considered in successful cases [26]. Ojha et al. [7] provided an update to the Chien et al. algorithm, with particular emphasis on the importance of multidisciplinary team assessment. In addition, they specifically recommended VFSS over FEES; particularly in patients less than 4 years old. Although, the authors did note that FEES may serve a unique role in identifying the pattern of spillage in select cases [7,36]. Finally, Ojha et al. provided their postoperative follow-up plan, including VFSS at 6 weeks, and depending on the results, further multidisciplinary team evaluation at 3-month intervals as needed.

Then, as the most recent update to their work, Wentland et al. [34] sought to reduce the total number of postoperative VFSSs. The multidisciplinary team recognized opportunities to both incorporate clinical evaluation of swallowing for healthy children with overt signs of aspiration and delay radiographic evaluation in children with a specific subset of comorbidities. In doing so, they noted a 43% reduction in the number of postoperative VFSSs obtained after cleft repair [34].

CONCLUSION

Although endoscopic repair remains the gold standard for management of Type I and Type II laryngeal clefts, the diagnostic and therapeutic role of injection laryngoplasty is evident. Though transoral robotic-assisted endoscopic repair appears well tolerated and feasible, broader implementation of this technology remains limited. Incorporating validated outcome measures (e.g. interpretation of MBS/VFSS, quality-of-life scales), recorded and followed over time, will improve both patient care and research efforts moving forward. Furthermore, the development and refinement of best practice algorithms can help standardize treatment and improve decision-making. A sample algorithm, incorporating evidence from prior studies and suggesting future management options is provided (Fig. 4).

Acknowledgements

None.

Financial support and sponsorship

None.

Conflicts of interest

There are no conflicts of interest.

REFERENCES AND RECOMMENDED READING

Papers of particular interest, published within the annual period of review, have been highlighted as:

* of special interest
** of outstanding interest

25. Describes the largest case series of pediatric transoral robotic surgery, including seven patients with laryngeal cleft, and confirms the feasibility and safety of repair. 

Provides the largest case series (n = 68) of Type I cleft patients undergoing injection laryngoplasty and further supports for its diagnostic and therapeutic role in management.


Presents a comprehensive approach to MBS interpretation, including utilization of validated outcome measures (i.e. Penetration–Aspiration Scale and Functional Oral Intake Scale).


Quantifies the amount of ionizing radiation received by children with laryngeal cleft during the total course of treatment and highlights the need to address this balancing measure with further research.


Discusses evidence-based modifications to a previous management algorithm and notes successful reduction in the number of VFSSs after implementation.
