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Single and Multi Stage Pediatric Laryngotracheal Reconstruction

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Introduction

Laryngotracheal reconstruction (LTR) is now accepted as the standard of care for established paediatric laryngotracheal stenosis. It can be adapted to address almost all laryngeal stenoses including Grade IV lesions, although it may not always be the most appropriate treatment. Cartilage graft augmentation is the tried and tested technique of airway reconstruction, which has now been used for over thirty years. A majority of paediatric otolaryngology departments will have the bulk of their experience with this technique. As in all airway surgery, decision making is at least as important as the actual surgery. This review therefore covers the aetiology and prevention of acquired laryngotracheal stenosis and discusses cartilage augmentation in classical (multistage) LTR with a covering tracheostomy and the more recently developed single-stage LTR. The reader is reminded that early lesions may be addressed endoscopically thus avoiding cartilage grafting altogether.

Acquired Laryngotracheal Stenosis

Aetiology

Paediatric laryngotracheal stenosis may occur in patients who have required intubation, often following premature birth. 90% of acquired subglottic stenoses have a history of intubation[6]. However, stenosis may develop after other forms of internal or external airway injury (TABLE 1). The endotracheal tube causes pressure necrosis and mucociliary stasis in the subglottic tissues leading to mucosal oedema and ulceration. The ulcer then deepens giving rise to exposed cartilage *(*FIGURE 1*)*, with subsequent infection and perichondritis which may progress to chondritis and cartilaginous necrosis. Granulation tissue typically forms in the areas of ulceration and fibrous tissue is deposited in the submucosa[11,14].

Histological studies have shown acute mucosal injury occurs invariably after intubation of the infantile larynx although injury progression is transient, with healing commencing within a few days with rapid improvement and completion of healing in most cases by 30 days[11]. The underlying aetiology of laryngotracheal stenosis is multifactorial and endoscopic studies have

not identified any consistent factors in the development of post-intubation subglottic stenosis in neonates[2]. Airway trauma may also be caused by tube movement in a restless patient, orotracheal tube placement or instrumentation from repeated intubations. Other sources of airway inflammation including nasogastric tubes[10] or local bacterial infection may compound the inflammatory response and subsequent fibrosis. Gastro-oesophageal reflux and systemic factors including chronic illness, immunosuppression and dehydration, also increase the susceptibility of the laryngeal mucosa to injury. A congenital element is present in some patients as a result of their anatomy. Infants with a smaller sized cricoid may have a tendency to develop stenosis following intubation.

Prevention

Neonatal care has significantly improved since the early 1970's, however laryngotracheal stenosis continues to occur in approximately 1% of paediatric patients after intubation. Low-irritant endotracheal tubes are now used, with the safest materials being polymeric silicone and polyvinyl chloride. Nasal endotracheal intubation can helps minimise tube movement. The parallel-sided straight tubes is preferableare generally preferred for long term neonatal intubation. It is important to avoid trauma during airway instrumentation and this is achieved via gentle tissue handling and preparation of the patient to provide relaxation at intubation. It is essential to use an appropriately sized tube which allows a leak at 20cm H_2O pressure. Although paediatric patients appear to tolerate longer periods of intubation than adults, airway injury and stenosis is still more likely after longer periods of intubation. A careful approach to surgery on the paediatric larynx is paramount and aggressive endoscopic interventions ought to be avoided in benign lesions with the use of staged procedures if necessary, for example in pathology involving the anterior commissure. Unavoidable high tracheostomies should be revised as soon as practical and tracheostomy should allow maximal preservation of native tracheal cartilage via the smallest tube possible to establish a safe, stable airway.

Treatment Options for Laryngotracheal Stenosis

Early lesions may be treated via medical therapy, endoscopic surgery or anterior cricoid split. (TABLE 2). Medical therapy includes treatment of any underlying conditions including infection or gastro-oesophageal reflux, which would hinder laryngeal recovery. Oral, intravenous or inhaled steroids and adrenaline nebulisers can all help optimise the airway. Endoscopic treatment is being increasingly utilised and is beneficial in addressing soft, immature and mild forms of stenosis. Techniques include cold steel, carbon dioxide and KTP laser and balloon dilatation. Mitomycin C may be used as an adjunct. Established laryngotracheal stenosis is best treated with cartilage augmentation laryngotracheal reconstruction (LTR), with cricotracheal resection being reserved for the most severe cases of airway stenosis.

Multistage vs Single-Stage Laryngotracheal Reconstruction

The development of the single stage procedure attracted substantial interest as it has several potential benefits over the traditional multi-stage LTR [5]. Tracheostomy insertion may be avoided or if present, the tube may be removed during the reconstruction. Tracheostomy closure with the reconstruction eliminates a potential source of infection whilst addressing the stoma site in the same procedure. The stages of airway stabilisation, healing and decannulation, which typically take several months in the traditional procedure, are compressed into a short period of postoperative endotracheal intubation, typically lasting around 7 days. The problems of longer-term stenting are avoided.

It must be acknowledged that although there is only a single open procedure, multiple subsequent endoscopic procedures are often required to monitor postoperative progress and further optimise the airway whilst healing occurs. Perioperative airway complications may occur and there is potential for unplanned extubation. There is also an inherent earlier reliance upon the newly reconstructed airway at planned extubation and if respiratory

compromise develops after extubation, the endotracheal tube may need to be replaced. Tracheostomy reinsertion may be required. Reintubation of these children is not always straight-forward and may potentially damage the cartilage reconstruction, if performed by inexperienced staff. Postoperative care must be undertaken in a paediatric intensive care unit, with associated issues of bed availability, cost and potential for complications to occur as a result of the required prolonged intubation.

Principles of Cartilage Augmentation and Stenting

Animal studies have confirmed the presence of viable cartilage following laryngotracheal reconstruction, with repiratory epithelium found lining the graft in the majority of cases[1,23]. Histological examination after LTR with anterior cartilage grafting in rabbits, also showed rapid epithelialization of the graft. Although the original graft underwent progressive necrosis and resorption, neochondrification occurred rapidly providing the graft with excellent structural support[13]. The cartilage used in LTR is successfully incorporated into the laryngeal framework and grows with the child. There have been no adverse effects reported on laryngeal growth. [6]

LTR is traditionally performed as a multi-stage procedure with a tracheostomy insitu. This surgery aims to safely create a stable airway, which is age-appropriate in size and preserves or restores normal laryngeal function. Cartilage grafts are used to augment the airway during LTR and thus address the underlying stenosis, whilst stents stabilise and support the reconstruction whilst healing occurs. The single-stage procedure was developed as an extension of the experience gained from anterior cricoid split surgery, where it became apparent an endotracheal tube may be used as a short-term airway stent [8,15,21]. Numerous techniques may be employed to expand the airway in LTR including anterior and/or posterior cricoid splits, each of which may be grafted. Grafts are most commonly fashioned using autogenous costal cartilage, which has been found to be a robust and reliable grafting material. Auricular and thyroid alar cartilage may also be used.

Assessment and Decision Making Factors

Each paediatric patient must undergo a comprehensive assessment to formulate an individualised treatment plan. Specific details of the history and examination are not covered in this review but assessment ought to document the effect of the airway stenosis on the child. This includes the degree of respiratory compromise, general condition of the child and any coexisting diagnoses which may affect the airway at other levels, for example Pierre Robin sequence, craniofacial anomalies, choanal atresia or chronic lung disease.

A rigid microlaryngoscopy and bronchoscopy during spontaneous ventilation remains the gold standard for endoscopic evaluation. This allows for close, systematic inspection of the airway at all levels using an appropriately sized laryngoscope with a telescope or operating microscope, under laryngeal suspension. The cricoarytenoid joints are palpated to test passive mobility and examine for interarytenoid scarring. The Hopkins rod telescope allows for accurate assessment of the subglottis and tracheobronchial airway in addition to the remainder of the larynx. Photographic documentation and/or video recording are useful (FIGURE 2). Features of the stenosis including the anatomical level, length and consistency (soft/firm), site (anterior +/or posterior or circumferential), maturity (active inflammation/oedema) and presence of granulations, fibrosis or scarring, must be carefully established. The suprastomal area should also be examined in cases with a pre-existing tracheostomy. The airway is formally sized and the stenosis is graded using the Myer-Cotton Grading system[17], to assist in treatment planning (FIGURE 3). A dynamic assessment of the airway ensures any co-existing tracheomalacia, vocal cord immobility or laryngomalacia is detected.

The single stage procedure has the potential to provide a decannulated stable airway, sooner than the traditional multistage procedure and is thus an appealing option. Treatment options for each child must be considered on a case-by-case basis. Single stage procedures are ideally

considered when an anterior graft is performed with or without a posterior cricoid split, although posterior grafts have been performed in single stage procedures. LTRs with a combination of anterior and posterior grafts have been reported to have a higher reintubation rate although their subsequent outcomes are comparable with the multistage procedure.[12] Single stage procedures have been shown to have poorer outcomes in patients with tracheal obstruction or tracheomalacia particularly in those aged less than 4 years.[12] The procedure is also contraindicated in patients whose airway anatomy makes reintubation technically difficult particularly in the emergency situation (e.g. craniofacial or vertebral anomalies) and in children with ongoing neurological deficits or chronic lung disease which preclude decannulation.

Babies weighing more than 4kg or with gestational ages of greater than 30 weeks have been found to have a greater chance of successful extubation and eventual airway patency[16]. Very small babies have more complications at extubation which may occur as a result of the co-morbidities frequently present in these infants. In addition the physical dimensions of their airway leave little margin for any degree of airway compromise. These patients may be better managed by an initially conservative course whilst they grow or via a multistage reconstruction, ensuring a stable airway throughout the lengthy healing process.

Multistage LTR is more suitable for the severe grades of stenoses. A trend toward higher reintubation rates and tracheostomy insertions after single stage reconstructions, has been reported for severe Grade 3 or 4 stenoses.[12] It is also preferable for patients with poor respiratory reserve, multilevel stenoses or those with underlying factors which may compromise normal healing, for example ongoing reflux or systemic problems, and who would thus require long-term stenting.

Classical Laryngotracheal Reconstruction using Costal Cartilage Graft with a Tracheostomy

The patient is positioned supine with the neck extended. The tracheostomy tube is replaced with an appropriately sized armoured cuffed endotracheal tube. The skin is prepared and the patient is draped, keeping the donor site separate from the neck. It is the preference of the senior author to first harvest the costal cartilage graft, to minimise contamination of the clean donor site. By convention the graft is taken from the right submammary region adjacent to the bony-cartilaginous junction. A transverse incision is made and the rib margins identified. The bony-cartilaginous junction is visualised and the adjacent cartilaginous portion of the rib is carefully transected. A 3-4cm segment of cartilage is elevated in a subperichondrial plane, taking care to avoid injury to the underlying pleura. Haemostasis is achieved and a leak test performed. Although uncommon, any pleural defect can thus be identified and repaired immediately. The wound is closed in layers with a subcuticular skin suture.

A transverse skin incision is made superior to the pre-existing stoma site and subplatysmal flaps are elevated from the superior margin of the thyroid cartilage to the tracheostomy site. A midline laryngofissure is performed (FIGURE 4A). The extent of this incision is dependent on the site and extent of the airway pathology. For isolated subglottic stenosis, the airway is opened from just below the vocal cords (level of the cords correlates with the midpoint between thyoid notch and cricoid in paediatric patients), through the anterior cricoid and upper tracheal rings to release the stenotic segment. At this stage a posterior cricoid split may be performed, if indicated (FIGURE 4B). Posterior glottic stenosis is addressed by carefully dividing the glottic scar via the posterior laryngofissure. The incision is extended superiorly into the interarytenoid region through the fibrosed interarytenoid musculature and inferiorly for ~1cm into the tracheoesophageal septum. The split should be splayed open with scissors to prove that the division is complete.

Once the stenosis is adequately released, the residual anterior and/or posterior defects may be assessed and measured to shape the cartilaginous augmentation graft. The graft is fashioned into a boat-shape with dimensions matching the defect (FIGURE 4C). The senior author uses

flanged grafts which will “lock” into position and thus aid in securing the graft and minimise the likelihood of graft displacement into the airway (FIGURE 5). Care is taken to bevel the superior luminal lip of the graft, to avoid ~~any airway prolapse~~excess cartilage obstructiong the lumen. A stent is used if a posterior graft is needed. The stent can be preformed or fashioned from a Portex tube with the upper ends welded together, to lie between the vocal cords. The inferior end sits just above the tracheostomy (FIGURE 4D). The stent is anchored with a robust, nonabsorbable transfixion stitch, to prevent any displacement. Grafts may be secured via interrupted vicryl sutures to provide additional stability in the immediate postoperative period. An airtight seal ensures that tracheal secretions do not bathe the graft which helps minimise graft infection. A layered closure is performed and a Penrose drain is inserted.

Postoperative care is undertaken in a ward environment, with staff who are experienced in the care of airway cases. A chest radiograph is taken to exclude any pneumothorax or pneumomediastinum. Prophylactic antibiotics and antireflux medications are used in the postoperative period. Stents are usually removed 4-6 weeks after reconstruction, followed by a check microlaryngoscopy 7-10 days later. Decannulation is then arranged and occurs in a monitored ward environment, provided the airway is assessed as adequate.

Stents

Stents are utilised in airway expansion surgery to provide a framework for mucosal healing. They give stability, minimise the possibility graft displacement and help prevent scar contracture. It is preferable to avoid stenting if possible and they can typically be avoided if there is only an anterior graft. Stenting is important if bulky submucous scarring is removed or thick glottic webbing is divided. They do have inherent risks, for example a Montgomery T tube has the dual purpose of stenting the reconstruction and providing a patent airway. Blockage of this form of stent has potentially disasterous consequences and parents must be trained in emergency procedures for removal and replacement with a tracheostomy.[22] Stents may also lead to an increased risk of aspiration in children with marginal feeding skills.

Despite these concerns however, they are well tolerated by patients and very useful in complicated cases.

Single Stage Laryngo-tracheal Reconstruction

The operative technique is very similar to the classical laryngotracheal reconstruction except the reconstructed airway is stented by an endotracheal tube in the postoperative period. If a posterior graft is needed and there is no tracheostomy, an extended laryngofissure is formed to allow a small tube to be placed into the distal airway, below the posterior graft. The tracheostomy site may be incorporated into the reconstruction. Postoperative care is provided in the Paediatric Intensive Care Unit, with an endonasally placed endotracheal tube remaining in position for around 7 days or until a low pressure leak is observed.[20] At Great Ormond Street Hospital, the common practice is to wean sedation as tolerated with the ideal situation being that of a spontaneously breathing, awake child, provided they are calm and tolerate the tube well.[19] Children undergo microlaryngoscopy and downsizing of the endotracheal tube under steroid cover after 7 days (FIGURE 6), with a trial of extubation the following day provided the reconstructed airway appears favourable. A further progress microlaryngoscopy is performed after 7-10 days to inspect the airway and remove any granulomas. Postoperative care differs in surgical centres and may include varying levels of sedation either alone or in combination with neuromuscular paralysis. Meticulous postoperative care is essential, particularly to avoid unplanned extubation and the associated morbidity of reintubation.

Outcomes

The overall success of LTR has traditionally been measured by decannulation rates. Excellent outcomes have been reported for single stage reconstruction with extubation/decannulation rates of 84-96%[7,9,15,16,21] being comparable to the classical staged procedure which has reported decannulation rates of 92%[7]. A group of 190 children undergoing single stage LTR were examined by the Cincinnati group. 29% required reintubation, 15% of the overall group required tracheostomies however only 4% of the

overall group remained tracheostomy dependent. 5% required a further operation before successful decannulation[12]. Outcomes also vary according to the grade of stenosis present. LTR enables approximately 90% to be decannulated for Grade I and II, 80% for Grade III and 50% or poorer for Grade IV stenoses.[4,7,18] Voice outcomes are also important but difficult to study as the younger patients may not have any voice to assess preoperatively due in part to their developmental stage and also having a tracheostomy in a narrowed airway. Overall, a normal or near-normal voice is achieved in about 50% of children after LTR[3]. Voice can be compromised by anterior commissure blunting, vocal cord immobility and supraglottic phonation, however the preoperative pathology also plays a significant role in whether a normal voice is achievable in the postoperative period.

TABLE 1: Aetiology of Acquired Laryngotracheal Stenosis

INTERNAL TRAUMA
- Intubation injury

EXTERNAL TRAUMA
- Blunt
- Penetrating

OTHER
- Trauma
  - Post laryngeal surgery - High tracheostomy, glottic web, supraglottic collapse
  - Burns
- Chronic infection e.g. Tuberculosis, Syphilis
- Chronic Inflammation
- Systemic - Sarcoidosis, SLE, Pemphigus, Wegeners
  - Reflux
- Laryngeal Neoplasm
  - Primary lesion - Chondroma, fibroma, carcinoma
  - Secondary involvement - Tumour infiltration, radionecrosis, postoperative scarring

TABLE 2: Selection of Treatment Modality

TREATMENT MODALITY	CRITERIA/PATIENT FACTORS
Laryngeal Rest / elective period of intubation	• Early stenosis - Oedema only/No fibrosis • "Small" babies – age and weight • Laryngeal inflammation– address any reversible component e.g.reflux, infection.
Endoscopic Treatment of granulations and early stenosis: Cold steel rather than CO2 laser, Balloon dilatation, Mitomycin C, endoscopic cricoid split	• Early/"soft" stenosis/granulation tissue/primarily oedema without fibrosis • Limited/isolated pathology - Grade I Stenosis • "Seal flipper" granulations • In patients not appropriate for open surgical intervention due to oxygen requirements, neurological co-morbidities, craniofacial anomalies
Anterior Cricoid Split	• Repeated failure of extubation in neonate with isolated subglottic stenosis • >1500g • no active respiratory/cardiac co-morbidities • <30% oxygen requirement
Laryngotracheal Reconstruction – Cartilage Augmentation	• Symptomatic laryngotracheal stenosis • Most appropriate for Grade II and III • >2000g
Cricotracheal Resection	• Severe Grade III or Grade IV Stenosis • At least 2mm from cords • Long segment stenosis extending into trachea • Salvage after failed LTR
Tracheostomy	• Medically unsuitable for above techniques • Failure of above techniques

FIGURES LEGEND

Figure 1 - Loss of subglottic mucosa and exposed subglottic cartilage following intubation of neonatal airway.

Figure 2 – Acquired subglottic stenosis following neonatal intubation.

Figure 3 – Myer-Cotton Grading system of Laryngeal Stenosis.

Figure 4 – Operative procedure for Laryngotracheal reconstruction using autologous costal cartilage graft

a. Midline laryngofissure is performed.

b. Posterior cricoid split may be required.

c. Boat shaped anterior cartilaginous graft is fashioned and sutured in place.

d. Stent is positioned appropriately.

Figure 5 – Flanged graft used for anterior cartilaginous augmentation.

Figure 6 – Appearance of subglottic airway at 7 days post-single stage LTR with anterior graft and posterior split.

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