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Congenital anterior glottic webs with subglottic stenosis: Surgery using perichondrial keels

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ABSTRACT

Four children with severe congenital anterior glottic webs required surgical reconstruction of their laryngeal airway to either avoid a tracheotomy or allow tracheotomy decannulation. The technique of re-establishing a glottic inlet allows both normal respiration, good cough and a satisfactory voice outcome. It utilizes an autogenous graft with perichondrium acting as an overlay keel to minimize glottic web reformation. All children were found to have Shprintzen syndrome.

Objective: The objective of this case series was to document the method of surgically using this perichondrial keel for airway reconstruction. It also examined the subsequent outcome and associated complications that were encountered.

Methods: A prospective analysis of four cases from 2001 to 2008 created a database of information. All cases were classified using the Cohen staging system. They were treated with the same surgical technique using auricular or costal cartilage graft with attached perichondrium, but the postoperative course was tailored to each individual case.

Results: All four children were successfully treated with removal or avoidance of a tracheostomy. All had an associated subglottic stenosis treated, and had no major complication requiring revision tracheotomy. They did not have any respiratory complications, and they produce a satisfactory voice albeit still slightly husky.

Conclusion: All four cases had Shprintzen syndrome, and confirms the need to screen for VCFS in children with an anterior glottic web. Tracheotomy is still the gold standard of treatment in severe congenital anterior glottic webs. The described technique offers another good option to the paediatric airway surgeon in managing this condition.

1. Introduction

Congenital anterior glottic webs are an uncommon finding in patients with respiratory distress and stridor. Fleischmann [1] first observed in 1882 this condition during an autopsy of an infant. Since then, the condition has been reported by Holinger with 16 patients [2], Benjamin who presented 19 anterior glottic web cases over a 12 year period [3], Cohen with 51 children over a 32 year period [4] and Wyatt et al. with 15 cases [5]. The presenting complaints include lack of voice, biphasic stridor, recurrent croup or pneumonia.

During the 6th week of embryological life, the developing laryngeal opening is obliterated by actively proliferating epithelial tissue, arising from the two arytenoid swellings and anteriorly by the epiglottic eminence. The definitive opening of the glottis begins with the vacuolization of the epithelial lamina and subsequent autolytic dissolution of the epithelium. It is theorized that glottic webs arise as a result of failure of one of these mechanisms, producing a glottic web that is anterior in position and has an associated subglottic stenosis (Fig. 1). Dr Seymour Cohen proposed a classification system of glottic webs as tabled (Table 1).

Surgical methods to correct glottic webs have been well described, with either endoscopic or external techniques [1–5]. In extensive glottic webs, it is universally agreed that some form of keel at the anterior commissure is necessary for best results. Keels described have been made from Teflon, plastic, silicone rubber and silastic. This case series describes our recent experience using autogenous perichondrial tissue as a form of keel to minimize web recurrence.

2. Methods

At the Children's Hospital at Westmead, a database of children with laryngeal webs have been previously created. A database of

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Table 1
Glottic web classification as described by Cohen [4].

Severity	Extent of glottis	Subglottic involvement	Visible cords	Symptoms
Type 1	<35%	Little or none	Present	Clear airway, mild voice hoarseness
Type 2	35–50%	Thin anterior web with minimal subglottic involvement	Present	Hoarse or weak cry, airway compromise only with exertion
Type 3	50–75%	Thick to thin web, extends to lower border of cricoid	Cords appear fused anteriorly and thins out posteriorly	Marked vocal dysfunction, weak and whispery voice, moderate airway obstruction
Type 4	75–90%	Uniformly thick web, extends to lower cricoid	Complete fusion, no visible cords	Aphonic, severe airway obstruction, requires tracheotomy

information was created to document patient characteristics, associated conditions, genetic analysis, laryngoscopy findings, surgical technique utilized, outcome scores looking specifically at airway stridor, cough and airway protection. The information was obtained with the approval of the Ethics Committee of this hospital.

3. Results

Since 1984, 109 patients have been diagnosed with a glottic web. 35 (32.1%) had a congenital anterior glottic web, 37 (33.95%) had congenital posterior web and 37 (33.95%) had acquired webs. From 2001 to 2008, four consecutive paediatric cases (ages 0–16) of congenital anterior glottic webs with subglottic stenosis (Cohen Type 3 and 4 (Table 1)) requiring an external approach laryngofissure were documented. The indication for surgery was to either decannulate from a previous tracheostomy or to avoid an imminent tracheotomy. The same surgeon was involved in all four cases.

3.1. Case one

A 3 month old child was presented for assessment of her airway after being noted to have a hoarse cry and biphasic stridor since birth. She had her patent ductus arteriosus (PDA) corrected but the stridor did not improve with anti-reflux measures. More recently, she had been experiencing choking cyanotic episodes numbering three times per week. Laryngoscopy demonstrated a thick anterior glottic web occupying at least the anterior two thirds of the membranous portion of the vocal cords (Cohen type 3), and associated subglottic stenosis. A screening fluorescence in-situ hybridization (FISH) test for a deletion at chromosome 22q11.2 locus was positive for velocardiofacial syndrome(VCFS) or Shprintzen syndrome.

After division of the cricoid cartilage and lower half of the thyroid cartilage, the web was divided in the mid-line under telescopic guidance by a second operator. The posterior edge of the cut web was gently sutured forward at the level of the glottis with 6/0 Maxon to the anterior cut edge of the thyroid cartilage incision on both sides. This relies on the mobility of the remaining mucosal tissue over the vocal ligament.

The remnant glottic tissue anterior to this was reflected inferiorly and also sutured to the thyroid ala laterally. This was performed gently so as to maintain a vocal fold at the level of the anterior commissure. Matching the sutures on each side of the reconstructed glottis allow precise re-approximation of the glottis.

An appropriate sized nasotracheal tube was inserted as a stent. A composite graft of cartilage with perichondrium taken from the cymba conchae was used to stent the subglottic stenosis, with a tongue of perichondrium left between the new glottic folds (Fig. 2). She was intubated for 7 days and extubated successfully. However her airway continues to be good 7 years later, and though her voice has good strength, she continues to have an articulation disorder and undertakes speech therapy regularly.

3.2. Case two

A 4 year old girl had been followed by our department since the age of 2 months. She presented initially with airway obstruction, respiratory distress, a diagnosis of atrial septal defect (ASD) and ventricular septal defect (VSD). She had a laryngoscopy at age 27 days, confirming a thick anterior glottic web with a small chink in the posterior commissure. The stenosis extended 15 mm into the subglottis. She received a tracheostomy which she retained until the reconstruction. She had no recognizable voice. Genetic studies (FISH) confirmed a deletion of chromosome 22q11.2. The laryngoscopy confirmed a Cohen Type 4 web, with a 75% subglottic stenosis.

The new vocal folds were created as described in the first case. A costal cartilage graft was harvested and shaped to fit the subglottic defect. However, a significant segment of perichondrium on the undersurface of the graft was made to extend above the glottic folds once the graft was inserted into the subglottis. This acted as

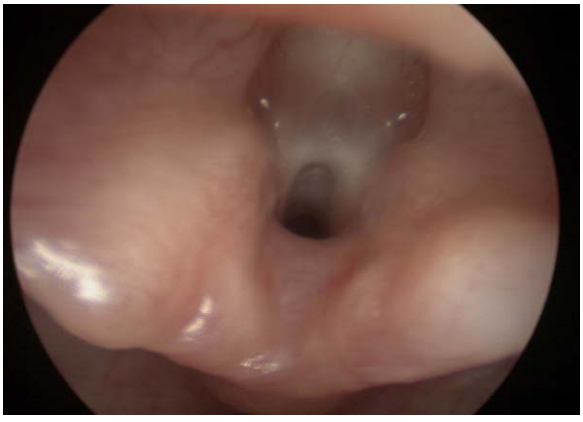

Fig. 1. Type 4 glottic web in neonate.

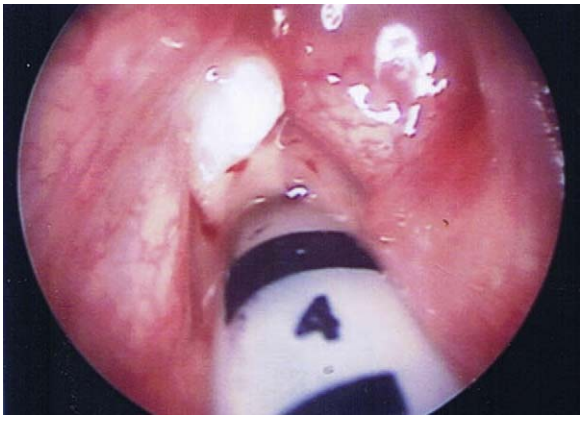

Fig. 2. Divided glottic web with perichondrium lining one cord.