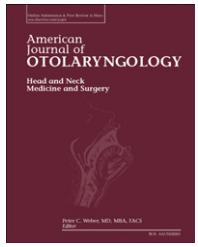
Available online at www.sciencedirect.com

ScienceDirect

www.elsevier.com/locate/amjoto



Type IV congenital laryngeal web: Case report and 15 year follow up

*Brendan Sorichetti, BSc, John P. Moxham, MD, FRCSC, Frederick K. Kozak, MD, FRCSC**

Division of Pediatric Otolaryngology, Head & Neck Surgery, BC Children's Hospital, University of British Columbia, Vancouver, BC, Canada

ARTICLE INFO

Article history:
Received 21 October 2015

ABSTRACT

A five day old patient with mild VACTERL syndrome had repair of a type IV congenital laryngeal web with successful decannulation 76 days later. Voice and respiratory outcome is good with follow up 15 years later. This case presents a rare clinical finding of a type IV laryngeal web successfully repaired with a keel and subsequent long term follow up during an era when it was suggested that repair be delayed until 18 months of age at the earliest.

1. Introduction

Laryngeal webs are rare and either present congenitally or are acquired secondary to trauma. Congenital webs comprise less than 5% of all congenital anomalies of the larynx [1]. A congenital web is a malformation in which abnormal tissue fails to be resorbed between the laryngotracheal sulcus and the primitive pulmonary sac which makeup the respiratory tap. Webs vary in appearance due to their size and thickness and the resultant degree of airway occlusion [2].

Clinical symptoms attributed to laryngeal webs range from breathing difficulties such as stridor and airway obstruction to dysphonia or an inaudible voice as a result of vocal obstruction [3,4]. Diagnosis involves fiberoptic laryngoscopy or direct micro laryngoscopy to fully evaluate the extent of the web. The location of the web commonly occurs at the anterior glottis and may extend posteriorly with some extension to the subglottis [5]. Cohen separated laryngeal webs into four categories depending on the amount of occlusion and severity of symptoms [6] (Table 1).

The primary goal of surgical therapy for a web relates to airway management followed secondarily by voice rehabilitation. After assessing the severity of the web a laryngofissure, resection of the web and placement of a keel has been the accepted form of treatment first appearing in 1950 and championed by Montgomery in 1970 in those cases requiring intervention [7,8]. Since that time, excision of the laryngeal web by cold steel or CO_2 laser, followed by keel insertion has been the main form of repair [9–13]. Although there are a variety of materials used for keels, the primary goal of placing this foreign body between the freshly divided web edges is to prevent reformation of scar tissue. Alternative surgical treatment for relatively thin laryngeal webs involves excision and separation of the web with the lower mucosal flap being folded up and the upper flap being folded down to cover the wound on the inferior and superior surfaces of the vocal cord, thereby negating the use of a keel [14–16]. This technique is technically demanding and not suitable for thicker laryngeal webs as the greater amount of scar tissue can easily lead to re-adhesion. The treatment and diagnosis of anterior glottic webs remain a therapeutic challenge and no single approach has been universally accepted.

1.1. VACTERL syndrome

VACTERL syndrome is a pattern of non-random congenital malformations which includes vertebral defects, anal atresia, tracheo-esophageal fistula with or without esophageal atresia,

* *Corresponding author at:* Division of Otolaryngology, University of British Columbia, Division of Pediatric Otolaryngology, BC Children's Hospital, K2-184, ACB, Department of Pediatric Surgery, 4480 Oak Street, Vancouver, BC, Canada V6H 3 V4.
E-mail address: fkozak@cw.bc.ca (F.K. Kozak).

http://dx.doi.org/10.1016/j.amjoto.2015.11.001
0196-0709/

Table 1 – Classification of laryngeal webs (types 1–4) [6].

Category	% Occlusion of the Glottis	Complications	Appearance
Type I	≤35%	- Mildly abnormal voice with some hoarseness	- Vocal cords remain visible
Type II	35%–50%	- Airway symptoms are uncommon - The voice is husky and weak - Typically associated with cricoid abnormalities	- Thick anterior webbing
Type III	50%–75%	- Cricoid abnormalities are expected. - Airway symptoms are severe and often require an artificial airway. - There are significant voice abnormalities	- Thick anteriorly - Difficulty locating the vocal chords
Type IV	75%–90%	- Severity of this condition requires immediate airway management. - Patient is unable to produce an intelligible voice and can be considered aphonic	- Vocal chords are now truly unidentifiable

radial and renal dysplasia, cardiac malformations and limb anomalies [17,18]. The presence of VACTERL association occurs in approximately 1/10,000–1/40,000 live births, and requires at least 3 component features and the absence of evidence of an overlapping condition [19,20]. In VACTERL syndrome, 50–80% of malformations involve the esophagus and the trachea and laryngeal webs are rarely associated with this syndrome [21–23].

1.2. *Embryological development of laryngeal webs*

Congenital laryngeal webs are believed to be caused by a malfunction during embryological development [24–26]. The failure of the epithelial lamina to recanalize has been thought to give rise to the range of disorders that include laryngeal webs and stenosis.

Other theories have proposed that abnormalities of subglottic extension can be attributed to the epithelial lamina having an abnormal inferior extension into the subglottis and remaining after embryogenesis [4]. Another potential scenario involves a ventral extension of the infraglottis called the vestibulotracheal duct. The failure of this duct to develop can cause a migration of epithelial lamina precartilaginous cells into the area which would form the anterior subglottis. These migratory cells result in the formation of a laryngeal web and associated subglottic stenosis [27].

2. Case report

A 2 day old female with VACTERL syndrome (Table 2) had repair of an imperforate anus with the anesthesiologist reporting difficulty intubating the patient and subsequently placed the endotracheal tube (ETT) at the laryngeal inlet. On day 5 the otolaryngology service was consulted as the patient had an abnormal cry and intermittent biphasic stridor. Direct laryngoscopy in the operating room noted a thick type IV laryngeal web covering 95% of the larynx (Fig. 1). To underscore the significant severity of this web, a representation of the web from a superior view of the vocal cords can be seen in Fig. 2. A grade-2 subglottic stenosis was also present and the trachea was normal.

Intubation with a 2.5 ETT was unsuccessful so a CO_2 laser was used to divide the midline of the posterior aspect of the cords until a 3.0 ETT could be inserted. Web thickness was approximately 4.5 mm. With the airway secured, the patient underwent a tracheostomy. The decision was made to ameliorate the web immediately and it was carefully lysed along the midline using a CO_2 laser. A cricoid split and laryngofissure with a keel insertion was undertaken.

The keel was removed two months later and the vocal cords had healed nicely with a small amount of granulation tissue along their surface. The patient still had a degree of mild subglottic stenosis. From a laryngeal standpoint the patient has done very well. The patient presents with a good voice, mildly hoarse and no further intervention has been required over the past fifteen years. A thin anterior web still exists and consideration to lyse it is under discussion. The larynx is seen in Fig. 3, 14 years post repair.

3. Discussion

Beginning in the 1950s, treatment of a laryngeal web involved surgical excision and placement of a keel [8]. Since that time there have been advancements in the surgical approach to treating this entity depending on the type and thickness of the web. With the introduction of the CO_2 laser in the 1970s, the use of the laser has also been employed. Whether the web is acquired or congenital, the purpose of introducing a keel is to create a partition between

Table 2 – Symptoms present in the newborn period and sequelae in the patient born with VACTERALs.

	Vertebral Anomalies	ARMs (Anorectal Malformations)	Cardiac Anomalies	Tracheo-Esophageal Anomalies	Renal Anomalies	Limb Anomalies	Other Findings
Description	Congenital scoliosis and mild torticollis	Imperforate Anus Operated on 2nd day of life	Mitral VSD Multiple ventricular septal defect and persistent ductus arteriosus	Type IV glottic web and laryngeal pouch	None	None	Signs of hearing impairment *Apparent later in life