

Congenital laryngeal webs: From evaluation to surgical management

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Abstract

Objective. To discuss the presentation, evaluation, and management of congenital laryngeal webs. **Methods:** The Institutional approved this retrospective review of all patients who were diagnosed with laryngeal webs at our institution within the past 10 years. **Results:** There were 3 type I and II webs and 7 type III and IV webs; All 3 children with type I and II laryngeal webs recovered after a single endoscopic procedure, while children with type III and IV laryngeal webs usually had a record of multiple visits to the hospital with multiple treatments. In all 7 children with type III and IV webs, a tracheotomy was performed. The average age of the patients who underwent tracheotomy was 4.86 months(range,1-11mo). Six of the seven children (85.7%) with type III and IV webs displayed subglottic stenosis. The ultimate treatment for such patients was open laryngoplasty. **Conclusions:** children with type III and IV laryngeal webs were more likely to have subglottic stenosis and needed to undergo tracheotomy earlier. They usually had a record of multiple visits to the hospital with multiple treatments. Open laryngoplasty combined with the implantation of a T-tube and reconstruction of the cricoid cartilage using hyoid bone may play a crucial role in the treatment of congenital laryngeal webs with subglottic stenosis.

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Conclusions: children with type III and IV laryngeal webs were more likely to have subglottic stenosis and needed to undergo tracheotomy earlier. They usually had a record of multiple visits to the hospital with multiple treatments. Open laryngoplasty combined with the implantation of a T-tube and reconstruction of the cricoid cartilage using hyoid bone may play a crucial role in the treatment of congenital laryngeal webs with subglottic stenosis.

Keywords: congenital laryngeal webs, T-tube, open laryngoplasty, hyoid bone reconstruction of the cricoid cartilage, subglottic stenosis

Succinct key points:

Children with type I and II laryngeal webs were usually older when they presented to the hospital.

Children with type III and IV laryngeal webs were more likely to have subglottic stenosis and needed to undergo tracheotomy earlier.

Children with type III and IV laryngeal webs usually had a record of multiple visits to the hospital with multiple treatments.

The implantation of a T-tube and reconstruction of the cricoid cartilage play a crucial role in the treatment of congenital laryngeal webs with subglottic stenosis

Children with type III and IV webs may also present with other problems of the throat or the whole body, including subglottic stenosis, vocal cord paralysis.

Background

The congenital laryngeal web is a rare form of clinical laryngeal malformation, which was first proposed by Fleischmann in 1882. This condition is formed by the growth of abnormal tissues in the glottic portion of the larynx and accounts for approximately 5%[1] of congenital structural malformations of the larynx. At 8-10 weeks of gestation, the congenital laryngeal web is generally associated with embryonic developmental disorders that may occur alone or with other clinical syndromes, such as 22q11.2 deletion syndrome. The clinical manifestations of this condition may differ, depending on the degree of involvement of the laryngeal tissues and the degree of obstruction in the trachea.

Patients with congenital laryngeal webs clinically present with vocal disturbances, hoarseness, wheezing, or breathlessness. This condition is most commonly found in the anterior coalition of the vocal folds and often combined with stenosis below the glottis. In 1985, Cohen[2] proposed a typology for the congenital laryngeal web, which included the following types: i) type I, a thin, membranous laryngeal web involving <35% of the vocal folds; ii) type II, laryngeal web tissues involving 35%-50% of the glottic portion; iii) type III, laryngeal web tissues comprising 50%-75% of the glottic portion and involving the lower glottic portion; and iv) type IV, laryngeal web tissues involving more than 75% of the glottic portion and completely blocking the glottis.

Materials and methods

The retrospective analysis was conducted on a total of 10 children diagnosed with congenital laryngeal webs. These children were admitted to our department between 2010 and 2020. Of the 10 children, 3 had type I and II laryngeal webs (Figure 1-D), while the other 7 children had type III and VI congenital laryngeal webs (Figure 1-A, B, C). Of the 3 children with type I and II laryngeal webs, all were females, whereas in seven children with type III and IV laryngeal webs (Table 1), six were males, and one was female. The oldest child with a type I and II laryngeal web was five years old at the time of presentation, while the youngest child was only 1 year and 9 months old. These children clinically presented with persistent hoarseness and no obvious respiratory distress. All three children did not undergo a tracheotomy, and all of them recovered after a single endoscopic procedure. The oldest children with type III and IV laryngeal webs in our study were aged 2 years and 2 months old at first presentation to our hospital, while the youngest child was 1 month old. The average age of children with type I and II laryngeal webs when they came to our hospital was 43 months, while that of children with type III and IV webs was 13 months. For all seven children with type III and IV congenital laryngeal webs, tracheotomy was performed early, at an age ranging between 1 and 11 months. The average age of these patients when they underwent tracheotomy was 4.86 months. In one of the seven children, we observed a combined atrial septal defect, while in another case, both atrial septal defects and laryngomalacia were observed. One child had vocal cord paralysis, and six children of seven (85.7%) displayed subglottic stenosis. Among these six children, four had an area of subglottic stenosis greater than 90% (Cotton Grade III), while the other two had 40% (Cotton Grade I) subglottic stenosis.

All 3 children with type I and II laryngeal webs recovered after a single endoscopic procedure. The other seven children with type III and IV laryngeal webs eventually recovered in our hospital through open laryngoplasty and were successfully extubated. Before surgery at our hospital, these children had undergone multiple

endoscopic or open surgical procedures, which included a maximum of five and a minimum of two endoscopic procedures. However, all the previous procedures were unsuccessful in removing the tracheal tube. The average number of operations performed on these patients in other hospitals before they came to our hospital was 2.85. For the one child without subglottic stenosis, the surgical method was T-tube implantation. For the other six children with subglottic stenosis, the surgical method was T-tube implantation combined with reconstruction of the cricoid cartilage. In five of the patients, cricoid cartilage was reconstructed with free hyoid bone, while in one patient, cricoid cartilage was reconstructed with costal cartilage. Preoperative enhanced CT of the neck was performed in all cases (Figure 2), which suggested varying degrees of stenosis in the glottic portion and below the glottis. The youngest of the seven children with type III and IV laryngeal webs when they underwent open laryngoplasty was 1 year and 1 month old; in contrast, the oldest was 2 years and 11 months old. The average age at surgery of the seven children was 24 months. In all seven cases, a T-tube was implanted intraoperatively, and in the six children with subglottic stenosis, the cricoid cartilage was incised to remove the scar tissue under the glottis. The cricoid cartilage was reconstructed using autologous rib cartilage or hyoid bone (one rib cartilage and five hyoid bone). In one of the cases who had vocal fold paralysis, the right vocal fold was removed, and a T-tube was placed through the original tracheal incision. The T-tube was placed slightly above the glottis and acted as a support to prevent adhesions. Children who present with choking while consuming fluids after surgery should be fed a soft or semiliquid diet. Additionally, this problem could be solved by dietary exercises. The tube was placed for 3 months in one case, 8 months in another case, and 6 months in five cases. The average time was 5.86 months. After placing the T-tube for a certain period, the children returned to the hospital for a follow-up visit and for a change in the regular tracheal tube. After one month of blockage, all seven children were successfully extubated.

Open laryngoplasty (T-tube implantation + hyoid bone reconstruction of the cricoid cartilage)

The surgery was performed under general anesthesia with an anesthetic cannula inserted in the place of the tracheal tube in the neck of the child (Fig. 3-A). The cricoid cartilage, thyroid cartilage, and hyoid bone were exposed. A portion of the hyoid bone was obtained and trimmed into a pike shape (Fig. 3-B, C). The affected cricoid cartilage was incised to remove the subglottic stenotic lesion tissues, the anesthetic cannula was removed, and a T-tube was inserted (Fig. 3-D). The T-tube was positioned slightly above the glottis using intraoperative laryngoscopy (Fig. 3-F). Afterward, the cricoid cartilage was repaired and reconstructed with the hyoid bone (Fig. 3-E), and the incision was sutured. A schematic diagram of the operation process is shown in Figure 4.

Results

All cases were followed up for over 2 years. We found that three children with type I and II laryngeal webs had recovered through a single endoscopic procedure, and their hoarseness was relieved after the surgery. Seven children with type III and IV laryngeal webs had their tracheal tubes successfully removed. These children did not resume dyspnea during the follow-up period, but some children still presented with hoarseness.

Ethical considerations

This study was approved by the Institutional Ethics and Research Committee of Our Hospital.

Discussion

Infants with dysphonia and weak crying after birth should be highly suspected of having congenital laryngeal webs. Additionally, infants who present with persistent hoarseness, recurrent laryngeal wheezing, or acute laryngitis after birth should be admitted to the Otorhinolaryngologic Department. In cases where the laryngeal web is excluded, laryngoscopy may be performed to confirm the diagnosis. The development of fiberoptic laryngoscopy and bronchoscopy has provided great improvement in the diagnosis of congenital laryngeal webs and other laryngeal malformations. These techniques may help to clarify the presence and extent of the laryngeal web obstructing the glottis. Thus, the local scarring and adhesion caused by blind operation under an ambiguous diagnosis can be avoided.

The laryngeal web was first proposed by Cohen, whose clinical typology offered some guidance for its clinical treatment. Cohen claimed that type I and II laryngeal webs were often found without any comorbidities, while type III and IV laryngeal webs were frequently combined with other structural malformations of the larynx or presented as a manifestation of the clinical syndrome. In our clinical practice, the most common throat complication was subglottic stenosis. Other complications included vocal cord paralysis, laryngomalacia and atrial septal defects. The most common syndrome associated with laryngeal webs was 22q11.2 deletion syndrome [3]. The type I and II laryngeal webs often involved relatively thin tissues, where the performance of microscopic endoscopic surgery produced an excellent outcome. This treatment improved the hoarseness and allowed the patient to speak in an almost normal voice. However, type III and IV laryngeal webs were generally combined with subglottic stenosis, requiring open surgery to achieve a better outcome. This resolved the child's breathing difficulties, and it also allowed them to speak in a normal voice. Open surgery can be performed in a variety of ways. In our hospital, open laryngoplasty combined with a T-tube and reconstruction of the cricoid cartilage was performed to treat type III and IV laryngeal webs. This procedure showed excellent results. The T-tube was placed for 6 months, slightly above the glottis, to prevent adhesions of the vocal cords and to support and enlarge the tracheal lumen and better address subglottic stenosis. However, the T-tube may cause discomfort, such as choking and coughing early after surgery. Thus, such patients need to be given a soft or semiliquid diet. In children with subglottic stenosis, the lingual bone or autologous rib cartilage can be used to reconstruct the cricoid cartilage. This surgery method can resolve the child's breathing difficulties and help in the successful removal of the tracheal tube. All children were successfully extubated. We recommend using the hyoid bone to reconstruct the cricoid cartilage, as it can be obtained in the same surgical area, reducing unnecessary trauma. We have advanced the age of surgery to approximately 2 years old, shortening the time for tracheostomy to open the tube, greatly improving the quality of life of the children, and reducing the nursing burden and living burden of the children's families. However, the child may still have postoperative hoarseness, which may require further rehabilitation. Such patients could be followed up with further rehabilitation to restore a normal speaking voice with high-quality daily life.

The successful treatment of the laryngeal web relies on the resolution of dyspnea and the acquisition of a normal voice. Patients with type III and IV laryngeal webs generally showed more difficulty in obtaining a normal speaking voice. The clinical reports available on voice quality in children with laryngeal webs are relatively scarce. Moreover, the few clinical reports that are available on the evaluation of voice quality have been mainly presented from the physician's own competent judgment. Additionally, it is difficult to obtain objective data on voice quality from pediatric patients. In a clinical case report covering 22 children with laryngeal webs, Tery used his subjective judgment to describe the postoperative outcome of the children's voice quality [4]. Here, they treated the laryngeal web with a T-tube and found that 90% of the children had fair voice quality after surgery, with their daily lives not being affected. In subsequent studies, we need to follow up on the children who presented with laryngeal webs into adolescence and adulthood. Thus, we can obtain objective data to evaluate the quality of such patients' voices.

In 2010, Goudy reported 18 cases of congenital laryngeal web over a period of 25 years in their hospital [5]. This study covered the largest number of clinical cases to date. However, most of them were type I and II laryngeal webs, with only one case being a type IV laryngeal web. In the past 22 years, Lawlor reported a total of 16 cases of congenital laryngeal webs, which included two cases of type IV laryngeal web[6]. In the last 10 years, a total of seven children with type III and IV laryngeal webs were admitted to our hospital. Satisfactory results were achieved through open laryngoplasty, which was combined with T-tube implantation and reconstruction of the cricoid cartilage.

Conclusion

Children with type I and II laryngeal webs mainly presented with hoarseness and discomfort without any obvious respiratory distress. Children were usually older when they were admitted to the hospital. The patients with type I and II laryngeal webs recovered through one simple endoscopic surgery, while children with type III and IV laryngeal webs mainly exhibited hoarseness and respiratory distress, which required

an early tracheotomy. These children usually had a record of multiple visits to the hospital with multiple treatments. The ultimate treatment required for such patients was open laryngoplasty, combined with the implantation of a T-tube, and reconstruction of the cricoid cartilage by hyoid bone may play a crucial role in the treatment of congenital laryngeal webs with subglottic stenosis. T-tube implantation is effective in preventing the re-adhesion of the vocal cords. The recommended duration for the T-tube implant is 6 months. The recommended age for this surgery is 2 years old. The most common comorbidity of type III and IV laryngeal webs was subglottic stenosis, which was likely combined with other laryngeal diseases, such as vocal cord paralysis and laryngomalacia, along with systemic diseases, such as atrial septal defects.

List of abbreviations: CT computed tomography

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Figure-1 A 、 B 、 C Laryngoscopic photograph of the patient with laryngeal web type III and IV,D Laryngoscopic photograph of the patient with laryngeal web type I and II

Figure 2 A 、 B CT photo of the patient with laryngeal web type I and II,A Soft tissue shadow is seen in front of the vocal cords,B No stenosis is seen under the glottis .C 、 D CT photo of the patient with laryngeal web type III and IV,C The glottis segment is narrow,D The subglottic area is narrow.

Figure-3 The surgery was performed under general anesthesia with an anesthetic cannula inserted in the place of the tracheal tube in the neck of the child (Fig. 3-A). A portion of the hyoid bone was obtained and trimmed into a pike shape (Fig. 3-B,C). The cricoid cartilage affected was incised to remove the subglottic stenotic lesion tissues, the anaesthetic cannula was removed and a T-tube was inserted (Fig. 3-D). the T-tube should be slightly above the glottis (Fig. 3-F). the cricoid cartilage was repaired and reconstructed with the hyoid bone (Fig.3-E),The photo of T-tube(Fig.3-G)

Figure-4 The cricoid cartilage affected was incised to remove the subglottic stenotic lesion tissues, Afterwards, the cricoid cartilage was repaired and reconstructed with the hyoid bone.

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