

Congenital laryngeal webs:From evaluation to surgical management

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Abstract

Objective. To discuss the presentation, evaluation, and management of congenital laryngeal web. **Methods.:** The Institutional Board of Ethics of shanghai Children's Hospital approved this retrospective review of all patients who were diagnosed with laryngeal web at our institution within the past 10 years. Charts mined for age at presentation, presenting symptoms, degree/location of web, associated syndromes, number/type of surgical procedures, and postoperative outcomes. **Results.:** Ten patients were included (6 male, 4female). There were 3 type I \ II webs, 7 type III \ IV webs; Children with type I and II laryngeal web were mainly presented with hoarseness and discomfort without any obvious respiratory distress($P \geq 0.05$). Children were usually older when they were presented to the hospital($p < 0.05$), All the 3 children with type I and II laryngeal web recovered after a single endoscopic procedure. While children with laryngeal web type III and IV usually had a record of multiple visits to the hospital with multiple treatments($p < 0.05$) All of these 7 type III \ IV webs , the tracheotomy was performed early, The ultimate treatment required for such patients was open laryngoplasty. **Conclusions:**There is a need for alertness regarding the neonatus exhibiting hoarseness and weak crying. To achieve a satisfactory outcome, it is essential to perform accurate preoperative staging and assessment along with an appropriate choice of treatment and placement of the laryngeal stent.

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Key words :

congenital laryngeal webs \ T-tube \ open laryngoplasty \ hoarseness \ tracheotomy

Succinct key points:

1. Ten patients who has congenital laryngeal web were included .
2. There were 3 type I \ II webs, 7 type III \ IV web .
3. All the 3 children with type I and II laryngeal web recovered after a single endoscopic procedure.
4. children with laryngeal web type III and IV usually had a record of multiple visits to the hospital with multiple treatments.
5. The ultimate treatment required for such patients was open laryngoplasty.

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 the abnormal tissues in the glottic portion and accounts for approximately 5% [1] of the 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, which depends on the degree of involvement of the laryngeal tissues and the degree of obstruction in the trachea.

Patients with congenital laryngeal web are clinically presented 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, which involves a thin, membranous laryngeal web involving <35% of the vocal folds, ii) type II including laryngeal web tissues involving 35%-50% of the glottic portion, iii) type III, which involves the laryngeal web tissues comprising 50%-75% of the glottic portion, and the involvement of usually the lower glottic portion, and iv) type IV including the laryngeal web tissues with more than 75% of the glottic portion, completely blocking the glottis.

Materials and methods

The retrospective analysis was conducted on a total of 10 children diagnosed with the congenital laryngeal web. These children were admitted to our department between 2010 and 2020. Of 10 children, 3 had type I and II laryngeal web(Figure1-D) while the other 7 children had type III and VI congenital laryngeal web(Figure1-A \ B \ C). Of the 3 children with type I and II laryngeal web, all were females, whereas, in seven children with type III and IV laryngeal web(Table-1), six were male, and one was female. The oldest child with type I and II laryngeal web was five years old at the time of presentation, while the youngest one was only 1 year and 9 months old. The children were clinically presented with persistent hoarseness and no obvious respiratory distress. Although three children at our hospital did not undergo a tracheotomy, all of them recovered from a single endoscopic procedure. The seven children with type III and IV laryngeal web in our study were aged 2 years and 2 months old at the time of their presentation to our hospital, while the youngest one was 1 month old, with a mean age of 13 months. The tracheotomy was performed early, at an age ranging between 1 and 11 months, in all seven cases. In one of the seven children, we observed a combined atrial septal defect, while in another case, both atrial septal defect and laryngomalacia were observed. Six children out of the seven displayed subglottic stenosis, while one had vocal cord paralysis. Among the six children, four had an area of subglottic stenosis greater than 90%(cotton III°), while the other two had 40% (cotton Ideg)subglottic stenosis.

All the 3 children with type I and II laryngeal web recovered after a single endoscopic procedure. The other seven children with type III and IV laryngeal web eventually recovered in our hospital through open laryngoplasty and were successfully extubated with the tracheal tube. The preoperative enhanced CT of the neck was performed in all cases(Figure-2), which suggested varying degrees of stenosis in the glottic portion and also below the glottis. The youngest of the seven children with laryngeal web type III and IV who received open laryngoplasty was of age 1 year and 1 month; in contrast, the oldest was of 2 years and 11 months, with the majority of children being around 2 years of age. In all seven cases, a T-tube was implanted intraoperatively, whereas, in the six children with subglottic stenosis, the cricoid cartilage was incised to remove the scar tissue under the glottis. This cricoid cartilage was reconstructed using the autologous rib cartilage or hyoid bone (one rib cartilage and five hyoid bone). In one of the cases that had vocal fold paralysis, the right vocal fold was removed, and a T-tube was placed through the original

tracheal incision. This tube was placed for 3 months in one case, 8 months in another case, and 6 months in another five cases. The T-tube was placed slightly above the glottis, which acted as a support to prevent the adhesions. The children presented with choking while consuming fluids after surgery should be fed a soft or semi-liquid diet. Also, this problem of choking on food could be solved by dietary exercises. After placing the T-tube for a certain period, the children had to return to the hospital for a follow-up visit and also for a change in the regular tracheal tube. After one month of blockage, all seven children 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.

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 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 was positioned using intraoperative laryngoscopy, and the T-tube should be slightly above the glottis (Fig. 3-F). Afterwards, the cricoid cartilage was repaired and reconstructed with the hyoid bone (Fig. 3-E) and the incision was sutured.

We mention that the reporting guideline has been followed in this study.

Statistics

SPSS version 17.0 was used in all data analyses. A difference with a p-value <0.05 was considered as statistically significant.

Results and Conclusion

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

Children with type I and II laryngeal web were mainly presented with hoarseness and discomfort without any obvious respiratory distress ($P > 0.05$). Children were usually older when they were presented to the hospital ($p < 0.05$), and it was usually identified when their families found out that their voices had been hoarse for a long time. The patients with laryngeal web recovered through a simple endoscopic surgery ($p < 0.05$). However, children with type III and IV laryngeal web mainly exhibited hoarseness and respiratory distress, which required an early tracheotomy. The most common comorbidity of laryngeal web type III and IV was subglottic stenosis, which was probably combined with other laryngeal diseases such as vocal cord paralysis and laryngomalacia along with systemic diseases such as atrial septal defect. Children with laryngeal web type III and IV usually had a record of multiple visits to the hospital with multiple treatments ($p < 0.05$). The ultimate treatment required for such patients was open laryngoplasty and We suggest that these children should receive open laryngoplasty at around 2 years of age. The T-tube implantation is effective in preventing the re-adhesion of the vocal cords. However, it may cause discomfort, such as choking and coughing early on after the surgery. Thus, such patients need to be given a soft or semi-liquid diet. The recommended duration for the T-tube implant is 6 months. In children with subglottic stenosis, the lingual bone or autologous rib cartilage can be used simultaneously to reconstruct the cricoid cartilage. We recommend using the hyoid bone for reconstruction as it can be obtained in the same surgical area, reducing unnecessary trauma. Simultaneously, an open laryngoplasty can be performed for children having a combination of vocal cord paralysis. Open laryngoplasty can resolve the child's breathing difficulties and help in the successful removal of the cervical tracheal tube. We have advanced the age of surgery to 1 year 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.

Discussion

Infants with dysphonia and weak crying after birth should be highly suspected of the congenital laryngeal web. Also, infants who are presented with persistent hoarseness, recurrent laryngeal wheezing, or acute laryngitis after birth should be admitted to the Otorhinolaryngologic Department. In case of the exclusion of the laryngeal web, laryngoscopy may be adopted to confirm the diagnosis in newborns who exhibit weak crying and/or breathlessness after birth. The development of fiberoptic laryngoscopy and bronchoscopy has provided a great improvement in the diagnosis of congenital laryngeal web 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 brought out by the blind operation under an ambiguous diagnosis can be avoided.

As reported, the laryngeal web was often found with complications involving other laryngeal malformations. Also, in our clinical practice, some children with laryngeal web exhibited a combination of laryngomalacia and vocal cord paralysis. The laryngeal web was first proposed by Cohen, whose clinical typology offered some guidance for its clinical treatment. A claimed that type I and II laryngeal web were often found without any comorbidities while type III and IV laryngeal web were frequently combined with other structural malformations of the larynx or were presented as a manifestation of the clinical syndrome. The most common syndrome associated with the laryngeal web was the 22q11.2 deletion syndrome [3]. The type I and II laryngeal web often involved relatively thin tissues, where the performance of microscopic endoscopic surgery could produce a great outcome. This treatment improved the hoarseness and allowed the patient to speak in an almost normal voice. However, type III and IV laryngeal web were generally combined with subglottic stenosis, requiring open surgery to achieve a better outcome. This could firstly resolve 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, the open laryngoplasty was combined with T-tube and hyoid reconstruction of the cricoid cartilage, which was performed to treat the laryngeal web of type III and IV. This procedure showed excellent results. The T-tube was placed for 6 months, slightly above the glottis, to prevent the adhesions of the vocal cords. To support and enlarge the tracheal lumen and better address the subglottic stenosis, the cricoid cartilage was reconstructed using the hyoid bone. All children were successfully extubated, but some of them exhibited slight hoarseness even after surgery. 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 web generally showed more difficulty in obtaining a normal speaking voice. The clinical reports available on voice quality in children with laryngeal web 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. Also, it is difficult to obtain objective data of voice quality from pediatric patients. In a clinical case report covering 22 children with laryngeal web, 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 the subsequent study, we need to follow up on the children presented with the laryngeal web even in their adolescence and adulthood. Thus, we can obtain objective data to evaluate the quality of such patients' voices.

In 2010, Goudy reported 18 cases of the 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 the laryngeal web of type I and II, with only one case being laryngeal web type IV. In the past 22 years, Lawlor reported a total of 16 cases of the congenital laryngeal web, 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 web was admitted to our hospital. Satisfactory results were achieved through open laryngoplasty, which was combined with intraoperative T-tube and hyoid reconstruction of the cricoid cartilage. In conclusion, performing laryngo-

plasty combined with the implanting of a T-tube and reconstruction of the cricoid cartilage by hyoid bone may play a crucial role in the treatment of congenital laryngeal web with subglottic stenosis, providing good clinical outcomes.

Conclusion

There is a need for alertness regarding the neonatus exhibiting hoarseness and weak crying. Children with type I and II laryngeal web can recover through a simple endoscopic surgery, While children with type III and IV laryngeal web may required an early tracheotomy, And the ultimate treatment required for such patients was open laryngoplasty. To achieve a satisfactory outcome, it is essential to perform accurate preoperative staging and assessment along with an appropriate choice of treatment and placement of the laryngeal stent.

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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

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