Anomalies of the Larynx

12

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12.1 Introduction

Laryngeal anomalies can be encountered by pediatric surgeons as a cause of respiratory distress, especially in infants. Even if many of these anomalies are among the field of competence of otolaryngologists, pediatric surgeons must be aware of the different conditions that can affect the pediatric larynx, often in association with other surgical pathologies. Laryngeal anomalies may represent a real challenge, and they are a paradigm of condition in which a team approach is rewarding. During the last years, in some Centers, as in Gaslini Institute, airway teams have been created to manage these complex patients, including among the others pediatric and cardiac surgeons, otolaryngologists, pulmonologists, anesthesiologist, neonatologists, intensivists, radiologists, gastroenterologists, etc. [1]. Also, in centers without a structured airway team, pediatric surgeons can be called at the bed of these patients just to perform an endoscopic airway evaluation or a tracheostomy, or they are involved as the patients have multiple malformations, including some surgical interest. There are pediatric surgeons who have developed a particular interest in this field, and they can manage these conditions deeply, often in cooperation with the specialists of the airway team. In our opinion, every pediatric surgeon must know the main laryngeal anomalies, be ready at recognizing them, and offer to the patients the proper treatment or derive them a more specialized center. A common surgical procedure such as a tracheostomy must be correctly performed, and only the knowledge of the underlying condition and where and how to perform a tracheostomy is crucial for the chance of success of the subsequent treatments.

In this chapter, we will treat briefly the more common laryngeal anomalies from a pediatric surgeon's perspective. Some of the laryngeal anomalies will be treated in the tracheal anomalies chapter to avoid repetitions.

12.2 Classification

We can distinguish congenital and acquired laryngeal anomalies. The incidence of the congenital anomalies is around 1 in 10,000 to 1 in 50,000 live births [2]. The most common of these is laryngomalacia, accounting for more than 60%

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G. Mattioli et al. (eds.), *Pediatric Thoracic Surgery*, https://doi.org/10.1007/978-3-030-74668-1_12

of them. The other two more common congenital conditions are congenital stenosis and vocal cord palsy. Among the uncommon anomalies, we mention webs or atresia, hemangiomas, laryngoceles, and laryngeal clefts.

Among the acquired lesions, the post-intubation stenosis is by far the most common.

An important issue related to laryngeal stenosis (congenital or acquired) is the classification of severity. Classically, the stenoses are classified in four degrees of severity, based on the percentage of the lumen involved by the stenosis:

Grade 1: 0–50%; Grade 2: 50–75%; Grade 3: 75–99%; Grade 4: 100% (no lumen).

Monnier modified the Myer Cotton classification by adding a letter to each degree, according to the presence of comorbidities (b); glottic involvement (c); both comorbidities and glottis involvement (d). For example, stenosis of 80% involving the vocal cord and the subglottic region in a normal baby will be grade 3c; complete stenosis starting from above the vocal cord up to the first tracheal ring in a Down's syndrome and cardiopathic patient will be classified as grade 4d.

12.3 Clinical Presentation, Diagnosis, and Treatment

Clinically, many of the laryngeal anomalies will present similarly: inspiratory stridor, retractions, croup, repeated respiratory infections, or acute and severe onset of respiratory distress, sometimes associated with transient respiratory conditions.

The key to the correct diagnosis is represented by the endoscopy. The other investigations, in particular radiological, may be sometimes useful to complement the endoscopic findings, but they cannot substitute the endoscopy. In our experience, many patients with respiratory symptoms undergo unnecessary or inconclusive CT scan or MRI. It is important to underline that in most cases endoscopic evaluation will be the only tool that will prompt the physician to distinguish among very different conditions with similar clinical aspects. Endoscopy should be performed by experts, as the patient has to be properly sedated and assisted, and this can be very challenging in a patient with an unknown respiratory condition. Basically, two types of airway endoscopy are available and are both useful: flexible and rigid. Through flexible endoscopy, we can evaluate easily and without a big risk of airway trauma the vocal cord mobility, the dynamics of the airway during spontaneous quiet breathing, and forced inspiration and expiration. Rigid endoscopy will show us in a more detailed way the anatomic features of the airway with a big magnification. Both evaluations are useful and ideally should be performed.

- Laryngomalacia:

  It is the most frequent cause of stridor in an infant. Many infants are labeled with the diagnosis of "laryngomalacia" (LM) just because of their inspiratory stridor, but this is incorrect, as other less frequent conditions could give the same symptoms. The diagnosis of LM is not possible without flexible endoscopy, which shows the typical supraglottic obstruction of the larynx from arytenoids, epiglottis, or both. Usually, LM is due to very short aryepiglottic folds (LM type 1). This causes an inward prolapse of arytenoids during inspiration. Less frequently, the obstruction is due to an omega-shaped epiglottis (LM type 2) or a posterior movement of the epiglottic toward the laryngeal lumen (LM type 3). Sometimes there is a combination of more types of LM.

  There is a natural tendency of LM to improve with the age, so in many cases, infants with stridor without other matters of concern (growth retardation, apneas, cyanosis, feeding problems) can be treated conservatively with simple observation, waiting for a spontaneous improvement. In more severe cases, endoscopic supra-glottoplasty is the treatment of choice that can be performed with CO_2 laser or with cold instruments and is able to resolve virtually all cases of LM. Supra-glottoplasty consists in cutting the aryepiglottic folds (in LM type 1) or reshaping the lateral margins epiglottis (in LM type 2) or fixing the epiglottis anteriorly to the tongue (in LM type 3). In

our Institute, ENT surgeons correct LM usually with CO_2 laser, with the patient in spontaneous breathing, at the end of the procedure the patient is sent to the ward, without the need of intensive care treatment.

- Vocal cord paralysis.

Vocal cord paralysis (VCP) can be congenital (idiopathic) or acquired, usually after surgical procedures (on the neck, mediastinum, heart, and big vessels) or neurological conditions. VCP can be unilateral or bilateral. The latter usually causes significant inspiratory dyspnea, as vocal cords (VC) are not abducted during inspiration, thus causing the obstruction. In bilateral VCP, the voice is usually not much impaired, while the patients with monoliteral VCP are frequently dysphonic. The diagnosis is based on flexible endoscopy with the patient awake or very lightly sedated. The correct diagnosis of VCP, in particular bilateral, requires experience. If sedation is too deep, impaired movement of VC can be due to the sedation itself. The endoscopist should therefore be sure that the patient is breathing alone and sufficiently awake so that the movement of the VC cannot be influenced by any drugs. Sometimes, bilateral VCP can be missed, as the VC moves symmetrically during the spontaneous breathing in a passive way for an inward negative pressure (like a suction) generated by the inspiration. In patients with the normal movement of VC, they spread during inspiration and move medially during expiration, while in patients with bilateral VCP, we observe the reverse: VCs are approximated toward the midline during inspiration (for the abovementioned suction effect), and they open more during expiration, spread apart by the air flowing out the trachea.

Regarding treatment, most bilateral VCP classically requires a tracheostomy. To avoid tracheostomy, bilateral endoscopic cricoid split and balloon dilatation seems a promising technique for neonates with congenital bilateral VCP. After the procedure, a big endotracheal tube is left as a stent for 10–15 days, then the patient is extubated, and usually does not require a tracheostomy.

Unilateral VCP usually does not require any urgent treatment, as the respiratory distress is absent or very little. The issue is more on the voice quality.

It is well known that VCP, both congenital and acquired, can improve spontaneously, so it is usually accepted as a conservative treatment, waiting for spontaneous improvement. The point is how much time has one to wait for it (in our experience spontaneous recovery can occur even after 6 or more years). To evaluate the recovery of VC movements, classically endoscopic evaluations are repeated every 6–12 months. More recently, ultrasound evaluation has been demonstrated to be useful for detecting VC movement in a less invasive way [3]. Another tool for evaluating VC function is the measurement of the evoked potentials through a little electrode inserted in the laryngeal muscles during an endoscopy. This gives us information about the innervation of the muscles and possibility of recovery.

The treatment of VCP is controversial. Many endoscopic techniques have been described, trying to improve the glottis space and improve airflow. Among them, posterior cordotomy [4], VC lateralization [5], and arytenoid latero-abduction [6] are the most popular. Another option is represented by a posterior cartilage graft that can be performed either endoscopically [7] or through an open approach (Figs. 12.1 and 12.2).

- Congenital laryngeal stenosis (Fig. 12.3):

Congenital subglottic stenosis is defined as a subglottic diameter of less than 4 mm in a neonate and less than 3 mm in a preterm baby. Congenital stenosis is much less frequent than acquired stenosis. The cricoid cartilage is abnormal in size or shape. The most frequent types of congenital subglottic stenosis are elliptic cricoid; thick anterior lamina of cricoid ring; and generalized thickening of cricoid ring. Clinical presentation is usually biphasic stridor or recurrent episodes of croup. Some cases are asymptomatic as the stenosis is not critical (grade 1) and become clinically evident during an episode of airway infection. The diagnosis is made during endoscopy

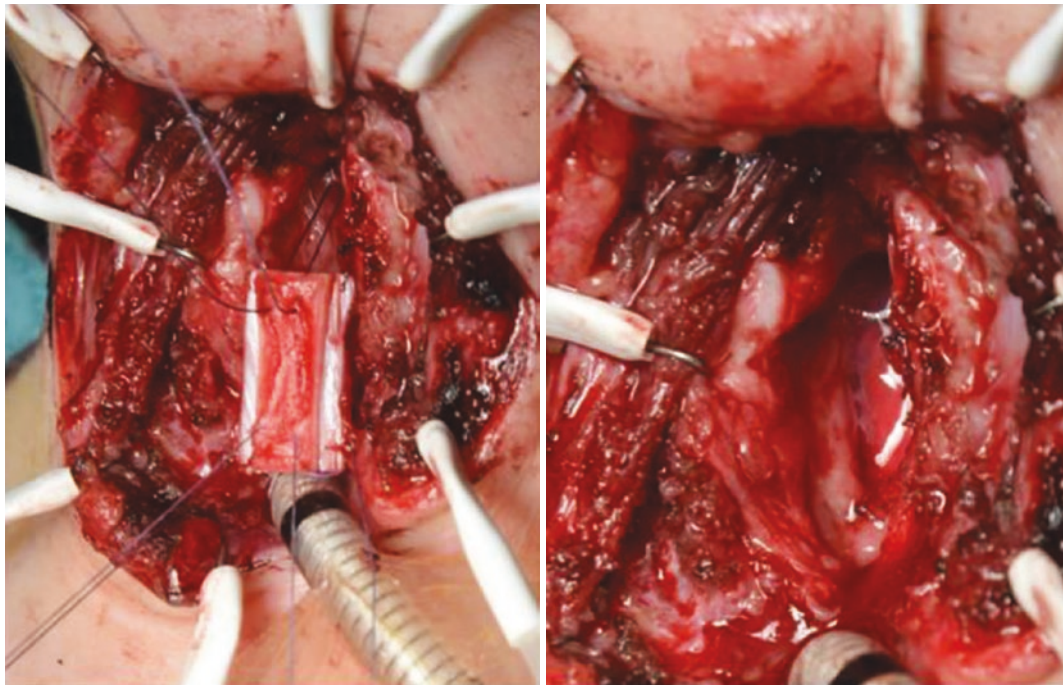

Fig. 12.1 Posterior cartilage graft for treatment of a congenital vocal cord paralysis: the graft before (left) and after (right) suture to the posterior cricoids plate

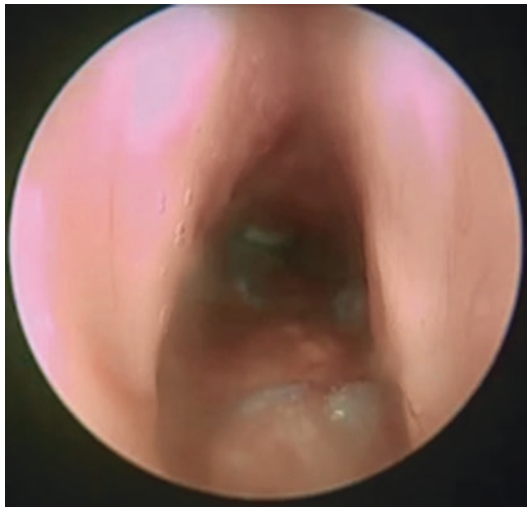

Fig. 12.2 Endoscopic view of a posterior graft

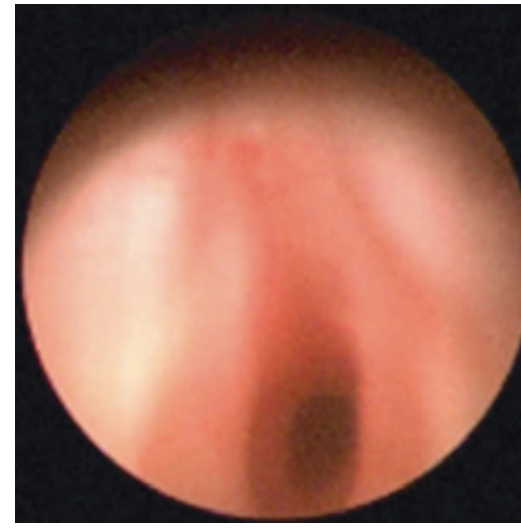

Fig. 12.3 Congenital laryngeal stenosis

(Fig. 12.4). It is important to calibrate the diameter of the stenosis with a tube in order to classify it into one the four degrees. Congenital subglottic stenosis does not respond to endoscopic treatment, as the cartilage is anomalous, and this cannot be modified endoscopically. Open laryngotracheal reconstruction with an anterior and posterior cartilage graft or partial crico-tracheal resection are the treatments of choice for symptomatic stenosis.

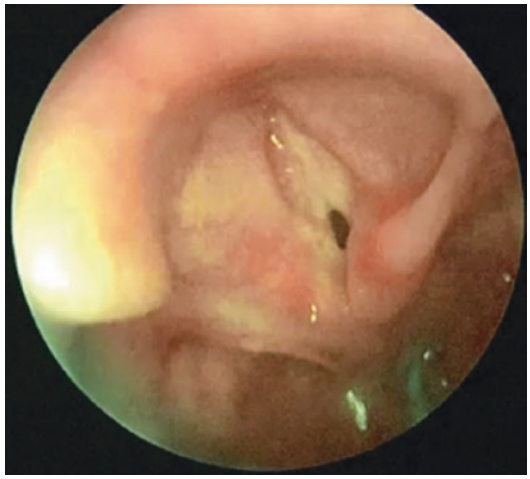

Fig. 12.4 Laryngeal web type 4

- Acquired laryngotracheal stenosis: see the chapter on tracheal malformations.
- Laryngeal clefts: see the chapter on tracheal malformations.
- Laryngeal webs and atresia:

  Laryngeal webs (LW) are due to defects of recanalization of the primitive larynx. Laryngeal atresia is the extreme form of web and is incompatible with life. LW are classified according to their severity in 4°:

  Type 1 is a thin anterior web occluding less than 35% of the glottis, and there is no subglottic involvement; type 2 is a slightly thicker web occluding 35–50% of the glottis and extending sometimes minimally in subglottic region; type 3 is a thick web occluding 50–75% of the glottis and always extending in subglottic region; and type 4 occludes 75–90% of the glottis with significant cartilaginous subglottic stenosis (Fig. 5).

  The symptoms are related to voice quality (from mild hoarseness in grade 1 to aphonia in grade 4) and in more severe grades respiratory distress is present.

  The treatment can be performed endoscopically to improve the quality of the voice in grades 1 and 2. In grade 3, an open approach is sometimes required if the subglottic component of the malformation is significant. In case of prevalent glottic web without subglottic significant stenosis, endoscopic approach can be preferred. In type 4 an open approach is always required with the aim of resolving not only the glottic web but also the subglottic stenosis (Fig. 12.4). In many of LW grade 4, a tracheotomy is required before the repair.

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