Reconstructive Surgery of the Laryngotracheal Junction

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ABSTRACT

The laryngotracheal junction is an anatomical region with special pathophysiological features. This review presents clinical pictures and malformations that manifest pre-dilectively at this localisation in children and adolescents as well as in adults. The diagnostic procedure is discussed. The possibilities of surgical reconstruction are presented depending on the pathology and age of the patient.

Contents

1. Clinical anatomy

The term "laryngotracheal junction" originates from the clinical observation of some of the peculiarities that this section of the upper airway shows, which is only a few centimeters long. Anatomically, it is the zone that begins cranially at the inferior border of the subglottic shelf of both vocal folds and extends over the cricoid cartilage to the first tracheal rings. The caudal boundary is not clearly defined. This area takes on special significance as a predilection site for some clinical pictures that either predominantly or even exclusively occur here. These include infantile "pseudocroup" in acute viral infections, true croup in the setting of diphtheria, respiratory manifestations of granulomatosis with polyangiitis (formerly Wegener's disease), idiopathic progressive subglottic stenosis, and the numerically particularly significant scarred subglottic stenosis, such as after long-term intubation, tracheostomy, or caused by other trauma. The laryngotracheal junction thus describes the actual subglottis and the beginning of the cranial trachea as a biologically homogeneous but anatomically heterogeneous unit. The submucosal tissue on the cricoid plate seems to be of special importance. The cricoid has the anatomical peculiarity of being the only section in the airway to be completely cartilaginous over the entire circumference. Contrary to its name, the cricoid cartilage is usually elliptically shaped. Dorsally, it forms the articular surfaces for the arytenoid cartilage and thus supports the dorsal glottis. It is elastically connected to the thyroid cartilage via the cricothyroid membrane; by means of contraction of the cricothyroid muscles, the only laryngeal muscle innervated by the superior laryngeus muscle, it tilts the arytenoid cartilages dorsally and thus pretensions the vocal folds (frame tension). At the level of the cranial cricoid cartilage, the recurrent nerve leaves its sulcus between the trachea and the esophagus and, after branching in a number of different ways, moves to the back of the cricoid cartilage plate in order to reach the interior of the larynx via its cranial edge.

2. Diagnostics

The leading symptom of laryngotracheal junction disease is inspiratory stridor. However, it is not possible to draw any further conclusions about the type and extent of the stenosis auditorily. Simple indirect laryngoscopy often misses subglottic changes, and transnasal flexible endoscopy offers greater diagnostic certainty. Nevertheless, despite typical symptoms, the correct diagnosis is surprisingly often made rather late; the misdiagnosis of bronchial asthma, in particular, is not uncommon. The most reliable diagnostic measure is endoscopic examination under short anesthesia with the aid of rigid optics or surgical microscope. Only in this way can all details be comprehensively identified and taken into account. Another advantage is the possibility of palpatory examination or manipulation. Preoperative diagnostic endoscopy is the most important measure for correct diagnosis and, consequently, for correct selection of the therapeutic procedure. Its importance, as well as the importance of precise, accurate and reproducible execution, can therefore hardly be overestimated.

The simplest and often most sufficient technique is brief inspection in apnea. In more complex situations or simultaneous interventions, jet ventilation should be preferred as a reliable and cost-effective procedure. Alternatively, intermittent intubation with a thin tube is possible. In this case, care should be taken to perform a primary endoscopy before the first intubation to avoid tube-related changes that may thwart an accurate diagnosis. Provided airway stenosis is investigated with some regularity, more advanced ventilation techniques should be available to safely manage even highly complex ventilation situations. These include tubeless supraglottic interposed simultaneous high- and low-frequency jet ventilation, apnea oxygenation with high-flow oxygen, and the Ventrain system. Specially adapted jet ventilation devices ensure good oxygenation over longer periods even in very young children and neonates.

The patient is positioned in the same way as for microlaryngoscopy: with the head extended and the neck flexed (sniffing position). The often observed simultaneous extension of the neck and support of the shoulder area with a positioning pillow should be avoided, as they complicate the exposure of the larynx [7].

The anesthesiologist's spatula laryngoscope is used to expose the larynx, and visualization is performed with a 0 ° optic of 30 cm length, correspondingly shorter in children. Routine video projection and recording is highly recommended. It allows all participants to be informed simultaneously about the current examination and thus about any risk situations. The interdisciplinary discussion of the case on the basis of the recorded high-resolution videos is extremely helpful for exact diagnostics and therapy planning.

After optimizing the image parameters and aspirating secretions, a slow camera movement is carried out from supraglottic to carina and back. The topographic relation of the pathology can be reliably established by markings on the endoscope via a fixed point (row of teeth).

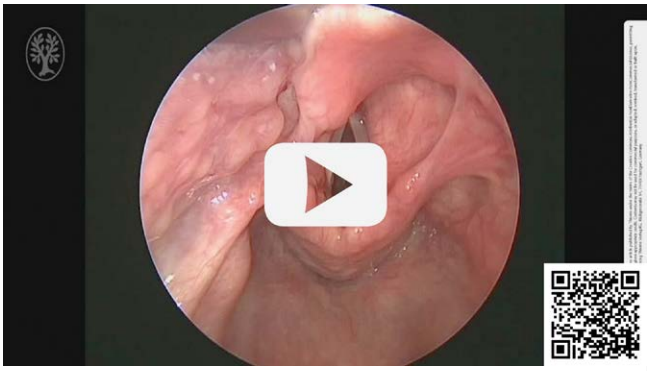
Assessment of airway stenosis should follow a structured and reproducible procedure. The most important parameters here are severity of stenosis, stenosis length, involvement of cricoid cartilage and vocal fold level, consistency and degree of activity. A recent consensus paper from the ELS provides a good guide for the evaluation of airway stenosis [17]. Based on the Cotton-Myer classification, an ELS classification was created that is easy to use and memorize (▶ **Table 1**), yet provides more information and higher prognostic value [27].

▶ **Table. 1** Classification of an airway stenosis according to the European Laryngological Society (ELS). Sub-locations. Supraglottic/glottis/subglottic/tracheal. The sum of the sub-locations is given with a letter (a = 1 location, b = 2 locations etc.). Comorbidity: the presence of cardiopulmonary concomitant diseases is coded with " + ". Example: an isolated subglottic stenosis with 60 % obstruction in a patient with COPD corresponds to grade IIIa + .

Grade I	Grade II	Grade III	Grade IV
< 50 % Narrowing of the cross-sectional area	50–70 % Narrowing of the cross-sectional area	70–99 % Narrowing of the cross-sectional area	Complete atresia
Number of sub-localizations	Number of sub-localizations	Number of sub-localizations	Number of sub-localizations
comorbidity	comorbidity	comorbidity	comorbidity

In adults, thin-slice computed tomography (with contrast enhancement if necessary) is the primary imaging modality, especially for stenoses of the laryngotracheal junction. Special attention is given to changes in the cricoid cartilage and cricoid plate. Magnetic resonance imaging is often good at visualizing the near pathognomonic submucosal soft tissue thickening in progressive idiopathic subglottic stenosis. Three-dimensional reconstructions may facilitate rapid assessment of the topography of the stenosis.

In infancy and especially toddlerhood, on the other hand, imaging techniques are only of importance for special questions due to the weak contrast between cartilage structures and surrounding soft tissue.



▶ **Video. 1** Scarred subglottic stenosis after intubation ventilation.

3. Airway stenosis in adults

3.1 Intubation-related stenosis

In adults, the development of tracheal stenosis by intubation is a rare event (▶ **Video. 1**). In addition to the duration of tube insertion [20], any intubation trauma is of particular importance, as it may occur especially in emergency situations. The underlying pathomechanism has not been conclusively clarified, but a multifactorial genesis can be suspected. Additional risk factors are an individual disposition and esophago-laryngeal reflux. Differentiation, especially from idiopathic subglottic stenosis, can be difficult. As a basic rule, if there is a time interval of more than 2 years between intubation and the first appearance of symptoms, a connection must be considered unlikely. Typically, intubation-associated stenosis manifests at the level of the cricoid cartilage and is short-stretched. In the early stage of stenosis formation, before completion of complete scarring, endoscopic therapy may be successful.

▶ **Video. 2** Typical picture of an idiopathic progressive subglottic stenosis (IPSS). This disease occurs nearly exclusively in females of child-bearing age.

3.2 Idiopathic progressive subglottic stenosis (IPSS)

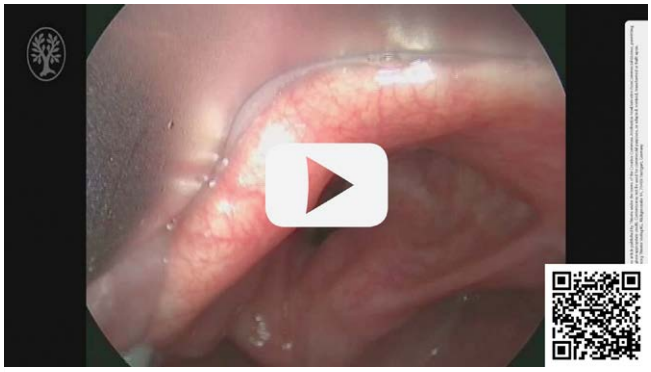
IPSS affects almost exclusively women of childbearing age (▶ **Video. 2**). The symptomatology of inspiratory stridor usually develops over years, but accelerated courses are possible [29]. A precipitating event must be ruled out by careful history taking; intubation more than 2 years ago may be considered etiologically insignificant. Particular attention should be paid to exclusion of systemic disease, especially polyangiitis with granulomatosis (Wegener's disease). Accelerated disease progression is typical during pregnancy. In combination with the clear gender predisposition, this has always been interpreted as an indication of a possible connection with the metabolism of female sex hormones. Studies to date have been contradictory in this regard, but recently there has been evidence that an imbalance between different types of estrogen and progesterone receptors may be etiologically significant, the importance of which for wound healing has been documented elsewhere [4–6]. The endoscopic appearance is inconsistent, but always shows an inconspicuous epithelial surface, mostly with submucosal, corkscrew-like scar strands. Almost pathognomonic is an increase of submucosal tissue especially in the area of the cricoid cartilage plate, which cannot be explained nosologically. Numerous etiologic models are discussed in the literature, ranging

from esophagotracheal reflux to microtrauma due to coughing attacks to chronic mycoplasma infections [3, 8, 9]. However, none of these explanatory models can plausibly explain the clinical presentation, the course of the disease, and especially the sex distribution. Proposals for therapy are correspondingly inconsistent. Some authors interpret IPSS as fibrotic inflammation in the sense of a limited or localized systemic disease, from which it is concluded that surgical repair does not adequately address the pathophysiologic processes. However, the application of endoscopic procedures, usually consisting of laser-based interventions, possibly in combination with high-pressure balloon dilatation as well as intralesional injections with corticosteroids, show rather disappointing results in many series with, in particular, high treatment frequency with only short symptom-free intervals [19, 23].

Open surgical reconstruction aims to remove the zones of pathology as completely as possible. The procedure of choice is cricotracheal resection (CTR, see section on surgical procedures) – corresponding data in the literature show predominantly good to very good results [1, 2, 10]. On the other hand, however, it is more time-consuming and the risk of complications should not be underestimated, especially in this patient group, which is distinguished from other forms of stenosis by a higher tendency to re-stenosis.

3.3 Tracheostomy

The most common form of tracheostomy-associated tracheal stenosis is the so-called "A-frame" deformity (▶ **Video. 3**). The cause is a loss of tracheal anterior wall as it develops iatrogenically due to an overly generous resection (especially with a necrotized Björk flap) or after perioperative infection with subsequent necrosis. The lack of anterior wall stability leads to instability of the tracheal sidewalls, which then medialize in the shape of the letter A. The apt term "pseudoglottic" stenosis has also been coined for this condition. Another significant risk factor is injury to the cricoid during tracheostomy, leading to chronic perichondritis with subsequent stenosis. Depending on the individual situation, other patterns of injury are of course encountered. The initially suspected higher frequency of laryngotracheal stenosis after dilated puncture tracheotomy cannot be verified neither statistically nor by our own observations [14].



▶ **Video. 3** The so-called "A-frame" deformity of the tracheal develops typically after closure of a tracheostoma.

3.4 Systemic diseases

In the majority of cases, the diagnosis of systemic diseases can be confirmed by a detailed history or by the presence of other manifestations. In particular, polyangiitis with granulomatosis may result in a larval initial finding in the subglottic larynx [28]. Endoscopic diagnosis shows a typical picture in most cases, whereas both biopsies and serological parameters may not always provide clarity, especially in the early phase. To clarify idiopathic subglottic stenosis, which can be difficult to differentiate from larval systemic diseases, a complete diagnosis in the sense of an exclusion procedure is obligatory.

4. Airway stenoses in pediatric patients

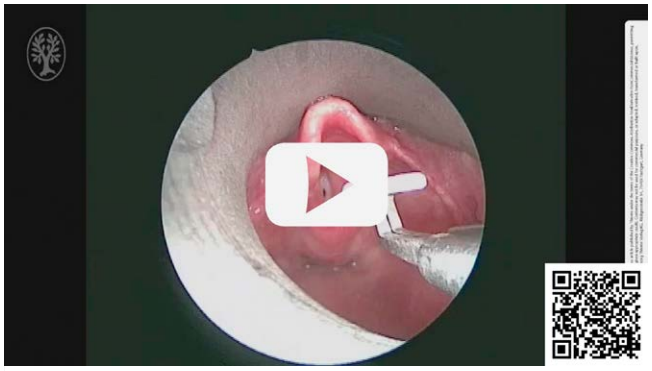
4.1 Subglottic stenoses

The cricoid cartilage is the only section in the airway that is completely cartilaginous and already physiologically has the smallest cross-section. Together with numerous other factors, this makes the cricoid a predilection site for the development of stenoses. Accordingly, the largest number of all acquired and congenital clinical pictures that can lead to a narrowing of the lumen are found here. Intubation-associated lesions remain by far the largest group.

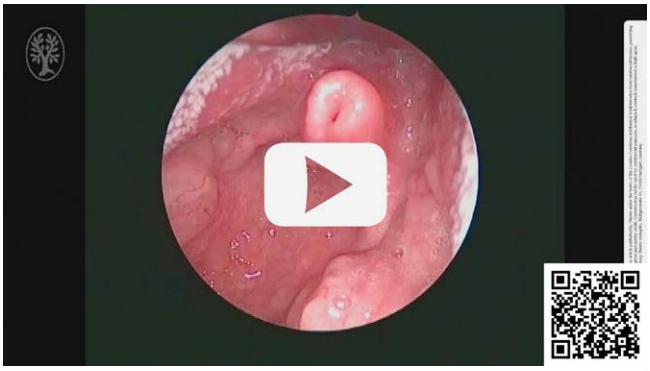
In perinatal medicine, the avoidance of a tracheostoma is a high objective. Unlike in adult intensive care, longer intubation times are also acceptable for this purpose. This is not only based on the understandable desire to avoid additional traumatization and stigmatization of the child and the relatives. Rather, neonatologists are aware that an early tracheostomy poses a considerable risk to the child. This is true not only for the surgical procedure itself, which is technically quite different from procedures performed at older ages and is challenging even for a skilled surgeon. For even after successful creation and healing of a tracheostoma, a significant risk remains: The combination of small neck dimensions with mostly well-developed subcutaneous adipose tissue and overall low reserve during apneic episodes leads to a high risk compared to accidental cannula dislocation. In infants, after removal of the tracheostomy tube, the tracheostomy canal is immediately functionally occluded by the surrounding soft tissues of the neck. Not infrequently, this is unnoticed until the compensatory reserve is already depleted. Therefore, deaths due to obstruction or dislocation of a cannula in an otherwise irritation-free tracheostoma occur again and again. Unlike at any other age, the tracheostoma in infants therefore poses a risk that requires special attention. From this derives the need for permanent monitoring of respiration. Because of this increased mortality, the insertion of a tracheostoma at the age of up to 18 months should only be undertaken in the absence of alternatives. This article presents other, possibly also costly appearing therapy procedures, which should be intensively applied and exploited in order to come as close as possible to the goal of tracheostoma avoidance.

4.2 Glottic stenoses

Interarytenoid fibrosis is a typical but widely unknown complication after intubation. Scarring in the posterior commissure results in mechanical fixation and/or ankylosis of the arytenoid cartilages. Differentiation from neurogenic vocal fold arrest is clinically difficult; testing passive mobility of the arytenoid cartilages during microlaryngoscopy is the diagnostic gold standard (▶ **Video. 4**).



▶ **Video. 4** Fixation of the arytenoid cartilages by interarytenoid fibrosis.



▶ **Video. 5** Typical laryngomalacia with curled epiglottis and mucosal hypertrophy of the arytenoid cartilages with shortened aryepiglottic fold. The video shows the insufficient laryngeal entrance during spontaneous breathing.

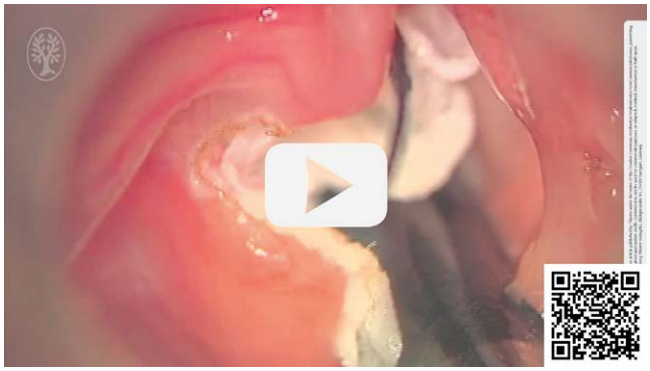
Surgical therapy, as with all glottic stenoses, must aim to achieve the best possible compromise between voice and breathing. Although tracheostomy largely avoids this dilemma, it is mostly perceived as unacceptable, so some voice deterioration is accepted as the price of tracheostomy avoidance. Numerous individual factors determine the choice of surgical approach; in most cases, at least a subtotal arytenoidectomy is required. If this is not sufficient, an expansion of the cricoid cartilage plate in the sense of a posterior laryngotracheal reconstruction (LTR, see section on Surgical Procedures) must be performed. Only in carefully selected cases of mild interarytenoid fibrosis surgical scar separation by laser division alone can be sufficient, but this must then always be combined with passive laterofixation.

4.3. Supraglottic stenoses

4.3.1 Laryngomalacia

The most common cause of stridor in newborns is changes in the supraglottis leading to a tendency to collapse on inspiration. The term laryngomalacia has become accepted as a generic term describing the functional interaction of immaturity of the juvenile larynx resulting in instability of the epiglottis and relative hypertrophy of the mucosa in the region of the arytenoid cartilages. There is also usually a shortening of the aryepiglottic folds in the sense of a discrete malformation. The unstable epiglottis and hypertrophic mucosa of the arytenoid cartilages can be sucked into the glottis by the flow of respiratory air (▶ **Video. 5**).

Laryngomalacia is found to be the primary cause in up to 60 % of infants with stridor. The stridorous breathing sound typically begins on the day of birth or a few days later. The worsening of symptoms is particularly noticeable with exertion, such as drinking or crying. The voice is completely unaffected. The breathing sound may be position-dependent and usually decreases in a prone position. Inspiratory stridor, which is usually low frequency, usually increases during the first months of life and reaches a maximum 3 to 6 months after birth if untreated. As the child grows, there is usually a spontaneous improvement in the breath sound, so that in more than 90 % of cases there are no longer any abnormalities after the 18th month of life. However, in approximately 10 % of cases, chronic airway obstruction can lead to apneas, impaired feeding with failure to thrive, weight loss, and pulmonary distress. This severe form of laryngomalacia requires further evaluation to consider possible surgical treatment options. However, milder forms may also be an indication for minimally invasive therapy, especially if the parents are constantly concerned about the breath sounds they perceive as threatening.



▶ **Video. 6** The surgical treatment of laryngomalacia depends on the findings while the main steps comprise a reduction of the arytenoid mucosal and cutting the aryepiglottic folds.

The different forms of laryngomalacia have found expression in numerous classifications. The generic term supraglottoplasty covers all measures to stabilize the laryngeal entrance. The cutting of the mostly shortened aryepiglottic folds leads to a reduced tendency to prolapse in the direction of the glottis due to the natural elasticity of the epiglottis. In cases of mucosal hypertrophy in the region of the cartilagines cuneiforme and corniculata, their reduction is indicated with strict sparing of the posterior commissure. In rare cases, it is necessary to “trim” the epiglottis by means of reduction of the mucosa at its free edges.

Supraglottoplasty (▶ **Video. 6**) is a reproducible procedure of manageable difficulty. Optimal exposure and maximum tissue-conserving technique are essential. Complications can only occur if the procedure is too aggressive. The local tissue reaction remains so low that extubation is possible immediately after the procedure. The occurrence of dysphagia is rarely observed and then only transitory.

The most serious complication is the development of supraglottic stenosis due to excessive scarring. The best prophylaxis consists of restrained mucosal resection and meticulous protection of the posterior commissure.

The high success rates of more than 95 %, the minimal invasiveness, and the possibility of simultaneous performance during diagnostic endoscopy make supraglottoplasty an elegant and effective procedure that should be indicated at a low threshold.

5. Malformations

5.1 Glottic web

Congenital "web" describes a congenital synechia of both vocal folds, typically affecting the ligamentous portions of the glottis. The diagnosis is usually obvious endoscopically, but less pronounced findings are not infrequently missed on flexible endoscopy because the vocal folds appear to be passively attached to each other. Although it usually appears as a delicate and short-stretched membrane, it is a complex malformation of varying extent that always reaches the cricoid cartilage anterocaudally (▶ **Video. 7**). Therapy depends on the extent of the finding: if the glottic stenosis is respiratory, normalization of breathing is the primary goal. In the long term, however, the goal of minimally disrupted voice production must be considered. Therefore, transection or even piercing alone is not adequate and should be used only in an emergency. Inadequate sole transection of the membrane may lead to an increase of stenosis up to complete atresia of the glottis. The treatment of choice is endoscopic separation of the synechia with passive insertion of a placeholder. Tracheostomy, even transitory, should be avoided. This procedure is readily available when the patient weighs about 10kg or more. Up to this point, a transitory glottic dilatation may be useful. Primary definitive reconstruction is only indicated in individual cases of simultaneous laryngeal malformations, especially subglottic stenosis.

5.2 Congenital vocal fold immobility

Congenital bilateral vocal fold immobility is probably neurogenic in origin. A precise pathophysiologic explanation is not known. The position of the immobile vocal folds may vary widely and provide for different clinical distress in the children.

Even for the experienced examiner, assessment of vocal fold mobility in the newborn can be very difficult and represents one of the few clear indications for flexible endoscopy in the awake infant. Comanifestation of vocal fold immobility and laryngomalacia occurs regularly. Therefore, when evaluating stridor in laryngomalacia, the mobility of the vocal folds should be assessed.

Electromyography is not useful at this age.

The spontaneous onset of glottic motility has been described and demonstrated several times in our own patients. Therefore, ablative glottic dilatations should not be considered until after 12 months of age. Up to this time, depending on the clinical symptoms, the insertion of a tracheostoma may be unavoidable, although this is associated with a considerable morbidity, especially in this age group. In individual cases, a transitory lateralization of a vocal fold by means of a traction cord can improve respiration and help to avoid placing a tracheostoma.

5.3 Congenital subglottic stenosis

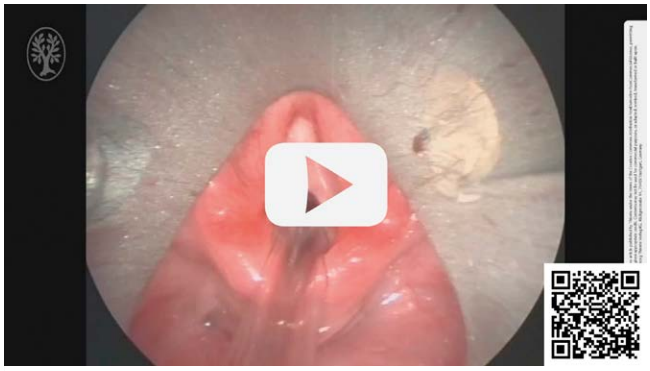
Genuine hypoplasia of the cricoid cartilage results in a reduced diameter which, depending on its severity, can lead to primary obstruction of the newborn's breathing. If this obstruction leads to emergency intubation of the infant after birth, the stenosis is later almost indistinguishable from intubation trauma.

Congenital cricoid stenosis is characterized by predominantly inspiratory stridor occurring immediately postpartum or a few weeks after birth. External causes and especially intubation are typically not present. For diagnosis, rigid endoscopy with preserved spontaneous breathing is the tool of choice. This must be done with special care, as only mild additional mucosal swelling can lead to complete occlusion of the remaining lumen. Imaging techniques are not helpful in diagnosis.

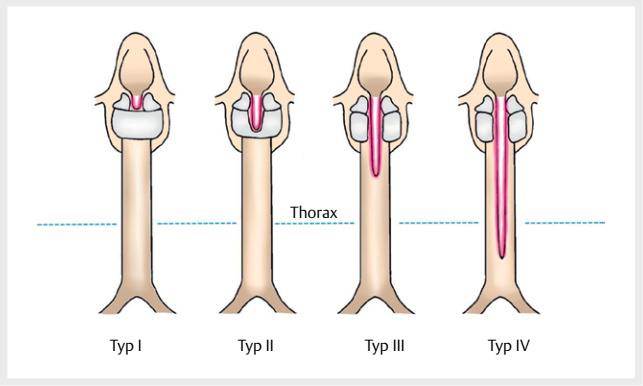
If intubation is unavoidable, it should be performed with the thinnest possible tube and for a short time. Tracheostomy can and should be avoided as much as possible, since cricoid dilatation is possible immediately after diagnosis. The gold standard in the treatment of congenital cricoid stenosis is laryngotracheal reconstruction (LTR-S, see section on surgical procedures) with autologous thyroid cartilage (▶ **Fig. 1a-c**). This is also possible in premature infants from 1,000 g birth weight.

5.4 Dorsal laryngotracheal cleft

A cleft in the dorsal larynx becomes clinically conspicuous with aspiration, recurrent respiratory infections and failure to thrive [16]. The edges of the clefts passively attach to each other; palpation of the cricoid cartilage plate with active spreading of the posterior



▶ **Video. 7** The congenital synechia of both vocal folds is a complex malformation with different severity that always reaches the cricoid cartilage antero-caudally.



▶ **Fig. 1** Classification of laryngeal clefts according to Benjamin and Inglis.

commissure is crucial for diagnosis; and is only possible during microlaryngoscopy. Testing for the presence of a laryngeal cleft should also always be considered during endoscopic diagnosis of esophageal atresia due to a coincident association of the two [13].

The clinical classification of cleft severity was defined by Benjamin and Inglis (▶ **Fig. 1**) on the basis of anatomic and prognostic criteria and has proven effective in practice.

More extensive forms with cleft formation beyond the cricoid (type III according to Benjamin and Inglis) or extending into the intrathoracic trachea (type IV) usually lead to respiratory insufficiency already in the neonatal period due to recurrent aspirations and pulmonary infections because of the lack of separation of esophagus and airway over a long distance. Posterior dislocation of the endotracheal tube is a typical sign of intubation, which is usually necessary; often only endoscopic intubation is successful.

Type IV clefts are extremely rare and usually associated with other malformations or genetically defined syndromes [24].

Reported comorbidities include prematurity, other tracheobronchial malformations, microgastria, and various gastrointestinal or cardiac malformations in addition to neurological impairments [24]. These require a multidisciplinary diagnostic and therapeutic approach that usually includes, among many other things, tracheostomy and chronic home mechanical ventilation and percutaneous tube feeding usually via PEJ over the first years of life. However, the severity of other tracheobronchial malformations as well as comorbidities in other organ systems should be meticulously analyzed before a decision for surgical therapy is made ab initio.

Mild forms (grades I and II according to Benjamin and Inglis) can be approached endoscopically (▶ **Video. 10**). If the entire cricoid cartilage plate is affected (grade III), open reconstruction via laryngofissure offers much higher chances of success. Surgical reconstruction of low-grade laryngeal fissures is performed immediately after diagnosis regardless of age if there are no other contraindications (e. g., low body weight in premature infants).

Neonates with type IV clefts require initial intensive care stabilization and sequential therapeutic approach based on clinical tolerance and stability. The historical mortality of more than 90 % could be reduced to less than 20 % in specialized centers. Tracheostoma and surgical correction of at least the intrathoracic tracheal portion of the fissure to secure the tracheal cannula position, as well as the creation of a gastrostoma with jejunal limb, are first emergency measures. These procedures require the use of a heart-lung machine or ECMO, for which a certain body weight must first be achieved. Surgical closure of the laryngeal fissure is performed in a further step usually about 2 to 3 months later depending on the clinical course.

6. Neoplasms

6.1 Hemangiomas

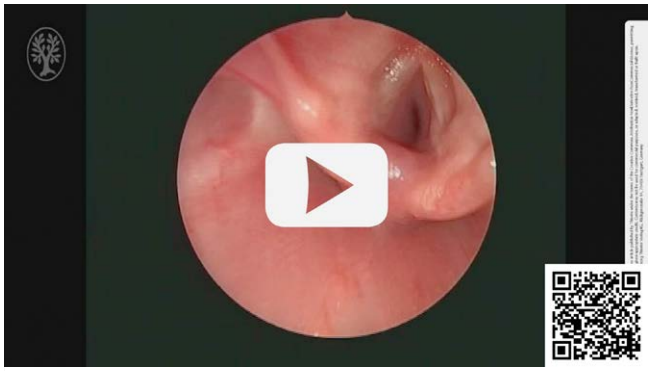
The congenital neoplasms are predominantly hemangiomas, which occur preferentially in the subglottis. Laser surgical therapy of connatal hemangiomas (▶ **Video. 8**) in the airway, which was frequently indicated in the past, has now been completely superseded by drug therapy with propanolol. The clinical appearance of subglottic hemangiomas is often relatively nonspecific and thus difficult to distinguish from other lesions. Because of the localization, sampling is prohibited. Therefore, in case of clinical suspicion, probatory weight-adapted drug treatment with a beta-blocker is the therapy of choice. If the suspicion is correct, an initial improvement can be expected within a few days. In the few cases that do not respond to conservative therapy, the reconstructive surgical options described in the following sections can be applied.

6.2 Cysts of the Morgagni recess/laryngoceles

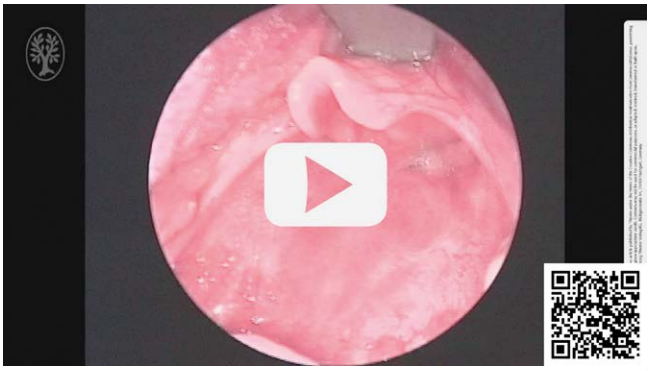
An outpouching of the Morgagni recess between the vocal fold and the false vocal fold can assume considerable proportions and lead to obstruction of the airway. Such an outpouching is encapsulated in the sense of a cyst in infancy. Therapy in most cases involves marsupialization of the cyst. This is usually adequately drained thereafter.

6.3 Subglottic cysts

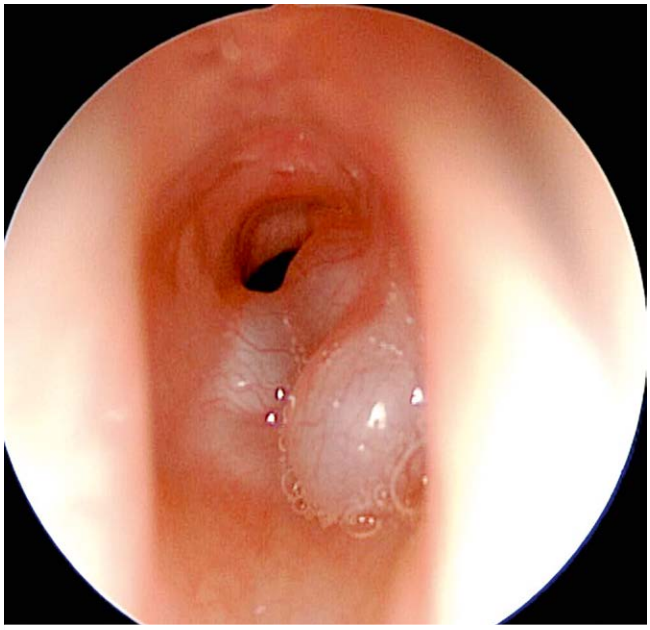
Subglottic cysts are presumably degenerative, distended mucosal glands that may typically occur in the laryngotracheal junction in premature infants (▶ **Fig. 2**). These are singular but can also be found in several locations and can cause significant respiratory obstruction if they increase in size. Therapy includes marsupialization and, if necessary, balloon dilatation of the lumen in the affected area. Because these are usually very young infants, collaboration



▶ **Video. 8** Subglottic stenosis in the context of hemangioma. After induction of drug therapy, improvement may be expected within few days.



▶ **Video. 9** Glottic papillomatosis in an infant.



▶ **Fig. 2** Subglottic cysts in a former premature infant.

between anesthesiologists and otolaryngologists during the procedure can be very challenging.

6.4 Papillomatosis

Juvenile papillomatosis of the upper respiratory tract is a relatively rare disease, but should always be considered in the differential diagnosis of stridor in infancy (▶ **Video. 9**). Due to the complexity of the clinical picture, it cannot be presented in depth within the scope of this paper.

7. Surgical procedures

7.1 Endoscopic therapy options in airway stenoses

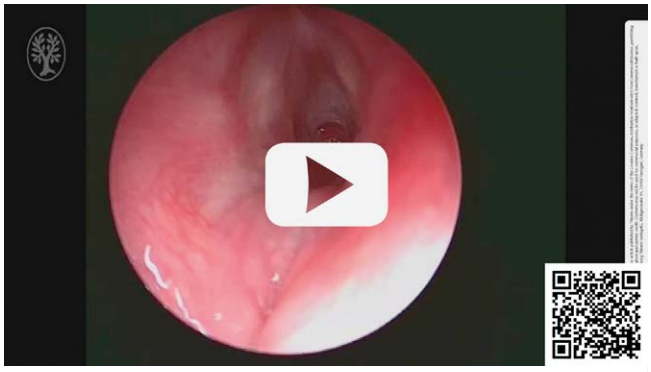
Endoscopic therapy procedures have the great advantage that they can be directly followed by the diagnostic steps. Another advantage is the lower morbidity compared to open surgical procedures.

On the other hand, they are particularly challenging for the entire interdisciplinary team, especially with regard to airway management. In the case of very young children or high-risk interventions, very closely coordinated cooperation between anesthesiologists and surgeons is therefore indispensable. This includes precise planning, including the postoperative phase, of the treatment and video-based presentation to the entire team in the operating room.

7.2 High-pressure balloon dilation

In this procedure, high-pressure balloons are applied and inflated at the level of the stenosis during direct (micro)laryngoscopy (▶ **Video. 11**). The products available on the market serve a very narrow therapeutic niche and are correspondingly expensive. The characteristics of the various balloons are defined by their approvals and are offered in different sizes and pressures.

▶ **Video. 10** Dorsal laryngeal cleft reaching into the cervical trachea (type III).



▶ **Video. 11** Subglottic stenosis as a sequela of intubation ventilation in an infant prior to balloon dilation.

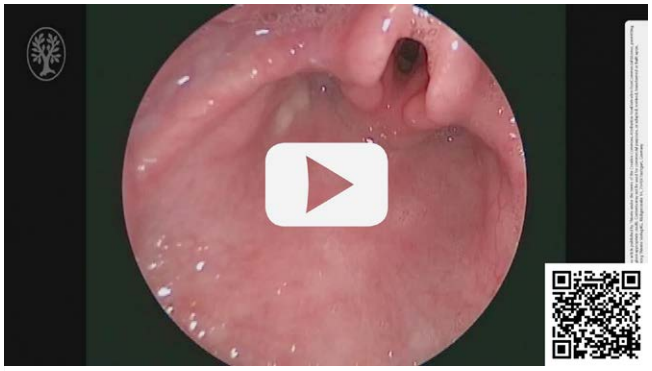
The procedure is technically simple and low-risk. Compared to classical bougienage techniques, the success rate is much higher because balloon dilatation involves an eccentric, radial application of force, which avoids tangential shear forces. Therefore, balloon dilatation is considered a low-threshold initial procedure with a wide range of indications [32]. As a basic rule, the fresher the stenosis, the better the results (▶ **Video. 12**). Prior to dilatation, targeted incision of circular scars to improve scar destruction may be helpful.

In the case of completely scarred stenoses, dilatation alone is unsuccessful. However, balloon dilatation has also established itself as an adjuvant measure after open reconstructions, because it is excellent for squeezing out postoperative edema and for stopping incipient restenosis at an early stage.

Lateral displacement of a vocal fold/laterofixation

Bilateral vocal fold obstruction may be congenital or acquired.

Congenital immobility is highly variable in clinical expression, depending on the position of the immobile vocal folds. Treatment is required when the general development of the child is impaired. In principle, the onset of mobility of the vocal folds can be expected in the course of the first years of life. Since the individual spontaneous improvement tendency cannot be predicted, the aggressiveness of therapy is based on the child's distress.



▶ **Video. 12** Minimal residual subglottic stenosis after balloon dilation performed twice in the infant of ▶ **Video. 11.**

Acquired glottic immobility may be the result of surgical procedures on the heart or esophagus. The cause of the immobility is trauma to the neural supply and, depending on its extent, may be irreparable. In these cases, permanent endoscopic glottic dilation may be necessary.

Passive lateralization can help gain time and avoid a tracheostomy if spontaneous improvement could be expected.

Lateralization of one vocal fold is performed under endoscopic or microscopic control by means of a thread looped around the vocal process. It is pulled through laterally and either tied to the skin via a silicone button or hidden subcutaneously. In permanent ablative glottic dilation, part of the dorsal glottis of the affected side is resected by laser surgery prior to lateralization.

7.3 Use of laser

The use of different laser techniques for the treatment of stenosis in children has been discussed intensively and controversially in the past. In the meantime, all major centers agree that the role of lasers in the pediatric airway is very limited. It is true that with careful indication and technically accurate application, low-grade subglottic stenoses can often be treated successfully. However, this is contrasted by a high recurrence rate, which should in no case lead to repeated laser surgery, as otherwise structural changes in the surrounding cartilage are induced, leading to an increase in the degree of stenosis as well as in the length of the stenosis. In the worst case, the chances of success for open reconstruction are severely compromised. Therefore, in case of restenosis after laser treatment, an open procedure should be used for recurrent treatment. In the past, a variety of laser types were propagated, but the broadest experience and best results were achieved with the CO_2 laser, which has the least thermal concomitant damage and is particularly precise to use.

The use of a CO_2 laser is typically carried out by means of coupling to the optical system of a surgical microscope. The use via an optical fiber does not provide any additional advantages and may only appear useful in individual cases. The advantages lie in the bloodless and contact-free procedure, which allows the resection of certain areas very precisely. Typical applications include arytenoidectomy or ablative procedures for glottis dilation. Supraglottoplasty can also be performed using CO_2 lasers (▶ **Video. 6**).

In infants, laser use is limited by the requirement to protect the ventilation tube. The risk of tube burn should not be underestimated. This risk can be eliminated by using jet ventilation.

Use of a “shaver”

The so-called "shaver" is an oscillating knife specially developed for airway surgery with simultaneous aspiration. It sucks in soft tissue and simultaneously cuts it off in a minimally invasive manner. The "shaver" is very useful in the removal of small changes in the mucosa, such as in papillomatosis, and allows fine work under endoscopic control.

8. Open reconstruction of airway stenoses

Until the 1970s, the cricoid cartilage of the larynx was considered a "no-man's land" that should be avoided surgically. During the great diphtheria epidemics of the late 19th century it was observed that even mild affections of the laryngotracheal junction could cause high-grade airway stenosis, which could almost never be successfully reconstructed with the means of the time. Furthermore, the cricoid cartilage was apostrophized as the foundation of the larynx, which to weaken was associated with a high risk of permanent laryngeal stability. In addition, the risk of damaging the recurrent laryngeal nerves was considered to be almost inevitable with procedures on the cricoid cartilage plate. In children, surgery on the cricoid cartilage was not ventured until the early 1990s, the prevailing opinion being that the risk of growth disturbance was high.

Basically, two different surgical strategies can be distinguished for interventions at the laryngotracheal junction.

Ablating procedures ("resection") aim to completely remove the pathology and reconstruct the airway through an end-to-end anastomosis. They are technically demanding and have a serious potential for complications. In return, they offer significantly superior success rates over all other techniques and are usually feasible on a single-stage basis. Ideally, they allow simultaneous resection of a preexisting tracheostoma.

Augmentative procedures ("reconstruction") leave the stenosis-forming tissue in place and aim to dilate it by inserting cartilage grafts, mostly from the rib. They are less complex technically and offer a broader range of indications, but produce poorer results compared with resecting procedures. They are typically performed 2-staged, i. e., with the tracheostoma in place, which increases the patient burden but reduces the risk to the airway. By leaving the tracheostoma in place, this procedure allows a newly created lumen of the larynx, e. g., in case of atresia, to be splinted by a stent for a longer period of time.

In individual cases, a combination of the above strategies in the sense of an extended reconstruction of the airway may also be necessary.

The introduction of high-pressure balloon dilatation in recent years has further improved postoperative follow-up after airway reconstructive surgery and thus the chances of success, because it allows gentle treatment of incipient restenosis or postoperative scarring even in a fresh anastomotic scar.

8.1 Cricotracheal resection (CTR)

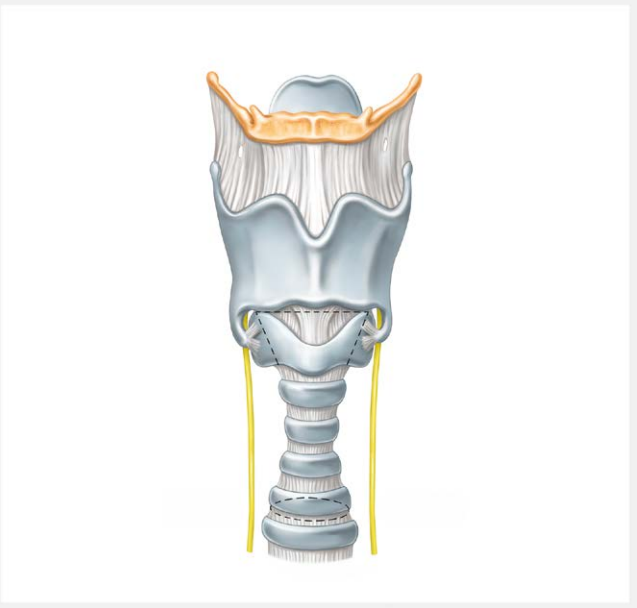
For laryngotracheal stenoses with involvement of the cricoid cartilage but without affection of the glottic level, cricotracheal resec-

tion is the procedure of choice. This is true regardless of the underlying etiology and extent of pathology. Cricotracheal resection (CTR) was first described for adult patients almost simultaneously and independently by Pearson [21] and Grillo [11], but its translation to the pediatric population did not occur until the late 1980s by Monnier [18].

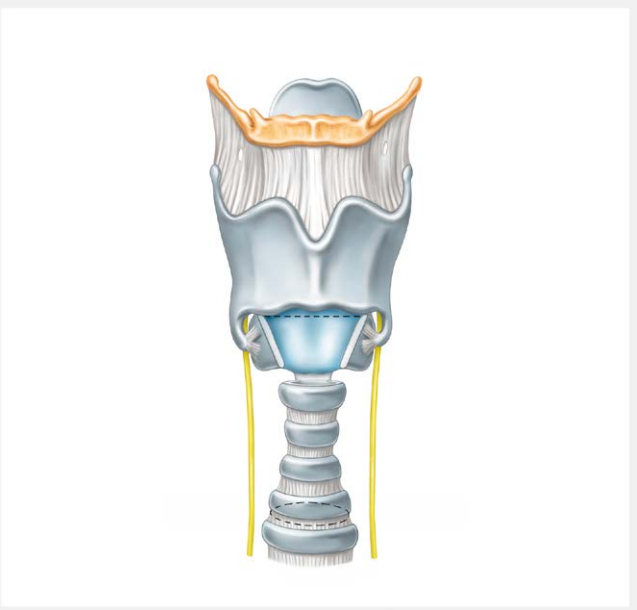
The basic principle is complete resection of the laryngotracheal stenosis (▶ **Fig. 3**) including the cricoid arch with complete exposure (▶ **Fig. 4**) of the cricoid plate [31]. After appropriate mobilization maneuvers, the distal trachea is partially adapted to the remaining cricoid, predominantly to the thyroid (▶ **Fig. 5**) (thyrotracheal anastomosis). A preexisting tracheostoma is usually included in the resection unless 2–3 healthy tracheal cartilage rings remain between the caudal stenotic margin and the cranial tracheostomal margin, which are available for anastomosis. As a rule, the procedure is performed in a single stage, i. e. without a new tracheostomy.

Whenever possible, ventilation is initially performed via a laryngeal mask. The cricoid, thyroid and trachea are carefully exposed via a cervical approach. Depending on the expected length of the resection, a supralaryngeal release is performed by incising off the infrahyoid muscles. Dissection of the lateral trachea is performed strictly along the cartilage, reliably avoiding damage to the recurrent nerves. The use of surgical loupes is highly recommended, especially for this substep. Below the stenosis, the trachea is incised and the distal tracheal stump is intubated. The cranial incision is made in the membrana cricothyroidea, the cricoid cartilage arch is transected obliquely on both sides (▶ **Fig. 6**), and the mucosa on the cricoid cartilage plate is incised depending on the level of the stenosis. The cricoid cartilage plate is exposed to its caudal edge, and the stenotic segment is separated from the esophagus and resected in toto (▶ **Fig. 7**). Complete resection of the entire pathologic portion is of high importance, as is generous mediastinal mobilization of the distal trachea. The diamond drill is used to thin out the cricoid cartilage plate to remove any residual pathology as well as to gain additional space (▶ **Fig. 8**). The distal trachea is now placed tension-free on the cricoid cartilage plate and anastomosed with absorbable suture material to the lateral aspects of the cricoid, the mucosa of the interarytenoid region, and the thyroid (▶ **Fig. 9**). This creates a primary epithelialized anastomosis while avoiding exposed cartilaginous surfaces (▶ **Fig. 9**). Prior to final knotting of the anastomotic anterior wall, the ventilation tube is removed from the distal trachea and the laryngeal mask still in place is reused for ventilation. The main advantage here is the immediate check for any air leaks, which must be meticulously closed. With few exceptions, spontaneous breathing is recovered on the table, postoperative intubation is of no benefit. Chin-chest sutures make physiologic breathing difficult and do not provide additional safety for anastomotic integrity. They should therefore be considered obsolete.

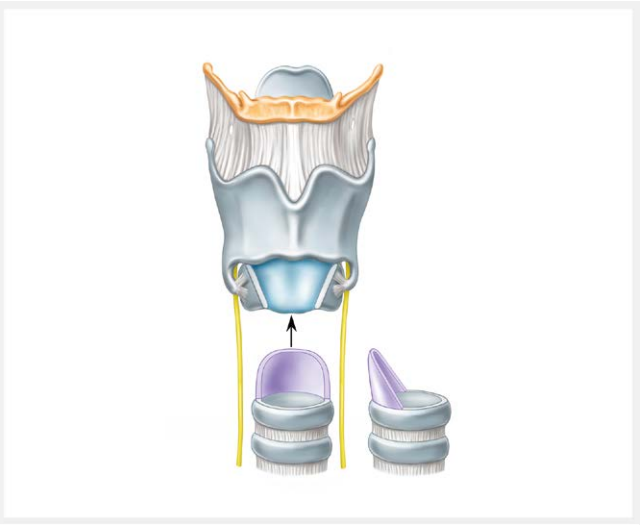
The success rate of a technically accurate and correctly indicated CTR is well over 90 %, which has been reproduced by various working groups [12, 26, 33]. Nevertheless, this is a complex, three-dimensional procedure of considerable difficulty that requires a learning curve that should not be underestimated. Despite the excellent final results, complications are not uncommon. Minor endoscopic corrections, such as removal of fibrin layers or granulations, are required in almost 2/3 of all patients, mostly only once. Severe complications such as anastomotic insufficiencies or cartilage necrosis are very rare exceptions [22]. Despite the anatomical proximity, injuries of the recurrent nerves are rare events due to accurate and precise surgical technique.



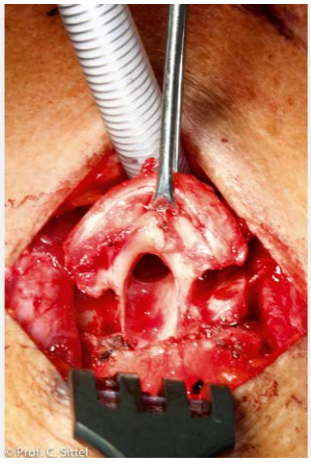
▶ **Fig. 3** Principle of cricotracheal resection (CTR). The stenosis of the laryngo-tracheal junction is exposed and mobilized under preservation of the nerves. Source: Created by S. Burger on behalf of the Klinikum Stuttgart



▶ **Fig. 4** Principle of cricotracheal resection (CTR). The stenosis of the laryngo-tracheal junction is resected completely under preservation of the dorsal plate of the cricoid cartilage. Source: Created by S. Burger on behalf of the Klinikum Stuttgart



▶ **Fig. 5** Principle of cricotracheal resection (CTR). The trachea is mobilized and anastomosed into the preserved laryngeal structure. Source: Created by S. Burger on behalf of the Klinikum Stuttgart

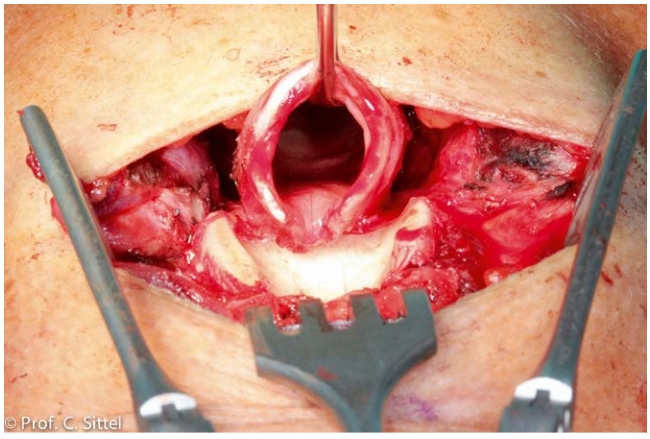


▶ **Fig. 6** Intraoperative steps of CTR. Exposition of the cricoid cartilage.

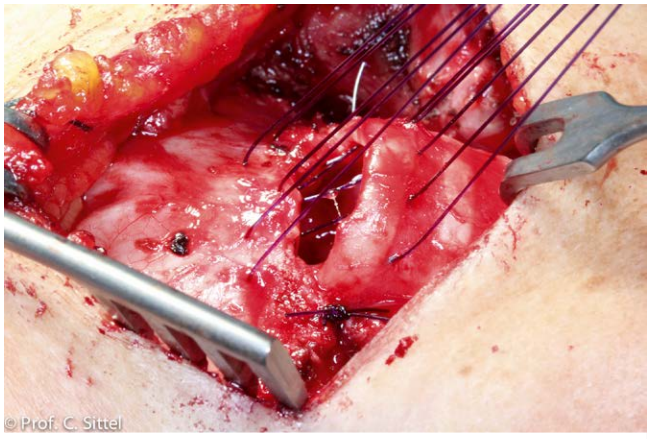
8.2 Tracheal segment resection

Tracheal segment resection is the procedure of choice for stenoses of the cervical and mediastinal trachea without cricoid cartilage involvement, which, however, occur much less frequently compared to subglottic stenoses. If the cricoid is involved, segmental resection is a contraindication! The unrecognized and/or disrespected extension of the pathology into the level of the cricoid regularly leads to restenosis, if nevertheless a pure segmental resection is performed instead of the actually indicated CTR. In the narrower sense, this is not an operation at the laryngotracheal junction, so this surgical technique will be described only briefly.

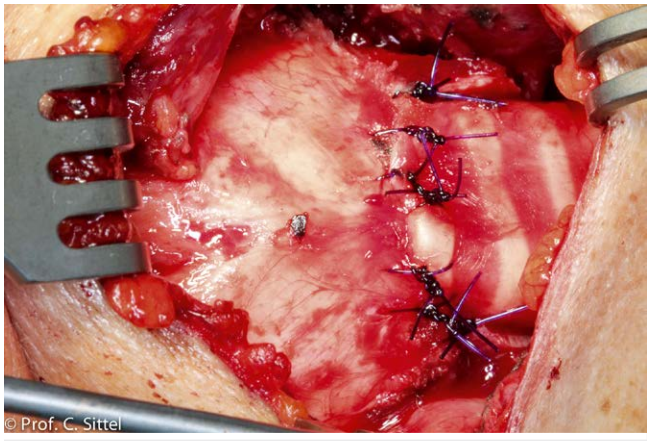
Since isolated tracheal stenoses without involvement of the cricoid cartilage are comparatively rare, but tracheal segment resection as "transverse resection" is often used as a synonym for tracheal surgery, it can be assumed that the indication is often made too uncritically. This is probably the reason why tracheal segment resection has an unjustified reputation as a procedure with little chance of success. With correct indication and precise technical implementation, however, the success rates are excellent.



▶ **Fig. 7** Intraoperative steps of CTR. Exposition of the cricoid cartilage plate after resection of the stenosed area.



▶ **Fig. 8** Intraoperative steps of CTR. Beginning of anastomosis between trachea and larynx, sutures in place, not yet tied



▶ **Fig. 9** Intraoperative steps of CTR. End of anastomosis between trachea and larynx.

The access route is almost exclusively transcervical, even for stenoses quite close above the bifurcation. Optimal exposure of the stenotic area with circumferential preparation of the entire tracheal circumference is essential. Separation between the esophagus and the pars membranacea is obligatory. Strict attention must be paid to adequate mobilization of the distal and proximal trachea to ensure a tension-free anastomosis, depending on the length of the resection. After complete resection of all pathological parts, the anastomosis is performed with absorbable suture material (PDS 2.0 or 3.0). Postoperative intubation should only be performed in absolutely exceptional cases; chin-chest sutures are obsolete.

8.3 Laryngotracheal reconstruction (LTR)

More than 100 years ago, in the case of diphtheria-related subglottic stenosis, which was endemic at the time, an attempt was made to achieve dilatation of the stenosed airway by permanent insertion of a placeholder. The name of Rethy is associated with the technique of additional splitting of the cricoid cartilage, by which the splinting dilatation was to be facilitated. In the 1970s, Cotton first described a technique in which the split of the cricoid cartilage was fixed anteriorly and posteriorly with an autologous graft of rib cartilage [15]. Under the term laryngotracheal reconstruction, this procedure represented the gold standard in the treatment of subglottic and glottic stenosis for the following 3 decades. The introduction of cricotracheal resection also in children has relativized the importance of the procedure. However, it remains the procedure of choice for all cases involving the vocal fold level. Even in cases of higher grade interarytenoid fibrosis that cannot be treated promisingly by endoscopy, laryngotracheal reconstruction (LTR) remains the best form of therapy.

Compared with cricotracheal resection, LTR is a technically less complex procedure that can be learned much more quickly [30]. The possible complications, although not less frequent, are less dramatic. These are probably also the most important reasons why, especially in childhood, CTR, which is fundamentally more promising, has only been able to prevail over laryngotracheal reconstruction in terms of numbers at particularly specialized centers. Another reason is the increasing number of complex stenoses involving the glottic level.

Autologous transplantation of rib cartilage is reliably possible in childhood; with increasing age, wound healing disorders with resorptions or rejections occur increasingly. Nevertheless, LTR can be a quite reasonable treatment option until the 4th decade of life, when resecting procedures are not possible. However, the indication must be made very carefully.

The harvesting of rib cartilage is usually technically feasible after the age of 2. In the first 18 months of life, a special form of LTR with the use of autologous thyroid cartilage can be performed [25] (see section LTR-S).

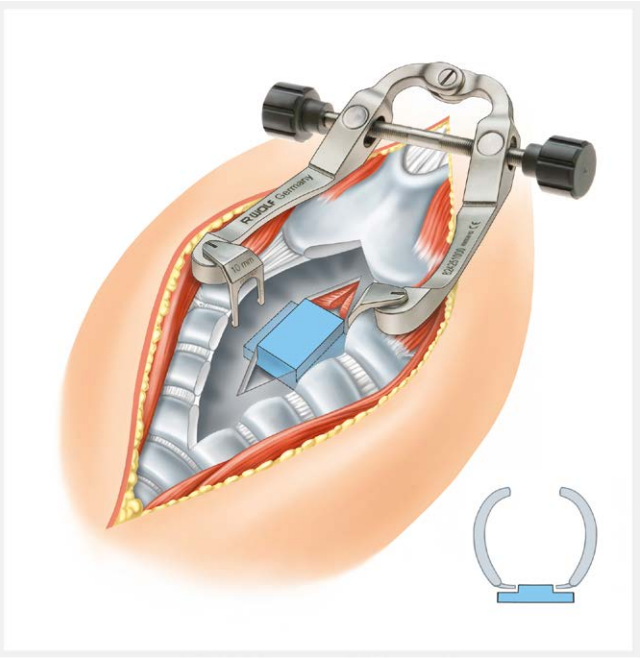
There are different modifications for the LTR depending on the author, so only the basic features will be described here.

The thyroid, cricoid, and cranial trachea are visualized via an external approach at the level of the cricoid cartilage. Unlike CTR, circumferential preparation is not required, so there is no significant risk to the recurrent nerves. After meticulous exposure of the laryngeal structures, the medial incision of the cricothyroid membrane, the cricoid, and the upper 1–3 tracheal cartilage rings is made. Depending on the exact location of the stenosis, a partial or even total laryngofissure must also be performed. For atraumatic exposure, the use of a special laryngofissure spreader is recommended (▶ **Fig. 10**). After clear exposure of the cricoid cartilage plate, the incision is made along the entire length until there is a clear gap between the two halves of the cricoid cartilage. If necessary, any additional interarytenoid fibrosis present must be separated with microscissors, preserving a cranial mucosal boundary. The posterior graft is prepared in a winner's rostrum-shaped fashion with the "1st place" facing lumenally, and should remain covered with rib perichondrium to reduce the tendency for granulation. To accommodate the graft, the posterior perichondrium of the cricoid cartilage plate is undermined. The graft is then successfully inserted with significant preload; in the vast majority of cases, additional fixation by suturing is not necessary or useful. The inherent tension of the cricoid cartilage results in a primary stable fit of the rib cartilage (▶ **Fig. 11**). In an analogous manner, a second graft is inserted into the split cricoid, extending into the cranial trachea or the caudal thyroid, depending on the individual situation (▶ **Fig. 12**).

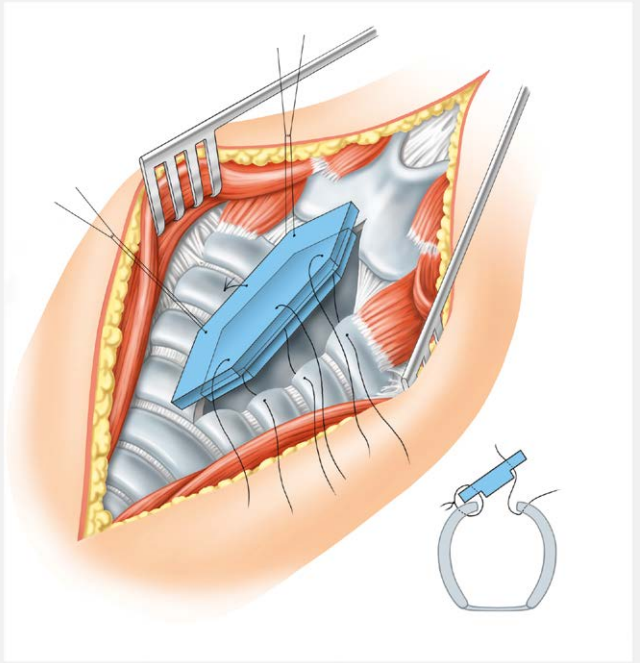
LTR is a modular procedure that can also be used anteriorly only or posteriorly only. Anterior LTR alone does not require stenting and can usually be performed without a tracheostomy. Posterior LTR and combined anterior and posterior LTR, on the other hand, should usually be performed in two stages, i. e., with temporary tracheostomy or preservation of an existing tracheostoma.

Postoperative stenting to splint the reconstructive result is used with increasing restraint. The Montgomery T-tube not infrequently leads to considerable problems due to granulation formation and pressure lesions. The "LT mold" developed by Monnier represents a considerable improvement in all respects, but unfortunately is still not commercially available. Alternatively, the Rutter stent is currently the best available solution and is used by us as standard. This stent can be individually shortened, but it is not adapted to

▶ **Fig. 10** Surgical retractor developed specifically for the exposition of laryngeal fissure. The branches are available in different lengths and may be changed according the anatomical depth. Source: Richard Wolf GmbH



▶ **Fig. 11** Principle of dorsal laryngo-tracheal reconstruction (LTR). A cartilage graft of the rib is inserted in the split cricoid cartilage plate. The exposition is most successful by means of the specifically developed laryngeal fissure retractor. Source: Created by S. Burger on behalf of the Klinikum Stuttgart



▶ **Fig. 12** Principle of anterior laryngo-tracheal reconstruction (LTR). A cartilage graft of the rib is inserted frontally in the split cricoid cartilage. Source: Created by S. Burger on behalf of the Klinikum Stuttgart

the laryngeal anatomy. Overall, however, the issue of postoperative stenting, especially in children, is still not satisfactorily resolved in many aspects.

Granulation and edema formations occur relatively regularly on the lumen-facing graft surfaces. A control endoscopy with the option of granulation removal should be performed regularly after about 3 weeks. Persistent granulations can often be improved very well by topical application of mitomycin-C (dosage 2 mg/ml). Finally, when successful airway reconstruction is confirmed during a control endoscopy, the stepwise decannulation procedure can be performed. Usually, this is done by first gradually reducing the cannula size (downsizing) followed by unplugging the cannula. The final step is taping of the tracheostoma prior to surgical tracheostoma closure. In adulthood, this process can usually be significantly shortened.

9. Special surgical procedures in pediatric patients

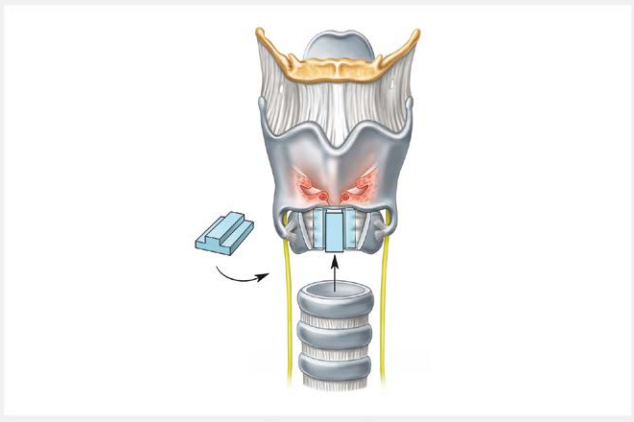
9.1 Extended CTR

In cases of pathological changes in several segments involving the glottis (multilevel stenoses), a combination of CTR and LTR may be indicated. In this case, the subglottic portion is resected to just below the level of the subglottic slope of both vocal folds. The glottic stenosis is widened by splitting the cricoid cartilage plate with insertion of a rib cartilage graft in the sense of a posterior LTR (▶ **Fig. 13**). The thyrotracheal anastomosis allows extensive mucosal coverage of the cartilage graft, which prevents granulation formation. Due to the extensive destabilization of the laryngeal skeleton, postoperative stenting becomes necessary and thus also the preservation or reinsertion of a tracheostoma below the anastomosis, which is closed bilaterally after normalization of respiratory function (▶ **Video. 13**). shows multilevel stenosis in a child, (▶ **Fig. 13**) demonstrates the principle of reconstruction, ▶ **Video. 14** shows the healed situation after extended CTR.

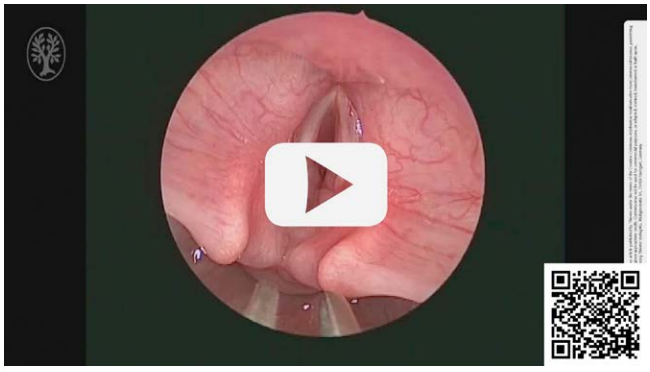
9.2 Laryngotracheal reconstruction with autologous thyroid cartilage

Today, this procedure can be considered the gold standard in the treatment of connatal cricoid cartilage stenosis. However, the consistently positive experience has also led to its use in other indications. In submucosal extirpation of subglottic connatal cysts, LTR-S is a useful prophylactic measure to avoid scarring stenosis. However, stenosis because of intubation-associated scars can also be treated in infancy with surprisingly good results.

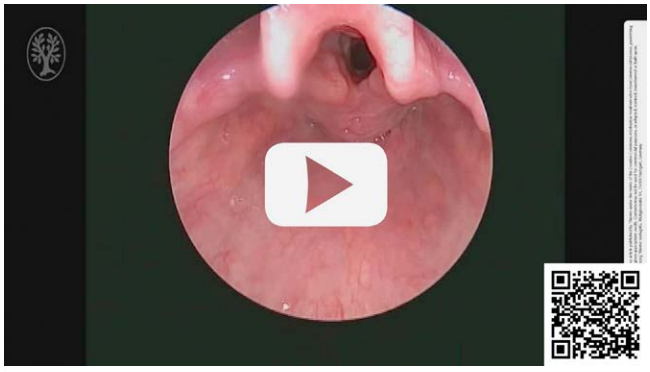
In this procedure, an approximately 2x3 mm piece of cartilage is harvested from the free upper margin of the thyroid cartilage via an external approach (▶ **Figs. 14–16**). This can be done elegantly in the same surgical area without an additional incision. The graft is inserted into the split cricoid cartilage and fixed with sutures. Postoperatively, the child remains intubated for 2–4 days. This procedure is successful in over 95 % of cases and is associated with minimal additional morbidity [25]. Of particular importance, however, is that all reconstruction options remain undiminished, both classic laryngotracheal reconstruction with autologous rib cartilage and cricotracheal resection. Even if this approach does not suc-



▶ **Fig. 13** Principle of extended CTR. The procedure uses the advantages of the combination of CTR and LTR for the treatment of multilevel stenoses with glottic involvement. Source: Created by S. Burger on behalf of the Klinikum Stuttgart



▶ **Video. 13** Preoperative condition of a toddler with multi-level stenosis with glottic fixation. Extended CTR was indicated.



▶ **Video. 14** Postoperative condition after extended CTR in a toddler.

ceed in reconstructing a permanently adequate airway, the procedure can still be very valuable in avoiding a tracheostoma in the first months of life and in bringing the child with adequate translaryngeal breathing to an age when open reconstructions can be used with very good prospects of success and an acceptable risk profile.

The procedure is only slightly invasive and significantly less stressful than, for example, a complete cricotracheal resection, a significant advantage in the often multimorbid, syndromic, or for other reasons such as prematurity unstable small patients.

LTR-S is therefore the procedure of choice for the treatment of subglottic airway stenosis up to 18 months of age, almost regardless of the underlying etiology. In an astonishingly large number of cases, it is even sufficient as a sole measure to reconstruct a permanently adequate and co-extensive airway.

9.3 Reconstruction of a dorsal laryngeal cleft

Surgical closure of a dorsal laryngeal cleft is a surgical challenge. The risks for the success of the procedure are not so much to be seen in the technical execution, but much more in the biological peculiarities of the anatomical region. The mucosal gap to be closed is located between the respiratory and the alimentary tracts and is accordingly stressed with each swallow or breath. As a result, healing is subject to great risk. In order to reduce the risk of regurgitation of gastric juice, it is regularly recommended to perform a fundoplicatio or to insert a feeding tube prior to closing the gap.

Another challenge is the age of the patients. In the vast majority of cases, treatment takes place in the first year of life. The majority of patients have additional malformations that increase the complexity of the therapeutic strategy.

The surgical strategy depends on the extent of the finding. Type I and type II laryngeal clefts are usually amenable to endoscopic exploration, whereas most type III and all type IV clefts require external access and the creation of a tracheostoma. Type IVb clefts with extension caudal to the upper thoracic aperture or laryngotracheal clefts require intraoperative extracorporeal membrane oxygenation (ECMO), otherwise exposure of the surgical site is not possible.

9.4 Endoscopic closure of dorsal laryngeal clefts

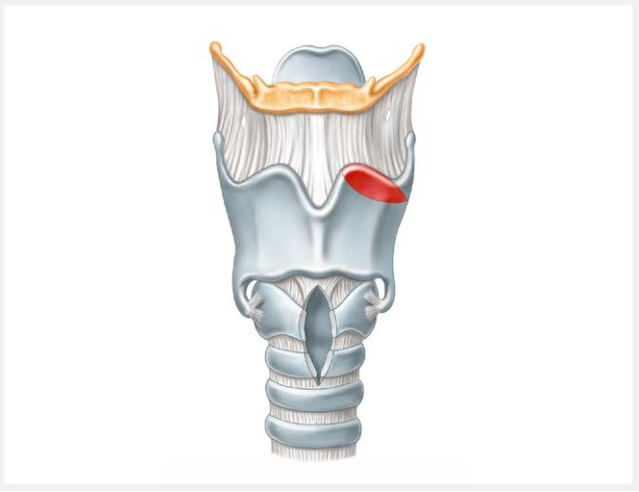
Regardless of the access route, the principle of gap closure is the same: the mucosa of the open gap between the larynx and esophagus/hypopharynx must be slit and then closed in two layers, laryngeally and pharyngeally, using absorbable single-knot sutures (▶ **Fig. 17**).

Endoscopic procedures are technically demanding and require very close cooperation between the surgeon and the anesthesia team. In most cases, the patients are very young and do not have a tracheostoma. Therefore, surgery is only possible with the use of supraglottic jet ventilation. Alternatively, a temporary tracheostomy may be unavoidable.

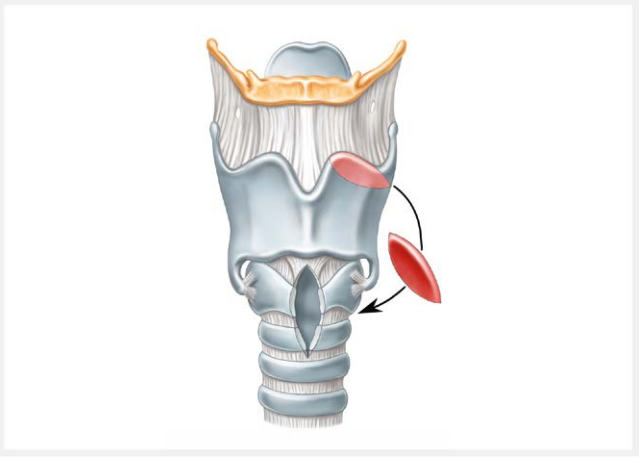
Adjustment of the dorsal larynx is best accomplished using a Lindholm laryngoscope. There is one manufacturer of jet ventilators on the market that allows controlled ventilation of infants from 1,000 g body weight using a technology specially developed for this age group.

Free and complete exposure of the gap is an essential prerequisite for optimal mucosal suturing. This is best achieved in our hands by using a vocal fold spreader (▶ **Fig. 18**), which is locked inside the Lindholm tube with the handle facing upwards.

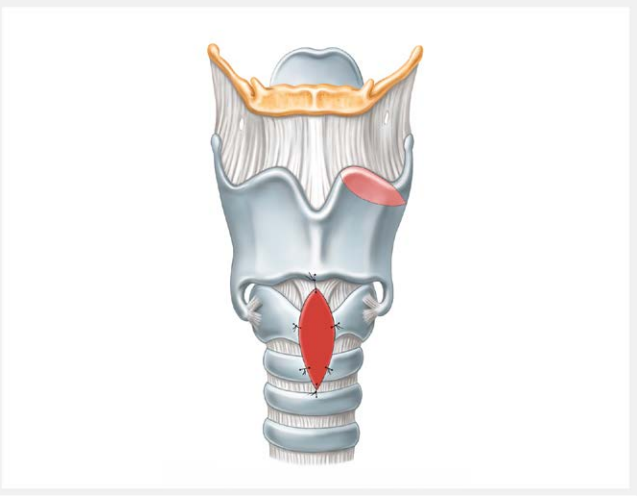
The mucosal slitting is performed under microscopic control using a CO_2 laser, whereby care must be taken to minimize carbo-



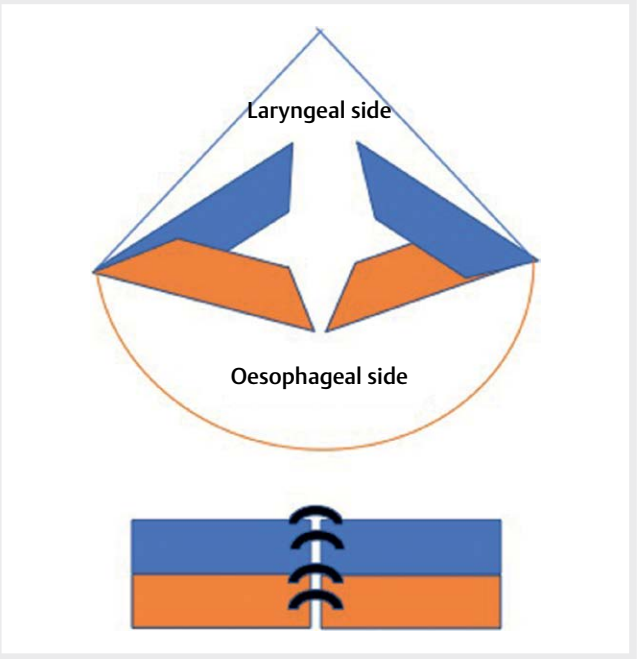
▶ **Fig. 14** Principle of autologous LTR with thyroid cartilage – first step. Source: Created by S. Burger on behalf of the Klinikum Stuttgart



▶ **Fig. 15** Principle of autologous LTR with thyroid cartilage – second step. Source: Created by S. Burger on behalf of the Klinikum Stuttgart



▶ **Fig. 16** Principle of autologous LTR with thyroid cartilage – last step. This intervention is very helpful in infants with cricoid cartilage stenosis and it is associated with low morbidity. Source: Created by S. Burger on behalf of the Klinikum Stuttgart



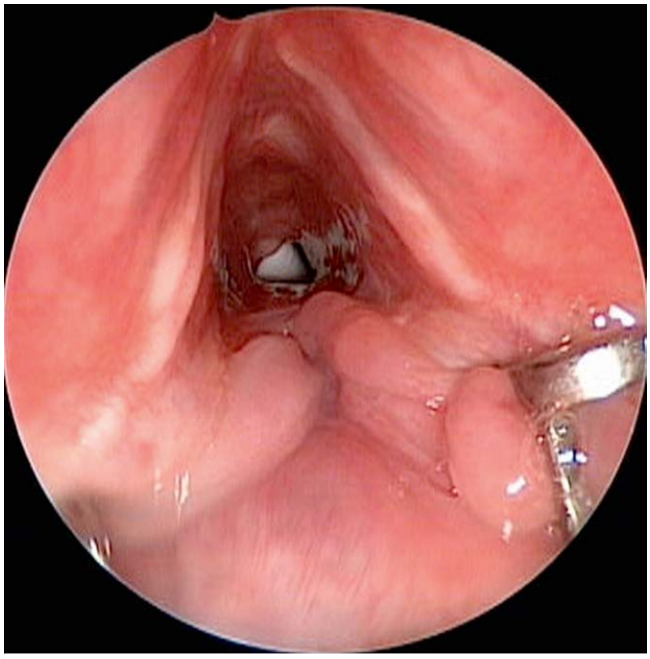
▶ **Fig. 17** Principle of two-layer closure of a laryngeal cleft. The separation of the laryngeal mucosa from the esophageal mucosal layer prior to suture of the cleft edges is central for a successful intervention.

nization of the cut edges. The free mucosal edges are to be handled atraumatically so as not to additionally jeopardize the healing of the anastomosis. The absorbable sutures are advanced and then immersed one by one, using a knot pusher. The technique requires patience and consideration for the respiratory needs of the child, which must be closely coordinated with the anesthesia team.

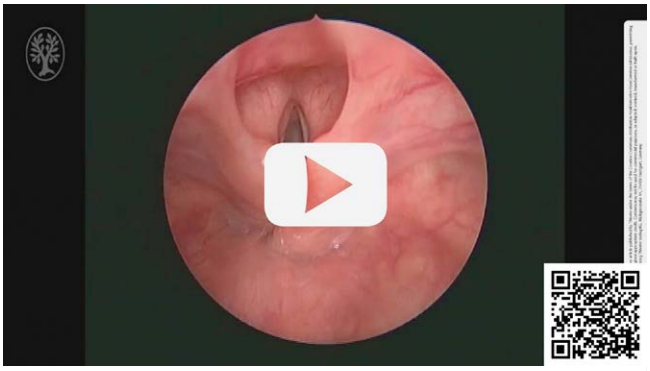
If possible, insertion of a naso-gastric feeding tube should be avoided, as it poses additional risks to the sutures due to mechanical stress and as a guide for gastric juice.

9.5 Open closure of dorsal laryngeal clefts

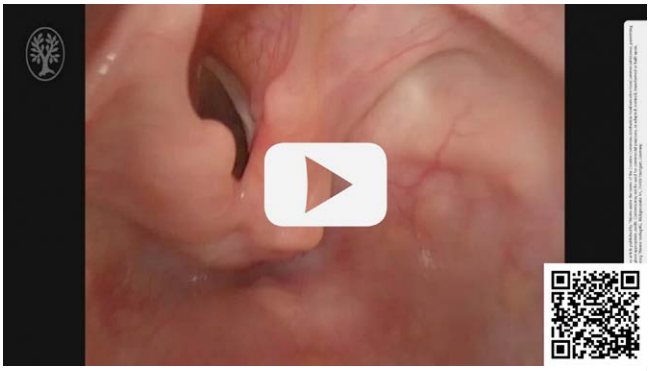
The vast majority of type III and all type IV laryngeal clefts require an open approach. For this purpose, the trachea is opened up to the caudal fundus of the fissure, as well as the cricothyroid cartilage, the cricothyroid membrane and the thyroid, in each case strictly in the midline. This complete laryngofissure must be performed exactly in the center in the area of the anterior commissure under visualization with surgical loupes, then later synechia of the vocal folds is generally not to be feared. After insertion of the laryngofissure retractor, the overview of the dorsal fissure formation is excellent. After decongestion with topical suprarenin, the lateral portions are separated into an anterior and a posterior flap. Particular attention must be paid to complete de-epithelialization of the cleft, especially in the fundus of the cleft, as this is the predilection site



▶ **Fig. 18** Exposition of a dorsal laryngeal cleft of type III using a vocal fold spreader. This instrument is also used for checking the passive mobility of the glottis.



▶ **Video. 15** Preoperative endoscopy of a laryngeal cleft type III.



▶ **Video. 16** Postoperative endoscopic control after closure of a laryngeal cleft type III via laryngofissure.

for recurrence. Then the two posterior mucosal flaps are adapted first, followed by the two anterior mucosal flaps with single button sutures. If sufficient mucosa is available and especially in revisions, a graft can be interposed, such as sternal or tibial periosteum, more rarely fascia lata, temporalis fascia or perichiondrium from the ear cartilage.

Closure of the laryngofissure is level-adjusted and almost never leads to wound healing problems. Postoperative stenting is not necessary, but the creation of a tracheostoma is. A gastric tube should be avoided in order to prevent additional foreign body pressure on the reconstruction. The child should already have been treated with a PEG or PEJ beforehand. Closure of the tracheostoma is possible only after normalization of breathing and swallowing. Children tube-fed immediately after birth often need many months to relearn the physiological swallowing process, even if the anatomical situation could be reconstructed ad integrum. ▶ **Video. 15** shows the endoscopic diagnosis of a type III dorsal laryngeal fissure prior to reconstruction via laryngofissure, ▶ **Video. 16** the same case during control endoscopy after wound healing has been completed. In both cases, instrumental palpation is important to accurately determine the extent or to detect remaining fistulas or insufficiencies.

9.6 Two-stage closure of laryngotracheal type IV clefts

The primary focus of care for neonates with type IV laryngotracheal clefts, which typically extend just anterior to the tracheal carina, is respiratory stabilization, achieving a safe airway as soon as possible, and preventing aspiration. These are often premature infants with correspondingly low birth weight, low tolerance to invasive procedures, and frequent multimorbidity due to malformation of other organ systems. Therapeutic decisions should be made in severe cases only after multidisciplinary discussion including a palliative care perspective.

A first step is the creation of a gastrostoma with a jejunal part both to drain gastric secretions and prevent gastroesophageal reflux and to provide jejunal nutrition. Surgical correction of the intrathoracic tracheal cleft portion can be accomplished with adequate safety only with the use of ECMO or cardiopulmonary machine, for which a body weight of approximately 2,000 g is usually required. The creation of a tracheostoma only makes sense if the tracheal cleft is closed at least up to the level of the stoma to allow a safe change of cannulae. The creation of a tracheostoma is therefore combined with the closure of the tracheal portion of the cleft. Via a right-lateral thoracic approach, the two lateral portions of the fissure are adapted using a technique analogous to the one described in the previous section. Usually, the present tissue material proves to be redundant with, as a consequence, often a persistent tracheomalacia symptomatology. Another problem is the possible formation of steps at the transition of the reconstructed pars membranacea to the intact one at the fundus of the former fissure. In case of an unfavorable choice of the tracheal cannula length, the cannula tip may come to rest in this area, which may lead to relevant obstructions and mucosal lesions. Endoscopic control of the surgical success as well as of the cannula position is therefore already indicated intraoperatively.

Surgical closure of the laryngeal and any residual tracheal cleft can then usually be performed after 6–12 weeks, depending on the stability and thriving of the child. The surgical procedure corresponds to the one described in the previous chapter.

In our experience, tracheal cleft recurrences are rare and typically occur at the junction of the laryngeal and thoracic cleft [24]. In contrast, tracheomalacia is almost obligatory and may require long-term ventilation.

In the course of often several years, after stabilization of the tracheomalacia and growth of the child, weaning from home ventilation and later decannulation is possible.

A pronounced failure to thrive despite probing of high-caloric food via jejunostoma is observed in many children with type IV cleft. In this case, additional caloric food fortification is required. Swallowing and esophageal function may remain compromised for a long time. Recurrent aspiration and subsequent respiratory infections may accordingly persist despite good surgical success. Nevertheless, recurrence of cleft should be excluded.

Conflict of Interest

The authors declare that they have no conflict of interest.

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