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Review

Upper airway anomalies in children: From diagnosis to treatment

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Abstract: Endoscopy of the airways plays a crucial role to investigate respiratory symptom and signs in newborns and children. Upper airway anomalies include malformations and acquired conditions found from the nose to the larynx. Common anomalies of the upper airways include laryngomalacia, vocal cord paralysis and subglottic stenosis. In the child their clinical presentation is often characterized by dysfunction in breathing, feeding or phonation. Most severe cases of congenital anomalies can present with respiratory distress at birth. Endoscopy is important not only for detection and diagnosis but also for treatment that is tailored to each condition depending on disease and severity. This review describes the commonest congenital or acquired anomalies of the upper airways seen in children. (www.actabiomedica.it)

Key words: airway malformations, stridor, children, endoscopy, laryngomalacia, cleft, stenosis, vocal cords

Introduction

Over the last years, the role of airway endoscopy in young age has significantly expanded. Appropriate instruments like rigid and flexible bronchoscopes have been developed for the small airways of children and endoscopy has been widely used to diagnose and treat several clinical pictures. The flexible bronchoscope plays a crucial role to investigate the cause of stridor and other respiratory symptoms or signs and to perform alveolar broncholavage. On the other side, the rigid bronchoscope, equipped with an adequate operating channel, is usually used for longer or interventional procedures and to extract inhaled foreign bodies (1-3). The rigid instrument has a lateral arm used for ventilation, which makes the procedure safer especially in children, more prone to hypoxic complications (4). However, the use of rigid bronchoscope is less common among specialists and its use is usually limited to a few national centers (5).

This review aims to provide an overview of the commonest malformations and diseases affecting the upper airway including a description of clinical presentation, endoscopic diagnosis and treatment.

Definition and symptoms

The upper airway extends from the nose to the larynx. This small tract of the airways plays a crucial role in breathing, feeding and phonation. Its evaluation requires specialists experienced in endoscopy since these anomalies need to be assessed under direct visualization during spontaneous breathing.

Congenital and acquired anomalies of the upper airways commonly result in obstruction. In response to upper airway obstruction, the child develops high

negative intrathoracic pressure that worsens the dynamic collapse of the soft extrathoracic airways.

Children are more prone to airway obstruction for several reasons: larger and prominent occiput that leads to flexion of the neck when the child is in supine position, larger tongue that can obstruct the air flow, anterior position of the larynx and soft and omega-shaped epiglottis. In addition, children's airways are generally softer and can easily collapse. When upper airway obstruction occurs, the child presents increased work of breathing that in some cases can rapidly progress to respiratory failure. Typical symptoms of upper airway obstruction are:

a. *snoring:* irregular inspiratory noise usually due to vibrations of the uvula, soft palate and tongue. When the obstruction is severe, snoring can be associated with episodes of apnea. The commonest causes of snoring are pharyngeal collapse, adenotonsillar hypertrophy and congenital malformations of the palate, tongue, jaw or mandible.
b. *stridor:* breathing sound of variable intensity produced by the vibration of tissues in response to turbulent air flow through a narrow segment of the airways. Stridor is one of the commonest indications for airway endoscopy in infants and children. Usually, this symptom suggests a narrowing of the larynx or of the extrathoracic part of the trachea but sometimes is the result of a foreign body in the esophagus. The stridor is usually *inspiratory,* however, in some children with subglottic lesions (particularly if the upper trachea is involved), an *expiratory* stridor may be heard. Biphasic stridor suggests a severe and fixed airway obstruction that can be located anywhere from the glottic plane to the mid-tracheal region. In neonates, the commonest causes of *biphasic stridor* are vocal cord paralysis and subglottic stenosis (6,7). In general, stridor intensity is directly related to the degree of airway obstruction. However, a sudden decrease in intensity may indicate a decrease in airflow due to the worsening of the obstruction. Conversely, when the obstruction is relatively mild, breathing can be quiet at rest, but stridor can develop during crying or activity due to increased airflow.
c. *hoarseness:* with or without stridor, hoarseness suggests an abnormality of the vocal cords. If present at birth or soon after birth, it suggests a structural (e.g. laryngeal web) or functional (e.g. vocal cord paralysis) abnormality. Intermittent hoarseness of varying degrees is mostly due to edema, whereas progressive hoarseness may be due to vocal cord lesions such as nodules or papillomas. Hoarseness associated with acute inspiratory stridor suggests narrowing of the glottis.

Choanal atresia

Choanal atresia is the commonest congenital anomaly of the nose with an incidence of approximately 1 in 7.000 live births. In most cases choanal atresia is due to a bony occlusion or, more rarely, to a membranous obstruction. While bilateral atresia can cause severe respiratory distress soon after birth with dyspnea, cyanosis, and asphyxia (newborns are obligate nasal breathers), the more common unilateral obstructions (mainly on the right side) are often detected later in life. Some children present with feeding difficulties and persistent rhinorrhea; respiratory symptoms tend to decrease during crying. The diagnosis of this anomaly is confirmed by flexible endoscopy; computed tomography (CT) is often necessary to delineate the exact site of the obstruction and define the nature of the defect.

Choanal atresia can be associated with syndromes like Charge syndrome (coloboma of the eye, heart defect, choanal atresia, delayed growth or development, genital hypoplasia and ear abnormalities or deafness) or Treacher-Collins syndrome (8-10). Another nasal anomaly is the stenosis of the pyriform aperture. Surgery is the gold standard treatment for bilateral atresia, can be performed transnasally with the endoscope and it is followed by the insertion of temporary stents.

Nasal masses

Congenital mass-like nasal lesions are rare. These lesions range from cystic masses (dermoid cysts, or

meningoceles) to more solid lesions such as hemangiomas, neurofibromas, teratoma and gliomas. Encephalocele and gliomas are commonly associated with midline defects such as cleft palate (8,9).

Craniofacial congenital abnormalities involving upper airways

Pierre Robin sequence consists of micrognathia, glossoptosis and cleft palate, resulting in pharyngeal obstruction. The severity of the airway obstruction varies widely. As the jaw grows forward with age, especially during the first 6 months of life, airway and feeding problems gradually resolve. Prone positional therapy may help but in more severe cases other options are considered like placement of a tube intranasally positioned in the distal oropharynx, the application of non-invasive ventilation or surgery with mandibular distraction. Pierre Robin sequence may be associated with other congenital malformations (e.g. Stickler's syndrome) (11,12).

A number of other craniofacial syndromic abnormalities can affect the patency of the nose and pharynx (13). These dysmorphic syndromes can be characterized by maxillary or mandibular hypoplasia and include craniofacial dysostosis (Crouzon syndrome), mandibular-facial dysostosis (Treacher Collins syndrome) and acrocephalosyndactyly (Apert syndrome) (10). In trisomy 21, the narrow nasopharynx with associated adenotonsillar hypertrophy, macroglossia and decreased muscolar tone may result in significant airway obstruction (14). True macroglossia is the cause of upper airway obstruction in patients with Beckwith-Wiedemann syndrome (exomphalos-macroglossia-gigantism syndrome) (15).

Macroglossia

Macroglossia can manifest with stridor, dysphagia and difficulty in emitting sounds and/or words.

Macroglossia may be:

a. *true macroglossia:* due to tumors (hemangiomas, lymphangiomas, neurofibromas, etc.), vascular malformations or associated with other abnormalities like in Beckwith-Wiedemann syndrome.
b. *relative macroglossia*: no identifiable cause of macroglossia, as in trisomy 21 (14,15).

Indications for surgery are issues with feeding, persistent drooling, speech impediments, orthodontic problems and psychological difficulties resulting from abnormal cosmetic appearance. When performed, aim of surgery is to reduce the tongue bulk while preserving a normal shape and improving function. The usual approach is anterior wedge resection but a variety of other techniques have been described. Tongue reduction consists of a central V shaped incision in the mobile tongue, 10 mm forward of the circumvallate papillae line. Sizing of the resected area is planned to obtain the tongue tip behind the dental arch. Rare surgical complications include postoperative oedema and wound dehiscence (16,17).

Adeno-tonsillar hypertrophy

Adenotonsillar hypertrophy is one of the main causes of airway obstruction in childhood and can be associated with obstructive sleep apnea (OSA) syndrome that include snoring, sleep apnoeas, restless sleep and mouth breathing (18). This condition can further aggravate airway obstruction in patients with pharyngeal collapse.

Pharyngeal collapse

Pharyngeal collapse may be due to a reduction in pharyngeal neuromuscular activity and/or anatomical narrowing of the airways by pharyngeal structures. Complete airway obstruction in the oropharynx during inspiration is usually accompanied by laryngomalacia or other concomitant airway abnormalities. Generalized hypotonia is a frequently associated systemic finding in patients with neurological impairment and is characterized not only by obstructive respiratory symptoms but also by feeding difficulties with dysphagia and failure to thrive.

In patients with pharyngeal collapse, gastroesophageal reflux (GER) is promoted by the negative intrathoracic pressure that occurs in the inspiratory phase.

The degree of sedation during the endoscopic investigation is an important element to consider during the dynamic assessment of the pharyngeal tone. Some authors have proposed the definition of "incoordinate pharyngo-laryngomalacia" for cases of severe laryngomalacia with complete collapse of supraglottic structures including pharyngeal collapse (14,19,20).

Masses of the tongue base

Cysts or masses arising from the dorsal surface of the tongue are rare but can cause significant airway obstruction, usually with concomitant feeding difficulties and oral cavity bleeding. Because some cysts vary in size over time, symptoms may be intermittent and related to progressive increase of the volume. Typically, symptoms increase during upper airway infections. Differential diagnosis includes lingual thyroid tissue, mucus retention cyst and thyroid duct cyst. The latter generally tends to expand towards the anterior wall of the neck and many times is not associated with respiratory distress (21).

Laryngomalacia

Laryngomalacia is a collapse of the supraglottic structures during inspiration, resulting in intermittent obstruction of airflow and stridor. Laryngomalacia is characterized by:

- omega-shaped epiglottis (Figure 1)
- short ariepiglottic folds
- various degree of prolapse of the arytenoids and inter-arytenoid mucosa (Figure 2)
- antero-posterior displacement of the epiglottis (22).

The incidence of the disease is unknown but it has been considered as the commonest cause of persistent stridor in neonates and infants. However, it may also occur in a later-onset form in children or even in adults (23-25). The pathogenesis is unknown, but a delay in neurofunctional maturation of the larynx has been suggested (25). Usually, clinical symptoms vary depending on the onset of the disorder. When laryngomalacia starts in infancy, the main respiratory

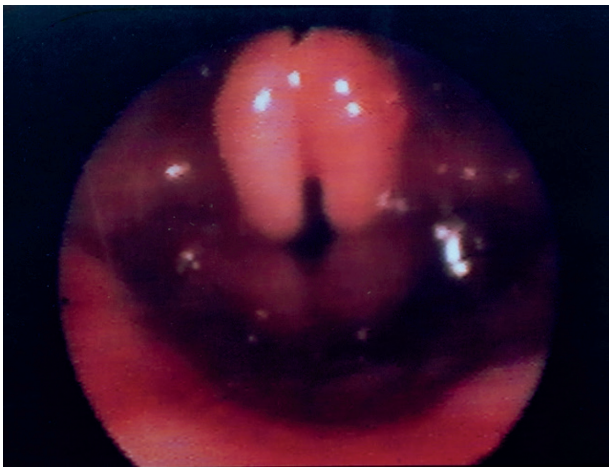

Figure 1. Larynx with omega-shaped epiglottis in 1-year-old boy. Image property of P.S.

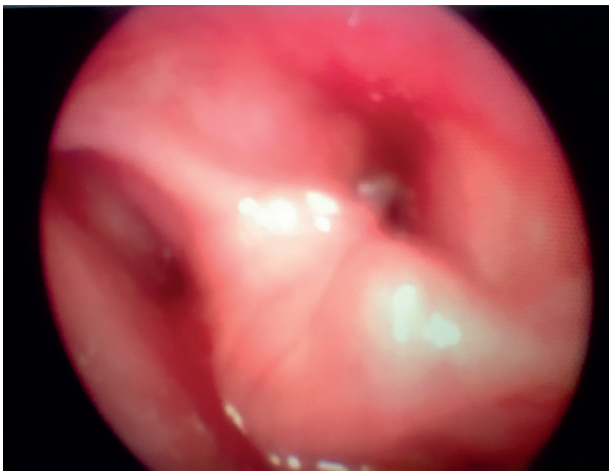

Figure 2. Laryngomalacia in 18-months-old boy. Arytenoids prolapse during inspiration, reducing the respiratory space, and cause inspiratory stridor. Image property of P.S.

symptom is inspiratory stridor. This occurs in the first 4-6 weeks of life and usually resolves within two years of age. When the onset occurs later in life, issues in swallowing, sleeping and exercise tolerance may be associated. Typically, stridor is exacerbated by crying, agitation, sucking and supine position. Severe forms may be associated with dysphagia, GER, microaspiration episodes, failure to thrive, feeding difficulties, cyanosis, intermittent respiratory obstruction and eventually heart failure if associated with OSA (14).

Diagnosis is made by flexible bronchoscopy with patient awake or mildly sedated to assess inspiratory

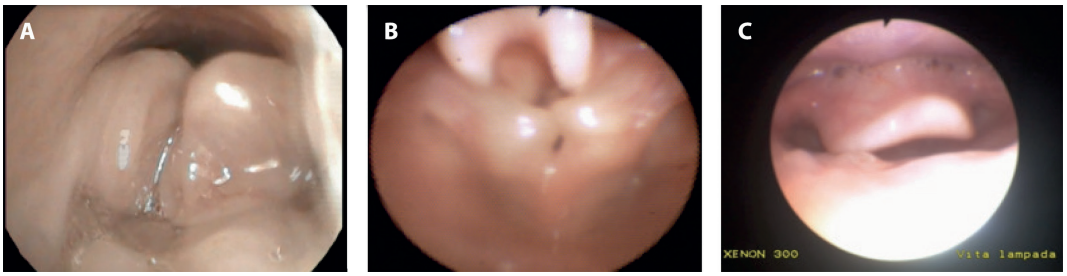

Figure 3. Classification of laryngomalacia: type 1, abundant and floppy mucosa overlying arytenoids prolapses during inspiration occupying the respiratory space (A); type 2, short aryepiglottic folds associated with long omega shaped epiglottis which curls on itself (B); type 3, retroflexed epiglottis collapsed posteriorly during inspiration (C). Image property of P.S.

collapse of the supraglottic structures. In case of bronchoscopy, assessment of the larynx should be performed before the application of lidocaine since topical anesthesia can exacerbate laryngomalacia. Different classifications have been proposed (22) according to the laryngeal component involved, but the commonest identifies three different types of laryngomalacia (Figure 3):

- *type 1*, prolapse of the mucosa overlying the arytenoids;
- *type 2*, short ariepiglottic folds;
- *type 3*, anteroposterior tilting of the epiglottis.

The laryngeal component may be also associated with the pharyngeal component, usually in the form of collapse. Depending on respiratory impact, the following conditions can be recognized:

- *mild* form, characterized by intermittent and isolated inspiratory stridor, associated with minimal respiratory work, normal development and growth of the child. It generally resolves in the first two years of life. A regular follow-up and parental reassurance is usually sufficient to guarantee a correct home management.
- *moderate* form, characterized by continuous stridor and significant increase in respiratory work and retractions. Although most symptoms resolve within the first 2 years of life, regular clinical assessments to exclude failure to thrive and polysomnography to exclude OSA must be performed over time. Radiological diagnostics with esophagogram can be considered to exclude associated vascular anomalies. Regular follow-up with serial endoscopies is needed.
- *severe* form, characterized by continuous inspiratory stridor, failure to thrive and various degrees of respiratory failure requiring oxygen support or respiratory assistance. These forms may be associated with severe GER, exacerbated by the inspiratory effort, which can further aggravate respiratory symptoms. A surgical approach performed with a variety of endoscopic techniques is generally required to reduce the redundant or obstructive supraglottic tissue. Depending on the defect, surgical treatment can range from the reduction of the arytenoid pyramids to supraglottoplasty or epiglottopexy. Surgery does not necessarily resolve the stridor completely but can significantly improve the respiratory dynamics. Temporary tracheostomy may be required in case of severe laryngomalacia that is not alleviated following supraglottoplasty (26).

Laryngeal stenosis

Laryngeal stenosis can be congenital or acquired. Stenosis is defined as a narrowing of the larynx at the level of the cricoid where the luminal diameter is less than 4 mm in a full term infant or less than 3 mm in a premature infant (27-29).

Congenital causes include subglottic stenosis, laryngeal web and atresia, vocal cord paralysis, subglottic

Table 1. European Laryngological Society (ELS) classification system for benign laryngotracheal stenosis. The ELS score consists of three components: 1, Myer–Cotton grade of airway obstruction, marked from I to IV; 2, number of affected airway subsites, marked from "a" to "d", and 3, presence of comorbidities, marked with a "+" (30).

ELS classification score				
1		**2**		**3**
Myer–Cotton grade	**Grade of airway obstruction**	**Extension**	**N° of affected airway subsites**	**Comorbidities**
I	<50%	**A**	1	Yes **(+)** or No **(-)**
II	51-70%	**B**	2	
III	71-99%	**C**	3	
IV	Complete obstruction	**D**	4	

hemangioma and laryngeal cleft with redundant mucosa (28).

In contrast, 90% of the acquired forms include subglottic stenosis occurring after intubation. Other less common causes include iatrogenic complications from endoscopic laryngeal interventions, benign tumors, caustic or thermal injuries, external blunt force injuries or trauma, chronic inflammatory disorders or idiopathic causes (27). Signs and symptoms clearly depend on the degree of stenosis. The degree of stenosis can be described by Myer Cotton classification (29):

- *grade I*, obstruction <50% of the lumen;
- *grade II*, obstruction of 51-70% of the lumen;
- *grade III*, obstruction >71% of the lumen;
- *grade IV*, no appreciable lumen, atresia.

In 2015, the European Laryngological Society (ELS) published a consensus paper proposing a scoring system, which integrates grade and extension of stenosis with the presence of concurrent comorbidities to provide a more comprehensive evaluation of the patient (Table 1) (30).

Congenital subglottic stenosis can be a cause of stridor in neonates. This type of stenosis can be membranous, characterized by soft tissues thickening in the subglottic area resulting in bilateral symmetrical narrowing of the subglottic space, or cartilagineous, due to a malformation of the cricoid cartilage, resulting in a circumferential stenosis of variable appearance, ranging from a normal to an abnormal shape. Depending on the degree of the stenosis, children can show a different clinical spectrum ranging from severe respiratory distress at birth to inspiratory or biphasic stridor within the first few months of life. Patients with milder stenosis may be asymptomatic until they experience an upper respiratory infection causing critical airway obstruction.

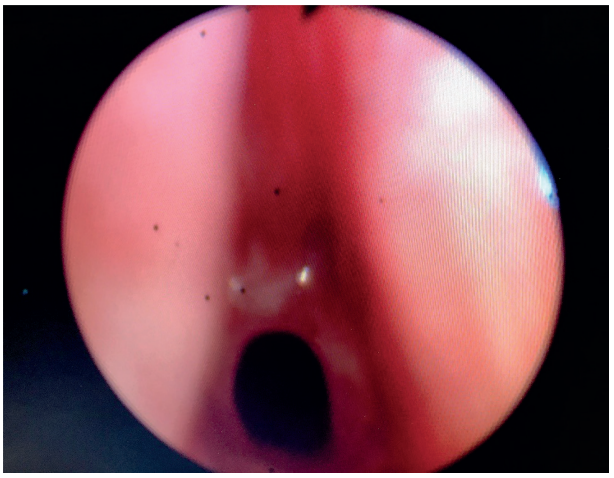

Figure 4. Subglottic stenosis after intubation in a 2-year-old child. The inflammation is demonstrated by edema and hyperemia of the mucosa. Image property of P.S.

Acquired subglottic stenosis is more common than the congenital form and is the result of a trauma on the airways such as prolonged endotracheal intubation or tracheostomy. In infants and children, the most vulnerable part of the airways is the cricoid cartilage, where damage is most likely to be found. When the injury involves the glottic plane, glottic-subglottic stenosis appears and hoarseness or aphonia may occur. Failure to extubate may be the initial clinical presentation of acquired subglottic stenosis (Figure 4) (31).

A similar condition, which must be included in the differential diagnosis, is subglottic edema. In this case, the subglottic space appears symmetrically edematous, it often occurs after rigid bronchoscopy and may be associated with supraglottic edema.

An accurate preoperative diagnostic work-up of subglottic stenosis is important in order to obtain crucial information that may have an impact on the postoperative outcome. Information that must be collected include mobility of the vocal cords, presence of glottic and/or supraglottic scar tissue, fixation of the cricoarytenoid joints, possible additional tracheal damage like stenosis or malacia related to tracheostomy, granuloma or scar tissue, obstruction related to OSA, difficulties in swallowing with or without chronic aspiration, severe GER, eosinophilic esophagitis or associated congenital abnormalities.

For patients who require intervention, endoscopic treatment with laser is considered for stenosis of grade I and II, anterior cricoid split or open laryngotracheal reconstruction are instead considered for high-grade stenosis (32,33).

Congenital laryngeal webs

Congenital laryngeal webs are rare. They usually occur at the level of the glottis (75%) but can also appear in supra and subglottic regions. They result from a failure of recanalization of the primitive larynx between the 8th and the 10th week of embryogenesis. Congenital subglottic stenosis, tracheoesophageal fistula or syndromes like 22q11 microdeletion can coexist (34).

These membranes may be complete or incomplete and vary in thickness. Different types of glottic membranes can be distinguished, depending on their extent. An acquired lesion is usually called "synechia". Symptoms are variable and range from hoarseness to aphonia, biphasic stridor to respiratory distress of varying degrees.

Cohen's classification is used to score this type of lesion (35):

- *type 1*, involves 35% or less of the glottic lumen;
- *type 2*, varies between 35 and 50%;
- *type 3*, varies between 51 and 75%;
- *type 4*, involves 76% or more of the glottic lumen.

In 1992, McCaffrey proposed a classification to describe the extent of stenosis among the most commonly involved sub-sites: glottis, subglottis and trachea (36):

- *stage I*, stenosis is located in the subglottic area or trachea, with a craniocaudal extension <1 cm;
- *stage II,* limited to the subglottic area and has a craniocaudal extension >1 cm;
- *stage III,* involvement of the subglottic area and trachea;
- *stage IV,* extension to the glottis with fixation or paralysis of at least one vocal cord.

Surgical treatment of laryngeal webs is usually postponed until the age of 3 or 4 years (27). Thin membranous glottic webs can be treated with endoscopic techniques (incision with instruments or with carbon dioxide laser, followed by dilations) while major webs and those involving the subglottis region require open laryngo-tracheal reconstruction (37).

Paralysis of the vocal cords

This is the third most common cause of stridor in children and the second most common laryngeal abnormality.

Unilateral vocal cord paralysis can result from traumatic birth or thoracic surgery and most often affects the left side (Figure 5).

Bilateral paralysis may be idiopathic or due to causes like birth trauma with hypoxia, associated severe malformation of the central nervous system (e.g. Werdnig-Hoffman disease, brainstem dysgenesis) or conditions resulting in increased intracranial pressure (e.g. Arnold-Chiari malformation, myelomeningocele, hydrocephalus) (38). The rare paralysis in abduction is due to impairment of the adductors ("open glottis") and is characterized by aphonia and aspiration. On the other side, the more common paralysis of the vocal cords in adduction ("closed glottis") is due to abductor paralysis and may present with hoarseness, stridor and respiratory distress. In case of unilateral paralysis the child may be asymptomatic or present mild stridor and occasional aspiration.

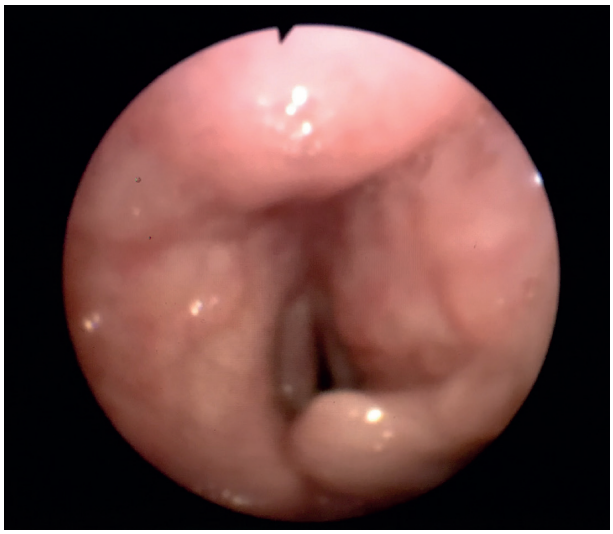

Figure 5. Right cord paralysis. Image property of P.S.

The diagnosis of unilateral vocal cord paralysis can be difficult to establish if the examination is not performed when the patient is awake or only mildly sedated.

Airway endoscopy shows:

- the lack of opening, in the inspiratory phase, of one or both vocal cords;
- the positioning in an anterior plane of the affected arytenoid;
- the hypoplasia of the paralyzed cord and the compensatory mechanism of the contralateral, whose arytenoid rotates and overlaps the other in a mechanism of conjunction.

In the diagnostic stage, in addition to endoscopy, a complete neurological evaluation and cerebral magnetic resonance should be performed in the neurological forms. In the peripheral forms, a cardiological evaluation of any expansive endothoracic and mediastinal processes is required.

Therapeutically, if the condition is well tolerated and the growth of the child is adequate, no procedure is necessary. In more severe cases, a temporary tracheotomy or endoscopic surgery including vocal cord lateralization procedures, arytenoidectomy, and laser cordotomy is performed (14).

With regards to vocal cords, an acquired condition resulting in stridor and that sometimes requires endoscopy is *vocal cord dysfunction*. Vocal cord dysfunction is a dysfunction in which the vocal cords show paradoxical adduction during inspiration or during inhalation and exhalation. The resulting symptoms and signs are: sudden onset of dyspnea, tightness in the throat or chest, inspiratory or biphasic stridor or wheezing and coughing. This abnormality is often found in association with asthma and gastroesophageal reflux; exercise and physical activity are also known as triggers of acute episodes (39). The duration of the attacks may vary from few minutes to days. Important clues in the differential diagnosis of asthma are inspiratory dyspnea, absence of symptoms at night, no oxygen desaturation and absent or only marginal response to bronchodilator therapy. The spirometry curve can show variable inspiratory flattening indicative of extrathoracic airway obstruction (40). Diagnosis is often difficult and endoscopy should be conducted during symptoms without anesthesia.

Hemangioma of the larynx

Hemangiomas are the most common tumors in childhood and are a rare cause of stridor in newborns. Lesions often show rapid growth up to 6-10 months, with involution usually at the age of 18 months. They may occur in the supraglottic (less common) or subglottic (more common) area.

Most subglottic hemangiomas are localized unilaterally with or without posterior extension and often in patients with other cutaneous hemangiomas of the head or the neck (41) (Figure 6). Endoscopic recognition may be difficult as the lesion may be covered with normal epithelium.

Hemangiomas are generally asymptomatic at birth and tend to manifest in the first months of life with biphasic stridor, predominantly expiratory, barking cough and sometimes failure to thrive. Larger hemangiomas may also occur at birth.

Propranolol has been demonstrated to be a well-tolerated medication to treat hemangiomas in infancy. The majority of patients do not need treatment beyond 18 months of age. Potential side effects may include hypotension, bradycardia and hypoglycemia.

Contraindications for starting propranolol are hypoglycaemic episodes, cardiac arrhythmia,

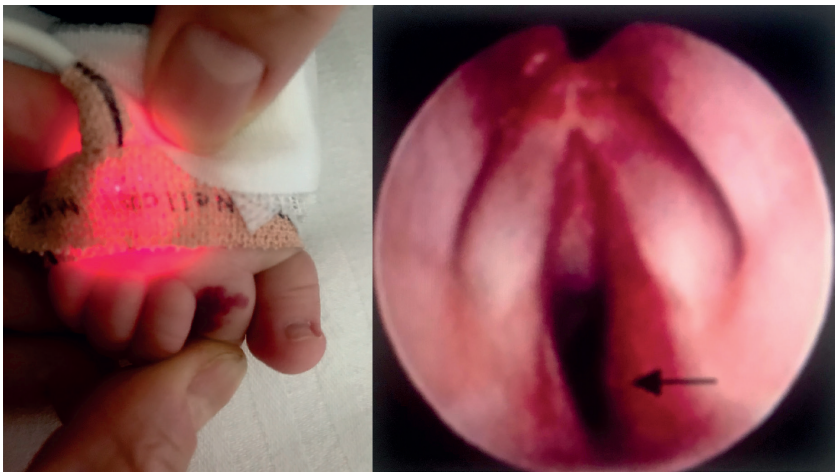

Figure 6. Cutaneous and subglottic manifestation of hemangioma (right vocal cord) in a neonate. Image property of P.S.

hypersensitivity to propranolol, recurrent wheezing and abnormal values of pressure or heart rate.

Pre-treatment investigations include cardiovascular and respiratory assessment, kidney, liver and thyroid blood examinations (42).

Laryngotracheal cleft (LTC)

LTC is a rare disorder that simultaneously involves the upper airway and the digestive tract. This disorder originates from a fusion defect in the midline of the tracheoesophageal wall during embryonic development. It can be associated with trachesophageal fistula, anal atresia, cleft lip, Meckel's diverticulum, tracheal and bronchial stenosis, cardiovascular defects. Males are more affected than females. Although the defect is rare, it can be burdened with considerable mortality and morbidity (43-45).

The Benjamin-Inglis classification (46) identifies 4 types of LTC:

- Type 1, interarytenoid defect up to the level of the vocal cords;
- Type 2, the defect involves the cricoid cartilage posteriorly;
- Type3, the defect involves the cricoid cartilage posteriorly and extends to the cervical trachea;
- Type 4, defect extension to the thoracic trachea (Figure 7).

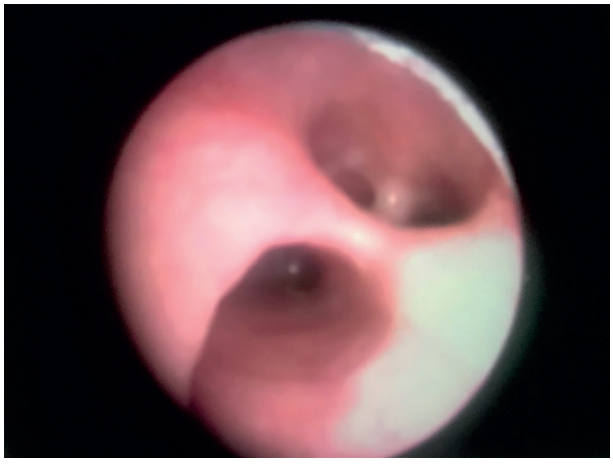

Figure 7. Endoscopic view of type 4 laryngotracheal cleft extending to thoracic trachea (trachea with carina in the upper part of the image and esophageal lumen in the lower part). Image property of P.S.

LTC grade 3 and 4 are considered life-threatening diseases.

The onset is particularly evident during feeding, when inhalation causes cyanosis, coughing and choking. Inspiratory stridor may be associated, mostly due to the collapse and redundant mucosa of the supraglottic structures that reduce the patency of the airway lumen in the inspiratory phase. The concomitant presence of expiratory stridor indicates an association with tracheomalacia.

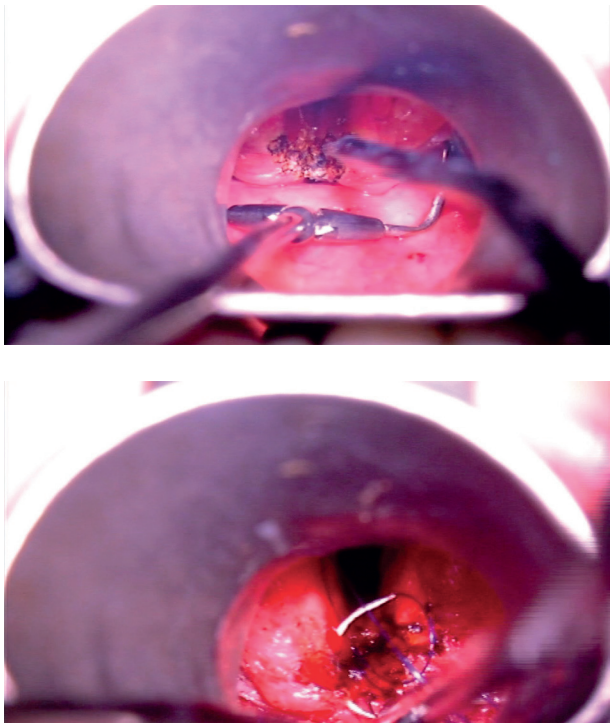

Figure 8. Endoscopic treatment of laryngotracheal cleft grade 2 with suspension microlaryngoscopy: laser cruentation of the cleft portion and suture with stitches. Image property of P.S.

Diagnosis is made with esophagogram and airway endoscopy, which allows the correct definition and extension of the defect. The endoscopic examination is performed in microlaryngoscopy with flexible bronchoscopy and inspection of the interarytenoid space with rigid optics at 0°. The rigid bronchoscope is ideal for exploring the posterior portion of the glottis, the subglottic region and the posterior part of the first tract of trachea.

Endoscopic approach with closure of the defect is usually adequate in LTC type 1 and 2 (Figure 8) while type 3 and 4 must be treated with open surgery (47).

Laryngeal cyst

Supraglottic cysts, commonly located in the ariepiglottic folds or epiglottis, are usually congenital. In contrast, subglottic cysts are usually acquired following an airway trauma like intubation (Figure 9).

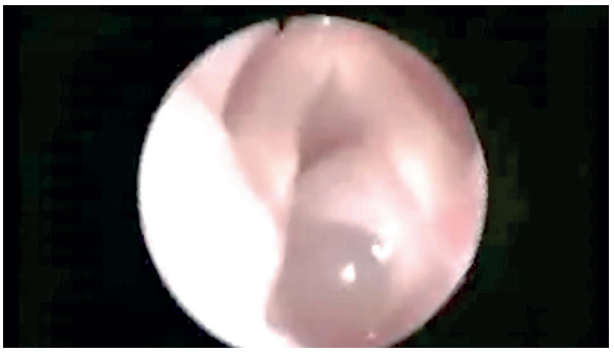

Figure 9. Ductal subglottic cyst showing translucid surface and clear mucus content. Image property of P.S.

The appearance of cysts varies considerably: while some are covered by thin and easily recognizable mucous membranes, others appear as a submucosal mass. A laryngocele is a rare special form of laryngeal cyst, which originates from the laryngeal ventricle, consists of an air-filled saccule and can be difficult to diagnose endoscopically (27). Newborns commonly present with stridor, hoarseness, weak crying or aphonia and sometimes feeding difficulties. They are often prone to bacterial infection with typical abscess.

Ductal or mucus retention cysts are the most common in children. They are caused by mucus retention in the duct of the submucosal glands. They are often detected in patients with a history of intubation or traumatic/inflammatory disease. Saccular cysts are distinguished from laryngocele because the lumen is not connected with the larynx, is not air-filled and is covered by normal mucosa. Saccular cysts cause stridor with dyspnea, cyanosis and difficulty in feeding from the first days of life. Glosso-epiglottic cysts are capable of generating respiratory obstruction by compression of the epiglottis and they simulate laryngomalacia-like symptoms that are exacerbated in supine position (14).

Treatment options include conservative medical management, cyst aspiration, endoscopic excision, microlaringoscopy or bronchoscopy marsupialization, deroofing with CO_2 laser and microdebrider. Some authors advocate for complete excision of the cyst by CO_2 laser after decompression by aspiration. Surgical

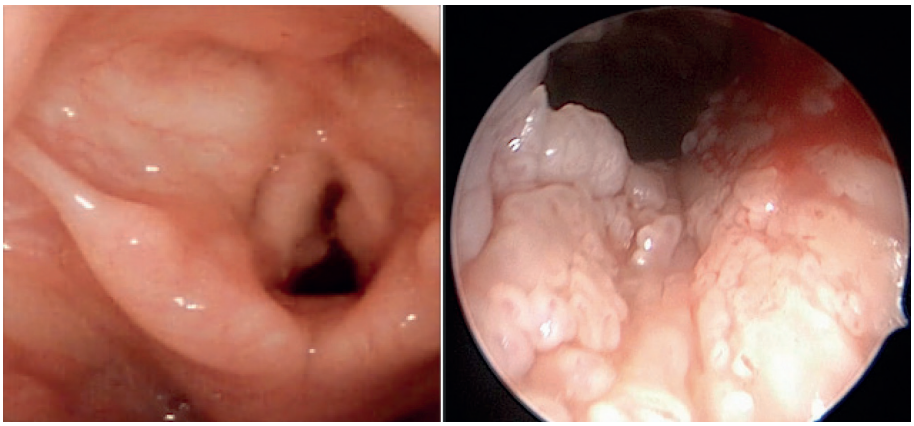

Figure 10. A 5-year-old boy with multiple vocal cord papillomas. On the right, papillomas in the trachea. Image property of P.S.

removal through an external approach is conducted in patients with recurrence of the cyst or in case of large cysts extending beyond the larynx (48,49).

Laryngeal papillomatosis

Laryngeal papillomatosis is a rare disease, but it represents the commonest benign neoplasm of the larynx in childhood. It is caused by Human papillomavirus types 6 and 11 and is characterized by the proliferation of squamous papillomas (50). The clinical course is often unpredictable. Spontaneous regression is possible but a spread of papillomatosis throughout the respiratory tract with associated significant morbidity and mortality can occur. Vocal cords are usually the first and predominant sites to be affected. The lesions typically appear as multiple, sessile or pedunculated, irregular and friable masses that may also extend into the airways (Figure 10). Presenting symptoms are usually hoarseness and stridor, less commonly cough or difficulty breathing.

Laryngeal lymphangiomas

Laryngeal lymphangiomas are rare lymphatic vessel malformations. Patients can be asymptomatic or present with snoring, stridor, increasing respiratory distress and airway obstruction. Endoscopy is crucial for diagnosis and less severe cases can be used to perform laser ablation (11).

Conclusions

In newborns and children respiratory signs and symptoms can be determined by congenital or acquired anomalies of the airways. After a detailed medical history and clinical examination, endoscopy is essential to investigate the presence of upper airway obstruction and make a precise diagnosis. A descriptive flowchart of the anomalies found in the nose, mouth, pharynx, larynx and vocal cords is presented in Figure 11. When performed by experienced operators, endoscopy is a safe procedure and can sometimes provide an effective treatment.

Conflicts of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

Author Contribution: conceptualization (PS), methodology (PS, VF, NM), investigation (PS, RB, SA), data curation (PS, VF, NM, LB), original draft preparation (PS), review and editing (VF, DC, RB, SE, MM).

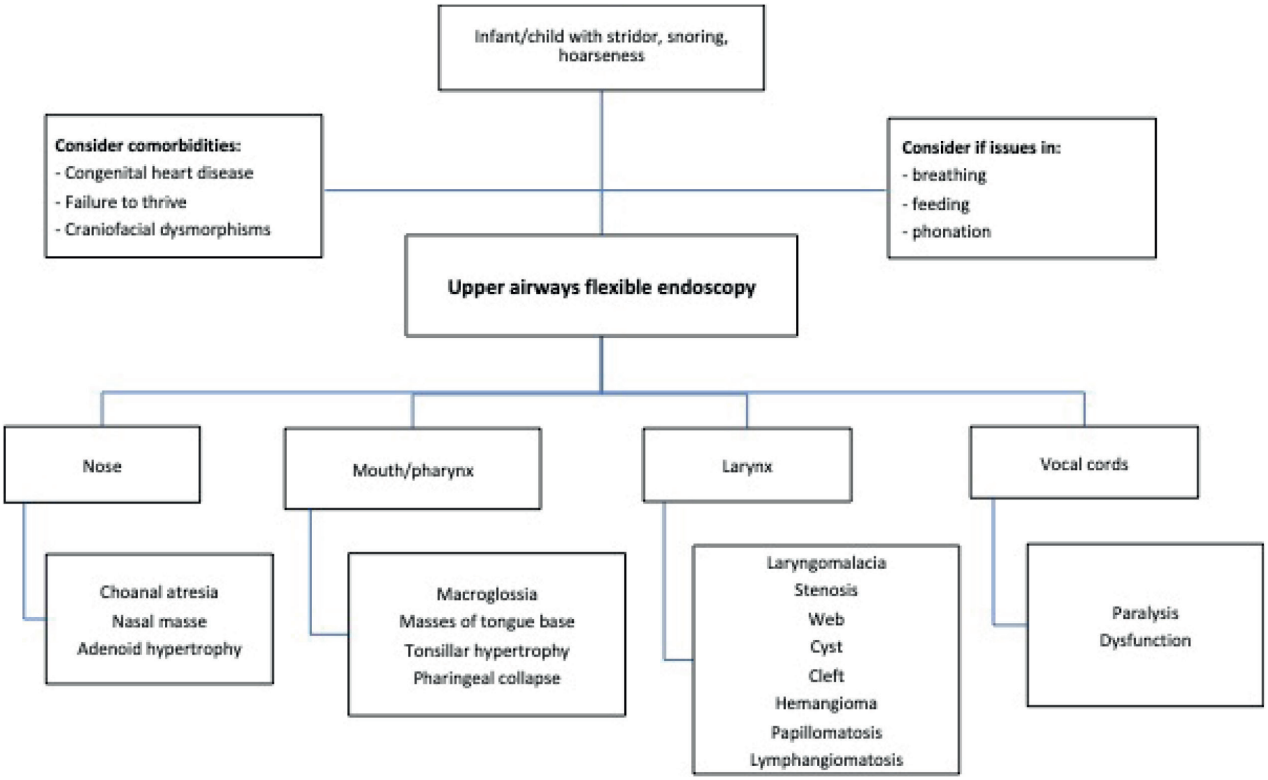

Figure 11. Flowchart of the anomalies found in the nose, mouth, pharynx, larynx and vocal cords.

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