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Message from Chairman Pediatric Respiratory Society, Delhi NCR

It has been a long association of more than eighteen years since its inception in 2005 when a small group of learned paediatric pulmonologist working in Delhi-NCR and general paediatricians came together with the common interest to promote the teaching and learning in the field of paediatric pulmonary medicine.

Over the period of time this family grew to an academic association and under the guidance of respected Dr. A K Sharma, we got it registered as **"Paediatrics Respiratory Society (PRS)"**. Working with the PRS group in different capacity as Finance Secretary, Honorary Secretary, Vice president and now as President has been a great learning and an enlightening experience for me.

To start with we had our bimonthly clinical meeting at various academic institutions and with the wisdom and support of our four pillars Dr. K Chugh, Dr. G R Sethi, Dr. S K Kabra and Dr. Varinder Singh we unfolded our wings to achieve the new heights. The group members have been on the forefront in promoting the knowledge and awareness about diagnosing and treating the respiratory diseases as per the guidelines to help the little patients reap the benefits of the latest developments in medicine. Also, PRS members have been instrumental in formulating and establishing the national and international guidelines on Acute Respiratory Infections, Asthma and allergic disorders and disseminating this knowledge at nation-wide level. The exceptional work done by respected Dr. Sangeeta Sharma, Dr. Varinder Singh and dear Dr. Ankit Parikh in the field of Paediatric tuberculosis has got recognition the world over and also by WHO.

To continue with the idea of promoting the latest in the field of paediatrics pulmonary medicine; PRS has started the publication of its E- journal and I am highly thankful to Dr Anil Sachdev for providing his tireless services as Editor-in-Chief and Dr. Neetu Talwar, Dr. Vineet Sehgal and Dr. Dhiren Gupta for supporting this endeavour.

This issue is dedicated to the Upper Airway Diseases which often remains ignored by the parents and thus under diagnosed and also undertreated. Though not contributing to significant mortality but none the less this group of diseases are responsible for significant morbidities and thus require due attention, recognition and appropriate management.

Our annual academic event as **Delhi Respicon 2023** is scheduled to be held on Aug 20, 2023 at AIIMS New Delhi. With more than two hundred registered delegates and distinguished faculty members, oral paper presentations by participants and about thirty-five poster paper presentations, we expect it to be a great academic feast.

Our team will continue to put in the best possible efforts to deliver a good learning experience through various conferences, CME's and workshops for all PRS members.

Regards

Dr Tilak Raj Dangwal

Editorial Board

INDEX

Acute Upper Airway Obstruction

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Abstract

Acute upper airway obstruction is not uncommon in pediatric age group especially in infants and early childhood. Majority of causes are acquired and infectious and sometimes are life threatening. Relevant history with quick clinical examination, prompt diagnosis, early institution of appropriate treatment and multidisciplinary approach can save catastrophic results. Advanced airway management may be needed.

Keywords: Airway obstruction; Stridor; Croup; Life threatening event

Introduction

Upper airway obstruction (UAO) is a common but potentially serious problem in pediatric age group. The upper airway extends from the nasal passages to the lower end of the cricoid cartilage (subglottis). Upper airway obstruction refers to conditions where the upper airway is obstructed leading to respiratory distress. The etiology varies from simple nasal blockage in a newborn to near fatal epiglottitis in a child. Even with partial airway occlusion, symptoms can be severe. Patients typically present with tachypnea, stridor, and grunting, although infants with complete obstruction may have apnea, and their condition can deteriorate quickly to cardiopulmonary arrest. (1) The findings from the history and physical examination are often nonspecific, leaving clinicians to rely on imaging findings to identify the cause for acute obstruction. In one study, severe upper airway obstruction accounted for 3.3 % of all pediatric intensive care unit (PICU) admissions. (2) Stabilisation of the infant and early recognition of the etiology helps in minimising complications and ensuring appropriate definitive therapy.(3, 4)

Pathophysiology

There are several anatomical peculiarities in the airways of a child, which makes it more prone to obstruction. These 'developmental disadvantages' are

- Relatively larger and prominent occiput causing flexion of neck in supine position
- Relatively larger tongue
- Anterior and cephalad position of the larynx
- Soft and omega shaped, vertically positioned epiglottis.
- Narrowest portion of the airways is at the level of the cricoid cartilage which is non-distensible(5).

Slight reduction in the calibre of the small cause severe airway obstruction (Poiseuille's law: $R=8\eta L/\pi r^4$). But during periods of turbulent airflow (as in case of a struggling child), the resistance to airflow becomes inversely related to fifth power of the radius of the airways. (5) As infants are obligate nose breathers, nasal block can precipitate significant respiratory compromise. During inspiration, the intraluminal pressure in the upper (extra-thoracic) airway becomes negative, causing collapse of the airways. The resultant narrowing makes the airflow turbulent producing inspiratory stridor. During exhalation, on the other hand, the intraluminal pressure exceeds the atmospheric pressure and the pressure exerted by the surrounding tissue, dilating the airway in cases of dynamic obstruction and improving the airflow. The configuration of the glottis may also predispose it to collapse during inspiration than exhalation(4). Stridor is the most common clinical manifestation of upper airway obstruction and described in table 1.

Table 1. Character and causes of stridor

Stridor	Characteristic auditory finding of upper airway obstruction
Inspiratory	Extrathoracic lesions (e.g., laryngomalacia, vocal cord lesion)
Expiratory	Intrathoracic lesions (e.g.,tracheomalacia, extrinsic compression)
Biphasic	Fixed lesions (e.g., Croup, laryngeal mass or web)

Stridor should be differentiated from stertor (a low-pitched inspiratory snoring sound typically produced by nasal or nasopharyngeal obstruction) and wheeze (musical, high pitched, polyphonic/monophonic and usually during expiration).

According to the 'Holinger's laws' of airway obstruction (5), in a child with noisy breathing, if the noise is worse during sleep, the obstruction is nasal or pharyngeal. If the symptoms are worse when the child is awake or exacerbated, the obstruction is typically laryngeal, tracheal or bronchial.

Etiology

UAO, congenital or acquired, has been classified into acute and chronic onset as mentioned in Table 2,3. Introduction of Diphtheria and Haemophilus influenzae vaccinations have dramatically reduced incidences of laryngeal diphtheria and acute epiglottis.

Table 2. Infectious and non-infectious causes of upper airway obstruction

Acute	**Chronic**
Infections	**Infections**
• Laryngotracheitis/ Laryngotracheobronchitis (LTB) or croup • Acute epiglottitis • Bacterial tracheitis • Laryngeal diphtheria • Retropharyngeal abscess • Tonsillar/ peritonsillar abscess • Ludwig's angina • Infectious mononucleosis	• Adenotonsillar hypertrophy • Chronic tonsillitis
Non-infectious	**Non-infectious**
• Airway foreign body • Angioneurotic edema • Airway trauma (penetrating/ blunt) • Airway burns (caustic/ thermal) • Vocal cord paralysis • Vocal cord dysfunction	• Choanal atresia/ stenosis • Laryngomalacia • Tumor of larynx (hemangioma, papilloma, cystic hygroma) • Vascular ring • Tracheal/ subglottic stenosis • Craniofacial anomalies (e.g., Pierre-Robin sequence) • Dysmorphic syndromes (e.g., Crouzon syndrome, Treacher-Collins syndrome)

Table 3. Congenital causes of upper airway obstruction in children

	Malformation	Characteristics
Nose	Nasal deformities	Choanal atresia or agenesis, septum deformities, turbinate hypertrophy, vestibular atresia, or stenosis.
Pharynx	Craniofacial anomalies	Anomalies causing facial retrusion are associated with upper airway obstruction, including Crouzon, Pierre Robin, and Apert syndromes.
	Tongue	Macroglossia and glossoptosis.
Larynx	Laryngomalacia	Most common cause of chronic stridor in infants. Almost all patients present by 6 weeks of age. Symptoms are more pronounced after upper respiratory infections.
	Laryngeal webs	75% located in the glottic area. Complete webs cause respiratory distress at birth and partial webs produce stridor, weak cry, and different degrees of respiratory distress. Associated anomalies are common.
	Laryngeal cysts	If located in supraglottic area, may cause respiratory distress and stridor.
	Laryngeal clefts	Characterized by abnormal communication between the larynx and pharynx, sometimes extending downward between the trachea and esophagus. Patients may present with aspiration, cough, swallowing difficulties, respiratory distress, hoarse cry, or occasionally with stridor; often associated with other congenital anomalies.
	Subglottic hemangioma	Presents as with stridor and respiratory distress, usually worsening during the first few months of life. Often associated with cutaneous hemangiomas.
	Subglottic stenosis	May be congenital but more often acquired secondary to intubation. Usually located 2 to 3 mm below the glottis.
	Vocal cord paralysis	Idiopathic or secondary to a neurologic disorder (including Chiari II malformation, hydrocephalus, meningomyelocele, hypoxic cerebral palsy, and cerebral hemorrhage)
Trachea	Tracheal stenosis	Usually presents with stridor or both stridor and wheezing. If stenosis is significant, respiratory distress occurs.
	Vascular rings or slings	74% of vascular rings are symptomatic. The airway compression usually is intrathoracic, causing expiratory stridor. Associated anomalies are common.
	Tracheomalacia	Often associated with other congenital anomalies. May be secondary to a vascular ring or cysts. Worsens with upper respiratory infections, crying, coughing, or feeding. May cause severe spells with cyanosis.
Bronchi and distal airways	Bronchogenic cyst	May occur at any point throughout the tracheobronchial tree. Typically present during childhood with recurrent coughing, wheezing, or pneumonia, but may become symptomatic during infancy or adulthood or present as an incidental finding on chest radiographs.

Approach to Acute Upper Airway Obstruction

Acute UAO is most often medical emergency requiring rapid assessment with simultaneous stabilisation with adequate oxygenation and ventilation support. Any child with respiratory distress with or without noisy breathing may have airway obstruction. Stridor is the most pertinent clinical sign of upper airway obstruction. It is normally inspiratory but can be both expiratory and inspiratory. However severe degree of obstruction can present as silently as airflow is near absent.

Croup

Croup (laryngotracheitis) is a respiratory illness characterized by inspiratory stridor, barking cough, and hoarseness. The parainfluenza viral infection of the supra-glottis known as croup occurs in 3% of children 6 months to 3 years of age. (6) Children with croup have a barking cough, inspiratory stridor, hoarseness, and respiratory distress, symptoms that typically have an abrupt onset at night. (7) Severity assessment of croup is done by the 17-point Westley's clinical scoring system but is cumbersome in busy clinic or casualty settings. (Table 4) (8) 'Steeple sign' is the radiological hallmark seen in neck radiograph but is seldom required for diagnosis. Oral or systemic steroids are main stay of therapy and are equally effective. (9) They are effective in all severity of croup and irrespective of duration of disease . (10) Glucocorticoids reduced symptoms of croup at two hours, shortened hospital stays, and reduced the rate of return visits to care(9). Recent Cochrane data base suggested that a smaller dose of 0.15 mg/kg of dexamethasone may be as effective as the standard dose of 0.60 mg/kg. (11) Noninferiority was demonstrated for both low-dose dexamethasone(0.15mg/kg) and prednisolone(1mg/kg) compared to standard dose dexamethasone in recently concluded RCT. (12) The use of nebulized epinephrine either racemic or L-epinephrine is effective for treatment of croup. (13) The dose includes 0.5mg/kg of L-epinephrine (Maximum 5mg) in 1:1000 concentration or 0.05mg/kg of 2.25% racemic epinephrine diluted in 3-4 ml of normal saline. Antibiotics, steam inhalation, short acting β2 agonists, antihistaminic, cough suppressants and mucolytics have not been found to be effective. Heliox may not be more effective than 30% humidified oxygen for children with mild croup, but may be beneficial in the short term for children with moderate croup treated with dexamethasone. The effect of heliox may be similar to 100% oxygen given with one or two doses of adrenaline. Most cases of croup are mild in severity. Hospital admission rate is <5%. (14)

Table 4. Wesley Clinical Score (8)

Clinical features			t
Westley Score	**Severity**	**Description**	**Management**
≤ 2	Mild	Occasional Barking cough No stridor at rest Mild or no Retractions	▪ Home Treatment Symptomatic care including antipyretics and oral fluids ▪ Outpatient treatment Single dose or oral dexamethasone 0.15mg/kg to 0.6mg/kg (maximum 16mg) or oral prednisolone 1mg/kg
3 - 7	Moderate	Frequent barky cough, stridor at rest, mild to moderate retractions	▪ Single dose or oral dexamethasone 0.6mg/kg (maximum 16mg) ▪ Nebulized epinephrine ▪ Hospitalization is generally not needed but may be warranted for persistent or worsening symptoms after treatment with glucocorticoid and nebulized epinephrine
8 to 11	Severe	Frequent barky cough, stridor at rest, marked retractions, significant distress	▪ Single dose of oral/IM/IV dexamethasone 0.6 mg/kg (maximum 16 mg) ▪ Repeated doses of nebulized epinephrine¶ may be needed ▪ Inpatient admission is generally required unless marked improvement occurs after treatment with glucocorticoid and nebulized epinephrine.
≥ 12	Impending respiratory failure	Depressed level of consciousness, stridor at rest, severe retractions, poor air entry, cyanosis or pallor	▪ Single dose of IM/IV dexamethasone 0.6 mg/kg (maximum 16 mg) ▪ Repeated doses of nebulized epinephrine may be needed ▪ Intensive care unit admission is generally required ▪ Consultation with anesthesiologist or ENT surgeon may be warranted to arrange for intubation in a controlled setting

Acute epiglottitis

Vaccination against *H. influenzae* type B has resulted in a lower incidence of epiglottitis and lower mortality associated with epiglottitis and pneumonia in children. (15,16) There is sudden onset of high-grade fever, difficulty in swallowing with drooling of saliva and preferential seating posture (Sniffing position). Often the mnemonic 4D's (Drooling, Dysphagia, Dysphonia and Dyspnea) is used to describe this condition. There is no barking cough which differentiate from croup. Most common organism responsible is Haemophilus influenzae, the epidemiology is changing with immunisation practices and other agents (Streptococcus and Staphylococcus) coming to the forefront. Patients with an aggressive disease course are typically male, have dyspnea or stridor, and present with edema of the epiglottis or aryepiglottic folds. (17) Elevated C-reactive protein levels and hyperglycaemia are also typical. X-ray neck lateral view may show typical thumb sign. Management includes hospitalization and intravenous 3rd generation cephalosporins (ceftriaxone) with supportive care. Rifampicin prophylaxis is recommended for close contacts of proven H. influenzae infections, at a dose of 20 mg/kg (maximum 600 mg) once daily for 4 d. Some experts even suggest elective intubation to avoid complications of emergency intubation. (18)

Bacterial tracheitis

Also known as the pseudomembranous croup or bacterial croup, it is most commonly caused by S. aureus. The median age of affected children is around 5 y. The typical history is of a child initially having symptoms of mild LTB deteriorating rapidly to develop increasing respiratory distress, toxic appearance, orthopnea and dysphagia(4). Endoscopy (flexible or rigid), done under general anesthesia, has a very important role in both diagnosis and management by removal of the exudative pseudomembrane and might also avert the need for intubation(4). Need for endotracheal intubation has been reported to be between 38 and 100 %. (19) Choice of IV antibiotics is usually a combination of 3rd generation cephalosporin (e.g., Ceftriaxone, Cefotaxime) and antistaphylococcal penicillin (e.g., Cloxacillin) for 10–14 d. (20)

Laryngeal diphtheria

It is caused by toxigenic strains of *Corynebacterium diphtheriae*. Classically, it presents as an insidious onset fever, sore throat, cervical lymphadenopathy and neck edema (bull neck). Laryngeal form is typically an extension from pharynx in children. A characteristic thick, dirty, adherent grey membrane which bleeds on attempted removal clinches the clinical diagnosis. A throat

swab should be sent for Albert stain and culture for bacteriological confirmation. The American Academy of Pediatrics recommended medical management comprises of Diphtheria antitoxin 20,000–40,000 U and antimicrobial therapy with penicillin or erythromycin given either orally or parenterally for 14 d. Active immunisation is also advised during convalescence.

Retropharyngeal/ Parapharyngeal abscess

Higher incidence rates in preschool children depends on both infectious and anatomical factors. In the first years of life, upper respiratory tract infections and cervical adenitis are significantly more common. Moreover, in the same period of life the paramedian chain of lymph nodes in the retropharyngeal space is prominent, whereas it tends to involute after the fifth years of age. Retropharyngeal and parapharyngeal abscesses are typically caused by the suppuration and perforation of lymph nodes, which serve as a drainage pathway of body sections of the upper respiratory tract. (21,22) Clinical course is with high fever, sore throat, dysphagia, neck pain with limitation of movement of neck or torticollis. A lateral neck radiograph may reveal widening of prevertebral soft tissue and air-fluid level in the retropharyngeal region, though the investigation of choice is a contrast CT scan to establish the diagnosis and also to plan the management. Management comprises of surgical drainage with IV antibiotics especially in cases with compromised airway. However increasing number of cases is being successfully treated with antimicrobials alone. (23) Broad spectrum antibiotics are covering S. aureus are first line of therapy, choice being Ampicillin-Sulbactam or Clindamycin for methicillin sensitive *Staphylococcus aureus* (MSSA) and Vancomycin or Linezolid for suspected methicillin resistant *Staphylococcus aureus* (MRSA) infection with total duration (IV+oral) being typically 14 d. (24)

Peritonsillar abscess

Peritonsillar abscess is a localised infection in the peritonsillar space (between the fibrous capsule of the tonsil and the superior pharyngeal constrictor muscle); it usually follows acute tonsillitis and is polymicrobial in etiology with both aerobic and anaerobic bacteria being involved. (25) It is more frequently diagnosed among adolescents. (26) The clinical diagnosis can be strongly suspected on the base of severe unilateral sore throat, cervical lymphadenopathy, tonsillar or pharyngeal exudates, uvular deviation toward the unaffected side and upper airway obstruction. (27) Apart from airway stabilization, these children are managed with surgical interventions coupled with IV antibiotics. (28) Most commonly employed antibiotics are a combination of 3rd generation cephalosporin with Clindamycin, with other alternatives being Meropenem, Imipenem and Piperacillin-Tazobactam. (29) Systemic review does not support the addition of metronidazole in

the first-line therapy. (30) Three surgical options are possible: needle aspiration, incision and drainage, and abscess tonsillectomy. Tonsillectomy in PTA is controversial: it should be considered in patients with recurrent tonsillitis, obstructive sleep apnea or in case of the failure of other techniques. (31)

Ludwig's angina

Ludwig's angina is characterized by cellulitis of the floor of the mouth (submandibular, sublingual, and submental spaces) all of which result in a posteriorly displaced tongue. It is commonly triggered by oral sepsis. Ludwig's angina causes substantial edema, distortion, and airway obstruction, making it a potentially life-threatening disease. The most common causes of Ludwig's angina are dental infections, followed by sialadenitis, peritonsillar abscess, abscess involving the parapharyngeal space, traumatic injuries to the oral cavity, and mandibular fractures. In the past, before the introduction of antibiotics, the mortality rate for Ludwig's angina exceeded 50%. (32) Early recognition and treatment for Ludwig's angina are of paramount importance due to the myriad of complications that can occur in association with Ludwig's angina. Known complications of Ludwig's angina include carotid arterial rupture or sheath abscess, thrombophlebitis of the internal jugular vein, mediastinitis, empyema, pericardial effusion, osteomyelitis of the mandible, subphrenic abscess, aspiration pneumonia, and pleural effusion. Airway management is the first step in the medical management of Ludwig's angina as airway compromise is the leading cause of death. Previous findings in a retrospective review advocated the use of elective awake tracheostomy as a much safer method than endotracheal intubation. (33) Early antibiotic therapy is of critical importance for successful treatment. Penicillin G, metronidazole, or clindamycin are good choices as initial coverage. Moreover, intravenous steroids and nebulized adrenaline use have been shown to allow for easier intubation avoiding tracheostomy or cricothyroidotomy and allowing for increased penetration of antibiotics into the fascial spaces by reducing edema and cellulitis. (34) Conservative management with intravenous antibiotics is associated with a risk of airway compromise that is nearly 10 times as high as that in patients who receive early surgical drainage (26.3% vs. 2.9%)(35). Surgery is indicated for patients who develop abscesses and are unresponsive to antibiotics and medical management. This is usually accomplished by decompressing the submental, submandibular, and sublingual spaces by external incision and drainage (33). Overall, the mortality associated with Ludwig's angina is approximately 8%. (36)

Airway foreign body

Foreign body aspiration (FBA) is a life-threatening pediatric emergency. The injuries, which are causes of morbidity and mortality in all age groups, are seen mainly in children under 3 years old especially male and is the fourth leading cause of accidental death in this group and the third in infants under 1 year. (37) Commonest inhaled objects are food materials(especially peanuts) followed by toys parts. (38) Severity of clinical presentation depends on age, type of object inhaled, time since inhalation, location of object and degree of obstruction. The classical tried is cough, wheeze and unilateral decreased breath sounds is present only in 57% cases. (39) In case of complete airway obstruction, Heimlich maneuvre in older children and alternate back blows and chest compressions in infants, should be attempted. Plain radiography of neck and chest can be normal or presence of foreign body, unilateral hyperinflation, collapse, mediastinal shift can be seen. Virtual bronchoscopy has no role in acute setting(40). In case of partial obstruction, rigid bronchoscopy is the preferred procedure for both diagnosis and management with success rate of 95% and complication rate of 1%. (39)

Angioedema

It is characterised by Angioedema is characterized by (i) sudden, pronounced erythematous or skin-colored deep swelling in the lower dermis and subcutis or mucous membranes, (ii) tingling, burning, tightness, and sometimes pain rather than itch,(iii) resolution slower than that of wheals (can take up to 72 h). (41) It can be hereditary, acquired, drug induced, or idiopathic. (42) (Table 5) Lips swelling, urticarial rash and itching are common clinical findings while recurrent episodes suggest hereditary angioedema (HAE). Table 5 describes clinical subtypes of angioedema with laboratory profile.

Table 5. Subtypes of angioedema and related laboratory anomalies (42)

Diagnosis	C4 level	C1inhibitor function	C1 inhibitor level	C1q level
Hereditary angioedema				
Type I	Low	Low	Low	Normal
Type II	Low	Low	Normal	Normal
Acquired angioedema	Low	Low	Normal or low	Low
ACE-inhibitor associated angioedema	Normal	Normal	Normal	Normal

ACE denotes angiotensin-converting enzyme and C1q a protein that is part of the classic complement pathway

The most common genetic form of HAE in up to 85% of cases is caused by low levels of C1 esterase inhibitor (C1-INH) protein, leading to a bradykinin-mediated increase in vascular permeability. During an attack of HAE, abortive treatment with C1-INH replacement is most commonly described, however, icatibant, ecallantide, or fresh frozen plasma are also used. (43) Anaphylaxis is treated with intramuscular adrenaline, fluid support, H1 antagonists, ß2 agonist inhalation and intravenous corticosteroids. (44) The consensus guidelines recommend screening children from families with a history of hereditary angioedema and all offspring of an affected patient. (45) Because the diagnosis is often delayed, a low threshold for screening in the emergency department is recommended, particularly in patients with a family history of angioedema. (44,45)

Conclusion

Acute upper airway obstruction in children is a serious condition and sometimes life threatening. Early diagnosis and initiation of medical treatment with airway management are lifesaving. Early inclusion of pediatric intensivist and otorhinolaryngologist is often needed in the management of UAO.

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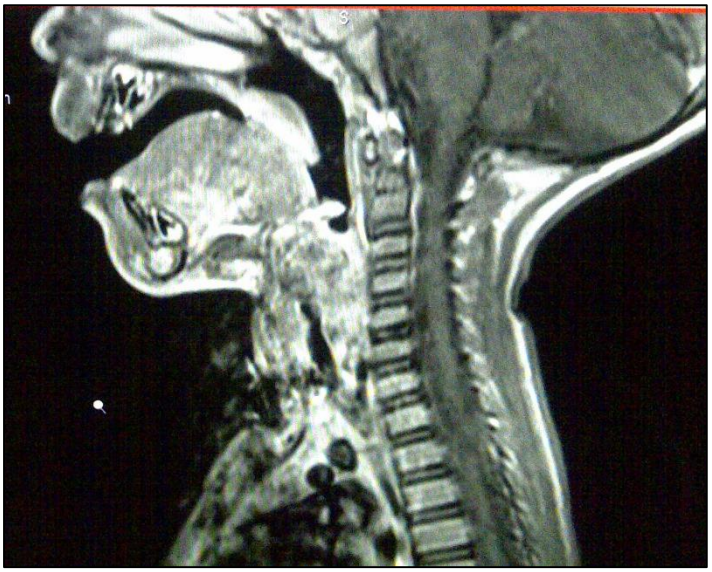
QUIZ

Your diagnosis please?



A rare cause of upper airway obstruction in a 6 months old male child presented with progressive stridor and acute on chronic respiratory failure.

Chronic Upper Airway Obstruction in Children

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Abstract

Chronic upper airway obstruction in children frequently mimics other conditions such as asthma or recurrent pneumonia. Clinical features such as stridor may lead to diagnosis. We will review the different causes of chronic upper airway obstruction and how to manage them. The management of UAO is based on the location and nature of the lesion.

Keywords: Airway obstruction; Stridor; Laryngomalacia; Adenoid hypertrophy

Introduction

Upper airway obstruction (UAO) is defined as occlusion or narrowing of the upper airways (nasal cavities, oral cavity, pharynx, and larynx) leading to compromise in ventilation. (1) The presentation of obstruction may be acute to chronic. It often mimics other respiratory conditions such as asthma or recurrent pneumonia. Here, we discuss the pathophysiology and management of chronic UAO. Causes of chronic UAO are classified depending upon the site of obstruction:

1. **Nasal cavity**

i. Choanal atresia

Choanal atresia is a congenital disorder in which the paired openings of nasal cavity to the nasopharynx are occluded by membranous soft tissue, bone or a combination of both. (2) The basis of this anomaly is due to failed recanalization of the nasal fossa at the time of fetal development - such as persistence of buccopharyngeal membrane, nasobuccal membrane of Hochstetter or the partial resorption of the nasopharyngeal mesoderm. (3) It can occur unilaterally or bilaterally. If unilateral, it manifests as unilateral mucopurulent discharge. If bilateral, the neonate is unable to breathe, presenting with respiratory failure. It should also be suspected in a neonate who turns cyanotic with feeding, that improves on crying. Since newborns are obligate nasal breathers, obstruction of airway may present as an acute emergency. Many patients present with chronic recurrent nasal discharge on the affected side. Treatment is usually surgical, however, endoscopic endonasal technique has less surgical complications with improved results.

ii. ***Nasal turbinate hypertrophy*** secondary to allergic rhinitis, nasal polyposis, old retained nasal foreign bodies or deviated nasal septum cause symptoms of obstruction.

iii. ***Juvenile nasopharyngeal angiofibroma*** **-** It is a vascular tumor that occurs in the posterior nasal cavity in adolescent males. The common presentations are chronic unilateral nasal obstruction and painless, unprovoked epistaxis.

2. Oral Cavity and Pharynx

Congenital causes of UAO at the level of the oral cavity include retrognathia (maybe idiopathic or associated with Pierre Robin Sequence, Treacher Collins syndrome, Goldenhar syndrome), glossoptosis, macroglossia (Beckwith Wiedemann Syndrome, storage disorders) and lymphovascular malformation. Acquired causes at this level include angioedema, Ludwig angina (cellulitis arising from dental infection at the floor of mouth). Potential traumatic causes include penetrating neck injury, burns/inhalation injury, or caustic injury, all of which can cause edema of oral cavity.

i. ***Adenotonsillar hypertrophy*** **-** It results from growth of lymphoid tissue along the Waldeyer's ring (enlarged adenoids and tonsils) in the nasopharyngeal region(figure 1). The commonest cause of adenoidal hypertrophy is recurrent infections caused by bacteria such as *Haemophilus influenza*, *Streptococcus pneumoniae*, *Streptococcus pyogenes* and *Staphylococcus aureus*. Frequent allergies and exposure to gastric juice as in gastroesophageal reflux disease, also promote acute or chronic inflammation associated with hypertrophy.

The clinical manifestations are chronic runny nose and post nasal discharge – as the inflamed adenoids are covered with mucopurulent secretions, there is passage of discharge which flows down to the back of the pharynx. Other symptoms include snoring, nasal obstruction, altered breathing pattern and obstructive sleep apnea. Otitis media with effusion, sinusitis, dental malocclusions, speech delay are known co – morbidities.

Initial approach to management is antibiotics such as amoxycillin for acute adenoiditis. Intranasal corticosteroids such as fluticasone propionate or mometasone are used in chronic/ recurrent adenoiditis. It has been found efficacious in reducing the size of adenoidal tissue on long term use, on a daily basis.

Adenotonsillectomy is indicated in children with obstructive sleep apnea or features of obstruction not relieved by medical management.

ii. ***Obstructive sleep apnea*** **(OSA) -** OSA occurs due to repetitive partial or complete cessation of airflow as a result of closure and re-opening of the upper airway during sleep. The presenting complaints are - increased effort of breathing, chest caving in odd sleep

positions, head extension, choking, gagging in sleep, mouth breathing, morning headaches, dry mouth, thirst, halitosis, difficult to control or worsening of epilepsy. (4) Due to this they have sleep disordered breathing and fluctuations in oxygen saturation. They also have increased risk of neurocognitive problems, metabolic syndrome and cardiovascular diseases. Diagnosis is made with the help of sleep related questionnaire, polysomnography, apnea - hypopnea index.

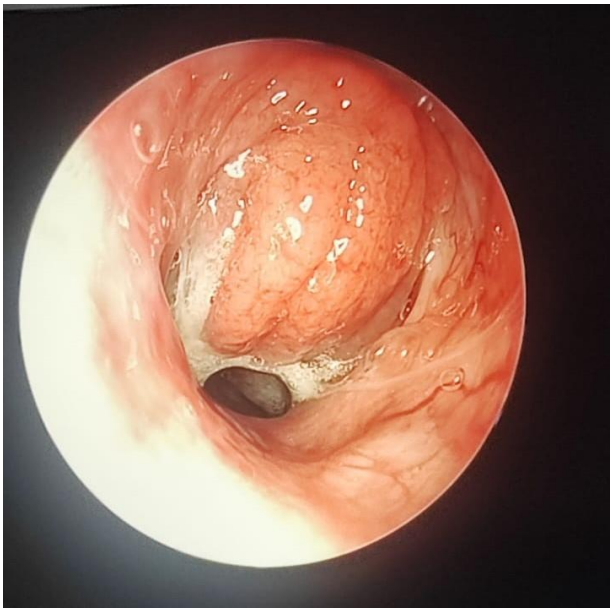

Figure 1. Endoscopic view of adenoidal hypertrophy. (Photo courtesy. Dr Ekta Narang, Assistant Professor Pediatrics, Chacha Nehru Bal Chikitsalaya, Delhi)

3. Vascular obstructive causes

i. *Vascular tumor*- Infantile hemangioma of infancy affects up to 5% of children and has a higher female preponderance. It can obstruct the airway when located in the oral cavity, pharynx or larynx. (5) Subglottic hemangioma represents 1.5% of congenital anomalies of the larynx. It is typically associated with segmental cutaneous hemangioma and presents as recurrent/persistent/severe croup without skin lesions. The mean age of diagnosis of an airway hemangioma is around 3.6 months. A subglottic hemangioma can be diagnosed on awake fiberoptic laryngoscopy and confirmed with angio- CT as an intensely contrast- enhancing, lobular, well delimited mass. For acute stridor due to subglottic hemangioma, short course of oral steroids significantly relieves symptoms. Propranolol used to manage subglottic hemangioma growth by inducing vasoconstriction, apoptosis or programmed cell- death and reduction of the vascular endothelial growth factors. It is usually started at a dose of 2 mg/kg/day in 3 divided doses. (6)

ii. Vascular malformations -

***Cystic hygromas*-** Head and neck lymphatic malformations may be classified based on De Serres Staging. (7) Radiologically, they may also be classified as microcystic (<2cm) and macrocystic (>2 cm) or mixed. (8) They may remain asymptomatic, but depending on site of lesion they may have mass effect. Sometimes there may be hemorrhage inside the lesion or superadded infection which can lead to rapid increase in size. If present in the neck, some children present with signs of impending respiratory failure due to obstruction and may require noninvasive ventilation and tracheostomy. Definitive treatment may require staged multimodal strategy, surgical excision and/or sclerotherapy.

4. Larynx

i. ***Laryngomalacia*-** Laryngomalacia occurs due to dynamic airway collapse of supraglottic structures into the laryngeal airway during inspiration (figure 2). It usually presents as an inspiratory stridor with suprasternal retractions in young children. Laryngomalacia is the most common congenital laryngeal anomaly. Often symptoms are not present at birth and affected children typically develop noisy breathing in the first 2 weeks of life with progressive worsening in the first 2 to 4 months. Severe laryngomalacia manifests as: respiratory failure due to upper airway obstruction, regurgitation of feeds, poor weight gain and obstructive sleep apnoea. Sometimes laryngomalacia may be associated with laryngeal clefts, tracheomalacia, vocal chord palsy, subglottic stenosis or as part of a syndromic disease.

Types of Laryngomalacia are as follows (9):

1. Inward collapse of aryepiglottic folds, primarily the cuneiform cartilages which are often enlarged
2. Long, tubular epiglottis (pathologic exaggeration of the normal omega shape)
3. Anterior, medial collapse of arytenoid cartilages
4. Posterior inspiratory displacement of epiglottis against the posterior pharyngeal wall or inferior collapse to the vocal cords
5. Short aryepiglottic folds

In most cases parental re-assurance is needed along with advice on feeding position and anti-reflux measures and prevention of recurrent respiratory tract infections. In severe cases, surgical intervention such as supraglottoplasty or rarely tracheostomy may be needed.

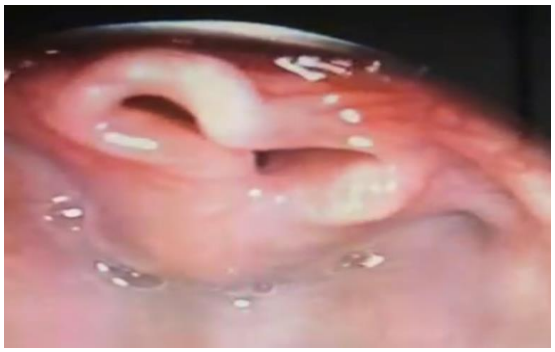

Figure 2. Endoscopic view of laryngomalacia (Photo courtesy. Dr Ekta Narang, Assistant Professor Pediatrics, Chacha Nehru Bal Chikitsalaya, Delhi)

ii. ***Vocal cord paralysis-*** It may be of following types:

Bilateral vocal cord palsy –It is usually idiopathic in nature. It may occur as a post intubation or surgical complication. Here, the vocal cords lie in paramedian position, leading to inspiratory stridor. Bilateral paralysis presents as an airway emergency with a near normal phonation and a high-pitched inspiratory stridor. Frequent aspirations lead to recurrent pulmonary infections. Management in 50% cases require tracheostomy.

Unilateral VCP is described as limited abduction of the vocal cord from the midline with deep inspiration. Unilateral VCP is caused primarily by iatrogenic injury to the left recurrent laryngeal nerve during cardiac surgery or following a difficult delivery requiring forceps. The left side is most commonly affected, likely because the longer course of the left recurrent laryngeal nerve presents greater opportunity for injury. It presents as noisy breathing, weak/feeble cry, choking on feeds and respiratory distress.

Certain neurologic syndromes such as Arnold Chiari malformation, polyneuropathy (Guillain-Barré syndrome), brainstem encephalitis may also present with vocal cord paralysis. (10)

iii. ***Congenital subglottic stenosis-*** It is also a commonly encountered laryngeal anomaly. It is defined as a diameter of less than 4 mm of the cricoid region in a full-term infant, and less than 3 mm in a premature infant. It results from incomplete recanalization of the laryngotracheal tube during the third month of gestation. It is of two types based on embryological defect. The most common type is the membranous congenital subglottic stenosis caused by submucosal hypertrophy with excess fibrous connective tissue. The other type is the cartilaginous type which results from an abnormal shape of the cricoid cartilage. (11) Severe subglottic stenosis results in biphasic stridor and laboured breathing. Flexible bronchoscopy establishes the diagnosis. Severe cases may necessitate endotracheal intubation and tracheostomy.

Laryngotracheoplasty may be required to achieve decanulation. In this procedure the subglottis is augmented with a rib or a thyroid ala cartilage graft. Milder cases often resolve on their own as the child grows.

Conclusion

Chronic upper airway obstruction can be due to congenital and acquired causes. In most cases flexible endoscopy establishes the diagnosis. Management is based on the location of obstruction and its degree of severity and require multidisciplinary approach.

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Unilateral Vocal Cord Palsy Associated with Congenital Heart Disease: A Case Report and Review of Literature

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Abstract: Stridor in infancy is not uncommon and presentation in a child may vary from noisy breathing alone to severe respiratory distress, feeding difficulty and apnea.The commonest cause of stridor is laryngomalacia. We described a 2-month-old female child presenting with stridor along with hoarse and weak cry for 1 month of age along with ventricular septal defect (VSD) with left to right shunt with right ventricular hypertrophy (RVH) with mild pulmonary arterial hypertension (PAH). This article reviews the literature regarding the presence of stridor in association with cardiac defect and discussed the pointer on history and examination suggesting a diagnosis other than laryngomalacia.

Keywords: Stridor, Vocal cord palsy, congenital heart disease.

Introduction

Noisy breathing is commonly found in children, especially newborns and infants. When a child or infant presents with stridor it is important to do a detailed clinical evaluation for likely underlying etiology, to assess for severity of obstruction and the risk of progression to respiratory failure to plan timely and appropriate intervention.Common causes of stridor in infants include laryngomalacia, tracheal stenosis, micrognathia, tracheo-esophageal fistula (TEF) and foreign body aspiration(1). We hereby describe an unusual cause of stridor in a young infant.

Case

A 2-month-old, immunized for age, exclusively breastfed girl was brought with complaints of noisy breathing, change in voice quality and weak cry noticed for the past 1 month. There was an additional history of suck rest suck cycle with feeding diaphoresis for

the past 1 month. The child had cough, coryza and fever for 3 days and fast breathing for 1 day. The noisy breathing did not vary with sleep, crying, feeding, or positional change. Cough was wet sounding, non spasmodic, non paroxysmal and was not associated with post-tussive vomiting

Born vaginally the child was full term, appropriate for gestational age (birth weight 2.8kg), and cried immediately at birth. There was history of 2 episodes of cough, coryza and fast breathing lasting 2-3 days each without the need for hospitalization. In between episodes, suck rest suck cycle and diaphoresis while feeding persisted. There was no associated lethargy, excessive irritability, rash, ear discharge or abnormal body movement. On examination, the child was failing to thrive (weight 2.4kg) with a respiratory rate of 62/min, a heart rate142/min. Oxygen saturation was 80% on room air and 96% at oxygen at 2litre per minute via nasal prongs. General physical examination did not reveal pallor, cyanosis, icterus, clubbing, edema or lymphadenopathy. Signs of rickets, including frontal bossing and rosary were present without signs of other vitamin deficiency. Respiratory system examination revealed subcostal and intercostal retractions. Auscultation revealed biphasic stridor along with occasional wheeze. Cardiovascular system examination revealed a pansystolic murmur best heard at the left lower sternal border. There was no hepatosplenomegaly.

Hemogram revealed hemoglobin 10gm%, total leucocyte count of 12000 (polymorphs 48%, lymphocytes 48%).C-reactive protein (0.8mg/l) and procalcitonin (0.4ng/ml) were normal. Serum Vitamin D level was12.2 ng/ml. Serum calcium was 8.5 mg/dL with normal ionized fraction. Liver and renal function tests were normal. Chest radiograph (Figure 1) revealed cardiomegaly with pulmonary infiltrate

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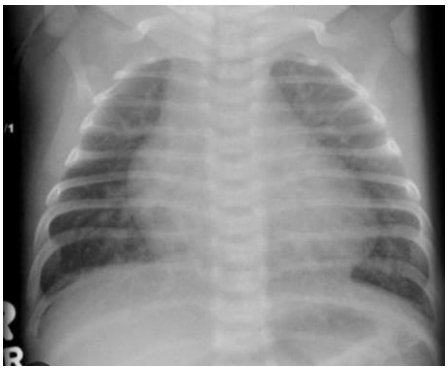

Figure 1 Chest radiograph AP view showing cardiomegaly and streaky perihilar and paracardiac infiltrates

The possibility of acyanotic congenital heart disease with community-acquired severe pneumonia with rickets with stridor was considered. Oxygen was continued via nasal prongs and antibiotics were initiated. Fever and fast breathing improved within 2 days. ECHO revealed muscular ventricular septal defect (VSD) of size5.85mm with the left to right shunt with dilated right ventricle with mild TR with mild PAH.

Flexible bronchoscopy evaluation revealed normal pharynx and epiglottis. Left side arytenoid and left vocal cord movement were markedly reduced compared to right side (Figure 2). Subglottis, trachea, carina and bilateral main bronchi were normal.

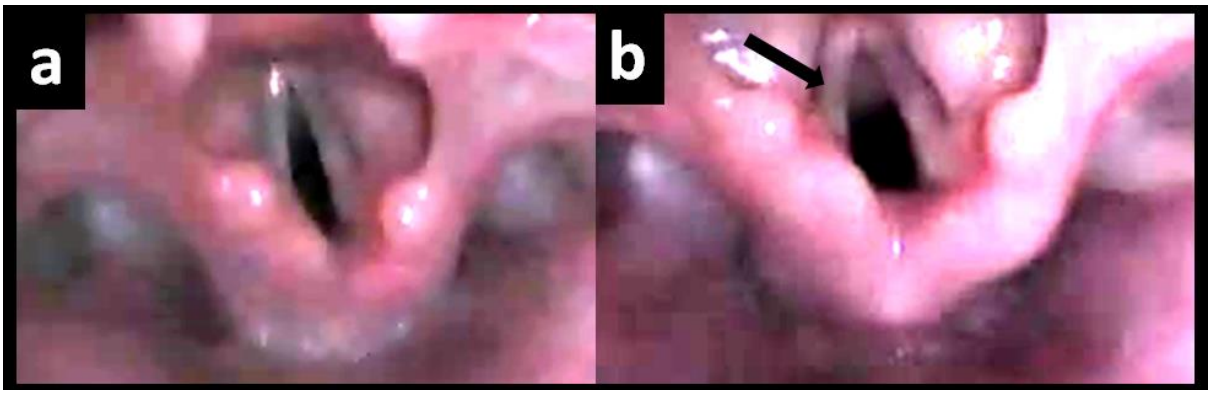

Figure 2. Vocal cords during adduction and abduction: (A) During adduction; (B) Decreased movement of left sided vocal cords (arrow) during abduction

The final diagnosis of ventricular septal defect (moderate) with left to right shunt with community-acquired pneumonia and left-sided vocal cord and arytenoids palsy with rickets was considered.

Discussion

The common causes of stridor in infancy are laryngomalacia, congenital subglottic stenosis, vocal cord palsy (VCP) or vascular compression of trachea. VCP accounts for 10-15% of cause of stridor (1).VCP may be unilateral or bilateral. Bilateral VCP is mostly idiopathic but may also be associated with neurological disease or birth trauma. Among neurological causes, Arnold Chiari malformation associated with meningomyelocele and hydrocephalus is common. Bilateral paralysis is diagnosed early compared to unilateral palsies (2) because of aphonia and more severe respiratory symptoms which may merit tracheostomy. Bilateral VCP related to birth injury may improve over two weeks in the presence of neuropraxia and may require longer in axonotmesis.(1)

Unilateral VCP predominantly affects the voice character while other respiratory symptoms are typically mild. It is caused primarily by iatrogenic injury to the left recurrent

laryngeal nerve (RLN) during cardiac surgery or following a difficult delivery requiring forceps. However, it also may result from nerve compression by a mediastinal mass or enlarged cardiothoracic structures.(2) Right and left RLNs are branches of vagus nerve that descend into neck and thorax.Right RLN branches at the level of subclavian artery and hook around it. Left RLN has a longer course compared to right, as it travels downward and loops around arch of aorta.It then ascends in tracheo-esophageal groove anterior to trachea and posterior to aortic arch and the left lobe of thyroid where it enters into larynx.As it passes under the aortic arch it passes through a limited triangular space formed by ligamentum arteriosum, aortic arch and left pulmonary artery. The nerve is vulnerable at this point and is at risk of compression by enlargement of any these. Left sided vocal cord palsy is more common due to this longer course of left RLN.

The first case of vocal cord paralysis in associated with cardiovascular pathology was described by Norbert Ortner wherein RLN was compressed by dilated left atrium associated with mitral stenosis. Stocker et al coined the term “cardio-vocal syndrome” noting the occurrence of left VCP.(3)

In infants and children cardio-vocal syndrome associated with CHD includes patent ductus arteriosus (PDA),double outlet right ventricle with atrial septal defect, VSD, thoracic aneurysm, surgical repair of coarctation of aorta and PDA ligation (4,5).

Other investigations which may help exclude other causes include a CECT scan covering the base of the skull to the mid-thorax and, MRI head. They may help identify masses causing compression, aberrant vessels, mediastinal masses and associated neurological anomalies.

Cardiopulmonary CT angiography is required for diagnosis of aberrant vessel or vascular ring. Role of flexible bronchoscopy to diagnose the cause of stridor in CDH is extremely important and its role to document correction of cause during recovery. (6) Evaluation of a child with stridor should include assessment of symptom severity and need for urgent intervention and detailed history and examination to indicate likely etiology of stridor,

If severity of stridor does not merit immediate intervention, a detailed history should include: age of onset, events preceding the onset of stridor, prenatal course and natal history, aggravating and relieving factors (like variation in stridor with position, sleep, cry, feeding), previous surgical procedures, ICU stays requiring prolonged intubation of airway, change in severity of stridor over time and whether child is thriving well. A focused examination should also be done which should include respiratory rate and distress, need for oxygen, phase of respiration wherein stridor is maximum, presence of cyanosis, apneic pauses, drooling, voice

character, voice volume, dysmorphism (especially pertaining to the craniofacial region), in addition to the pulmonary and cardiac examination.

Stridor in children is most commonly due to laryngomalacia accounting for upto 70% of cases. The pathogenetic mechanism is the collapse of supraglottic structures during inspiration. The stridor in laryngomalacia typically appears within initial several weeks of life. There is variation in stridor intensity with changes in the position of body, head and neck, crying, and feeding. Symptoms typically resolve in first 1 to 2 years of life without need for any surgical intervention. Severe laryngomalacia may have associated feeding difficulties, choking and cyanosis.

A study in Sydney by Davidson et al described 3 patients with VCP and associated CHD. Patient 1 was a full-term born child presenting at 5 weeks of age with a hoarse voice, weak cry, tachypnoea and a pansystolic murmur (PSM). After two months of medical management with diuretics, the VSD was restricted, and the voice and cry normalized with normal vocal cord movement on videoscopy. The second patient born full term had hoarse cry notice at two weeks of life and was diagnosed with congenital left VCP by videoscopy. Following surgical closure of VSD, the symptoms resolved. The third patient was born full term and presented at 6 weeks of age with a hoarse voice, weak cry, increased work of breathing, failure to thrive and a PSM. After surgical closure of the VSD symptoms improved but ongoing decreased left vocal cord movement was seen on videoscopy at 12 weeks of age (7).

As discussed above clinical evaluation may suggest the likely etiology. In the current case the early onset of stridor, weak cry in addition to change in character of voice suggest a diagnosis other than laryngomalacia. Considering the presence of a cardiac defect, it was the likely cause of stridor. Treatment of unilateral palsy depends on severity of symptoms and associated underlying defects. Most cases of idiopathic unilateral VCP resolve over 6-12 months without surgical intervention. Treatment of underlying cardiac defect may results in an improvement in stridor.

Conclusion

When a patient presents with a very early onset of stridor, change of voice character and volume, a diagnosis other than laryngomalacia should be considered. Detailed history and examination may suggest the likely underlying cause of stridor.

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Quiz Answer

Fibrosarcoma of Larynx - completely obliterating upper airway. Emergency airway management should be planned in the operating room.