Case Report

Indian Journal of Emergency Medicine
Volume 10 Number 2, April - June 2024
DOI: http://dx.doi.org/10.21088/ijem.2395.311X.10224.5

Stridor in Er!!!

Ashish Nair[1], Athira C.N.[2], Jerry Mathew[3]

How to cite this article:
Ashish Nair, Athira C.N., Jerry Mathew. Stridor in Er!!!. Ind J Emerg Med. 2024;10(2):69-74.

Abstract

5-month child presenting with high grade fever with cough and noisy breathing to the EMD. On evaluation the baby appears to be in stridor and respiratory distress. Urgent chest X-ray AP view (Fig. 1) showed haziness in the right mid and lower zone. Airway was secured followed by CECT thorax suggestive of long segment tracheomalacia with aberrant innominate artery compression. Confirmation was done with a flexible bronchoscope followed by a suctioning to maintain luminal patency. In this article, we report our experience of Tracheomalacia with aspiration pneumonia in paediatric age group.

Keywords: Stridor; Tracheomalacia; Intrathoracic Airway; Brachiocephalic Artery; Upper Airway Obstruction; Flexible Bronchoscope.

INTRODUCTION

Stridor is a sign of upper airway obstruction. In the paediatric age group, abnormal passage of air produces stridor. The cause of stridor can be located anywhere in extra-thoracic airway (nose, pharynx, larynx, and trachea) or the intrathoracic airway (tracheobronchial tree). Stridor may be caused by inflammation/infection or inhalation of foreign body (acute) or chronic. It may be congenital or acquired. Laryngomalacia is considered as the most common cause of chronicity in stridor in children younger than two years. It has a male-to-female (M:F) ratio of 2:1.[1] This may be due to an intrinsic defect or delay in maturation of supporting structures of the larynx. The airway may get obstructed partially during inspiration due to prolapse of the flaccid epiglottis, arytenoids and aryepiglottic folds. Inspiratory stridor usually worsens when the child is lies in a supine position or when crying or agitated, or when an upper respiratory tract infection occurs.[2] The role of a Critical care physician when faced with an infant with noisy breathing is: (1) to evaluate the severity of respiratory compromise and the requirement for immediate intervention for prevention of respiratory failure; (2) to evaluate the history along with clinical examination to assess whether a significant lesion is suspected and, if so, timely referral of the child to an ENT surgeon for an upper and lower airway bronchoscopy; (3) to evaluate the management strategies of the underlying lesion and to collaborate with other experts for follow-up and further management of the child.

Author's Affiliation: [1,3]Senior Resident, Department of Critical Care Medicine, [2]Resident, Department of Anaesthesiology, Bharti Vidyapeeth Hospital, Pune 411043, Maharashtra, India.

Corresponding Author: Athira C.N., Resident, Department of Anaesthesiology, Bharti Vidyapeeth Hospital, Pune 411043, Maharashtra, India.

E-mail: drathiracn@gmail.com

Received on: 19-02-2024

Accepted on: 22-05-2024

CASEREPORT

A 5-month-old male child was brought to the ER with history of high-grade fever which was continuous in nature associated with chills and rigor since 4-5 days. H/o running nose and cough in the past 1 week. Patient was in respiratory distress with noisy breathing. The mother gives a history of recent history of hospital admission with 15 days of ventilator support and 28 days hospital admission. Patient was diagnosed with long segment tracheomalacia with h/o aspiration pneumonia and respiratory failure. Patient had been referred to tertiary center for further evaluation.

Causes of Stridor in Children According to Site of Obstruction

Nose and pharyank

Choanal atresia

Lingual thyroid or thyroglossal cyst

Macroglossia

Microganathia

Hypertrophic tonsils/adenoids

Retropharyngeal or peritonsillar abscasess

Laryns

Laryngomalacia

Laryngeal web, cyst or laryngocele

Intubation

Foreign body

Cystic hygrome

Subglottic hemangioma

Laryngeal papiloma

Angioneurotic edema

Laryngospasm (hypocalcemic tetany)

Psychogenic striod

Trachea

Tracheomalacia

Table 1: Causes of Large Airway Compression

Cause	Examples	Notes
Primary Extreme prematurity	Often involves all of the tracheobronchial tree	Can be related to prolonged positive pressure ventilation Has a natural history of recovery but can take years
Syndromic	Noonan syndrome Chitayat syndrome[85] Down syndrome Ehlers Danlos syndrome CHARGE syndrome Skeletal dysplasias involving the cartilaginous structures	Length and degree of malacia can be very variable even within the same syndromic phenotype Clinical impact can vary from the trivial to severe
Idiopathic		
Secondary External large vessel compression	Double aortic arch Pulmonary artery sling Absent pulmonary valve Severe pulmonary artery hypertension	Malacia can persist at the site of compression even after the vessel has been moved away from the airway wall
Cardiac compression	Dilated cardiomyopathy Following heart transplant in infant	
Mediastinal mass	Teratoma Inflammatory myofibroblastic tumour Bronchogenic cyst	There is controversy as to whether a large normal thymus will ever compress the airway structure and cause malacia Current wisdom is No
Post-surgery	Tracheo-oesophageal fistula repair Following slide tracheoplasty for long segment complete rings Following removal of tracheostomy Fetal balloon insertion for congenital diaphragmatic hernia	TOF repair will frequently leave a defined area of residual tracheal malacia often at the level of the fistula Occasionally removal of the complete tracheal rings will unmask distal bronchomalcia
Infection/inflammatory disorders	Following tracheitis Stevens-Johnson syndrome	Severe staphylococcal tracheitis or bronchitis can leave long lasting malacia

Table Cont...

Injury	Button battery ingestion injury Delayed removal of inhaled foreign body with infection	Erosion of battery from oesophagus into the trachea can be rapid and leave lasting cartilaginous destruction with an acquired tracheo-oesophageal fistula
Skeletal deformity compression	Severe pectus excavatum - especially when in association with a scoliosis Scoliosis	These deformations will often impact on a bronchus - with the left main bronchus draped over the deviated spine and trapped between pulmonary artery and descending aorta

Bacterial tracheitis

On Examination

A 4 kg child in respiratory distress with HR-155/' RR-50/' Temp-103F, CFT-<3sec, dehydration present. Saturation on room air was 94-95% On auscultation B/lcrepitations were heard with coarse conductive sounds, no mumurs heard. Active alert child but lethargic child.

Intervention

Immediately taken on O2 support via nasal prongs (target SpO2->95%), N/G secured.

ABG was suggestive of respiratory acidosis with compensatory metabolic alkalosis.

IV fluids ISO-P @15cc/hr, started on Elores 150mg iv bd, Inj. PCM and Meftal for fever control, nebulization with Levolin (0.31 mg q4hrly) and Budecort (0.5mg q12hrly). Later colistin nebulisations was added after the culture reports were available.

Urgent chest X-ray AP view (Fig. 2) showed haziness in the right lower and mid zone. Lateral view could not be taken.

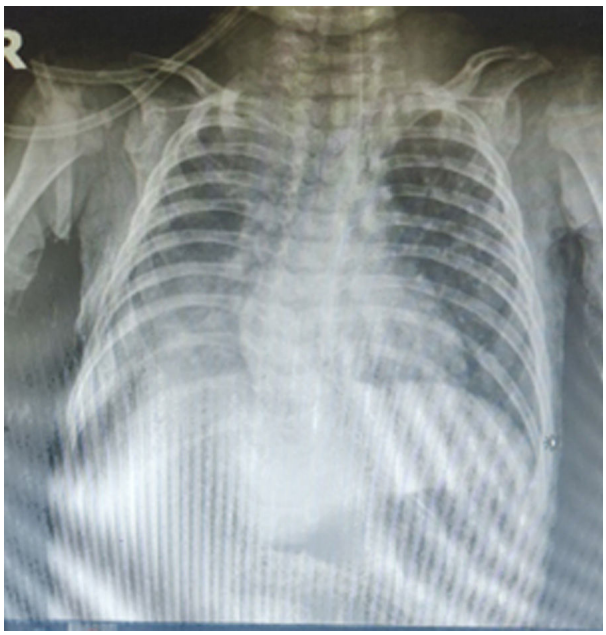

Fig. 2: Chest X-Ray Ap

Airway was secured with 3.5no. ETT and patient was shifted for urgent CECT thorax.

CECT thorax suggestive of displacement of trachea towards the right side. Aberrant origin of the innominate artery (brachiocephalic trunk) on the left side of the Aortic Arch causing mild anterior indentation/ extrinsic compression on the trachea at trachea at the level of the thoracic inlet s/o possible innominate artery compression of the trachea. Multiple resolving patchy areas of air space consolidation with surrounding groung glass opacification in the postero basal segments of bilateral lower lobes-possible aspiration pneumonitis.

Bronchoscopy was done using paediatric bronchoscope suggestive of long segment tracheomalacia starting just above carina.

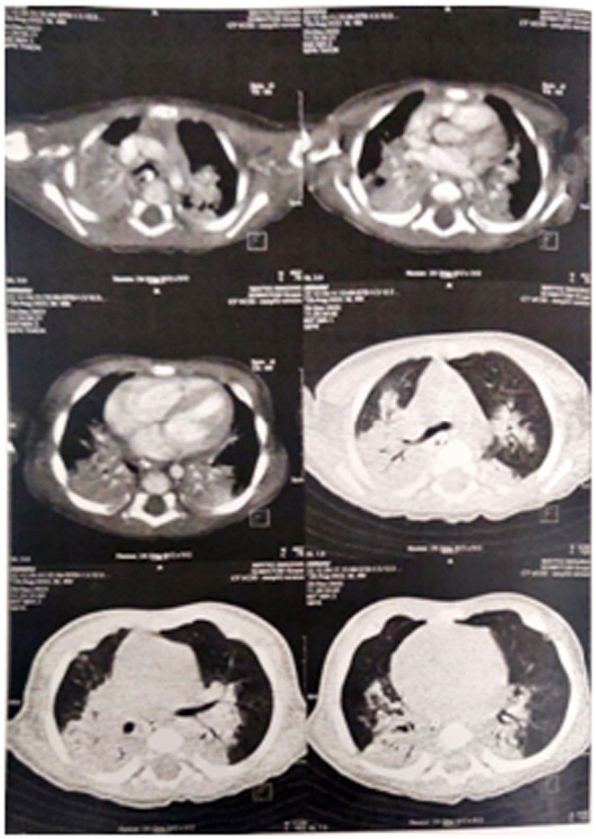

Fig. 3: CECT thorax (contrast)

Table 2: Symptoms

Symptom	Reported frequency in TBM
Cough	18-100%
Dyspnoea	59%
Wheeze	20-49%
Decreased exercise tolerance	25-35%
Symptoms of gastro-oesophageal reflux	26-53%
Apnoeic events including near death attacks	15-82%
Recurrent Infections	8-59%
Stridor	3-100%
Failure to thrive	11-24%

Challenges

- Aspiration pneumonia
- Paediatric airway emergency
- Choice of treatment of Tracheomalacia

Procedure

After adequate preparation of patient and checking bronchoscope. It is mandatory to keep one size larger and one size smaller than the estimated one. A tracheostomy set is always kept ready for emergency purpose.

Flexible Bronchoscopy was done under sedation (Fig. 4). Pooled secretions were removed and samples for staining and cultures were sent. Suctioning of intraluminal secretion was before the end of the procedureperformed until clear and luminal patency confirmed.

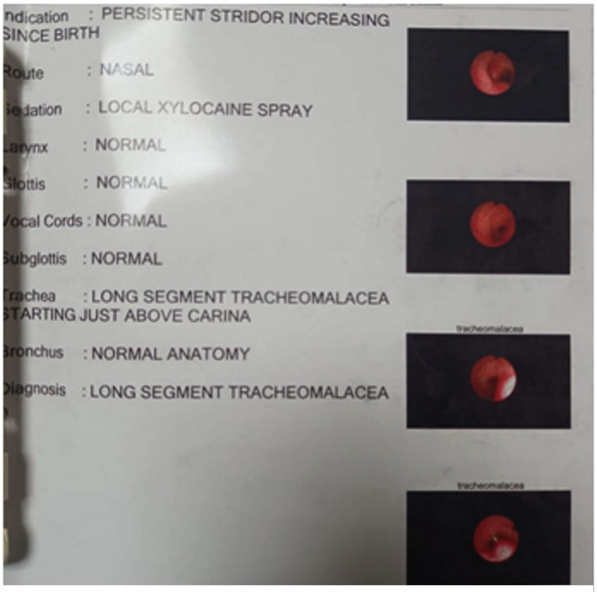

Fig. 4: Bronchoscope

DISCUSSION

Stridor is caused by abnormal air passage during breathing and often considered as the most prominent sign suggestive of upper airway obstruction. It is usually heardin the inspiratory phase (typically due to supraglottic or glottic obstruction) but may also occur in the expiratory phase (arising from obstruction at or below glottic level and/or due to severe upper airway obstruction). Stridor due to congenital anomalies may present from birth or may develop within days, weeks or months.

Malacia derived from the Greek word lakajὸ (malakos), means softness. Tracheomalacia (TM) is a condition where the walls of the trachea andbronchi develop abnormal flaccidity leading to collapse of the airway during expiration. Malacia can be confined either to the trachea – tracheomalacia (TM), or bronchi – bronchomalacia (BM), but may also involve the entire airway. Most authors report a male: female (M: F) ratio of about 2:1 in TBM.[4] The loss of airway wall rigidity is most prominent with external compressing forces as seen during intra-thoracic expiratory effort and is mostly associated with a softening of the cartilaginous architecture of the airwayrings or may also present with the congenital absence of supporting rings.[3]

The presenting symptoms of airway obstruction present with varying severity. Some children present with stridor, cough or recurrent chest infections while some are ventilator dependent. Signs of tracheomalacia include cyanosis, a prolonged expiratory phase, stridor (which may be bi-phasic) respiratory distress and signs of infection. Sternal or intercostal retraction may be apparent when the condition is severe. Respiratory distress usuallymanifests as grunting, open mouth breathing and head retraction. The Common presenting symptoms are summarised in Table 2 which highlights the highly variable nature of this reporting.[3]

Chest radiograph can show signs of pneumonia/ pneumonitis. The lateral neck radiograph is usually taken with extension of the neck and during inspiratory phase so that the pharyngeal soft tissues are not confusedwith a retropharyngeal mass. Computed tomographic (CT) scan and magnetic resonance imaging (MRI) may be obtained to visualize the anatomy of the airway and the surrounding soft tissue structures, including any evidence of vascular compression or anatomical compression.

Direct examination of the airway is often necessary to confirm the diagnosis and is essential in evaluation of children with persistent stridor. Flexible fibreoptic bronchoscopy is commonly used in the evaluation of airways in children. However, use of rigid bronchoscopy performed under general anaesthesia (GA) gives a better view of the airway, especially the subglottic view. Rigid bronchoscopy also helps in tissue biopsy and removal of foreign bodies.

A complete blood count is useful if an infection is suspected. Determination of the ESR is helpful in assessing for the presence of an infection. Depending on the degree of respiratory distress, ABG determination may be necessary to assess the degree of hypoxia and ventilatory status of the patient. An electrocardiogram (ECG) and echocardiogram (2D-Echo) are indicated if significant heart murmurs are present or when structural heart disease is suspected in patients.

Treatment of stridor should be directed to the underlying cause. The airway should be secured immediately in children with severe respiratory distress or actual airway obstruction. This can be done via endotracheal intubation. After adequate ventilation, which is achieved by intubation, tracheostomy may be performed if deemed necessary. Supportive measures like oxygen, humidified air, intravenous fluids, suction and aerosol treatments with steroids and beta-adrenergic drugs may be used.

Innominate Artery Compression Syndrome

Innominate artery compression syndrome, also known as ***brachiocephalic artery compression syndrome***, is a rare cause of tracheal stenosis which is seen in the paediatric population. Abnormal compression of the anterior aspect of the trachea occurs as the brachiocephalic artery crosses it. Children with age less than 3 years with stridor, breathing difficulties, asthma, apnoeic spells, and recurrent respiratory infections should be suspected of this conditio.[6] The brachiocephalic artery usually originates from the left of the trachea in normal children without obstructive symptoms or associated tracheal compression.[5] The artery subsequently impinges/compresses the trachea as it traverses anteriorly at the level of the thoracic inlet.[5]

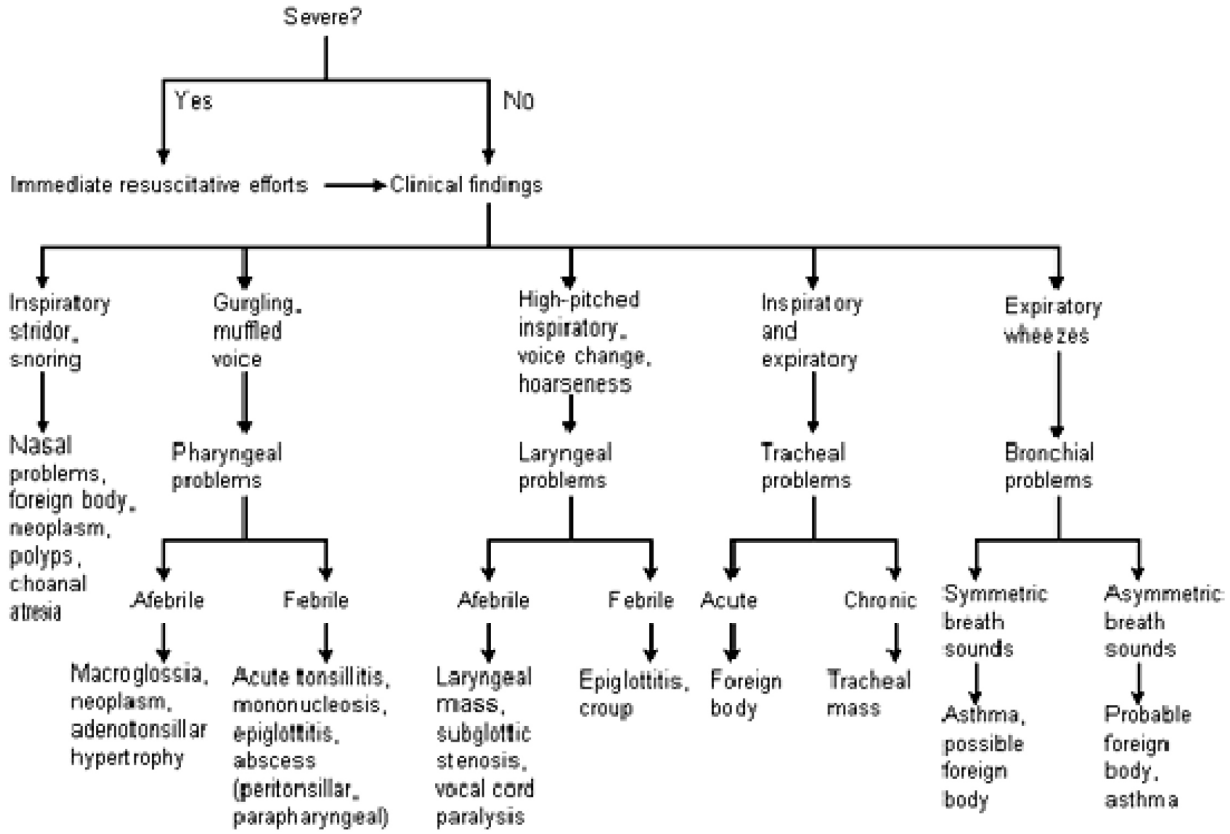

Fig. 5: Algorithm for Stridor

Treatment Options for Malacia in Children

The treatment plan will be tailored to the individual child, the underlying pathology, the skills and experience of the team and with a holistic approach to other pathologies and clinical concerns. ***Tracheostomy*** is a feasible option where the area of narrowing is confined to a section of the trachea and will be splinted by the tracheostomy shaft and buy time for natural improvement or surgical repair. ***Aortopexy***–Hitching up the anterior vascular anatomy and attaching it to the back of the sternum (with removal of the thymus) can support the airway. This procedure ideally suits a limited section of malacia following repair of a tracheo-oesophageal fistula. ***Sleeve Resection*** is done less commonly but can be considered where there is a congenital absence of one or two rings leading to an area of significant malacia. Placement of ***External Splinting*** involves a technique where the malacic airway is suspended by sutures to a rigid prosthesis placed around either the affected bronchus or the trachea. Insertion of an ***Internal Stent***. In Candidates for stent insertion, the main indication for stenting is failure of medical and surgical treatments with persistent severe symptoms. In practice, stenting is most often suggested for children who are ventilator dependent and has to date often been considered an intervention of last resort.

CONCLUSION

Tracheobronchomalacia (TBM) is associated with significant problems foraffected individuals, their families, and the clinical team. In the recent literature, the incidence of TBM is probably increasing due to improved survival of premature infants and patients with congenital heart disease and OA/TOF. TBM has been associated with increased morbidity and longer stays on intensive care following surgery for congenital heart disease. Each case is unique and often complex and patients are bestmanaged by an interdisciplinary team. A stepwise approach to treatment is usually adopted, starting with medical management before consideration of tracheostomy, other surgical options or stent insertion.

Directions for Future Research

- A better understanding of the underlying pathogenesis for the malacic process to guide the development of growth factors for cartilaginous strength and integrity.
- A paediatric classification for degrees of malacia to assist with further research and management.
- The use of 3D printing to design and print bespoke stents for the growing child.
- The use of bioabsorbable materials for both internal stents and external splints that will have a predetermined absorbable period for use in transient conditions and growing airways.

REFERENCES

1. Simon NP. Evaluation and management of stridor in the newborn. ClinPediatr. 1991; 30:211-6.
2. Clough J. Managing stridor in children. Practitioner. 1995;239(1557):724-8
3. Wallis C, McLaren CA. Tracheobronchial stenting for airway malacia. Paediatric Respiratory Reviews. 2018 Jun 1;27:48-59.
4. Masters IB, Chang AB, Patterson L, Wainwright C, Buntain H, Dean BW, Francis PW. Series of laryngomalacia, tracheomalacia, and bronchomalacia disorders and their associations with other conditions in children. Pediatric pulmonology. 2002 Sep;34(3):189-95.
5. Koivikko A, Puhakka H, Vilkki P. Innominate Artery Compression Syndrome. ORL. 1976; 38(3):187-92. doi:10.1159/000275274 - Pubmed
6. Fawcett SL, Gomez AC, Hughes JA *et. al.* Anatomical variation in the position of the brachiocephalic trunk (innominate artery) with respect to the trachea: a computed tomography-based study and literature review of Innominate Artery Compression Syndrome. Clin Anat. 2009;23 (1): 61-9. doi:10.1002/ca. 20884 - Pubmed citation.