ARTICLE ORIGINAL / ORIGINAL ARTICLE

URGENT UPPER AIRWAY OBSTRUCTION IN PEDIATRIC POPULATION
A 13-Year Experience in a Tertiary Care Center

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ABSTRACT ● The objective of this study is to report the main causes and presentations of urgent upper airway obstruction in children, and to analyze factors delaying appropriate diagnosis and management. All cases of pediatric urgent upper airway obstruction between January 2003 and June 2016 were studied. Demographic data, clinical characteristics, and management were analyzed. Twenty-two patients were included (mean age 13.3 months). Most frequent etiologies were vocal cord paralysis (18%) and sub-glottic foreign bodies (18%). In five cases, initially suspected diagnosis was different than final diagnosis. It is important not to miss the correct diagnosis and to manage severe cases in adequately equipped tertiary care centers.

Keywords : upper airway obstruction; children; urgent; emergency management

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RÉSUMÉ ● L'objectif de cette étude est de rapporter les principales causes et présentations cliniques des obstructions respiratoires hautes chez les enfants afin d'analyser les facteurs retardant le diagnostic et la prise en charge. Tous les cas pédiatriques d'urgence respiratoire haute admis entre janvier 2003 et juin 2016 ont été revus. Les données démographiques, les caractéristiques cliniques et les différents traitements ont été analysés. Vingt-deux patients ont été inclus (âge moyen : 13,3 mois). Les étiologies les plus fréquentes étaient la paralysie des cordes vocales (18%) et les corps étrangers sous-glottiques (18%). Dans cinq cas, le diagnostic suspecté initialement était différent du diagnostic définitif. Il est important de vérifier le bon diagnostic et de traiter ces cas d'urgence le plus rapidement possible dans un centre de soins tertiaires adéquat.

Mots-clés : obstruction respiratoire haute; pédiatrique; urgence respiratoire

INTRODUCTION

Upper airway obstruction is a frequent cause of consultation in the pediatric population that needs immediate treatment. Diagnosis is not always evident on history and physical examination, making management of these cases challenging.

The most important clue towards upper airway obstruction is the presence of stridor along with dyspnea. In fact, narrowing of the airway at any level from the nose/oral cavity to the distal bronchi causes increase in velocity to allow the same volume of air to pass [1]. When velocity increases, the Venturi effect causes further narrowing and turbulent flow that results in rhythmic vibrations of the airway and production of the stridor.

The upper airway in children is much smaller and narrower than that of adults. Hence small changes in the radius at this level account for large modifications in the cross sectional area due to the exponential relationship between them (area = π*radius2). This puts the pediatric population at greater risk of severe upper airway obstruction from relatively otherwise benign conditions [2].

Therefore, underestimation or misdiagnosis of upper airway obstruction can put the child in a life-threatening situation [3].

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Upper airway obstruction can be categorized into supraglottic, glottic or subglottic depending on characteristics of the stridor and accompanying symptoms and signs. It is therefore essential to emphasize the importance of creating well-defined diagnostic work-up plans and management algorithms for severe cases of upper airway obstruction in children.

The present study is a review of all cases of severe upper airway obstruction that were managed in our institution from January 2003 to June 2016. In addition to reporting the major causes and clinical presentations of upper airway obstruction in this series, the main objective is to analyze the factors delaying appropriate diagnosis when present and to critically review the treatment plan.

MATERIALS AND METHODS

All pediatric upper airway obstruction cases treated in Hôtel-Dieu de France Hospital, Beirut, Lebanon, from January 2003 to June 2016, were retrospectively reviewed. Inclusion criteria were:

1) Patients presenting to the emergency department with acute upper airway obstruction
2) Age < 16 years

Patients presenting with lower, intrathoracic airway obstruction (i.e. bronchial foreign bodies, lower respiratory tract infections, lower respiratory tract congenital malformations) were excluded from the study.

Data regarding clinical presentation, etiology, diag-

nostic tests, treatment procedures, and follow-up were retrieved from hospital charts. A descriptive statistical analysis of the data was done using SPSS, and a review of the English and French literature regarding upper respiratory obstruction in children was performed. Finally, a plan for emergent management and treatment of upper airway obstruction in children was proposed.

RESULTS

Twenty-two patients were included in the study and data concerning demographic information, clinical presentation, work-up, diagnosis, treatment and follow-up was extracted from their charts. This data is presented in Table I.

Mean age was 13.3 months and male patients were more frequent with a sex ratio of 2:1. The most frequent etiologies were vocal cord dysmotility disorders (18%) and sub-glottic foreign bodies (18%) followed by piriform aperture stenosis and bilateral choanal atresia, 13.5% of cases each. Other etiologies were: laryngomalacia, glottic web, sub-glottic stenosis, sub-glottic hemangioma, tracheomalacia, tracheal stenosis, tracheal granuloma and Pierre-Robin sequence. In five cases (23%) the initial suspected diagnosis was different than the final diagnosis; one case of bilateral vocal cord paralysis was initially diagnosed as laryngomalacia, a case of sub-glottic stenosis was initially treated as asthma, a case of sub-glottic foreign body was first diagnosed as a sub-glottic stenosis, and two cases of piriform aperture stenosis were first diagnosed as choanal atresia.

TABLE I SUMMARY OF 22 CASES OF CHILDREN WITH URGENT UPPER AIRWAY OBSTRUCTION

Case	Age (mos)	Sex	Presentation	Final Diagnosis	Confirmatory Investigations	Treatment	Follow-up	Remarks
1	2	M	Inspiratory stridor; Subxyphoid retraction (SXR) & Suprasternal retraction (SSR)	Laryngomalacia	Flexible naso-pharyngo-laryngoscopy (FL)	Intubation & Steroids + Partial epiglottectomy	N/A	Prematurity
2	1.3	F	Inspiratory stridor with dyspnea & Respiratory distress	Sub-glottic hemangioma	Rigid bronchoscopy (RB)	Intubation + Steroids + β-blockers	N/A	
3	3	M	Inspiratory stridor SSR & Cyanosis	Tracheal granuloma	RB	Surgical removal	N/A	Neonatal pneumonia with intubation
4	1.5	M	Inspiratory stridor SXR & SSR	Congenital bilateral vocal cord paralysis	FL	Tracheotomy	Decanulated at age 2 years	Intubation for 2 weeks at birth Cosanguin parents
5	19	M	Inspriatory stridor with aphonia since birth. Severe respiratory distress during episode of URI	Glottic web	FL	Intubation + Surgery after stabilization	Extubated at day 3 post-op	
6	0	M	Intubation at birth for severe respiratory distress Stridor at extubation on day 3	Bilateral vocal cord paralysis	FL	Supportive treatment	Died at 42 days	Polymalformative syndrome Cosanguin parents
7	0	F	Inspiratory stridor at birth followed by severe respiratory distress at day 8	Bilateral vocal cord paralysis	FL	Intubation	Tracheotomy at 7 weeks	Polymalformative syndrome Cosanguin parents
8	0	F	Stridor and respiratory distress since birth SSR	Bilateral vocal cord paralysis	FL	Tracheotomy	Decanulation at 2 years 70% improvement of vocal cord mobility	
9	9	M	Biphasic stridor of 4 months duration On admission severe respiratory distress following URI	Tracheomalacia	RB. Thoracic angioscan: no extrinsic compression	Supportive treatment	N/A	
10	168	M	Inspiratory stridor & SSR since age 3 years Respiratory distress following URI	Sub-glottic stenosis	Thoracic CT scan & RB	Tracheotomy followed by laryngoplasty	Decanulation at 3 months post-op	

11	18	M	Inspiratory stridor, respiratory distress after a penetration syndrome	Sub-glottic foreign body	RB	Extraction	N/A	
12	12	M	Inspiratory stridor SSR & Respiratory distress	Sub-glottic foreign body	RB	Extraction	24h ICU	
13	0	F	Respiratory distress at birth Difficulty in introducing NG tube 6	Piriform aperture stenosis	Sinus CT scan	Drilling	N/A	
14	36	M	Inspiratory stridor. Respiratory distress. Polypnea. SSR. Penetration syndrome	Sub-glottic foreign body	RB	Extraction	N/A	
15	9	M	Dyspnea since birth Severe respiratory distress Retromandibular, suprasternal, subxyphoid and intercostal retraction	Piriform aperture stenosis	Sinus CT scan	Drilling	N/A	Patient had surgery for piriform aperture stenosis at 4 months of age in another hospital with recurrence. Mega-incisive
16	0	M	Severe respiratory distress at birth Intercostal retraction & SXR	Tracheal stenosis	Thoracic CT scan & RB	Intubation followed by tracheoplasty	N/A	
17	0	F	Cyanosis Failure to pass NG tube bilateraly Retromandibular retraction	Choanal atresia	Sinus CT scan	Endoscopic surgery at birth	Removal of calibration tubes on day 5 followed by respiratory distress Flexible fibroscopy showed granuloma Endoscopic removal of granuloma	
18	0	M	Cyanosis Failure to pass NG tube bilateraly Retromandibular retraction	Choanal atresia	Sinus CT scan	Endoscopic surgery at birth	Respiratory distress following removal of calibration tubes Repermeabilization by endoscopic surgery	
19	0	F	Respiratory distress at birth Nasal intubation impossible Oral intubation	Choanal atresia	Sinus CT scan & FL	Endoscopic surgery at 7 days of life	N/A	
20	1	M	Respiratory distress at birth. SSR. SXR. Retromandibular retraction. Failure to pass NG tube bilaterally	Piriform aperture stenosis	FL	Drilling	N/A	Mega-incisive & absent superior lip frenula
21	12	M	Inspiratory stridor. SSR. Cyanosis	Sub-glottic foreign body	FL	RB & Foreign body extraction	N/A	
22	2	F	Inspiratory stridor since birth Respiratory distress following inhalation pneumonia	Pierre-Robin sequence	Chest X ray Cardiomegaly FL: glossoptose & cleft palate	Tracheotomy	N/A	Prematurity

mos: months **M**: male **F**: female **SXR**: subxyphoid retraction **SSR**: suprasternal retraction **URI**: upper respiratory infection
FL: flexible naso-pharyngo-laryngoscopy **RB**: rigid bronchoscopy **CT**: computed tomography **N/A**: not available

Flexible laryngoscopy and rigid bronchoscopy were the main confirmatory tests in almost all cases. Five cases (23%) needed emergent intubation, while four (18%) needed tracheotomy. Surgery (laryngoplasty or tracheoplasty) was the treatment in 11 cases (50%) while 2 cases (9%) were managed with supportive therapy. During the follow-up period, only one death was reported.

The main variables are summarized in Table II.

DISCUSSION

Upper airway obstruction in children may be overlooked but it can also cause severe respiratory distress. An early diagnosis is very important and helps avoiding significant complications and sometimes death. Obstructive symptoms range from mild stridor to severe dyspnea with episodes of apnea, suprasternal and subcostal retractions, and tachypnea. In addition, hypoxia leads to cyanosis while hypercapnea leads to sudation and drowsiness, all of which are very distressing for parents. In our series the most frequent presentation was inspiratory stridor and suprasternal retraction, which was in accordance with the previously published data [2].

TABLE II
DEFINITIVE DIAGNOSIS, CONFIRMATORY TESTS AND TREATMENT OF 22 CHILDREN WITH URGENT UPPER AIRWAY OBSTRUCTION

Variables	N	%
Sex Ratio (Male/Female)	15/7	-
Mean Age (Months)	13.35	-
Definitive Diagnosis		
Piriform aperture stenosis	3	*13.64*
Bilateral choanal atresia	3	*13.64*
Pierre-Robin sequence	1	*4.54*
Laryngomalacia	1	*4.54*
Glottic web	1	*4.54*
Bilateral vocal cords paralysis	4	*18.18*
Sub-glottic foreign body	4	*18.18*
Sub-glottic stenosis	1	*4.54*
Sub-glottic hemangioma	1	*4.54*
Tracheomalacia	1	*4.54*
Tracheal stenosis	1	*4.54*
Tracheal granuloma	1	*4.54*
Wrong Suspected Diagnosis	5	*22.73*
Confirmatory Diagnostic Tests		
FL	10	
RB	8	
Sinus CT	5	
Thoracic CT	2	
Thoracic angioscan	1	
Chest X-ray	1	
Immediate Management		
Intubation	5	*22.73*
Tracheotomy	4	*18.18*
Surgery	11	*50.00*
Supportive	2	*9.09*

FL: flexible naso-pharyngo-laryngoscopy **RB**: rigid bronchoscopy
CT: computed tomography

Level of obstruction affects the clinical presentation in upper airway obstruction in children. In fact, retractions, stridor, voice, and feeding are important clues [1]:

- Nasopharyngeal and oropharyngeal obstruction typically produces stertorous noises.
- Supraglottic airway is collapsible which is responsible for inspiratory Ventouri effect resulting in a high-pitched inspiratory stridor, suprasternal retractions, muffled voice and possible feeding problems.
- Glottic and subglottic airways are relatively rigid, thus strongly affected by absolute cross-sectional area. Obstruction at this level produces an inspiratory or biphasic intermediate pitched stridor, supraclavicular and intercostal retractions. On another hand, glottic obstruction is characterized by dysphonia versus a barking cough in subglottic obstruction.

Initial assessment in an emergency setting should be focused on vital signs and on patient's stability. The general condition of the child and physical exam findings are key elements to assess the degree of severity. In upper airway obstruction pulse oximetry can be falsely reassuring and should not be relied upon to establish treatment plan [2, 3]. In fact, oxygen saturation will not drop until late in the process once respiratory fatigue has already occurred. Hence, the most critical aspect of the initial evaluation of a child with upper airway obstruction is to look for signs of respiratory fatigue and eminent respiratory failure such as: cyanosis, diaphoresis, diminished retractions, diminished respiratory effort, gasping. When eminent respiratory distress is suspected, securing the child's airway becomes the top priority [4].

In an emergency room setting, mobilizing specialty support (critical care team, otolaryngology) should be done in parallel to establishing the airway. In children where difficult intubation is suspected (history of previous difficult or traumatic intubation, craniofacial deformities, syndromic child, etc.) fast action and anticipation of the next step in case of failure are always warranted [5].

After initial history and physical examination, flexible endoscopy is part of the initial assessment whenever an upper airway obstruction is suspected. However, in unstable children or those with severe presentation this should be done with high caution due to the risk of laryngospasm and deterioration during flexible endoscopy examination. In extreme cases, this should be done in the operating room with the child still awake and an emergency intubation cart ready. Rigid bronchoscopy under general anesthesia is complementary to flexible endoscopy and is necessary for evaluation of the airway distal to the vocal cords [3].

An algorithm for management of urgent upper airway obstruction in children is proposed in Figure 1. This algorithm was based on our institution's experience as well as data from other published series [4,5].

In the published literature the most frequent causes of

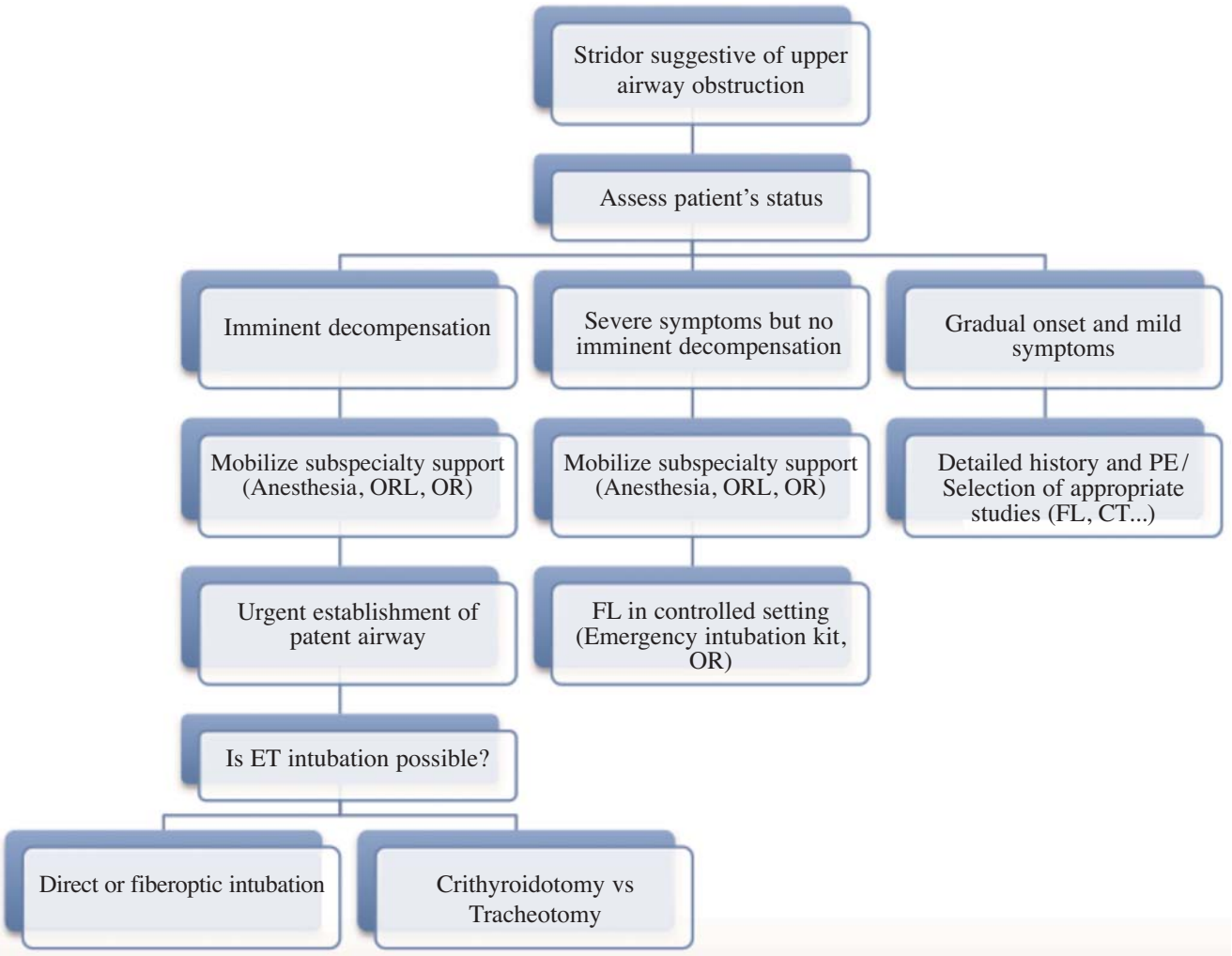

Figure 1. Algorithm for management of children with urgent upper airway obstruction

ORL: otolaryngology **OR**: operating room **FL**: flexible naso-pharyngo-laryngoscopy **CT**: computed tomography **ET**: endotracheal

stridor in children are laryngomalacia accounting for 60% of cases, followed by vocal cord dysmotility disorders, subglottic stenosis, sub-glottic foreign bodies and tracheomalacia which all together account for 30% of cases [1].

In our series, which was restricted to children presenting with urgent upper airway obstruction, the most frequent etiologies were vocal cord dysmotility disorders and sub-glottic foreign bodies. This could be due to the fact that the majority of cases of laryngomalacia present with a stridor but are not severe enough to cause eminent upper airway obstruction [6]. In fact, the most frequent cause of congenital stridor manifesting within the perinatal period is congenital laryngomalacia with a reported incidence of 35-75%. Eighty percent of infants will have mild to moderate laryngomalacia with minor inspiratory stridor and are managed conservatively, while the remaining 20% will have severe laryngomalacia and will require surgical intervention [6-9].

Vocal cord paralysis is the second most common congenital laryngeal anomaly. Bilateral forms are usually idiopathic but can in certain cases be secondary to Arnold-Chiari or other central nervous system anomalies. Unilateral paralyses are not always symptomatic whereas bilateral forms can be associated with a high-pitched inspiratory stridor, near normal phonation and sometimes airway emergencies. Diagnosis is done via flexible fiberoptic nasopharyngoscopy. Treatment includes a tracheotomy kept in place for a minimum of 18-24 months, followed by vocal cord lateralization procedures, unilateral arytenoidectomy, or laser cordotomy when decanulation is unsuccessful [10,11].

Regarding non-congenital causes of upper airway obstruction in children it is important to look for inciting traumatic events such as endotracheal intubation. Indeed, iatrogenic stricture and granulation tissue formation, after endotracheal intubation, are the most common etiologies of subglottic stenosis [12,13].

Delayed diagnosis is an important cause of failure in

managing these urgent cases resulting in irreversible consequences and even death. This situation can be due to undiagnosed or misdiagnosed cases. In our series, out of the five cases in which the initial working diagnosis was different than the final diagnosis, four were referred from other hospitals. This is a very important argument in favor of recommendations that incite to manage and treat urgent upper airway obstructions in children in a tertiary care center equipped with all the necessary resources and infrastructures [2,14,15].

CONCLUSION

Urgent upper airway obstruction in children is a life-threatening condition that should be managed promptly but also with extreme caution. Physicians must make sure not to miss the correct diagnosis as this may cause irrecoverable delays in treating the patient and should also limit the use of invasive investigations in unsafe and non-prepared settings as this can result in severe deterioration and worsening of the patient that cannot be properly managed.

REFERENCES

1. Greinwald J, Cotton R. Pathophysiology of stridor and airway disease. In: Van De Water T, Staecker H, editors. Otolaryngology Basic Science and Clinical Review. New York, NY: Thieme Medical Publishers, 2006: 212-24.
2. Boudewyns A, Claes J, Van de Heyning P. Clinical practice: an approach to stridor in infants and children. Eur J Pediatr 2010 Feb; 169 (2): 135-41.
3. Hartnick CJ, Cotton RT. Stridor and Airway Obstruction. In: Bluestone C, Stool S, Alper C et al., editors. Pediatric Otolaryngology, Vol 2. 4th edition. Philadelphia, PA: Saunders; 2003: 1437-47.
4. Yataco JC, Mehta AC. Upper airway obstruction. In: S. Raoof, American College of Physicians, editors. Manual of Critical Care. New York: McGraw Hill Medical, 2009: 388-97.
5. Luten RC, Kisson N. The difficult pediatric airway. In: Walls RM, Murphy MF, Luten RC, et al, editors. Manual of Emergency Airway Management, 2nd ed. Williams & Wilkins: Lippincott, Philadelphia, 2004: 236.
6. Thompson DM. Abnormal sensorimotor integrative function of the larynx in congenital laryngomalacia: a new theory of etiology. Laryngoscope 2007; 117 (6 Pt 2, Suppl 114): 1-33.
7. Birch DA. Laryngeal stridor in infants and children: a study of 200 cases. J Laryngol Otol 1961; 75: 833-40.
8. Lewison M, Lira D. Apnea in the supine position as an alerting symptom of a tumor at the base of the tongue in small infants. J Pediatr 1965; 60: 1092-3.
9. Hsieh WS, Yang PH, Wong KS, Li HY, Wang ECR, Yeh TF. Vallecular cyst: an uncommon cause of stridor in newborn infants. Eur J Pediatr 2000; 159: 79-81.
10. Daniel S. The upper airway: Congenital malformations. Paediatric Respiratory Reviews 2006; 7: S260-S263.
11. Nicollas R, Triglia JM. The anterior laryngeal webs. Otolaryngol Clin North Am 2008, 41: 877-88.
12. Schroeder JW Jr, Holinger LD. Congenital laryngeal stenosis. Otolaryngol Clin North Am 2008; 41; 865-75.
13. Ho AS, Koltai PJ. Pediatric tracheal stenosis. Otolaryngol Clin North Am 2008; 41: 999-1021.
14. Wiatrak B. Congenital anomalies of the larynx and trachea. Otolaryngol Clin North Am 2000; 33: 91-110.
15. Evans KN, Sie KC, Hopper RA, Glass RP, Hing AV, Cunningham ML. Robin sequence: from diagnosis to development of an effective management plan. Pediatrics 2011 May; 127 (5): 936-48.