DOI: 10.4274/buchd.galenos.2021.87160
J Dr Behcet Uz Child Hosp 2022;12(1):101-106

Case Report

Rare Causes of Stridor: Not All Stridors are Croups

Stridorun Nadir Nedenleri; Tüm Stridorlar Krup Değildir

Serdar Al[1], Nevin Uzuner[1], Suna Asilsoy[1], Gizem Atakul[1], Özge Atay[1], Özge Kangallı[1], Büşra Acun[2], Yüksel Olgun[3], Taner Erdağ[3], Özkan Karaman[1]

[1]Dokuz Eylül University Faculty of Medicine, Department of Pediatric Allergy and Clinical Immunology, İzmir, Turkey
[2]Dokuz Eylül University Faculty of Medicine, Department of Pediatrics, İzmir, Turkey
[3]Dokuz Eylül University Faculty of Medicine, Department of Otorhinolaryngology, İzmir, Turkey

ABSTRACT

Stridor is a considerable finding, which arises from congenital or acquired causes, that might have a good prognosis or might be a life-threatening cause, and hence requires rapid assessment and intervention. Stridor is mostly considered as croup in pediatric emergency units. A multidisciplinary approach might be required for an accurate diagnosis. The patient's age, time and severity of initial symptoms, response to treatment, and coexistent findings assist in differential diagnosis. Detecting the underlying cause may be difficult and time consuming. In this study, we intended to present cases that we assessed regarding the etiological reasons presented with chronic stridor and to present remarkable characteristics of them in diagnosis and differential diagnosis, by considering the literature. Patients diagnosed with subglottic stenosis, subglottic hemangioma, laryngomalacia and laryngeal web were presented with distinctive history and clinical findings. Patients with intermittent respiratory problems coexisting with stridor can be diagnosed typically through an adequate medical history, a well-enough physical examination, and radiological methods. Flexible endoscopy provides a safe and complete examination of the children's airway.

Keywords: Stridor, dyspnea, infant, hemangioma, subglottic stenosis

Received: 18.03.2021
Accepted: 25.10.2021

Corresponding Author
Serdar Al MD
Dokuz Eylül University Faculty of Medicine, Department of Pediatric Allergy and Clinical Immunology, İzmir, Turkey
drserdaral@gmail.com
ORCID: 0000-0002-4846-6761

ÖZ

Stridor, doğuştan veya edinsel nedenlerden kaynaklanan, iyi bir prognoza sahip veya yaşamı tehdit edebilen, bu nedenle hızlı değerlendirme ve müdahale gerektiren önemli bir klinik bulgudur. Stridor, pediatrik acil servislerinde çoğunlukla krup, larenjit veya astım olarak değerlendirilir. Doğru bir teşhis için multidisipliner bir yaklaşım gerekebilir. Hastanın yaşı, başlangıç semptomlarının zamanı ve şiddeti, tedaviye yanıtı ve birlikte bulunan bulgular ayırıcı tanıya yardımcı olur. Altta yatan nedeni tespit etmek zor ve zaman alıcı olabilir. Bu çalışmada, kronik stridor ile ortaya çıkan etiyolojik nedenler açısından değerlendirdiğimiz olguları literatür eşliğinde sunmayı ve tanı ve ayırıcı tanıda dikkat çekici özelliklerini sunmayı amaçladık. Subglottik darlık, subglottik hemanjiyom, laringomalazi ve laringeal web tanısı alan hastalar, ayırt edici öykü ve klinik bulgular ile sunuldu. Stridor ile birlikte görülen aralıklı solunum problemleri olan hastalar tipik olarak ayrıntılı tıbbi öykü, dikkatli fizik muayene ve radyolojik yöntemlerle teşhis edilebilir. Fleksible endoskopi, çocukların hava yolunun güvenli ve eksiksiz bir şekilde incelenmesini sağlar.

Anahtar kelimeler: Stridor, solunum sıkıntısı, süt çocukluğu, hemanjiom, subglottik stenoz

Cite as: Al S, Uzuner N, Asilsoy S, Atakul G, Atay Ö, Kangallı Ö, Acun B, Olgun Y, Erdağ T, Karaman Ö. Rare Causes of Stridor: Not All Stridors are Croups. J Dr Behcet Uz Child Hosp. 2022;12(1):101-106

INTRODUCTION

Stridor may be the clinical manifestation of many problems related to the upper airway. The age of the patients, accompanying examination findings, the persistence of clinical findings and treatment responses should be the main markers for differential diagnosis.

Stridor is divided into two groups as acute and chronic based on its onset and duration. Stridor with an acute onset manifests within minutes, hours, or days (subacute) and usually progresses rapidly. Foreign body aspiration, anaphylaxis, bacterial tracheitis, epiglottitis, laryngotracheitis (croup), retropharyngeal abscess, peritonsillar abscess and airway burns may cause acute stridor.

Chronic stridor is typically caused by a congenital or acquired structural abnormality that leads to obstruction of the upper airway from inside or outside. Congenital causes include vocal cord paralysis, laryngomalacia, tracheomalacia, bronchogenic cyst, vascular ring, laryngeal malformations, infantile hemangiomas, and

subglottic stenosis. Stridor may develop due to acquired vocal cord dysfunction, respiratory papillomatosis, vocal cord paralysis, subglottic stenosis, laryngeal spasm due to hypocalcemia, and tumor compression [1-3].

In this study, we presented patients with stridor who were admitted to the hospital and misdiagnosed, such as croup, laryngitis or asthma. We aimed to emphasize their remarkable characteristics in the differential diagnosis in the light of literature.

Written informed consent from the patients' parents or guardians was obtained to publish cases in this study.

CASE REPORTS

Case 1

A 45-day-old male infant presented with respiratory distress. He had been intubated for 31 days following birth. On physical examination (PE), tachypnea, stridor and long expiration time were detected. No pathological finding was found at ear, nose, and throat (ENT) examination and direct laryngoscopy. Flexible endoscopy, performed under general anesthesia, revealed 95% stenosis in the subglottic region (Figure 1-A).

Case 2

An 8-month-old male patient was admitted to our clinic with difficulty sucking during an upper respiratory tract infection. Stridor and wheezing were noted when the patient was crying and excited. He was hospitalized for bronchiolitis three times. A hemangioma, which narrowed the airway passage by 50% in the subglottic region, was detected through flexible endoscopy during ENT examination (Figure 1-B).

Case 3

A 2-month-old female patient presented with respiratory distress and wheezing. Her wheezing and stridor increased when she cried since she was born, and she was more comfortable when she slept. On PE, tachypnea, stridor, intercostal and substernal retractions, and bilateral rales in lungs were observed. Laryngomalacia was assessed by flexible bronchoscopy in ENT examination (Figure 1-C).

Case 4

A 3-month-old male was referred for investigation of the etiology of stridor. Despite three direct laryngoscopic examinations and one bronchoscopic examination in another hospital, the exact cause of the stridor could not be elucidated.

On PE, wheezing and stridor were detected. No pathology was detected via direct laryngoscopy during the ENT examination. A hyperintense region was detected at the rima glottides level by computed tomography (CT) of the neck. A hemangioma covered with normal mucosa at the subglottic level was detected just below the vocal cords during the flexible laryngoscopic examination performed under general anesthesia (Figure 1-D).

Case 5

A 45-day-old male was intubated at birth due to wheezing and respiratory distress. The infant stayed in neonatal intensive care unit NICU for 30 days, 11 days of which he was intubated. On PE, he had mild stridor, wheezing and suprasternal retractions. Weight gain was at the lower threshold. In the ENT examination, which was performed under anesthesia, the glottic level was normal during flexible bronchoscopy; however, irregular web and stenosis were detected in the subglottic region (Figure 1-E).

Demographic and clinical data of all patients are given in Table 1.

DISCUSSION

Stridor arises from the turbulent airflow through the narrowed airway. Usually, it is heard during inspiration, and stridor that also occurs on expiration is termed biphasic stridor. It suggests severe, constant airway obstruction at the level of the subglottis, glottis, or upper trachea.

Stridor due to congenital anomalies may be present from birth or develop with time. In the event of narrowing at lower degrees, there might be no stridor during rest; however, due to increased activity (e.g., excitement or crying) and increased airflow rate, stridor may occur [4].

The feature of the sound heard provides further and very important insights. Hoarseness indicates an abnormality in the vocal cords. The stertor, another form of noisy breathing, evokes snoring, and essentially it is an inspiratory, low-pitched, secretory, and snoring-like sound. Pathologies that may lead to obstruction of the oropharynx, nasopharynx, and hypopharynx may usually cause stertor [5].

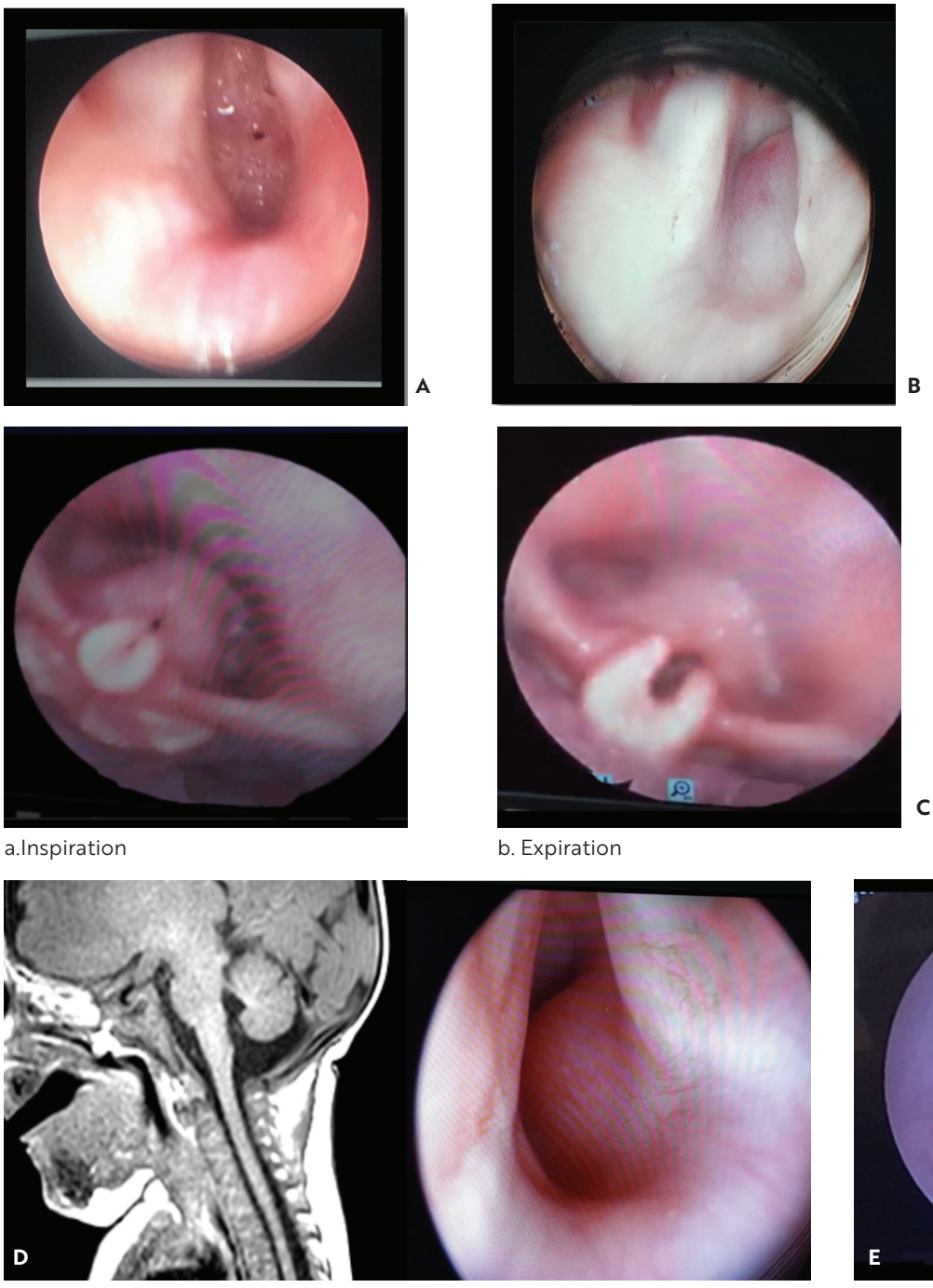

Figure 1. (A) Flexible bronchoscopy subglottic stenosis causing 95% stenosis, **(B)** Hemangioma, observed via flexible bronchoscopy, causing 50% stenosis. **(C)** Laryngomalacia, in the first picture (a), the epiglottis and arytenoids collapse during inspiration and close the glottic opening, causing inspiratory stridor. In the second picture (b), the glottis is open during expiration, and no abnormal sound is heard, **(D)** Hemangioma in neck tomography and bronchoscopy, **(E)** Subglottic stenosis and web in bronchoscopy

Table 1. Demographic and clinical findings of the patients

	Case 1	Case 2	Case 3	Case 4	Case 5
Diagnosis	Subglottic stenosis	Subglottic hemangioma	Laryngomalacia	Subglottic hemangioma	Laryngeal web, stenosis
Age/gender	45 days/male	8 months/male	2 months/female	2,5 months/male	45 days/male
Time of birth	On term	On term	On term	34 GW	On term
Birth weight (grams)	3,300	3,200	3,000	2,100	3,340
Onset time of the complaints	At birth	2.5 monts	Since birth, it has increased for the last 1 month	2.5 monts	Since birth
Complaints and symptoms	Tachypnea, stridor, longer expirium	Bronchiolitis attacks, snoring, stridor when excited or crying	Stridor when crying and lying down, snorre reflux	Stridor, wheezing	Wheezing, mild stridor, suprasternal retraction
Symptom severity	Severe	Mild, worsens with infection	Mild, worsens with infection	Moderate	Severe
Comorbidity	Mild MR, ASD	Absent	Physiological GER	Absent	Absent
Growth arrest	Slow weight gain	Absent	Absent	Absent	Slow weight gain
Duration of hospitalization	Constant	Short-term hospitalization for bronchiolitis 3 times	Absent	Neonatal pneumonia for 10 days in the neonatal period	30 days
Intubation	Intubated for 31 days	Absent	Absent	Absent	Intubated for 11 days
Treatment	Tracheostomy	Beta-blocker theraphy	Follow-up	Beta-blocker therapy	Tracheotomy
Health condition of the patient during follow-up	Tracheostomy cannula Acceration of growth	Partial recovery	Decrease in complaints	Partial recovery	Tracheostomy cannula Acceleration of growth

GW: Gestational Week, MR: Mitral regurgitation, ASD: Atrial septal defect, GER: Gastroesophageal reflux

Laryngomalacia is the most common cause of congenital stridor. It typically resolves spontaneously at the age of 12 to 24 months. Children with cor pulmonale, pulmonary hypertension, hypoxia, apnea, recurrent cyanosis, growth retardation, pectus excavatum, stridor causing respiratory failure, or laryngomalacia with severe neck/chest retractions are potential candidates for surgical treatment [6].

Laryngeal webs are rare congenital anomalies that arise from failure to dissolve the epithelial layer that shelters the laryngeal opening in normal development. Moreover, it can also be induced by trauma to the airway (intubation). These webs should be considered a lighter version within the spectrum of laryngeal atresia. 90% of the congenital laryngeal webs have been anterior glottic, while 2% are supraglottic, and 7% are subglottic. Patients with the laryngeal web generally present respiratory distress and a weak or high-pitched cry during infancy. Treatment of the laryngeal web depends on the degree of airway obstruction, whereas anterior webs in the vocal cords can be treated with simple dissection; a laryngeal stent or tracheotomy might be required in some cases [7]. Case 5 had complaints starting from birth and had 30 days of hospitalization, 11 days of which he was intubated. Although his complaints from birth onwards and the absence of adhesions in the vocal cords primarily suggested a congenital laryngeal web, it was also considered that it could have developed secondary to trauma as he was intubated for 11 days. Tracheotomy was conducted due to the presence of the web in the subglottic region and partial stenosis. Accelerated weight gain and amelioration in respiratory symptoms were detected during the follow-ups after the treatment.

Congenital subglottic stenosis is the constricting the lumen of the trachea in the cricoid area that is considered to be caused by incomplete canal formation of the

cricoid ring. It may cause biphasic stridor if it is severe in the newborn. We should note that congenital subglottic stenosis is distinguished from gained subglottic stenosis because of the lack of a history of trauma or instrumentation and relatively less severe symptoms. Less than half of the children with congenital subglottic stenosis need the administration of tracheostomy [8]. Stenosis may develop due to injury to the glottis and surrounding structures following endotracheal intubation in neonates. Acquired subglottic stenosis is more common among infants who have been intubated for a week or ≥3 times.

Endoscopy is required to identify the underlying reason and also to determine the level of the airway obstruction. Children with severe subglottic stenosis may require a tracheostomy for treatment. Decannulation may be possible as the child grows up or following reconstructive surgery [9]. Although acquired causes seem to be more common, the patient (case 1) had severe hypoxia and respiratory distress that started with birth. Very severe stenosis was detected during the flexible endoscopy of the ttic and cannulated by tracheotomy. A recovery in respiratory symptoms and accelerated weight gain were observed throughout post-cannulation follow-ups.

Subglottic hemangiomas may lead to airway obstruction. They usually become apparent around three months and cause biphasic stridor. Nowadays, the first option in treatment is propranolol. Twelve to 18 months of treatment is adequate, but it is necessary to be careful against potential adverse effects [10].

Case 2 and case 4 are the cases that we diagnosed with subglottic hemangioma. The complaints of case 2 started when he was two months old, and his stridor became permanent with the gradual growth of the subglottic hemangioma. During this period, the patient was hospitalized due to respiratory distress and received treatment for bronchiolitis. The fact that subglottic hemangiomas lead to biphasic stridor and cause expiratory manifestations makes differential diagnosis challenging for physicians.

Although the laryngoscopic examination was performed four times and one flexible bronchoscopic examination was performed due to the chronic stridor of case 4, the hemangioma, which usually looks bluish and red, could not be detected since it was covered with normal mucosa. As the current clinical manifestations indicate airway obstruction, neck CT was performed, and a hyperintense region was observed at and below the glottis level. The diagnosis could be achieved through flexible endoscopic examination, performed under general anesthesia.

Causes of congenital stridor are frequently evaluated with the misdiagnosis of croup, laryngitis, asthma, and wheezing infants [1,2,11,12]. Congenital causes of stridor can be life-threatening. Therefore, patients with chronic and recurrent symptoms should be evaluated carefully.

In conclusion, the medical history, including type and time of onset of stridor, whether it is inspiratory or biphasic, the progression of symptoms over time, the increase in breathing need in infants, such as crying, excitement, and eating, sleep-wakefulness relationship, perinatal problems and endotracheal intubation, should be questioned for differential diagnosis. Patients with intermittent respiratory problems coexisting with stridor can be diagnosed typically through an adequate medical history, a well-enough PE, and radiological methods. Flexible endoscopy provides a safe and complete examination of the children's airway and should be conducted under general anesthesia on cases with suspected airway pathology if it is indispensable.

Acknowledgements: Thanks to Prof. Dr. Özden Anal for extensive English editing.

Informed Consent: Written informed consent from the patients' parents or guardians was obtained to publish cases in this study.

Peer-review: Externally and internally peer-reviewed.

Author Contributions

Surgical and Medical Practices: S.A., N.U., S.As., G.A., Ö.A., Ö.K., B.A., Y.O., T.E., Ö.Kar., Concept: S.A., S.As., Y.O., T.E., Ö.Kar., Design: S.A., S.As., G.A., Ö.A., Ö.K., Y.O., T.E., Ö.Kar., Data Collection and/or Processing: S.A., N.U., S.As., G.A., Ö.A., Ö.K., B.A., Y.O., T.E., Ö.Kar., Analysis and/or Interpretation: S.A., N.U., S.As., Ö.Kar., Literature Search: S.A., S.As., Writing: S.A., S.As., Ö.Kar.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Nguyen TVT, Le BBT. Stridor or wheezing? Congenital laryngomalacia misdiagnosed with infantile asthma in young children: a case report. MedPharmRes. 2020;4(3):21-3. doi:10.32895/ump.mpr.4.3.5

2. Derinöz O, Şişmanlar T. An unusual cause of stridor: Congenital laryngeal web. Turk Pediatr Ars. 2018;53(3):185-8. doi: 10.5152/TurkPediatriArs.2017.3922.

3. Uribe-Parra JD, Lozano-Triana CJ, Lopez-Cadena AF, Landinez-Milan G. Rare chronic stridor: Case report and literature review. Rev Fac Med. 2017;65(3):531-6. doi:10.15446/revfacmed.v65n3.56010.

4. Pfleger A, Eber E. Assessment and causes of stridor. Paediatr Respir Rev. 2016;18:64-72. doi: 10.1016/j.prrv.2015.10.003.

5. Shatz A, Goldberg S, Picard E, Kerem E. Pharyngeal wall collapse and multiple synchronous airway lesions. Ann Otol Rhinol Laryngol. 2004;113(6):483-7. doi: 10.1177/000348940411300613.

6. Boudewyns A, Claes J, Van De Heyning P. Clinical practice : An approach to stridor in infants and children. Eur J Pediatr. 2010;169(2):135-41. doi: 10.1007/s00431-009-1044-7.

7. Rutter MJ, Dickson JM. Congenital Malformations of the Larynx. In: Elden LM, Zur KB, eds. Congenital Malformations of the Head and Neck. Springer Science+Business Media New York 2014; 2013:1-193. doi:10.1007/978-1-4419-1714-0

8. Hartnick CJ, Cotton RT. Congenital laryngeal anomalies: Laryngeal atresia, stenosis, webs, and clefts. Otolaryngol Clin North Am. 2000;33(6):1293-308. doi: 10.1016/S0030-6665(05)70282-6.

9. Downing GJ, Kilbride HW. Evaluation of Airway Complications in High-Risk Preterm Infants: Application of Flexible Fiberoptic Airway Endoscopy. Pediatrics. 1995;95(4):567-72. Available from: https://link.gale.com/apps/doc/A16899154/AONE?u=anon~9c2a69be&sid=googleScholar&xid=9bdfdcdf

10. Bush A, Abel R, Chitty L, Harcourt J, Hewitt RJ, Nicholson AG. Congenital Lung Disease. In: Wilmott RW, ed. Kendig ' s Disorders of the Respiratory Tract in Children 9th Edition. Ninth Edit. Elsevier Ltd; 2019:1086-251.

11. Atmaca S, Ünal R, Şeşen T, Kılıçarslan H, Ünal A. Laryngeal Foreign Body Mistreated as Recurrent Laryngitis and Croup for One Year. Turk J Pediatr. 2009;51(1):65-6. Available from: http://citeseerx.ist.psu.edu/viewdoc/download?doi=10.1.1.602.8188&rep=rep1&type=pdf

12. Ramly B, Khan A. Case Report Stridor, Is It Always Laryngomalacia In An Infant? Malaysian J Paediatr Child Heal. 2017;23:55-9. Available from: https://mpaeds.my/journals/index.php/MJPCH/article/view/46/28