

Isolated congenital tracheal stenosis: A rare and deadly condition

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Received: 22 May, 2019

Accepted: 28 July, 2019

ABSTRACT

Introduction: Congenital tracheal stenosis (CTS) is a serious and rare condition. In most cases, CTS is associated with cardiopulmonary abnormalities; however, isolated CTS is present in 10%–30% of patients. The severity of the disorder is dependent on the symptoms, which correlate with the CTS classification.

Case presentation: We discuss our findings in an infant who presented with severe respiratory compromise where incidental intra-operative findings revealed CTS with no cardiopulmonary abnormalities. Because of a lack of resources in the emergency department, we created a tracheostoma and inserted an endotracheal tube.

Conclusion: The main aim in treating CTS is to secure the airway and provide sufficient oxygen.

KEYWORDS

Congenital, Tracheal stenosis, Deadly condition

INTRODUCTION

Congenital tracheal stenosis (CTS) is a rare disorder that may be associated with cardiopulmonary abnormalities. Diagnosis is almost always made during infancy secondary to respiratory obstruction.¹ CTS is a spectrum of disorders involving narrowing of the airway with lesions commonly located at the complete tracheal rings with variable length, location, and severity of the luminal narrowing.^{2,3}

However, isolated CTS is present in 10%–30% of patients.⁴ We experienced a 3-months-old baby girl, who presented with upper airway obstruction requiring an emergency tracheostoma and direct laryngoscopy with rigid tracheoscopy, which revealed severe isolated CTS. In this case report, we explained the rarity of this disorder, which is a significant challenge, and discuss the relevant management.

CASE REPORT

A 3-months-old infant was initially treated for pneumonia

secondary to upper respiratory tract infection. She experienced noisy breathing beginning at 1 month of age that worsened during crying and resolved spontaneously; no cyanosis was noted. Clinical examination revealed expiratory stridor with no cyanosis, and without syndromic facies. On day three of admission, the infant developed worsened respiratory symptoms requiring supplemental oxygenation. Multiple attempts at intubation were performed with one apparent life-threatening event requiring cardiopulmonary resuscitation for 1 minute. We finally able to intubate the patient with a 3.5-mm endotracheal tube, and a chest X-ray revealed that the endotracheal tube could not be advanced further than the T1-T2 level. Echocardiography was performed at bedside revealed no abnormality. However, the patient continued to deteriorate, and emergency tracheostomy was performed to secure the airway. In this of emergency setting, computed tomography (CT) of the neck and thorax was not possible prior to the surgery.

Intraoperatively, we intubated the patient with a 3.0-

DOI: 10.1002/ped4.12148

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mm endotracheal tube, which we could advance further than the 3.5-mm tube. We performed tracheostomy, but unfortunately, inserting the smallest (3.0-mm) tracheostomy tube failed. Therefore, we created a tracheostoma between second and third tracheal rings, and inserted a 2.0-mm endotracheal tube through the tracheostoma, which we anchored 3 cm from the tracheostoma margins (Figure 1).



FIGURE 1 Tracheostoma and modified endotracheal tube size 2.0 mm placement.

Direct laryngoscopy and rigid tracheoscopy performed in the emergency room revealed normal appearance of bilateral vocal cords, epiglottis, arytenoid cartilages, and the subglottis (Figure 2). No subglottic narrowing or granulation was noted; however, tracheal narrowing was present circumferentially from the first ring, with 80% narrowing at the third to fourth rings (Figure 3), which was suspected to be funnel-type stenosis. In addition, the distal part of the endotracheal tube was located at the level of the middle of fourth tracheal ring, and it was difficult to pass the rigid scope beyond the fourth ring.



FIGURE 2 Direct laryngoscopy and rigid tracheoscopy revealed normal appearance of bilateral vocal cords, epiglottis, arytenoid cartilages, and the subglottis.

We explained the condition thoroughly to the patient's parents post operatively, and emphasized that the patient required further management at a specialized center with the necessary expertise and equipment to perform the required surgical procedure; however, there was no nearby center with the expertise. The patient's parents understood the severity of their daughter's condition, but did not elect further management, and requested no further intervention and no active resuscitation. The patient died the next day secondary to worsening respiratory distress.

DISCUSSION

CTS is a rare and life-threatening condition with a mortality rate of 44%–79%.⁵ The estimated incidence of CTS is two per 100 000 live births;⁶ however, the true incidence of CTS is unknown because many infants die before the diagnosis is made.⁷ CTS is usually associated with other congenital anomalies, seen as cardiopulmonary anomalies in the majority of patients.^{8,9} More than 75% of

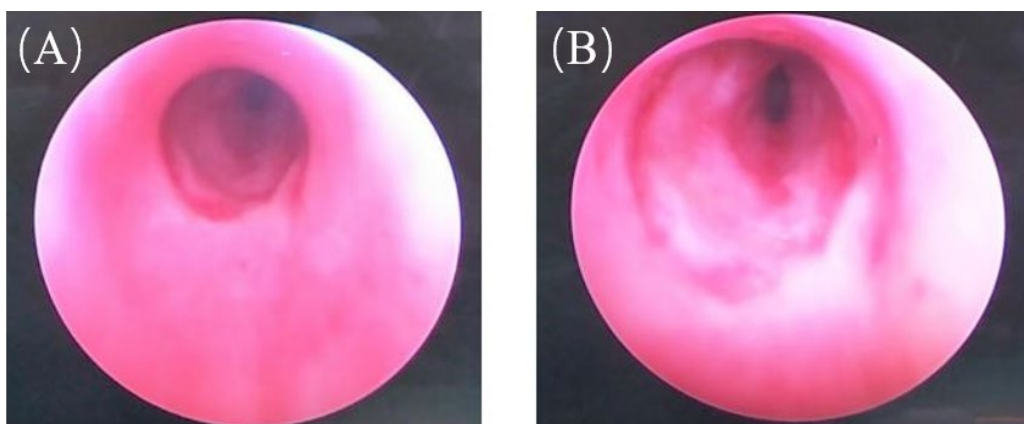


FIGURE 3 Tracheal narrowing was present circumferentially from the first ring (A), with 80% narrowing at the third to fourth rings (B).

patients with CTS are complicated with other anomalies,¹⁰ and most commonly, with pulmonary arterial sling, cardiac defects, and lung hypoplasia or agenesis. Fortunately, isolated CTS presentations are very rare.

CTS is characterized by narrowing of the tracheal lumen, and Cantrell and Guild¹¹ classified CTS by anatomical criteria into three types: generalized, funnel-like, and segmental stenosis. Classification using functional symptoms was introduced in 2003 as mild: asymptomatic or occasional symptoms; moderate: respiratory symptoms without respiratory embarrassment; and severe: severe symptoms including respiratory embarrassment.¹² Our patient was classified anatomically as having funnel-like stenosis considering we were unable to pass the endotracheal tube beyond the fourth tracheal ring, and functionally, she had severe symptoms.

It is believed that pediatric patients are able to tolerate 50% tracheal narrowing before becoming symptomatic.¹³ Airway obstruction in patients with CTS can be exacerbated by spasms, inflammation, or direct trauma after airway manipulation or surgery.¹³

CTS management varies depending on the severity, airway pathology, and the involvement of other anomalies, and a multidisciplinary approach is usually necessary for success.⁷ Generally, if the patient is asymptomatic, conservative management is preferred, and in severe cases surgical intervention is the treatment of choice.¹⁴ The surgical approach may differ according to the surgeon and the symptom severity, and may include resection and primary anastomosis, patch tracheoplasty with nontracheal autologous tissue, slide tracheoplasty, and tracheal transplant with a cadaveric tracheal homograft.^{4,15,16} However, in our patient, because of the lack of resources and the urgency we created a tracheostoma and replaced the tracheostomy tube with a 2.0-mm endotracheal tube. The child was then ventilated, with no desaturation.

In conclusion, CTS is a rare and life-threatening condition, and early diagnosis and intervention increase the survival rate. A method of prenatal diagnosis is needed; however, no prenatal surgical procedures have been proven satisfactory. Patient management varies according to the center and the available facilities; however, the main aim of surgical intervention in emergent patients is to secure the airway and provide sufficient oxygen.

Data on the treatment and long-term prognosis in CTS are lacking. Further studies and higher numbers of cases are needed, and active efforts with a multidisciplinary approach to discover the underlying cause are essential.

CONFLICT OF INTEREST

None.

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How to cite this article: Arunathan R, Zainal Ariffin AH, Khor KG, Tan SN. Isolated congenital tracheal stenosis: A rare and deadly condition. *Pediatr Invest.* 2019;3:191-193. <https://doi.org/10.1002/ped4.12148>