

Case Report

Innominate artery compression syndrome of the trachea in a 12-month old

12 aylık bir bebekte trakeanın brakiosefalik arter kompresyon sendromu

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ABSTRACT

Presentation: A 12-month-old female presents after an episode of difficulty breathing with apnea and cyanosis. Parents performed chest compressions for 20 seconds until she woke up. This is her third episode of apnea over the last 10 months. Parents also describe chronic biphasic stridor at baseline since she was 2 months old. The stridor is not positional or associated with eating, but worsens when ill. She does not spit up after feeding and there is no stertor.

Physical Exam: T 36.2 C, HR 140 beats/min, BP 119/77 mmHg, RR 30 breaths/min, SpO2 98% on room air. She is well-appearing with mild congestion and biphasic stridor that is audible without a stethoscope.

Evaluation: Respiratory panel positive for rhinovirus, which was positive 3 weeks ago after a previous apneic event. Chest x-ray unremarkable. No signs of aspiration noted on evaluation by speech language pathologist. Video EEG was normal. EKG showed sinus tachycardia. CTA of the chest revealed significant compression of the trachea where the right brachiocephalic (innominate) artery passes over the trachea.

Diagnosis: She underwent bronchoscopy which showed tracheomalacia with tracheal compression to approximately 90% obstruction, consistent with a diagnosis of innominate artery compression syndrome (IACS). This patient underwent reimplantation of the innominate artery with post-operative bronchoscopy that showed significant improvement in tracheal compression. She presented one month later with apneic events and repeat bronchoscopy showed tracheomalacia with posterior wall prolapse. She then underwent posterior tracheopexy and subsequently had a bronchoscopy which showed no collapse or dynamic compression.

Discussion: Innominate artery compression of the trachea can cause biphasic stridor, respiratory arrest, and growth faltering. The degree of compression can range from asymptomatic to severe obstruction. Respiratory illnesses often cause stridor and can influence the index of suspicion for a rare condition like IACS. Surgical intervention is warranted in children who present with life-threatening apneic events.

Keywords: ear nose throat, cardiology, tracheal compression

ÖZET

Başvuru: 12 aylık kız hasta, apne ve siyanoz ile seyreden solunum güçlüğü epizodu sonrası başvurdu. Ebeveynler, hasta uyanana kadar 20 saniye göğüs kompresyonu uyguladı. Bu, son 10 ay içinde yaşadığı üçüncü apne epizodudur. Ebeveynler ayrıca 2 aylıkken başlayan kronik bifazik stridor tanımlamaktadırlar. Stridor pozisyonel değildir veya yemek yeme ile ilişkili değildir, ancak hasta olduğunda kötüleşir. Beslenmeden sonra kusma yoktur ve stertor yoktur.

Received: 19.02.2025 · Accepted: 10.04.2025 · Published: 13.02.2026

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Cite this article as: Taylor I, Van Meurs A, Tam R. Innominate artery compression syndrome of the trachea in a 12-month old. *Pediatr Acad Case Rep.* 2026;5(1):15-8.

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Fizik Muayene: Ateş 36,2°C, kalp hızı 140 atım/dakika, kan basıncı 119/77 mmHg, solunum hızı 30 nefes/dakika, SpO2 oda havasında %98. İyi görünümlü, hafif konjesyon ve stetoskop olmadan duyulabilen bifazik stridor mevcut.

Değerlendirme: Solunum paneli rinovirüs için pozitif, bu virus 3 hafta önce önceki apneik olay sonrasında da pozitif. Akciğer grafisi normal. Konuşma-dil patoloğu tarafından yapılan değerlendirmede aspirasyon bulgusu saptanmadı. Video EEG normaldi. EKG sinüs taşikardisi gösterdi. Göğüs BT anjiyografisi, sağ brakiosefalik (innominat) arterin trakea üzerinden geçtiği yerde trakeada belirgin kompresyon ortaya koydu.

Tanı: Hastaya bronkoskopi yapıldı ve yaklaşık %90 obstrüksiyon ile trakeal kompresyon gösteren trakeomalazi saptandı, bu innominat arter kompresyon sendromu (IACS) tanısı ile uyumluydu. Bu hasta innominat arter reimplantasyonu geçirdi ve ameliyat sonrası bronkoskopi trakeal kompresyonda belirgin düzelme gösterdi. Bir ay sonra apneik olaylarla başvurdu ve tekrar bronkoskopi posterior duvar prolapsusu ile trakeomalazi gösterdi. Daha sonra posterior trakeopeksi yapıldı ve ardından yapılan bronkoskopi kollaps veya dinamik kompresyon göstermedi.

Tartışma: Innominat arter trakea kompresyonu bifazik stridor, solunum arresti ve büyüme geriliğine neden olabilir. Kompresyon derecesi asemptomatikten ciddi obstrüksiyona kadar değişebilir. Solunum yolu enfeksiyonları sıklıkla stridora neden olur ve IACS gibi nadir bir durum için şüphe endeksini etkileyebilir. Yaşamı tehdit eden apneik olaylarla başvuran çocuklarda cerrahi müdahale gereklidir.

Keywords: kulak burun boğaz, kardiyooloji, trakeal kompresyon

INTRODUCTION

Innominate artery compression syndrome (IACS) of the trachea is a significant cause of airway obstruction in infants and children and may cause biphasic stridor, cyanosis, and respiratory arrest. The degree of compression varies, as does the symptom severity, ranging from asymptomatic to severe obstruction resulting in apnea and the need for airway intervention. [1] The exact prevalence of abnormalities involving mediastinal great vessels is unknown, as most cases are asymptomatic, but is estimated to be up to 3% of the population. [1,2] Infants and children with tracheal vascular compression may also present with chronic cough, dysphagia, and growth faltering.

CASE REPORT

The patient is a 12-month-old female presenting after an episode of difficulty breathing with subsequent apnea and cyanosis. Her pulse was not checked during the episode, but her parents performed chest compressions for 20 seconds until she woke up. This is her third episode of apnea after respiratory difficulty over the last 10 months. The first two apneic events coincided with confirmed respiratory syncytial virus (RSV) and rhinovirus infections, respectively. Parents report removing curdled milk from the back of her mouth during these episodes.

Parents also describe chronic biphasic stridor at baseline since her RSV infection at two months of age. The stridor has been present most days over the last 10 months and is not positional, but it does worsen when she is ill. It is not associated with drinking or with eating

solids. She does not spit up excessively after feeding and there is no significant stertor while asleep. She has a slight gross motor delay and significant growth faltering after two months of age. However, she is otherwise developmentally appropriate, up to date on vaccinations, and has no other significant past medical or surgical history.

After this episode of apnea, she was initially brought to a local emergency department, where she was administered dexamethasone, nebulized albuterol and ipratropium, and racemic epinephrine. Given the need for subspecialty care, she was air-transferred across state lines to a large pediatric center. Her previous two apneic episodes were managed at the same local emergency department and she was discharged home given resolution of her respiratory difficulty. For previous presentations, she has received steroid courses, antibiotics, and acid suppression therapy, as well as multiple unremarkable chest x-rays. No diagnosis was found.

Her presenting vital signs at the large pediatric center are as follows: T 36.2 [E1] C, HR 140 beats/minute, BP 119/77 mmHg, RR 30 breaths/minute, SpO2 98% on room air. On examination, she is well-appearing with mild congestion, coarse breath sounds, and biphasic stridor that is audible without a stethoscope. Her chest x-ray shows low lung volumes but is otherwise normal. Her complete blood count with differential is unremarkable apart from white blood cell count of $18 \times 10^9/L$ and platelet count of $872 \times 10^9/L$. Complete metabolic panel is unremarkable. The respiratory panel is positive for rhinovirus, although she did test positive for rhinovirus three

weeks before this presentation after her last apneic event.

The differential for this patient's recurrent apneic events and stridor based on initial presentation includes reflux with aspiration, laryngomalacia, tracheomalacia, complete tracheal rings, tracheoesophageal fistula (TEF), or vascular compression. Additional causes of apneic events were considered, including seizure, arrhythmia, or hypertrophic cardiomyopathy, although these would not classically be associated with chronic stridor. Reflux and aspiration are common in infants and were initially considered in this patient given the reports of milk in the back of her throat after the apneic episodes, but this was considered less likely as the events do not occur after feeds. In the hospital, there were no overt signs of aspiration noted on evaluation by speech-language pathologist. [3] Laryngomalacia or tracheomalacia were also considered, and pediatric otolaryngology was consulted to further evaluate for these pathologies. However, laryngomalacia was thought to be less likely given that the stridor was not positional. Her biphasic stridor and apneic events could be explained by complete tracheal rings, TEF, a vascular ring, or other form of vascular compression, which were evaluated for with chest computed tomography angiography (CTA). The differential for apneic events also includes seizures, so continuous video electroencephalogram monitoring was performed for approximately 24 hours and the results were normal. An electrocardiogram was obtained and showed sinus tachycardia, reassuring against arrhythmias as the cause of her apneic events. Hypertrophic cardiomyopathy was also considered unlikely given her reassuring family history and physical examination without murmur.

CTA of the chest revealed significant compression of the trachea at the level of the thoracic inlet, where the right brachiocephalic (innominate) artery passes over the trachea, with no evidence of vascular ring or sling (Figure 1). She then underwent microlaryngoscopy and bronchoscopy, which showed severe tracheomalacia with dynamic mid-tracheal compression to approximately 90% obstruction. The pediatric cardiothoracic surgery team was consulted for recommendations regarding surgical intervention for tracheal compression by the innominate artery. A pre-operative echocardiogram was obtained, demonstrating normal cardiac anatomy and function.

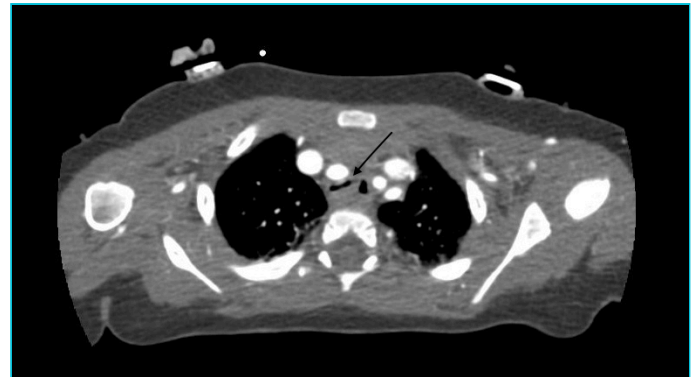


Figure 1. CT angiography of the chest demonstrating compression of the trachea by the right brachiocephalic artery.

DISCUSSION

Definitive treatment for IACS of the trachea is largely a surgical operation. The two most widely described surgical approaches are aortopexy with innominate artery suspension and innominate artery reimplantation. [4,5] Reimplantation may provide greater and more immediate improvement in airway obstruction as well as improved safety outcomes when compared with aortopexy. [6] No standardized imaging-based criteria exist to guide decisions for surgical intervention; this decision is largely clinical, and becomes urgent when patients present with recurrent apnea. [7] Additional surgical strategies that have been described for the treatment of acquired tracheomalacia caused by innominate artery compression. [8,9] Of note, there has been some controversy associated with the diagnosis, as it may be unclear whether symptoms are caused solely by mass effect from the artery or if tracheomalacia is contributing, as vascular compression can weaken the cartilaginous rings of the trachea over time. [7,10] Regardless, intervention is warranted in children, such as this one who presents with potentially life-threatening apneic events.

The patient underwent reimplantation of the innominate artery with the pediatric cardiothoracic surgery team and tolerated the surgery well with no apparent complications. Post-reimplantation bronchoscopy with the pediatric otolaryngology team showed significant improvement in tracheal compression decreasing from 90% to 65% obstruction. She did not have ongoing stridor or apneic events in the immediate postoperative period and was discharged home on postoperative day two. She re-presented one month later with additional apneic events during tantrums, at which time repeat CTA of the

chest demonstrated relieved airway compression after repositioning of the right brachiocephalic artery (Figure 2). Repeat bronchoscopy was performed which showed continued tracheomalacia with prolapse of the posterior wall, especially with increased work of breathing or agitation. She underwent posterior tracheopexy with the pediatric general surgery team and subsequently had a bronchoscopy, which showed no collapse or dynamic compression. She had no additional events in the hospital during this second admission and was discharged home on postoperative day seven.

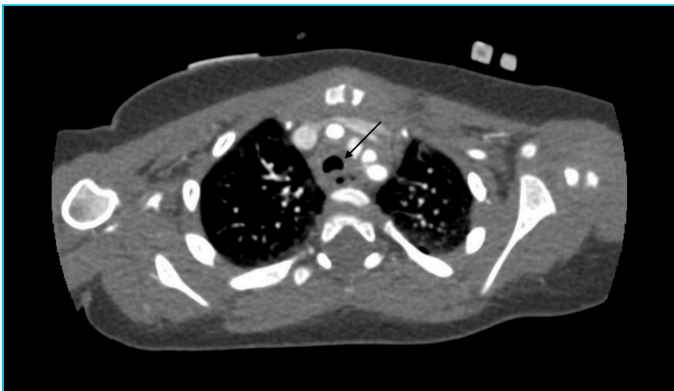


Figure 2. CT angiography of the chest demonstrating relieved airway compression after surgical repositioning of the right brachiocephalic artery.

CONCLUSION

Vascular compression of the trachea should be suspected in children presenting with unexplained chronic respiratory distress, such as recurrent stridor and/or episodes of apnea, and dysphagia and/or growth faltering should heighten concern. The diagnosis of IACS relies heavily on imaging studies and bronchoscopy, which can help visualize dynamic tracheal compression. Surgical intervention is warranted in some patients with innominate artery compression syndrome of the trachea, and a multidisciplinary approach with surgical subspecialists helps develop a definitive treatment plan. Respiratory diseases often cause stridor and can influence the index

of suspicion for a rare condition like IACS. When subspecialty resources are constrained, a repeat presentation of apnea with parental complaint of chronic stridor warrants early evaluation.

Patient Consent Form / Hasta Onam Formu

The parents' of this patient consent was obtained for this study.

Conflict of Interest / Çıkar Çatışması

The authors declared no conflicts of interest with respect to authorship and/or publication of the article.

Financial Disclosure / Finansal Destek

The authors received no financial support for the research and/or publication of this article.

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