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Successful case report of congenital long-segment complex tracheal stenosis treated with sliding tracheoplasty associated with pulmonary artery vascular ring

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Abstract

Background Congenital tracheal stenosis is a rare structural obstructive disorder affecting a segment of the trachea. It is often caused by complete or nearly complete cartilaginous rings narrowing the airway. Approximately 50% of cases are associated with abnormal left pulmonary artery rings.

Case presentation We present the case of a 4-month-old female infant with complex congenital tracheal stenosis associated with a left pulmonary artery vascular ring and congenital heart disease. The patient underwent successful surgical intervention involving repair of the congenital heart defect and reimplantation of the pulmonary artery, followed by sliding tracheoplasty in one surgery procedure. Postoperative outcomes were favorable, with successful extubating and resolution of respiratory symptoms.

Conclusions Sliding tracheoplasty proved an effective treatment option in this complex case, highlighting its reproducibility and favorable outcomes in managing congenital tracheal stenosis.

Keywords Case report, Tracheal stenosis, Pediatric, Heart defects

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Background

Congenital tracheal stenosis is a structural obstructive disorder affecting a segment of the trachea, which can be short or long [1]. This condition, affecting 1 in 64,500 live births, is often caused by complete or nearly complete cartilaginous rings narrowing the trachea [1, 2].

Approximately 50% of cases are associated with an abnormal left pulmonary artery ring. Clinically, symptoms typically appear a few months after birth and include predominant biphasic stridor, cyanosis, chest retractions, and recurrent pneumonia [3, 4].

In this article, we describe a successful case involving a patient with congenital long-segment tracheal stenosis associated with a vascular ring. The patient was treated surgically in a single procedure with excellent results.

Case presentation

A female infant, 4 months old, was referred to our center with a diagnosis of congenital heart disease under investigation. The patient was born as one of the twins in her mother's first pregnancy, with prenatal care since the first month, prenatal ultrasounds without alteration. She was delivered by cesarean due to a premature rupture of membranes of 16 h. With 2370 g of weight and 46 cm in length, a gestational age assessment score corresponds to 37 weeks of gestation. Upon hospital discharge, she had no complications.

A heart murmur was detected during the initial evaluation at 1 month of age, leading to follow-up with pediatric cardiology. An echocardiogram revealed a 3-mm patent foramen ovale, a 2×4 mm ventricular septal defect, and moderate pulmonary hypertension. The patient is being treated with spironolactone. She presented to the emergency department with a sudden onset of paroxysmal cough, dyspnea, decreased appetite, and fatigue. She denied fever or altered mental status, prompting a visit to the corresponding zone hospital, where she was hospitalized and started on a diagnostic protocol.

A nano tomography revealed the presence of a gastroesophageal and retro bronchial sling of the left pulmonary artery.

The patient was referred for evaluation by the pediatric cardiology service and for a bronchoscopy. On arrival, she presented with respiratory deterioration and signs of septic shock, requiring ventilatory support and admission to the intensive care unit. During her hospitalization, she completed the study protocol and was evaluated by a multidisciplinary team: pediatric cardiology, pediatric thoracic surgery, and pediatric cardiothoracic surgery.

Preoperative diagnoses included congenital long-segment tracheal stenosis with the presence of an accessory bronchus (Grillo type III), sling of the left pulmonary artery, accessory right lobar bronchus, auricular septal defect (ASD), VSD, and severe pulmonary hypertension (Fig. 1). A bronchoscopy confirmed the accessory right bronchus and unpassable tracheal stenosis (Fig. 2).

During surgery, repair of the ASD, VSD, and reimplantation of the pulmonary artery were performed without

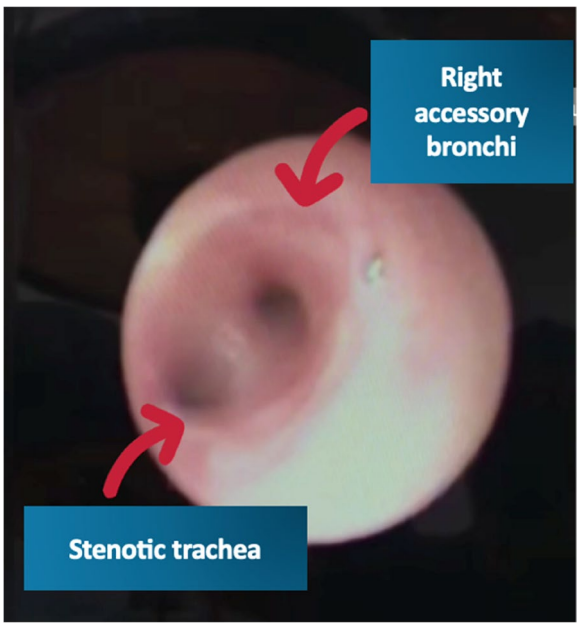

Fig. 2 Pre-surgical bronchoscopy. Accessory right bronchi, with neonatal bronchoscope, which was unable to pass through the stenotic trachea

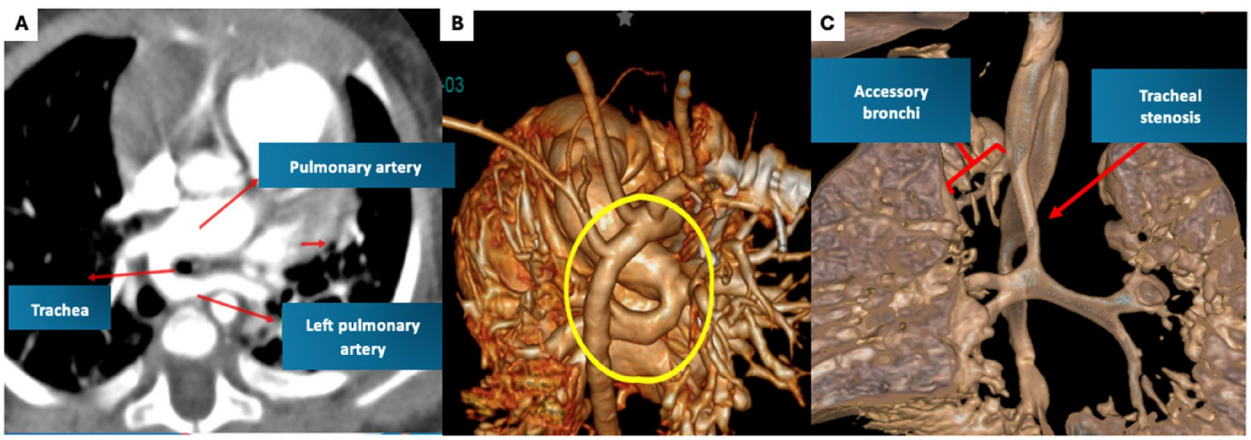

Fig. 1 Angiotomography and reconstruction. **A** This study shows the descending aorta behind the left pulmonary artery ring. **B** The image of the reconstruction. **C** In a coronal view, the accessory tracheal bronchi and the stenosis area range from the emergence of the right tracheal bronchus to the carina

complications in the first surgery timing by the pediatric cardiothoracic surgery team.

Exposure of the surgery field is achieved through midline sternotomy, dissecting through planes until the sternum is reached and divided using an oscillating saw. Dissection continues through planes to release any adhesions, followed by pericardial incision and placement of non-absorbable suture references to allow exposure of the middle mediastinum. An anatomical exploration is conducted, confirming the pre-surgical diagnosis. Unfractionated heparin is administered, achieving an activated clotting time (ACT) > 400, with a decrease in temperature to 30 °C. The ascending aorta is cannulated with a 12-Fr cannula, and venous cannulation is performed in the superior and inferior vena cava using 7–0 polypropylene sutures. Total cardiopulmonary bypass is established without complications.

A right atriotomy is performed through the incision, identifying the atrial and interventricular communication. The interventricular communication is closed primarily with 6–0 Prolene sutures, and the secundum-type atrial communication is closed primarily with 6–0 Prolene sutures. The right atriotomy is closed with 6–0 Prolene sutures. The left pulmonary sling is dissected from the surrounding structures, clamps are placed, and it is cut. It is then reimplanted on the lateral aspect of the pulmonary trunk using 7–0 Prolene sutures.

Subsequently, the tracheal stenosis was addressed through a sliding tracheoplasty. An accessory right bronchus and complete stenosis of seven tracheal rings up to the level of the carina were found. First, a dissection of the posterior aspect of the trachea was performed, and a silk suture was placed to facilitate better mobilization. A circumferential incision was made at the midpoint of the stenosis. In the distal segment, a longitudinal incision was made on the anterior aspect of the trachea to the carina. A longitudinal incision was made in the proximal segment's posterior aspect. The two tracheal stumps were approximated, sliding one over the other, and anastomosed with simple PDS 5–0 sutures, starting with the posterior aspect. A pneumatic test was performed with no evidence of air leakage. Tissue adhesive was applied above the anastomosis site. The anastomosis was performed anterior to the pulmonary vessels and the accessory bronchus; no surgical procedure was performed on the accessory bronchus.

The sliding tracheoplasty was completed in 41 min without complications, with a total pump time of 171 min and an aortic clamp time of 130 min (Figs. 3 and 4).

The patient was managed in the PICU, with mechanical ventilation in pressure-controlled mode, PIP 17, PEEP 6, FiO_2 85%, and respiratory rate (RR) 40; gasometry showed respiratory alkalosis, allowing for a reduction in ventilatory parameters and successful extubation was achieved on postoperative day 17. Follow-up studies showed adequate airway patency (Fig. 5), and no stenosis was observed on bronchoscopy before discharge. The patient continues to be followed up and shows clinical improvement at 1 year of age. They were currently undergoing follow-up bronchoscopies, which showed adequate

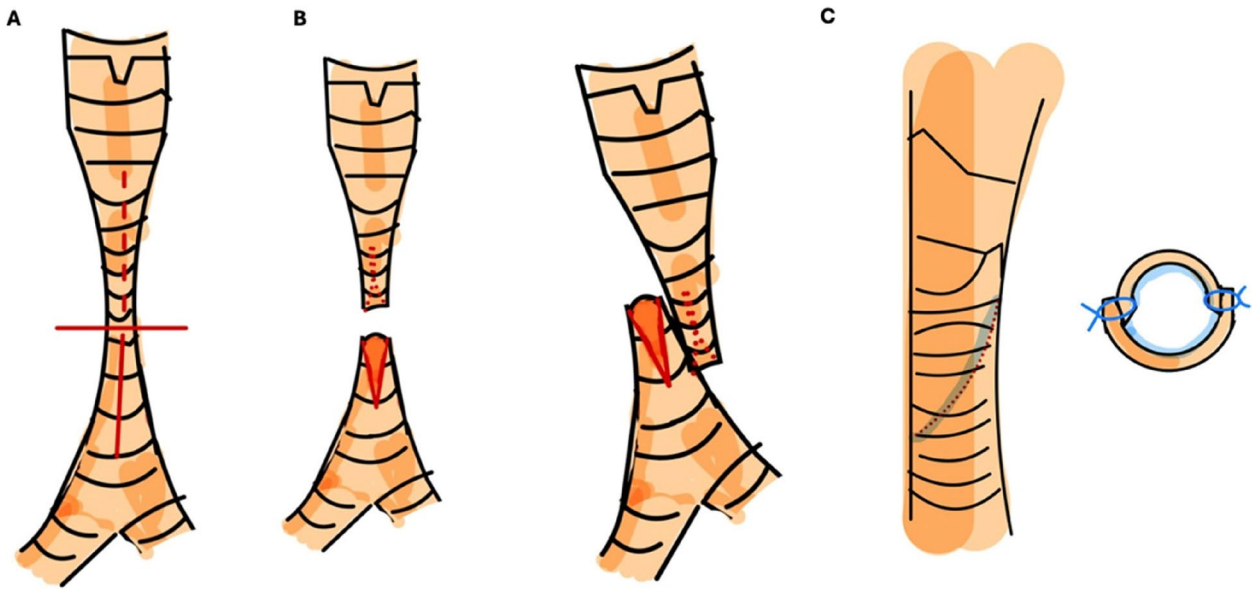

Fig. 3 The sliding tracheoplasty technique. **A**, **B** Longitudinal incision on the proximal trachea and transversal incision on the distal trachea before and after. **C** Final anastomosis from lateral view and axial view. A scheme made by the author

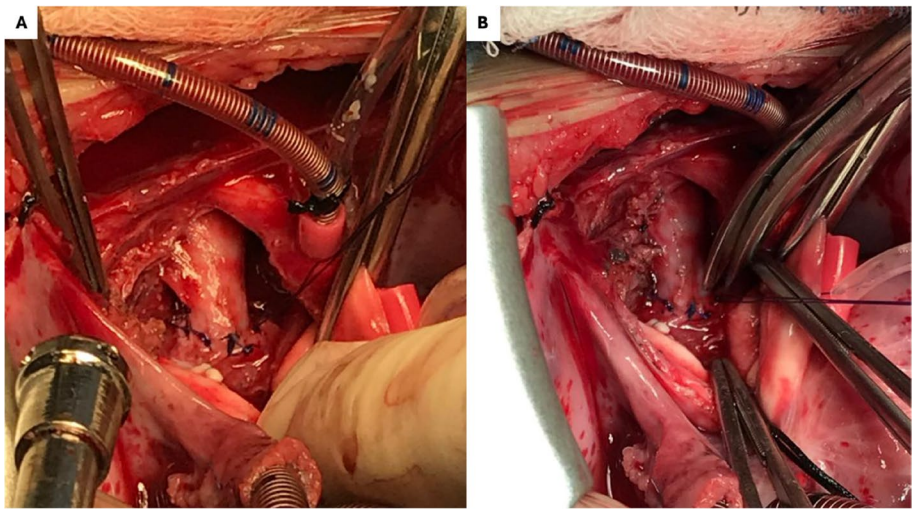

Fig. 4 The sliding tracheoplasty technique image. **A** Anterior anastomosis of the trachea. **B** Lateral view of the anastomosis on the trachea

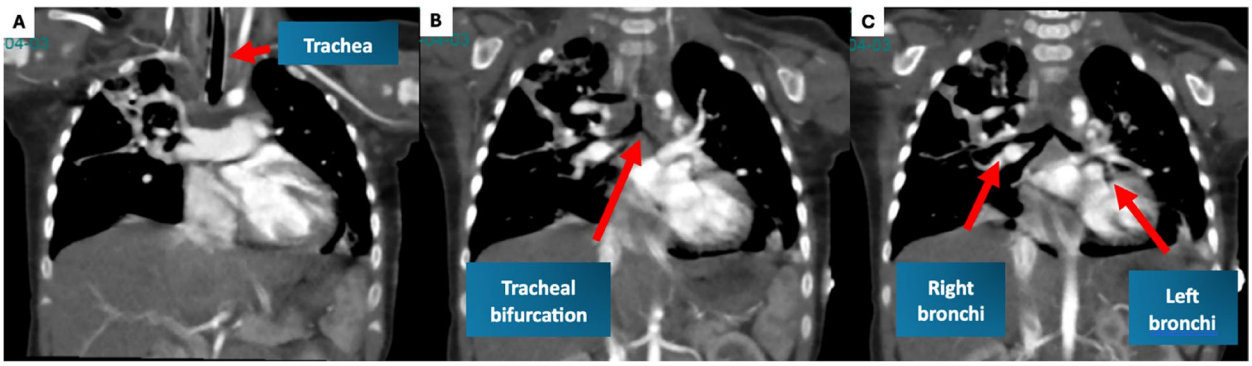

Fig. 5 Angiotomography. **A** A coronal view on the sixth day after surgery, showing adequate air passage at the tracheal level. **B** Tracheal bifurcation. **C** Left and right permeable bronchi

passage of the bronchoscope at the level of the tracheal plasty without evidence of stenosis. He engages in play and movement activities without the need for supplementary oxygen.

Discussion

Congenital tracheal stenosis is a rare and potentially life-threatening anomaly that often necessitates invasive mechanical ventilation and definitive tracheal reconstruction. Patients typically present with inspiratory stridor, expiratory wheezing, apnea, feeding difficulties, recurrent pneumonia, upper respiratory tract infections, and episodes of cyanosis [2, 5]. Up to 60% of cases are reported to have associated congenital malformations, including vascular rings, congenital heart diseases, tracheoesophageal fistula, and hemivertebrae [6].

Diagnosis is sometimes complicated; it can be achieved through chest radiographs, bronchoscopy, CT scans, and MRI [1]. CT is reported to be superior to MRI in providing higher spatial resolution with more apparent tracheal contours and shorter scanning times. Congenital tracheal stenosis consists of complete cartilaginous rings. It can be classified according to Grillo classification: type I—generalized tracheal hypoplasia, where nearly the entire trachea is stenotic, with only average cranial rings 1–3; type II—funnel-shaped tracheal narrowing, where the standard segment varies in location and length but always assumes a funnel shape from cranial to caudal; type III—segmental tracheal stenosis, characterized by short-segment stenosis at various levels of the trachea, sometimes below an anomalous right upper lobe bronchus; and type IV—bridge bronchus stenosis is a variant of type III, in which the anomalous right upper lobe bronchus lies near the carina via horizontally branching bronchi, and the stenotic bridge bronchus connects the proximal trachea to the rest of the lungs [7, 8].

Various approaches exist for managing congenital tracheal stenosis, from observation to various surgical treatments [8]. The patient's age and symptoms during initial evaluation are crucial in determining the treatment course. Patients are generally categorized into three groups.

Patients with mild stenosis (narrow posterior membranous trachea, asymptomatic or with occasional symptoms) allow for observation over a variable period, monitoring of airway growth, and accompanying symptoms. Patients with moderate stenosis (absence of posterior membranous trachea, complete tracheal rings, symptomatic but without critical airway compromise) require short-term surgical intervention. Patients with severe stenosis (complete tracheal rings, highly symptomatic, and critically compromised airway) can be further subdivided into type A, without associated anomalies, and type B, with associated anomalies, necessitating immediate intervention [9].

Traditional surgical treatment options include endoscopic dilation with or without stent placement, tracheal resection with end-to-end anastomosis for short-segment stenosis, and sliding tracheoplasty, as well as its modified version for long-segment stenosis, along with the placement of biological grafts, with varied success rates [10]. Surgical intervention depends on the surgeon's preference; however, sliding tracheoplasty is currently the treatment for severe congenital stenosis, as it allows for a loss of only 50% of the stenotic trachea's length. This procedure is typically performed via median sternotomy with ECMO or extracorporeal circulation [5, 11]. The original sliding tracheoplasty technique described by Tsang and colleagues in 1989 [2] does not include carinal repair. It is generally not considered an option for infants with involvement of both proximal bronchi, for whom the modified technique is recommended [12, 13].

Concomitant anomalies, such as atrial and ventricular septal defects, can be corrected simultaneously. Complex anomalies, such as Tetralogy of Fallot, pulmonary atresia with ventricular septal defect, and complete atrioventricular septal defects, are recommended for staged repair [7]. However, the decision between simultaneous or staged repair is controversial; simultaneous repair is believed to be more physiological, but surgeries requiring prolonged cardiopulmonary bypass are generally associated with a poor prognosis [4, 14].

A retrospective observational study conducted from 1998 to 2018 described that staged repair will be offered in patients with complex abnormalities, especially if they are newborns, premature, or weigh less than 5 kg. They can be operated on during the same hospitalization, following the correction of the most limiting pathology first, followed by the next anomaly. After a cardiovascular surgeon establishes ECMO or extracorporeal circulation, any cardiovascular anomalies are corrected, and a pediatric surgeon performs tracheoplasty. The reported criteria for surgical intervention in congenital tracheal stenosis include symptoms like persistent wheezing, recurrent respiratory infections, respiratory failure requiring endotracheal intubation, an inability to wean off mechanical ventilation, a degree of stenosis like long-segment stenosis and a segmental diameter of less than 50% of the average value, a potentially life-threatening dyspnea, and the need for endotracheal intubation [2, 4, 15].

Urgent surgery is suggested in cases with severe symptoms or repeated respiratory infections after controlling inflammation. Elective surgery is recommended for mild symptoms, managed conservatively at first, with surgical correction delayed until 6 months of age [2].

Factors associated with prolonged intubation have been reported to include low body weight and concurrent cardiovascular abnormalities. Factors associated with a higher likelihood of death include low body weight, preoperative advanced ventilation, prolonged extracorporeal circulation time, and the presence of granulation tissue [3, 16].

Heterogeneous factors can influence the outcomes, like postoperative tracheomalacia and restenosis. Associated factors of adverse outcomes include additional cardiovascular anomaly, carinal stenosis, and bronchial stenosis with concomitant carinal stenosis and compression. Preoperative morphological evaluation is of surgical importance, and with adequate preoperative recognition and close surveillance postoperatively, the intermediate outcomes in patients with the ring-sling complex are satisfactory [17, 18].

In the previously reported case, a patient with complex type 3 long-segment congenital tracheal stenosis, concurrent congenital heart disease with VSD and ASD, was surgically intervened in a single surgical session by the pediatric cardiothoracic service and pediatric thoracic surgery due to respiratory urgency with correction of the cardiac anomaly and the pulmonary artery sling.

Conclusion

We chose to perform a sliding tracheoplasty because we deemed it the best option given the case's complexity. We achieved excellent surgical and postoperative outcomes, a successful extubating process, and were discharged home without respiratory difficulty. Sliding tracheoplasty is a reproducible procedure with favorable outcomes in complex patients, as demonstrated in the previously described successful case.

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Authors' contributions
ELC and VSVG were responsible for data collection, management, sequence alignment, and initial manuscript drafting. JMF contributed to the clinical aspects of the study and provided patient treatment. JFN and JMU participated in patient treatment, sequence alignment, and manuscript drafting. CMLV and DAO contributed to the study design, performed the statistical analysis, and provided critical input. MMRG and SBG contributed to the study design, coordination, and manuscript drafting. All authors have read and approved the final manuscript.

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Availability of data and materials
The data and materials used in this study are available upon request. Due to restrictions on sharing the data publicly, a direct link to the dataset cannot be provided. However, all relevant data and materials necessary to replicate the study's findings will be made available to the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
This article presents a case report involving a pediatric patient with tracheal stenosis in a long segment and other cardiac defects. The study complied with the ethical principles outlined in the Declaration of Helsinki. Written informed consent was obtained from the patient's parents or legal guardians to publish their cases and accompanying images. As per their guidelines, the Hospital Inner Committee of the Mexican Institute of Social Security waived the need for ethics approval (17 CI 09 015 042). The confidentiality of patient information was strictly maintained, and all data were de-identified before analysis. This study adheres to the ethical standards of our institution and ensures the privacy and rights of the patients involved.

Consent for publication
Written informed consent was obtained from the patient's parents or legal guardians to publish their cases and accompanying images.

Competing interests
The authors declare that they have no competing interests.

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