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CASE REPORT

Pierre Robin sequence and double aortic arch: a case report☆

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Summary We present an 8-day-old female with two admissions for respiratory failure. On the first admission, the diagnosis of Pierre Robin sequence (PRS) and laryngomalacia was made after assessment with chest radiography, echocardiography, and flexible fiberoptic laryngoscopy. Four days after discharge, the child presented with stridor and respiratory distress, and a new cardiac murmur was noted after admission. Repeat echocardiography, with confirmatory direct laryngobronchoscopy, revealed a double aortic arch (DAA) with distal tracheal compression. This case illustrates the necessity of a complete otolaryngic evaluation, including direct laryngobronchoscopy, to search for a synchronous airway lesion in any neonate with severe respiratory distress associated with stridor.

1. Introduction

Pierre Robin sequence (PRS) and double aortic arch (DAA) are two conditions that are known causes of respiratory distress in the newborn period. We describe an infant whose diagnosis of DAA was delayed as a result of the concomitant presence of PRS and laryngomalacia. This case highlights the need for diagnostic persistence when one is faced with confounding clinical data that incompletely explains the patient's symptoms. It also identifies DAA as another congenital cardiovascular defect associated with PRS.

2. Case presentation

An 8-day-old infant female, born at 36 weeks estimated gestational age, presented to the Emergency Department (ED) with the chief complaint of 4 days of nasal congestion and "difficulty breathing".

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There was no report from the parents of any antecedent trauma, feeding intolerance, hoarseness or change in the quality of her cry. The parents did report "noisy breathing" since birth, which varied with position and was mostly associated with inspiration. In the ED, the patient received nebulized albuterol for wheezing and two normal saline boluses for poor perfusion. She remained tachypneic and mottled, so she was screened for sepsis with a complete blood count, and blood, urine and cerebral spinal fluid cultures. During the lumbar puncture, while being held in the side-lying and flexed position, the baby became apneic and cyanotic. She was resuscitated quickly, and was electively intubated prior to her transfer to the Pediatric Intensive Care Unit (PICU) due to persistent respiratory distress. Upon close physical assessment after her admission and stabilization, it was noted that she had features consistent with PRS, i.e. glossoptosis, micrognathia, high-arched palate, and monophasic inspiratory stridor, which was noted prior to intubation. No cardiac murmurs were appreciated on initial and follow-up exams during this admission. A detailed family history was notable for a deceased brother with Cornelia de Lange syndrome (CDLS). Our patient had no findings suggestive of CDLS aside from her micrognathia.

Admission venous blood gas while intubated on an FiO_2 of 0.80 revealed the pH was 7.21; $PaCO_2$, 75 mmHg; PaO_2, 47 mmHg; and SaO_2, 100%. Her serum bicarbonate was 30 mmol/L. She was assessed for other syndromic features, specifically Stickler syndrome and Velo-cardio-facial syndrome (VCSF), and none were found. Her renal ultrasound, head ultrasound, chest X-rays and echocardiogram were read as normal and chromosomal studies were sent. She weaned quickly off the ventilator to nasal cannula by hospital day # 2. After extubation, persistent positional inspiratory stridor was noted, particularly while lying supine. The otolaryngology (ENT) service performed flexible fiberoptic laryngoscopy (FFL) which showed mild laryngomalacia and peri-arytenoid edema and erythema. The subglottis and trachea were not visualized due to the limitations of bedside FFL. These findings were felt to be consistent with laryngopharyngeal reflux. She remained hemodynamically stable throughout this hospitalization and never required inotropic support. Her respiratory syncytial virus washing and viral and bacterial cultures all returned negative. Upon discharge on hospital day # 6, she was noted to have mild residual stridor that was associated with supine positioning and inspiratory in nature. She was feeding well and discharge was arranged. Her parents were given instructions for prone positioning, close parental monitoring, and empiric gastroesophageal reflux treatment with an H2 blocker, and she was expected to progressively improve as she grew.

Four days after her discharge from the PICU, she presented again to the ED with tachypnea, inspiratory stridor, retractions, and evidence of poor per-

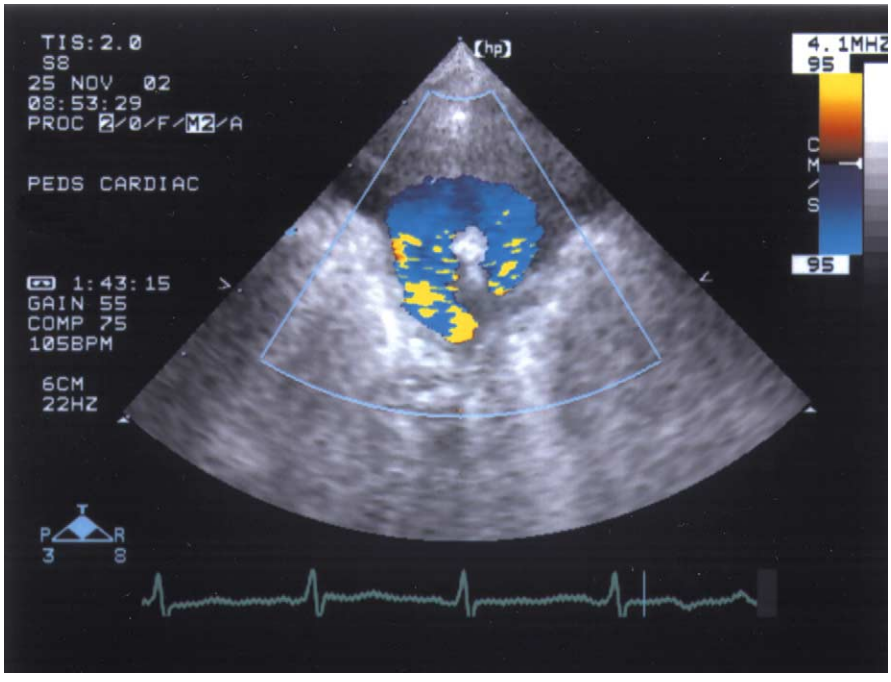

Fig. 1 Color doppler image of great vessels via echocardiogram in our patient with double aortic arch. Vascular ring is clearly demonstrated surrounding the trachea.