

When Wheezing is NOT Asthma: Recognizing Scimitar Syndrome

Kasey Lin¹, Kevin Lin¹, K K Patterson²

¹ Third Year Dental Student, Stony Brook University School of Dental Medicine, Stony Brook NY

² Faculty, Department of Orthodontics and Pediatric Dentistry, Stony Brook University School of Dental Medicine

ABSTRACT

A comprehensive physiologic assessment of a pediatric patient's upper and lower airways is crucial for ensuring safe and effective dental procedures. Asthma, a common sign being wheezing upon exhale, can impact airway function and influence dental treatment planning. Scimitar syndrome, a rare congenital anomaly affecting the cardiac and pulmonary systems, presents with specific abnormalities like right pulmonary artery and lung hypoplasia. Radiographic imaging reveals abnormalities of the pulmonary vein in the shape of a Turkish sword, or scimitar. Symptoms may include congestive heart failure, failure to thrive, and respiratory issues. Dental professionals should consider Scimitar syndrome in the differential diagnoses for patients of all age groups experiencing recurrent respiratory symptoms including wheezing not alleviated with bronchodilator therapy.

SYNDROME OVERVIEW

- The name "scimitar syndrome" originates from the distinct radiographic appearance of anomalous right pulmonary veins connecting to the inferior vena cava.
- Scimitar syndrome comprises a triad of characteristics: (1) curvilinear vascular density in the right lower zone; (2) hypoplastic right lung; and (3) dextroposition of the heart.¹
- Symptoms may manifest in infancy or later in childhood/adulthood. The infantile form typically presents within the first two months of life with symptoms such as tachypnea, recurrent pneumonia, failure to thrive, and signs of heart failure. This form of the syndrome is severe, often associated with significant cardiac abnormalities and pulmonary hypertension, resulting in a poor prognosis. In contrast, the diagnosis of the adult form is often incidental, and some patients may remain entirely asymptomatic.³

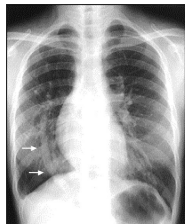


Figure A. Chest x-ray showing an abnormal shadow in the right lower lung field (arrows).⁴

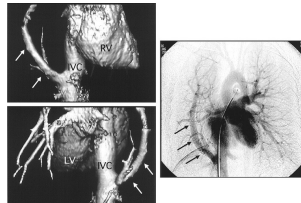


Figure B. Top left, 3D CT in right anterior oblique view. An anomalous vein can be visualized unequivocally (arrows). Bottom left, 3D CT in posteroanterior view. Arrows indicate the anomalous vein. Right, Late phase of pulmonary arteriogram with digital subtraction showing the anomalous pulmonary vein (arrows). IVC indicates inferior vena cava; RV, right ventricle; and LV, left ventricle.⁴

CASE REPORT: MEDICAL HISTORY

- 3-year-old Hispanic female presented with mother to SBU SDM April 2018
- Medical history included pulmonary hypertension, patent ductus arteriosus, partial anomalous right pulmonary veins pathognomonic of Scimitar Syndrome, and ectopic atrial tachycardia.
- Medication: Symbicort 2x/day
- Supplement: multivitamin with fluoride
- Surgical History:
 - 12/2014 Cardiac surgery for PDA repair and reimplantation of right pulmonary vein into left atrium
 - Diaphragmatic hernia repair
 - 5/12/2015 Removal of right lower lobe of lung
- No antibiotic prophylaxis required per cardiologist consulted on 10/2023

CASE REPORT: DENTAL HISTORY

- 3-year-old Hispanic female presented in 2018 for an initial appointment referred by outside dental provider due to uncooperative behavior for check up with severe early childhood decay. Patient was not a candidate for general anesthesia due to extensive cardiac history and weight per anesthesiologist. Treatment provided was SDF applications to teeth #B, D, E, F, L, S, and T.
- Patient returned in 2023 for a recall appointment followed by treatment planning appointment due to significant crowding and decay. Clinical exam and radiographs revealed: B root tip, H-F, I-MOL, 19-O, K-gross decay, L-root tip, S-gross decay, T-gross decay



Figure C. Panoramic Image September 2023

- Orthodontist reviewed the following treatment plan: extract remnants of primary teeth, Hyrax expander, deliver LLHA appliance, and monitor growth and development.
- Patient was cleared on 10/2023 for dental treatment by physician with the use of local anesthesia with epinephrine and the use of nitrous oxide without restriction.
- Pediatric dental resident completed the following:
 - Extractions of # C, H, I, L, K, S, and T
 - Sealants # 3, 14, 30
 - Composite Restoration # 19-O

LITERATURE REVIEW

- Scimitar syndrome is more prevalent in females and can exhibit familial patterns. The exact cause of the syndrome remains unclear, but it is believed to stem from a fundamental developmental abnormality affecting the entire lung bud in early embryogenesis.³
- Scimitar syndrome consists of multiple anomalies including right lung hypoplasia, dextroposition of heart, right lobe sequestration, horseshoe lung, pulmonary artery hypoplasia, anomalous pulmonary venous drainage from right lung to inferior vena cava (IVC) rather left atrium - Partial Anomalous Pulmonary Venous Return (PAPVR) and anomalous systemic arterial supply to right lung from aorta or its branches. These components lead to bronchiectasis and pulmonary hypertension which display clinically as wheezing.²
- To avoid progression to right ventricular failure, surgically rerouting the anomalous right pulmonary veins and repairing the associated cardiac defects, such as an ASD are warranted.³

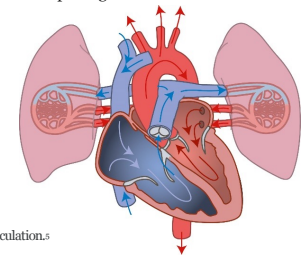


Figure D. Typical Pulmonary and systemic circulation.⁵

CONCLUSIONS

- Multidisciplinary care team consisting of pediatric pulmonologist, cardiologist, cardiothoracic surgeon and physiotherapist are essential in comprehensive medical care of persons with Scimitar syndrome.
- Dental professionals should be mindful of the final diagnosis in any child or adult patient with recurrent wheezing not alleviated by bronchodilator therapy, as all wheezes are not "asthma."

REFERENCES AND DISCLOSURES



Please scan QR code to access references.

The authors report no financial or other disclosures for this case report and discussion of Scimitar syndrome.