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Two-Dimensional Echocardiographic Diagnosis of Pulmonary Artery Sling in Infancy

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The vascular anomaly in which the left pulmonary artery arises from the right pulmonary artery and passes posteriorly and leftward between the trachea and the esophagus is termed a pulmonary artery sling. Two-dimensional echocardiograms were performed in five infants with this anomaly and successfully identified it in four, including one patient with truncus arteriosus communis.

The subxiphoid long-axis sweep was useful in identifying the origin and initial course of the left pulmonary artery, and short-axis subxiphoid views showed both its origin from the right pulmonary artery and its initial posterior course. Angulation toward the cardiac apex displayed the right pulmonary artery in cross section anteriorly and the left pulmonary artery in cross section posteriorly. A transducer orientation midway between the subxiphoid long- and short-axis positions was helpful in distinguishing a large right upper lobe branch of the right pulmonary artery from a pulmonary artery sling. The precordial short-axis plane displayed the origin and initial posterior and leftward course of the left pulmonary artery, while the bifurcation of the main pulmonary artery, usually easily seen in this view, could not be demonstrated.

Two-dimensional echocardiography offers a rapid, noninvasive diagnosis of pulmonary artery sling in infants.

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Pulmonary artery sling is an unusual and serious anomaly in which the left pulmonary artery arises from the right pulmonary artery and passes posteriorly and leftward between the trachea and esophagus, eventually reaching the left hilum. Affected infants generally present with respiratory distress during their first weeks or months of life, and mortality remains high despite current surgical techniques (1–3).

The diagnosis may be suspected or established with the aid of plain chest X-ray film (4), barium swallow (5), computed tomography (6), bronchoscopy (7), bronchography (8) or pulmonary arteriography (1–3). We present our experience with the use of two-dimensional echocardiography in the diagnosis of pulmonary artery sling in five infants.

Methods

Patients. A review of echocardiographic, surgical and autopsy files from January 1981 through June 1983 disclosed five patients with a confirmed diagnosis of pulmonary artery sling who had undergone cardiac ultrasound evaluation. In one patient (Case 1) the condition was diagnosed prospectively by echocardiography before other studies were performed. In a second patient (Case 2), the diagnosis was made by echocardiography after barium swallow, which had demonstrated anterior indentation of the esophagus. A third patient (Case 3) was transferred to our institution after diagnosis by pulmonary arteriography; the diagnosis was confirmed by two-dimensional echocardiography. A fourth patient (Case 4) was transferred to our institution at 1 month of age with a diagnosis at cardiac catheterization of truncus arteriosus communis and a right aortic arch. After corrective surgery the infant could not be weaned from the ventilator because of pronounced dyspnea and wheezing. Repeat cardiac catheterization demonstrated a pulmonary artery sling. In retrospect, the pre- and postoperative two-dimensional echocardiograms clearly demonstrated the pulmonary artery anomaly, which had not been appreciated initially. The final patient (Case 5) presented soon after birth with respiratory distress. A chest X-ray film showed a hypoplastic right lung with resultant hyperinflation of the left lung and dextrocardia. Echocardiographic examination failed to disclose cardiac anomalies. The infant died of progressive respiratory failure, and a pulmonary artery sling was discovered at autopsy.

Echocardiography. Two-dimensional echocardiograms were performed using either a Picker Echoview 80

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CI or a Diasonics Cardiovue 100. Standard subxiphoid long- and short-axis sweeps, as well as precordial long- and short-axis views, were employed. The subxiphoid long-axis sweep was initiated with the ultrasound sector in a transverse plane at the level of the diaphragm. The transducer was then angled superiorly and anteriorly. Particular attention was directed to the origin and course of the pulmonary arteries as they passed through the ultrasound plane. The transducer was then rotated 90° clockwise and a second slow sweep was performed from the plane of the venae cavae to the cardiac apex. The right pulmonary artery was carefully followed from the right hilum to its origin from the main pulmonary artery. A third subxiphoid sweep with the transducer rotated midway between the long- and short-axis positions was frequently useful for demonstrating the right upper lobe branch of the right pulmonary artery and, in the patient with truncus arteriosus communis, the initial course of the anomalous left pulmonary artery. A standard parasternal short-axis view was also employed to demonstrate the pulmonary artery anatomy.

Results

Cases 1 to 3. *Subxiphoid long-axis view.* The echocardiographic appearance of a pulmonary artery sling in a subxiphoid long-axis view varied depending on the angle of the proximal portion of the left pulmonary artery. If the initial course of this artery was superior and posterior, it was seen in its long axis arising from the right pulmonary artery (Fig. 1). If its initial course was directly posterior or inferior and posterior, it appeared as a circular or elliptical structure that, by slow cranial angulation of the transducer, could be shown to originate from the right pulmonary artery.

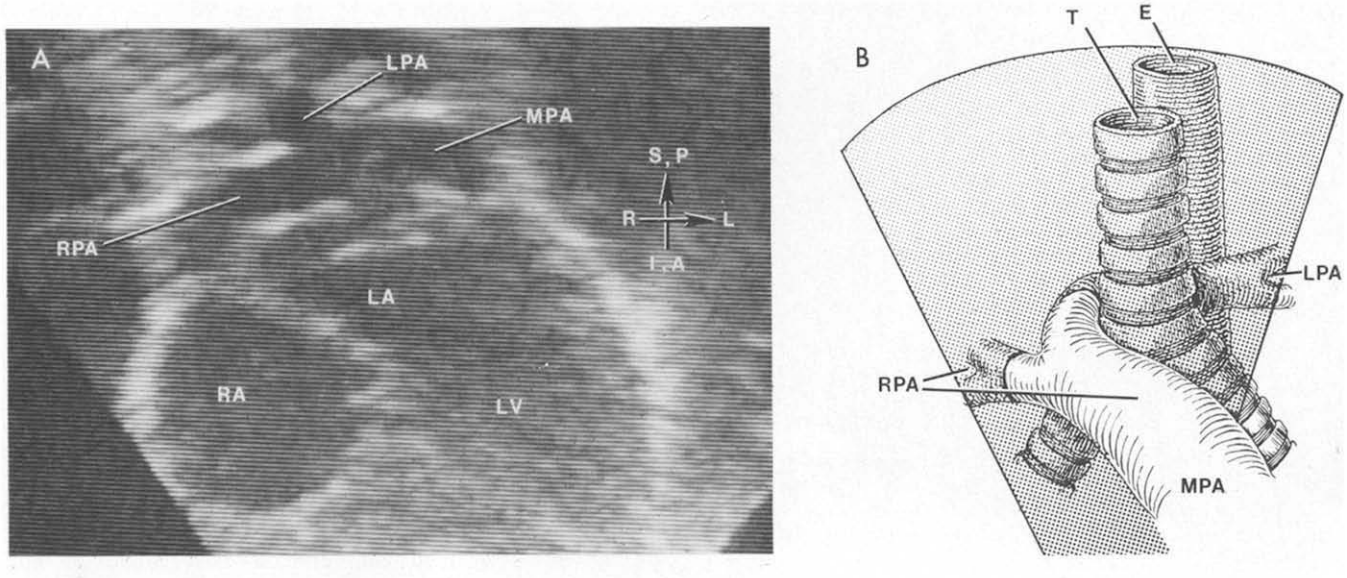
Subxiphoid short-axis view. In the subxiphoid short-axis view a vascular structure could be seen coursing posteriorly from the right pulmonary artery close to the position where the right pulmonary artery crosses behind the superior vena cava (Fig. 2). Angling the transducer toward the cardiac apex demonstrated the right pulmonary artery in cross section anteriorly and the left pulmonary artery in cross section posteriorly (Fig. 3).

Parasternal short-axis view. In the parasternal short-axis plane, the main and right pulmonary arteries could be seen in their usual locations, and the left pulmonary artery could be identified originating from the right pulmonary artery and coursing posteriorly and leftward (Fig. 4). The retrotracheal portion of the left pulmonary artery could not be visualized in any view because of interference from the intervening air column. In all echocardiographic sweeps, this artery was absent from its usual location.

Case 4. The right upper lobe branch of the right pulmonary artery and the anomalous left pulmonary artery could be imaged simultaneously in this patient with truncus arteriosus communis. With the transducer positioned midway between the subxiphoid long- and short-axis views, the upper lobe branch of the right pulmonary artery could be seen directed superiorly and to the right, while the anomalous left pulmonary artery was demonstrated posteriorly and to the left.

Case 5. The overlying hyperinflated left lung appeared to prevent identification of the pulmonary artery anatomy

Figure 1. Echocardiographic sector (**A**) and graphic representation (**B**) of the subxiphoid long-axis view appearance of a pulmonary artery sling. The left pulmonary artery (LPA) can be seen arising from the right pulmonary artery (RPA) and coursing superiorly (S) and posteriorly (P) in its proximal portion. Angulation of the transducer inferiorly would demonstrate the left pulmonary artery directed inferiorly (I) and leftward (L). A = anterior; E = esophagus; LA = left atrium; LV = left ventricle; MPA = main pulmonary artery; R = right; RA = right atrium; T = trachea.



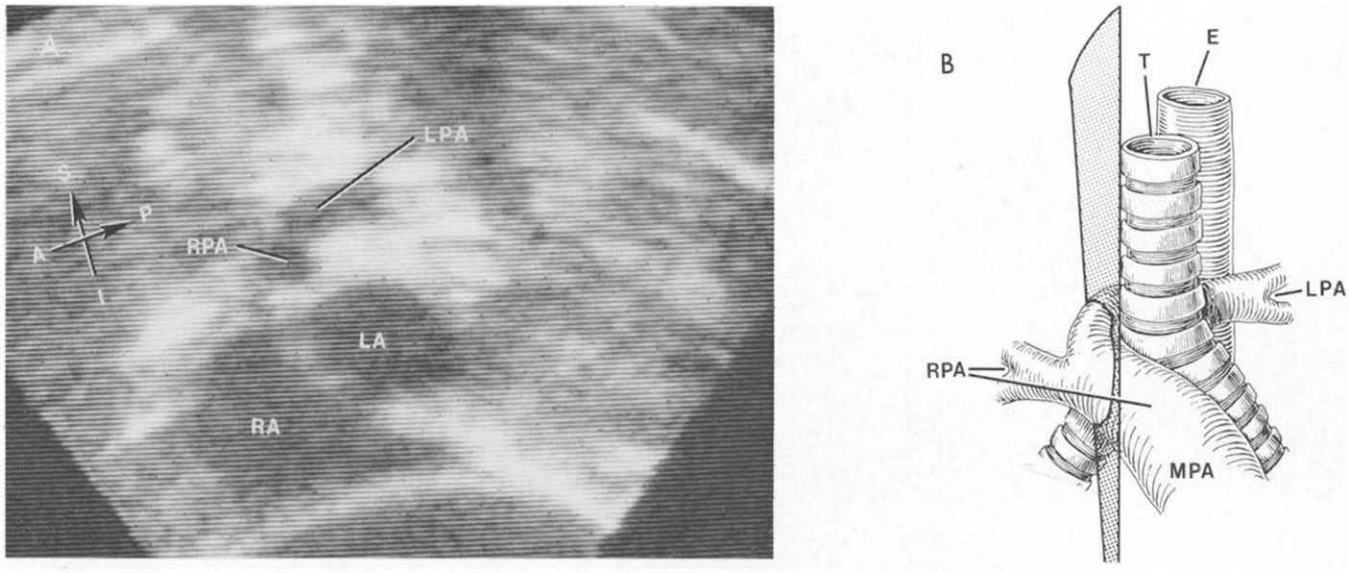


Figure 2. Echocardiographic sector view (**A**) and graphic representation (**B**) of the subxiphoid short-axis plane demonstrating the origin of the left pulmonary artery (LPA) from the right pulmonary artery (RPA) and the initial posterior course of the left pulmonary artery. Abbreviations as in Figure 1

in this patient, the only one whose condition was not diagnosed by two-dimensional echocardiography.

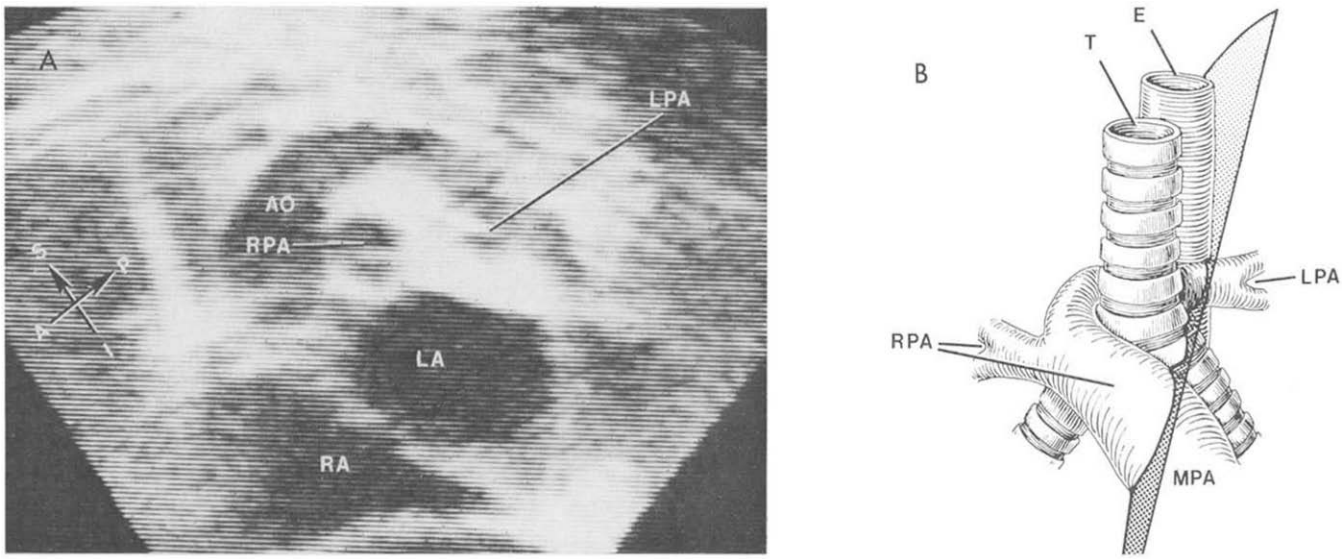
Discussion

Review of reported cases of pulmonary artery sling reveals an inhomogeneous group. Occasional reports of affected adults with minimal or no symptoms have appeared (9,10); more commonly, symptoms of respiratory distress develop soon after birth.

Tracheobronchial anomalies. Among infants with pulmonary artery sling, at least half have tracheobronchial abnormalities, particularly the presence of complete cartilaginous rings and associated hypoplasia of the distal trachea and bronchi (1,7,11). These infants would be expected to exhibit continued stridor despite relief of extrinsic vascular compression. Infants without airway malformations, whose symptoms relate solely to the vascular anomaly, are likely to derive considerable benefit from surgery.

Associated cardiovascular anomalies. Cardiovascular anomalies accompany pulmonary artery sling in approxi-

Figure 3. Echocardiographic sector view (**A**) and graphic representation (**B**) of the subxiphoid short-axis plane obtained by angling the transducer slightly toward the cardiac apex from the sector shown in Figure 2. The right pulmonary artery (RPA) is shown in cross section anteriorly and the left pulmonary artery (LPA) is shown in cross section posteriorly. AO = aorta; other abbreviations as in Figure 1



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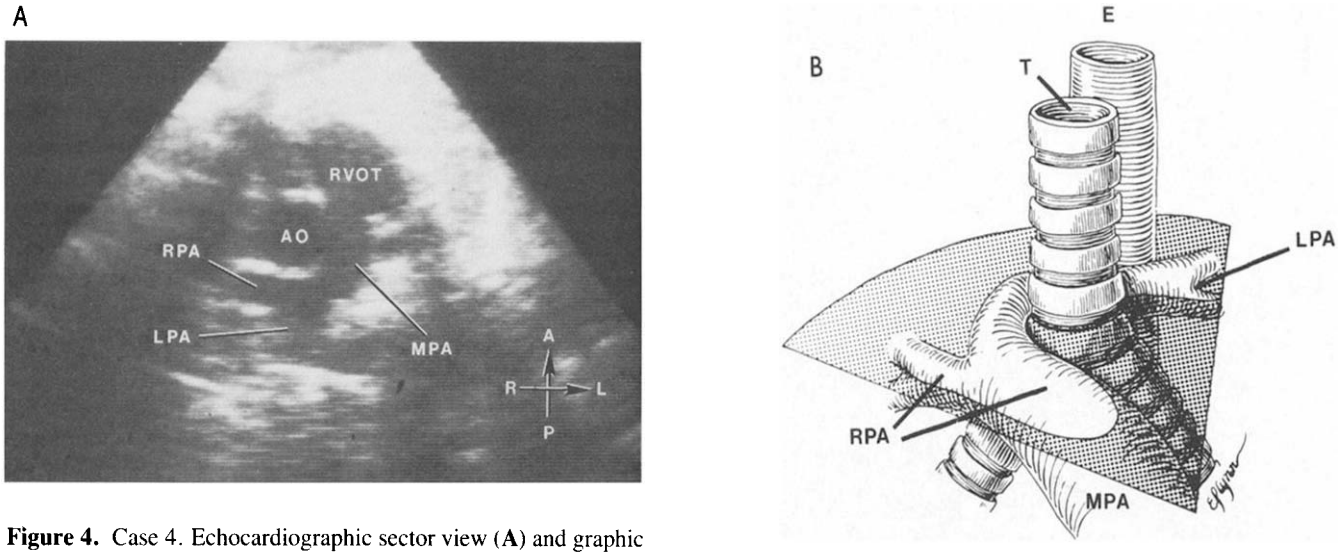

Figure 4. Case 4. Echocardiographic sector view (**A**) and graphic representation (**B**) of a pulmonary artery sling in the parasternal short-axis view. The left pulmonary artery (LPA) is seen arising from the right pulmonary artery (RPA) and coursing posteriorly (P) and leftward (L). The main pulmonary artery does not bifurcate in the usual fashion and the left pulmonary artery is absent from its usual location. AO = aorta; RVOT = right ventricular outflow tract; other abbreviations as in Figure 1.

mately 50% of reported cases and undoubtedly influence the surgical outcome (1–3). The most common associated malformations are patent ductus arteriosus, atrial septal defect, persistent left superior vena cava and ventricular septal defect. Conotruncal anomalies appear to be rare; a single case of tetralogy of Fallot with pulmonary artery sling has been described (12). We believe that our Case 4, the patient with truncus arteriosus communis, is the first to be reported with such an association.

Diagnostic methods. The diagnosis of pulmonary artery sling may be suspected on a plain chest X-ray film on the basis of evidence of obstructive emphysema, atelectasis, mediastinal shift or tracheal narrowing (4,7,8). Supporting evidence may be obtained by demonstrating anterior indentation of a barium-filled esophagus (5). Some authors (7,8) have advocated bronchographic and bronchoscopic studies of the tracheobronchial tree, which provide information that carries considerable prognostic significance. Cardiac catheterization and pulmonary arteriography provide the most sensitive and specific means of demonstrating a pulmonary artery sling and may be helpful in ruling out other cardiovascular defects.

We have found that with currently available ultrasound technology and careful technique, two-dimensional echocardiography is capable of demonstrating a pulmonary artery sling. Multiple subxiphoid and precordial echocardiographic views contribute to the diagnosis. It seems probable that echocardiography will prove to be a useful screening examination for patients in whom the diagnosis of pulmonary artery sling is being considered, obviating the need for more cumbersome or invasive studies such as barium swallow or computed tomography. If greater anatomic detail of the vascular structures is required, one may proceed directly to pulmonary arteriography. Bronchoscopy should also be considered to evaluate the tracheobronchial tree.

Echocardiographic pitfalls. Although a large upper lobe branch of the right pulmonary artery may simulate the echocardiographic appearance of a pulmonary artery sling in the subxiphoid long-axis sweep, clockwise rotation of the transducer should demonstrate the posterior and leftward course of a pulmonary artery sling as opposed to the superior and rightward course of a right pulmonary artery branch. Further rotation to the subxiphoid short-axis plane should demonstrate both the anterior right and the posterior left pulmonary artery in cross section, an echographic finding virtually pathognomonic for a pulmonary artery sling.

An additional echocardiographic pitfall may lie in the misinterpretation of a patent ductus arteriosus or a left atrial appendage as a normally positioned left pulmonary artery. Although this problem was not encountered in our patients, it probably could be resolved with the aid of contrast echocardiography or Doppler interrogation of the structure. A venous injection of saline solution would fail to opacify a left atrial appendage or a patent ductus arteriosus in the absence of right to left atrial or ductal shunting. Doppler examination would demonstrate flow from the aorta to the pulmonary artery through a patent ductus arteriosus (in the absence of pulmonary artery hypertension) and little or no flow in a left atrial appendage.

Suprasternal notch imaging. Although not employed in the examination of our five patients, suprasternal notch echocardiographic views should be similar to short-axis subxiphoid images and could be helpful in identifying the

initial posterior course of the anomalous left pulmonary artery as well as distinguishing a patent ductus arteriosus from a normally positioned left pulmonary artery.

Summary. Pulmonary artery sling should be considered in the differential diagnosis of any infant with unexplained stridor. Two-dimensional echocardiography appears to offer a rapid noninvasive means of diagnosis of this potentially fatal congenital anomaly.

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References

1 Sade RM, Rosenthal A, Fellows K, Castaneda AR. Pulmonary artery sling. J Thorac Cardiovasc Surg 1975;69:333–46

2. Grover FL, Norton JB, Webb GE, Trinkle JK. Pulmonary sling. J Thorac Cardiovasc Surg 1975;69:295–300.

3. Koopot R, Nikaidoh H, Idriss FS. Surgical management of anomalous left pulmonary artery causing tracheobronchial obstruction J Thorac Cardiovasc Surg 1975;69:239–46.

4. Williams RG, Jaffe RB, Condon VR, Nixon GW. Unusual features of pulmonary sling. Am J Radiogr 1979;133:1065–9.

5 Gumbiner CH, Mullins CE, McNamara DG Pulmonary artery sling. Am J Cardiol 1980;45:311–5.

6. Rheuban KS, Ayres N, Still JG, Alford B. Pulmonary artery sling: a new diagnostic tool and clinical review Pediatrics 1982;69:472–5.

7 Cohen SR, Landing BH. Tracheostenosis and bronchial abnormalities associated with pulmonary artery sling. Ann Otol Rhinol Laryngol 1976;85:582–90

8 Siegel MJ, Shackelford GD, McAlister WH Tracheobronchography in the evaluation of anomalous left pulmonary artery. Pediatr Radiol 1982;12:235–8.

9. Kale J, Rafferty RE, Carlton RW Aberrant left pulmonary artery presenting as a mediastinal mass Arch Intern Med 1970;125:121–5.

10. Dumler MP A rare cause of dysphagia JAMA 1966;197.513–4

11. Berdon WE, Baker DH, Wung JT, et al. Complete cartilage-ring tracheal stenosis associated with anomalous left pulmonary artery: the ring-sling complex. Pediatr Radiol 1984;152:57–64.

12. Jacobson JH, Morgan BC, Anderson DH, Humphreys GH. Aberrant left pulmonary artery: correctable cause of respiratory obstruction. J Thorac Cardiovasc Surg 1960;39:602–12