Tetralogy of Fallot with Severe Pulmonary Valvar Stenosis and Pulmonary Vascular Sling (Anomalous Origin of the Left Pulmonary Artery from the Right Pulmonary Artery)

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SUMMARY. A patient with the rare combination of tetralogy of Fallot with severe pulmonary valvar stenosis and pulmonary vascular sling is presented. The limitations imposed by pulmonary artery hypoplasia on the display of pulmonary vascular sling by conventional imaging techniques are discussed.

KEY WORDS: Pulmonary vascular sling — Anomalous origin of left pulmonary artery — Tetralogy of Fallot — Echocardiography — Ultrasonography

Anomalous origin of the left pulmonary artery from the right pulmonary artery or pulmonary vascular sling is a rare cause of respiratory distress in the newborn. However, it may be associated with significant morbidity, and, unrecognized, may lead to early death from the effects of chronic or recurrent respiratory infections.

We review a case of tetralogy of Fallot with severe pulmonary valvar stenosis and pulmonary vascular sling discovered at postmortem examination.

Case Report

This 1.3-kg infant was born at a gestational age of 32 weeks by spontaneous vaginal delivery. Soon after birth she was noted to have multiple congenital anomalies including duodenal atresia, anal atresia, and a single pelvic kidney. Additionally, the diagnosis of congenital heart disease consisting of tetralogy of Fallot with severe pulmonary valvar stenosis, hypoplastic pulmonary arteries (2-3 mm), and extensive collaterals was made by echocardiography.

On December 16, 1988, she underwent correction of the duodenal and anal atresia. Postoperatively, she developed hypoxemia and underwent a difficult reintubation of her trachea, thought to be related to the presence of subglottic stenosis or complete tracheal rings. Bronchoscopic evaluation by the otolaryngologists demonstrated distal tracheal stenosis. She underwent cardiac catheterization which demonstrated small confluent pulmonary arteries apparently supplied by large collateral vessels from the descending aorta to the left lung (Fig. 1).

After a period of stabilization, she was taken to the operating room for placement of a right modified Blalock-Taussig shunt utilizing a 3-mm Gore-Tex tube graft. There were no intraoperative complications. Postoperatively, positive pressure mechanical ventilation was continued. Nutritional support was provided by intravenous alimentation. Multiple attempts at extubation of the airway and liberation from ventilatory support were unsuccessful, associated with cyanosis, agitation, and apparent increase in bronchospasm. She could not be resuscitated following one such episode.

Postmortem examination of the heart and great arteries demonstrated the presence of a previously unsuspected anomalous origin of the left pulmonary artery from the right pulmonary artery (pulmonary vascular sling) in addition to tetralogy of Fallot with severe pulmonary valvar stenosis (virtual pulmonao' atresia), right aortic arch with aberrant left subclavian artery, and large systemic-to-pulmonary collaterals (Fig. 2). The right modified Blalock-Taussig shunt was intact and patent at necropsy.

Discussion

The presence of a pulmonary vascular sling is suggested in the newborn period by the early appearance of symptoms of respiratory distress, which may be the presentation of atelectasis or obstructive emphysema. Since the anomalous left pulmonary artery passes between the trachea and esophagus, symptoms of esophageal compression are usually not present. There are known anomalies of the tracheobronchial structures including complete carti-
laginous rings and hypoplasia of the distal trachea and bronchi which occur in approximately 50% of cases [1, 3, 9]. In addition, there may be associated cardiovascular anomalies which are variable, and also occur in approximately 50% of cases [4, 6, 9]. A single previous case associated with tetralogy of Fallot and pulmonary atresia was reported in a case series by Jacobson et al. in 1960 [5]. The occurrence of other conotruncal anomalies appears to be extremely rare. Yeager et al. [10], in a review of five cases of pulmonary artery sling, described a unique case of truncus arteriosus with associated anomalous left pulmonary artery. We believe that the current case is the only one with tetralogy of Fallot and severe pulmonary valvar stenosis in association with anomalous origin of the left pulmonary artery from the right pulmonary artery to be described in the era of modern diagnostic imaging.

The traditional approach to the diagnosis has relied on the observation of a mass between the trachea anteriorly and the esophagus posteriorly, sometimes visible on the plain chest roentgenogram. In addition, there may be unequal aeration of the lung fields [2]. Although not specific for pulmonary vascular sling, the presence of an oblique anterior indentation on the esophagus during barium esophagography is highly suggestive. Subsequent pulmonary angiography provides the standard method for outlining the aberrant branching pattern of the pulmonary arteries.

The association of hypoplasia of the pulmonary arteries in lesions with diminished blood flow across the right ventricular outflow tract is well known. In the case reported, the combination of prematurity, small size, associated tetralogy of Fallot with severe pulmonary valvar stenosis, and small pulmonary arteries made adequate angiographic display of the branching pattern of the pulmonary arteries impossible.

The review by Yeager et al. [10] demonstrated the utility of two-dimensional echocardiography utilizing multiple subxiphoid and precordial echocardiographic views in the diagnosis of pulmonary vascular sling.

Detailed retrospective scrutiny of the preoperative angiography demonstrated that the origin of the left pulmonary artery was indeed to the right of the trachea, indicated by the radioopaque marker of the endotracheal airway seen in Fig. 1. It is likely that this was not seen initially, since the left pulmonary artery filled first from an aorticpulmonary collateral vessel. Similarly, exhaustive review of the preoperative echocardiograms proved that the anomalous branching pattern and retrotracheal course of

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**Fig. 1.** Anteroposterior angiogram from an injection through a pigtail catheter in the upper descending aorta. The diminutive right and left central pulmonary arteries are well seen. The origin of the left pulmonary artery can be seen arising from the right pulmonary artery, and to the right of the trachea, which is marked by the endotracheal tube. *ETT,* endotracheal tube; *LPA,* left pulmonary artery; *RPA,* right pulmonary artery.

**Fig. 2.** Left posterior oblique photograph which has been printed backwards to display the anatomy as in the two-dimensional echocardiogram. The retrotracheal left pulmonary artery is clearly seen. The right modified Blalock-Taussig shunt is partially hidden under the right aortic arch; the proximal right pulmonary artery can be seen clearly at the origin of the left pulmonary artery. The anomalous left subclavian artery is seen as the fourth branch of the aortic arch. In this postmortem specimen, the left pulmonary artery has been flattened, giving the false impression that its diameter was larger than it measured in vivo. *Ao,* aorta; *LAA,* left atrial appendage; *LPA,* left pulmonary artery; *MPA,* main pulmonary artery; *RPA,* right pulmonary artery; *S,* right modified Blalock-Taussig shunt; *T,* trachea.