CONCURRENT VASCULAR RING AND OCCULT LEFT PULMONARY ARTERY ASSOCIATED WITH VENTRICULAR SEPTAL DEFECT: A REPORT OF AN UNCOMMON CONSTELLATION

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A vascular ring resulting from an anomalous right subclavian artery arising from the descending thoracic aorta in association with a right ligamentum arteriosus and left aortic arch is uncommon. Concurrent occult left pulmonary artery and

additional intracardiac defects are rare. We report the case of a patient who underwent successful repair of a ventricular septal defect (VSD) with relief of a vascular ring and reconstruction of a discontinuous left pulmonary artery.

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Fig 1. **A**, Intraoperative finding of vascular ring and occult left pulmonary artery (LPA). ARSA, anomalous right subclavian artery; RCA, right coronary artery; LCA, left coronary arteries; LSA, left subclavian artery. **B**, Elevation (arrow) of main pulmonary artery (MPA) flap for reconstruction of left pulmonary artery. **C**, Completed repair with relief of vascular ring.

Clinical summary. A 3.56-kg 3-week-old boy born at 37 weeks' gestation had intermittent dyspnea, pallor, and diaphoresis necessitating supplemental oxygen and continuous positive airway pressure through the nose. He was dependent on nasogastric nutrition because oral feeding aggravated his respiratory symptoms. Chest radiograph showed decreased left lung volume and markings. Trans-thoracic echocardiography confirmed a left aortic arch and a large perimembranous VSD. There was no coarctation or patent ductus arteriosus. No left pulmonary artery was visualized. Barium contrast radiography revealed posterior indentation suggestive of aberrant right subclavian artery or double aortic arch. Cardiac catheterization confirmed the VSD and demonstrated the aberrant origin of the right subclavian artery from the proximal descending thoracic aorta. Right ventricular contrast injection opacified a large main pulmonary artery and a right pulmonary artery, both with normal branching patterns. Pulmonary vein wedge injection revealed a discontinuous left pulmonary artery with normal intrapulmonary branching. The child was taken to the operating room on a semielective basis because he

could not be weaned from ventilatory support after catheterization.

Intraoperative findings revealed a hyperinflated right lung, a distended main pulmonary artery, and no intrapericardial left pulmonary artery. Intrapleural dissection demonstrated a distal tubular left pulmonary artery with ligamentous continuity arising from the undersurface of the aortic arch. On the right side there was a ductal ligament coming off the retro-esophageal anomalous right subclavian artery, which was arising from the descending thoracic aorta. This, combined with the left-sided aortic arch, effectively formed a vascular ring (Fig 1, A).

In the operation the right-sided ligamentum was divided and bypass was established. A pericardial window was created behind the left phrenic nerve to allow left pulmonary artery reconstruction. After aortic crossclamping and antegrade cardioplegic arrest the VSD was repaired transatrially with a bovine pericardial patch. The left pulmonary artery was disconnected from the left ligamentum. All ductal tissue was excised.

The anterior wall of the main pulmonary artery was developed as a flap hinged on the left posterolateral aspect to

bridge the gap to the left pulmonary artery (Fig 1, *B*). The left pulmonary artery was connected with the tongue of main pulmonary artery with interrupted 7-0 Prolene suture (Ethicon, Inc, Somerville, NJ) for the posterior layer. The anterior surface was created with a hood of autologous pericardial tissue held down by continuous 7-0 Prolene suture. The complete repair is illustrated in Fig 1, *C*. The chest was stented open and the patient was transferred to the intensive care unit. During the next 96 hours the patient required moderate inotropic and ventilatory support. The sternum was closed on the third postoperative day, and the patient was extubated on the seventh postoperative day. At follow-up after discharge the patient is progressing well and tolerates oral feeding without cyanotic episodes. Postoperative echocardiograms have consistently revealed adequate flow into the left pulmonary artery with good biventricular function.

Discussion. The rare combination of vascular ring and absent left pulmonary artery resulting in acute respiratory distress has been recently reported.¹ We are not aware, however, of any previous case reports depicting the combination of a left aortic arch with aberrant right subclavian artery to produce a complete vascular ring with a right ligamentum and an absent left pulmonary artery. A right-sided ductus or ligamentum producing a complete vascular ring in association with a left aortic arch is usually associated with a right descending aorta.² Our case represents a rare anatomic picture. Timing of initial presentation and severity of symptoms depend on the degree of tracheoesophageal compression.

Accurate delineation of the anatomy is imperative. Barium contrast radiography and transthoracic echocardiography to exclude intracardiac defects are compulsory. On a barium contrast radiograph a pulmonary artery sling produces an anterior indentation on the esophagus, whereas a vascular ring results in a posterior indentation. In difficult cases computed tomographic scanning or magnetic resonance imaging may be used in combination with bronchoscopy to assess the site and severity of tracheal stenosis. In our case angiography was needed to confirm the presence of the aberrant right sub-

clavian artery as a substrate for the complete vascular ring and to locate the left pulmonary artery.

McKim and Wiglesworth³ have demonstrated that hilar and intrapulmonary branches of the so-called absent pulmonary artery are in fact invariably present. A further study defined 2 patterns on the basis of the presence or absence of the intrapericardial pulmonary artery.⁴

In concurrence with our policy of single-stage repair for most complex defects, we proceeded with corrective repair in this case. The absence of inflow into the pulmonary artery should prompt an early intervention to restore flow if the distal vascular and gas exchange apparatus is to be preserved. In experimental animals ligation of the left pulmonary artery at birth caused hilar and axial pulmonary arteries to become atretic and often obliterated by 4 to 6 months of age despite satisfactory bronchial perfusion.⁵

We report for the first time a case of a complete vascular ring comprising an aberrant right subclavian artery arising from a left descending aorta and a right ligamentum arteriosus, with associated absent left pulmonary artery and VSD. This constellation of defects should be approached electively at the time of presentation, and single-stage repair can provide gratifying results.

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