Case Reports

Large Fetal Pulmonary Arteriovenous Fistula: Impact on Pulmonary Development

M.W. Russell, C. Gomez, C. Nugent, J. Christiansen
University of Michigan, F1310 MCHC/Box 0204, 1500 E. Medical Center Dr., Ann Arbor, MI 48109-0204, USA

Abstract. A case report of a patient with a large pulmonary arteriovenous fistula and valvar pulmonary stenosis is presented. The fistula was diagnosed prenatally and its effect on intrauterine cardiovascular growth and development documented. Due to concerns about massive intrapulmonary shunting potentially causing profound cyanosis after delivery, an EXIT (EX-utero Intrapartum Treatment) procedure was used to transfer the infant from placental to extracorporeal membrane oxygenation (ECMO) support. Severe pulmonary microvascular disease resulted in prohibitive pulmonary hypertension despite surgical ligation of the fistula. Prenatal and postnatal hemodynamic assessments of the fistula are presented and are compared to the pathologic findings.

Key words: Pulmonary arteriovenous fistula — In utero diagnosis — pulmonary development — Valvar pulmonary stenosis

Pulmonary arteriovenous fistulas are congenital communications between the pulmonary arterial and pulmonary venous systems that usually present with exertional dyspnea and cyanosis in the third or fourth decades of life [9]. The timing of the onset of symptoms is primarily dependent on the percentage of cardiac output that passes through the fistula. Extremely rarely, the fistula is of sufficient hemodynamic significance to cause profound cyanosis, acidosis and death in the neonatal period [4, 6]. In the present case, the development of a fetus with an unusually large pulmonary arteriovenous fistula (PAF) is tracked from 23 weeks gestation through delivery. The implications of this connection on pulmonary vascular development and on pre- and post-natal care are discussed.

Correspondence to: M.W. Russ; email: mruss@umich.edu

Fig. 1. Fetal echocardiographic image obtained during diastole at 23 weeks gestational age. Note the four chamber cardiac enlargement (RA, LA, RV, and LV denote the right and left atria and ventricles). Also indicated are the arteriovenous malformation (AVM), the descending aorta (Ao), right lung (RL) and placenta (P).

Case History

A patient presented to our institution with a 23 week gestational age fetus that had been determined to have marked four-chamber cardiomegaly on a screening obstetrical ultrasound. At the initial presentation there was severe, cardiomegaly with a cardiothoracic ratio of 0.73 (normal < 0.35), good biventricular function, markedly dilated right and main pulmonary artery, valvar pulmonary stenosis, a reverse-oriented patent ductus arteriosus, and an abnormal vascular connection in the right posterior lung field. A gradient of 42 mm Hg was obtained across the vascular connection that appeared to be multiple, small arteriovenous channels adjacent to the pleural surface. During the course of the pregnancy, there was progressive four chamber cardiac enlargement (Fig. 1), continued dilatation of the main and right pulmonary artery, and gradual formation of a pulmonary venous lake. However, there were never indications of impending heart failure; there was normal umbilical vessel and inferior vena cava flow patterns and no significant effusions.

Due to concerns about pulmonary arterial and venous hypoplasia, airway compromise, pulmonary parenchymal hypoplasia
was marked hypoplasia of the right middle, right upper and left pulmonary artery, and no significant contrast could be visualized in the pulmonary venous system of the left lung or the uninvolved segments of the right lung. Due to the large size and short length of the communication, non-surgical intervention via coil occlusion was determined to be higher risk than surgery.

The patient was taken for surgical ligation of the right lower lobe pulmonary artery proximal to the fistula. The friable nature of the ductus arteriosus necessitated institution of cardiopulmonary bypass and complete repair of the cardiac anomalies. The fistula, which arose from the very dilated main right pulmonary artery and drained directly into the left atrium via a large sinus cavity, was ligated and divided. The right pulmonary artery was reduced in size to relieve the tracheobronchial compression caused by its severe dilatation. Examination of the right upper, middle and lower lobe and left main pulmonary arteries showed them to be quite hypoplastic, accepting only 3-4 mm dilators. A valvotomy was performed to separate the fused raphe of the thickened pulmonary valve (Fig. 3). After rewarming, the patient was weaned from cardiopulmonary bypass and noted to have a pulmonary arterial pressure approximately 20 mmHg suprasystolic. The lungs accepted tidal volumes of 6-8 cc/kg with minimal end-tidal CO₂ return suggesting pulmonary parenchymal hypoplasia. Therefore, the patient was placed back on ECMO support and returned to the intensive care unit.

Two additional days of ECMO support failed to yield any progress in decreasing the amount of cardiopulmonary support required. The lung fields were extremely congested on chest X-ray and any weaning of the ECMO flow was accompanied by hypotension, hypoxia and decreasing mixed venous oxygen saturation. After discussion of the short and long-term prognosis with the family, the decision was made to discontinue ECMO support. Shortly after discontinuing support, the patient expired and an autopsy was performed.

On pathologic examination, the left pulmonary artery and left lung were noted to be markedly hypoplastic. There was diffuse bilateral congestion with hyaline membrane formation. The small arteries were noted to have marked medial hypertrophy and rare thrombi (Fig. 4).

Discussion

PAFs are thought to occur either secondary to incomplete degeneration of the vascular septae between the arterial and venous plexuses at the level of the pulmonary bud [1] or to a defect in the terminal capillary loops allowing dilatation and formation of thin-walled vascular sacs fed by a single artery and drained by a single vein [3]. Only rarely are they sufficient size to cause symptoms in the perinatal period. If a PAF is hemodynamically significant in the perinatal period, it is commonly fatal [7]. With prompt diagnosis and treatment, several cases of neonatal PAFs have been successfully treated by resection of the involved lung segment [2, 5]. However, it is evident that the present case had a greater compromise of pulmonary parenchymal and vascular development.

What is striking about this case are the secondary effects of the fistula on the growth of the heart and on the development of the pulmonary vascular system.