Selective Cyanosis of the Right Arm
Isolation of Right Subclavian Artery from Aorta with Bilateral Ductus Arteriosus and Pulmonary Hypertension

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SUMMARY. A sixteen-month-old child presented with cyanosis of the right arm. Investigation revealed bilateral persistent ductus arteriosus with isolation of the right subclavian artery from the aorta. Pulmonary vascular resistance and pulmonary arterial pressure were elevated so that the right subclavian artery received desaturated blood from the right pulmonary artery via the persistent right ductus arteriosus.

KEY WORDS: Bilateral ductus arteriosus — Pulmonary hypertension — Isolation of the right subclavian artery

Persistence of bilateral ductus arteriosus is embryologically an unusual occurrence and has been associated with interruption of the aortic arch [2, 4] or serious intracardiac defects [2-4, 6]. The following case is that of bilateral ductus arteriosus without associated cardiac anomalies that presented with the unusual physical finding of cyanosis of the right arm.

Case Report

The patient was born at 36 weeks' gestation and remained in the hospital for the first month of life because of "feeding difficulties." Recurrent episodes of respiratory distress and wheezing were noted, and the mother reported that the child's right arm frequently became cyanotic. An evaluation at 6 months of age revealed no significant murmur and a chest film at this time was normal. Continued respiratory problems required six hospitalizations. At 16 months of age congestive heart failure was recognized and further cardiac evaluation was obtained.

At that time the cyanosis of the right arm was obvious in relation to the rest of the body. A systolic ejection murmur was heard at the upper left sternal border, and the pulmonary component of the second heart sound was increased in intensity. Peripheral pulses were normal and blood pressures were equal in both arms. A chest roentgenogram showed cardiomegaly and prominent pulmonary vascularity that was equal on both sides. The ECG was compatible with right ventricular hypertrophy and right atrial enlargement.

At cardiac catheterization a left ductus arteriosus was demonstrated by the course of the catheter and an aortogram (Fig. 1). A right ductus arteriosus was demonstrated by a pulmonary artery angiogram and by a right brachial artery retrograde arteriogram (Fig. 2). The latter injection also filled the right vertebral artery, a normal subclavian artery branch. The right and left pulmonary arteries were in continuity. The aortic pressure was 95/50 mm Hg; left pulmonary artery pressure, 83/40 mm Hg; and right pulmonary artery pressure, 83/52 mm Hg. Because of differing oxygen saturations in each pulmonary artery, the pulmonary blood flow and pulmonary vascular resistance calculations were approximations. The calculated pulmonary vascular resistance was 50% of systemic resistance, and pulmonary blood flow was twice the systemic flow. The elevated pulmonary resistance was partially reversible with the use of tolazoline hydrochloride.

Because the shunt was predominantly left to right, ligation of the left ductus arteriosus was performed. Postoperatively, the child experienced intractable right heart failure and persistent cyanosis, which was partially alleviated by oxygen. A repeated catheterization three months after surgery revealed the pulmonary vascular resistance to be 80% of systemic resistance, but partially responsive to tolazoline as well as oxygen. Despite continued oxygen administration, the patient died nine months after surgery.

Autopsy

Abnormal findings were limited to the cardiovascular system and lungs. Pulmonary vascular obstructive disease was present with medial hypertrophy and luminal obliteration of vessels in both lungs equally. The right ventricular muscle was hypertrophied, with a thickness twice that of the left ventricle. There were no intracardiac anomalies. The left ductus arose in the usual manner and had been surgically ligated. The right ductus was continuous with the right subclavian artery, which was isolated from the aorta. The aortic arch was intact with otherwise normal branches.
Fig. 1. Injection of contrast material through a catheter located in the aortic arch demonstrates filling of the pulmonary artery through a left ductus arteriosus.

Fig. 2. Injection of contrast material into the right brachial artery demonstrates filling of the right pulmonary artery through a right ductus arteriosus.

Fig. 3. A Embryology of the normal left aortic arch. B Embryology of the case presented showing bilateral ductus arteriosus with the right subclavian artery arising from a persistent right ductus arteriosus. C Embryology of bilateral ductus arteriosus with persistence of the right fourth arch and isolation of the right pulmonary artery (PA).

Discussion

Symmetrically arranged aortic arches undergo accurately defined changes in the young embryo (Fig. 3A). Branches of the sixth arches become the right and left pulmonary arteries. In left aortic arch the connection between the right sixth arch and the right dorsal aorta is normally obliterated. This connection on the left persists as the ductus arteriosus. The right subclavian artery normally develops as a branch of the right dorsal aorta and connects to the aorta via the fourth right arch. In the case presented (Fig. 3B), the right fourth arch was obliterated and the right subclavian artery developed as a branch of the right dorsal aorta and connected to the pulmonary artery via the right sixth arch. Ultimately, the