We report the case of a male neonate who had a prenatal diagnosis of Fallot’s tetralogy. He presented with respiratory distress during the second week of life. Chest x-ray showed an enlarged right ventricle and pulmonary edema. Echocardiography demonstrated characteristic features of Fallot’s tetralogy. However, cardiac catheterization disclosed that, in this case of Fallot’s tetralogy, the left pulmonary artery had an anomalous origin in the ascending aorta. At 23 days of age, the patient underwent total surgical correction of the defects, during which a direct anastomosis was employed without conduit placement. 10 days later, he was discharged. We present the results of a literature review of the epidemiological, physiopathological, clinical, diagnostic, and surgical characteristics of this rare condition.

Key words: Cardiac surgery. Fallot’s tetralogy. Congenital heart defect.
PA trunk, along with direct closure of the ascending aorta and resection of the left PA and reimplantation on the fourth day of life, surgical repair was performed, consisting of an infundibular-valvular or subvalvular stenosis.

ventriculography showed a large subaortic ventricular septal defect with a heterologous pericardium patch. The infundibular muscle and anomalous bands of the RV were resected through an atrial approach. Postoperative echocardiography showed a gradient of 17 mm Hg at the left PA and RV pressures at 40% of systemic pressures. One week later, the patient was discharged.

**DISCUSSION**

Anomalous origin of a PA branch from the aorta is an extremely rare conotruncal malformation, accounting for only 0.12% of all congenital heart diseases. In most cases, the anomalous branch originates on the posterolateral wall of the ascending aorta, near the aortic valve. In 15%, the origin is distal, near the base of the innominate artery. Anomalous origin of the right branch is 5-6 times more frequent than the left. Most articles on anomalous origin of a PA branch from the aorta describe isolated clinical cases except for 2 series, which review 12 and 16 cases, respectively.

This condition can present alone or in association with other congenital heart defects; the most frequent is persistent ductus arteriosus. The heart disease most frequently associated with left-sided anomalous origin is Fallot’s tetralogy. Association with microdeletion of chromosome 22q11, as occurred in our patient, has only been described in one case, indicating that this anomaly pertains to the group of conotruncal cardiac malformations caused by anomalous development of the neural crest cells.

Only 9 cases of anomalous origin of the PA from the left aorta associated with Fallot’s tetralogy have been reported; the case we describe is the first reported in the Spanish medical literature.

The clinical presentation includes heart failure, heart murmurs, tachypnea, dyspnea, and repeated respiratory distress. Heart failure is the predominant symptom, with this being particularly serious when associated with coarctation or interruption of the aortic arch.

Two-dimensional color Doppler echocardiography is used for the initial diagnosis, although up to 15% of cases can go undetected. In our case, we could not detect the anomalous origin, probably because both branches were very close, as observed in Figure (B and D). The Doppler examination may be suggestive of anomalous origin, visualizing systodiastolic flow in the left PA, a phenomenon that we probably did not observe due to the elevated pulmonary vascular resistance of the newborn. Cardiac catheterization continues to be the definitive test for diagnosis, along with providing useful information for surgical planning.

Catheterization shows the systemic pulmonary pressures in the 2 arteries. The abnormally connected lung is perfused at a systemic pressure, whereas the other is exposed to the entire cardiac output from the RV. When
the pulmonary resistances drop during the neonate period, the pressure and flow within the anomalous PA increase, causing overcirculation and leading to pulmonary hypertension. The development of pulmonary hypertension in the lung normally irrigated by the pulmonary artery may be due to vasoconstrictive circulating substances or crossed neurogenic factors.

Early surgery within the neonatal period is preferable, as it would prevent pulmonary occlusive vascular disease that can develop quickly from age 3 months. The surgery consists of direct anastomosis of the anomalous branch to the pulmonary artery trunk. It may be necessary to expand the operated area with an autologous pericardium patch or interposition of a homograft. Techniques that use autologous tissues to widen and lengthen the anomalous origin of a PA branch from the aorta may be associated with a lower incidence of restenosis than direct anastomosis techniques, although the reported series are too small to clearly demonstrate this. At the present time, mortality is virtually nonexistent.

The most frequent postoperative complication is stenosis of the anastomosis (10.6%), which can develop months after surgery. Patients with severe restenosis may require surgical angioplasty, balloon dilatation or stent placement.

Anomalous origin of a PA branch from the aorta is a rare entity in which prompt diagnosis and surgery are essential to prevent irreversible vascular pulmonary disease. Follow-up to detect restenosis, which is frequent, is important.

REFERENCES


