Pediatr Cardiol 23:474–475, 2002
DOI: 10.1007/s00246-002-1433-4

Pediatric Cardiology

Transposition of the Great Arteries and Aortopulmonary Window in the Same Patient: Clinical Report and Follow-Up

V. Duca, G. Sulliotti, C. Maggio, G. Corsello
Divisione di Neonatologia e Terapia Intensiva Neonatale, Ospedale "V. Cervello", Università di Palermo, Via Trabucco, 180, 90146 Palermo, Italy

Transposition of the great arteries (TGA) has been reported in combination with several congenital defects. Only one case of TGA has been described in association with aortopulmonary window (APW) [1]. This patient died soon after surgery due to hemodynamic complications and pulmonary hypertension.

We report a case showing this rare combination with favorable clinical evolution. A full-term newborn presented with a systolic murmur, tachicardya, and tachipnea shortly after birth. Echocardiography revealed atrial situs solitus, concordant atrioventricular connections, and discordant ventricular–arterial connections (Fig. 1). The great vessels' spatial relationship was anteroposterior. An APW, originating from the posterior side of the ascending aorta, was seen entered the pulmonary artery trunk just below its bifurcation (Fig. 2). An unrestrictive patent foramen ovalis was also present. An arterial switch operation (ASO) was performed on day 26 with reimplantation of the coronary arteries in the usual fashion. The patent foramen ovalis was only partially closed, leaving a 4-mm opening (fenestrated) to allow for right-to-left shunting in the event of a severe pulmonary hypertensive crisis. On postoperative day 2 a pulmonary hypertensive crises did occur. She was successfully extubated on postoperative day 4. On day 16 after surgery she developed a triventricular posthemorrhagic hydrocephalus, be-

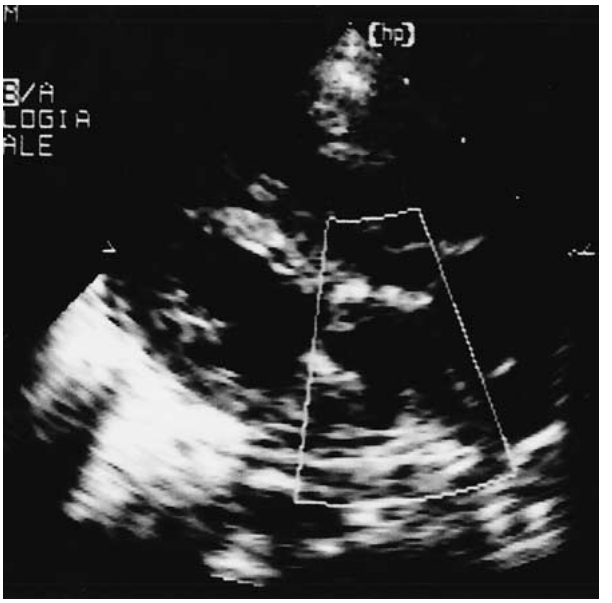

Fig. 1. Parasternal long axis: pulmonary artery arising from the left ventricle and large aortopulmonary connection.

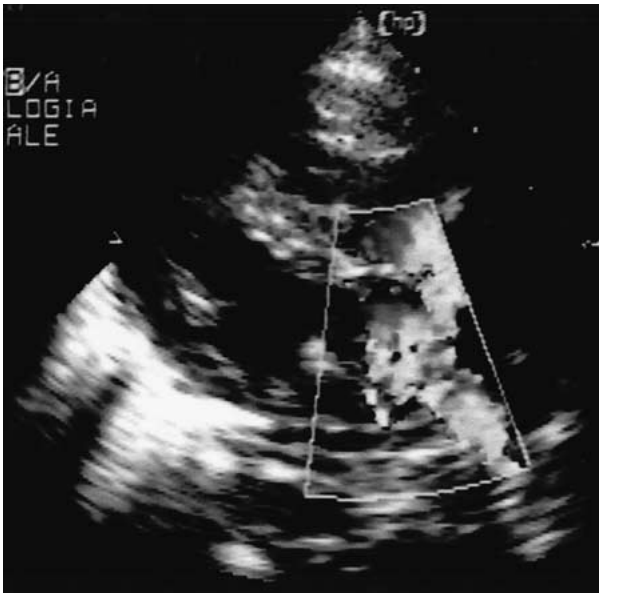

Fig. 2. Parasternal long axis: color doppler evidence of shunting throught the aortopulmonary window.

Correspondence to: G. Corsello

lieved to be secondary to a bleeding episode which may have occurred during the prolonged surgical procedure. A ventriculoperitoneal shunt was successfully inserted. Nine months after the neurosurgical procedures, she is stable and shows a normal developmental pattern.

The combination of TGA with APW has been previously reported in one patient who died in the postoperative period secondary to severe pulmonary hypertension. Although several techniques of patch closure and coronary flow diversion may be adopted in such conditions, a conventional ASO was safely performed in our patient. Patients with isolated TGA or APW are at risk for pulmonary hypertension postoperatively. The pulmonary capillaries become rapidly muscolarized and can react to various stimuli. In our patient, the risk of severe pulmonary hypertension was higher because of the combination of two defects. In order to minimize the manifestations of this potential adverse event, a 4-mm fenestration at the atrial level was left. Our opinion is that both early identification of complex heart defects and surgeon's experience are the main factors in performing a precocious and successful surgery, reducing the risk of complications.

Reference

1. Krishnan P, Airan B, Sambamurthy S. Rajani M, Rao IM (1991) Complete transposition of the great arteries with aortopulmonary window: surgical treatment and embryological significance. *J Thorac Cardiovasc Surg 101*:749–751