Case reports

Isolated anomalous origin of the right pulmonary artery from the ascending aorta (so-called "hemitruncus"): diagnosis and complex management in a newborn

Michele D'Alto, Carlo Pisacane, Giuseppe Santoro, Berardo Sarubbi, Emanuele Romeo, Maria Giovanna Russo, Raffaele Calabrò

Department of Pediatric Cardiology and Grown-Up Congenital Heart Disease (GUCH), Second University of Naples, Naples, Italy

Key words:
Ascending aorta; Congenital heart disease.

We report a case of an anomalous origin of the right pulmonary artery (RPA) from the ascending aorta diagnosed at echocardiography at 13 days of age. The diagnostic clue was relieved in the suprasternal and parasternal high short-axis views, showing aorto-RPA continuity with a systolic flow in the left pulmonary artery and a systo-diastolic flow in the RPA. At 34 days of age the infant was submitted to surgery during which a direct end-to-lateral anastomosis without conduit interposition was performed. During the short-term follow-up the patient developed RPA stenosis at the anastomosis site and underwent percutaneous stent implantation.

(Ital Heart J 2004; 5 (1): 61-63)

Received May 27, 2003; revision received October 20, 2003; accepted November 3, 2003.

Address:

Dr. Michele D'Alto

Via D. Fontana, 81
80128 Napoli
E-mail: mic.dalto@tin.it

An anomalous origin of one pulmonary artery from the aorta is an uncommon congenital heart anomaly almost always lethal without surgical correction[1-6]. It more commonly involves the right pulmonary artery (RPA) than the left and is more frequently associated with other cardiac anomalies such as patency of the arterial duct and tetralogy of Fallot[1,7]. Despite its rarity, it should be considered in any newborn with congestive heart failure and an increased blood flow[8-11]. An anomalous origin of the RPA could be prenatally diagnosed[12].

A 13-day newborn male, 3.2 kg in weight, was admitted to our Department due to tachypnea and tachycardia. At clinical examination he showed precordial hyperactivity, a gallop rhythm and a grade 2/6 systo-diastolic murmur at the high left sternal edge. The arterial pressure was 75/45 mmHg. The liver was palpable about 3 cm below the right-costal margin.

Chest radiography showed a moderate cardiomegaly and accentuated bilateral pulmonary vascular markings.

The 12-lead ECG showed sinus tachycardia (180 b/min), right-axis deviation and right ventricular hypertrophy.

Two-dimensional color Doppler echocardiography showed situs solitus of the viscera and atria and normal atrioventricular and ventricular-great vessel relations. Both ventricles were moderately dilated. Right ventricular systole was severely dysfunctional and the pressure in this was high (subsystemic). At the right oblique subxiphoid, suprasternal (Fig. 1) and parasternal high short-axis (Fig. 2A) views a continuity between the aorta and RPA was clearly shown. The diagnostic clue was relieved at color flow imaging, showing a systolic flow in the ascending aorta and in the left pulmonary artery (Fig. 2B) while a systo-diastolic flow in the RPA (Fig. 2C). This different pattern was very helpful in the diagnosis of an anomalous origin of the RPA from the ascending aorta. It was consequent to the different resistance of the systemic and pulmonary beds: the flow from the ascending aorta to the RPA was continuous because a significant pressure gradient persisted during diastole.

The patient was submitted to cardiac catheterization prior to surgical intervention. Left ventricular angiography confirmed that the RPA arose from the ascending aorta (Fig. 3). The systolic pressure in the right ventricle and left pulmonary artery approximated the systemic pressure.

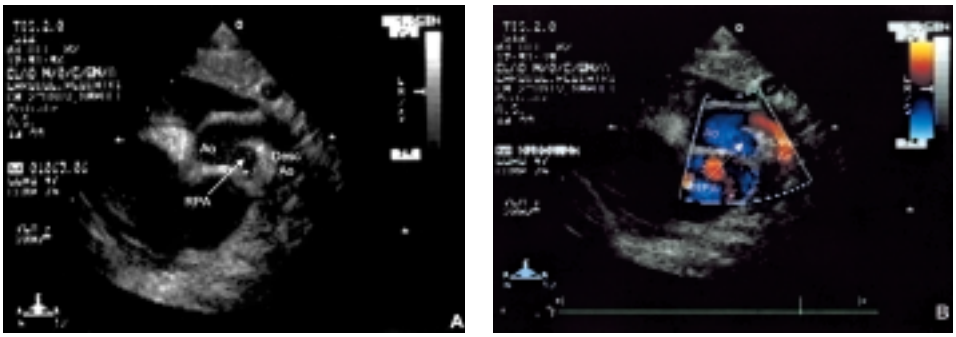

Figure 1. *Suprasternal view: two-dimensional* (A) *and color Doppler* (B) *imaging clearly show a continuity between the aorta (Ao) and the right pulmonary artery (RPA).*

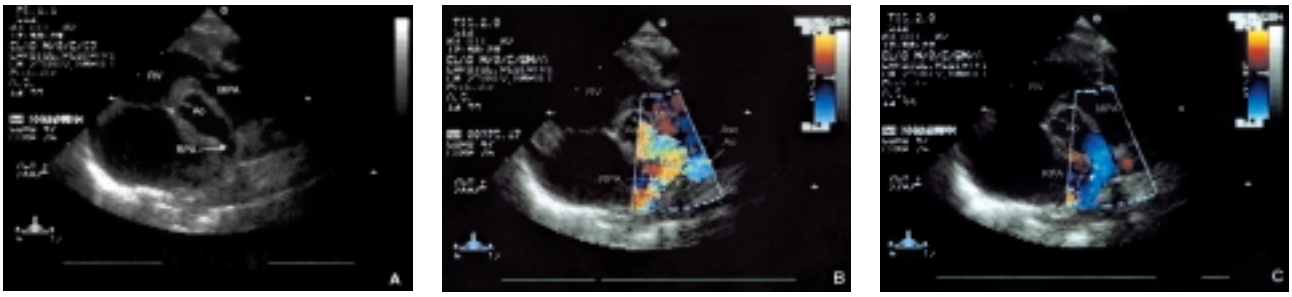

Figure 2. *Parasternal high short-axis view* (A) *shows a continuity between the aorta (Ao) and the right pulmonary artery (RPA). At color flow imaging, the systolic flow in the ascending Ao and in the left pulmonary artery* (B) *and a systo-diastolic flow in the RPA* (C) *may be clearly seen. MPA = main pulmonary artery; RV = right ventricle.*

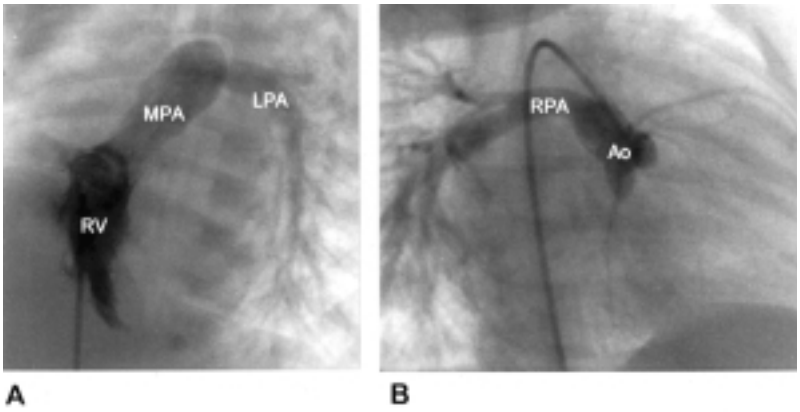

Figure 3. *Left ventricular angiography (*A: *left anterior oblique 60°, cranial 20°;* B: *right anterior oblique 30°, cranial 20°) confirms that the right pulmonary artery (RPA) arises from the ascending aorta (Ao). LPA = left pulmonary artery; MPA = main pulmonary artery; RV = right ventricle.*

At 34 days of age, the patient underwent a successful surgical reconstruction of the continuity between the main pulmonary artery and the anomalous RPA with an end-to-lateral anastomosis using cardiopulmonary bypass during hypothermic cardioplegia (28°C). A postoperative echocardiography demonstrated normal flow to the RPA.

The patient was discharged 7 days after surgical intervention.

At follow-up, echocardiography showed a critical restenosis at the site of the RPA anastomosis prompting us to perform cardiac catheterization with stent implantation (Jomed 12 mm, Beringen, Switzerland) dilated to 7 mm (Fig. 4) at the age of 6 months. Should restenosis occur, the implanted stent could be redilated up to 14 mm. Four months after stent implantation the child is alive and in good clinical conditions.

In the long-term follow-up, the anatomical and functional patient status could be assessed by means of computed tomographic scan and lung perfusion scintigraphy.

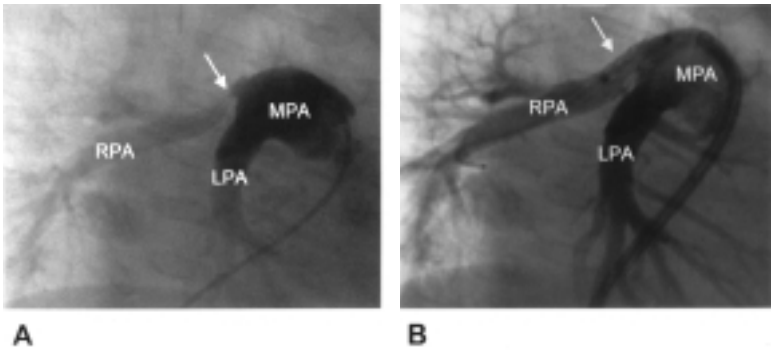

Figure 4. *The figure shows a critical restenosis at the site of the right pulmonary artery (RPA) anastomosis before stent implantation* (A) *and its resolution after stent implantation* (B). *LPA = left pulmonary artery; MPA = main pulmonary artery.*

References

1. Aru GM, English WP, Gaymes CH, Heath BJ. Origin of the left pulmonary artery from the aorta: embryologic considerations. Ann Thorac Surg 2001; 71: 1008-10.
2. Abu-Sulaiman RM, Hashmi A, McCrindle BW, Williams WG, Freedom RM. Anomalous origin of one pulmonary artery from the ascending aorta: 36 years' experience from one centre. Cardiol Young 1998; 8: 449-54.
3. Fucci C, di Carlo DC, Di Donato R, Marino B, Calcaterra G, Marcelletti C. Anomalous origin of the right pulmonary artery from the ascending aorta: repair without cardiopulmonary bypass. Int J Cardiol 1989; 23: 309-13.
4. Miyazaki K, Murashita T, Kubota T, Matsuzaki K, Miyazaki YJ, Yasuda K. Neonatal repair of anomalous origin of the right pulmonary artery from the ascending aorta. A case report and review in the literature. J Cardiovasc Surg 2000; 41: 863-8.
5. Prifti E, Frati G, Crucean A, Vanini V. A modified technique for repair of the anomalous origin of the right pulmonary artery from the ascending aorta. Eur J Cardiothorac Surg 2002; 22: 148-50.
6. Salaymeh KJ, Kimball TR, Manning PB. Anomalous pulmonary artery from the aorta via a patent ductus arteriosus: repair in a premature infant. Ann Thorac Surg 2000; 69: 1259-61.
7. Lin MH, Shen CT, Wang NK, Lin YM, Jeng CM. Magnetic resonance imaging of anomalous origin of the right pulmonary artery from the ascending aorta in association with ventricular septal defect. Am Heart J 1996; 132: 1073-4.
8. Kocis KC, Midgley FM, Ruckman RN. Aortic arch complex anomalies: 20-year experience with symptoms, diagnosis, associated cardiac defects, and surgical repair. Pediatr Cardiol 1997; 18: 127-32.
9. Matsubayashi K, Ueda Y, Ogino H, Matsumura M. A case of anomalous origin of the pulmonary arteries: right pulmonary artery from the descending aorta and the left pulmonary artery from the ascending aorta. Pediatr Cardiol 1999; 20: 298-300.
10. Tagliente MR, Troise D, Milella L, Vairo U. Isolated anomalous origin of left pulmonary artery from the ascending aorta. Am Heart J 1996; 132: 1289-92.
11. Trapali CJ, Thanopoulos BD. Severe right ventricular dysfunction in a neonate with aortic origin of the right pulmonary artery. Pediatr Cardiol 1998; 19: 425-7.
12. Jung MJ, Yoo SJ. Prenatal diagnosis of anomalous origin of the right pulmonary artery from the ascending aorta. Cardiol Young 2002; 12: 186-8.