Case reports

A rare form of interrupted aortic arch

Gabriella Agnoletti, Adele Borghi, Francesco Annecchino*

*Division of Cardiology, *Division of Cardiac Surgery, Ospedali Riuniti, Bergamo, Italy*

Key words:
Aortic atresia; Double aortic arch; Interrupted aortic arch; Persistent ventral aorta.

We report the case of a newborn with DiGeorge syndrome and aortic atresia associated with a complex anomaly of the aortic arch, interpreted as interrupted aortic arch type C, a persistent right ventral aorta and an aberrant right innominate artery. At 8 months the child underwent Norwood palliation with interposition of an 8 mm PTFE tube between the pulmonary trunk and the descending aorta and of a 3.5 mm shunt between the junction of the right ventral aorta to the left carotid artery and the right pulmonary artery. At 11 months he had substitution of the PTFE tube and bidirectional cavopulmonary anastomosis; he is now waiting for biventricular correction.

(Ital Heart J 2001; 2 (3): 228-230)

Received August 28, 2000; revision received December 21, 2000; accepted December 22, 2000.

Address:

Dr.ssa Gabriella Agnoletti

Via C. Battisti, 43C
25073 Bovezzo (BS)
E-mail:
gabriellaagnoletti@libero.it

Introduction

Interrupted aortic arch is a rare anomaly, sometimes associated with complex heart defects such as an aortopulmonary window, truncus arteriosus and transposition of the great arteries[1]. Interrupted aortic arch has also been described in association with aortic atresia[2,3] and in the setting of a double aortic arch[4]. However, to our knowledge, the association of an interrupted aortic arch with aortic atresia and persistence of the contralateral ventral aorta has never been described previously. We report a case of an infant with DiGeorge syndrome, aortic atresia, a complex anomaly of the aortic arch interpreted as interrupted aortic arch type C and persistence of the right ventral aorta, left descending aorta and aberrant right innominate artery.

Case report

A 2.920 kg, newborn male was referred, at the age of 4 days, for complex congenital heart disease. Facial features (micrognathia, short palpebral fissures, telecanthus) suggested DiGeorge syndrome. The respiratory rate was 60/min, the heart rate 160 b/min, and the systemic arterial pressure 90/60 mmHg. Peripheral pulses were normal; moderate hepatomegaly was present. At auscultation a single second sound and a grade 3/6 systolic murmur at the lower sternal border were identified.

Echocardiographic findings showed situs solitus, levocardia, concordant atrioventricular alignment, normal left and right ventricles, a large nonaligned ventricular septal defect, aortic atresia, severe hypoplasia of the ascending aorta, normal origin of the coronary arteries and continuation of the pulmonary artery into the descending aorta through a patent ductus arteriosus. The aortic arch was not visualized.

Cardiac catheterization, performed on day 6, showed two normal ventricles, a large nonaligned ventricular septal defect, aortic atresia, hypoplasia of the ascending aorta and interruption of the aortic arch after the left carotid artery (Fig. 1). The left subclavian and left vertebral arteries and the right innominate artery, dividing into the right subclavian and right carotid arteries, originated from the left descending aorta (Figs. 1 and 2). The ascending aorta was connected to the right carotid artery through an anterior vessel interpreted as a persistent right ventral aorta (Figs. 1 and 3).

At 12 days the child underwent Norwood palliation with interposition of an 8 mm PTFE tube between the pulmonary trunk and the descending aorta and of a 3.5 mm shunt between the junction of the right ventral aorta to the left carotid artery and the right pulmonary artery. Repeat cardiac catheterization showed good reconstruction of the aortic arch and a patent Blalock Taussig shunt. At 11 months the child underwent substitution of the PTFE tube and bidirec-

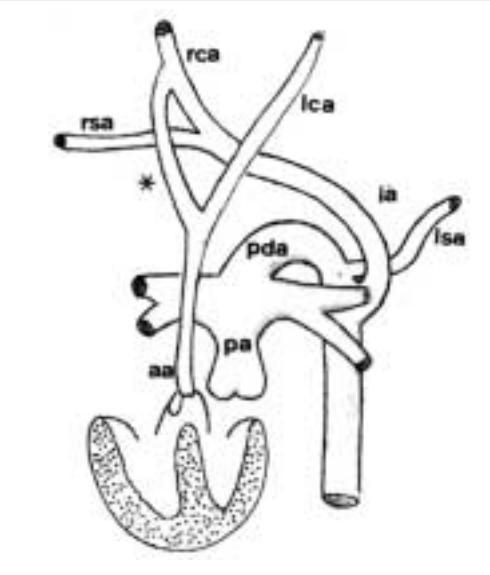

Figure 1. *Schematic representation: the ascending aorta continued into the right ventral aorta and left carotid artery. The aortic arch was interrupted between the left carotid and the left subclavian arteries. The ventral aorta was connected with the right carotid artery which, in turn, originated together with the right subclavian artery, from an aberrant right innominate artery. aa = ascending aorta; ia = innominate artery; lca = left carotid artery; lsa = left subclavian artery; pa = pulmonary artery; pda = patent ductus arteriosus; rca = right carotid artery; rsa = right subclavian artery. * ventral aorta.*

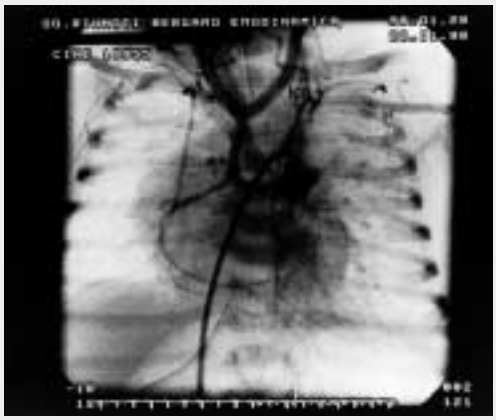

Figure 2. *Anteroposterior view. Stop flow injection in the descending aorta. A small ascending aorta was shown to give rise to two normal coronary arteries and to connect with the left common carotid artery and the right ventral aorta. Abbreviations and symbol as in figure 1.*

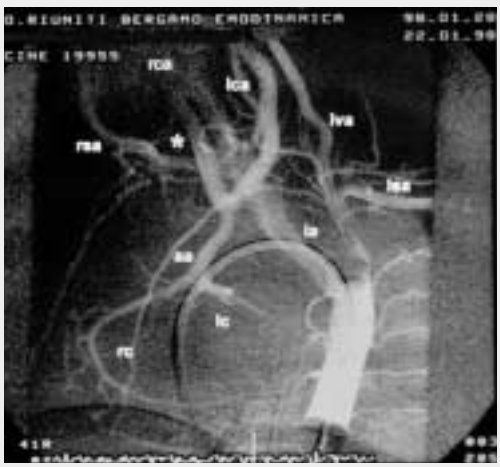

Figure 3. *Left anterior oblique view. Stop flow injection in the descending aorta. The ascending aorta continued into the right ventral aorta and the left carotid artery. The ventral aorta was connected with the right carotid artery. The aortic arch was interrupted between the left carotid and the left subclavian arteries. The right innominate artery originated from a left descending aorta. lc = left coronary artery; lva = left vertebral artery; rc = right coronary artery. Other abbreviations and symbol as in figure 1.*

tional cavopulmonary anastomosis. The child presents with normal body growth and is now waiting for biventricular correction. Genetic analysis confirmed DiGeorge syndrome (karyotype 46 XY, ishdel (22)(q11.2q11.2)(TUPLE 1-).

Discussion

Although interrupted aortic arch has been described in association with aortic atresia[2,3] and in the setting of a double aortic arch[3], to our knowledge the association of an interrupted aortic arch, aortic atresia and persistence of the contralateral ventral aorta has never been described previously.

A right aortic arch with an aberrant left innominate artery is a rare, but well known entity[5]; however, as far as we know a left interrupted aortic arch with a right aberrant innominate artery has not been reported.

In the case described, a left interrupted aortic arch coexisted with a persistent right ventral aorta, a right aberrant innominate artery and aortic atresia. The sequence of the brachiocephalic vessels was left carotid, left subclavian and right innominate artery. Embryologically this anomaly could be explained by regression of the right embryonic arch proximal to the left common carotid artery, with persistence of the right dorsal aortic root. The right dorsal aorta would supply the right seventh intersegmental artery (distal right subclavian) and the right third arch (right common carotid artery). The left ventral aorta would connect with the left third aortic arch, while the left fourth aortic arch would be reabsorbed. This case could not be interpreted as a double aortic arch with interruption of the left arch, as no persistent right dorsal aorta could be identified.

An interrupted aortic arch is often associated with major cardiac defects and multisystem noncardiac malformations. Linkage to deletion of 22q11.2[6] has already been shown. This deletion was also present in our patient.

References

1. Serraf A, Lacour-Gayet F, Robotin M, et al. Repair of interrupted aortic arch; a ten year experience. J Thorac Cardiovasc Surg 1996; 112: 1150-60.
2. Atik E, Cury P, Albuquerque AM. Aortic atresia with aortopulmonary window and interrupted aortic arch. Int J Cardiol 1998; 66: 217-21.
3. Ohye RG, Kagisaki K, Lee LA, et al. Biventricular repair for aortic atresia or hypoplasia and ventricular septal defect. J Thorac Cardiovasc Surg 1999; 118: 648-53.
4. Shufold WH, Sybers RG, Weens HS. The angiographic features of double aortic arch. AJR Am J Roentgenol 1972; 16: 125-40.
5. Garti IJ, Aygen MM. Right aortic arch with aberrant left innominate artery. Pediatr Radiol 1979; 8: 48-50.
6. Van Mierop LHS, Kutsche LM. Cardiovascular anomalies in DiGeorge syndrome and importance of neural crest as a possible pathogenic factor. Am J Cardiol 1986; 58: 133-7.