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Case report

Tetralogy of Fallot whit a "contralateral" ductus arteriosus

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Summary. Two neonates were taken shortly after birth to our unit with a prenatal diagnosis of [S,D,S] Tetralogy of Fallot with pulmonary atresia and "unusual" aorta to pulmonary connection. The echocardiogram confirmed the main diagnosis showing: a left aortic arch with a vascular connection between the right innominate artery and the origin of the right pulmonary artery in patient A; and right aortic arch with a vascular connection between the left innominate artery and the origin of the left pulmonary artery in patient B. (www.actabiomedica.it)

Key words: congenital heart disease (CHD), aortic arch, Tetralogy of Fallot

Case Report

According to the Edwards hypothetical double arch model, each embryo has two ducti arteriosi, derived from the right and left sixth pair of aortic arches, respectively. Usually, when a left aortic arch develops the left ductus arteriosus (homolateral) persists, whether when a right aortic arch develops the right ductus persists instead. Nevertheless rarely, the homolateral ductus regresses, and the contralateral may persist as an alternative, thus connecting the base of the subclavian artery (the one branching from the innominate artery) and the pulmonary bifurcation (1).

We present two neonates with prenatal diagnosis of [S,D,S] Tetralogy of Fallot with pulmonary atresia and left aortic arch with right ductus arteriosus: between the right subclavian artery and the origin of the right pulmonary artery in patient A; and right aortic arch with left ductus arteriosus: between the left subclavian artery and the origin of the left pulmonary artery in patient B (Fig. 1).

Both patients successfully underwent central BT-Shunt and ductus ligation and completed the repair successfully after one year and 18 months respectively.

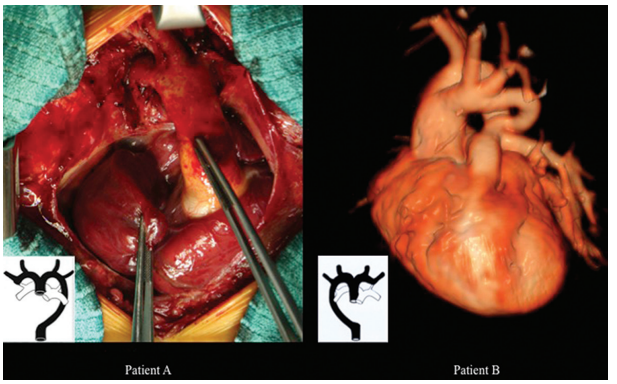

Figure 1. Patient A: Surgical view. Patient B: 3D Computerized tomographic angiography

"Contralateral" ductus arteriosus means: right ductus arteriosus in a left aortic arch (Patient A) and left ductus arteriosus in right aortic arch (Patient B).

Embryologically there are potentially two ducti arteriosi: one "usual" or "homolateral" to the aortic arch, coming off the aortic isthmus; a second one "unusual" or "contralateral" to the aortic arch, coming off the innominate artery.

Therefore patient A has a left aortic arch (to the left of the trachea, riding the left bronchus), with a

right ductus arteriosus from the right subclavian artery to the right pulmonary artery (to the right side of the trachea) (2). Patient B instead has a right aortic arch (to the right of the trachea, riding the right bronchus), with a left ductus arteriosus running from the left subclavian artery to the left pulmonary (to the left side of the trachea) (3).

Discussion

Differential diagnosis in such patients should be performed between a small tubular and very high aortopulmonary window, Major Aorto-pulmonary collateral artery (MAPCA), persistent 5th aortic arch (systemic to pulmonary connection variant), homolateral or controlateral ductus arteriosus. MAPCA is a not PGE1 responsive tortuous vascular structure originating from the descending aorta and with a persistent 5th aortic arch, "systemic-to-pulmonary variant", which runs from the distal ascending aorta to the pulmonary bifurcation, but parallel or homolateral to the aortic arch (same tracheal side) (4).

Persistent 5th aortic arch appears as a vascular structure running inferior and parallel to the "real" aortic arch from the innominate artery to the left subclavian artery (5).

It can be associated with major congenital heart malformations involving the systemic or the pulmonary circuits.It usually has no clinical significance but can be either, beneficial as in systemic outflow tract obstructions or cause hemodynamic compromise when associated with a significant left to right shunt (6).

An early diagnosis may improve outcomes in such kind of anatomy and should be every time checked to avoid complication during staged approach for Tetralogy of Fallot/PA-VSD and could be associated with important multisystem morbidity and mortality (7, 8).

Conflict of interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article

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