Cardiology in the Young (2011), **21**, 468–470
doi:10.1017/S1047951111000217

Brief Report

Anomalous origin of the right coronary artery from the pulmonary artery associated with tetralogy of Fallot: description of the pre-surgical diagnosis and surgical repair

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Abstract Anomalous origin of the right coronary artery from the pulmonary artery is a rare congenital defect. We describe the case of an infant with anomalous origin of the right coronary artery from the pulmonary artery in association with tetralogy of Fallot. This patient had a pre-operative echocardiographic diagnosis, which was confirmed by angiography, and later underwent a successful surgical repair.

Keywords: Congenital cardiac disease; echocardiography; cardiovascular surgical procedures

Received: 6 September 2010; Accepted: 6 January 2011; First published online: 2 March 2011

THE ANOMALOUS ORIGIN OF THE RIGHT CORONARY artery from the pulmonary artery is an extremely rare congenital cardiac defect. Most frequently presenting in isolation, it can also be associated with other congenital cardiac malformations, and, if not diagnosed, it complicates the surgical correction. We present the case of an infant with tetralogy of Fallot in whom an anomalous origin of the right coronary artery from the pulmonary artery was diagnosed at the echocardiographic evaluation preceding the corrective surgery and confirmed by angiography.

Case report

A 4-month-old infant with tetralogy of Fallot was admitted to our department for surgical repair. The diagnosis, which was made on prenatal ultrasound and confirmed at birth, consisted of tetralogy of Fallot with severe anterior and rightward infundibular septal deviation and a peak systolic gradient across the right ventricular outflow tract of almost 100 millimetres of mercury. Despite the severe narrowing of the right ventricular outflow tract, the oxygen saturation was 88% on room air and no hypoxic spells occurred during the first months of life.

The electrocardiogram and chest X-ray were consistent with the diagnosis of tetralogy of Fallot.

A complete echocardiographic examination revealed, in addition to the anatomical features of the main malformation, an unusual brightness of the right ventricular endocardium, suggestive of endocardial fibroelastosis (Fig 1a). Furthermore, multiple colour spots within the myocardium of the interventricular septum were evident at colour flow evaluation (Fig 1c). These unexpected findings led the examiner to carefully assess coronary artery anatomy. From a short-axis view at the base of the heart, the left ostium appeared to arise from a clockwise-rotated aortic root in a posterior position. Colour flow mapping showed diastolic antegrade flow from the aortic sinus towards the left main coronary artery and the anterior-descending and circumflex arteries. The origin of the right coronary artery from the aorta was not visible, and diastolic reverse flow was evident with colour flow interrogation of its proximal segment (Fig 1b). The turbulent

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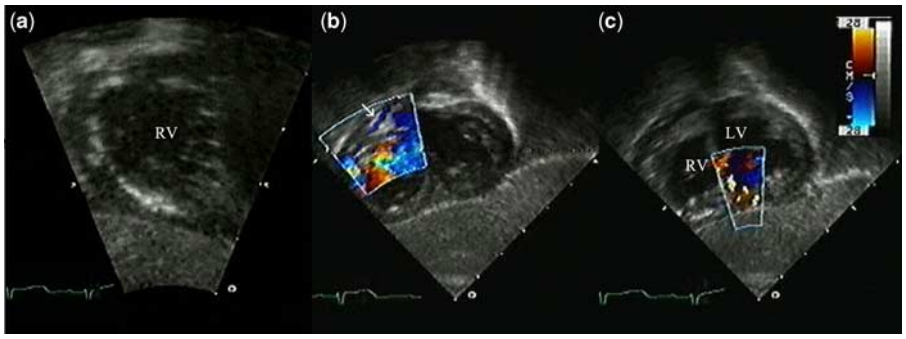

Figure 1.
*Subxiphoid short-axis view: (**a**) endomyocardial fibroelastosis of the diaphragmatic wall and of the lateral free walls of the right ventricle (RV); (**b**) colour Doppler shows diastolic reverse flow in the proximal right coronary artery (arrow); (**c**) colour Doppler shows the intercoronary collateral circulation between the circumflex and the right coronary artery (RV = right ventricle; LV = left ventricle).*

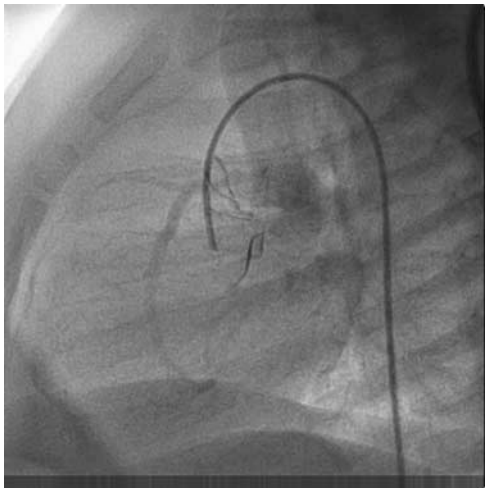

Figure 2.
Aortogram in latero-lateral projection showing the pulmonary origin of the right coronary artery.

flow in the main pulmonary artery, due to pulmonic valve stenosis, masked the reverse flow from the anomalous right coronary artery into the low-pressure pulmonary bed. An aortogram confirmed the echocardiographic diagnosis of anomalous origin of the right coronary artery from the pulmonary artery (Fig 2).

Surgical tetralogy of Fallot repair included bicuspid pulmonary valve commissurotomy; right ventricular outflow tract myotomy/myectomy; trans-atrial ventricular septal defect patch closure; reconstruction of the pulmonary valve cusp with 0.1 polytetrafluoroethylene membrane; and patch augmentation of the main pulmonary artery.

The right coronary artery was isolated and dissected from the pulmonary trunk and subsequently re-implanted into the aorta with 8.0 polypropylene sutures. The surgical report described the anomalous right coronary artery as being thin-walled and dilated. The post-operative course was uneventful, and the infant was extubated 24 hours after surgery and discharged home on post-operative day 7.

Discussion

The anomalous origin of the right coronary artery from the pulmonary artery is a rare congenital defect affecting 0.0023 live births.[1] Even more unusual is the association with tetralogy of Fallot, which appears in the literature only in individual case reports.[1–8] As far as we know, this is the first case in which the diagnosis was established by echocardiography before surgery. Nowadays, the practice of performing surgical corrections of complex congenital malformation in infancy without the aid of an angiographic confirmation of the clinical diagnosis is widespread. Unfortunately, in some cases, this practice can lead to underdiagnosis of rare associated malformations, and thereby compromise the surgical result.

In foetal life, blood flows from the pulmonary artery to the anomalous coronary artery because of the elevated pulmonary vascular resistance, giving sufficient oxygen delivery to the myocardium. After the falling of the pulmonary vascular resistance, in the first days of life, a process of collateralisation might produce a steal phenomenon into the coronary circulation. Moreover, in our case, the infundibular stenosis and consequently the right ventricular hypertrophy might have increased the myocardial demand, influencing the potential for myocardial ischaemia.[3]

The electrocardiogram was not helpful in the diagnosis of ischaemic lesions, showing the classical electrical finding of tetralogy of Fallot (right axis deviation and right ventricular hypertrophy) without any ST-T- or T-wave alterations suggestive of ischaemia. Therefore, only a careful

https://doi.org/10.1017/S1047951111000217 Published online by Cambridge University Press

echocardiographic study evaluating the direction of blood flow into the right coronary artery could give us the clue to the diagnosis. The unusual hyperechoic changes of the right ventricular endocardium and the abundant colour flow spots into the interventricular septum, due to the well-developed intercoronary collateral circulation, acted to raise the suspicion of the examiner.

In conclusion, what this successful case taught us is that it is only by a critical and thorough evaluation of all the clinical and echocardiographic aspects of each individual patient that fatal errors can be prevented. Furthermore, we suggest that even in centres that have adopted a routine non-invasive pre-surgical diagnostic strategy, cardiac catheterisation with angiography is always mandatory when clinical data do not fit the echocardiographic diagnosis.

Acknowledgement

The authors have no financial and/or conflict of interest to declare.

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