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Double aortic arch in a patient with Fallot's tetralogy

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A THREE-MONTH-OLD FEMALE CHILD WITH tetralogy of Fallot was catheterized in the course of preoperative preparations for corrective surgery. At previous echocardiography, we had detected a left-sided aortic arch, and did not suspect the presence of a double aortic arch. Her history revealed mild respiratory stridor, with no cyanotic spells. Physical examination showed the typical findings of tetralogy of Fallot, with mild cyanosis and saturations of oxygen of 90 percent at rest. Cytogenetic analysis revealed a microdeletion of the short arm of chromosome 22.

An angiogram following an injection in the right ventricle (Fig. 1) showed right-to-left shunting across the ventricular septal defect into the overriding aorta, right ventricular hypertrophy, and subpulmonary and valvar pulmonary stenosis. Additionally, it revealed the presence of a double aortic arch, with both arches being patent, and of about equal size. The angiogram performed in the left ventricle (Fig. 2), confirmed left-to-right shunting across the ventricular septal defect, and provided a more detailed view of the double aortic arch. It also showed the descending thoracic aorta to be left-sided.

At open-heart surgery, the ventricular septal defect was closed, and the obstructed right ventricular outflow was relieved by pulmonary valvoplasy and patch augmentation of the ventriculo-pulmonary junction. The right aortic arch was divided at the origin of the right subclavian artery. Recovery was uncomplicated, and follow-up at 18 months showed mild pulmonary

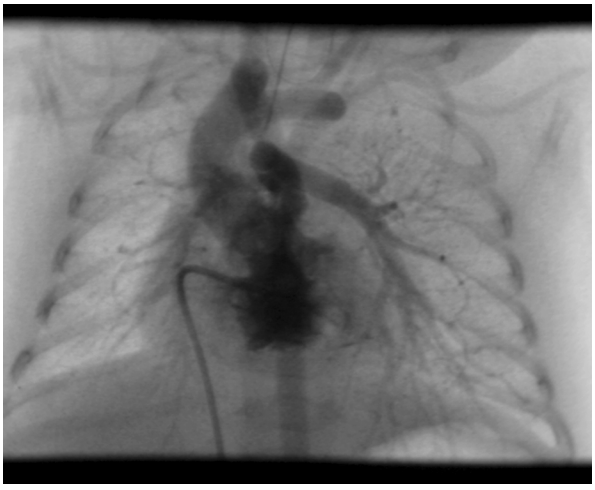

Figure 1.

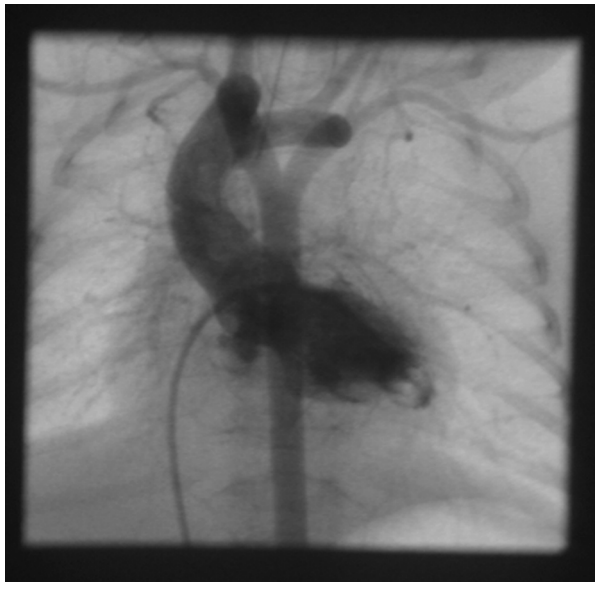

Figure 2.

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valvar stenosis and some regurgitation, but no residual intracardiac shunting.

Double aortic arch is itself a rare malformation, which occurs infrequently in combination with Fallot's tetralogy. It is possibly more common among patients with associated deletion of chromosome 22q11, who have a higher incidence of abnormal branching of the aorta.[1] The importance of diagnosis lies in its recognition preoperatively in the setting of tetralogy, particularly in the current era of early primary correction without invasive preoperative investigations. We still perform diagnostic cardiac catheterization routinely as a preoperative investigation for infants under consideration for primary repair of tetralogy of Fallot. In this instance, our policy permitted the important associated anomaly to be diagnosed in timely fashion.

Please refer to the Cambridge Journals Online website at: http://journals.cambridge.org/abstract_PII for associated animated movie clip.

Reference

1. McElhinney DB, Clark BJ, Weinberg PM, et al. Association of chromosome 22q11 deletion with isolated anomalies of aortic arch laterality and branching. J Am Coll Cardiol 2001; 37: 2114–2119.

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