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Brief Report

Aortic stenosis after uncomplicated surgical repair of tetralogy of Fallot

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Abstract Tetralogy of Fallot is only rarely associated with aortic valvar disease. We present a child who had uncomplicated repair of tetralogy of Fallot at 16 months of age, and who developed mild aortic stenosis three years later, with mild to moderate aortic incompetence being seen two years subsequent to that event. No aortic valvar disease had been noted prior to surgery. No intervention is planned at this stage.

Keywords: Acquired disease; echocardiography; aortic valvar insufficiency

Tetralogy of Fallot is rarely associated with aortic valvar disease.[1–3] We present a child who, subsequent to uncomplicated surgical repair of tetralogy of Fallot, gradually developed significant aortic stenosis and incompetence. This stenosis had not been present at the time of the surgical repair.

Case Report

Our female patient was born by normal vaginal delivery at 36 weeks of gestation to healthy and unrelated parents after an uneventful pregnancy. At neonatal examination, an ejection systolic murmur was heard at the lower left sternal edge. Examination was otherwise normal and she was pink. An electrocardiogram and chest X-ray were normal, but echocardiography revealed the anatomy of tetralogy of Fallot with a left-sided aortic arch. She remained well but, at fourteen months of age, was admitted with a cyanotic spell. Oral propranolol was started pending surgery. Total repair was carried out 2 months later at a tertiary centre. Repair consisted of resection of the obstructed right ventricular outflow tract, and closure of the ventricular septal defect. The postoperative course was uneventful. Serial follow-up showed mild residual obstruction in the right ventricular outflow tract, with a stable gradient of 35 mm of mercury, and mild pulmonary regurgitation. After a further period of three years, however, mild aortic stenosis was found (Fig. 1), with a stable gradient of 25 mmHg across the left ventricular outflow tract. After a further 2 years, mild to moderate aortic incompetence was also noted. There has not been any change in degree of stenosis or regurgitation over the past three years, nor has there been any progressive left ventricular dilation as monitored with serial M-mode echocardiography. No intervention is required in the near future, but the patient will continue to have annual follow-up.

Discussion

Aortic stenosis is rarely associated with of tetralogy of Fallot.[1] To date, to the best of our knowledge, there are only four reported cases in the literature. The first cases were reported in 1968 by Glancy et al.[2] These included rheumatic fever, endocarditis, and a bifoliate aortic valve without stenosis. Ino et al.[3] reported a neonate who died following percutaneous balloon valvoplasty of the stenotic aortic valve. Tetralogy of Fallot was discovered at postmortem There are also two reports of prenatal diagnosis of Fallot's tetralogy with absent pulmonary valve syndrome in association with aortic stenosis.[4,5] Our patient is unique, as the aortic stenosis was not present at the time of operation, but developed later on, and was subsequently associated with moderate aortic regurgitation.

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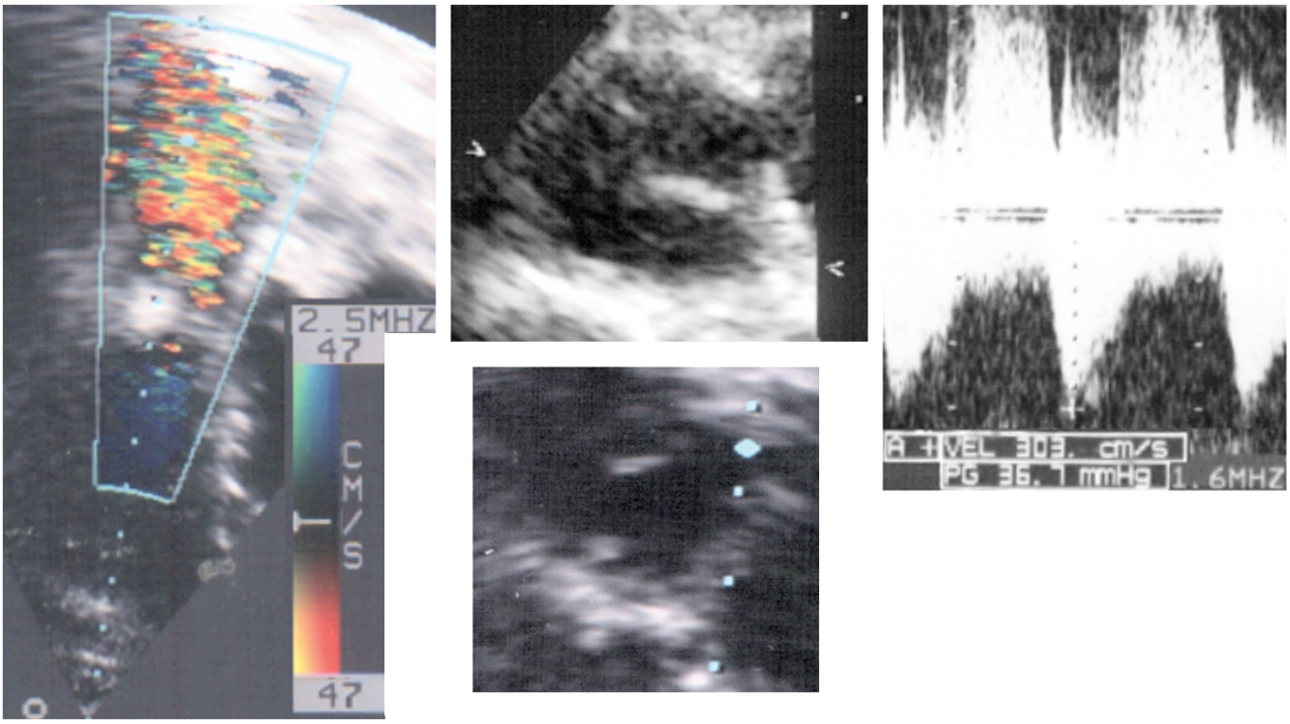

Figure 1.
Echocardiography of aortic valve, combined with colour flow Doppler (upper left), reveals turbulence distal to the valvar leaflets. The cross-sectional images in short axis show a valve with three leaflets (upper middle), the leaflets doming as they open (upper lower). The Doppler signal confirms the presence of a stenotic and regurgitant valve (right panel).

Aortic regurgitation is known to develop late after surgical repair of tetralogy, especially if repair is delayed to beyond the first decade of life.[6] This was not the case in our patient. Chugh et al.[7] reported 46 out of 62 individuals with aortic regurgitation after surgical correction. In three-quarters of these, the regurgitation was mild, and in only two patients was the regurgitation severe enough to require aortic valvar surgery. Niwa et al.[8] have identified potential risk factors relating to dilation of the aortic root in this setting. These were male sex, pulmonary atresia, right aortic arch, and delayed total correction, irrespective of previous palliation. Our patient had none of these risk factors. Our patient is also of interest in that the aortic valvar disease is not worsening, but careful follow-up will be required. This is being done annually, both clinically and with echocardiography.

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