Correction of type C atrioventricular canal associated with tetralogy of Fallot¹

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Bastos, P, de Leval, M, Macartney, F, and Stark, J (1978). *Thorax*, 33, 646–648. Correction of type C atrioventricular canal associated with tetralogy of Fallot. Type C complete atrioventricular canal associated with tetralogy of Fallot in a 5-year-old child was successfully corrected. Repair of the atrioventricular canal was combined with infundibular resection and the insertion of an outflow tract patch across the pulmonary valve ring. The postoperative course was uneventful, and the child is well one year after operation.

The association of complete atrioventricular canal with tetralogy of Fallot is a rare combination occurring in less than 5% of patients with complete endocardial cushion defects (Fisher *et al*, 1975). The operative risk for patients with isolated tetralogy of Fallot or atrioventricular canal is now acceptable. In the past two years we have operated on 89 children with tetralogy of Fallot, with four deaths (4.5%), and 15 children with complete atrioventricular canal, with three deaths (20%). The results for total correction of patients with both these anomalies have been poor, and only a few successfully corrected patients have been reported (d'Allaines *et al*, 1969; Fisher *et al*, 1975; Mills *et al*, 1976; Pacifico, 1977; Zavarella *et al*, 1977).

The purpose of this report is to present a 5-year-old child with type C atrioventricular canal associated with tetralogy of Fallot. Both defects were successfully corrected.

Case report

The patient, a baby girl, was born after a 39-week gestation. The pregnancy was complicated by pre-eclamptic toxæmia. The mother, aged 40, had had nine previous pregnancies, five of which ended in miscarriage. The clinical features of Down's syndrome were recognised at birth and confirmed by chromosome studies. The child developed heart failure and was treated with digoxin. At 11 weeks of age cyanosis was noticed, and she started to have occasional cyanotic spells. She was referred

to our hospital at the age of 16 months. A clinical diagnosis of tetralogy of Fallot was confirmed by cardiac catheterisation and biventricular angiocardiology, and so a left Blalock-Taussig shunt was performed. The child did not improve after the operation, and no shunt murmur was audible. A Waterston shunt was performed, and this produced significant improvement.

Cyanotic spells recurred one year later, however, and treatment with propranolol was started. She was readmitted for assessment before corrective surgery at the age of 3½ years. The angiocardiology features of tetralogy of Fallot were confirmed. The arterial oxygen saturation was 86%, but the child was having occasional cyanotic spells. The presence of an atrioventricular canal was not suspected, although in retrospect this could have been diagnosed from the angiocardiology. The posterior attachment of the mitral component of the valve was abnormally positioned, and a common atrioventricular orifice straddling the ventricular septum was visible on the lateral right ventricular angiocardiology. The left Blalock-Taussig shunt was not patent, and the aortopulmonary shunt was functioning but small. Two days after cardiac catheterisation the child developed a right-sided hemiplegia and therefore corrective surgery was deferred until a year later when she had recovered completely from this.

On examination, the features of Down's syndrome were noted. She was cyanosed and had clubbed fingers. There was a 2/4 systolic murmur at the left sternal edge and a continuous shunt murmur audible at the right side of the sternum and over the right chest. Her haemoglobin was

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18.4 g/dl and PCV 53%. The electrocardiogram showed a long PR interval, P pulmonale, a QRS mean axis of $+210^\circ$ with a clockwise loop in the frontal plane, and severe right ventricular hypertrophy. The radiographic heart size and lung markings were normal.

The operation was performed in February 1977 when she was 5 years old and weighed 15 kg. Propranolol was discontinued ten days before operation. The Waterston shunt was dissected and temporarily occluded. Cardiopulmonary bypass was instituted and the temperature of the perfusate lowered to 25°C . The aorta was crossclamped and the aortic root perfused with cold (4°C) Ringer's solution. The aorta was then opened and the Waterston shunt closed with mattress stitches. Inspection of the anatomy through the right atrium showed a free floating anterior common leaflet without chordal attachment to the ventricular septum (type C complete atrioventricular canal of the Rastelli classification (Rastelli *et al*, 1966)). Typical features of Fallot's tetralogy were also found, that is to say an anteriorly deviated infundibular septum producing infundibular and valvar pulmonary stenosis and an overriding aorta.

A vertical right ventriculotomy was performed and an infundibular resection carried out. The ventriculotomy was extended across the hypoplastic pulmonary valve annulus, and a woven Dacron patch was sutured into the left pulmonary artery and the ventriculotomy. After this, repair of the atrioventricular canal was performed through the right atrium using the technique described by Rastelli and associates (Rastelli *et al*, 1968). The common leaflet was divided into the mitral and tricuspid components. The cleft in the mitral valve was closed with fine single stitches and then a patch of Dacron velour was sutured to the right side of the ventricular septum. Special care was taken not to create subaortic narrowing, which appeared to be a possibility because of the dextroposed aorta that was overriding the ventricular septum. The mitral and tricuspid portions of the common leaflet were then attached to the patch at the expected level of the upper margin of the ventricular septum with single mattress stitches of 5-0 ethiflex. The atrial portion of the defect was closed with the upper part of the Dacron patch using a continuous prolene stitch. The coronary sinus was left on the left side of the patch to avoid the conducting tissue.

The right atrium was closed and the aortic clamp removed after evacuation of air from the ascending aorta and left ventricle. When the patient was rewarmed, bypass was discontinued without difficulty. Intracardiac pressures were

satisfactory (see table). The patient was intubated with a nasotracheal tube and ventilated for 48 hours. Weaning from the ventilator was achieved through a constant positive airway pressure system. Inotropic support was given for the first two days. The postoperative course was uneventful.

Table Intracardiac pressures (mmHg)

	At cardiac catheterisation (3 October 1975)	At operation: after repair (24 February 1977)
Right atrium	$\bar{8}$	$\bar{15}$
Right ventricle	90/8	35 systolic
Pulmonary artery	40/20	
Left atrium	$\bar{8}$	$\bar{15}$
Left ventricle	90/8	90 systolic
Aorta	100/70	90 systolic

She was discharged on the 16th postoperative day, and was taking digoxin, frusemide, and potassium supplements. Her subsequent progress was excellent, the diuretics were discontinued, and at present (12 months after operation) she is pink, fully active, and shows no signs of heart failure. The electrocardiogram shows sinus rhythm and the chest radiograph shows only moderate cardiomegaly.

Discussion

The association of complete atrioventricular canal and tetralogy of Fallot was first described by Bull in 1885 and more recently by Mouquin *et al* in 1956. It is a rare combination that would appear to occur more often when the atrioventricular canal is of type C (Fisher *et al*, 1975; McCabe *et al*, 1977). McMullan *et al* (1972) described four patients with valvar and infundibular stenosis in their group of 27 patients with complete atrioventricular canal treated surgically. Not enough details were provided to say whether these had tetralogy of Fallot associated with atrioventricular canal or just valvar or infundibular pulmonary stenosis.

These combined malformations could be correctly diagnosed before operation. Sometimes, as in our case, the complete diagnosis may be missed. Suspicion should be raised in patients with tetralogy of Fallot and features of Down's syndrome. A loud systolic murmur, radiating more widely than in patients with tetralogy, and a separate systolic murmur from mitral or tricuspid regurgitation may be present (Tandon *et al*, 1974). Left axis deviation in the electrocardiogram with a counter-clockwise loop in the frontal plane

should give rise to suspicion. Our patient, as well as those reported by d'Allaines *et al* (1969) and Mills *et al* (1976) showed an electrocardiographic pattern usually associated with tetralogy of Fallot.

One of us (FJM) has recently reviewed the angiocardigraphic findings in 91 patients with persistent common atrioventricular canal. It is in fact quite often possible to diagnose the presence of a common atrioventricular orifice on right ventricular angiocardiology alone, particularly when there is a right-to-left shunt at ventricular level, as in tetralogy of Fallot. If there is any doubt, however, about the identification of separate mitral and tricuspid valves on right ventriculography, left ventriculography should be carried out, preferably in the left and right anterior oblique projections, with or without cranio-caudal tilt. Such a policy not only renders it unlikely that the presence of a common atrioventricular orifice will be missed, but also delineates better any additional defects in the ventricular septum.

Careful assessment of the anatomy at operation is important if a successful correction is to be achieved. The parietal band of the crista is deviated anteriorly, and the aorta overrides the septum. According to Lev and colleagues (1961), these are the anatomical landmarks that make it possible to distinguish between a complete atrioventricular canal associated with tetralogy of Fallot and atrioventricular canal with pulmonary valvar or infundibular stenosis. In patients with infundibular stenosis the aorta does not override the septum and the parietal band is not deviated. Repair of the ventricular component of the endocardial cushion defect is more difficult in patients with tetralogy of Fallot. As pointed out by Zavarella *et al* (1977), the defect in the anterior part of the ventricular septum can be overlooked, which would result in a residual right ventricular-aortic communication. Also, care must be taken not to narrow the outflow tract of the left ventricle, a complication made more likely by the biventricular origin of the aorta in these patients.

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