Case reports

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Absent pulmonary valve syndrome *Surgical correction with pulmonary arterioplasty*

P SYAMASUNDAR RAO, GERALD M LAWRIE

From the Department of Paediatrics and Baylor Heart Team, King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia

SUMMARY A 13 month old child with classic features of absent pulmonary valve with tetralogy of Fallot underwent successful surgical repair by closure of the ventricular septal defect, relief of the right ventricular outflow tract obstruction, and partial resection and plastic repair of the aneurysmally dilated pulmonary arteries. Cardiac catheterisation data and clinical follow up for more than 18 months after the operation indicated excellent results. It is suggested that plastic repair of the aneurysmally dilated pulmonary arteries along with closure of the ventricular septal defect and relief of the right ventricular outflow tract obstruction should be performed early, perhaps between 1 and 2 years of age.

Absent pulmonary valve syndrome is a rare congenital heart defect and constitutes 3-5% of cases of tetralogy of Fallot. Usually the syndrome is associated with ventricular septal defect and pulmonary stenosis. Occasionally, however, it may be associated with other defects—namely, atrial septal defect, ventricular septal defect, patent ductus arteriosus, endocardial cushion defect, double outlet right ventricle, and Marfan's syndrome—or it may be found without any other congenital cardiac malformations.¹ The main features of absent pulmonary valve syndrome are pulmonary insufficiency because of the absence or rudimentary nature of the pulmonary valve cusps and massive dilatation of the main and the major branch pulmonary arteries, sometimes to aneurysmal proportions, producing varying degrees of compression of the tracheobronchial tree.

When absent pulmonary valve syndrome presents in infancy the respiratory problems are severe and the prognosis is extremely poor.¹ The recommendations for treatment of such infants vary from medical treatment and pulmonary physiotherapy (because of high mortality for complete repair)² to complete intracardiac repair at any age,³ with varying types of treatments in between.⁴⁻⁸ These include aneurysmorrhaphy of the ipsilateral pulmonary artery,⁵ suspension of repaired (aneurysmectomy) pulmonary artery to the retrosternal fascia,⁶ division of the right pulmonary artery and reconnection with the main pulmonary artery,⁷ and division of the right pulmonary artery via a tubular prosthetic graft in the

pulmonary artery and anastomosis with the superior vena cava.⁸ Even in older children there is much controversy about the choice of operation: some workers^{2,3} do not recommend routine pulmonary valve replacement at the time of intracardiac repair whereas others⁹ suggest that the pulmonary regurgitation should be corrected by valve placement at the time of total surgical correction. We report a 13 month old child with absent pulmonary valve syndrome and moderate respiratory distress in whom total surgical correction, which included closure of the ventricular septal defect, relief of pulmonary stenosis, and plastic repair of the pulmonary arteries, was successful. Cardiac catheterisation data and clinical follow up for 18 months after the operation suggest excellent results.

Case report

A 13 month old boy weighing 8.5 kg was investigated because of recurrent respiratory tract infections and a cardiac murmur. The child was not cyanotic but was in mild respiratory distress. Pertinent cardiac findings included increased right and left ventricular impulses, single second sound, a long grade 3/6 ejection systolic murmur along the left sternal border, and a grade 2-3/6 early diastolic decrescendo murmur heard best at the left upper sternal border. The electrocardiogram showed right ventricular hypertrophy. A chest x ray film showed mild cardiomegaly, slightly increased

Fig. 1 (a) Preoperative chest radiograph showing moderate cardiomegaly, minimally increased pulmonary vascular markings, and considerably dilated right pulmonary artery (arrows). (b) A selected frame from a right ventricular cineangiogram obtained before operation showing marked dilatation of the main, right, and left pulmonary arteries.

Table Findings on cardiac catheterisation

	Before operation		After operation	
	O ₂ saturation (%)	Pressures (mmHg)	O ₂ saturation (%)	Pressures (mmHg)
Site				
IVC	57	—	61	—
SVC	52	—	54	—
RA	57	a=6, v=5, m=2	58	a=6, v=6, m=4
RV	80	90/6	58	35/7
PA	85	25/10	58	25/5
PV	98	—	—	—
LA	98	a=10, v=15, m=7	—	—
LV	98	100/13	—	—
Ao	98*	100/60, m=77	95†	—
Qp:Qs	3:2		1:0	
PVRI	1:3		1:5	

*The aorta (Ao) was entered from the right ventricle via the ventricular septal defect.

†Taken from a femoral artery puncture at the time of postoperative cardiac catheterisation.

IVC, inferior vena cava; LA, left atrium; LV, left ventricle; PA, pulmonary artery; PV, pulmonary vein; PVRI, pulmonary vascular resistance indexed to body surface area (m²); Qp:Qs, ratio of pulmonary to systemic flow; RA, right atrium; RV, right ventricle; SVC, superior vena cava; a, a wave; v, v wave; m, mean.

Fig. 2 (a) Chest radiograph obtained one year after operation showing mild cardiomegaly, if any, and near normal sized pulmonary arteries. (b) An angiographic frame showing the size of the pulmonary arteries after the operation. The catheter tip is located at the junction of the right ventricle with the main pulmonary artery. This cine was obtained at a higher magnification factor (6") than the preoperative film (9").

pulmonary vascular markings, and a prominent right pulmonary artery (Fig. 1a). Cardiac catheterisation data showed a large increase in oxygen saturation at the level of the right ventricle, normal aortic saturation, increased right ventricular pressure to almost systemic level, with a 65 mmHg pressure gradient across the pulmonary valve, and an increased ratio of pulmonary to systemic flow (Table). Selective cineangiography showed a large ventricular septal defect, moderate to severe pulmonary stenosis, and considerable dilatation of the main, left, and right pulmonary

arteries (Fig. 1b). A patent foramen ovale was also diagnosed.

Under a total cardiopulmonary bypass the ventricular septal defect was closed using a double velour Dacron patch through a right atriotomy. The patent foramen ovale was also closed. The pulmonary artery and the right ventricular outflow tract were opened through a longitudinal incision. The pulmonary valve cusps were rudimentary and consisted of a small peripheral rim of valve tissue. The incision was extended cephalad in both directions over the right and left main pulmonary arteries up to the origin of the upper lobe branches. Segments of between $\frac{1}{2}$ and 1 cm of the anterior walls of the main, right, and left pulmonary arteries were resected, and the pulmonary arteries were closed with a running 4-0 Prolene suture. The right ventricular outflow tract and the pulmonary valve annular area were enlarged with a pericardial patch. The postoperative course was uneventful.

Cardiac catheterisation performed 10 days after the operation showed no evidence of a ventricular septal defect and no appreciable right ventricular outflow obstruction. The repaired pulmonary arteries appeared near normal (Fig. 2b). There was only minimal pulmonary insufficiency. The child was discharged home the next day. Postoperative follow up for about 18 months showed no symptoms, normal growth, a grade 1-2/6 early diastolic decrescendo murmur, and near normal sized pulmonary arteries on the chest radiograph (Fig. 2a).

Discussion

Patients with absent pulmonary valve syndrome generally fall into two main clinical categories^{1,9}: those who present in early infancy with severe cardiorespiratory distress and who may not survive beyond infancy (group 1); and those who may have mild symptoms during infancy and survive infancy (group 2). Our patient may belong to the second group; he had classic features of absent pulmonary valve syndrome as described in the published work.¹ The child underwent successful surgical correction by patch closure of the ventricular septal defect, relief of the pulmonary stenosis, and partial resection and plastic repair of the aneurysmally dilated pulmonary arteries. Although there is agreement with regard to closure of the ventricular septal defect and relief of right ventricular outflow tract obstruction, there is considerable debate as to whether valve insertion at the pulmonary artery annulus should be performed at the time of intracardiac repair.^{2,3,9}

Ilbawi *et al.*⁹ presented data on 10 group 2 patients and suggested that the pulmonary valve should be replaced at the time of intracardiac repair. The age of

the patients with pulmonary valve insertion at the time of primary repair varied between 6 and 11 years. Deterioration of right ventricular function secondary to persistent pulmonary valve regurgitation in the subgroup without pulmonary valve replacement was cited as the reason for their recommendation.⁹

Pulmonary valve regurgitation produced experimentally in dogs,¹⁰ produced by pulmonary valvotomy for isolated pulmonary stenosis,¹¹ or produced after repair of tetralogy of Fallot,^{12,13} as well as isolated "naturally occurring" pulmonary regurgitation¹⁴ may be well tolerated for years. This is the main reason why most workers do not recommend pulmonary valve replacement routinely but reserve it for patients with raised pulmonary artery pressure. Recent reports,^{15,16} however, have shown that a small percentage of patients require pulmonary valve replacement to relieve pulmonary regurgitation several years after repair of tetralogy of Fallot. This and the deterioration of patients with absent pulmonary valve syndrome without previous pulmonary valve replacement,^{3,9} would tend to support the need for pulmonary valve replacement. But, the currently used pulmonary valve substitutes such as aortic valve homografts and porcine valves have inherent problems of their own^{17,18} making them unsuitable alternatives. The right ventricular dysfunction pointed out by Ilbawi *et al.*⁹ is a result of long term pulmonary regurgitation, which is present since birth. If the total repair is done within the first two years, as in our case, the right ventricle may be protected to some degree. The plastic repair of the pulmonary arteries, as performed in our case, has not been performed regularly in most, if not all, of the large series.^{2,3,9} Furthermore, there are technical difficulties and size limitation in using valved conduits in young infants. For these reasons we suggest that plastic repair of the aneurysmally dilated pulmonary arteries along with closure of ventricular septal defect and relief of right ventricular outflow tract obstruction should be performed early, perhaps between 1 and 2 years of age in patients with absent pulmonary valve syndrome. Although follow up results at the end of 18 months are encouraging, a longer period of follow up and experience with more cases may be necessary to confirm these observations and support our recommendations.

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Requests for reprints to Dr P Syamasundar Rao, Department of Paediatric Cardiology, King Faisal Specialist Hospital and Research Centre, PO Box 3354, Riyadh, 11211 Saudi Arabia.