THE 'RASTELLI' OPERATION – RESULTS AT THE SINGAPORE GENERAL HOSPITAL

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ABSTRACT

Reconstruction of the right ventricular outflow tract in patients with right ventricle-pulmonary artery discontinuity with a valved conduit is well established. The aim of this study was to review our results at the Singapore General Hospital. Between October 1989 and May 1993, 13 patients underwent the 'Rastelli' operation. The mean age at definitive repair was 68 months. Ten patients had pulmonary atresia with ventricular septal defect, two patients truncus arteriosus (one patient each with type 3 and type 2) and one patient had tetralogy of Fallot with absent pulmonary valve syndrome. In 12 patients cryo-preserved pulmonary artery homografts with a mean diameter of 21mm (19-23mm) were used, and a 11mm composite Hancock dacron valved conduit in one patient. There were no operative deaths. The mean duration of follow-up is 37 months (23-48 months). All patients were well and in NYHA class I on recent follow-up. On echocardiographic assessment 12 patients had trivial or no gradient across the right ventricular outflow tract, while one patient had a gradient of 25mmHg. The "Rastelli" operation can be undertaken with low mortality and morbidity and early results are encouraging.

Keywords: *Rastelli operation, pulmonary atresia, pulmonary artery homograft, truncus arteriosus*

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INTRODUCTION

The use of extra-cardiac conduits has enabled successful palliation of patients with right ventricle-pulmonary artery discontinuity. Although some patients with pulmonary atresia and ventricular septal defect have adequate confluent pulmonary arteries, they, more often present with hypoplastic central pulmonary arteries. The latter would require creation of systemic to pulmonary artery shunt(s) to induce pulmonary artery growth and palliate cyanosis. In addition, these patients may have major aorto-pulmonary collateral arteries that supply a substantial portion of the lung. In these patients, it is necessary to recruit these vessels by anastomosing them to the central pulmonary arteries, thereby unifocalising the pulmonary blood flow to provide as large a vascular bed as is possible prior to final repair.

In this report we describe our results in patients undergoing the 'Rastelli' procedure at the Singapore General Hospital.

MATERIALS AND METHODS

From October 1989 until May 1993, 13 patients underwent the 'Rastelli' procedure at the Singapore General Hospital. The mean age at definitive correction was 68 months (range 4-118 months), and the mean weight at operation was 15.5 kg (range 3.6-26.6 kg). The mean height at correction was 105.82 cm (range 46-165 cm). There were 9 male and 4 female patients in our series. The mean pre-operative haemoglobin concentration was 17.16 gm/dl (range 11.8-23.9 gm/dl). The mean haematocrit was 50.6% (range 38.8-62.3%).

The 13 patients, included 10 patients with pulmonary atresia with a ventricular septal defect, two of whom had transposition of great arteries in addition; 2 patients with truncus arteriosus (one patient each with type 3 and type 2) and one patient had tetralogy of Fallot with absent pulmonary valve syndrome.

Pre-operative echocardiography and cardiac catheterisation were performed in all patients.

Initial palliation was necessary in 9 patients. One patient with a type-3 truncus arteriosus underwent pulmonary artery banding at 8 months of age. In the group of patients with pulmonary atresia and ventricular septal defect, 3 patients underwent bilateral modified Blalock-Taussig shunts (the age at which the first shunt was performed was 2 weeks, 6 months and 2 years respectively and the age at which the second shunt was performed was 31 months, 21 months and 6 years respectively). In 5 patients unilateral, modified Blalock-Taussig shunts were performed, one of whom required bilateral unifocalisation of multiple aorto-pulmonary collateral arteries.

Surgical technique

All patients were given phenoxy-benzamine (1mg/kg body weight) just before cardio-pulmonary bypass was initiated to achieve uniform cooling and reduce afterload peri-operatively. Operative repair was carried out under cardio-pulmonary bypass using a single aortic cannula and bi-caval cannulation with caval snaring. The rate of flow was maintained at 2.4 l/min/m^2 for patients over 10 kg in weight and 150 ml/kg/min for those under 10kg. Systemic hypothermia was used in all patients with the nasopharyngeal temperature maintained at 25°C. The aortic cross clamp was then applied and cold crystalloid cardioplegia infused through the aortic root at 20 minute intervals to induce and maintain cardiac arrest. The left atrium was vented through an opening in the interatrial septum in all patients. A vertical incision was then made in the right ventricular outflow tract, and through this opening, infundibular resection and closure of the ventricular septal defect were performed. The ventricular septal defect was repaired using a dacron patch with interrupted mattress plegetted prolene sutures. The patient was then rewarmed. The atrial septal

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opening (patent oval foramen or surgically created defect) was then closed and the cross-clamp released. The right atrial incision was then repaired. The pulmonary outflow tract was constructed during re-warming with heart beating.

The right ventricle to pulmonary artery connection were made in 12 patients with cryo-preserved pulmonary artery homograft. The mean diameter of the conduit was 21mm (range 19-23mm). For the proximal anastomosis, the posterior margin of the homograft was directly sutured to the superior margin of the right ventricular opening, and a measured tailored pericardial patch was then sutured to the anterior margin of the homograft and the opening in the right ventricular cavity to complete the conduit. In one infant with a type-2 truncus arteriosus, a 11mm Hancock-dacron valved composite conduit was used.

The mean duration of cardiopulmonary bypass was 205 minutes (range 165-298 min), and the mean duration of aortic cross-clamp was 109 minutes (range 62-179 min).

RESULTS

There were no operative deaths. The median stay in the intensive care unit was 3 days (range 2-15 days). The median hospital stay was 8 days (range 7-30 days). One patient had supraventricular tachycardia controlled successfully with digoxin. Three patients had major infections (urinary tract in one patient and respiratory in 2 patients) that prolonged their hospital stay. All patients are well at recent follow-up. One patient is on oral digoxin while the rest require no medication. The mean period of follow-up is 27 months (range 12-38 months). All patients had regular post-operative clinical and echo-cardiographic assessment of the pulmonary conduit and right ventricular function. Ejection systolic murmurs were present (grade 2/6 or 3/6) in 5 patients. Only one of these patients (who had a transposition of the great artery, pulmonary atresia and ventricular septal defect) had a gradient across the right ventricular outflow tract of 25mmHg. The remaining patients showed trivial or no gradients.

DISCUSSION

Since the description by Rastelli and colleagues[1] in 1969 of an intraventricular tunnel repair (left-ventricle to aorta), with closure of proximal pulmonary artery and restoration of right ventricle to pulmonary artery continuity with a valved extra-cardiac conduit, for patients with transposition of great arteries with ventricular septal defect and left ventricular outflow tract obstruction, similar procedures have been collectively referred to as the 'Rastelli' operation. This series reviews our experience in these patients who may be divided into two subgroups: one with a diminished pulmonary blood flow related to pulmonic stenosis or pulmonary atresia and the other with increased blood flow as is seen with truncus arteriosus.

The feasibility of surgical correction in patients with pulmonary atresia with ventricular septal defect depends, among other things, on the size and distribution of the central and branch pulmonary arteries, the presence of aorto-pulmonary collateral arteries and the number of broncho-pulmonary segments they supply. The presence or otherwise of central pulmonary arteries determines the degree of difficulty in palliating these patients and considerable effort is taken to demonstrate their presence. This can be done during cardiac catheterisation, either by opacification via the arterial duct or by selective pulmonary venous injection. More recently nuclear magnetic resonance imaging (NMRI) techniques have been used and inability to demonstrate the central pulmonary arteries during NMRI is considered evidence of their absence[2]. Multiple aorto-pulmonary

Table I – Patients and results

SL No	Age (Months)	Sex	Diagnosis	Previous Palliation	Preop Hb gm/dl	Pulmonary Homograft Size (mm)	Stay in ICU (days)	Postop RVOT gradient (mmHg)	Current NYHA Class
1	52	M	PA/VSD	RBTS	19.3	20mm	3	Trivial	1
2	35	M	PA/VSD	–	16.1	22mm	2	Nil	1
3	56	M	PA/VSD	RBTS	15.8	21mm	2	Trivial	1
4	59	M	PA/VSD/MAPCA	–	16.7	22mm	3	Trivial	1
5	52	F	PA/VSD	RBTS/LBTS	15.8	19mm	6	Trivial	1
6	108	F	PA/VSD/MAPCA	LBTS/RBTS Bilateral Unifocalisation	14.8	21mm	8	Nil	1
7	106	F	PA/VSD	RBTS/LBTS	20.3	20mm	3	Nil	1
8	48	M	PA/VSD	RBTS	23.9	20mm	3	Trivial	1
9	91	M	PA/VSD/TGA	LBTS	18.0	21mm	13	25mmHg	1
10	80	M	PA/VSD/TGA	RBTS	16.1	21mm	7	Trivial	1
11	4	M	Truncus Arteriosus Type II	–	14.3	11mm Hancock Dacron Conduit	6	Nil	1
12	88	F	Truncus Arteriosus Type III	Pulmonary Artery Banding	16.3	21mm	2	Nil	1
13	108	F	FT/APVS	–	11.8	23mm	2	Nil	1

PA : Pulmonary atresia
VSD : Ventricular septal defect
RBTS : Right Blalock Taussig Shunt
NYHA : New York Heart Association
APVS : Absent Pulmonary Valve Syndrome
MAPCA : Multiple Aortopulmonary Collateral Arteries
TGA : Transposition of Great Arteries
RVOT : Right Ventricular Outflow Tract
LBTS : Left Blalock Taussig Shunt

collateral arteries are frequently present and may be the sole blood supply for a substantial portion of lung tissue. These collaterals arise from the descending thoracic aorta, and may anastomose with either a central pulmonary artery or a normally connected lobar branch, or else often after branching enter the hilum continuing distally with the airways to supply the alveolar bed. In an elegant study by Sheila Haworth and Fergus J Macartney[3], 91/209 broncho-pulmonary segments were found solely connected to collateral arteries.

The preliminary management of these patients is therefore focussed on enhancing growth of the central arteries and unifocalising the intra-parenchymal vessels. Growth enhancement is best achieved with prosthetic aorto-pulmonary shunts and central shunts are now preferred, especially in patients with small pulmonary arteries, as they produce uniform growth of the central pulmonary arteries and also because of pulmonary artery obstruction and distortion that can occur with peripheral shunts[4]. As it has been shown that pulmonary arterial growth on shunting is better in younger patients[5], early palliation is required to achieve optimal results. We now undertake early unifocalisation and shunting, and 4 patients are currently awaiting total repair.

In patients with pulmonary atresia with ventricular septal defect, we prefer to undertake definitive correction when the patients are 12 to 15 kg in weight, as we can then use a larger sized conduit. We use cryo-preserved pulmonary homografts 3-5 mm larger than the minimum size required, as derived from the Birmingham formula, to reduce the frequency of conduit replacement due to size discrepancy secondary to later somatic growth. This may be responsible for the low gradients recorded in our patients on follow-up. However, excellent results[6] (94% actuarial freedom from reoperation in patients over one year) have been achieved in patients in whom the conduits used approximated the child's expected pulmonary valve annulus diameter.

Patients may be clinically well despite serious conduit obstruction. There may be no symptoms in over 50% of patients. This makes periodic clinical and echo-cardiographic evaluation imperative as development of dysfunction in the pulmonary ventricle is a poor prognostic feature[8]. Conduit replacement is therefore indicated in asymptomatic patients, if the pressure in the pulmonary ventricle nears systemic values[9], and in symptomatic patients showing ventricular dysfunction and/or important atrio-ventricular valve regurgitation[8].

In patients with truncus arteriosus, we now undertake neonatal repair. One patient who underwent staged repair, did so in a period prior to the establishment of a paediatric service at the general hospital. Conduit replacement is frequently needed in patients undergoing correction in the neonatal period. Between 60% and 65% of patients in reported literature[8,10,11] require conduit replacement at a median interval of 4 years.

In conclusion, we have shown that the Rastelli operation can be done with low morbidity. We hope that our practice of undertaking the definitive repair at an age when a conduit larger than 18mm in diameter can be used will reduce the frequency of their replacement.

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