Persistent Truncus Arteriosus Operated During Infancy: Long-Term Follow-Up

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SUMMARY. Between July 1974 and October 1988 19 consecutive infants (mean age 2.7 months, range 5 days to 11.7 months) underwent surgical correction for persistent truncus arteriosus by one surgeon (J.L.M.). Continuity between the right ventricle and pulmonary arteries was achieved with an antibiotic-sterilized aortic homograft (diameter 13–18 mm) together with patch closure of the ventricular septal defect. There were 3 early postoperative deaths (16%): 1 patient had severe aortic regurgitation, the other 2 had preoperative cardiac arrests. Of the latter, 1 had suffered severe cerebral damage, and the other developed recurrent pulmonary hypertensive crises and low cardiac output. The 16 survivors have been followed for 3.1–17.3 years (mean 7.8 years). Four patients required subsequent replacement of the homograft for stenosis, aortic valve replacement for regurgitation, or both (3.0, 4.0, 8.5, and 12.0 years postoperatively). Of the 16 survivors, 15 are in NYHA class I. Of the 13 patients who have not had aortic valve surgery, 9 have no evidence of stenosis or regurgitation. In the 14 children with the original homograft the median of the residual peak gradient across the right ventricular outflow tract is 15 mmHg (range 10–40 mmHg), and no patient has severe homograft regurgitation at follow-up. Repair of persistent truncus arteriosus has been achieved with low early mortality and no late mortality, which reflects excellent long-term function of the homograft. Furthermore, if truncal valve function is good at presentation, patients are unlikely to require aortic valve surgery.

KEY WORDS: Cardiac surgery — Congenital heart defects — Truncus arteriosus

Persistent truncus arteriosus accounts for 1.2% of all congenital heart malformations and presents with congestive heart failure or cyanosis during the neonatal period (60%) or before 12 months of age (97%) [12, 13]. It requires surgical treatment during infancy; otherwise about 80% of these patients die within the first year of life [16]. The truncal valve morphology and function vary and are important factors when determining outcome [1, 2, 10]. The perioperative risk of pulmonary hypertension in children with truncus arteriosus is well recognized [4, 5, 7, 9, 14, 16]. The immediate results of surgery are therefore largely dependent on the function of the truncal valve and the pulmonary vascular reactivity. However, once the early postoperative period is safely over, there may be residual hemodynamic abnormalities that influence long-term outcome. Our study was designed to assess noninvasively the long-term functional results in patients undergoing surgical repair of truncus arteriosus during infancy.

Materials and Methods

We have reviewed a group of 19 consecutive infants who underwent repair of truncus arteriosus between July 1974 and October 1988 performed by one surgeon (J.L.M.). There were 11 female and 8 male patients. Fifteen patients had truncus arteriosus type 1 and four had type 2. In 2 patients with type 1 truncus arteriosus there was also interruption of the aorta type B. The age at operation ranged from 5 days to 11.7 months (mean 2.7 months). The weight at operation was between 2.2 and 8.5 kg (mean 4.0 kg).

Surgical technique included surface cooling with total circulatory arrest at 16°C and bypass rewarming. The ventricular septal defect was closed with a Dacron patch and an antibiotic-sterilized homograft anastomosed first to the main pulmonary
artery or its branches and then to the right ventriculotomy. The original size of the homografts used was between 13 and 18 mm [median 16 mm]. Three homografts (two 17 mm and one 14 mm in diameter) were tailored during the operation: One cusp and the adjacent aortic wall were excised; thus the actual size of these three homografts was smaller. In 2 patients with interrupted aortic arch, this anomaly was repaired at the same time.

Results

There were 3 early postoperative deaths: One patient had severe aortic regurgitation, and the other two had preoperative cardiac arrests. Of the latter, one had suffered severe cerebral damage and the other developed recurrent pulmonary hypertensive crises with a low cardiac output. Of the 16 survivors, 9 (56%) experienced at least one episode of pulmonary hypertensive crisis during the early postoperative period. There were no late deaths.

The 16 survivors have been followed for 3.1-17.3 years, with a mean follow-up of 7.8 years (Fig. 1). Four patients have required aortic valve or homograft replacement 3.0, 4.0, 8.5, and 12.0 years after operation, respectively. Replacement of the homograft was performed in 2 cases. In 1 of these patients there was severe stenosis of the homograft valve with a gradient of 50 mmHg. The other required aortic valve replacement for severe regurgitation, and the homograft was electively replaced by a larger one. There were 2 more patients with severe aortic regurgitation who had their aortic valve replaced. The aortic valve was replaced in all 3 patients with a St. Jude Medical valve. One other patient who had type 2 truncus arteriosus developed stenosis of the right pulmonary artery that required patching at the age of 4 months.

Assessment of the functional result and capacity has been performed in surviving patients by clinical, echocardiographic, color flow Doppler, and exercise testing. Semiquantitative criteria were used for the Doppler and color flow Doppler assessment of the homograft and aortic valve regurgitation [3, 11].

Long-Term Function of the Homograft

Two patients had undergone homograft replacement: 1 for significant stenosis of the homograft valve and the other one electively during aortic valve replacement. The 14 surviving patients with the original homograft have a gradient between the right ventricle and the pulmonary artery ranging from 10 to 40 mmHg with a median of 15 mmHg at follow-up (Fig. 1).

Only 3 children had no evidence of homograft regurgitation at follow-up; mild homograft regurgitation was found in 11 children and moderate regurgitation in 2. No patient developed severe homograft regurgitation on clinical, echocardiographic, and color flow Doppler evaluation (Table 1).

Long-Term Function of the Aortic (Truncal) Valve

Three patients with severe regurgitation had aortic valve replacement (Table 2), and in all of these patients the truncal valve at original presentation was severely malformed and regurgitant. Of the 13 patients who have not had aortic valve surgery, 9 have no evidence of stenosis and regurgitation. Three