Surgical experience with persistent truncus arteriosus in symptomatic infants under 1 year of age

Report of 13 consecutive cases

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SUMMARY Between January 1974 and November 1980, 13 symptomatic infants under 1 year of age with persistent truncus arteriosus type I or II underwent surgery. Pulmonary artery banding was performed in 10 cases, with five deaths. Among the survivors, one developed severe pulmonary vascular disease and only two underwent late intracardiac repair. Primary total correction was performed in three infants and all are well, though one required conduit replacement five and a half years after the initial procedure. Recently, antibiotic sterilised homograft conduits, rather than heterografts, have been preferred as extracardiac conduits in infancy. Early intracardiac repair followed, if necessary, by later conduit replacement appears to have significant advantages over "conventional" pulmonary artery banding and late total correction. Concern remains on the treatment of choice in infants under 3 months of age.

Persistent truncus arteriosus is a rare congenital malformation accounting for less than 2% of all the congenital heart defects seen in the first year of life.¹ In the great majority of infants born with this malformation, there is a large left-to-right shunt, with large pulmonary blood flow,² though normal or low pulmonary flow can be present,³ resulting from pulmonary obstruction. Deformities of the truncal valve are often present, resulting in different degrees of truncal incompetence.⁴⁻⁵ The natural history of patients with persistent truncus arteriosus is poor. Almost all present during the first months of life with intractable congestive cardiac failure.²⁻³ More than 60% die before the age of 6 months⁶⁻⁷ and more than 80% before 1 year.⁸⁻⁹ In the survivors, pulmonary vascular disease increases progressively after the age of 2 years and one-third are inoperable by the age of 4 years.

In the surgical management of infants with persistent truncus arteriosus and intractable congestive cardiac failure, the choice lies between a two-stage procedure involving banding of either the main pulmonary trunk or the individual pulmonary arteries,

followed by late total repair,³⁻¹⁰ or a primary "total correction".¹¹⁻¹⁵

In this report, we review the surgical experience in infants under 1 year of age with persistent truncus arteriosus and severe congestive cardiac failure at the Royal Liverpool Children's Hospital during the past seven years.

Patients

Between January 1974 and November 1980, 13 consecutive infants with persistent truncus arteriosus type I and II⁶ underwent operation at the Royal Liverpool Children's Hospital. Pulmonary artery banding was performed in 10 cases and primary intracardiac repair in three. The age at presentation ranged from 1 day to 10 months. All had signs of severe congestive cardiac failure, associated in three cases with mild cyanosis. Cardiac enlargement with increased pulmonary vascularity on the chest x-ray film was present in all patients. Cardiac catheterisation and angiocardiology were included in the pre-operative evaluation of each case. The arterial oxygen saturation ranged from 83% to 98% (mean 91%). In one patient, who presented at 10 months of age, the pulmonary vascular resistance was found to be increased ($R_p/R_s=0.44$), whereas, among the other

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cases, the ratio between the pulmonary and systemic resistances ranged from 0.08 to 0.16 (mean $R_p/R_s = 0.13$). Mild regurgitation of the truncal valve was present in five cases and moderate regurgitation in two. A systolic pressure gradient between the common trunk and the pulmonary arteries was present in four infants. The principal indications for surgical treatment were uncontrolled congestive cardiac failure in 12 patients and an increasing pulmonary vascular resistance in one infant. The age at operation ranged from 2 weeks to 12 months (mean 4.1 months) (Fig. 1) and the weight from 2.1 kg to 7.9 kg (mean 4.1 kg).

Surgical approach and results

PULMONARY ARTERY BANDING (10 patients)

These infants ranged in age from 14 days to 12 months (mean 3.4 months) and in weight from 2.1 to 7.9 kg (mean 3.9 kg). A left lateral thoracotomy was used in all. In three cases the main pulmonary artery was constricted, while right and left branches were banded individually in seven instances. There were four hospital deaths (Fig. 1), three of which occurred among the six cases operated on during the first three

months of life, while only one occurred among the four older cases, and all resulted from low cardiac output. One infant died four months after operation in intractable congestive cardiac failure.

Fig. 1 Age and weight at operation. □ pulmonary artery banding; ■ early deaths after pulmonary artery banding; ● primary total repair.

Fig. 2 Truncal angiogram (2A) showing severe valve regurgitation, 3.7 years after banding of main pulmonary artery. Postoperative chest x-ray film (2B) of the same case, 2.9 years after secondary total repair. RV, right ventricle; LV, left ventricle; CT, common trunk; RPA, right pulmonary artery. The arrow points towards the truncal valve.

Of the five long term survivors, two have undergone late intracardiac repair. In the first patient, preoperative cardiac catheterisation at 4 years of age showed a 60 mmHg gradient across the main pulmonary artery band, an arterial oxygen saturation of 90%, and a pulmonary to systemic resistance ratio of 0.16. Severe regurgitation of the truncal valve, which was of mild degree at the first study, was present (Fig. 2A). At operation, continuity between the right ventricle and the pulmonary artery was achieved using a 20 mm Hancock conduit and the truncal valve was replaced with a heterograft prosthesis. The child is well 34 months after the operation. Fig. 2B shows the postoperative chest x-ray film of this case.

The second patient was reinvestigated at 5 years of age after banding of the individual pulmonary arteries for type II truncus at 6 months of age. The right pulmonary artery was almost totally occluded at the site of the band and the left pulmonary artery was severely narrowed (Fig. 3). The systemic oxygen saturation was 52%. The total pulmonary blood flow was considerably reduced, with unequal distribution between the lungs, the right pulmonary artery being more severely restricted than the left. The pulmonary vascular resistance was normal in the left lung, while the right pulmonary artery was not entered. At operation, at 6 years of age, both pulmonary arteries were

found to be severely hypoplastic. Endothelial proliferation at the site of banding was almost completely occluding the lumen of the right pulmonary artery, while the left branch was patent to a 4 mm Hegar's dilator. Distal to the bands, the tissue was extremely friable. Plastic enlargement of both pulmonary arteries was performed inserting gussets of preserved calf pericardium after incising the vessels across the banded areas down to the first lobar branches. A 20 mm aortic root homograft was interposed between the right ventricle and the distal pulmonary arteries. This patient is making a satisfactory recovery one month after operation, though the heart shadow is moderately enlarged on the chest x-ray film and antifailure treatment has been required to control right ventricular failure.

Two other survivors of banding have been restudied. At 4 years of age, one patient was found to have an arterial oxygen saturation of 69% with elevation of pulmonary vascular resistance to suprasystemic levels and further surgical treatment is considered inadvisable. The second patient, with type II persistent truncus arteriosus underwent banding of the individual pulmonary arteries at 2 months of age, and was restudied at 32 months. Both distal pulmonary arteries were at low pressure and the arterial oxygen saturation was 65%. Moderate to severe truncal valve regurgitation was present. The remaining survivor of pulmonary artery banding has not yet been restudied, but clinical progress has been satisfactory.

PRIMARY INTRACARDIAC REPAIR (three patients)

One infant, with persistent truncus arteriosus type II, was operated on in 1975 at 4 months of age, weighing 4.1 kg. Deep hypothermia and circulatory arrest were employed. The pulmonary arteries were detached individually from the truncus and continuity between the "double barrelled" pulmonary arteries and the right ventricle was established with a 12 mm Hancock conduit. Satisfactory recovery (Fig. 4A) was followed four years later by dyspnoea and signs of right heart failure. At cardiac catheterisation, five and a half years after the operation, a 60 mmHg gradient across the conduit was demonstrated (Fig. 5). Reoperation was performed in September 1980. At repair, the discrepancy between the heart size and the size of the conduit was evident (Fig. 4B) and there was no evidence of functioning of the heterograft valve leaflet tissue (Fig. 6). The conduit was removed and replaced with a 20 mm antibiotic sterilised aortic root homograft conduit. The patient has made a satisfactory postoperative recovery and he is doing well two months after operation.

In the two more recent patients, deep hypothermia

Fig. 3 Truncal angiogram 4.2 years after individual banding of both pulmonary arteries. The left pulmonary artery is severely narrowed at the site of banding (white arrow). The right pulmonary artery is not visualised and it was found almost completely occluded at operation.

Fig. 4 (A) Early postoperative chest x-ray film of the infant who underwent total repair at 4 months of age. (B) Chest x-ray film of the same patient five years later. The discrepancy between the heart size and the size of the prosthetic valve ring is evident.

Fig. 5 Right-oblique (A) and posteroanterior (B) view of pulmonary angiogram of the same case illustrated in Fig. 4. The narrowing across the conduit was more pronounced at the distal end (white arrow).

with low flow perfusion was preferred. In one of them, 8 months old, with persistent truncus arteriosus type I, the main pulmonary artery was detached from the common trunk while in the other, 6 months old, with persistent truncus arteriosus type II, the origins of both pulmonary arteries were detached from the posterolateral aspect of the truncus with a flange of wall tissue. In both cases, continuity between the right ventricle and the pulmonary artery was established with fresh, antibiotic sterilised aortic homograft conduits 16 and 18 mm in diameter, respectively. Both infants, with a follow-up of seven

and eight months, are growing well and are symptom free.

Discussion

In 1977, the Mayo Clinic group described the ideal candidate for total repair of persistent truncus arteriosus as a patient of between 5 and 12 years of age, with normal or mildly raised pulmonary resistance.¹⁰ With the natural history of persistent truncus arteriosus in mind, it is evident that only a small minority of cases

Fig. 6A, B Heterograft valve leaflets almost completely disappeared five and a half years after insertion of a 12 mm Hancock conduit in the same case as Fig. 4 and 5.

could ever be included in this ideal management programme.

Poirier *et al.*³ and Marcelletti *et al.*¹⁰ found a positive correlation between the age of patients at the time of repair and hospital mortality and consequently proposed palliative pulmonary artery banding for severely symptomatic infants. Though previous palliation was not related to hospital mortality in the experience of the Mayo Clinic,^{2 10 16} other authors have identified a number of problems which may follow pulmonary artery banding in this group of patients.¹⁴ Pulmonary artery banding is not always effective in preventing the development of pulmonary vascular disease,² the growth of the pulmonary arteries may be compromised,¹⁷ and the waiting period between palliation and correction is associated with an appreciable cumulative mortality.¹²

These results are confirmed by our own experience. Of the 10 infants who underwent pulmonary artery banding, five died after operation (50% mortality) and one developed severe pulmonary vascular disease. Though two cases have undergone successful late intracardiac repair, in one patient the obstruction at the site of the banding and the distal hypoplasia of both pulmonary arteries were considerable and the procedure performed, though life saving, may not prove to be entirely successful. Pulmonary artery banding can therefore be considered to have given a satisfactory result in only two of nine cases, with one patient awaiting reinvestigation.

The improved surgical results in the treatment of right ventricle to pulmonary artery discontinuity by the use of extracardiac valved conduits in infancy¹³

have shown the possibility of early intracardiac repair of persistent truncus arteriosus in the first year of life. This has been confirmed by the results achieved by Turley *et al.*¹⁸ and Stark *et al.*¹⁹ in this age group, with hospital mortality rates of 21% and 46%, respectively. All our three patients who underwent primary intracardiac repair survived and are doing well. It should be noted, however, that these three patients who underwent primary intracardiac repair were older than the patients who were banded (Fig. 1). In the report of Stark *et al.*¹⁹ three of the six deaths among 13 infants undergoing primary repair were in patients of less than 3 months of age. Though the age distribution of the patients with persistent truncus arteriosus reported by Turley *et al.*¹⁸ is not stated, four of the six deaths (among 29 patients submitted to primary repair) were also among patients under 3 months of age. Thus, primary intracardiac repair can be achieved in patients with persistent truncus arteriosus over 3 months of age with an acceptable mortality, but the mortality rate in patients less than 3 months of age remains high. Nevertheless, our present results and those of others^{2 12 14 17} show also that pulmonary artery banding does not provide a satisfactory solution for these patients. In our series in fact, among the five deaths which occurred in infants submitted to banding, four were in cases under 3 months of age at the time of operation.

The decision whether a valve bearing Dacron prosthesis or an aortic homograft conduit should be used to establish continuity between the right ventricle and the pulmonary artery remains controversial. Marcelletti *et al.*¹⁰ identified the use of an aortic

homograft as a significant operative risk factor, and Appelbaum *et al.*¹² and Ebert *et al.*¹³ referred to their preference for synthetic grafts in this group of patients. Though several reports have been published on late failure of homograft conduits^{20 21} it has been recognised that late dysfunctions were mainly a consequence of the techniques of sterilisation and preservation²² rather than of the tissue itself; meanwhile one of our cases (Fig. 6) showed that the function of heterograft valve leaflets could be severely impaired in a medium term period. In our unit, fresh, antibiotic sterilised aortic homograft conduits have proved satisfactory in early and medium term function in re-establishing the right ventricle-pulmonary artery continuity.²³ Therefore, they have been routinely used in our more recent experience.

Appelbaum *et al.*¹² and Ebert²⁴ emphasised the need to insert a relatively small conduit in infants, in order to avoid kinking or excessive mediastinal compression. This problem may be related more to the type rather than to the size of the conduit itself. When an aortic homograft is employed, a larger size conduit can be accommodated in the small mediastinum, minimising sternal and mediastinal compression and, it is hoped, delaying the time of conduit replacement. Another problem recently raised by Ebert,²⁴ of the need of a long ventriculotomy whenever a large conduit is used in infants, and, therefore, postoperative right ventricular impairment, has been avoided by making high transverse incisions into the outlet tract of the right ventricle, avoiding excision of the muscular edges of the ventriculotomy.

Although the use of an extracardiac conduit to achieve a physiological circulation in a small infant cannot be regarded as a total repair,¹⁸ it appears to provide a better chance of survival than conventional palliation. Our results and those of others^{14 15 18} confirm that intracardiac repair of persistent truncus arteriosus can be performed in severely symptomatic infants and that this "open palliation" carries a lower risk than conventional pulmonary artery banding and later "total" repair. Concern remains on the treatment of choice in infants during the first three months of life, in whom poor results have been achieved whatever technique has been used.

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