The Fontan procedure, which was designed to eliminate the possibility of improving long-term left ventricular function by early complete correction.

Summary
The Fontan procedure was applied to correct 13 cases of tricuspid atresia, all with concordant arterial connections. Seven of the patients had had a total of ten previous operations to create palliative shunts. There were 3 early deaths, all due to low cardiac output states, but no late deaths. The postoperative management of the survivors was not unduly complicated, except in the 1st patient. Complete heart block developed transiently in 2 patients. The results are classified as good to very good in all 10 survivors. Our experience supports the opinion that the Fontan procedure gives good symptomatic relief in patients with tricuspid atresia. We discuss the possibilit of improving long-term left ventricular function by early complete correction.

Patients and methods
Thirteen consecutive patients, aged 18 months to 30 years (mean 12.7 years), were operated upon at Groote Schuur Hospital and the Red Cross War Memorial Hospital for Children, Cape Town, between August 1975 and August 1981. Of these patients, 8 were male and 5 female. All patients had concordant atrioventricular and ventriculo-arterial connections.

Seven of the patients had previously undergone a total of ten palliative operations. Four of these patients had functioning Blalock shunt; 1 of these had previously had a Glenn shunt constructed elsewhere but this was no longer functional. Another 3 patients had a functioning Waterston shunt, 1 after a Blalock shunt had failed. One patient had had a pleurodesis done at the age of 2 years. Severe cyanosis and/or progressive shortness of breath were the indications for surgery. All patients were in normal sinus rhythm pre-operatively and satisfied all criteria for the Fontan procedure, i.e. they had large pulmonary arteries, low pulmonary artery pressure and pulmonary vascular resistance, and a well-contracting left ventricle as seen on angiography.

Operative technique
All operations were performed through a median sternotomy and any adhesions present were freed. Both venae cavae were performed through a median sternotomy.

The atrial septal defect was always repaired through the inferior vena cava. The left-to-right shunt at ventricular level was repaired by closing the ventricular septal defect by a direct stitch in 3 patients and by means of Dacron patch in another 3. The ostium infundibuli was closed by a direct stitch in a further 3 patients and with a Dacron patch in 1. One patient had severe pulmonary valve stenosis and this was obliterated by a direct stitch. In the remaining 2 patients there was no shunt at ventricular level, i.e. there was no ventricular septal defect. The methods used to obtain right atrium-to-pulmonary artery continuity are diagrammatically depicted in Figs 1 - 4. In 4 patients the right atrial appendage could be attached to the rudimentary right ventricle or to a large infundibular chamber; anteriorly this anastomosis was completed by the use of a pericardial patch (Fig. 1 a, b).
A valveless conduit was used between the right atrial appendage and rudimentary right ventricle in 6 patients, except that in 1 of these patients a prosthetic valve, a size 31 Saint Jude valve, was incorporated in the atrial appendage because of the relatively large right ventricular chamber and for fear of regurgitation into the right atrium in systole (Fig. 2).

In 2 patients, who had previously undergone operations for a Waterston shunt, a valved conduit containing a Björk-Shiley prosthesis was used to obtain continuity between the right atrial appendage and the right pulmonary artery (Fig. 3). In 1 patient a valved conduit containing a porcine xenograft (Hancock) was anastomosed between the right atrial appendage and the main pulmonary artery (Fig. 4).

**Results**

**Hospital mortality**

There were 3 deaths in hospital (23%). The first one was in a 31-year-old female patient, early in this series. A Carpentier-Edwards porcine xenograft had been inserted into the inferior vena cava, the atrial septal defect closed, and a valveless conduit used between the right atrial appendage and the rudimentary right ventricle. There was severe cardiac failure postoperatively, requiring massive doses of positive inotropic support. One week later respiratory distress developed and the child was then given artificial ventilation. A large pleural effusion was drained with a chest drain. Gradual deterioration with uncontrolled failure, ureaemia, wound dehiscence and sepsis followed, with death in the 3rd postoperative week.

The second hospital death was that of a male patient aged 7 years, with a functioning Waterston shunt. At the previous operation he had bled intrapericardially and we found the adhesions extremely difficult to resect. He also had atrial septal defects, one a coronary sinus defect that was quite difficult to close. He was in severe failure due to low cardiac output after discontinuation of bypass and remained in this condition in spite of full supportive therapy, including ventilation, until he died 2 days later.

The third death was of a 11/2-year-old female infant, the last patient in this series. In view of the good results with the Fontan operation thus far, and in an attempt to eliminate the need for initial palliative procedures, complete correction was undertaken once it had been established that there would be no need for prosthetic valves or conduits in the repair (Fig. 1 a, b). Pulmonary valve stenosis was present and was relieved by open commissurotomy. A size 10 Hégar dilator could then go through the valve. On discontinuation of the bypass the heart took over the circulation poorly, with a very high venous pressure of 25 mmHg and low cardiac output. A few hours later the child also became anuric and it was then decided to enlarge the pulmonary valve ring and main pulmonary artery with a pericardial patch. Improvement was only temporary, and the next day we decided to return the patient to the operating theatre and construct a superior vena cava-to-right pulmonary artery (Glenn) shunt. This would have taken 50% of the load of the failing right atrium. This procedure did improve the patient's condition a little but she died the following day when some of the positive inotropic support was reduced.

**Complications**

Severe right heart failure was seen in only 3 of the remaining 10 patients: 1 presented with severe ascites, sacral oedema and ankle oedema, the other 2 with pleural effusions. All were managed successfully with diuretics alone.

A 4th patient developed low cardiac output failure with cardiovascular collapse and cyanosis 1 week after surgery. He was successfully treated by ventilation, diuretics and positive inotropic support.

Complete heart block was present in 2 patients. They were temporarily paced and both returned to sinus rhythm a few days after operation.

Fast atrial fibrillation was observed in 1 other patient, 8 days after operation, but caused no cardiovascular embarrassment at all; electrical cardioversion was successful.

Deep-vein thrombosis developed on the 10th postoperative day in the right leg of a 13-year-old boy. This was confirmed by a phlebogram, and was managed with intravenous heparin initially followed by the administration of oral anticoagulants. His lesion was corrected by the method shown in Fig. 1, and he will not require long-term anticoagulation.

**Late results**

Three patients were from Europe — 1 from Rumania, 1 from Italy and 1 from Greece. All 3 were well, in sinus rhythm, in good haemodynamic condition and had clear chest radiographs 1 month after discharge from hospital, i.e. at their first outpatient follow-up visit and prior to their return to their respective countries.

The very first patient operated on is now 23 years old and doing very well. She is married and earlier this year consulted a specialist physician in her home town regarding the possibility of starting a family. Her haemodynamic status was found to be good, with no elevation of venous pressure and no liver enlargement, and she was in sinus rhythm. The prosthetic
xenograft valve in the inferior vena cava was functioning well and no calcification was seen on the radiograph.

All the other patients attend clinics and at their last visit were found to be in good or excellent condition. Only 1 is still mildly cyanosed, but is otherwise a healthy active young boy. Those with prothetic valves are maintained on anticoagulants. None of these patients has, as yet, been recatheterized.

**Discussion**

The early results with the Fontan procedure showed a significant morbidity and mortality. As experience increased and when the procedure was simplified (by omitting the use of vena-caval valves) results improved markedly.¹ ² ³ ⁴ ⁵ ⁶ ⁷ ⁸ ⁹ ¹⁰ This procedure was then applied to other forms of complex congenital cardiac malformations, with equally good results.⁹ ¹⁰

We have so far only used this method to treat 'classic' tricuspid atresia. As experience with this procedure increased, we modified the methods used. Inferior vena-caval valves were used only on the first 2 patients. Valve conduits containing a porcine xenograft valve were used only once, early in this series; their use was discontinued when their durability in children was found to be uncertain.

The venae cavae are cannulated separately through the lateral wall of the right atrium near their opening so as to minimize traumatic interference with right atrial contraction. The right atrium is opened only through the right atrial appendage and the atrial septal defect is closed through this opening, mostly with a patch of either Dacron or pericardium.

We believe that in this condition as much use should be made of the rudimentary right ventricle as possible. There is no doubt that it can contribute significantly in the propulsion of blood to the pulmonary artery. This chamber is opened longitudinally and the ventricular septal defect closed, either with a Dacron patch or with a direct suture if small. Often, however, there is a large infundibular chamber with a small os infundibuli, and we have elected in these cases to suture the os infundibuli rather than the ventricular septal defect, thereby avoiding the chance of heart block and also reducing ischaemic time and bypass time quite considerably.

There is no doubt that the correction of choice is anastomosis of the right atrial appendage to the rudimentary right ventricle, roofing this anastomosis with a piece of pericardium.⁹ This will allow for growth of the tissues and it can therefore theoretically be used in very young children, thus avoiding palliative surgery and its associated mortality and morbidity. This has been done successfully elsewhere¹¹ in a 1-year-old infant, the youngest reported in the literature. Our first attempt in this direction failed; in retrospect, failure was due to the fact that pulmonary valve stenosis was present and that in spite of an open commissurotomy this must still have given rise to considerable pulmonary outflow resistance.

When the right atrial appendage fails to reach the rudimentary right ventricle, the anastomosis can be carried out with a valvless Dacron or Gortex conduit of the largest possible calibre compatible with the patient's size.

When a shunt from the ascending aorta to the right pulmonary artery (Waterston) has been constructed previously, it is advisable in taking this down to place a valved conduit between the right atrial appendage and the right pulmonary artery. This distal anastomosis site is enlarged by opening the right pulmonary artery towards the bifurcation of the pulmonary artery. This results in a wide opening and also relieves any obstruction due to kinking or fibrosis. Often the ascending aorta has to be transected to achieve this reconstruction. If pulmonary valve atresia is present, a valved conduit can be placed between the right atrial appendage and the main pulmonary artery.

The results of the Fontan procedure are quite acceptable. Although this operation may be classified as only 'palliative', it gives adequate relief of symptoms and of cyanosis and the patient is able to lead a normal life. However, postoperative evaluation¹² ¹³ of these patients has shown that they all have abnormal haemodynamics even at rest, with more significant abnormality on exercise. The long-term implications of these findings remain uncertain.

Deterioration of left ventricular function in these patients could partly be due to systemic-to-pulmonary artery shunts that were functional over a number of years before the Fontan procedure was undertaken. It would therefore seem reasonable to avoid these shunts by undertaking correction in suitable candidates in infancy whenever the anatomy permits complete repair without the use of prosthetic material and valves. The criteria for the Fontan procedure will have to be even more strictly respected in these cases for a successful outcome.

We wish to express our sincere gratitude to the many members of the medical and nursing staff of Groote Schuur Hospital, the Red Cross War Memorial Children's Hospital and the University of Cape Town Medical School who contributed to the care of these patients, and also to Miss J. J. Bosman, of Groote Schuur Hospital, for sketching and photographing the diagrams and Mrs J. Park for secretarial assistance.

**REFERENCES**


