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FONTAN PROCEDURE FOR CORRECTION OF COMPLEX CONGENITAL CARDIAC ANOMALIES*

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Until recently, surgical treatment of complex congenital cardiac anomalies mainly consisted of palliative techniques for either reducing pulmonary blood flow by pulmonary banding procedure or increasing pulmonary flow by systemic-pulmonary shunt operations. It was only in 1968 that Fontan and Baudet[1] began successfully to treat tricuspid atresia (TA) by creating right atrio-pulmonary communications, and this was the beginning of a new era in the surgical treatment of complex congenital heart diseases.

The original Fontan procedure consists of the closure of the atrial septal defect (ASD) and the ventricular septal defect (VSD), the placing of a valved conduit between the right atrium and the pulmonary artery, anastomosis of the superior vena cava to the right pulmonary artery and placement of a valve in the orifice of the inferior vena cava. Although this original procedure was initially successful[2-4], it was subsequently modified by several leading cardiac surgeons[2-4].

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The possibility of physiological correction without placing a valve in the orifice of the inferior vena cava and anastomosis of the superior vena cava to the right pulmonary artery was realized[5,6]. Because of the disadvantages of valved conduits, the right atrial appendage was anastomosed to either the right ventricular outflow tract or to the main pulmonary artery[7].

The Fontan procedure was initially used for the treatment of tricuspid atresia (TA), but later it became the preferred method for providing a complete physiological correction in high risk complex cardiac anomalies too[8]. For instance, the surgical treatment of the univentricular heart with double atrioventricular (A-V) valves can be accomplished by the closure of the right A-V valve with a patch, by drainage of systemic venous blood into the pulmonary artery through right atrio-pulmonary anastomosis and by the closure of the pulmonary artery at the ventricular outflow thus enabling the functional single ventricle to pump blood into the aorta.

The Fontan procedure was utilized in our clinic on four patients, two with TA and two with univentricular heart. In this report we discuss this procedure in detail with special regard to surgical technique and results.

Material and Methods

A modified Fontan procedure was used on four patients at the Hacettepe Children's Hospital between 1980 and 1983. The ages of the patients ranged from 6-16 (mean 9) years.

Two of these patients had TA and the other two had univentricular heart (Table I). The hemoglobin concentration of these patients ranged from 15.8 to 22.4 (mean 20.2) g/dl.

TABLE I

Summary of Four Cases

Patient	Age (years)	Diagnosis	Palliative procedure	Definitive operation	Follow-up
A.T.	7	Tricuspid atresia	–	Direct anastomosis between the right atrium-right ventricular out-flow tract	Class I* after 3 years
Z.D	16	Tricuspid atresia	Left Blalock-Taussig shunt (1973)	Right atrium-pulmonary artery direct anastomosis.	Class II after 2 years
A.C.	7	Univentricular heart	–	Right sided A-V* valve was closed. Right atrium-pulmonary artery direct anastomosis	Class I after 6 months
S.S.	6	Univentricular heart	–	ASD and right A-V valve were closed. Right atrium pulmonary artery anastomosis	Class I after 4 months

A-V : Atrioventricular
Class : New York Heart Association Functional Class

The first patient had type I - b TA according to the Keith et al[9] classification. The preoperative pulmonary artery pressure of this patient was 15 mmHg.

The second patient had type III TA with a previously performed left Blalock-Taussig shunt and a pulmonary artery pressure of 10 mmHg.

The third patient had ventricular inversion with a double inlet left ventricular type univentricular heart and pulmonary stenosis. The aorta was located anteriorly and slightly to the left of the pulmonary artery. A pulmonary artery pressure of 15 mmHg was measured preoperatively.

The fourth patient also had a left ventricular type univentricular heart with a rudimentary outlet chamber. The great arteria relation was transposed, there was valvular pulmonary stenosis and the pulmonary artery pressure was less than 18 mmHg.

Operation was indicated in each patient because of worsening cyanosis or progressive exercise intolerance or both.

Operative Technique

After median sternotomy, the gross cardiac anatomy was confirmed again, and pressures were recorded. The previously performed Blalock-Taussig shunt in the second patient was not functioning. The superior vena cava was dissected slightly superior to the pericardial reflection and it was cannulated directly. This technique can facilitate access to the retroaortic structures including the superior aspects of the right and left atria and the right pulmonary artery.

The inferior vena cava was cannulated through the right atrium near the caval orifice. After cannulating the ascending aorta, a cardio-pulmonary bypass was established. Moderate systemic hypothermia (28 °C) was used and myocardial protection was achieved with infusion of cold potassium cardioplegia and topical cooling with iced saline solution.

In the first patient an incision into the right atrial appendix was performed and a 3 × 3 cm ASD was closed with a patch. The rudimentary right ventricle was then opened with a longitudinal incision at the outflow tract. Anything smaller than a 1 × 1 cm VSD was closed primarily. A flap was prepared from the anterior wall of the right atrial appendix and sutured to the incision in the right ventricle to form the posterior wall of the tunnel. The procedure was completed by suturing a pericardial patch to form the anterior wall of the tunnel. With this technique systemic venous blood was directed to the pulmonary artery via the right ventricular outflow tract[7] (Figure 1).

Similarly, in the second patient, a right atriotomy was made into the right atrial appendage, the ASD was closed primarily and the right atrial appendage was anastomosed to the main pulmonary artery with an anteriorly placed pericardial patch. The proximal pulmonary artery was divided and each end was sutured.

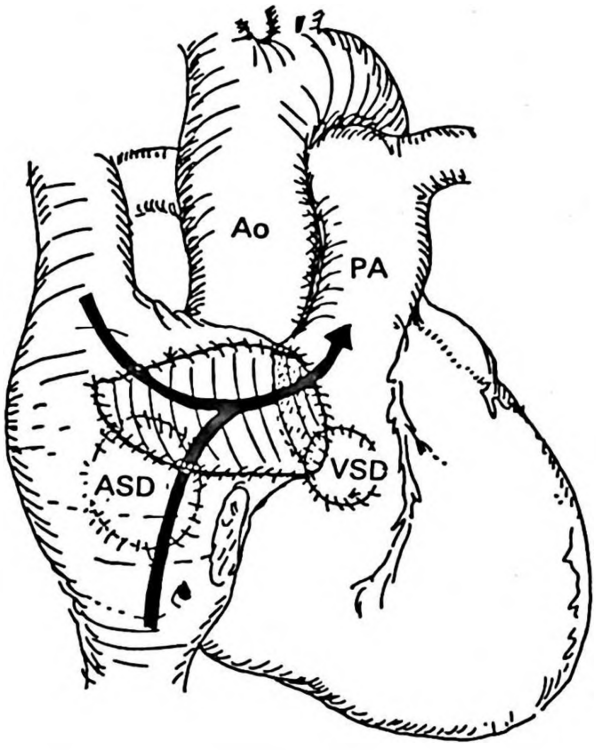


Figure 1

Modified Fontan procedure.
The direct anastomosis of the right atrial appendage to the ventricular outflow tract in combination with anterior enlargement by using a pericardial patch.

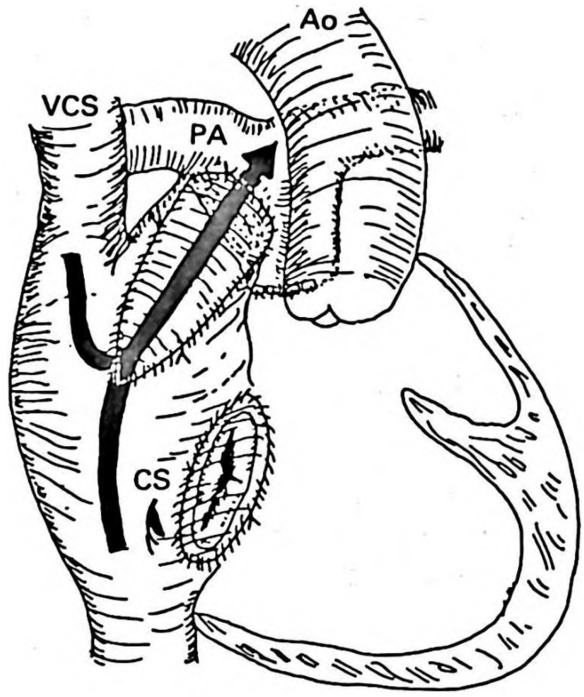


Figure 2

The direct anastomosis of the right atrial appendage to the pulmonary artery with anterior enlargement by using a pericardial patch and closure of the right A-V valve.

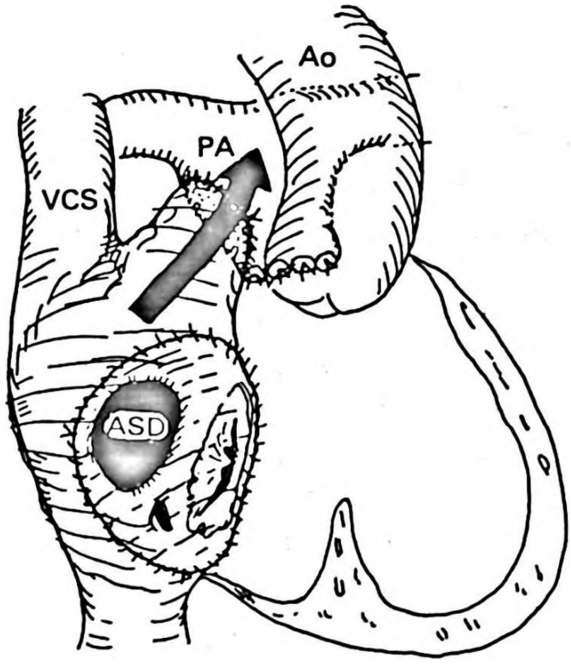


Figure 3

The direct anastomosis of the right atrial appendage to the pulmonary artery and both closure of the right A-V valve and ASD.

Key to Figures

Ao	: Aorta	VSD	: Ventricular septal defect
PA	: Pulmonary artery	CS	: Coronary sinus
VCS	: Vena cava superior	A-V	: Atrioventricular
ASD	: Atrial septal defect		

Approximately the same procedures were followed in the patients with a univentricular heart. The closure of the right A-V valve was achieved by suturing a dacron patch to the annulus. Sutures were placed at the base of the valve leaflets, and care was taken to keep them parallel to the annulus along the posterior and septal leaflets. Thus A-V node and conduction tissue were avoided (Figure 2).

In the fourth, there was a large ASD, so a single patch was used to exclude both the ASD and the right A-V valve. The suture line was carried along the atrial wall 1 cm above the annulus so as not to interfere with the conduction system (Figure 3).

The pulmonary artery was closed by direct sutures reinforced with Teflon pledgets passing through the annulus. The main pulmonary artery was opened with an incision 4 - 4.5 cm long, and the anastomosis which extended from the right pulmonary artery to the right atrium - pulmonary artery was constructed directly into the one and with an anteriorly placed pericardial patch into the other. An effort was made to keep the diameter of the anastomosis as large as possible.

Each patient was supported with a low dose of dopamine in the early postoperative period and extubated a few hours after the operation. We also used intermittent external compression of the lower extremities via airpants (Jobst).

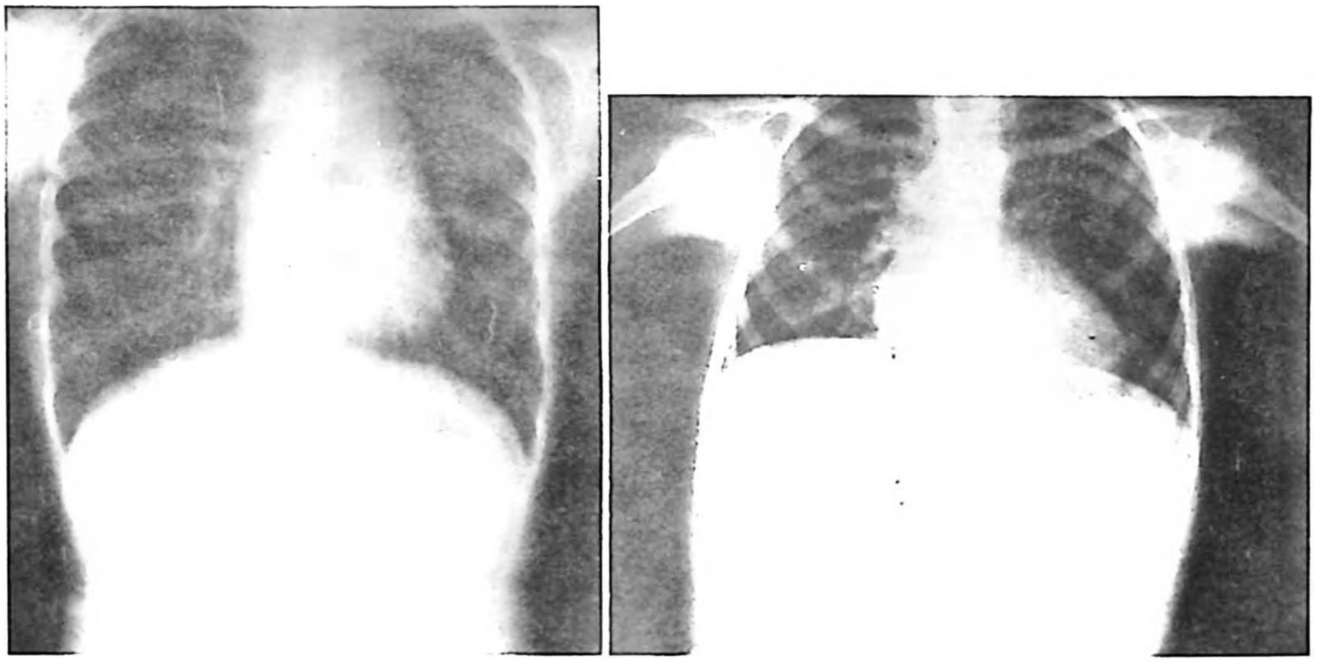
Results

The patients have been followed up from 4 to 36 months. There have been no deaths. One patient with TA showed atrial arrhythmias but this soon returned to sinus rhythm and sinus rhythm has continued in all patients (Figure 4).

Figure 4

Postoperative ECG shows sinus rhythm in the case of univentricular heart.

In the first two patients the liver became enlarged and often tender during hospitalization. There was also right-sided pleural effusion requiring multiple thoracocenteses. In the other two, the postoperative period was uneventful and the hemodynamic condition excellent. In these patients there was no pleural effusion, hepatomegaly, peripheral edema or ascites (Figure 5).



5 – a 5 – b

Figure 5

Showing the postoperative chest X-ray of the third patient (a) and the fourth patient (b).

The first two patients were stabilized with digitalis, diuretics and salt restriction within a few weeks and discharged from hospital in good condition. With the other two only digitalis was given.

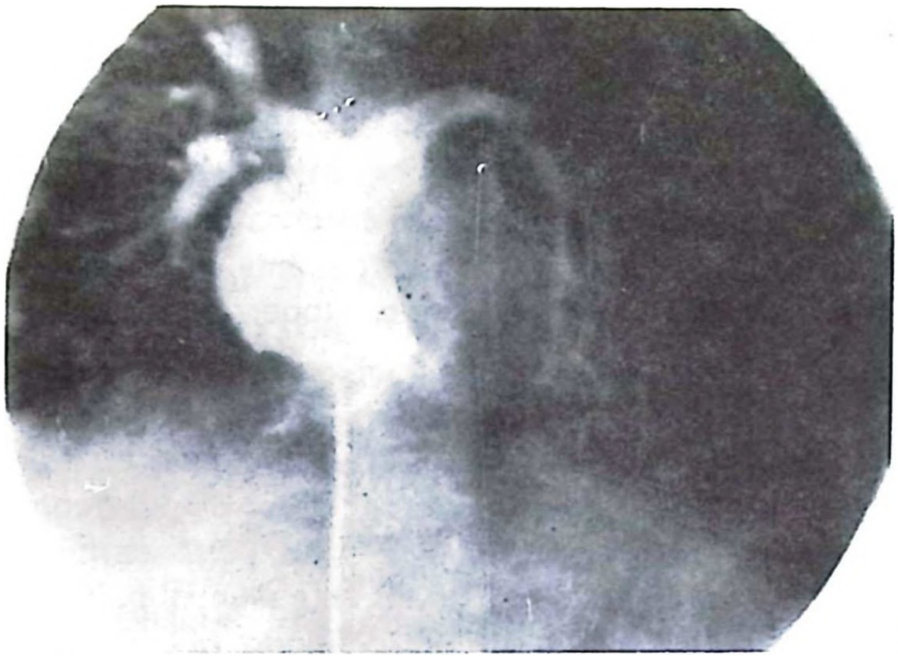
One of the patients with a univentricular heart was reevaluated by cardiac catheterization and angiocardiographically (Figure 6). The right atrial pressure was 10 mmHg and there was no caval regurgitation.

Postoperative check-ups have shown a gratifying functional improvement, with three of the patients completely unrestricted (New York Heart Association Functional Class I) and just one patient mildly restricted (Class II).

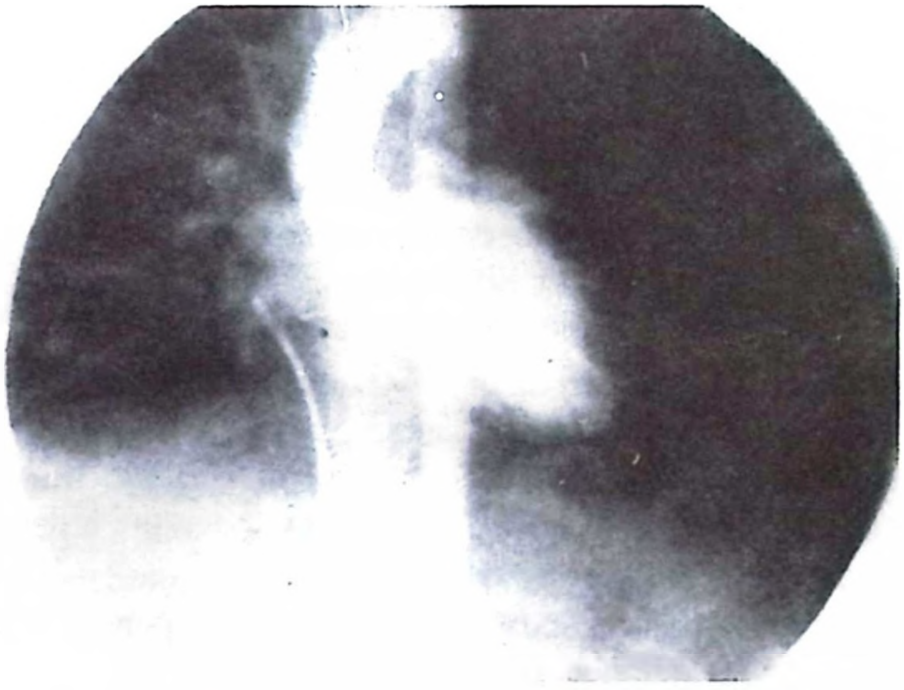
Discussion

After the first successful performance, by Fontan and Baudet[1], of a right atrium to pulmonary artery anastomosis in the treatment of TA, this technique has been used frequently for the relief of severe cyanosis and for improving the clinical status of selected patients with this anomaly. Subsequently this procedure was also used in the treatment of some more complex congenital cardiac malformations[8,10,11].

The Fontan procedure brings into being an atrial dependent circulation in which venous blood flows from the right atrium to the pulmonary artery without the aid of right ventricular contractions. With regard to this procedure, Choussat et al[12]



6 – a



6 – b

Figure 6

Illustrating the postoperative angiocardiographic findings of a case of univentricular heart. Contrast material injected into the right atrium was directly passing into the pulmonary artery (6 – a) and was filling the single ventricle and aorta in the recirculation phase (6 – b).

suggest that certain criteria should be observed so as to ensure a safe and effective operation. The most important of their ten criteria include: an age of 4-15 years, sinus rhythm, a mean pulmonary artery pressure of 15 mmHg or less, a pulmonary resistance of less than 4 U/m^2, a pulmonary artery-aortic diameter ratio of 0.75 or more, left and right pulmonary branches of an adequate size, no left-sided A-V valve incompetence and a left ventricular ejection fraction of 0.60 or more.

Atrial contractions were thought to be important for the blood flow to the pulmonary artery, so sinus rhythm should be preserved whenever possible. But it was reported that, in the presence of acceptable pulmonary vascular resistances some atrial arrhythmias including atrial fibrillation and junctional rhythm could be tolerated well[13,14]. Gale et al[15] reported that with 78% of their patients these criteria were not fulfilled but that they survived surgery. Our patients conformed to these criteria and we have had excellent early and late postoperative results with no mortality.

In the Fontan procedure, while constructing the right atrium pulmonary artery communication, the necessity for valved conduits has proved controversial. Various nonvalved and valved conduits have been used by different surgeons[16-19]. Although Fontan et al[20] still recommend them, experience has shown that the use of a conduit (valved or nonvalved) is not an essential part of the procedure[6-8,15,21]. Because of the long-term complications of conduit stenosis related to valve deterioration or fibrous peel thickening[7,8,21-23], anastomosis of the right atrium directly into the pulmonary artery or into the hypoplastic right ventricular outflow tract (in tricuspid atresia) has been performed and recommended[7,15,17,21]. Further, the minimal space available retrosternally and the lack of conduit growth limit their use among young patients. Due to the unpleasant long-term effects and the limited usefulness of them in children, with all our patients we have performed a direct anastomosis without a valved conduit and have obtained good hemodynamic results[23].

As regards surgical technique, attention should be paid to certain points in order to preserve the right atrial function and the sinus rhythm. Each vena cava should be cannulated directly and the right atrial incisions must be limited. The right atriotomy should be made largely into the right atrial appendage[15,19].

The ASD, if present, and the right atrioventricular valve in the case of a univentricular heart, should be closed via this opening. If ASD is large, this should be closed by using a patch[7,8,15,17].

In the direct communication between the right atrium and the pulmonary artery, the opening must be as large as possible[15]. The diameter of this anastomosis in our opinion should be 4 - 4.5 cm across. So as to obtain a sufficiently large anastomosis, the incision made on the atrial appendage can be extended to the free wall of the right atrium and the anterior wall of the communication may be enlarged by using a pericardial patch[7,8,15,22].

In our last two patients, in whom the anastomoses may be considered to be sufficiently large (approximately 4 - 4.5 cm), no hemodynamic problems such as peripheral edema, hepatomegaly, ascites, pleural effusion etc have been observed. But we consider that the early postoperative abnormalities due to right heart failure in our first two patients were related to the smaller communications (approximately 2-2.5 cm) between the right atrium and the pulmonary artery.

Another step in the Fontan procedure is the closing of the pulmonary artery at the valvular level. This can be achieved by simple ligation or division of the pulmonary artery and the closure of each end with running sutures[15,18]. From our experience with our last two patients we considered that the closure of the pulmonary artery, by means of sutures reinforced with Teflon pledgets, is both adequate and safe.

Recently, the Fontan procedure has been employed in the treatment of some other complex congenital cardiac anomalies in which the anatomic correction was either not possible or carried a high risk. The univentricular heart, double-outlet left ventricle with pulmonary stenosis, double-outlet right ventricle with tricuspid insufficiency, noncommitted VSD and pulmonary stenosis, ventriculoarterial discordance with VSD, pulmonary stenosis and A-V valve straddling are just some instances[8,10,11,17,18,21].

In particular, in cases of the double inlet univentricular heart, the correction consisting of closure of the right A-V valve and construction of an anastomosis between the right atrium and the pulmonary artery has been preferred to anatomic correction[8,18]. The anatomic correction (interventricular septation) carries a high risk because of the difficulties involved in the precise trimming of the interventricular patch and the possibility of conduction tissue injuries[18].

While closing the right atrioventricular valve, a prosthetic patch should be used and the sutures should be placed along the atrial wall 1-2 cm above the annulus in order to avoid the A-V node and the penetrating bundle[8] (Figure 7). For this purpose the right A-V valve and the ASD, if present, can be closed together by using a single patch. This was our method with one of our patients (Figure 3).

In the Fontan procedure, the use of a subpulmonic chamber (communication between the right atrium and the hypoplastic right ventricular out-flow tract – Björk modification) has been recommended for the contribution of the right ventricular contractions[7,19,22].

In the majority of patients with univentricular heart, there are the associated anomalies of pulmonary stenosis and malposition of the great arteries[24]. This means that, from the surgical point of view, anastomosis from the right atrium to the pulmonary artery can more easily be carried out.

The early mortality rate resulting from the Fontan procedure varies from approximately 4 to 26 percent. The most important factor leading to mortality is the relatively high pulmonary vascular resistance. Experience with this procedure has taught us that in the early postoperative period, there will often be such conditions as pleural effusion, hepatosplenomegaly, ascites and peripheral edema, the result of right heart failure; but these usually disappear during the first three or four weeks postoperatively[8,15,19,20].

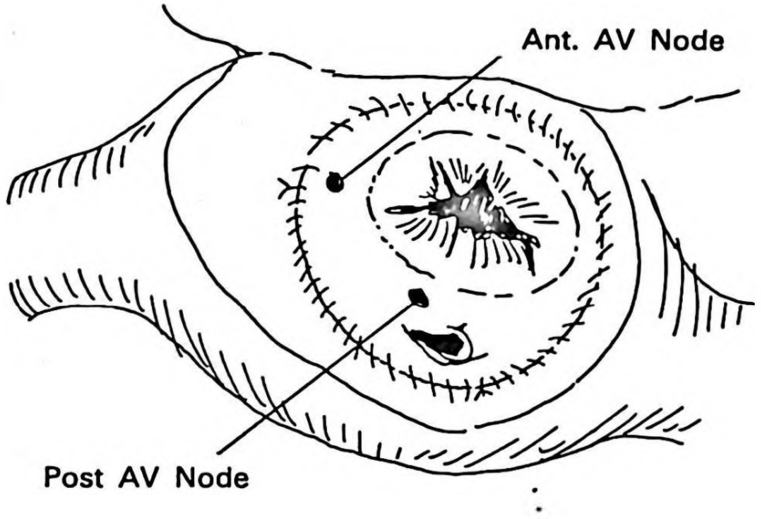


Figure 7
Placing of a prosthetic patch (See text).

Sometimes, however, serious problems of renal failure or of protein-losing enteropathy due to right heart failure have been observed in long-term follow-ups[8].

In cases of a univentricular heart, during the closure of the right A-V valve, A-V block has been reported with an incidence of approximately 30 percent[8]. The above mentioned methods reduce this ratio dramatically. Bearing in mind that the first Fontan procedure was reported in 1971 it is as yet difficult to determine the long-term results. This year Fontan has reported the results of his own series of 100 cases[20]. In this study, 94 of his patients were in Class I or II.

According to the average follow-up periods which in the literature range from several months to 4.5 years, the Fontan procedure can be performed for TA and univentricular heart with a low operative risk and with good hemodynamic results[8,15,20,21].

Summary

Recently, the Fontan procedure has been popularized for the correction of tricuspid atresia, univentricular heart and some other complex cardiac anomalies. In our department this procedure has been used on four patients, two of them with tricuspid atresia and the other two with univentricular heart. In this report, the indications, the surgical techniques and the results are all discussed in detail.

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