TOTAL CAVOPULMONARY CONNECTION IN A 20 YEAR OLD PATIENT: A CASE REPORT

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A 20 year old female patient with a single atrium, single ventricle, common atrioventricular valve, pulmonary artery atresia was treated successfully with total cavopulmonary connection (CPC). Previously, a right Blalock-Taussig shunt procedure had been performed by the same institute in 1976. Total cavopulmonary connection can provide an excellent definitive treatment for a variety of complex congenital heart lesions even on adults. To our knowledge this case is the oldest reported patient in our country for total CPC.

Key words: Congenital heart disease, cyanosis, total cavopulmonary connections.

In 1971, Fontan and Baudet reported the first clinically successful procedure which was totally bypassed the right side of the heart for the treatment of tricuspid atresia [1]. Since the original description, modifications of Fontan procedure have been described allowing its application to a variety of complex congenital lesions. Puga and De Leval described total cavopulmonary connection (CPC)(De Leval Procedure) as a valveless repair which avoids a direct atriopulmonary connection [2,3].

CASE REPORT

A 20-year-old female patient was admitted to the hospital of Hacettepe University School of Medicine because of cyanosis. In 1976 a right Blalock-Tausig shunt was performed due to insufficient distal pulmonary vasculature and since then she had not been seen by use. She was in functional class IV (NYHA). Physical examination revealed a pulse rate of 80/min in sinus rhythm, and a blood pressure of 110/70 mmHg. She had clubbed fingers and promient precordial bulging. A grade 1-2/6 systolic murmur was auscultated over the precordium.A chest roentgenogram showed cardiomegaly. Laboratory examination showed the following values: Hb: 21,1 gm/dl, Hct: 63.3%. Oxygen saturation was 28%. Cardiac angiography was performed and the diagnoses of a

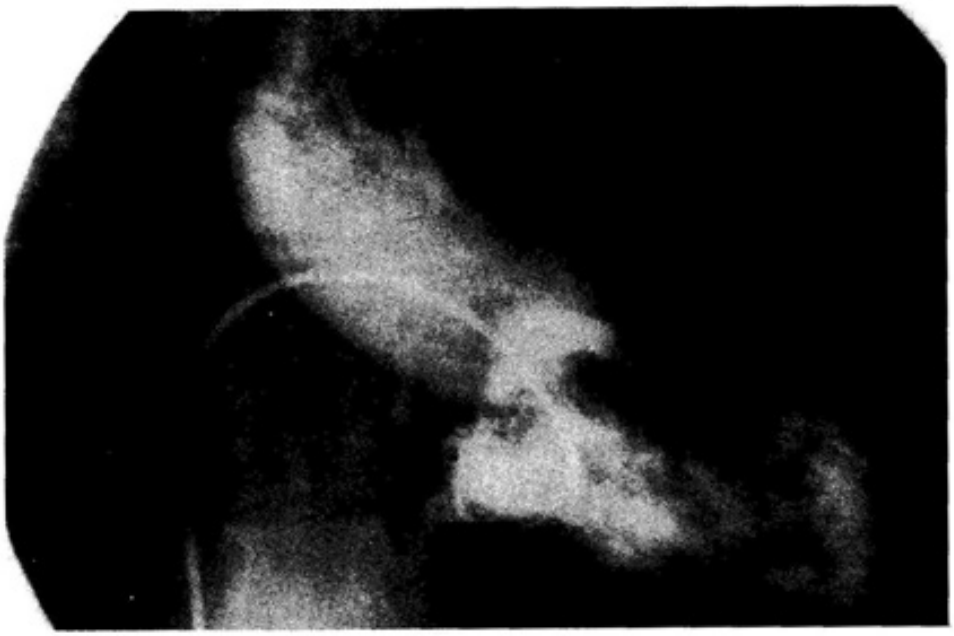

Figure 1: *Preoperative ventriculography displaying single ventricle, pulmonary artery atresia.*

single atrium, a single ventricle, pulmonary atresia, common atrioventricular valve was established (Fig.1).

The operation was performed using extracorporeal circulation (CPB) and direct bicaval cannulation. Before canulation the mean pulmonary artery (PA) pressure was 11 mmHg. The previously performed systemic-pulmonary shunt was ligated as CPB was initiated.Hypothermia, cold crystalloid cardioplegia and interrupted aortic crossclamping was used.

Figure 2: *Schematic drawing of the right atrium showing the tunnel and the cavopulmonary anastomosis. Superior vena cava (SVC), inferior vena cava (IVC), Aort (Ao), right pulmonary artery (RPA), left pulmonary artery (LPA).*

The right atrium (RA) was opened and the single atrium, single ventricle with common atrioventricular valve (Rastelli type C) was observed. A tunnel created by a Dacron patch around the orifices of the superior vena cava (SVC) and the inferior vena cava (IVC), using the posterolateral atrial wall as a posterolateral wall of the tunnel. The tunnel was fashioned to create a conduit of uniform diameter between the orifices of the SVÇ and IVC. The SVC was divided above the cavoatrial junction, and an end-to-side anastomosis was performed between the cardiac end of the SVC and the inferior aspect of the right PA. The distal orifice of SVC was anastomosed to the superior aspect of the right PA (Fig.2). She had an uneventful recovery and was followed up 3 days in ICU. She was in sinus rhythm and small dose of dopamine was given. Arterial oxygen saturation was 80% during intubation, and 70% 3 days after operation. The duration of intubation was 18 hours. The postoperative course was unexpectedly smooth, she was kept on daily dose of digitalis, dipyridamol, aspirin and warfarinsodium (coumadin) keeping the prothrombin time around 20 seconds as well as diuretic administration twice a week.

Postoperative cardiac catheterization and angiograpy demonstrated that the total cavopulmonary shunt was patent and that there was no pressure gradient at the site of the anastomosis (Fig.3). Mean pulmonary artery pressure was 12 mmHg, and mean SVC pressure was 14 mmHg. The arterial oxygen saturation was increased to 80%, and the patient improved to functional capacity class II (NYHA). No signs of ascite, pleural effusion, congestive heart failure, or arrhythmias had been observed. The patient was discharged from the hospital on the 10.th postoperative day.

DISCUSSION

There are several technical advantages to total CPC when compared with other Fontan modifications: The construction of the posterolateral tunnel avoids suture placement near the atrioventricular conduction pathways, and so decreasing the likelihood of the postoperative heart block. Reduction of turbulence prevents energy loss and minimizes the risk of atrial thrombosis, besides improving the hemodynamic performance [3,4]. Also, the risk of operative and early postoperative complications as ascites, pleural effusion, heart failure and heart blocks are less prevalant [3,5]. Although Fontan and modified Fontan procedures including total CPC provides excellent results between 4 and

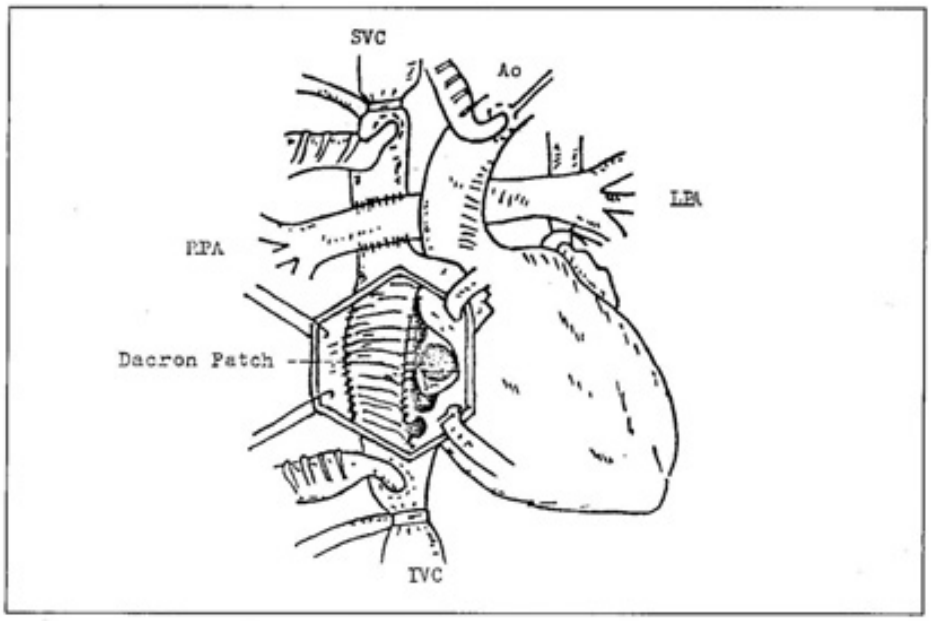

Figure 3: *Postoperative angiography showing the tunnel position.*

15 year-old patients [5,6], the results of older age group are quite acceptable. As far as our knowledge, our patient is the first and the oldest reported case for total CPC in our country.

De Leval procedure is an effective modification of the Fontan procedure which can be performed with relatively technical facility with low morbidity and mortality and may result in improved hemodynamics and reduced late arrhythmias, compared with other Fontan-type procedures [3,5].

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