

# Fontan operation, new results and modifications:

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## **Abstract:**

### **Back ground:**

Since the first Fontan operation, various techniques have been described. These methods have been changed during the time. Now new drugs are also being used beside surgery.

### **Case reports:**

Fifteen patients with mean age of 8 years and diagnosis of tricuspid atresia or D TGA and single ventricle were operated in our hospital. All of them, except one were operated with extra cardiac technique and fenestration was created in half due to CVP (In seven cases).

We had one mortality and three case of postoperative morbidities. The major complications were pleural and pericardial effusion and chylothorax. We treated two patients inflicted with effusion with oral Sildenafil sulfate (Viagra) therapy.

### **Conclusion:**

The mortality in our study was comparable to other studies. We prefer to have a fenestration between conduit and RA even with low CVP.

Oral Sildenafil therapy would reduce pulmonary artery pressure in complicated patients.

### **Keywords:**

Fontan operation, Fenestration, Sildenafil sulfate

### **Introduction:**

Since the first Fontan procedure which was accomplished for repair of tricuspid atresia by Fontan and colleagues in 1971, various techniques have been described. Fontan operation involved constructing a cavo-pulmonary (Glenn) anastomosis, in first patient, a direct anastomosis

between RA appendage and proximal end of the divided RPA was created and in other two patients who had discordant ventriculoatrial connection, an aortic allograft valved conduit was placed between RA and RPA. In all 3 patients a second allograft valve was inserted into IVC ostium. Foramen ovale was closed and pulmonary trunk ligated or divided. This procedure was modified with excision of pulmonary trunk with its intact pulmonary valve from the RV and anastomosis to RA appendage after closing the VSD and ASD.

After many reports of successful repairs, subsequently, direct RA-pulmonary artery connection used by Fontan in his first case has been modified and widely used. The extra cardiac conduit Fontan, in which IVC is disconnected from RA and connected by a prosthetic tube to the RPA outside the heart, has become popular in recent years and a bidirectional superior cavo – pulmonary connection completes the procedure. Then, it was realized that this operation was applicable to many other forms of univentricular, AV connection, for example single ventricle and other anomalies with one severely hypoplastic ventricle (1).

### **Methods and Materials:**

We operated 15 patients between four and 16 years old (mean age of 8 years old) with Fontan procedure between Jan 2005 to Jan 2007 in Rajaei heart center. There was no gender difference among them. Tricuspid atresia was the diagnosis in a half and DTGA and single ventricle in the other. All patients except one were operated with extra cardiac technique. In 3 out of fourteen patients with extra cardiac conduit, anastomosis of SVC to RPA was created by Gortex



tube (because of kinked SVC ), but in other 11 cases SVC was anastomosed to RPA directly. Fenestration was created between extra cardiac conduit and RA at the end of operation, whenever the CVP was higher than 20 mmHg ( in 7 Cases ).

All patients took ASA ( 80 mg Tab ) after operation and the one complicated with recurrent effusions, anticoagulated with Warfarin.

### **Results:**

Postoperative evaluation demonstrated one mortality and a few morbidities. The mortality rate was 6.3% ( 1 of 15 patients) and this patient died 24 hours after surgery due to low cardiac output without response to inotropic agents . This patient was not candidate for reoperation because of no clot formation in Fontan pathway and also existence of LA high pressure .

The overall morbidity was about 20% ( 3/15). We found severe and recurrent pleural and pericardial effusion in two patients (12.6%). there were no fenestration in these two and both were treated with oral sildenafil sulfate . The other complication was resistant chylothorax in one patient which did not respond to high calorie diet associated with water and salt restriction. We reoperated this patient with thoracotomy and thoracic duct ligation and the result was good .

Finally, we should mention that there was no indication for takedown in any of our patients.

### **Discussion:**

Fifteen patients with the diagnosis of tricuspid atresia ,DTGA, or single ventricle and at the mean age of eight years were operated with Fontan procedure and extra cardiac technique ( except in one case ) in our center , Fenestration was also created between extracardiac conduit and RA when CVP was higher than 20 mmHg at the end of operation . The overall mortality and morbidity were 6.3% and 20% respectively. The main complication were pleural and pericardial effusions and chylothorax. The two patients inflicted with pleural and pericardial effusions were treated with Sildenafil sulfate and the other patient with chylothorax needed re operation.

The previous surveys demonstrate variable results about conducting a fenestration between conduit and RA. The same show the influence of fenestration on early and late outcomes following the Fontan procedure. A study in

Mexico on 81 patients determined the global mortality of 28.4% and resulted in 2.8 times more risk of death in patients without fenestration (2).

49 subjects studied in USA showed improvement of short-term outcome with Baffle fenestration by decreasing pleural drainage, hospital length of stay and need for additional postoperative procedure (3).

In other words , some studies did not show any influence of routinely fenestration in extra cardiac Fontan operation. In one study in USA on 81 patients, fenestration was performed selectively in 32 cases. There were two operative death and prolonged pleural drainage occurred in 13 patients , 8 with fenestration and 5 without, therefore, fenestration is not necessary in most Fontan patients with extra cardiac technique and should not be performed routinely (4).

The other study in Brazil on 62 patients compared immediate and late results in patients with or without fenestration .

With fenestration, the mortality , pleural effusions, and pericardial effusions were 7.3% , 41.4% and 29.2% respectively but without fenestration these were 4.7%, 23.8% and 14.2 %respectively .

CVP was higher without fenestration ,and hospital stay were similar in both groups ,therefore these result indicated that atrial fenestration did not improve outcome of patients (5) .

We prefer to have a fenestration between conduit and RA even if CVP is lower than 20mmHg .

We should also mention that oral sildenafil sulfate could be very effective in lowering pulmonary artery pressure , especially in patients with some postoperative problematic symptoms. Other studies also determined similar results.

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