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**Later Fontan Patient**

Though most postoperative patients after Fontan are asymptomatic, but late symptoms occurring in an adult or during adolescent age are not uncommon.

## 18.1 Symptoms & Signs

Diminished exercise tolerance, fatigue, and palpitations are most probably related to:

- i. Obstructed Fontan circulation.
- ii. Ventricular dysfunction.
- iii. Increased A-V (atrioventricular) valve insufficiency and or
- iv. Arrhythmia.

Lower extremity edema or ascites with or without above symptoms are related to protein losing enteropathy (PLE).

*In an asymptomatic patient*, the physical examination is unremarkable with no significant murmurs but with presence of a single S<sub>2</sub> only on auscultation.

*In symptomatic patients* who present with significant hemodynamic abnormalities, the physical findings are salient as well as in patients with A-V valve insufficiency, in patients with significant pulmonary collateral blood flow, subaortic stenosis, and an obstructed Fontan.

*Obstructed Fontan circulation:* The jugular venous pressure is elevated.

BVF (bulbo-ventricular foramen) obstruction: A loud systolic ejection murmur is heard at the base.

A-V valve insufficiency: Apical pansystolic murmur or murmur at the left lower sternal border is heard.

## 18.2 Diagnosis & Management

The investigations commonly used to evaluate the symptomatic Fontan patient during the late postoperative period which varies by weeks and or several months include the following:

Chest x-ray, ECG, 2D echo, cardiac catheterization, Holter studies, and O<sub>2</sub> saturation.

The usual uneventful postoperative Fontan patient should have a normal or near normal oxygen saturation.

If the patient had significant cyanosis → persistent right-to-left shunt (if the patient had an atrial fenestration).

With the use of extracardiac conduits for the Fontan, only a few centers are performing fenestrations.

Fenestrations may be closed by a device in the cardiac catheterization laboratory.

Closure of a fenestration corrects cyanosis, improves oxygenation, reduces the need for anti-congestive medications, and prevents paradoxical embolization.

(Balloon occlusion of a fenestration, temporarily, may be useful in order to determine the effect of the closure on hemodynamics. If aortic pressure is stable and RA pressure does not increase significantly, it is considered appropriate to close the fenestration).

1. Leak of the IVC into right pulmonary artery baffle:

These baffle leaks are often small and do not cause significant cyanosis, and these can be closed by a transcatheter device.

2. Persistent left SVC:

It causes significant right-to-left shunting by draining venous blood into the coronary sinus or directly into the left atrium.

3. Intrapulmonary shunts through AV fistulae:

These are often encountered in patients with the bi-directional or classic Glenn anastomosis.

## 18.3 Arrhythmia

Atrial tachyarrhythmias are frequent in Fontan, especially in classic atrial-PA anastomosis. Usually, these are associated with atrial enlargement and right atrial thrombus.

Result in a high incidence of significant AV valve regurgitation and ventricular dysfunction.

(The incidence of arrhythmia may be decreased in extracardiac Fontan which requires less direct manipulation or suturing of the atrial tissue).

*Cardioversion:*

Sustained atrial arrhythmia usually requires cardioversion followed by anticoagulation for 4-6 weeks.

*Antiarrhythmic Drugs:*

Since recurrent arrhythmia is common, prolonged drug therapy is often required to maintain the sinus rhythm.

Before prescribing the drug for chronic use, take into consideration the following three limitations:

1. Drug induced sinus node and AV conduction system abnormalities.
2. Limited drug efficacy for chronic use.
3. Presence of ventricular dysfunction.

Amiodarone may be used in patients with poor ventricular function and refractory arrhythmias.

*Radiofrequency ablation:*

Electrophysiology studies have demonstrated that after the Fontan operation, reentrant atrial tachycardia occurs in the areas of non-conductive atriotomy scars surrounding the prosthetic material, adjacent anatomic landmarks such as the SVC or IVC, and the atrioventricular annulus. These zones may be amenable to interruption with a line of block between the surgical and anatomical barriers by radiofrequency energy. The results are variable in terms of maintaining the long-term sinus rhythm.

*Surgical ablation & Revision:*

In severe cases or in cases of failure of above treatment options, a surgical MAZE procedure with epicardial DDD pacing should be done. It is carried out in conjunction with revising the classic RA-PA Fontan to a lateral tunnel or extracardiac conduit Fontan. Results are favorable.

*Pacemaker implantation:*

Epicardial pacing is an option for patients with sinus node dysfunction or heart block and results are similar to patients with biventricular physiology.

## **18.4 Protein-Losing Enteropathy**

It is a serious and late complication, and the incidence ranges from 1.5 to 11%.

Stigmata: Fatigue, malaise, ascites, pleural effusions, diarrhea, hypoalbuminemia, and lymphocytopenia.

### 18.4.1 Etiology

It is not known though it is assumed to be due to an elevated systemic venous pressure in the Fontan circuit.

In one study, the postoperative systemic venous pressure was not significantly correlated with this complication, but perioperative cardiac injury, longer CPB time, and the single right ventricular anatomy were the only perioperative risk factors that were associated with development of PLE.

Other potential risk factors, such as postoperative systemic venous pressure, pre-Fontan measurements of systemic venous pressure, cardiopulmonary bypass time, single right ventricle anatomy, gender and age at the Fontan operation, anomalous venous drainage, atrioventricular valve regurgitation, pulmonary artery distortion, and pulmonary artery resistance were not significantly correlated with this complication.

### 18.4.2 Management

Cardiac catheterization: Perform at the onset of PLE to study any evidence of abnormal hemodynamics or an obstruction in the Fontan circuit.

*Symptomatic treatment:*

- i. Diet: High protein and high medium-chain triglyceride dietary intake.
- ii. Albumin infusion.
- iii. Diuretics.
- iv. Intestinal cell membrane stabilizers, such as oral steroids and heparin, showed improvement in some sporadic cases.

*Surgical procedures:*

- i. Fenestration: In some refractory cases, fenestration of the systemic venous baffle improved hemodynamics and lower the pressure in the systemic venous system.
- ii. Cardiac transplantation: In unresponsive patients and in dire circumstances, it is an ultimate indication.

## 18.5 Ventricular Dysfunction

*Diastolic ventricular dysfunction:* Poor diastolic compliance with abnormal ventricular relaxation in pediatric patients, if becomes progressive, would lead to later *systolic dysfunction* with ↓ systolic performance over a time.

Incidence of heart failure of systemic right or single ventricle is high in Fontan as compared to congenitally corrected TGA or post Mustard or Senning repair of TGA.

*Medical:* Diuretics, digoxin, and afterload reduction with ACE-inhibitors.

(At present little data are available regarding optimal medical treatment for this condition).

*Surgical:*

- i. Lateral tunnel or external conduit:

In severe ventricular dysfunction despite maximal medical therapy, if patient had classic RA-PA Fontan, it is converted into a lateral tunnel or extracardiac (external conduit) Fontan. It has been effective in increasing cardiac output and alleviating symptoms, and the procedure is done in conjunction with a MAZE procedure since chronic atrial arrhythmias are frequently associated with a Fontan failure.

ii. Cardiac transplantation:

It is the treatment of choice if other treatments were ineffective, and the results have been fair.

## **18.6 Thromboembolism**

Thromboembolic events are not infrequent (33%.) after the Fontan operation.

### **18.6.1 Predisposing Factors**

Atrial arrhythmias, sluggish blood flow, right-to-left shunts, and hypercoagulable states are commonly implicated. Though the role of anticoagulant treatment is not clear, following recommendations may be addressed.

### **18.6.2 Warfarin**

May be used for prevention, in patients with a dense spontaneous echo-contrast in the right atrium.

### **18.6.3 Antiplatelet Therapy**

Therapy was generally considered not effective.

### **18.6.4 Aspirin**

Patients treated with 81 mg of aspirin found no evidence of either thromboembolic events or asymptomatic thrombus on transthoracic echocardiography on a long term.

(Transthoracic echocardiography is a less sensitive test as compared to transesophageal echocardiography in the detection of an intracardiac thrombus).

## 18.7 Pregnancy

In normal pregnancy the cardiac output ↑ by 30-40%, intravascular volume ↑ by 50%, stroke volume ↑, heart rate ↑, systemic vascular resistance ↓, and pulmonary vascular resistance ↓.

Pregnant patients with Fontan have a limited hemodynamic reserve to adapt to the above changes, and the venous pressures are elevated. There is an increased risk of atrial arrhythmia, heart failure, and an increased incidence of spontaneous abortion and pre-term labor.

