Use of the Total Artificial Heart in the Failing Fontan Circulation

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Disclosures: None

Funding: None
The Fontan operation is the final planned palliative operation for children with functional single ventricles (SVs). This circulation is usually tolerated initially, but eventually fails secondary to chronic low cardiac output and elevated venous pressure.

Because of the passive flow of blood through the pulmonary vascular bed, inadequate preload is often the underlying cause of diminished cardiac output in the failing Fontan circulation.

In the failing Fontan circulation, a systemic VAD does not address this fundamental issue and will not necessarily reduce the systemic venous pressure. Indeed, filling and function of the VAD is likely to be limited by inadequate preload.
Implantation of a total artificial heart (TAH) will restore a “normal biventricular circulation” and reverse systemic manifestations of Fontan failure, improving patient candidacy for transplantation.
Patient

A 13 year-old male with pulmonary atresia and intact ventricular septum developed severe, refractory circulatory failure 10 years after an extracardiac Fontan operation.

Hospitalized at an outside institution with severely depressed left ventricular systolic function, respiratory failure, hepatic insufficiency with coagulopathy, and plastic bronchitis.

On transfer, the patient was in extremis with multi-organ failure and respiratory decompensation.

Echocardiography and catheterization confirmed severely depressed ventricular function with elevated Fontan pressures.

With informed consent from the family, a 70cc Syncardia TAH was implanted.
Extra-cardiac Conduit Fontan
Excision of Native Heart and Fontan Conduit

- SVC
- Reconstructed PA
- Remnant of RA
- IVC
- Aorta
- LA with cuff of ventricular muscle
Anastomosis of Atrial Connector to LA

Atrial Connector
Creation of Neo-Right Atrium
Creation of Neo-Right Atrium
Insertion of Arterial Connectors
Insertion of Artificial Left Ventricle
Completed Implantation
Postoperative Course

The postoperative course was complicated by bleeding and external compression of the neo-RA and reconstructed IVC that required re-exploration and eventual stent placement.
Compression of Neo-Right Atrium
Postoperative Course

Additional complications included rhabdomyolysis and renal failure necessitating dialysis. Therapy with inhaled tissue plasminogen activator (tPA) and rigid bronchoscopy were necessary to clear the airway of the casts resulting from plastic bronchitis.
Post-implantation Organ Function

Postoperative Day

AST (U/L)
Post-implantation Organ Function

Lipase (U/L) vs. Postoperative Day

- Postoperative Day: 0, 5, 10, 15, 20, 25, 30
- Lipase (U/L): 0, 500, 1000, 1500, 2000, 2500

The graph shows a significant increase in Lipase levels post-implantation, peaking around the 0-10 day mark, followed by a gradual decline towards baseline values by around the 30-day mark.
Cardiac output was well preserved and generally remained above 5 liters per minute. Over several weeks, the patient demonstrated recovery of all end-organ function, including resolution of plastic bronchitis.

The patient underwent rehabilitation therapy and was able to ambulate without assistance. On postoperative day 61, a suitable donor heart became available and the patient underwent orthotopic heart transplantation.
Completed Implantation
Beginning LA Anastomosis
Completed LA Anastomosis

SVC

IVC
Interposition Grafts for Caval Anastomoses

SVC

IVC
Completed Transplantation
Conclusions

This case represents the first successful use of a TAH as a bridge to heart transplantation in the failing Fontan circulation.

Implantation of a TAH can restore a “normal biventricular circulation” and reverse systemic manifestations of Fontan failure improving patient candidacy for transplantation.