

**Patient/Family Lay Summary:** FON Case Review Conference January 2022

**Coordinated by:** Texas Children's Hospital

**Topic:** Advanced Therapies for the Failing Fontan Circulation

### **The Problem**

Cardiac dysfunction can complicate the decision to achieve Fontan circulation as well as surgical repairs in patients with existing Fontan circulation. Recent data is encouraging for the use of mechanical devices to support heart function within the context of Fontan circulation. When single ventricle patients are marginal Fontan and heart transplant candidates, determining if a patient would do well with such devices and the timing of placement of such devices can be a challenge to their care team. Additionally, in patients who have been living with Fontan circulation and require further heart surgery, heart dysfunction can complicate such additional heart procedures, bringing such devices into consideration to help their heart recover in the setting of already complicated Fontan circulation. Different mechanical support devices have various advantages and disadvantages. We discuss the use of an Impella device in this scenario.

### **Clinical Course of the Cases Presented**

Case 1 is that of a pediatric patient prenatally diagnosed with an unbalanced atrioventricular septal defect and small aortic arch who was found to have concerning regurgitation of the atrioventricular valve (AVV) from birth. The patient underwent Stage I palliation consisting of Norwood operation with a Sano shunt, the postoperative course being complicated by cardiac dysfunction and significant AVV regurgitation. Despite achievement of Stage II palliation with a Glenn procedure at 4 months of age, the patient remained with concerning cardiac dysfunction and valve regurgitation, which persisted despite medical therapy optimization unto the time of traditional consideration for a Fontan procedure. The persistent heart dysfunction and valve regurgitation caused multiple admission for the patient and preempted ability to complete his palliation with a Fontan: ultimately he was listed for heart transplant. Progressive desaturation and decompensated heart failure ensued while the patient awaited a heart transplant and our center opted to place a HeartWare ventricular assist device and concurrently perform the Fontan operation in an effort to support his heart function and improve his desaturation respectively while he awaited heart transplant. Approximately 18 months later he underwent successful heart transplant.

Case 2 is that of a young adult with complex single ventricle anatomy who had undergone three-staged palliation culminating in a fenestrated extracardiac Fontan within the first

three years of life. Over the years he unfortunately developed severe AVV regurgitation, heart dysfunction, and Fontan-associated liver disease, which led to symptomatic swelling and recurrent hospitalization.

The valve regurgitation and dysfunction led to an untenable state of swelling at which time options included repair of the very leaky AV valve or heart transplantation. The patient did not wish to undergo a transplant and replacement of the AV valve was performed, with knowledge that his heart dysfunction may require the need for mechanical device assistance thereafter. Postoperative course became acutely complicated by ventricular tachycardia and complete heart block about a week after surgery, the latter for which a pacemaker was placed. His heart function deteriorated in the setting of his newly competent AV valve and the aforementioned arrhythmias and to improve cardiac output, an Impella device was placed. This device assisted in the recovery of his heart, allowed him to wean to oral medications, and permitted to move forward with all aspects of rehabilitation including extubation and mobilization.

### **Important Points, Lessons Learned, and Potential Solutions**

- Case 1: This was a unique case of concurrent construction of Fontan circulation *with* placement of a ventricular assist device, the decision for each driven by their respective effects to support this patient towards eventual heart transplantation.
  - Ventricular assist device (VAD) support of patients with Fontan circulation, especially when failure is due to ventricular dysfunction, has demonstrated favorable outcomes.
  - VAD support with Glenn circulation can be more challenging for multiple physiologic reasons.
  - In rare cases, when cyanosis and cardiac dysfunction precede Fontan completion, concurrent construction of Fontan circulation with placement of a ventricular assist device may be considered.
  - This approach may optimize candidacy for and clinical status going into eventual heart transplantation. It also may support growth and rehabilitation, maintain end organs, and ultimately allow for a bridge to transplantation.
- Case 2:
  - AV valve repair/replacement may be high-risk in Fontan patients due to potential postoperative challenges. If considered (alongside options such as transplantation), management strategies include:
    - In-hospital optimization to improve baseline status before intervention
    - Observation for declining function and awareness of abnormal heart rhythms
  - In certain situations, short-term Impella device placement can be considered to foster recovery of heart function after surgical re-interventions in Fontan patients

- Potential advantages of the Impella device for heart dysfunction include allowing patients to wean from IV medications and conversion to oral medications to support the heart as well as earlier patient mobilization and rehabilitation.