case report - congenital

Ventricular assist device in univentricular heart physiology

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Abstract

The use of mechanical cardiac assistance is well established as a bridge to orthotopic heart transplantation (OHT) or to recovery for patients with congestive heart failure, however, the experience in single ventricle (SV) physiology is still limited. We report two cases of mechanical assistance in patients with SV physiology: a 2-year old male with hypoplastic left heart syndrome who underwent Norwood Stage I and II followed by HF and a 4-year old female with a univentricular heart who developed a severe right ventricular dysfunction 2 years after a cavopulmonary shunt. Mechanical support utilizing ventricular assist devices (VADs) is considered a valid tool to bridge patients with congestive heart failure to either OHT or to recovery. Increasing experience and improved outcomes utilizing this technology in children with biventricular hearts have led to considering employing these devices in failing SV treatment. We present 2 cases of terminally ill children with SV who were assisted with a VAD.

Keywords: Congenital heart disease • Ventricular assist device • Univentricular hearts

CLINICAL SUMMARY

Patient 1

A 2-year old, 8 kg boy, with mitral and aortic valve atresia, hypoplastic left ventricle was referred to us for congestive heart failure. The patient underwent a Norwood–Sano at birth and a bidirectional cavopulmonary anastomosis procedure at 8 months. The aortic arch was reconstructed with pulmonary homograft, and no calcifications were found in subsequent surgical procedures. After 5 months, severe tricuspid insufficiency and right ventricular dysfunction appeared. A tricuspid valve repair using the De Vega technique was performed. At cardiac catheterization, a mean pressure of 18 mmHg in the cavopulmonary system and a mean atrial pressure of 15 mmHg were found. Extracorporeal membrane oxygenation (ECMO) was started due to clinical worsening. A systemic right atrial (inlet) and aortic cannulation (outlet) were placed using Berlin Heart EXCOR® (BH) cannulae. After 2 weeks, subsequent to lung recovery, we switched to a 25 ml BH support, utilizing the previously placed cannulae. Accordingly, clinical conditions improved, but mean pressures in the cavopulmonary system remained high. After 2 days of mechanical support, a donor heart became available and OHT was performed. The patient was extubated on postoperative day (POD) 3 and discharged home on POD 32. Sudden death occurred at home 6 months after OHT.

Patient 2

A 4-year old, 8.5 kg girl, with {S;D;S} double inlet right ventricle with mitral valve atresia, hypoplastic left ventricle and pulmonary stenosis with normal aortic and pulmonary artery relations underwent an atrial septectomy with bidirectional cavopulmonary anastomosis at 2 years of age. Her general clinical conditions remained stable for almost 2 years although she developed severe right ventricular dysfunction and worsening of tricuspid regurgitation. Cardiac catheterization showed a mean pressure of 12 mmHg in the cavopulmonary system with a transpulmonary gradient of 3 mmHg. Over the next few months, her heart-failure symptoms worsened and she was listed for OHT. However, her clinical conditions continued to deteriorate and a 25 ml BH was implanted with systemic right ventricular (inlet) and aortic cannulation (outlet). Her initial postoperative course was characterized by an immediate improvement of haemodynamics with pulse oximeter values of around 80% and a mean pressure of 10 mmHg in the cavopulmonary system. On POD 2 she was extubated and on POD 10 she was discharged to the ward. After 2 weeks from VAD implantation, a pump exchange was performed. Despite adherence to a standard anticoagulation protocol on POD 51 a large thrombus appeared on the outflow valve. This required another pump change with a major cerebral complication, namely a significant right-side cerebral ischaemia with left-side paralysis. Over the next few weeks, there was a partial neurological recovery. After 166 days of mechanical support, the patient unfortunately died of thromboembolic complications waiting for OHT.

DISCUSSION

Mechanical assist devices have proved to have the ability of promoting the recovery of haemodynamically strained organs and prolonging the life of OHT candidates [1]. This is of crucial importance in view of the shortage of organ donors. As previously reported, VADs in the failing single ventricle (SV) circulation have

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the potential to allow circulatory, metabolic and pulmonary recovery, however, paucity of data precludes recommendations regarding a preferred surgical technique and postoperative management [1–4]. Experience based consensus is useful to formulate guidelines on VAD assistance for SV physiology.

To our knowledge, VAD implants for failing SV physiology are described in several case reports: 6 patients with Fontan circulations, 2 with bidirectional cavopulmonary anastomosis and 2 with a systemic to pulmonary shunt. Patients with bidirectional cavo-pulmonary anastomosis, as in this case, are considered better candidates for mechanical assistance compared with patients with shunt-dependent or Fontan circulation [1, 2]. These patients can be supported with a systemic ventricular inflow and aortic outflow cannulation technique. In patients with right dominance, the inflow cannula is inserted on the diaphragmatic part of the right ventricle. This may be performed under transoesophageal echocardiography guidance, to allow positioning of the cannula as far as possible from the papillary muscles (Fig. 1). In hearts with a left dominance, the inflow cannula could be inserted in the left ventricular apex. A larger pump size is needed to support the systemic and pulmonary systems. This strategy has the potential advantage of maintaining systemic cardiac output in the presence of increased pulmonary vascular resistance, but is probably responsible for an increase mean cavopulmonary pressure [2, 4].

VAD support in the failing Fontan circulation poses several surgical options and challenges [3, 4]. These are variable and greatly influenced by individual patient characteristics. As previously described, the failing Fontan circulation can be supported in different ways such as: the use of a single LVAD when the cause of failure is ventricular dysfunction [3], a single RVAD, when the failing of single ventricle physiology appears with high venous pressures and multiorgan dysfunction but with normal ventricular function [4], and BVAD when the two mechanisms of failing physiology occur together. In the second option, implantation of RVAD requires a revision of the Fontan pathway to allow the separation of the systemic venous and pulmonarycirculations.

Ventricular cannulation is preferred as an inflow option because of lower thromboembolic risk. In one case (Patient 1), we used the atrial approach to avoid a second surgical procedure.

The management of patients with SV failure assisted with VADs remains challenging. Our experience, especially in cyanotic patients, demonstrates that the implementation of a strict coagulation protocol is crucial for a good outcome. For these reasons, we modified our anticoagulation protocol reported previously [5]. Postoperative anticoagulation is started with intravenous heparin and dipyridamole (10 mg/kg/day) 24 h after intensive care unit admission. Heparin infusion is titrated to adjust the partial thromboplastin time between 60 and 80 s. A chronic anticoagulation regimen is initiated on POD 3 with warfarin to keep the INR between 3.5 and 4. Aspirin (5mg/kg/day) is added. Management of end-stage failure of the SV circulation remains challenging, however the potential advantages and lack of reliable alternatives will lead to an increased use of VAD support in the future.

**Conflict of interest:** none declared.

**REFERENCES**