The use of a CircuLite micro-pump for congenitally corrected transposition of the great arteries

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Abstract

We report a case of a 49-year old male with a congenitally corrected transposition of the great arteries (ccTGA) implanted with a left atrial to right subclavian artery ventricular assist device (CircuLite) because of the failure of the anatomic right (systemic) ventricle. Additionally, elevated pulmonary pressures and peripheral vascular resistance (7.4 Wood units) prevented him from being put on the transplant list. The implant, performed off-pump through a right minithoracotomy, was uncomplicated and there were no adverse events. Within 1 month of the implant, there was a marked improvement in exercise tolerance and decreases in pulmonary pressures and resistance, so that the patient was able to return to work and became eligible for transplant listing. As of the time of writing, the patient has been supported for 10 months and is awaiting a heart transplant.

Keywords: Mechanical circulatory assistance • Congenitally corrected transposition of the great arteries • Minimal invasive

With improvements in palliative and corrective cardiac surgery in infants and young adults, more congenital heart disease patients are surviving to adulthood. However, in many instances, treatments delivered early in life do not provide a permanent solution, and a relatively large number of these patients require advanced cardiac care as they age [1]. Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart defect that accounts for <1% of patients with the congenital heart disease. In patients with this anomaly, the anatomic left atrium connects to the anatomic right ventricle (RV) from which the aorta originates and thus serves as the systemic ventricle. In a significant proportion of these patients, the anatomic RV dilates, hypertrophies and, eventually, becomes dysfunctional and patients present with end-stage heart failure [2].

Orthotopic heart transplantation is the treatment of choice in ccTGA, but is available to few patients. Ventricular assist devices (VADs), which play an increasingly important role in the treatment of end-stage systolic heart failure, could also be an alternative for failing ccTGA. However, the complex anatomy and physiology of ccTGA and the general lack of experience with VADs in congenital heart disease limit the widespread use of this application. In addition, currently available VADs connect to the heart via cannulae placed through the LV apex with the tip inserted 2–3 cm into the cavity. Although there are reports of these devices being used in the RV position, including case reports of ccTGA [3, 4], they may not be ideally suited for this application. This is because the anatomic features of the RV renders positioning of the inflow cannula more difficult than in the usual LV position.

However, the position and function of the atria are anatomic ally and physiologically normal in ccTGA. This suggests that an assist device using an atrial inflow cannula instead of a ventricu lar apical inflow cannula might prove advantageous. We report here the first successful use of the Synergy® Surgical System (CircuLite, Saddle Brook, NJ, USA) [5], which sources blood from the left atrium and returns it to the right subclavian artery, in a patient with ccTGA and severe heart failure.

The patient is a 49-year old male who had a congenitally corrected TGA and a ventricular septal defect that was closed at the age of 20 years (in 1981). A pacemaker was placed postoperatively because of an AV dissociation. He was asymptomatic for more than 20 years. A screening echocardiogram performed in 2003 revealed a grade III regurgitation of the anatomic tricuspid valve and a right (systemic) ventricular ejection fraction of ≏40%. Follow-up examinations over the ensuing years showed a progressive decline in systemic ventricular function and the development of severe pulmonary hypertension. Despite maximal tolerated medical therapy, his condition deteriorated rapidly in March 2011 when he presented with NYHA class IV symptoms, at which time the pacemaker was upgraded to a CRT device. However, a right heart catheterization showed continued worsening of haemodynamics (Table 1, pre-op data) and heart failure symptoms, including cachexia and reduced exercise capacity. The very high pulmonary pressures and pulmonary vascular resistance (PVR), however, prevented him from being listed for heart transplant. This left only mechanical support as an option both for improving the quality of life and, potentially, for reducing his PVR in order to improve candidacy for heart transplant.

The Synergy Surgical System was implanted in June 2011 in an off-pump operation through a small right-sided thoracotomy with the micro-pump positioned in a sub-pectoral pocket (Fig. 1). Pump
speed was increased to 23 000 rpm during the implantation under echocardiographic and haemodynamic monitoring. No further pump speed adjustments were necessary later on. After weaning from inotropic support, heart failure medication was started again (diuretics, ACE-inhibitor and beta-blocker). The patient was extubated within 24 h and discharged after 22 days. His symptoms improved from NYHA class II at the time of discharge. Objective evidence of improved clinical status was a VO2 max that increased from 9.6 preoperatively to 14 ml/kg/min within 1 month of implant which has been maintained. NT-proBNP levels decreased from 2227 ng/l preoperatively to 995 ng/l within 2 months. Follow-up right heart catheterizations showed significant and sustained improvements in all parameters and, most notably, a marked decline in PVR (Table 1). As a result, the patient has been listed and is awaiting heart transplant. At the time of this report, the patient has been on support for 10 months, is fully ambulatory and has returned to work. There have been no serious adverse events to date.

**DISCUSSION**

The scarcity of donor organs and the mortality risk of heart transplantation in congenital heart disease suggest that there could be an increased role for mechanical circulatory support in these patients. Case reports and small series have appeared, describing anatomic right (systemic) ventricular LVAD implants in patients with transposition of the great arteries and returning to work. This new VAD technology may provide a viable option for treating systemic heart failure in patients whose in situ invasion also has the advantages of simplifying and reducing risks of an eventual heart transplantation surgery, especially in the case of a patient who has already undergone a surgical procedure involving a sternotomy.

In our case, we implanted a small, partial support system whose inflow is placed in the left atrium in an off-pump procedure through a small right thoracotomy. Avoidance of mediastinum invasion also has the advantages of simplifying and reducing risks of an eventual heart transplantation surgery, especially in the case of a patient who has already undergone a surgical procedure involving a sternotomy.

The synergy system provides circulatory support with flows ranging from 1.5 to 4.25 l/min. For patients treated with this device for left heart failure, haemodynamic measurements have shown that, in addition to increasing the cardiac index by an average of ~1 l/min/kg, this approach also includes marked reductions in pulmonary wedge pressure, pulmonary artery pressures and PVR. As summarized, these effects were achieved in the present patient, thus allowing him to be listed for heart transplant. Overall, these effects were sufficient to improve exercise tolerance and allowed the patient to return to work. This new VAD technology may provide a viable option for treating systemic heart failure in patients with ccTGA.

**Conflicts of interest:** Daniel Burkhoff is medical director of CircuLite®. Bart Meyns is member of the scientific advisory board (SAB) of CircuLite®.

**REFERENCES**


