

Peer-reviewed Case Report

Seeing into the Future: HeartMate 3 to the Systemic Right Ventricle in a Completely Blind Patient with Congenitally-corrected Transposition of the Great Arteries

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Juhi Ramchandani,* Chirag Dipak Shah, Kashif Saleem, Maya Guglin, Roopa Rao

Indiana University School of Medicine, Indianapolis, IN

*Corresponding author: jramc001@gmail.com

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Abstract

A 24-year-old, blind male with congenitally-corrected transposition of the great arteries and systemic right ventricular failure presented with New York Heart Association class IV heart failure despite home inotropic therapy. He was not a transplant candidate due to psychosocial issues. He underwent a successful HeartMate 3 (Abbott Laboratories) placement with the implementation of techniques to guide home ventricular assist device care despite blindness.

Keywords: transposition of the great arteries, congenital heart defect, chronic heart failure, cardiac assist devices



Introduction

Congenitally-corrected transposition of the great arteries (ccTGA) is a rare congenital cardiac anomaly comprised of atrioventricular and ventriculoatrial defects. It makes up about 0.5% of congenital heart disease cases.¹ Unfortunately, up to 2/3 of patients with this anomaly will have clinical signs of heart failure by age 45, with some developing symptoms as early as the first decade of life.^{1,2} Newer ventricular assist devices (VADs) like the HeartMate 3 (HM3, Abbott Laboratories) show promising results in patients with advanced heart failure, with lower stroke rates, hospitalizations, device malfunction, and mortality than older devices such as the Heartmate II (HMII, Abbott Laboratories).^{3,4} Although the HM3 has been implanted in some adults, experience is still limited in patients with ccTGA.⁵⁻¹¹ Moreover, VAD care poses several challenges, especially for patients with visual impairment. There is a paucity of literature on VADs for blind patients with advanced heart failure.¹²⁻¹⁵ We present a unique case of HM3 implantation in a completely blind patient with ccTGA.

Case Report

A 24-year-old male with a history of depression and anxiety was diagnosed with ccTGA with an Ebstein anomaly at age 17 before engaging in sports. He was otherwise healthy and initially asymptomatic but had subsequent hospitalizations for recurrent supraventricular tachycardia after ablation, automatic implantable cardioverter defibrillator placement, syncope, and heart failure exacerbations from systemic right ventricular (RV) failure.

Unfortunately, five years after the diagnosis, he suffered a self-inflicted gunshot wound to his head in a suicide attempt, which rendered him completely blind. He was considered for advanced heart failure therapies, including transplantation and VAD, but initially did not qualify due to his psychosocial issues. His maintenance medications included metoprolol, sacubitril-valsartan, and furosemide; home milrinone therapy was initiated. We briefly considered tricuspid valve replacement and cardiac resynchronization therapy defibrillator placement. However, these procedures were not pursued because of concern for limited long-term benefits.

He presented with worsening functional status and heart failure symptoms; he was classified as New York Heart Association (NYHA) class IV status and Interagency Registry for Mechanically Assisted Circulatory Support level 2. In the interim, he significantly improved his psychological health, so VAD support was considered as a bridge to potential transplantation. His body mass index was 25.3 kg/m² (weight 84.6 kg, height 182.9 cm).

A transthoracic echocardiogram one week before VAD implantation showed a severely dilated systemic RV, severely reduced ejection fraction (< 20%), and moderate-to-severe tricuspid regurgitation (TR). Right heart catheterization before milrinone therapy showed elevated right-sided pressures (mean right atrial pressure = 8 mm Hg, RV = 25/5 mm Hg, pulmonary artery pressure = 24/14 [20] mm Hg, pulmonary capillary wedge pressure = 13 mm Hg, V wave = 14), a pulmonary artery pulsatility index of 1.25, an aortic pulsatility index of 2.5 (using non-



invasive blood pressure 103/70), a Fick cardiac output of 3.77 L/min, and a cardiac index of 1.9 L/min/m².

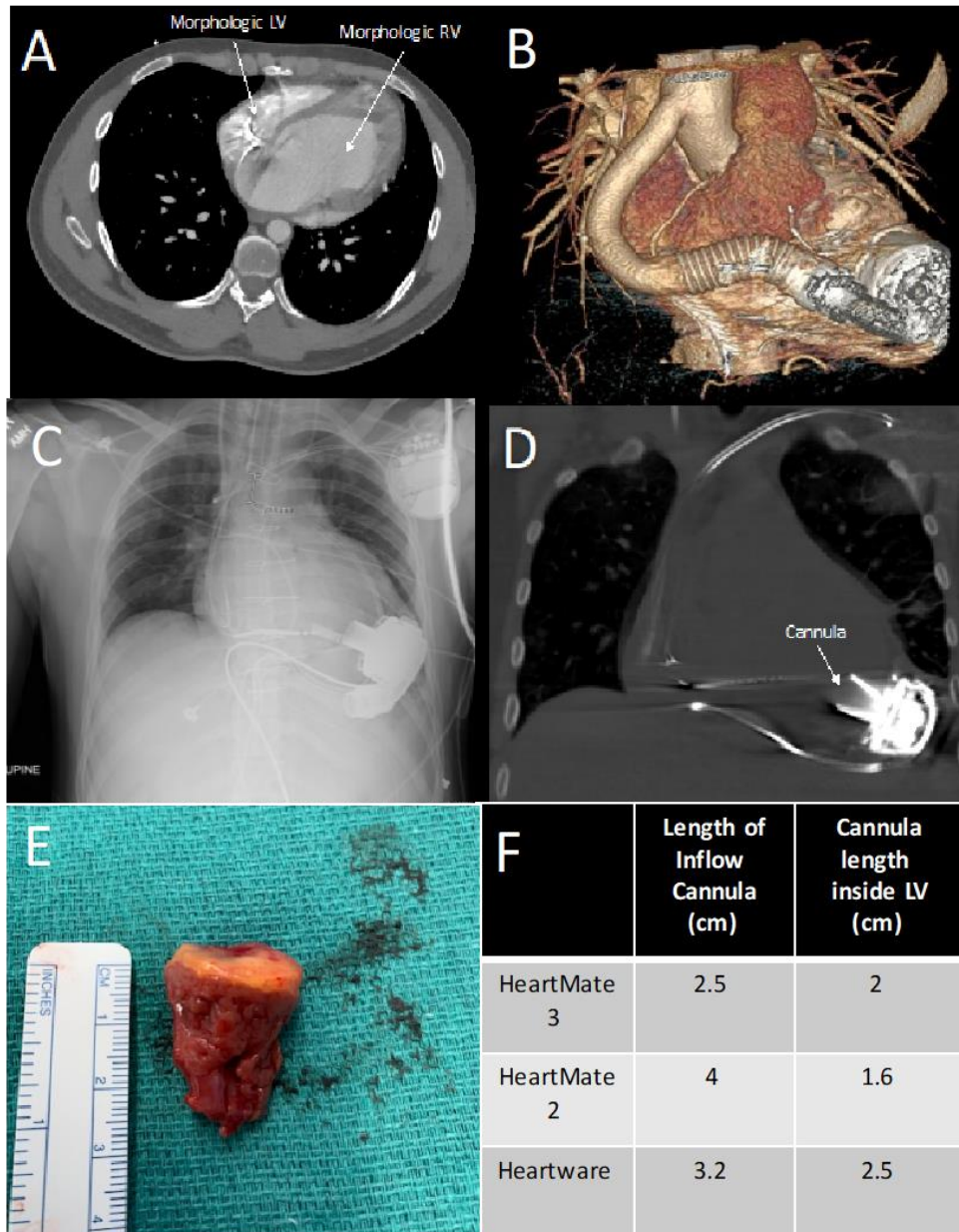


Figure 1. Orientation of the morphologic right ventricle (RV) and left ventricle (LV). Images of (A) the ventricular outflow track and (B) 3-D reconstruction, with (C) additional views of the cannula on chest X-ray and (D) coronal computed tomography. (E) 2.5 cm of the RV apical core was taken out to allow optimal positioning. (F) The table compares cannula lengths for different types of ventricular assist devices.



The patient's blindness was a major limiting factor to receiving a VAD. However, he could independently perform his daily activities, communicate effectively, and navigate tasks with minimal difficulty. His caregivers agreed to 24-hour supervision. After ensuring the patient and his caregivers understood the post-procedure requirements, we successfully implanted a HM3 device to the systemic RV (Figure 1, A-D). His RV had extensive trabeculations, which were resected as much as possible. The apical core measured 2.5 cm at the maximum length (Figure 1E). We used a transesophageal echocardiogram to guide placement of the inflow cannula. Despite severe TR, we decided not to intervene because of the complexity of the repair.

The procedure was complicated by ventricular fibrillation in the operating room, requiring multiple defibrillations and an amiodarone drip. The patient suffered from postoperative cardiogenic shock and needed inotrope and vasopressor support. Initially, he was managed at a lower speed (5200 rpm) to avoid suction events. However, he developed significant pulmonary edema complicated by respiratory failure and needed mechanical ventilatory support, probably because of his significant TR. In addition to diuresis, the VAD speed was gradually increased to 5700 rpm to allow for optimal unloading as the TR and pulmonary edema improved. We also treated him for potential superimposed bacterial pneumonia. He remained in the cardiac intensive care unit for 15 days before being transferred back to the cardiology service. He was eventually discharged home on postoperative day 28 in improved condition on room air. The VAD settings at the time of discharge were: speed = 5700 rpm; flow = 4.9 L/min; pulsatility = 3.1; and power = 4.3 W.

Additional accommodations were made to ensure success after discharge, including training sessions before and after VAD placement, the use of different velcro surfaces, maintenance of device orientation, and the use of phone applications. With these tools, our patient is doing well 2.5 years after the procedure. He can independently change the battery, connect the cords, and troubleshoot VAD alarms with ease. He is currently classified as NYHA class I.

Discussion

This case is unique in many ways. VAD placements in systemic RVs for adult patients with ccTGA typically involve Heartware Ventricular Assist Device systems (HVAD, Medtronic), HMII, and the Thoratec® VAD System (Abbott Laboratories).¹⁻⁴ Very few reports mention implantation of HM3 in adults with ccTGA.^{12,13,16,17} Two reports mention VAD placement in legally blind patients, one with an HMII in a patient with severe diabetic retinopathy and another with an HVAD in a patient with amaurosis after retinal detachment repair.^{14,15} Our case is the first to report implantation of a HM3 in a completely blind patient with ccTGA.

The challenge for VAD placement in this congenital anomaly is the presence of extensive trabeculations and moderator band, which can obstruct the cannula. The cannula length of each device is also variable (Figure 1F).^{16,17} Thrombosis is a major concern with the HMII and HVAD devices. Thus, we selected the HM3, which has greatly overcome this issue with a < 1% reported incidence of device thrombosis.¹⁸



Several techniques were implemented to overcome limitations associated with blindness. The patient and his family were trained before VAD implantation to ensure device familiarity. We ensured that the patient received caregiver supervision 24 hours a day in the postoperative period. Non-visual cues promoted VAD care at home, including the use of velcro surfaces (soft and rough sides) to distinguish between different attachments. The white cord and its attachment to the mobile power unit were identified using the soft side (loop) of the velcro (Figure 2A). The rough side (hook) of the velcro was attached to the driveline connection on the backup controller (Figure 2B), so that the patient could change controllers if needed. Tactile arrows on the battery clip and charger enabled him to correctly connect the batteries (Figure 2C). The patient was educated to orient the controller such that the cords and driveline

wires always exited to the patient's left, so that the battery button was on the top left. A free application called "Be My Eyes" facilitated visual cues to optimize VAD care. This application connects blind individuals with volunteers using live video calls on their cellphone. Our patient uses it to visualize and troubleshoot VAD alarms (Figure 2D).

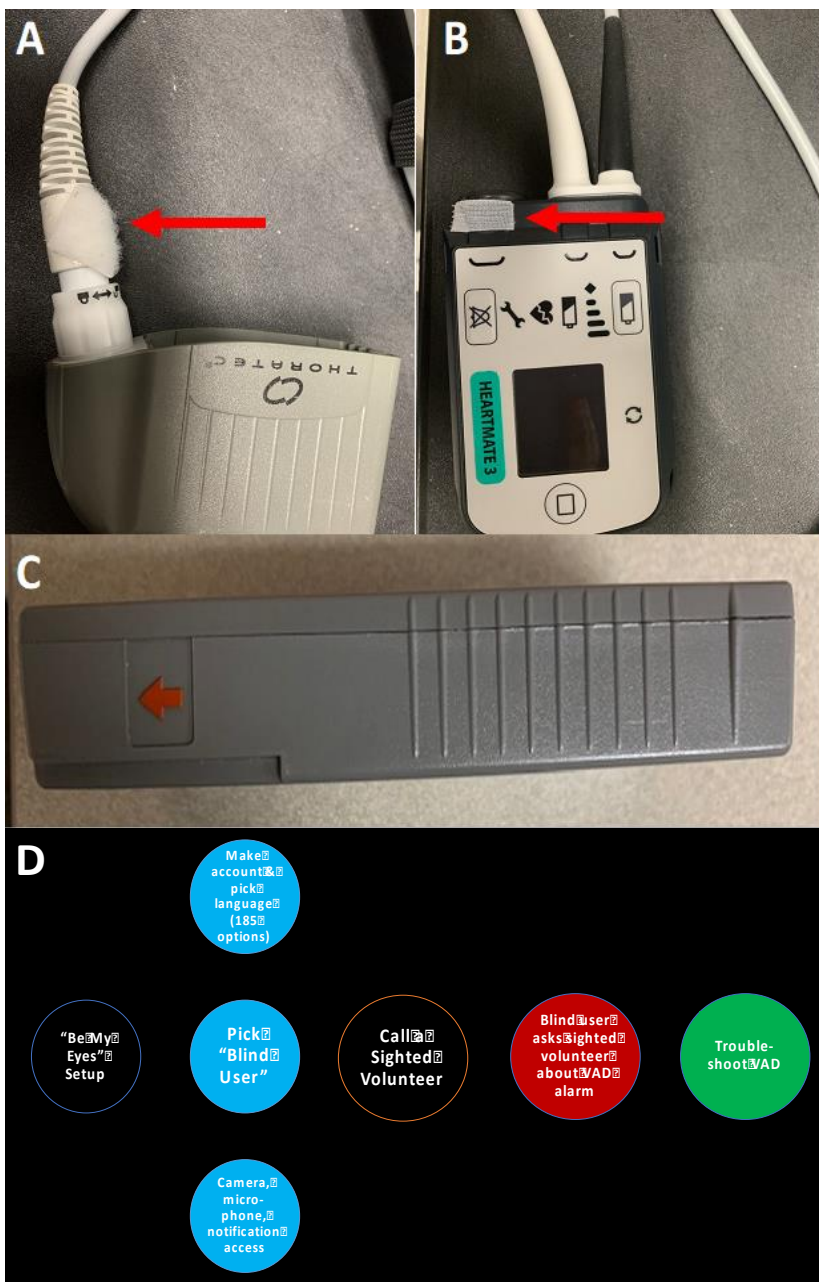


Figure 2. Device modifications and an application to overcome limitations associated with blindness. (A) The velcro loop side identifies the white cord, (B) the hook side identifies the driveline connection of the backup controller, and (C) tactile arrows assist when the battery is exchanged (D) Illustration of the setup for "Be My Eyes," an application that our patient uses to "Call a Sighted Volunteer" for ventricular assist device troubleshooting.

Conclusion



Literature on the use of the HM3 in adult patients with ccTGA is limited.^{14,17} This is the first case of HM3 implantation in a completely blind, adult patient with ccTGA. Further studies are needed to explore the application of HM3 and other novel techniques to ensure VADs as a viable therapeutic option for blind patients.

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