

Case Report

Orthotopic Heart Transplant in a Child with Nonverbal Autism

Bailey DB^{1*}, Schneider LM¹, Maeda K², Hollander SA², Shaw RJ¹, Rosenthal D² and Froehlich-Santino W¹

¹Department of Child and Adolescent Psychiatry, Stanford University, USA

²Department of Pediatric Cardiology Lucile Packard Children's Hospital, Stanford University, USA

*Corresponding author: Bailey DB, Department of Child and Adolescent Psychiatry, Stanford University, USA

Received: January 13, 2016; Accepted: March 22, 2016; Published: March 29, 2016

Abstract

Improved outcomes for heart transplantation for end-stage heart failure in pediatric patients, and a critical shortage of donor organs, makes it imperative to reserve heart transplantation for those who are most disabled by heart failure and most likely to derive the maximum benefit from transplantation. Careful consideration is required in the selection and training of candidates for the rigorous care required after transplantation. In addition to medical appropriateness, factors such as psychosocial stability and the presence of other severe chronic illness that could interfere with treatment adherence are considered in evaluating a candidate's suitability for transplant. This report illustrates the case of a child with nonverbal autism and dilated cardiomyopathy who received a successful orthotopic heart transplant following prolonged support with a Left Ventricular Assist Device (LVAD). The Success of the transplant and bridging medical interventions was supported by family involvement and clinical expertise of multiple specialties. This case provides evidence that children with Autism Spectrum Disorders (ASDs) should be considered for cardiac or other solid organ transplants. Although children with ASDs present unique challenges for medical interventions, like all patients, children with ASDs should be evaluated on a case-by-case basis. The presence of ASD (including nonverbal ASD) should not be an absolute contraindication for transplant.

Keywords: Autism; Heart; Transplant; Cardiac; Pediatric

Abbreviations

ASDs-Autism Spectrum Disorders; LVAD-Left Ventricular Assist Device; P-TRI-Pediatric Transplant Rating System

Introduction

According to recent registry reports from the International Society for Heart and Lung Transplantation, approximately 400 pediatric heart transplantation procedures are performed worldwide each year. The most common indication for heart transplantation in older children and adolescents is cardiomyopathy [1]. Autism is a neurodevelopmental disorder caused by abnormal brain development, manifesting in persistent deficits in social communication and restricted, repetitive patterns of behavior, interests or activities. ASDs tend to occur more often in people with certain genetic conditions, which may lead to greater risk for congenital conditions including cardiomyopathy [2]. Management of children with severe autism in the hospital setting and post-transplant can be challenging. Direct communication between the patient and team may be limited due to deficits in communication or lack of social motivation. Routines may be difficult to establish in the setting of emergent medical needs. Adverse responses to sensory stimuli may create significant challenges to utilizing medical devices. Furthermore, sensory seeking behaviors may further threaten the maintenance of such interventions. Individuals with ASDs may require greater assistance with daily activities, both during hospitalization and post-transplant.

Case Presentation

A nine-year-old male with autism (weight: 47kg, height: 148cm,

and BSA: 1.39 m²) presented to an outside hospital with a two week history of malaise and shortness of breath. An echocardiogram confirmed dilated cardiomyopathy with severely depressed ventricular function. Despite medical management, the patient progressed to cardiogenic shock, was transferred to an out-of-state facility capable of offering cardiac transplantation, and was immediately placed on ventricular assist extracorporeal membrane oxygenation. Given the patient's poor prognosis for recovery, it was decided that heart transplantation was the most appropriate treatment.

During pre-transplant evaluation, appropriate concerns arose regarding biopsychosocial factors, from both the medical team and family. The patient possessed no spoken language, though he had recently developed the ability to communicate short sentences through the use of a keyboard and demonstrated the ability to understand concepts appropriate to his age. He experienced particular difficulty with motor control, requiring assistance for many daily activities. Additionally, he possessed significant sensory sensitivities and had poor reactions to restrictive environments. He had in the past struggled with impulsivity including development. With these challenges, the team and family faced was maintaining an LVAD in the setting of a cardiovascular intensive care unit where distressing stimuli and a restricted environment were essential. Balancing these concerns, the patient had a supportive family who was involved in the medical management, committed to working with a large team of caregivers, and communicated effectively with providers. After presentation to the multidisciplinary selection committee, the patient was accepted for heart transplantation and underwent implantation of a Heartmate[®] II LVAD as a bridge to transplantation.

- 1.) Rooms
 - a. Private room if possible
 - b. Visible clock
 - c. Visual red disk timer (e.g. Time Timer©) in room
 - d. Schedule Board with Names of staff and Check Boxes for Daily Routine/Schedule
- 2.) Consistency
 - a. Assigned primary nurse/Minimize staff changes
 - b. Maintain as consistent of a routine as possible
- 3.) Sensory
 - a. Quiet sign on door
 - b. Minimize vital signs and monitoring. Schedule vitals and interventions at consistent times
 - c. Decrease monitor volumes
- 4.) Communication
 - a. Always introduce yourself and tell the child prior to doing anything
 - b. Use simple, concrete language
 - c. Try your best to make sure the child is listening to you
 - d. Use visual aids such as timers and pictures
- 5.) Disruptive Behavior/Aggression
 - a. Involve child psychiatry
 - b. Minimize stimuli (people, lights, monitors, noise)
 - c. Elbow immobilizers instead of restraints if biting or hitting self

Figure 1: In-Patient Hospital Protocol for Children with ASDs (Froehlich, Hoffman, and Lotspeich, unpublished).

Given the sensitive LVAD equipment, one critical step was to establish the safety and physical supervision. The family played an essential role in educating the medical staff about optimal ways to support the patient. Although a parent was always at the bedside, this patient was also provided with one-to-one aides to assist with mobility and transfer, and to provide respite as needed. Due to difficulties the patient experienced with transitions between unfamiliar staff members, the pool of the hospital staff was limited to provide greater consistency.

Social work was vital in providing support for the family. The child psychiatry team, aided with pretransplant psychological evaluation and provided support throughout the hospitalization. Individual therapy was conducted via typing and secure e-mails with the therapist in the room. This unique intervention helped the patient to communicate with the medical team, practice skills, and manage anxiety around procedures and transitions. Through typing, he was able to express his thoughts including fears about death, and to explore these feelings with his psychiatrist. Individualized interventions such as leaving the room door closed, scheduling quiet times, and allowing the patient to sit outside clinic rather than in busy waiting rooms were useful in minimizing stress reactions. The hospital, school, occupational therapy, physical therapy, and child life teams ensured continued education and maintenance of a regular schedule as was possible.

After eight months of hospitalization and maintenance with LVAD pump, the patient underwent orthotopic heart transplantation. The heart was implanted with a standard bicavalanastomosis. The patient completed outpatient management with limited complications, no significant rejection and was cleared for return home one year after the first presentation. Adherence with immune suppressant drug therapies has been excellent, with levels being at or near goal throughout the post-transplant course. This successful outcome can be credited to in part to an adaptive, and knowledgeable family that allowed for effective coordination between patient, family and specialty teams.

Discussion

Recent advances have led to the prolongation and quality of life

for pediatric patients after heart transplant. However, the process including preoperative, perioperative, and long-term postoperative stages, can present major psychosocial obstacles for families [3]. For children with ASDs, the process of transplantation and aftercare presents additional challenges. Success in the transplant treatment process requires resilience and ability to tolerate daily physically invasive procedures, unpredictable changes in routine, as well as the patient and family's ability to adhere to treatment regimens. Due to the limited availability of donor organs, transplant centers are faced with the challenging task of deciding which patients will benefit most from receiving a donor organ. Selection committees incorporate a range of biopsychosocial factors into their candidacy decisions. Teams utilize instruments such as the Pediatric Transplant Rating Instrument (P-TRI) [4]. The P-TRI identifies areas of psychosocial vulnerability or strength that help determine ability to complete the transplant process and maintain treatment adherence. Among the criteria evaluated in the P-TRI are factors related to illness such as knowledge about and motivation for transplant, treatment adherence, psychiatric history, family environment and psychosocial support. For children with ASDs, it is important to consider how social-communication and other challenges may affect these areas, and what mechanisms can be employed to overcome barriers. Children with ASDs may require special accommodations and multi-disciplinary care during hospitalizations. Hospitals may establish protocols for managing children admitted with ASDs. In the case described, a general in-patient protocol for children with ASDs was employed (Figure 1). It is a vital part of the transplant process to involve a multidisciplinary team including physicians, nurses, mental health professionals, social workers and rehabilitation specialists. Care conferences involving all disciplines should be held regularly and include the patient and family to discuss treatment plans. Teams and caregivers must remember the physical and psychological toll prolonged hospitalization takes on patients and families. Reciprocally, families must be willing to accept imperfections in systems of care. Flexibility and willingness to employ innovative measures tailored to the individual needs of each child are imperative. Although ASDs may present challenges to transplant, it is important to note that autism may uniquely prepare patients and families for the challenges involved in the organ transplant process. Families often already have

experience managing complex systems of care and time intensive treatment practices. They may already maintain strict healthcare routines and have an established support network to assist with care needs, and are often well-versed in daily sacrifice to accommodate the special needs of their children. In the evaluation of candidates for heart transplant, medical urgency and ability to maintain transplant care are important factors. While ASDs and other developmental delays may pose challenges in medical management, these disabilities should not be considered absolute contraindications to transplant. Many of the possible associated challenges may be overcome with strong support, including close family involvement and a dedicated interdisciplinary transplant team.

References

1. Benden C, Edwards LB, Kucheryavaya AY, Christie JD, Dipchand AI, Dobbels F, et al. The registry of the international society for heart and lung transplantation: sixteenth official pediatric lung and heart-lung transplantation report-2013. *J Heart Lung Transplant*. 2013; 32: 989-997.
2. Prevalence of Autism Spectrum Disorders - Autism and Developmental Disabilities Monitoring Network, 14 Sites, United States, 2010. *Surveillance Summaries, Morbidity and Mortality Weekly Report*; March 30, 2012; 1-19, CDC.
3. Gold, Kirkpatrick, Fricker, Zitelli. Psychosocial issues in Pediatric Organ Transplantation: The Parent's Perspective. *Pediatrics*. May 1. 1996; 77: 738-744.
4. Fung E, Shaw RJ. *Pediatric Transplant*. *Pediatr Transplant*. 2008; 12: 57-66.