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Review

The pediatric experience of living with a ventricular assist device

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ABSTRACT

With the increasing use of long-term mechanical circulatory support in the management of pediatric heart failure, there is a growing patient population with unique qualities and needs. The objective of this review is to better understand the experience of having a ventricular assist device from the perspective of the child. The literature findings were categorized into the following themes: the persistent threat of mortality, identification with the device, the perception of life pre-device, and caregiver strain. While there is a large area that remains to be studied in future research, these findings can be utilized in the development of education and support programs to effectively meet the needs of this unique population.

1. Introduction

Since the beginning of recorded human history our understanding of the heart has transcended its physiologic role. From the ancient Egyptian belief that the heart housed the eternal essence of an individual, to the modern practice of inscribing loved ones' initials inside the heart symbol, the heart carries tremendous metaphorical qualities [1]. We can literally feel a wide range of emotions that seem to emanate from the center of our chest and we notice the heart's automatic response to our joys and fears; given this intimate connection it is not surprising that the heart is so intricately linked with our humanity.

At the same time, we cannot escape that the heart is synonymous with and vital for life – indeed in most instances the presence of a heartbeat is the *definition* of life. A failing heart portends imminent death; as such a great deal of cardiovascular research over the previous decades has focused on supporting and optimizing cardiac function. At the most extreme, this includes the development of ventricular assist devices (VAD)—machines implanted in the body to augment or replace the intrinsic function of the heart. These devices are now routinely used in adults, and their use is becoming increasingly common in the pediatric population. Cardiovascular practitioners rightfully focus on how these devices function physiologically. However, just as we have a long heritage of appreciating the heart for more than its mechanical role, it is important to realize and appreciate the impact that placing a machine within the body to supplant this role has on an individual, especially when that individual is a child at variable stages within the developmental process.

2. Background

Pediatric heart failure (HF), whether secondary to congenital heart disease, myocarditis, or cardiomyopathy, is estimated to account for more than 20,000 hospitalizations each year in the United States alone [2]. While these numbers pale in comparison to the adult HF population, mortality in pediatric HF is disproportionately high. It is estimated that 46% of children diagnosed with HF will either undergo transplantation or die within five years of diagnosis [3].

The initial treatment for pediatric HF is aggressive medical management, with progression to heart transplantation in patients who are refractory to standard therapy. Yet many children cannot maintain adequate cardiovascular performance while awaiting transplant, and require discussion of mechanical circulatory support as a bridge to transplantation [4]. Prior to the early 2000s, this support was limited to extracorporeal membrane oxygenation (ECMO), which can provide only short-term support [5]. Due to the limited availability of donor hearts and the high morbidity associated with ECMO, the duration of support that ECMO can provide frequently falls severely short of the average time a patient spends on the transplant waitlist.

While long term mechanical circulatory support has been a growing tenet of HF treatment in the adult population for decades, size limitations and disproportionately large stroke volumes have precluded the pediatric HF population from benefiting from many of these devices [4–6]. For this reason, the development of the Berlin Heart EXCOR was a major contribution to pediatric medicine, as the first long term device with multiple pump and cannula size options. In 2000, this pneumatically driven, pulsatile-flow VAD was approved for use in Europe, and by

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2004 it was being used widely across the United States [6]. In 2011, it officially gained FDA approval, becoming the first FDA approved long-term device for children [7].

While the Berlin Heart notably increased the potential for longer durations of support, and thus, rate of survival to transplantation, the incidence of neurologic events has not been any lower than that of ECMO [3,8]. Additionally, due to the large size of the Berlin Heart's external drive unit, patients remain confined to the inpatient setting and restricted to minimal activity [9]. These drawbacks have propelled the industry toward continuous-flow devices [10,11]. While these devices have revolutionized HF management in the adult population, size limitations continue to impede progress in the development of continuous-flow VADs in the pediatric population.

As there is not yet a continuous-flow device approved by the FDA for use in pediatrics, the FDA has permitted the off-label use of adult devices in children and adolescents who meet specific size and weight restrictions [10,11]. Due to the lower morbidity associated with continuous-flow devices, as well as the potential for hospital discharge after device implantation, these devices have been implanted in pediatric HF patients with increasing frequency over the past decade [7,10,11].

With this progress, approximately 20% of pediatric patients who now undergo heart transplants are supported with a VAD prior to transplantation [12]. Yet support options still remain severely limited for infants and smaller children. In the United States, this limitation has led to the development of the Pumps for Kids, Infants, and Neonates (PumpKIN) trial funded by the National Institute of Health. The goal of this project is to promote the development of pediatric-specific continuous-flow devices. The first randomized trial, comparing the Berlin Heart to a new pediatric-sized continuous-flow device (Jarvik Infant 2015), has already begun enrollment [4,11]. With funds and resources dedicated to the development of pediatric-specific devices, the prognosis of pediatric heart failure could be very different in the upcoming years.

The number of pediatric patients supported with VADs will continue to increase, not only due to continued technological developments, but also as data continues to suggest that early timing of device implantation is associated with more favorable outcomes after transplant [4,5]. Likewise, the indication for device placement has broadened in recent years, with some centers utilizing VADs as support for myocardial recovery, prevention of secondary organ failure while determining transplant candidacy, and in some cases, as destination or palliative therapy [4,5,11,13–15]. Overall, the increased utilization and duration of VAD therapy will translate to more pediatric patients living with VADs in both the outpatient and inpatient settings [9,11,14].

As research and experience continue to propel our ability to utilize technology to improve outcomes for children with HF, it is important to remember that just as the heart itself has always held special meaning above and beyond its physiologic function, the act of mechanically supporting the heart has important psychological and emotional manifestations that must be considered. To adequately prepare for this growing patient population and develop centers prepared to meet the educational and psychosocial needs of these patients, it is necessary to have a deeper understanding of the experience of living with a VAD from the perspective of both the child and family. The aim of this review is to evaluate the current literature on the experience of pediatric VADs, identify the prevailing themes, and highlight the gaps in the literature that implore further research.

3. Current Body of Literature

Data on patient outcomes, adverse events, and quality of life in the adult HF population is relatively abundant in the literature, but the unique features and disease processes of pediatric HF make it difficult to extrapolate the findings to the pediatric population [11,13,14]. While individual pediatric centers have collected data on cohorts of patients with VADs since the early 2000s, a systematic database was not in existence for pediatric devices until the development of the PediMACS registry in 2011 [7,14]. While the PediMACS registry only collects data on patients in the United States, it is the largest database of its kind for this patient population. Since then, the data on patient outcomes, adverse events, and survival rates has grown substantially. However, there is still a paucity of literature evaluating measurements of quality of life, psychological health, and the impact on child development [11,16,17]. Many factors, such as pre-device health status, development stage, and whether the child is being managed as an inpatient or outpatient greatly influence the experience of living with a VAD. Yet within the few studies that have been conducted in the last several years, there are four prevailing themes about the pediatric experience that can be extracted and synthesized. We have categorized them as follows: *the persistent threat of mortality, identification with the device, the perception of life pre-device,* and *caregiver strain.*

4. The Persistent Threat of Mortality

The heart intermittently makes its presence known to most of us by racing in times of stress or pounding following intense exercise, but the constant presence of a VAD serves as incessant *memento mori* following implantation [1]. This is quite justified; although long-term VADs have significantly improved the survival rate in pediatric heart failure, from 55% with ECMO to 80% with continuous-flow devices, they are not without serious risks [5,7,8,12]. Over half of the pediatric patients with VADS in the US between 2012 and 2014 had at least one adverse event prior to device explantation or heart transplantation. What is more unsettling is that 28% of all the patients studied in the PediMACS registry incurred three or more adverse events [7]. The most common adverse event is infection, followed by device malfunction, bleeding, and neurologic insult by means of thromboembolism or hemorrhagic stroke [5,7,12,14]. While not all adverse events are equally detrimental or even fatal, these events have the potential to reduce a patient's quality of life and transplant candidacy.

Older children and adolescents become especially cognizant of these risks and the dire stakes at hand [15,17]. Not coincidentally, this older age group is also at higher risk of developing psychiatric symptoms, such as anxiety and depression [16]. A qualitative study of school-aged children and adolescents in Melbourne, Australia, who had been discharged home with long-term devices reported universal themes of fear, uncertainty, and a sense of doom [17]. Despite being trained on the steps to take in various emergency situations, the participants described feeling frightened of their device. They had a clear understanding of the grave implications of a device malfunction, and expressed a constant awareness that the device was keeping them alive. They also conveyed a constant state of uncertainty, not knowing if and when a donor heart would become available—or if they would even survive to transplantation.

These responses were strikingly similar to the findings of a phenomenological study from the University of Alberta, which reiterated the sense of doom and constant awareness of one's own mortality [15]. Despite attempts to continue social activities, preserve peer relationships, and maintain a sense of normalcy, the looming threats associated with the device remain at the forefront of patients' minds. The physical presence of the device only exacerbates the constant worrying. Unlike an implanted pacemaker, patients have to carry the external portion of their device with them wherever they go [15]. The short driveline, which may pull even while sitting in place, serves as a reminder of the continuous threats they face. Thus, it is often impossible for these children and adolescents to momentarily ‘forget’ about their device and relish the carefree moments of childhood.

5. Identification with the Device

As the heart is so synonymous with life, the implantation of a