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Review

Novel strategies for supporting challenging populations: Inpatient infant, developing toddler, successful school-age, & the autonomous adolescent

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ABSTRACT

Pediatric patients supported by ventricular assist devices form a growing and challenging population. They are high-risk, and heterogeneous (in terms of age, neurodevelopment stage, and underlying anatomy) and relatively low-volume. Standardization of care across the various ages and stages is difficult. Multidisciplinary teams managing these patients must have a good understanding of growth and development to provide optimal care and creative options for growth, development, and adherence in order to achieve improved outcomes for pediatric ventricular assist device patients.

1. Introduction

Pediatric ventricular assist device use is growing rapidly around the world, and with that comes many opportunities and challenges. According to Pedimacs, a national registry of pediatric ventricular assist device data, 37 hospitals across America utilize the database, and 241 patients were enrolled from September 2012 to December 2016 [1]. Caring for pediatric ventricular assist device patients is complicated in that there are many types of devices available to use in a very diverse population of patients ranging from infant through adolescent. As the use of ventricular assist devices becomes more commonplace, it is important to know how to manage these patients of all ages, stages, and anatomies.

2. Inpatient Infant

Infants have a high incidence of both congenital heart disease and cardiomyopathies. Congenital heart disease, the most common birth defect, affects nearly 1% of births, an incidence of around 40,000 annually in the US (of whom, 15,000 will require surgery early in life) [2]. Cardiomyopathies are less common, but infants less than 12 months old form the majority of 1 in 100,000 children in the U.S. [2]. Although the incidence of heart failure is high among infants, mechanical support is problematic. Infants carry the highest mortality rate of these mechanically supported patients [3]. Ventricular assist device options for the infant population include the widely-used FDA-approved pulsatile Berlin Heart EXCOR (Berlin Heart Inc.; The Woodlands, TX) for children greater than three kilograms. Additional options are continuous flow centrifugal pumps such as Pedimag (Abbott Vascular; Santa Clara, CA) or Rotaflow (MAQUET Cardiovascular; Wayne, NJ.). All of the currently available devices for infants require inpatient hospital care until the device is decommissioned due to patient transplant, recovery, or withdrawal. Appropriate selection of a device is critical to achieving positive outcomes. Points to consider in selecting the optimal device include: the etiology of heart failure, the anticipated duration of support, and likelihood of recovery, type of support (left ventricular, right ventricular, or biventricular support), and device availability. Device availability may be a particular challenge in pediatrics (especially free-standing children's hospitals), where cost consideration and the relative infrequent usage, often results in limited availability of highly specialized devices, including the total artificial heart and percutaneous mechanical support devices.

A thorough understanding of ventricular assist device biomechanics is a prerequisite to good outcomes. For example, understanding the prevalence and etiology of adverse events, such as stroke, allows the medical team to take a proactive and standardized approach to prevention of such events. Although available options for children are still limited, new devices are on the horizon, such as the Infant Jarvik 2015. This device, part of the PumpKIN (Pumps for Kids, Infants and Neonates) Trial, is the first continuous flow intra-corporeal ventricular assist device designed for children under 20 kg. Initially designed for support down to approximately three kilograms, design changes required to minimize *in vitro* hemolysis have increased the lower end of support in the trial to eight kilograms. This continues to leave smaller

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children without similar durable, dischargeable options, although whether the Infant Jarvik 2015 will be capable of supporting smaller children remains unclear.

The infant on a ventricular assist device faces many challenges. The primary objective of mechanical support remains maintenance of good end organ perfusion while minimizing the risk of adverse events associated with anticoagulation and general use of these devices, such as a hemorrhagic or ischemic stroke. Additional important considerations include supporting normal growth and development for the infant on a ventricular assist device. Growth requires adequate nutrition, which can be challenging in infants with both a history of poor systemic perfusion prior to device implantation and often persistent elevations of central venous pressure even with device support. Nutrition should be started slowly and with careful monitoring for signs of feeding intolerance. Appropriate calorie intake enables growth but is also important for wound healing following surgical procedures. Many will need an alternative feeding method, such as a nasogastric or nasojejunal, to maintain nutrition goals.

Developmental milestones to focus on for a hospitalized infant on a device include: smiling, looking at people, cooing, and turning head toward sounds. In later infancy: different types of crying, babbling, copying facial expressions, reaching for toys, rolling over and sitting up are expected [4]. During these times of hospitalization, allowing time to let families be close to their infant will support family bonding and is essential to the infant's emotional growth. As these milestones are best achieved through rehabilitation, it is important to have a routine schedule for physical, occupational, and speech therapy, to promote development and overall strength. Rehabilitation success can be slow, and safety of the baby and the device may seemingly counteract efforts for usual daily activities. In most cases, following the immediate surgical period, the cannulas are secure enough to eliminate the need for many activity limitations. It is important for a key member of the patient's team to know and communicate the surgical configuration of the cannulas in order to create a safe plan of care for rehabilitation.

Once it is known that the patient is stable enough to undergo rehabilitation there are several key actions that can be incorporated into the patient's care. Such actions include: placing the baby in age appropriate seats and chairs such as car seats or bumbo seats to facilitate the patient's ability to sit instead of lying in the hospital bed. Floor mats can be used to provide tummy and floor time. These actions help the infant work on head, neck, and core strength, and can be achieved with proper pump positioning and adequate supervision by appropriately trained staff. Encouraging parents to hold their child frequently, or take their child out for walks in a stroller or wagon will help with both the infant's development and infant-parent bonding. Routine ventricular assist device education and standard management protocols accessible to the multidisciplinary team, including the family, elevates the safe handling of patients on devices. Education for the inpatient infant on a device includes infection prevention and safe handling of device while allowing patient to have normal growth and development experiences, as mentioned. Gaining support for inventive and safe ways to handle the device, which will increase mobility of the infant, takes some initiative that will reap many rewards for the infant's development and outcome.

3. Developing Toddler

As children transition from infancy to toddlerhood the rapid acquisition of growth and motor development slows in favor of tremendous advances in intellection, social, and emotional functioning. Normal development for the toddler includes: walking, running, feeding self, saying single words and then sentences, and playing with others [4]. Additionally, nutrition can offer challenges for the toddler as they learn about many different food groups and have an inconsistent diet. Toddlers move around a lot, are more aware of themselves and their surroundings, and their desire to explore new objects and people increases. During this stage, toddlers will show greater independence, which is important when caring for toddlers who have a ventricular assist device.

In choosing mechanical support for toddlers, as in infants, the etiology of heart failure, anticipated duration of support, and type of support (left ventricular, right ventricular, or biventricular support) will influence the device choice. The Berlin Heart EXCOR, the Pedimag, or the Rotaflow each support the physiologic needs of a toddler. The older toddler has the potential for an internal continuous flow device such as the HeartWare HVAD ventricular assist device (Medtronic Cardiac Rhythm and Heart Failure Management; Mounds View, MN). Approved for patients with BSA of 1.5 m^2 or greater, this device has been used off-label in children as small as 13 kg as a bridge to transplant. However, fitting this device in a small child may require technical modifications to ensure proper cannula position and device function; thoracic cavity measurements may be necessary to ensure proper fit [3].

The Berlin Heart EXCOR is lightweight and has cannulas which exit transthoracically and offer more stability and opportunity for mobility and rehabilitation. Many children can easily walk about with this device. One of the biggest challenges includes making sure the cannulas and pneumatic air tubing stay untangled as the patient moves about. Clothing can help discourage the patient's curious hands from exploring the cannula and dressing sites and decrease the risk for infection. The Pedimag and Rotaflow pumps are heavier and need to be secured to equipment while the patient ambulates. Special caution needs to be taken when moving these patients to ensure there is never tension on the cannula. HeartWare HVAD equipment includes the controller and batteries that can be stored in the manufacturer patient packs, or the care team or family can pursue unique solutions for toddlers to carry this equipment. Specially made backpacks keep equipment secure and help these children develop their independence and balance, and improve the weight distribution of the equipment. HeartWare is an intrapericardial pump requiring an extracorporeal power source and pump controller, and this offers the ability to convalesce and rehab at home. To ensure the safest care for these toddlers, a unique plan assessing each child's development and capabilities needs to be evaluated. Training the family to care for their child out of the hospital can be worrisome in the beginning, but with continued support and education, a successful transition to home will occur.

4. Successful School-age

School-age child development, ages six to twelve, includes a lot of variability in growth and development [5]. Physical changes are apparent during this time, including onset of puberty, and with this comes more self-awareness. School-age children have smooth and strong motor skills but there remains much variability in their coordination, endurance, balance, and physical abilities. There can be large differences in height, weight and build among school-age children, and as early as the age of six, a sense of body image begins developing.

Factors that influence device choice include: patient size, anatomy, type of support (left ventricular, right ventricular or biventricular support), previous procedures, and expected duration of support. School-aged children have a weight range from 20 to 42 kg, with a BSA range from approximately 0.78 m^2 to 1.34 m^2, based on the interquartile range [6]. Device options for school-age children include temporary extracorporeal centrifugal continuous flow devices including the Pedimag and Centrimag, or the Rotaflow. Temporary extra-corporeal devices are only used in the inpatient setting either as a bridge-to-decision, recovery, to a more durable device, or cardiac transplantation. The para-corporeal pulsatile pump system, the Berlin Heart EXCOR can be used to support school-age children with pump sizes of 25 ml, 30 ml and 50 ml being most applicable to this age group. Both extra-corporeal and para-corporeal devices can be used as uni-ventricular or bi-ventricular support.