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Hypoplastic Left Heart Syndrome

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Hypoplastic Left Heart Syndrome (HLHS)

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Introduction

Congenital heart defects are the most common type of birth defects and can range from minor to severe. A severe and rare form of congenital heart disease is hypoplastic left heart syndrome (HLHS). HLHS consists of the underdevelopment of the left sided heart structures leading to the failure of the left ventricle to support the systemic circulation resulting in single ventricle physiology (Mussa & Barron, 2016). HLHS can be dangerous and life threatening if not treated within the first few days of life (Mussa & Barron, 2016). The Center of Disease Control and Prevention estimates that 1,025 babies are born each year in the United States with hypoplastic left heart syndrome (2019).

The topic of HLHS was chosen because healthcare providers need to have the knowledge of how to approach care for the HLHS patient population. As medical and surgical advancements continue an increased number of patients with HLHS are reaching adulthood. Therefore, to provide the best outcomes for patients with HLHS it is imperative that healthcare providers receive appropriate education on the epidemiology, pathophysiology, physiology, treatment plans, and the surgical and medical management of this complex patient population.

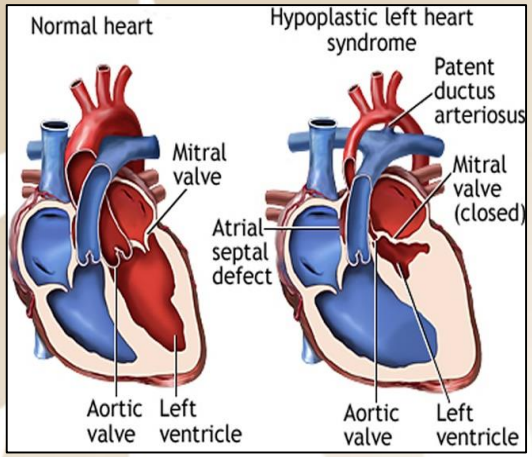

Figure 1. normal heart (left) and hypoplastic left heart (right). Retrieved from https ://healthjade.com/hypoplastic left heart syndrome/

Signs and Symptoms

Symptoms are present at birth or shortly after birth.

- Tachypnea
- Tachycardia
- Weak pulse
- Poor feeding
- Cyanosis
- Lethargy

(Mayo Clinic, 2018).

- Postnatally the newborn pulse oximetry screening can detect low oxygen levels in the blood, possibly indicating a sign of critical congenital heart disease (Centers of Disease Control and Prevention, 2019).

Pathophysiological Processes

Underlying Pathophysiology

- HLHS occurs during fetal cardiac development.
- HLHS has no known cause and is present at birth.
- No association with maternal age or parity (Mussa & Barron, 2016).
- There are associations with chromosomal abnormalities. (Mussa & Barron, 2016).
- "HLHS is slightly more common in males, but there is no ethnic or geographic association" (Benson et al., 2016, p.25).
- Evidence is suggesting a genetic component is likely but thought to be complex and multifactorial (Mussa & Barron, 2016).
- In HLHS "hypoplasia of the left ventricle (LV) and aorta are cardinal features, but the pathologic definition also includes atresia or stenosis of both the aortic and mitral valves" (Benson et al., 2016, p.25). Therefore, the left side of the heart can not support the systemic circulation (Mussa & Barron, 2016).
- For survival, the right ventricle must support the systemic circulation through the right to left flow of blood across a patent ductus arteriosus (PDA) (Mussa & Barron, 2016). Adequate mixing of oxygenated and deoxygenated blood at the atrial level through the atrial septal defect is also essential for survival (Figure 1) (Hass et al., 2017).
- The right ventricle is the systemic pumping chamber and cardiac output is dependent on the patent ductus arteriosus (PDA), which normally closes a few days after birth and is kept patent by intravenous infusion of prostaglandin E until surgery is performed. (Cheatham & Deyo, 2016, p.49).
- "A widely accepted hypothesis is that HLHS develops as a result of embryonic alterations in blood flow, such as premature narrowing of the foramen ovale or aortic valve obstruction" (Benson et al., 2016, p.26).
- "An alternative hypothesis for HLHS etiology focuses on the summation of the coordinated actions of signaling pathways and gene regulatory networks that guide the complex process of heart development" (Benson et al., 2016, p.26).

Significance of Pathophysiology

- HLHS is a single-ventricle physiology and persists throughout a patient's life (Donnellan & Justice, 2016).
- Complex and fragile patient population.
- HLHS is a fatal condition if left untreated.
- Without treatment HLHS would be responsible for 25-40% of all neonatal cardiac deaths (Mussa &Barron, 2016).
- Prevention of HLHS not possible.
- There is an increased risk of having a second child with congenital heart disease if the first child has HLHS (Health Jade, 2018).
- Prior to the 1980s HLHS was a universally lethal condition until 1982, when surgical management became available (Mussa & Barron, 2016).
- However, surgical management is not curative because the heart can never be "normal" (Mussa & Barron, 2016).
- The surgical management of HLHS requires "a series of three staged operations that focuses on utilizing the right ventricle to support the systemic circulation" (Mussa & Barron, 2016, p.75).
- Children born with HLHS after their first stage operation remain medically fragile until they have undergone the second stage operation (Nieves et. al., 2017).
- Life expectancy and exercise capacity is decreased (Hass et al., 2017).

Diagnosis

- Prenatally or postnatally
- Prenatal diagnosis can be detected by ultrasound in the second trimester of pregnancy.
- If HLHS is suspected by ultrasound a fetal echocardiogram can confirm diagnosis and show structural abnormalities and how the heart is working (Health Jade, 2018).
- A prenatal diagnosis by ultrasound allows for parent preparedness, prognosis, possible outcomes, and planning of postnatal management (Mussa & Barron, 2016).
- Prenatal diagnosis allows for better hemodynamic stability when compared with a postnatal diagnosis which improves neurodevelopmental outcomes (Mussa & Barron, 2016).
- Postnatal diagnosis is also confirmed by an echocardiogram.

- After confirmation of HLHS diagnosis all treatment options need to be explained to families to empower them to make an informed decision (Mussa & Barron, 2016).
- Treatment options range from surgery to no intervention (comfort care) (Mussa & Barron, 2016).
- Survival rates have increased over time for patients with HLHS due to advancements in surgical technique, postoperative management, and perfusion strategies (Donnellan & Justice, 2016).

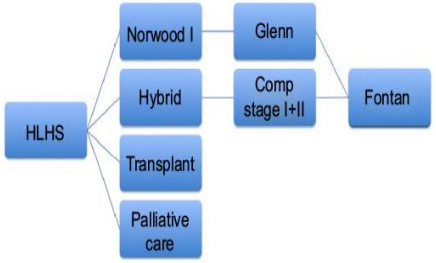

Figure 2. Displays the various surgical pathway for HLHS. This Photo by Unknown Author is licensed under CC BY-NC

Implications for Nursing Care

- Important to avoid early delivery when possible because early term of 37 to 38 weeks' gestation is associated with worse outcomes after cardiac neonatal surgery (Donnellan & Justice, 2016).
- Variation exists in both surgical and medical management in infants with HLHS (Donnellan & Justice, 2016).
- Postoperatively key nursing interventions include pain management and avoiding hyperventilation, which increases pulmonary blood flow and decreases systemic output, monitoring vital signs closely, temperature regulation, and identifying presence of acidosis (Draben, 2018).
- Fragile and complex patient population.
- Growth and nutrition must be continually assessed.
- Before discharge from the hospital and throughout continued follow-up appointments family education on a variety of topics is imperative due to the complexity of HLHS.
- Teaching the family to monitor their infant for signs and symptoms of deterioration and notify the healthcare team with any concerns is crucial for early detection of problems that may warrant treatment. (Nieves et al., 2017).
- Consider the stress burden to the family and offer resources such as counseling.
- Heart transplantation is not primary treatment because neonatal heart donors are rare and to wait for a suitable organ could lead to death. The series of three staged operations is preferred primary treatment (Mussa & Barron, 2016).
- Heart transplantation is a secondary option in children or adults who run into heart failure later in life (Mussa & Barron, 2016).
- HLHS requires lifelong routine follow-up visits with a cardiologist to monitor patient's progress (Centers of Disease Control and Prevention, 2019).
- As medical and surgical advancements continue an increased number of patients with HLHS are reaching adulthood.

Conclusion

HLHS is life threatening if not treated within the first few days of life. HLHS is a rare and severe form of congenital heart disease and requires a cohesive multidisciplinary team to bring forth best outcomes. The ongoing research in genetics brings hope to identifying a cause for HLHS. As medical and surgical advancements continue an increased number of patients with HLHS are reaching adulthood which creates a unique and challenging set of problems (Mussa & Barron, 2016).

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