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**Hypoplastic Left Heart Syndrome in Infancy and Early Childhood**

Hypoplastic left heart syndrome (HLHS) is a congenital heart defect defined by the underdevelopment of the structures of the left side of the heart, including the mitral valve, left ventricle, aortic valve, and aortic arch.1-4 In babies with HLHS, there will be an opening between the left and right atrium called an atrial septal defect (ASD).1-3 The abnormal mitral valve, connecting the right atrium and ventricle, prevents some of deoxygenated blood from flowing from the right atrium to the right ventricle and instead forces blood from the right atrium to the left atrium through the ASD.2 This blood bypasses the lungs and does not become oxygenated. The ductus arteriosus is a normal fetal artery connecting the pulmonary artery to the aorta. In a typically developing heart, this connection is no longer needed after birth and narrows and closes within the first few days of life. However, in HLHS, this connection is necessary to provide oxygenated blood to the body. If the connection remains open, it is referred to as the patent ductus arteriosus (PDA). Continuous intravenous prostaglandin is needed to keep the PDA open.2 Babies with HLHS will appear bluish in color due to mixing of oxygen-rich blood and oxygen-poor blood in the aorta.

HLHS occurs in .016-.036% of live births.1 While rare, this is a complex diagnosis responsible for 23% of cardiac deaths occurring in the first week of life.1 HLHS is typically diagnosed in utero and parents are given three options for post-birth treatment. Option 1 is comfort care where no surgical treatment is provided. Option 2 is placing the child on a heart-transplant waiting list, and Option 3 is to undergo a series of 3 surgeries.5 Without surgical intervention, death in imminent.1 It is estimated that 70% of newborns born today with HLHS will reach adulthood.4 This paper will focus on medical and therapeutic management in infancy and early childhood of HLHS.

The initial surgical procedure, the Norwood procedure, is performed shortly after birth and allows the right ventricle to pump both the lungs and body. It involves enlarging the aortic septal defect allowing blood from the right and left ventricle to mix. The PDA is then closed and a “new” aorta is formed by anastomosing the pulmonary artery to the aortic arch and descending aorta.2 This “new” aorta creates a pathway from the heart to the body.2 Finally, a shunt is placed from the right ventricle to pulmonary artery so blood can be pumped to the lungs.2 The right ventricle is pumping blood to the lungs and heart. Babies generally will need to be hospitalized for one month after surgery.

The second surgery, the bi-directional Glenn procedure, is performed between 4 and 10 months and involves anastomosing the superior vena cava to the pulmonary arteries.2 The shunt placed during the first surgery is ligated at this time. Blood entering the superior vena cava will be immediately directed to the lungs. The right ventricle is only responsible for pumping oxygenated blood to the body. The third and final surgery, the Fontan procedure, is performed between 18-24 months.2 The inferior vena cava (IVC) is anastomosed to the pulmonary artery so blood entering the IVC will be directed to the lungs. The end result of these three surgeries is rewiring the heart so deoxygenated blood is directed to the lungs and the properly functioning right ventricle takes over the duty of pumping oxygenated blood throughout the body. Throughout this extensive surgery process, the patient will be on a combination of medications that may include diuretics, pulmonary vasodilators, lusitropic agents, and anti-inflammatory agents in order to minimize venous pressure and pulmonary vascular resistance, enhance diastolic function, and minimize systemic inflammatory responses.4

Even with the successful completion of surgery, there is a risk of right ventricular dysfunction. Approximately one third of patients will die by the age of 25 from end-stage right ventricular failure.6 If the patient survives, HLHS and its reconstructive surgeries may cause cognitive impairment, speech and language abnormalities, motor delays, and learning disabilities.6 These sequelae are likely due to a combination of abnormal fetal blood flow, chronic cyanosis, cardiopulmonary bypass used during surgeries, hemodynamic instability, cardiac arrhythmias, and cerebrovascular accidents.4,6 Delay in motor development is partially due to hospitalization and immobilization after surgeries, and motor delay is common until adolescence. Physical therapy will be provided in the hospital and the home, when the patient is medically stable enough to be discharged from the hospital. Recognizing developmental delay early is necessary to support motor development in these children.6

**Evaluation, Treatment, and Outcome Measures**

Evaluation and treatment will vary based on the age of the child and the setting in which the child is being seen. Patients with HLHS spend a significant portion of their first year in the neonatal intensive care unit and the pediatric intensive care unit.7 In the hospital setting, pre-and postoperatively, the infant will likely be receiving numerous therapies including physical therapy, occupational therapy, and feeding therapy as tolerated based on medical stability.7,8 Physical therapist will largely focus on positioning and handling, parental education, and providing the baby with safe opportunities for movement, exploration, and play.7,8 The main goal will be increasing parental confidence in handling and encouraging the parent-child bond.7 Education interventions will include direction on appropriate sensory stimulation, appropriate touch, and reading behavioral cues.8 Therapeutic interventions may include the child turning head from side to side while visually tracking a brightly colored toy or facilitating and assisting rolling to sidelying to allow for positional change and opportunity to play in a gravity-minimized position.7 Throughout treatment, physical therapists need to be aware of medical status and the child’s tolerance to physical, visual, and auditory stimulation.8

When the child is medically stable between surgeries and after all three surgeries have been completed, the child will likely be receiving early intervention services in the home. The physical therapist’s role will be to facilitate and assist in developing gross motor milestones in order to explore and participate in his or her natural environment. The therapist will assess developmentally appropriate skills in various positions including prone, supine, sitting, quadruped, and standing. In a study conducted by Rajante et al., prone and standing skills were the most delayed skills in children with HLHS.6 This may be due, in part, to the recommendation that babies avoid the prone position for one month following open heart surgery. It has been suggested that parents of children with HLHS have increased stress and anxiety in regards to their child’s safety. This overprotective quality may lead some parents to avoid prone for longer periods of time, exacerbating gross motor delay as prone is critical position for developing antigravity movements in the first year of life.6

Motor development can be evaluated using the Alberta Infant Motor Scale (AIMS) or the Bayley Scales for Infant Development: gross motor subtest. The AIMS is a performance-based, observational measure of infant gross motor development from 0 to 18 months of age. The Bayley is a performance-based, observational measure of infant and child gross motor development from 0 to 42 months of age. Both scales have normative reference data and can be used to track progress over time. Both of these scales will give similar information, and therefore, it would be redundant to preform both with the same patient. The AIMS may be easier to use and requires less time to perform, but the Bayley is validated for a longer period of the child’s life and thus can better track data in a longitudinal manner. Therefore, the AIMS may be more appropriate for infancy in hospital setting, while the Bayley may be more appropriate for the early intervention setting where the child may be seen by the same therapist until 3 years of age. Both the AIMS and the Bayley have been used to demonstrate motor delay in children with HLHS compared to typically developing children.4,6 In the hospital setting, the AIMS and Bayley will not be the best to track day-to-day outcomes. Assessing and qualifying tolerance to treatment will be a better indicator of day-to-day progress.7,8

**Psychosocial Considerations**

Parents of children with HLHS have reported concerns related to learning medical terms and skills, disrupted parent-child relationship, and increased levels of parental stress and anxiety.9,10 Parents and children go through numerous hospital to home transitions during which parents to take over the child’s medical care including taking daily vital signs, monitoring nutritional status and intake, and symptom management.10 Continuous discharge teaching and planning done throughout the hospital stay gives parents increased confidence in caring for their child.10 Parents will benefit from frequent and repetitive verbal education and written educational materials.10 Providers need to be sensitive to the caregivers emotional state and ability to listen, understand, and retain information.10 Parents may find it difficult to bond with their child in the hospital environment. Physical therapists can play a key role in handling education and incorporating the parents into physical therapy sessions to increase parental confidence.10

Parental stress is multifactorial and is related to the number of surgical procedures that the child has undergone and the age of the child.9 Parents of infants and toddlers have more anxiety due to the majority of complications and surgeries for HLHS occurring in infancy and early childhood.9 Parents also have more stress when their child has additional medical comorbidities.9 Parents are dealing with the loss of the healthy baby and may require additional psychosocial resources and support.9,10

**Review of Literature**

The literature regarding the medical and surgical aspects of HLHS is comprehensive and has expanded in the last decades with the development of the serial surgery procedures. It is widely agreed upon that the three options for infants with HLHS are comfort care, heart transplant, or palliative surgical care. In 1970s and 1980s heart transplant was the best option for HLHS patients. Now that the survival rate with palliative care has increased and up to 25% of patients with HLHS die waiting on the transplant list, palliative care is typically the first-line treatment.11 Statistics regarding survival rates of palliative care vary and some authors report it as a bridge to transplant therapy, while others report it as a destination therapy. This inconsistency is likely due to lack of donor hearts.11 Patients with HLHS will likely only be put on this heart-transplant list when heart failure becomes unresponsive to other medical treatment.11

The literature regarding specific evaluation and treatment plans for physical and occupational therapies in the HLHS infant and toddler populations is far less extensive. It is well-documented that infants with HLHS experience oral feeding difficulties, motor delays, and neurologic and behavioral disturbances. Physical, occupational, and speech therapies are widely used for this population largely due to the potential complications and co-morbidities that come with being a critically ill, hospitalized infant. Therefore, research from NICU, PICU, and early intervention therapy services can be used to develop appropriate evaluation and treatment plans for HLHS patients undergoing palliative care or transplant. Most guidelines for infant and toddler hospital care emphasize parental education, position and handling techniques, and providing appropriate sensory stimulus based on patient tolerance. However, these guidelines rarely provide specific interventions, as each infant will respond differently and will need to be treated accordingly.

Citations:

1. Yabrodi M, Mastropietro CW. Hypoplastic left heart syndrome: from comfort care to long-term survival. *Pediatr Res*. 2017;81(1-2):142-149. doi:10.1038/pr.2016.194
2. Hypoplastic Left Heart Syndrome (HLHS) | Diagnosis and Norwood Procedure. https://www.cincinnatichildrens.org/health/h/hlhs. Accessed November 13, 2019.
3. Ohye RG, Schranz D, D’Udekem Y. Current therapy for hypoplastic left heart syndrome and related single ventricle lesions. *Circulation*. 2016;134(17):1265-1279. doi:10.1161/CIRCULATIONAHA.116.022816
4. Feinstein JA, Benson DW, Dubin AM, et al. Hypoplastic left heart syndrome: current considerations and expectations. *J Am Coll Cardiol*. 2012;59(1 Suppl): S1-42. doi:10.1016/j.jacc.2011.09.022
5. Palisano RJ, Orlin MN, Schreiber J. Spinal Cord Injury. In: *Campbell’s Physical Therapy for Children, 5th Edition*.; 2017:646.
6. Rajantie I, Laurila M, Pollari K, et al. Motor Development of Infants with Univentricular Heart at the Ages of 16 and 52 Weeks. Pediatric Physical Therapy. 2013;25(4):444–450. doi: 10.1097/PEP.0b013e3182a31704.
7. Cass L. *Physical Therapy Management and Treatment of an Infant with Hypoplastic Left Heart Syndrome in the PICU with a Strong Parent-Child Bond: A Case Report*. Iowa Research Online; 2018.
8. Ross K, Heiny E, Conner S, Spener P, Pineda R. Occupational therapy, physical therapy and speech-language pathology in the neonatal intensive care unit: Patterns of therapy usage in a level IV NICU. *Res Dev Disabil*. 2017;64:108-117. doi:10.1016/j.ridd.2017.03.009
9. Caris EC, Dempster N, Wernovsky G, et al. Anxiety Scores in Caregivers of Children with Hypoplastic Left Heart Syndrome. *Congenit Heart Dis*. 2016;11(6):727-732. doi:10.1111/chd.12387
10. March S. Parents’ perceptions during the transition to home for their child with a congenital heart defect: How can we support families of children with hypoplastic left heart syndrome? *J Spec Pediatr Nurs*. 2017;22(3). doi:10.1111/jspn.12185
11. Hsu DT, Lamour JM. Changing indications for pediatric heart transplantation: complex congenital heart disease. *Circulation*. 2015;131(1):91-99. doi:10.1161/CIRCULATIONAHA.114.001377