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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

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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

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Abstract

Background: Hypoplastic Left Heart Syndrome (HLHS) is a congenital heart defect where the left side of the heart is underdeveloped. Children born with HLHS often spend a significant amount of time in neonatal and pediatric intensive care units (NICU and PICU) at the beginning of their lives. This early time is also a very critical time for brain development as well as development of a strong parent-child bond. The PICU environment and medical instability of the infant disrupt this bonding and development. **Purpose:** The purpose of this case study is to present the outcomes of a child with HLHS having undergone multiple procedures and medical complications with a strong parent-child bond. **Case Description:** The child was a 6 month old male born with HLHS. He has been in the NICU and then PICU for his entire life undergoing various cardiac procedures and recovering from them requiring intubation and VA ECMO among other medical interventions. **Intervention:** This infant was receiving services from physical, occupational, speech/feeding, and respiratory therapy as well as child life on top of his cares from nursing. In addition to these specialties, this child was able to develop a strong parent-child bond as his parents were able to spend a majority of their time in the PICU. **Outcome Measures:** The Bayley Scales of Infant Development, Third Edition was used to assess the child's development in different categories compared to general population norms. His overall General Adaptive Composite Score was 75 indicating that he is borderline developmentally delayed. **Discussion:** This case highlights the perceived value of a strong parent-child bond in the outcomes of a medically fragile infant with HLHS in the PICU. In addition, this case study promotes the role of physical therapy in the care and management of this patient population.

Keywords: Hypoplastic left heart syndrome; congenital heart defect; pediatrics; intensive care unit; physical therapy; rehabilitation

Introduction

Hypoplastic Left Heart Syndrome (HLHS) is a congenital heart defect in which the left side of the heart is underdeveloped; i.e., the side of the heart that normally pumps blood to the body.¹ HLHS occurs in approx. 16-36 per 100,000 live births.² What once was a fatal condition can now be treated with a series of surgeries with the first one shortly after birth and two more following as the child grows. The first surgery, or less commonly heart transplant, is required soon after birth to treat this defect otherwise it will result in death. Current data reports that the five-year survival rate is approximately 65% even with surgical repair.³ Pediatric physical therapists will likely encounter these patients in neonatal and pediatric intensive care units (NICU and PICU), however there is limited research on optimal treatment or factors contributing to positive outcomes in this patient population. Thus, the purpose of this case study is to present the outcomes of a child with HLHS at 6 months of age after multiple procedures and medical complications, highlighting the potential role of a strong parent-child bond despite living in the PICU for his entire life.

Background and Surgical Treatment for HLHS

Infants with HLHS undergo a series of 3 heart surgeries to create a functional cardiopulmonary system. These surgeries are the Norwood, bidirectional Glenn, and Fontan.⁴ The Norwood surgery typically is performed within the first 2 weeks of life. In this surgery, the Atrial Septal Defect (ASD) is enlarged. ASD is another congenital heart defect commonly present in infants with HLHS; it is an abnormal communication between the left and right atria which results in blood shunting between atria. It is typically closed surgically however is left open purposefully in infants with HLHS for functional use of the atrial connection. Also during the Norwood, the pulmonary artery is transected, and half of it is connected to the aorta, and the aorta is reconstructed.⁴ This allows the right ventricle, which normally supplies the lungs only, to pump blood to both the lungs and the body. A Blalock–Taussig (BT) shunt is commonly inserted in addition to create a connection from the right subclavian artery to the portion of the pulmonary artery directed to the lungs to establish a source of pulmonary blood flow.⁴

The bidirectional Glenn procedure typically occurs when the child is 4-6 months old, however can be performed earlier than 4 months or delayed past 6 months depending on medical status of the infant. This surgery creates a direct connection from the superior vena cava to the pulmonary artery. With this, the work that the right ventricle has to perform is reduced because it allows the deoxygenated blood to flow directly from the body to the lungs. After this surgery, a child's blood oxygen saturation (SpO₂) typically ranges from 80-85% due to the continuous mixing of oxygenated and deoxygenated blood in the heart.

Finally, the Fontan procedure is typically performed when the child is 18 months – 3 years old. A direct connection is created from the inferior vena cava to the pulmonary artery, so that all returning deoxygenated blood from the body goes directly to the lungs. This allows the right ventricle to have the singular job of pumping oxygenated blood out to the body, and deoxygenated blood no longer mixes with oxygenated blood in the heart. The expected SpO₂ of a child after having all three surgeries returns to about 95%. Unfortunately, this reconstruction of pulmonary and systemic blood flow is not always sustainable for a lifetime, and many of these children will still need further surgeries or cardiac transplant.⁴

Children born with HLHS are bound to spend a significant amount of time in the neonatal and pediatric intensive care units at the beginning of their lives. Their vitals are constantly being monitored and they are being given a variety of medications. It is not uncommon for these infants to require measures such as mechanical ventilation to survive after birth and each surgery. Veno-Arterial Extracorporeal Membrane Oxygenation (VA ECMO) is an important life supporting measure taken to significantly reduce the work of the heart and lungs.⁵ However, it is a very invasive, high-risk measure that involves close monitoring of the infant. Not all infants with HLHS need VA ECMO, and the fact that an infant was placed on VA ECMO indicates that there was likely a period of time that he or she had very low levels of oxygen. This puts them at higher risk of neurodevelopmental delays.

Parent-Child Bond

A parent-child attachment or bond is described as the infant using the parent as a safe haven and secure base to explore from.⁶ A strong bond of an infant to a parent is important for healthy early neurodevelopment.² This initial bond sets the stage for future happiness, independence, and resilience. The purpose of this bond is to make the child feel safe, secure, and protected.⁷ The forming of this bond is made much more difficult and complicated when the child is in the PICU. The PICU is a very unnatural environment for both the infant and the parents. The infant requires medical care by the healthcare team, and the importance of the role of the parent in care of the infant can be neglected or forgotten at first. The parent role changes as they are no longer the sole caregiver, however it is still important for them to make this early bond with the child. Aside from the medical needs of the baby, trying to interact and bond with a baby can be difficult when medical staff is always close. The parents may not feel as comfortable being goofy or possibly singing to their infant if that is something they would typically do at home. Forming this bond in the PICU can be even more difficult for first time parents that don't have any past experience to reference.

This is where physical therapy and other rehabilitation providers can provide crucial education and assistance. PT's can support both the infant and the parents through this process to promote attachment. Using their knowledge to interact with the infant, PT's create a positive learning environment to the degree that the infant can cognitively handle. On the other end, PT's can educate parents on not only the importance of interaction, but different ways to interact with their child. With these skills, the parents can be more comfortable and confident in their role as a parent and can therefore provide a calm, stable support to their infant while he or she is constantly being poked and moved by strangers.

An infant with HLHS is not healthy enough to interact with his or her environment and family in the same way as an average developing infant would. Because of this, these children tend to fall behind in achieving motor, social-emotional, and cognitive milestones. Along with these milestones, early brain development has time sensitive periods of rapid development.⁸ This places patients with HLHS at higher risk for neurodevelopmental impairment. Accordingly, the goal of PT intervention for this population is often to facilitate the development of new skills and control as well as educate the parents on how to properly interact with their child positively while monitoring their medical instability. This case study provides an example of one patient, followed from 4 to 6 months of age, with HLHS.

Case Description

The patient in this case study was born with HLHS by C-Section at a pediatric hospital in the Midwest at 39 weeks and 1 day gestational age. He was diagnosed with HLHS in utero, therefore the medical team was prepared for his medical complexity from birth. He also had an ASD. He underwent the Norwood procedure with BT shunt at 8 days old. From there he was placed on VA ECMO for 2 days, and his chest was closed 4 days after the ECMO was decannulated. Unfortunately, he had to be placed back on VA ECMO for another 8 days just six days after having his chest closed. He underwent another open-heart surgery at 8 weeks old to increase the size of the shunt, and his chest was closed 1 week later. Extubation was trialed 9 days after chest closure, however he did not tolerate it and had to be re-intubated for 2 more weeks. The medical team again trialed extubation and transitioning to nasal intermittent mandatory ventilation (NIMV), but he required reintubation the following day. The medical team then found that he needed his Aorta to be repaired which required another open-heart surgery which was done at 13 weeks old. Three and a half weeks later, he was finally able to tolerate being extubated and transitioned to NIMV. After a long period of stabilization and taking things day by day, he was able to transition down to a regular nasal cannula for oxygen at 24 weeks old. The day after achieving this, an NG tube was placed so that he could begin the process of tolerating food in his stomach.

This patient was chosen to be the subject of this case study because he was doing very well neurodevelopmentally considering all of the medical procedures and complications he had endured in his life thus far. He was also chosen to highlight the perceived value of having a strong parent-child bond along with physical therapy in the PICU. A strong parent-child bond is considered an adjunct intervention to physical therapy in this case. Therefore, he was not picked specifically for the intervention because it was not an intervention directly provided by or controlled by physical therapy. It would be wonderful for every baby in the PICU to get as much parent-child bonding time as this little one received, however it is not realistic or possible for all parents of infants with HLHS to spend a significant amount of time in the hospital.

This case covers a 9-week period when the child was 4-6 months old in the PICU, and was relatively stable medically compared to his first three months of his life. The medical team was working on slowly weaning him off of oxygen and medications including paralytics and sedatives. At 6 months of age, he was fortunate to continue having stable heart function and oxygen saturation and did not yet require the bidirectional Glenn surgery. The patient's immediate family consisted of his mother, father, and a 3-year-old brother who lived only about 15-20 minutes away from the hospital. His mother was on maternity leave for an extended period of time and was often at the hospital to spend time with the patient. She and her husband were very involved and hands on in physical therapy sessions and interested in being educated on ways that they could constructively play with the patient outside of therapy time.

The main goal of the entire medical team and his parents was to help him get healthy enough to be discharged home for a time before having the bidirectional Glenn procedure, the second in the series of cardiac surgeries. The more specific goal of physical therapy was for him to achieve motor, social-emotional, and cognitive milestones to promote optimum early neurodevelopment.

Intervention

The patient in this study was receiving physical therapy, occupational therapy, and speech/feeding therapy as often as he tolerated. At 4 months of age, physical therapy was only able to work with him approximately 2-3 times per week due to medical instability. Typically, a PT session would be 5-20 minutes depending on his state and needs at the time. We would work with him in his crib. At the beginning of the session, we would work on tolerating play time. Starting simple, we would try to get him to move his head side to side by following toys. When he was looking to one side, we would let him try to turn himself into side lying, and then we would facilitate the turn as needed and spend some time in that position. We would then transition him to supported sitting. This support was generally given with one hand on his chest and one hand on his upper back. In this position, we would work on head and neck control. Due to multiple open-heart surgeries and sternal precautions, he was not able to work on prone time until 6 weeks post chest closure which was when he was 3 and a half months old, therefore there was some delay and catching up that he had to do with posterior musculature strengthening against gravity. At first, the goal was for him to tolerate being prone for 10 seconds at a time. Prone time was progressed from prone over a pillow to prone on a flat surface as tolerated. He didn't lift his head much at all in prone at 4 months old.

All of these position changes also challenged his vestibular system, which was considerably under-stimulated for the first part of his life when he was mostly on his back or slightly turned to the side. As an infant being challenged, it was common for him to become upset and start crying. Due to his limited heart function, it was important to calm him down before it affected his vitals. For all these reasons, tolerating position changes was a big physical therapy goal at 4 months of age. During sessions, we were constantly evaluating his ability to tolerate changing positions and staying in those positions for increasing lengths of time, acquiring of new skills, and his overall tolerance to physical therapy in terms of length of session.

At 6 months of age, he was able to be seen up to all 5 days of the week. A typical physical therapy treatment session would last would up to 45 minutes often depending on nap schedule and

whether or not he was weaning medications at the time. At this point in time, he was able to be transferred to a play mat on the floor for a change in scenery and another vestibular challenge. To acclimate to the floor, we would start with him in supine and encourage kicking of legs and reaching of arms with a balloon and other toys. At just about 6 months old, he was able to roll supine to/from side lying, but not consistently. We would facilitate transition to prop sitting in a transition that he would eventually learn how to perform himself. In sitting, he had achieved head control, so the main focus had transitioned to trunk control along with play in sitting. Prone positioning was also worked on during physical therapy sessions as well as a small amount of supported standing. Occasionally, PT would co-treat with Occupational Therapy such as when he was first put into a baby bathtub. Occupational therapy was able to monitor the sensory components of the task and physical therapy was monitoring positioning.

During all of these sessions, parental education was incorporated when his mother or father were present. At 4 months of age, he was just getting to the point that he could handle being held and interacted with more. As his medical status improved, his parents were able to spend more time holding him as well as playing and interacting with him. It is a fair estimate to say that one or both of them were there for an average of 8 hours every day. The patient is their second son, therefore they were familiar with playing with an infant. They were also very willing to learn how to interact with him while monitoring his vitals and managing lines. As they spent more time with him, they became in tune to his tolerance for interaction and were often seen holding or playing with him. They were always looking for suggestions from physical therapy as well as occupational and speech therapy as to how they could cognitively and physically challenge him during play. The nurse's and parents of this patient worked together as a tag team more than any other child on the floor. His parents were very fortunate to be able to open up their schedules in this way and spend a significant amount of time bonding with their child. Not all parents can afford to spend a lot of time in the PICU due to needing to work, taking care of other children, and long driving distance to the hospital.

It can be a challenge for parents in the PICU to become comfortable holding and maneuvering their baby that has multiple IVs, oxygen, and monitoring lines in place. This challenge has both physical and mental components. Mentally, there is a large amount of stress associated with having a child in the PICU. Physically, parents have to spend time growing their confidence in handling their medical fragile baby, especially if they have limited medical knowledge or are first time parents. The parents of this child took on this challenge in stride. It was apparent that they were there for the baby and they were there for each other as well. They made it through the hard times and were celebrating the small victories as they came while being realistic about the future.

Outcome Measures

Along with physical therapy re-evaluation within each session, the Bayley Scales of Infant Development, Third Edition (Bayley-III) was used to assess this patient's capabilities in motor, social-emotional, and cognitive skills. The children's hospital where this patient was observed uses the Bayley-III to assess all infants after having been placed on ECMO. It is a norm-referenced measure that is used for children ages 1-42 months.⁹ The Bayley-III does not provide any sensitivity or specificity data and few other articles provide this data. A 2015 article looked into the sensitivity and specificity of the cognitive and language sections of the Bayley-III in preterm children. It found that the sensitivity of mild-moderate cognitive delay was 29.4-38.5% and the specificity was 92.3-95.5%.¹⁰ In the language section, they found the sensitivity of mild-moderate language delay to be 40-46.7% and sensitivity to be 81.1-85.7%. The reliability and validity of the Bayley-III is considered to be insufficient based on the limited amount and quality of current research on the subject.⁹

The patient was tested at 6 months and 1 day old and his results are shown below in Table 1.

Table 1. Bayley-III Scores and Age Equivalents by Subtest.

Subtest	Total Raw Score	Scaled Score	Age Equivalent
Cognitive	24	7	5 months
Receptive Communication	9	9	5 months, 10 days
Expressive Communication	5	6	3 months, 20 days
Fine Motor	24	15	8 months
Gross Motor	20	7	5 months
Social-Emotional	64	9	Full mastery for his age
Adaptive Behavior	N/A	49	75 th percentile for his age

The results of his testing display that he was only slightly delayed in some of the categories aside from expressive communication. It is reasonable that his expressive communication would be delayed due to requiring long term intubation and sedation. His receptive communication skills may have been delayed due to being on sedatives for a long period of time as well. One likely factor contributing to his gross motor delay would be that he was unable to work on skills in prone positioning due to his sternal precautions. When assessing his cognitive abilities, he may have had difficulties because the items on the cognitive scale require motor abilities as well. His overall General Adaptive Composite Score on the Bayley-III was 75 indicating that he is borderline developmentally delayed.

A 2018 study assessed Bayley-II scores in infants who had survived 14 months with surgical intervention but did not require heart transplant. They found the scores of the infants to be significantly lower than normative means.³ This study cannot be directly compared to the patient in this case study because he was assessed with the Bayley-III at 6 months old instead of the Bayley-II at 14 months of age. However, it is telling that this patient population tends to score below normative means.

Cognitively, a normally developing 6 month old should be able to play with a single object and bang it, but he had not achieved that yet. With language, he was beginning to coo and he was sometimes calmed just by making eye contact with one of his parents. He was also turning his head towards a voice which is on track for a 4-5 month old.⁸ From a social perspective, he was listening to voices, smiling at his parents, and had maybe laughed once or twice according to his parents. This is typically achieved at 4-5 months.⁸ According to the Bayley-III, his social-emotional score at 6 months of age indicates that he is exhibiting more self-regulation as well as interest in the environment around him.

As for motor skills, in the supine position, he was able to kick a lot, but he was just starting to bring his feet to his mouth which is typical of a 4-5 month old.⁸ At 6 months, he was only able to sit without support when placed there and it was only for limited periods of time with close supervision which is considered average. He was just beginning to hold and manipulate toys at 6 months which is typically achieved at 4-5 months.⁸ He was more delayed with prone positioning activities, however this was to be expected due to his sternal precautions as noted earlier. These are just a few of his various skills. At 6 months of age, it is difficult to say what is truly delayed versus average or low average achievement when this patient was borderline for delay according to his Bayley-III scores and his skills were almost caught up to averages.

From a clinical perspective, his improvement was notable over time. Just during the time of observation he developed new skills and increased his tolerance to interaction which is shown in the frequency of treatment portion of Table 2 below. Some of this change was likely due to age and maturation independent of the intervention, however it required a lot of facilitation by therapists as well as time and attention during play with his parents focused on these goals to help him achieve. While he was behind norms for attaining some milestones, he was making progress as he was becoming more medically stable. He was also able to wean down his oxygen needs and medication doses such as Milrinone, which is used for blood pressure support, which is outlined in the table below. Weaning off of oxygen was especially significant for him because oxygenation is key with HLHS. The level of oxygen required would affect his ability to be discharged home. Once he was down to using a nasal cannula, he was able to start trying to feed orally which was another large step in the direction of progress.

Table 2. Clinical and medical changes over time.

	4 months old	5 months old	6 months old
Frequency of treatment	2-3x/week	3-4x/week	4-5x/week
Milrinone dose	0.5 mkm	0.25 mkm	discontinued
Oxygen needs	NIMV	Nasal CPAP-HFNC	Regular nasal cannula

Discussion

One important factor when working with infants with HLHS is that they have to be able to be calmed. The heart of a patient such as this doesn't have the reserves to tolerate high levels of stress, such as with crying and getting worked up. Although crying is common and a form of language for infants, too much crying and overexertion of the fragile heart is to be avoided. The weaning of medications and oxygen make his body work even harder, and he experiences withdrawal symptoms from some of the medications. This infant was often able to be calmed by his mother by being held and talked to. This reaction indicates a strong bond with his mother. Because his mother had spent so much time with him rocking, holding, and talking softly to him, the healthcare staff was also able to calm him down when parents were not around. Staff was able to use the holding, rocking, and talking to calm him as well. Of course, it was not quite the same as when mom held him. It was very beneficial to the child that she and her husband were able to spend more time with him than even his nurses. This ability to be calmed could contribute to overall medical stability of the child.

When an infant with HLHS in the PICU first becomes tolerant of increased interaction and movement, the physical and occupational therapists often begin providing treatment. They play a role in helping the parent find what calms their infant while in this critical, fragile state. PT's and/or OT's can educate the parents on incorporating a calming touch during nursing cares when the baby is too unstable to handle being held or touched more than just for necessary cares. Encouraging the parents to take an active role in cares can help build their confidence and remind them that they play a crucial role. We can help them understand that instead of stepping away when medical staff walks in, they should instead step up to understand how to provide for the infant better and be there to calm the infant around people that can be scary in the infant's eyes. From there, we progress to teaching parents how to interact with the baby while holding them as well as how to build up pillows and towels to create a supportive environment for the baby while lying in bed. As the infant progresses, PT's and OT's continue to be a source of education to the parents and an educated set of eyes and hands to evaluate and challenge the infant appropriately. In this way, we can help facilitate a strong parent-child bond which will only improve and optimize the health of the child.

This case study highlights the perceived value of a strong parent-child bond in the outcomes of a medically fragile infant with HLHS in the PICU. In addition, this case study promotes the role of physical therapy in the care and management of this patient population. Although physical therapy is not the main treatment of HLHS, it is a critical component of care in the PICU for development support and parent education. Physical therapists provide an important channel of family education that can help foster a strong parent-child bond and teach optimal calming techniques in an unfamiliar and fragile environment for the parents and child.

This case study is somewhat limited in the fact that it is not an easily reproducible intervention. It was an adjunct intervention rather than a direct physical therapy intervention however it made the overall care of this infant different compared to other infants in the PICU. From the healthcare caregiver point of view, it seemed that one likely factor to the overall progress of the infant despite significant medical setbacks was the strong parent-child bond, however it could be many other things. Another limitation is that this intervention does not currently and will not ever have any randomized controlled trials completed to determine the independent value of the parent-child bond on the outcomes of the

children in this patient population. However, any means to optimize their care may be of value to healthcare caregivers.

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