Fall 2014

Protein Losing Enteropathy following Fontan Palliation in the Single Ventricle Population

Alaina Dunkleberger
Otterbein University, Alaina.dunkleberger@otterbein.edu

Protein Losing Enteropathy following Fontan Palliation in the Single Ventricle Population

Alaina Dunkelberger, RN, BSN, CCRN
Otterbein University, Westerville, Ohio

Review of Single Ventricle Surgical Management

Congenital heart defects requiring single ventricle palliation are a rare but life-threatening occurrence. There are multiple defects resulting in single ventricle physiology including defects in which the right or left ventricle within the heart is either undeveloped (hypoplastic left heart syndrome, hypoplastic right heart syndrome), or the valve to the main pulmonary artery did not form (pulmonary atresia). These defects prevent the heart from supplying adequate blood flow to the lungs or body. Single ventricle congenital heart defects are not easily treated surgically due to their complexity in nature. Single ventricle surgical palliation typically involves three open-heart surgeries including the Norwood or Hybrid Procedure, the Bidirectional Glenn procedure, and finally the Fontan procedure, (Hardiman, 2013, p. 327).

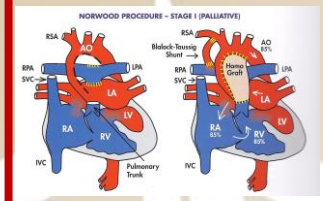

Figure 1. Hypoplastic Left Heart Syndrome anatomy (left). Norwood procedure anatomy (right).

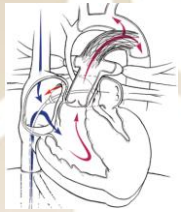

Figure 2. Hybrid procedure circulation.

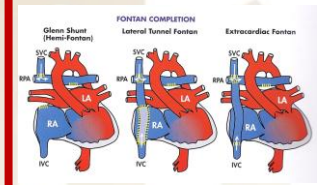

Figure 3. Bidirectional Glenn anatomy (left). Intracardiac fontan anatomy (middle). Extracardiac fontan anatomy (right).

Traditional Three Stage Single Ventricle Palliation

The Norwood procedure is the traditional operation performed during the neonatal period. This procedure converts the single operating ventricle into the main pumping chamber for both systemic and pulmonary blood flow. The pulmonary artery and aorta are combined, removing the main pulmonary artery from the branching pulmonary arteries. A shunt is placed between the neo-aorta and the pulmonary arteries to provide blood flow to the lungs, (Hardiman, 2013, p. 327).

Nationwide Children's Hospital in Columbus, Ohio, often performs the Galantowicz-Cheetham Procedure, also known as the Hybrid procedure, in place of the Norwood procedure. This procedure entails bands being placed around the pulmonary arteries to reduce pulmonary blood flow which encourages shunting back into the systemic system. A stent is then placed in the patent ductus arteriosus to maintain mixed oxygenated and deoxygenated blood being delivered to the body. Finally, a balloon atrial septostomy is performed to allow for more mixing of oxygenated and deoxygenated blood.This procedure requires less manipulation of the heart than the Norwood procedure and also allows the child to avoid cardiopulmonary bypass (Rowland, et al., 2008.)

The Bi-directional Glenn is performed four to six months after the Norwood procedure. The shunt between the pulmonary artery and the aorta is removed. The superior vena cava is then connected directly to the right pulmonary artery. This connection brings deoxygenated blood from the upper half of the body passively to the lungs, bypassing the right ventricle. (Hardiman, 2013, p. 327).

The Fontan operation is typically performed18-24 months after the bi-directional Glenn procedure. The inferior vena cava is connected to the right pulmonary artery. This, combined with the bi-directional Glenn, allows for all deoxygenated blood to flow passively to the lungs. The single ventricle is now only providing systemic perfusion (Hardiman, 2013, p. 328).

Protein-Losing Enteropathy

Protein-losing enteropathy (PLE) is a rare but serious condition that can occur following Fontan palliation. This disease occurs when protein from the body is "lost" or leaking into the intestinal tract. Hypoalbuminemia in generally the first indication of PLE (Sathiyasekaran, 2013). Diagnosis is then confirmed by the presence of fecal alpha 1-antitrypsin, proving the existence of blood protein below the pylorus in the gastrointestinal tract (Braampskamp, Dolman, & Tabbers, 2010).

PLE is said to occur in approximately 3–15% of patients after the Fontan operation and carries a high risk of mortality (Umar & DuBaise, 2009). Five and ten year survival rates after PLE diagnosis following the Fontan surgery were noted to be 48% and 30% respectively (Umar & DuBaise, 2009).

Risk Factors for Developing PLE

Pre-operative risk factors indicated include having a hypoplastic left ventricle and increased end-diastolic pressures. Longer operative times have been indicated in the development of PLE post-operatively. Post-operative echocardiograms showing a reduced ventricular area have been linked with the development of PLE (Umar & DuBaise, 2009). Post-operative renal failure is also related to the development of PLE in the future (Meadows & Jenkins, 2011, p. 370).

Theories on the Pathophysiology behind Protein-Losing Enteropathy

The pathophysiology of the development of protein-losing enteropathy has not been precisely determined but several theories exist. The lymphatic and cardiovascular system are closely related; elevated central venous pressures increases lymphatic production while simultaneously slowing lymphatic return (Meadows & Jenkins, 2011, p. 373). One theory is that the increased systemic venous pressures related to the passive blood flow to the pulmonary artery causes dilation of the lymphatic system within the gastrointestinal tract leading to leakage of protein into the gastrointestinal system (Umar & DiBaise, 2009). A second theory is the elevated systemic venous pressure alongside the impaired cardiac output from a single ventricle state combine to impair perfusion and oxygenation to the gastrointestinal system (Umar & DiBaise, 2009). The impaired blood flow and ischemia compromises the epithelium and leads to protein leaking into the lumen. "Gross and microscopic pathologic examination of the intestine in patients with protein-losing enteropathy demonstrates several characteristic findings consistent with lymphatic engorgement, stasis and insufficiency," (Meadows & Jenkins, 2011, p. 369). A third theory suggests that the chronic low cardiac output state of a person with single ventricle circulation causes inflammation; cytokines cause vasoconstriction and have been found at elevated levels immediately after Fontan palliation which is not surprising, but a study in 2006 found elevated cytokine levels years after the procedure (Ostrow, Freeze, & Rychik, 2006, p. 698). The pathophysiologic significance of these findings lies in the fact that there is no definitive causative source leading to the development of protein-losing enteropathy. The lack of a determined underlying cause creates difficulty for providers attempting to medically manage this population.

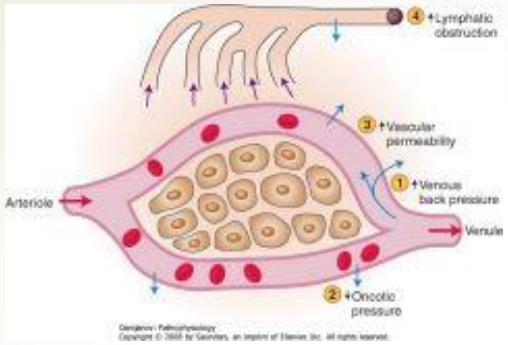

Figure 4. Depiction of the development of protein-losing enteropathy related to lymphatic and venous obstruction.

Clinical Manisfestations of Protein-Losing Enteropathy

The loss of protein into the gastrointestinal tract creates many life-altering and life-threatening clinical manifestations. Patients with PLE can become severely edematous which can inhibit respiration. Diarrhea may occur causing electrolyte imbalances as well as dehydration. Ascites develops as the leaking of protein continues; it can eventually become severe enough to cause discomfort and difficulty breathing, requiring abdominal paracentesis. Pleural effusions are a common occurrence in patients with PLE, requiring the placement of chest tubes or even chemical or surgical pleurodesis. Pericardial effusions can also occur which have the potential to become life-threatening (Caltepe & Comba, 2013).

Medical Management of Protein-Losing Enteropathy

Management of protein-losing enteropathy has proven to be inadequate for patients and frustrating for healthcare professionals. Supportive measures include dietary modifications (high protein, low fat), diuretics including Lasix and spironolactone, and intermittent albumin infusions (Braamskamp, Dolman, & Tabbers, 2009, p. 1183).

More specific treatment includes heparin therapy which has shown limited success (Meadows & Jenkins, 2011, p. 371). Heparin's mechanism of action in aiding in the resolution of PLE is unclear. A deficiency of enterocyte heparin in patients that showed success with heparin therapy has been suggested (Meadows & Jenkins, 2011, p. 371). Corticosteroids have been used as well with some limited success with 2-3 weeks of treatment, the mechanism of action in the disease remains unclear but is thought to be related to its anti-inflammatory effects (Meadows & Jenkins, 2011, p. 371). Adrenal suppression has been commonly found among patients with PLE, often requiring stress-doses of corticosteroids (Goldberg, Dodds, & Rychik, 2011, p. 79).

Surgical treatments include cardiac catheterization to relieve any obstructions to systemic or pulmonary blood flow. Creating a fenestration within the Fontan when systemic pressures are elevated to allow blood flow into the right atrium has shown some encouraging results; the creation of a fenestrated Fontan increased the patient's risk for stroke (Meadows & Jenkins, 2011, p. 371). Attempting to improve the low cardiac state that is unavoidable with single ventricle circulation via atrioventricular pacing when appropriate has shown some success at improving levels of serum protein (Goldberg, Dodds, & Rychik, 2010, p. 116). Cardiac transplantation is viewed as the final option for patients with unrelenting protein-losing enteropathy. Following cardiac transplantation, recurrence of PLE has been rare (Meadows & Jenkins, 2011, p. 372).

Nursing Implications

Nursing implications for protein-losing enteropathy are multi-faceted. The nurse should provide emotional support for the patient and their family. PLE is typically a chronic, fatal condition so the diagnosis can be devastating. The nurse can improve the experience for the patient and family by advocating for early palliative care involvement. There are several ethical issues revolving around the care of the single ventricle population, "…Because the neonate is incapable of participating in the decision-making process, the nurse must advocate for the child adequately and succinctly explaining all facets of each of the available treatment options to the parents," (Zeigler, 2003, p. 69). The nurse is responsible for the proper administration of prescribed therapies and educating the patient and/or their families on their use. The patient should be closely monitored for worsening symptoms, specifically respiratory failure related to ascites or pleural effusions. The nurse should be prepared to intervene in a medical emergency such as cardiac tamponade from a pericardial effusion. .

References

Braamskamp, M., Dolman, K., & Tabbers, M. (2010). Clinical practice: Protein-losing enteropathy in children. *European Journal of Pediatrics, 169*(10), 1179-85.

Caltepe, G., & Comba, A. (2013). Protein-losing enteropathy in children. *Turkish Pediatrics Archive / Turk Pediatri Arsivi, 48*(1), 7-12.

Goldberg, D., Dodds, K., & Rychik, J. (2010). Rare problems associated with the Fontan circulation. *Cardiology In The Young*, *20 Suppl* 3113-119. doi:10.1017/S1047951110001162
Goldberg, D., Dodds, K., & Rychik, J. (2011). New concepts: development of a survivorship programme for patients with a functionally univentricular heart. *Cardiology In The Young,21 Suppl* 277-79.

Hardiman, T. (2013). Hypoplastic left heart syndrome: an overview. *British Journal Of Cardiac Nursing, 8*(7), 325-331.

Meadows, J., & Jenkins, K. (2011). Protein-losing enteropathy: integrating a new disease paradigm into recommendations for prevention and treatment. *Cardiology In The Young, 21*(4), 363-377.

Additional Sources

Ostrow, A., Freeze, H., & Rychik, J. (2006). Protein-losing enteropathy after fontan operation: Investigations into possible pathophysiologic mechanisms. *The Annals of Thoracic Surgery, 82*(2), 695-700.

Rowland, D., Galantowicz, M., Corbitt, R., Cannon, M., Heskett, N., Warmbrier, J., & Baker, A. (2008). Parent's resource guide: Hypoplastic left heart syndrome. Retrieved from http://www.nationwidechildrens.org/Document/Get/37115

Sathiyasekaran, M. (2013). Protein-Losing Enteropathy: Treatment & prognosis in pediatrics. New Dehli, India: *Jaypee Brothers Medical.*

Umar, S., & DiBaise, J. (2009). Protein-losing enteropathy: Case illustrations and clinical review. *The American Journal of Gastroenterology, (105),* 43-49.

Zeigler, V. (2003). Pediatric ethics, issues, & commentary. Ethical principles and parental choice: treatment options for neonates with hypoplastic left heart syndrome. *Pediatric Nursing*, *29*(1), 65-69.

Figure 1 and Figure 3. "Hypoplastic Left Heart Syndrome (HLHS), or Hypoplasia of the Left Heart" [Image]. (2010). http://heartdefectsforeveryone.blogspot.com/2010/04/hypoplastic-left-heart-syndrome-hlhs-or.html

Figure 2. "First Stage Hybrid 1". [Image] (2008). www.nationwidechildrens/org/document/get/37115

Figure 4. "Pathophysiology" [Image]. (2011). http://fce-study.netdna-ssl.com/images/upload-flashcards/back/0/1/44210096_m.jpg

OTTERBEIN UNIVERSITY