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Protein Losing Enteropathy following Fontan Palliation in the Single Ventricle Population
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Theories on the Pathophysiology behind Protein-Losing Enteropehy

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. 371). One theory is that the increased systemic venous pressures related to the passive blood flow to the pulmonary artery cause dilation of the systemic venous system within the gastrointestinal tract leading to leakage of protein into the gastrointestinal system (Umar & DiBatte, 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 & DiBatie, 2009). The impaired blood flow and ischemia compromises the epithelial lining of the small intestine leading to the lumen. “Gross and microscopic pathologic examination of the intestine in patients with protein-losing enteropathy demonstrate several characteristic features in common with patients with the disease, including increased lymphatic flow, stasis, and muscular disruption.” (Meadows & Jenkins, 2011, p. 368). 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 in these patients. This can lead to protein leakage into the lumen. “Hepatic and elevated cytokine levels years after the procedure (Ostro, Freese & Rychik, 2006, p. 686).” The pathophysiologic significance of these findings lies in the fact there is no definitive causal 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.

Risks for Developing PLE

Pre-operative risk factors indicate that PROCEDURE may increase the risk of mortality (Umar & DiBatie, 2009). Five and six month survival rates following the Fontan surgery were noted to be 84% and 30% respectively (Umar & DiBatie, 2009).

Risk Factors for Developing PLE

Protein-Losing Enteropehy (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. Hypoproteinemia is generally the first indication of PLE (Sheyaarhawan, 2013). Diagnosis is then confirmed by the presence of fecal alpha-1 antitrypsin, a protein blocking the gastrointestinal blood below the pylorus in the gastrointestinal tract (Brueckmann, Doleman, & Tabbers, 2013).

PLE is said to occur in approximately 3–15% of patients after the Fontan operation and carries a high risk of mortality (Umar & DiBatie, 2009). Five and six month survival rates following the Fontan surgery were noted to be 84% and 30% respectively (Umar & DiBatie, 2009).

Traditional Three Stage Single Ventricle Palliation

The Norwood procedure is the traditional operation performed during the neonatal period. This procedure involves the single ventricle circulation that becomes the pulmonary circulation for both systemic and pulmonary arteries. Both the right and left pulmonary arteries are combined, removing the main pulmonary artery and surrounding the 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).

Nativoidre’s Children’s in Columbus, OH, perform a bidirectional Glenn in their Hybrid Procedure, also known as the hybrid procedure, in place of the Norwood procedure. This procedure entails bands being placed around the pulmonary arteries and combined ventricle to reduce pulmonary blood flow which encourages shutting back into the systemic system. A shunt is then placed in the patient 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 oxygenated and deoxygenated blood to flow. This procedure requires less time than the Norwood procedure and also allows the child to avoid prolonged bypass times (Rayok, et al., 2008.)

The Bi-directional Glenn is performed four to six months after the Norwood procedure. This procedure removes the ventricle circulation from the pulmonary artery and the aorta is the returned. The superior vena cava is then connected to the main pulmonary artery. This connection brings oxygenated blood directly from the blood body passively to the lungs, bypassing the right ventricle (Hardiman, 2013, p. 327).

The Fontan operation is typically performed after the development of the bio- directional Glenn procedure. The inferior vena cava is connected to the main pulmonary artery. This, combined with the presence of an atrial septal defect allows deoxygenated blood to passively flow to the lungs. The single ventricle is now only responsible for systemic perfusion (Hardiman, 2013, p. 328).

Pathological Manifestations of Protein-Losing Enteropehy

The loss of protein into the gastrointestinal tract creates many life-altering and life-threatening clinical manifestations. Patients with PLE can become universally edematous which can inhibit respiration. Oedema may occur causing electrolyte imbalances as well as dehydration. Acute dehydrations as well as the leaking of protein content can be acutely be overcome severe enough to cause discomfort and difficulty breathing, requiring a hospitalization. Pleural effusions are a common occurrence in patients with PLE requiring the placement of chest tubes or even chest wall or surgical procedures. Furthermore patients can also occur which have the potential to become life-threatening (Calleja & Comba, 2013).

Medical Management of Protein-Losing Enteropehy

Management of protein-losing enteropathy has proven to be inadequate for patients and families. Supportive care measures are the primary focus of care (Tabbers & DiBatie, 2009). Supportive measures include dietary consultation, including Labial and sironilactone, and intermittent albumin infusions (Brueckmann, 2013). The impaired blood flow and ischemia compromise the epithelial lining of the small intestine leading to the lumen. “Hepatic and elevated levels years after the procedure (Ostro, Freese & Rychik, 2006, p. 686).” The pathophysiologic significance of these findings lies in the fact there is no definitive causal 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.

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References