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Protein Losing Enteropathy following Fontan Palliation in the Single Ventricle Population

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The pathophysiology of 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 increase 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 vasculature within the gastrointestinal tract leading to leakage of protein into the gastrointestinal system (Umar & DiBalle, 2009). A second theory is the elevated systemic venous pressure along the inadequate cardio output from a single ventricle state combined to impair perfusion and oxygenation to the gastrointestinal system (Umar & DiBalle, 2009). The impaired blood flow and ischemia compromise the epithelial barrier to protein leaking into the lumen. “Gross and microscopic pathologic examination of the intestine in patients with protein-losing enteropathy demonstrates several characteristic histological changes including increased engorgement, stasis and muco-fibrosis.” (Meadows & Jenkins, 2011, p. 368). A third theory suggests that the chronic low cardiac output state of a person with single ventricle circulates influences calcium ion fluctuations; calcium ions cause vasoconstriction and have been found at elevated levels in patients with protein-losing enteropathy. Hyponatremia and elevated electrolyte levels years after the procedure (Ostrow, Freese, & Rychik, 2006, p. 1086). 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.

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 is generally the first indication of PLE (Sathiyasekaran, 2013). Diagnosis is then confirmed by the presence of focal alpha-1 antitrypsin, proving the existence of blood protein below the pylorus in the gastrointestinal tract (Brøeimp, Delman, & 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 & DiBalle, 2009). Five and six month survival rates for diagnosis following the Fontan surgery were noted to be 84% and 30%, respectively (Umar & DiBalle, 2009). The pathophysiologic nature of PLE is multifactorial in origin. The impaired blood flow and ischemia compromise the epithelial barrier to protein leaking into the lumen. “Gross and microscopic pathologic examination of the intestine in patients with protein-losing enteropathy demonstrates several characteristic histological changes including increased engorgement, stasis and muco-fibrosis.” (Meadows & Jenkins, 2011, p. 368). A third theory suggests that the chronic low cardiac output state of a person with single ventricle circulates influences calcium ion fluctuations; calcium ions cause vasoconstriction and have been found at elevated levels in patients with protein-losing enteropathy. Hyponatremia and elevated electrolyte levels years after the procedure (Ostrow, Freese, & Rychik, 2006, p. 1086). 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.

Medical Management of Protein-Losing Enteroapthy

Management of protein-losing enteropathy has proven to be inadequate for patients and families alike. Supportive measures include dietary restriction, diuretics, including Lasix and spironolactone, and intermittent albumin infusions (Braampskamp, DiBaise, & Tabbers, 2009). There are no specific measures to control or arrest the disease process. In general, support involves administration of corticosteroids (Goldberg, Dehli, & Rychik, 2010). Although developing PLE is a rare (Meadows & Jenkins, 2011, p. 372). Transplantation, recurrence of PLE has been encouraging results; the creation of a transatrial communication within the patent ductus arteriosus into the right atrium has shown some promising results. In a small pilot study, the creation of a fenestration in the right atrium increased the patient’s NAP for survival (Cheetham, Rowland, et al., 2008). Attempting to improve the low cardiac state and protein-losing enteropathy, there are few therapeutic choices for patients. Current treatment of PLE includes the use of corticosteroids. Corticosteroids are used as a bridging therapy in the development of protein-losing enteropathy. Following cardiac transplantation, recurrence of PLE has been noted (Rowland, et al., 2013, p. 372).

References


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Nursing Implications

Nursing implications for protein-losing enteropathy are to proactively assess children and families for this condition to ensure that the patient and family are provided with the proper information and support. These children and families experience chronic, fatal condition so the diagnosis can be traumatic for the patient and family. The nurse can improve the experience for the patient and family by advocating for early palliative care teams. These teams can provide ethical issues resolving around the care of the young patient with a fatal diagnosis. The nurse in in a unique position to advocate for the vulnerable treatment options to the parents,” (Ziegler, 2005, p. 69). The nurse is responsible for the proper care of patients, including accurate administration of prescribed therapies and education that are non-invasive and beneficial to their use. The nurse should be closely monitored for warning symptoms, specifically respiratory failure related to severe edema. In these cases the patient should be prepared to intervene in a medical emergency such as cardiac tamponade from a pericardial effusion.

Additional Sources


Figure 1 and Figure 3: “Hypoplastic Left Heart Syndrome: Hypoplastic Left Heart Syndrome.” (2010). http://www.nationalthoracic.org/get/37115

Figure 2. “First Stage Hybrid I.”. www.nationalthoracic.org/document/75786

Figure 4. “Pathophysiology” (2010). http://life.therapy.netdna-ssl.com/flash/144224/2009_05_26_00035📙