PEDIATRIC SURGERY *Update**
Vol. 53 No. 01 JULY 2019

TPN Peritoneal Extravasation

Umbilical venous catheters (UVC) in newborns provide intravascular access for administration of intravenous fluids, parenteral nutrition (TPN), drugs, transfusions and central venous monitoring especially for management of low birth weight infants. Improper placement of umbilical catheters is associated with complications that may lead to morbidity and mortality. The tip of the umbilical venous catheter should be positioned at the junction of the inferior vena cava and right atrium just at or above the level of the diaphragm (T7 to T9). Also, adequate blood return must be obtained before use. A normal positioned UVC between T7 and T9 can stay in use up to 14 days if intravenous access is in need, otherwise is advised to discontinue the UVC by day 10 and replace it with a percutaneously place intravenous central catheter. Improper placement of UVC can lead to intraperitoneal extravasation of TPN. This may also lead to vessel perforation or liver capsule disruption due to hepatic necrosis. The complication of intraperitoneal spillage has an insidious onset. The infant will develop abdominal distension, tenderness associated with acute pain, ascites, dehydration, hemoconcentration, characteristic induration of the abdominal wall due to infiltration of the soft tissue and acute renal failure from hypovolemia and hypertonicity of TPN. Abdominal ultrasound will demonstrate ascites. Paracentesis if performed reveals a cloudy fluid high in glucose, triglycerides and protein consistent with TPN. The differential diagnosis is chylous ascites with high chylomicrons and lymphocytes in a peripheral smear of fluid sample. Intraperitoneal TPN extravasation has also been reported after placement of femoral central a venous catheter. Management of TPN peritoneal extravasation should consist of catheter removal, paracentesis or peritoneal exploration to accomplished peritoneal lavage of the offending fluid. Prognosis is usually good if not associated with serious liver laceration with uncontrollable hemorrhage.

References:

1- Egyepong J, Jain A, Chow P, Godambe S: Parenteral nutrition--ascites with acute renal failure as a complication from an umbilical venous catheter in an extremely low birth weight infant. BMJ Case Rep. 2011 Apr 26;2011. pii: bcr0220113813. doi:10.1136/bcr.02.2011.3813.
2- Sztajnbok J, Troster EJ: Acute abdomen due to late retroperitoneal extravasation from a femoral venous catheter in a newborn. Sao Paulo Med J. 120(2):59-61, 2002
3- Adesanya O, Naqvi M: Term Neonate With Liver Laceration, Obstructive Uropathy, and Ascites-Secondary to Extravasation of Total Parenteral Nutrition: A Complication of Malpositioned
Umbilical Venous Catheter. Glob Pediatr Health. 2016 Oct 12;3:2333794X16670494. eCollection 2016.
4- Shareena I, Khu YS, Cheah FC: Intraperitoneal extravasation of total parental nutrition infusate from an umbilical venous catheter. Singapore Med J. 49(2):e35-6, 2008
5- Guzoglu N, Erdeve O, Yilmaz Y, Dilmen U: Intraperitoneal extravasation from umbilical venous catheter in differential diagnosis of neonatal chylous ascites. Acta Paediatr. 99(9):1284, 2010

6- Selvam S, Humphrey T, Woodley H, English S, Kraft JK: Sonographic features of umbilical catheter-related complications. Pediatr Radiol. 48(13):1964-1970, 2018

Alpha-1-Antitrypsin Deficiency

Alpha-1-antitrypsin is a glycoprotein produced primarily in the liver secreted under the influence of proinflammatory cytokines forming several globulins whose roles are to suppress additional tissue damage by neutralizing granulocyte elastase and proteinase primarily in the lung during infective and inflammatory process. Is responsible for protecting tissues against proteolytic damage by enzymes like neutrophil elastase and proteinase. It is also a tissue reparation inductor. Deficiency of alpha-1-antitrypsin (A1ATD) is a rare autosomal recessive co-dominant disorder most often seen in Northern European ancestry populations. A1ATD is the most common genetic cause of pediatric liver disease and transplantation. The defect in alpha-1-antitrypsin expression in the hepatocyte is followed by damage to the liver and/or lungs primarily. A1ATD can be asymptomatic or long-lasting and even permanently symptomatic depending in expression on the genotype and numerous exogenous factors such as infection, toxic or other damages to the liver and lungs. Liver damage occurs due to intrahepatocyte retention of alpha-1-antitrypsin and in the lung due to the lack of its protective effect. Early in life A1ATD causes a cholestatic syndrome, conjugated hyperbilirubinemia beyond the second week of life, associated with low birth weight and poor weight gain sometimes difficult to differentiate from biliary atresia. The liver damage is severe in only 1-2% of affected patients, most often has a slowly progressive character and juvenile cirrhosis develops in 15% of cases. In other organs A1ATD can cause pulmonary emphysema (chronic obstructive pulmonary disease), cytoplasmic anti-neutrophil cytoplasmic vasculitis and inflammatory necrotizing panniculitis in the skin. A1ATD individuals screened at birth who smoke develop COPD by age of 40 years. A1ATD is also associated with other diseases such as rheumatoid arthritis, sinusitis, nasal polyps, inflammatory bowel disease, peptic ulcer disease and diabetes. Management of A1ATD is limited. A diagnosis of A1ATD may have important implications including testing of family members, genetic counseling, smoking avoidance, avoidance of high risk occupations and consideration of augmentation therapy.

References:

1- Radlovic N, Lekovic Z, Radlovic V, Simic D, Topic A, Ristic D, Ducic S, Baletic A: Alpha-1-antitrypsin deficiency in children: clinical characteristics and diagnosis. Srp Arh Celok Lek. 142(9-10):547-50, 2014
2- Greulich T, Nell C, Hohmann D, Grebe M, Janciauskiene S, Koczulla AR, Vogelmeier CF: The prevalence of diagnosed alpha-1-antitrypsin deficiency and its comorbidities: results from a large population-based database. Eur Respir J. 2017 Jan 4;49(1). pii: 1600154. doi: 10.1183/13993003.00154-2016. Print 2017 Jan.
3- Aboussouan LS, Stoller JK: Detection of alpha-1 antitrypsin deficiency: a review. Respir Med. 103(3):335-41, 2009
4- Piitulainen E, Mostafavi B, Tanash HA: Health status and lung function in the Swedish alpha 1-antitrypsin deficient cohort, identified by neonatal screening, at the age of 37-40 years. Int J Chron Obstruct Pulmon Dis. 12:495-500, 2017
5- Khan Z, Venkat VL, Soltys KA, Stolz DB, Ranganathan S: A Challenging Case of Severe Infantile Cholestasis in Alpha-1 Antitrypsin Deficiency. Pediatr Dev Pathol. 20(2):176-181, 2017
6- Fregonese L, Stolk J: Hereditary alpha-1-antitrypsin deficiency and its clinical consequences. Orphanet J Rare Dis. 2008 Jun 19;3:16. doi: 10.1186/1750-1172-3-16.

Thromboelastography

Coagulation disorders can lead to intra- or postoperative bleeding associated with increased morbidity and mortality. The transfusion of blood products to correct coagulopathy disorders is guided by clinical judgement, standard laboratory testing (PT, PTT, platelets, bleeding time, etc.), and thromboelastography testing. Thromboelastography (TEG) is a viscoelastic hemostatic assay, a functional test of clot formation and degradation performed on whole blood, at a point of care (emergency department, intensive care units or operating room) for clotting system deficiency that analyzes each phase of the coagulation process. Blood from a patient is mixed with citrate and placed in a cup of the TEG machine connected to a computer. The cup oscillates and the coagulation process occurs. This testing rapidly generates numeric and graphic results that can lead to guided-directed intervention for correct of coagulation disorders. Five variables are measured using thromboelastography. They are: R (reaction time) quantify as the time until measurable clot is formed, usually between five and 10 minutes. Should the patient have a prolonged R time then management is with fresh frozen plasma transfusion. K time - time since initial clot formation until it has a 20 mm fixed strength, normally one to 3 minutes. An abnormal K-time means low fibrinogen levels and is managed with cryoprecipitate. Alpha angle - is the speed of fibrin accumulation, similarly, affected by fibrinogen levels. Normal alpha angle is 53-72 degrees. Again, an abnormal alpha angle is managed with cryoprecipitate. Maximum amplitude (MA) - consist of the highest vertical amplitude of the TEG tracing. MA reflects the strength of the clot normally reaching 50-70 mm. A narrow maximum amplitude is managed with platelets infusion. LY30 percentage - represent 30% decrease in the amplitude of the TEG tracing thirty minutes after maximum amplitude is obtained. It is a measure of fibrinolysis normally being zero and 8%. A higher than normal LY30 is indicative of a hypercoagulable state and may be managed with antifibrinolytics such as aminocaproic acid. TEG is superior to conventional coagulation testing in detecting early trauma coagulopathy reducing mortality. TEG should be available in trauma level I and II centers.

References:

1- Wasowicz M, Srinivas C, Meineri M, et al: Technical report: analysis of citrated blood with thromboelastography: comparison with fresh blood samples. Can J Anaesth 55(5): 284-9, 2008

2- Chan KL, Summerhayes RG, Ignjatovic V, et al: Reference values for kaolin-activated thromboelastography in healthy children. Anesth Analg 105(6): 1610-3, 2007

3- Rusell RT, Maizlin II, Vogel AM. Viscoelastic monitoring in pediatric trauma: a survey of pediatric trauma society memebers. J Surg Res. 214: 216-20, 2017

4- Wikkelso, A(1), Wetterslev J, Moller AM, Afshari A: Thromboelastography (TEG) or thromboelastometry (ROTEM) to monitor haemostatic treatment versus usual care in adults or children with bleeding. Cochrane Database Syst Rev. 2016 Aug 22;(8):CD007871. doi:10.1002/14651858.CD007871.pub3.

5- Figueiredo S, Tantot A, Duranteau J: Targeting blood products transfusion in trauma: what is the role of thromboelastography? Minerva Anestesiol. 82(11):1214-1229, 2016

6- Nogami K: The utility of thromboelastography in inherited and acquired bleeding disorders. Br J Haematol. 174(4):503-14, 2016

***Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP**
Professor of Pediatric Surgery, University of Puerto Rico - School of Medicine, Rio Piedras, Puerto Rico. Director - Pediatric Surgery, San Jorge Children's & Woman Hospital.
Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426.
Tel (787) 340-1868 and 999-9450 E-mail: *titolugo@coqui.net*
Internet: http://home.coqui.net/titolugo
**** PSU 1993-2019***
ISSN 1089-7739