Anesthetic Management of a Patient with Maple Syrup Urine Disease Undergoing Liver Transplantation from Living Related Donor

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ABSTRACT

Maple syrup disease is a rare metabolic condition affecting the pediatric population. It is related to reduction or deficiency of branched chain a-ketoacid dehydrogenase (BCKD). The mainstay of management is by strict dietary protein restriction with branch chain amino acid (BCAA) free diet along with supplementation of other essential amino acids and frequent monitoring of BCAA levels. However, the approach should be through multidisciplinary team addressing dietary requirement, growth and psychomotor development. This can be challenging due to compliance with such regimens as well as metabolic decompensations during physiological stress that can occur in fasting, surgery, infection and inflammation. Liver transplantation has been shown to enable MSUD patients to have unrestricted diet and normal BCAA post-operatively. Although MSUD is a rare disease, there is extensive literature published related to it from pediatrics, gastroenterology, hepatology and transplantation point of view. We describe a case report of peri-operative management of 4 years old MSUD patient undergoing living related liver transplantation (LRLT) with normal BCAA levels post-operatively as this type of surgery is relatively recent in addition to the limited data on the peri-operative management of MSUD patients undergoing surgical procedures.

Key words: Maple syrup urine disease, Liver transplant, Anesthetic management, Peri-operative

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