

Publication

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Maple syrup urine disease is a rare autosomal-recessive metabolic disorder caused by a deficit of oxidative decarboxylation of branched-chain amino acids. First symptoms appear in the neonatal period. Without treatment the disease is characterized by rapid progression of neurological symptoms. During stressful situations, such as infection or surgery, patients may experience severe ketoacidosis, rapid neurological deterioration and hypoglycemia. The perioperative management of a 26-year-old man with maple syrup urine disease is described, a review of the disease is given and anaesthesia-related implications are discussed.

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