Abstract Type: Case Report/Case Series

Unyime Ituk, M.B., B.S., F.C.A.R.C.S.I.; Terrence Allen, M.B., B.S., FRCA; Ashraf Habib, M.B., Ch.B., FRCA
Duke University Medical Center

Introduction: Glutaric aciduria type 1 (GA1) is an autosomal recessive disorder resulting from a genetic deficiency of glutaryl-CoA dehydrogenase, essential for the metabolism of lysine, hydroxylysine and tryptophan [1]. This deficiency results in the accumulation of glutaric and 3-hydroxyglutaric acids in the urine with secondary carnitine depletion. It is a rare metabolic disorder with an incidence of 1:30,000 [2]. It can be marked by episodes of neurologic crisis during which patients develop dystonia, athetosis & seizures. This is triggered by increased catabolism, such as fever, dehydration, fasting and surgery. Pregnancy & outcomes in parturients with GA1 are not well documented. We present a report on the anesthetic management of a parturient with GA1.

Case Report: A 23 year old lady, G1P0 with a BMI of 41kg/m2 and a history of GA1 presented for a scheduled cesarean section at 36 weeks gestation for marginal placenta previa. She was diagnosed with GA1 as an infant and has been on restricted protein diet, riboflavin and L-carnitine. Urine organic acids, plasma amino acids and plasma/Urine L-carnitine levels were monitored during pregnancy. In order to reduce the period of preoperative fasting, her surgery was scheduled as the first case of the day. She was admitted the night before and plasma L-carnitine, liver enzymes & serum glucose were measured. An IV infusion of L-carnitine 667mg in 10% Dextrose in NS @ 125ml/hr to prevent excess protein metabolism was started. An emergency metabolic protocol was in place in the event of a neurologic crisis. This involved administration of a loading dose of L-carnitine, riboflavin, dextrose 25%, treatment of vomiting, sedation and neuroprotection with phenobarbital. Serum lactate and glucose were measured before and after surgery. She had an elective cesarean delivery under spinal anesthesia. Hyperbaric bupivacaine 12mg, fentanyl 15mcg and preservative free morphine 150mcg were administered intrathecally. A prophylactic phenylephrine infusion was administered with 2 L of lactated ringers to prevent maternal hypotension. The L-Carnitine infusion was continued perioperatively until regular PO diet was resumed. She delivered a 2680g male infant with Apgar scores of 8 and 9 at 1 and 5 mins. Her plasma L-carnitine and Lactate were within normal limits 24 hours postoperatively. She remained asymptomatic and was discharged home on the 3rd postoperative day.

Discussion: The management goal of GA1 in the parturient is to minimize catabolic stress and prevent acute neurologic crisis, which can rapidly Result in injury to the basal ganglia. Reducing the period of starvation in conjunction with an L-Carnitine infusion and neuraxial anesthesia may have attenuated the catabolism associated with cesarean delivery [3]. It is important also to avoid drugs with extrapyramidal side effects in these patients.