Internet Journal of Medical Update

Journal home page: http://www.akspublication.com/ijmu

Letter to Editor

Glycogen storage disease type 3: a management challenge in pregnancy

Ojo Gbemisola Okunoye, Corinne Deakin and Simon Maguire

Department of Obstetrics and Gynecology, University Hospital of South Manchester, Wythenshawe , Hospital, Manchester M23 9LT, UK

Sir

Glycogen storage disease (GSD) type 3 is an inborn error of glycogen metabolism resulting from the deficient activity of glycogen debranching enzyme. It is associated with progressive liver disease, myopathy and risk of cardiomyopathy with an incidence of 1 in 100000 live births[1]. We describe a successful pregnancy outcome in a woman with GSD type 3 with an emphasis on the risk of peripartum cardiopulmonary complication.

A 34-year-old primigravida with known GSD type 3 was booked at 8 weeks. She was diagnosed with GSD type 3 in childhood and developed progressive skeletal myopathy, hypertrophic cardiomyopathy and liver cirrhosis in adolescent years. She developed liver failure at age 30 and underwent a successful liver transplant a year later. Antenatally she remained on tacrolimus 2 mg twice daily and her liver function tests remained normal. Serial echocardiogram did not reveal any evidence of deterioration of the previously diagnosed left ventricular hypertrophic cardiomyopathy and she had no symptoms suggestive of cardiac decompensation. Fetal wellbeing was satisfactory with a normal anatomy scan and good growth velocities on serial scans.

A multidisciplinary review at 34 weeks agreed on a planned delivery by elective caesarean section at 37 weeks with the aim of avoiding spontaneous onset of labor. This was based on the unpredictability of labor with associated tachycardia which, coupled with her underlying diastolic dysfunction, could precipitate pulmonary congestion. An earlier gestation of 37 weeks was chosen in view of her history of liver transplant, which carries a risk of spontaneous preterm labor.

An elective caesarean section was performed in cardiac theatre at 37 weeks under GA with continuous cardiac monitoring, a CVP, arterial line and transesophageal echocardiogram in place. A live baby boy weight 3130 gm was delivered in very good condition. The procedure was surgically uncomplicated with a blood loss of 350 ml. However, just after the delivery, she developed acute pulmonary oedema. She received intravenous frusemide and remained in the cardiac intensive care unit where she made good recovery and was transferred to the postnatal ward within 48 hours, prior to her hospital discharge on postnatal day 4.

Existing literature on pregnancy in women with GSD type 3 mainly involved pre-transplant patients and focused on antenatal prevention of hypoglycaemia with frequent cornflour supplements[2,3]. The risk of peripartum cardiopulmonary complication has not been highlighted in seemingly stable patients. The susceptibility to hypoglycaemia is not a concern in post liver transplant patients. The acute pulmonary oedema developed by our patient, with the benefit of invasive monitoring and meticulous fluid management, is believed to have been induced by the transient increase in intravascular volume caused by 'uterine autotransfusion' following uterine contraction after delivery of the baby. This physiologic process is usually unnoticeable in healthy parturients. Pre-operative anticipation and planning was critical to the successful outcome in this case. Clinicians looking after women with GSD type 3 should be aware that pregnancies in these women remain high-risk even post liver transplant.

REFERENCES

1. Hobson-Webb LD, Austin SL, Bali DS, Kishnani PS. The electrodiagnostic characteristics of Glycogen storage disease type III. *Genet Med*. 2010;12(7):440-5.
2. Lee P. Successful pregnancy in a patient with type III glycogen storage disease managed with cornstarch supplements. *Br J Obstet Gynaecol.* 1999;106(2):181-2.
3. Bolton SD, Clark VA, Norman JE. Multidisciplinary management of an obstetric patient with glycogen storage disease type 3. *Int J Obstet Anaesth*. 2012;21(1):86-9.

GO Okunoye
Email: okunoyezaza@yahoo.co.uk