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CASE REPORT

Anaesthetic management of an obstetric patient with Pompe disease

H. J. Cilliers,* S. T. Yeo, N. P. Salmon

Department of Anaesthesia, Hereford Hospitals NHS Trust, Hereford, UK

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Abstract Pompe disease (Glycogen storage disease type II) leads to abnormal glycogen deposition in various vital organs resulting in multiple systemic sequelae. We present the anaesthetic management for caesarean section of a 31-year-old parturient with known Pompe disease. The parturient had symptoms and signs of respiratory dysfunction and the pregnancy was complicated by preeclampsia. She underwent urgent caesarean section under regional anaesthesia resulting in the birth of a healthy baby girl. To our knowledge, this is the first reported case of both spinal anaesthesia for caesarean section and successful live birth in a patient with Pompe disease.

Introduction

Pompe disease (glycogen storage disease type II) is a hereditary condition seen in 1 in 40 000 live births that leads to dysfunction of the enzyme acid α-glucosidase. The subsequent build up of glycogen in body tissues causes the widespread systemic dysfunction that characterizes the disease. The main effects are progressive muscle weakness with eventual cardiorespiratory failure. The anaesthetic management of the disease in infants and children has been described, but there are no case reports of adults with the disease. This is the first case report to describe the anaesthetic management of a patient with Pompe disease for a caesarean section.

H. J. Cilliers, Senior House Officer, **S. T. Yeo**, **N. P. Salmon**, Consultant Anaesthetists, Hereford County Hospital.
* Correspondence to: H.J. Cilliers, Department of Anaesthesia, Hereford Hospitals NHS Trust, Union Walk, Hereford, HR1 2ER.
E-mail address: hjcilliersmail-medical@yahoo.co.uk

Case report

A 31-year-old parturient (G2,P1), known to have Pompe disease, presented during the 13th week of gestation for antepartum care. Her previous pregnancy three years before had been complicated by severe preeclampsia and intrauterine growth retardation. At 27 weeks of gestation her labour had been induced and she received epidural analgesia. As the fetus showed signs of distress, an emergency caesarean section had been performed. For this the epidural was extended with 5-mL boluses of 0.5% bupivacaine to a total of 25 mL. Block of cold and touch sensation extended to T4 bilaterally but patchy distribution necessitated conversion to general anaesthesia. This proceeded without complication. Unfortunately the baby died three days later due to complications of prematurity.

On this presentation she was highly compliant with a protein rich diet to counter her muscle weakness. She was 161 cm tall with a body mass index of 19 kg/m^2.

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Her exercise tolerance was good, although she had some difficulty climbing stairs and sitting up from a supine position. Subjectively her respiratory function was not impaired, but spirometry at 10 weeks of gestation indicated a mixed obstructive and restrictive pattern with no improvement on inhaled β-stimulants (Table 1). She had no other medical problems, although routine antenatal blood tests showed mild derangement of hepatic enzymes (aspartate aminotranferase (AST) 99 U/L, alanine aminotransferase (ALT) 130 U/L, lactate dehydrogenase (LDH) 934 U/L and alkaline phosphatase (ALP) 142 U/L). These were similar to levels recorded in her previous pregnancy. Renal function tests and an echocardiogram performed at 28 weeks were both within normal limits.

Pregnancy was uneventful until 31 weeks when preeclampsia developed. She was given labetalol 100 mg twice daily and aspirin 75 mg once daily. Her blood pressure remained slightly elevated at 140/90 mmHg until 35 weeks, when traces of protein appeared in her urine. She was referred for an anaesthetic opinion considering her Pompe disease, obstetric history and hypertension. A provisional plan was made for elective caesarean section at 38 weeks using a regional technique. The dose of labetalol was progressively increased to 200 mg three times daily.

Unfortunately her preeclampsia deteriorated, with increasing proteinuria and labile hypertension; she was admitted to hospital at 37 weeks. Her preeclampsia continued to worsen with a diastolic pressure of in excess of 100 mHg controlled only by i.v. hydralazine infusion. At this point the decision was made to deliver the baby by urgent caesarean section. Her full blood count, renal function and clotting were normal, with liver enzymes unchanged compared to early pregnancy.

Continuous arterial monitoring was established with a 20-gauge radial arterial cannula to facilitate the titration of antihypertensive medication. Oral sodium citrate was given to increase gastric pH. Combined spinal-epidural anaesthesia was chosen for the caesarean section. This was performed without complication in a sitting position at the L4-5 level with loss of resistance to saline using a needle-through-needle technique (16-gauge Tuohy and 27-gauge pencil point needles). Hyperbaric 0.5% bupivacaine 1.5 mL and fentanyl 20 μg were given intrathecally. An epidural catheter was then inserted leaving 5 cm in the epidural space. A further 5 mL of bupivacaine 0.5% was administered through the epidural catheter to ensure sufficient cephalad spread of the block. A dense block developed with loss of sensation to cold to T3, and loss of light touch at T8, 10 min after the spinal injection. The i.v. hydralazine infusion was stopped as a reduction in blood pressure was anticipated following the spinal injection. Compound sodium lactate solution was administered at 75 mL/h with further 100-mL boluses to maintain a systolic pressure >110 mm Hg. Vasopressors were avoided. Continuous cardiotocography confirmed fetal well-being throughout the conduct of spinal anaesthesia. A healthy 1980-g baby was delivered 8 min after skin incision. Apgar scores at 1 and 5 min were 9 and 9 respectively. Incremental i.v. boluses of oxytocin (Syntocinon) 2 units were given after delivery. Prophylactic intravenous amoxicillin 1 g with clavulanic acid 200 mg was also administered. Estimated total blood loss throughout the operation was approximately 500 mL. A total of 800 mL of i.v. fluid was given.

In the recovery ward the woman developed a non-specific frontal headache with mild photophobia. Two i.v. boluses of fentanyl 25 μg were administered 10 min apart for analgesia. During the episode her blood pressure was stable between 134/76 and 140/80 mmHg. She complained of no other neurological symptoms or signs, but was closely monitored for worsening of the symptoms. Her headache resolved within 2 h. As the effects of the spinal wore off, her diastolic pressure increased again, but stayed below 100 mmHg. The labetalol was weaned as the blood pressure returned to normal over the next two days. During this time she was managed in a high dependency area.

Post caesarean section analgesia was achieved with regular paracetamol, codeine phosphate and intramuscular morphine as required. Once her renal function was established as normal, regular diclofenac sodium was introduced to supplement her analgesia. She was slowly weaned off labetalol over the next few days and was discharged with a healthy baby girl four days post partum.

Table 1 Spirometry results

	Volume	% of predicted
Total lung capacity	4.28 L	87.3
Residual volume	2.14 L	150.1%
Vital capacity– erect	2.14 L	61.7%
Vital capacity – supine	1.49 L	
Expiratory reserve volume	0.84 L	68.1%
Forced vital capacity	2.12 L	60.8%
Forced expiratory volume in 1 s	1.85 L	61.0%
Peak expiratory flow	200 L/min	50%

Discussion

Pompe disease is a rare genetic disorder with dysfunction of acid α-glucosidase which causes abnormal deposition of glycogen in the lysosomes of body tissues.[1–3] To the best of our knowledge this is the first case report of a successful birth from a woman with