Management of labor and delivery in a woman with Morquio syndrome

C. Delgado, C. Kent, M. Sedensky, C. Ciliberto, R. Landau
Department of Anesthesiology and Pain Medicine, University of Washington, Seattle, WA, USA

ABSTRACT
Morquio syndrome, a congenital mucopolysaccharidosis, presents several challenges for the provision of effective labor analgesia. We report the case of a woman admitted for induction of labor who received an early epidural and subsequently required cesarean delivery. Optimal bilateral labor analgesia was not achieved despite multiple adjustments, and systemic analgesia was needed for cesarean delivery.

Keywords: Morquio syndrome; Analgesia; Obstetric; Cesarean delivery

Introduction

Morquio syndrome is a rare autosomal recessive lysosomal storage disorder, caused by a deficiency in N-acetylgalactosamine-6-sulfate sulfatase (GALNS), resulting in tissue accumulation of glycosaminoglycans and interference with cellular function.[1] Severe skeletal dysplasia is a hallmark of the disease. It also affects connective tissues, including the airway and heart valves, resulting in high mortality in infancy. Surgical intervention and recently available enzyme replacement therapy may prolong life expectancy.[2] Anesthesiologists are more likely to see adult patients with this condition, including women of childbearing age. To the best of our knowledge, this is the first description of neuraxial analgesia for induction of labor in a woman with Morquio syndrome, and of the challenges encountered in managing labor epidural analgesia and epidural anesthesia for cesarean delivery.

Case report

A 28-year-old G6P0 woman with Morquio syndrome was evaluated at the obstetric anesthesia high-risk clinic at 28 weeks of gestation. Previous obstetric history included two dilatations and evacuations under general anesthesia (GA), and three spontaneous abortions that required no intervention. She had undergone external fixation of a left femoral fracture and a left hip replacement under GA, but sustained a left sciatic nerve injury resulting in left foot drop and neuropathic pain requiring methadone treatment. Anesthetic airway records reported uneventful tracheal intubations with a grade 1 direct laryngoscopic view and cervical stabilization. She had also undergone uneventful epidural catheter insertion that had provided effective postoperative analgesia.

The woman weighed 56 kg and was 142 cm tall (body mass index 27.8 kg/m^2). Cardiopulmonary auscultation revealed no abnormality and airway examination was unremarkable (Mallampati class I, thyromental distance and mouth opening within normal ranges). There was a full range of motion of the cervical spine, but significant lumbar kyphosis and thoraco-lumbar scoliosis (Figs. 1 and 2). Absent dorsiflexion of the left lower extremity was noted with decreased sensation. Neurological examination was otherwise unremarkable.

A magnetic resonance imaging (MRI) scan performed one year before pregnancy confirmed kyphosis at the lumbar level with retrolisthesis at L1–2, anterolisthesis at L2–3, mild canal stenosis at T1–2 and T2–3, but no signs of thecal sac compression. There was moderate to severe canal stenosis and thecal sac compression in the lumbar levels. There was no evidence of atlanto-axial instability (Fig. 3). As neurologic examination remained stable after imaging, no new diagnostic procedures were requested. Echocardiogram was normal, with preserved ejection fraction, no valvular abnormalities and no evidence of pulmonary hypertension. Mild restrictive lung capacity was present (forced vital capacity, 68%).

Labor was induced at 39 weeks of gestation with a transcervical Foley catheter followed by intravenous oxytocin. At 3 cm cervical dilation, the patient reported a verbal pain rating score (VPRS) of 7 out of 10 and requested epidural analgesia. Using a loss-of-resistance to saline technique with a 17-gauge Tuohy needle (Epidural Catheterization Kit with FlexTip Plus®

Accepted August 2015
Correspondence to: C. Delgado, Department of Anesthesiology and Pain Medicine, University of Washington, Box 356540, 1959 NE Pacific Street, BB-1469 Seattle, WA 98195-6540, USA.
E-mail address: delgadou@u.washington.edu

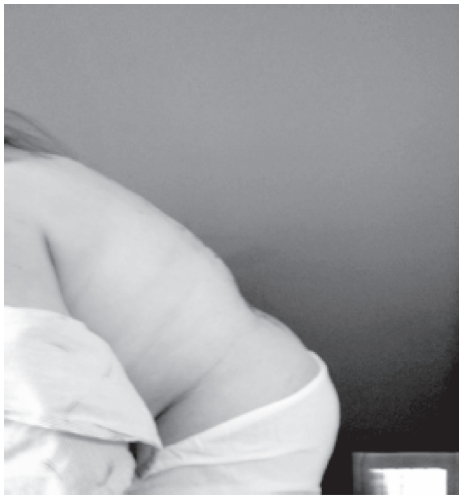

Fig. 1 Lumbar kyphosis

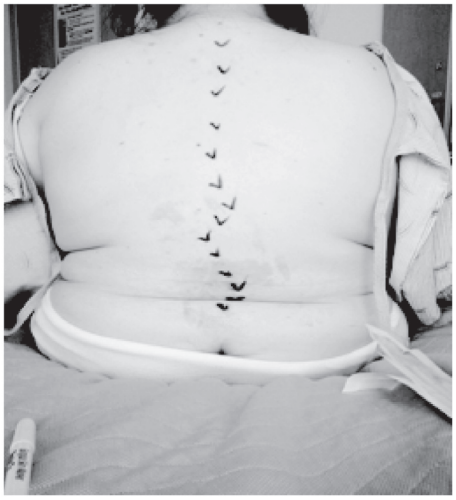

Fig. 2 Thoraco-lumbar scoliosis

Catheter, AK-05502-UWMC, Arrow International Inc, Reading, PA, USA), the epidural space was found at a depth of 4.5 cm at the presumed L4–5 interspace using a midline approach with the patient in the sitting position. Insertion was uneventful and 5 cm of catheter was left in the epidural space. After negative aspiration, a 2 mL bolus of 1.5% lidocaine with epinephrine 1:200 000 was given; sensory dermatomal assessment with pinprick revealed a unilateral block (absent pinprick to L1 on the left side). Although it is our usual practice to initiate epidural analgesia with incremental doses of 0.25% bupivacaine 5–10 mL, due to the anatomical abnormalities and mucopolysaccharidosis, lower dose requirements were anticipated and cautious incremental doses were used (Table 1). An initial dose of 0.25% bupivacaine 3 mL was given. Patient-controlled epidural analgesia (PCEA) using bupivacaine 0.0625% with fentanyl 2 μg/mL (continuous infusion 6 mL/h; bolus 3 mL; lockout 10 min) was subsequently started. This differs from our standard setting of continuous infusion at 10 mL/h, bolus 5 mL and 10 min lockout. The patient continued to report pain with contractions, primarily on the right side. A further dose of 0.25% bupivacaine 3 mL eventually established a bilateral dermatomal level from T10 to L3.

Over the next 6 h, the woman intermittently reported moderate to severe pain scores that required analgesic interventions as both epidural bolus doses and an increase in the infusion rate from 6 to 10 mL/h; the concentration was also increased to bupivacaine 0.125% with fentanyl 2 μg/mL at the same PCEA setting. Due to a predominantly one-sided block, the catheter was pulled back 1 cm, but this did not improve analgesia.

The epidural catheter was removed and a new catheter inserted at the same lumbar interspace using the same positioning and technique. Ultrasonography was not used, since experience of the provider and ease of previous placement deemed it unnecessary. The epidural space was found at 5.5 cm and 5 cm of catheter was threaded into the space. After a 3 mL dose of 1.5% lidocaine 1.5% with epinephrine 1:200 000, PCEA was restarted and proved more effective as the patient was unable to feel contractions. Three hours later PCEA was stopped at the patient's request as despite reporting back pain, she was unable to feel contractions. Analgesia was restarted after two hours when the patient reached full cervical dilatation.

After 17 h of labor without adequate progress, cesarean delivery was planned. Over approximately 20 min, 15 mL of 2% lidocaine with epinephrine 1:200 000 was administered in incremental doses, producing a significantly asymmetric sensory block. The epidural catheter was pulled back 1 cm and an additional bolus of 2% lidocaine 5 mL was given. Block symmetry with pinprick testing improved and the abdomen was prepared for surgery. However, sensory testing by the obstetricians before incision revealed an inadequate block on the right side of the abdomen. A bolus of 3% chloroprocaine 3 mL (chosen for its speed of onset and alternative metabolism by plasmatic hydrolysis) was administered. The anesthetic level was judged to be adequate for surgery 33 min after the first dose of lidocaine.

A healthy female baby was delivered. Oxytocin was started as an infusion (15 U in 0.9% saline 250 mL over 90 min) and produced adequate uterine tone. Preservative-free epidural morphine (Duramorph) 3 mg was given after delivery of the baby, which is our normal practice. The patient reported right-sided abdominal pain after uterine exteriorization, and epidural 3% chloroprocaine 5 mL was given. Intraoperative intravenous supplementation was considered necessary; ketamine 20 mg, midazolam 1 mg and fentanyl increments to