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Kyphomelic dysplasia, Pierre Robin Sequence and pregnant

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ABSTRACT

We present the anaesthetic management of a parturient with kyphomelic dysplasia and Pierre Robin Sequence who underwent elective caesarean delivery. Potential anaesthetic issues and management strategies are discussed.
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Introduction

Restricted growth, also referred to as dwarfism, is defined as an adult with a height of less than 148 cm.[1]

It may occur in more than 400 medical conditions,[2] all of which can be broadly classified using the trunk-to-limb ratio into two main types: proportionate short stature or disproportionate short stature.[3]

Kyphomelic dysplasia (KD) is a rare prenatal skeletal disease that causes disproportionate short stature. It is characterised by short bowed long bones, irregular

flared metaphyses, flared ribs and mild vertebral flattening.[4] Variable features include micrognathia, mid-face hypoplasia, cleft palate and the potential to develop cervical subluxation over time.[4]

Pierre Robin Sequence (PRS) comprises the triad of micrognathia, glossoptosis and airway obstruction.[5] It is often associated with cleft palate.

Case report

The patient was a 23-year-old primigravida with KD and PRS. She was 116 cm tall and had thoracic scoliosis, an exaggerated lumbar lordosis, a fixed flexion deformity of the hips, and was known to have spinal stenosis in the upper thoracic region. She had undergone a cleft palate repair as an infant but airway management details for this procedure were not available. She had received no subsequent anaesthetics. Airway assessment revealed significant micrognathia, an inter-incisor distance of 3.5 cm, a limited ability to protrude her jaw, a reduced thyromental distance, restricted neck extension and a Mallampati score of two. She had no other co-morbidities.

She was keen to have a vaginal delivery, but cephalo-pelvic disproportion rendered its success unlikely. In view of her short stature, spinal deformities and craniofacial abnormalities, safe and rapid anaesthesia with central neuraxial blockade or general anaesthesia (GA) for emergency surgery was considered challenging. After discussion with the obstetric team and the patient, it was agreed that the safest option was elective caesarean delivery at 37 weeks' gestation. She declined GA and a combined spinal-epidural (CSE) technique using a reduced spinal dose was planned. If unsuccessful, awake fibre-optic tracheal intubation and GA was planned, having all necessary equipment prepared and available in the operating room. A similar plan was made were she to present early for an emergency caesarean delivery.

She attended for elective caesarean delivery at 37 weeks' gestation. Standard monitoring was instigated, with a paediatric cuff used for blood pressure measurement. A 16-gauge venous cannula was inserted. Additional padding was used to support her hips while the CSE anaesthetic was performed with her in the sitting position, using a two-space technique, with full asepsis maintained. Spinous processes were palpable and ultrasound imaging of the spine was not required. The epidural component was sited at the L1/2 interspace with a 18-gauge Tuohy needle via a midline approach, using a saline loss-of-resistance technique. The epidural space was located at a depth of 5 cm. No test dose was administered. The spinal component was performed at the L2/3 interspace using a 25-gauge pencil-point needle. Locating the subarachnoid space was technically challenging but was achieved following re-angulation of the needle. Cerebrospinal fluid was freely aspirated

and 1.5 mL 0.5% hyperbaric bupivacaine with 200 µg diamorphine was injected intrathecally, following which she was positioned supine on the operating table with 20 degrees of left lateral tilt. She received crystalloid fluid co-loading, a variable-rate phenylephrine infusion and there were no clinically important changes in blood pressure. She developed a block to cold sensation bilaterally to the T4 dermatomal level. Surgery was uneventful and no epidural supplementation was required. A 2.87 kg neonate was born with Apgar scores of 9 and 9 at 1 and 5 minutes.

A lower threshold for blood transfusion was agreed pre-operatively due to the patient's estimated blood volume of approximately 4500 mL. Estimated blood loss was 400 mL and no transfusion was required. She received 1000 mL of crystalloid fluid during the caesarean delivery and her blood pressure was maintained within normal parameters.

Mother and baby were monitored in a recovery area for two hours before transfer to a midwife-led postnatal ward. Postoperative analgesia consisted of a short course of regular paracetamol and ibuprofen with supplemental oral morphine solution as required. No complications were encountered, and both mother and baby were discharged home approximately 24 h later.

Discussion

Achondroplasia is the most common cause of disproportionate short stature and most case reports describe patients with this genetic condition.[6] Parturients with restricted growth, in particular disproportionate short stature, are at an increased risk of surgical delivery,[1,6,7] primarily because of cephalopelvic disproportion[7–8] which causes malpresentations and inadequate fetal descent.[1] Cardiorespiratory compromise secondary to the impingement of a gravid uterus may also necessitate surgical delivery[7–8] and it has been suggested that there is an increased risk of uterine rupture.[1] Patients with restricted growth may have cervical spine instability[4] and tracheal intubation can be difficult.[7,9–10] Our patient also had features of PRS and based on our airway assessment we concluded that the safest method for securing the airway would be an awake fibre-optic intubation.

Central neuraxial block may be difficult due to spinal stenosis, a small epidural space, an exaggerated lumbar lordosis and kyphoscoliosis.[7–11] In one review article, it was attempted using a variety of methods for 12 parturients with restricted growth who underwent caesarean delivery. Difficulty with the neuraxial technique occurred in nine patients and six required multiple attempts.[12] Four examples of CSE anaesthesia for caesarean delivery were found in parturients with restricted growth in whom the subarachnoid space could not be identified;[11–13] one patient developed a high block after

epidural dosing which necessitated conversion to GA;[21] and one patient most likely had accidental intrathecal catheter placement.[12]

The spread of local anaesthetic has been reported to be unreliable. In one case, spinal anaesthesia with bupivacaine achieved adequate sensory block height in a parturient with achondroplasia requiring a caesarean delivery but the block receded rapidly intraoperatively, necessitating sedation and supplemental analgesia.[14] A separate article highlighted two restricted-growth patients who required conversion to GA as a result of an inadequate epidural sensory block.[12]

We considered a titratable technique to achieve central neuraxial block to be the best option for our patient and a two-space CSE technique was used. The epidural component was sited first so that it could be used to achieve anaesthesia if the subarachnoid space proved inaccessible. Appropriate dosing for spinal anaesthesia is difficult to calculate in these cases. Danelli et al.[15] performed a prospective, blinded study to determine the minimum effective dose of hyperbaric bupivacaine required for caesarean delivery but the study did not include patients with restricted growth. These investigators derived a formula of 0.06 mg of bupivacaine per cm height to be the minimum effective dose in 95% of parturients. Sarma et al.[16] used this formula to good effect to achieve satisfactory spinal anaesthesia in a patient with achondroplasia who required an emergency caesarean delivery.

We found 10 published case descriptions of parturients with restricted growth having a caesarean delivery under an adequate spinal anaesthetic.[12,16–21] In all these cases bupivacaine was used at an average dose of 0.064 mg per cm height. In our patient we used 7.5 mg of bupivacaine which represents 0.064 mg per cm height.

Restricted growth has varied heterogenicity and each parturient should be assessed on an individual basis when planning for delivery. We cannot conclude that this provides a one-size-fits-all in this situation. However, the formula discussed may offer a starting point in determining a bupivacaine dose for spinal anaesthesia in such patients, particularly when time does not allow for a slowly-titrated central neuraxial block or when there are concerns about airway management.

Operative delivery was considered the most likely outcome had our patient laboured naturally and if an emergency caesarean delivery had then been required, the timely securing of the airway or provision of central neuraxial block was likely to have been difficult and potentially detrimental to a positive outcome. Early communication and advanced planning for an elective surgical delivery in this case led to a good outcome.

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