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CASE REPORT

Achondroplasia: anaesthetic challenges for caesarean section

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ABSTRACT
Pregnancy in women with achondroplasia presents major challenges for anaesthetists and obstetricians. We report the case of a woman with achondroplasia who underwent general anaesthesia for an elective caesarean section. She was 99 cm in height and her condition was further complicated by severe kyphoscoliosis and previous back surgery. She was reviewed in the first trimester at the anaesthetic high-risk clinic. A multidisciplinary team was convened to plan her peripartum care. Because of increasing dyspnoea caesarean section was performed at 32 weeks of gestation. She received a general anaesthetic using a modified rapid-sequence technique with remifentanil and rocuronium. The intraoperative period was complicated by desaturation and high airway pressures. The woman's postoperative care was complicated by respiratory compromise requiring high dependency care.

Keywords: Achondroplasia; Caesarean section; General anaesthesia; Remifentanil; Rocuronium

Introduction

Dwarfism is defined as a failure to attain a height of 148 cm in adulthood.[1] It arises as a result of more than 200 medical conditions whose origins can be genetic, constitutional or metabolic. The most common type of dwarfism is achondroplasia occurring in 0.5–1.5 per 10 000 live births,[2] accounting for approximately 70% of cases of dwarfism. The typical appearances of achondroplasia include disproportionate dwarfism plus several craniofacial, central nervous system, spinal, respiratory and cardiac anomalies. Of adults with achondroplasia, 10–15% have a fixed, angular thoracolumbar junction kyphosis of sufficient severity to be of neurological consequence and likely to impact on respiratory and cardiac function.[3] These characteristic features can lead to increased difficulty with airway management during general anaesthesia but also make neuraxial anaesthesia potentially hazardous. When coupled with anaesthetic risks encountered during the third trimester of pregnancy such as aspiration of stomach contents, decreased cardiorespiratory reserve and supine hypotension, anaesthetic management of the gravid achondroplastic patient with marked kyphoscoliosis for caesarean section poses significant challenges and requires meticulous planning involving a multidisciplinary team (MDT).

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Case report

A 20-year-old nulliparous achondroplastic woman measuring 99 cm in height was referred to the high-risk anaesthetic clinic at 11 weeks of gestation. Her history was significant for severe thoracolumbar kyphoscoliosis and extensive spinal surgery on two previous occasions. This included anterior spinal strut surgery at four years of age and further spinal fusion with vascular rib grafting at the age of 15. Postoperative recovery was complicated by respiratory failure secondary to right lower lobe pneumonia and required admission to the intensive care unit for prolonged ventilation via a tracheostomy.

At the anaesthetic clinic she was noted to be dyspnoeic on mild activity and had significant supine hypotension. She weighed 36 kg with a body mass index (BMI) of 36 kg/m^2. Examination of the chest showed normal heart sounds and good bilateral air entry. On airway assessment she had a Mallampati 2 score, with a thyromental distance of >6.5 cm, normal jaw protrusion, and a full range of neck movement. Review of previous anaesthetic charts revealed a Cormack and Lehane grade 1 view at laryngoscopy and easy bag-valve-mask ventilation and intubation with a 6.5 mm reinforced tracheal tube. Despite the extensive scarring over her entire back, the spinous processes were palpable from L2 to L5. No anatomical or functional abnormalities of the upper airway were noted on nasoendoscopy performed by an otolaryngologist. Lung function tests revealed decreased lung volumes with a vital capacity of 0.83 L without airflow obstruction.

Baseline blood gases were normal. An overnight sleep study excluded sleep apnoea with no oxygen saturation abnormality observed. Cardiac assessment demonstrated baseline sinus tachycardia of 120 beats/min with a normal echocardiogram. Magnetic resonance imaging and computerised tomography scans performed before pregnancy showed a normal craniocervical junction, severe kyphosis from T9 to L1 measuring 140 degrees and a low lying conus medullaris terminating at the mid L3 level.

An MDT was convened which included anaesthetists, obstetricians, midwives and neonatologists. Following discussion with the patient and the MDT, it was decided to perform a caesarean section at 30–36 weeks of gestation due to anticipated cephalo-pelvic disproportion.[1] General anaesthesia was considered the safest option given her extensive spinal surgery, respiratory compromise and exaggerated supine hypotension. This also reflected the woman's preference. An anaesthetic management plan including calculated drug dosages, fluid management and ventilation strategies was documented and circulated.

A family history of deep vein thrombosis (DVT) warranted a thrombophilia screen. Despite negative results, but in consideration of the woman's immobility due to dysponoea, it was decided to begin enoxaparin 20 mg daily at 20 weeks as prophylaxis against DVT. This was continued uninterruptedly six weeks into the postpartum period and then stopped.

Due to worsening respiratory compromise exacerbated by severe anxiety, a scheduled caesarean section was performed under general anaesthesia at $32 + 4$ weeks of gestation. The woman received preoperative steroids to promote fetal lung maturity, and antacid prophylaxis. An experienced anaesthetic, obstetric and neonatal team were in attendance. Intravenous access was achieved and non-invasive blood pressure monitoring applied with an appropriately sized paediatric cuff. Baseline blood pressure was 115/45 mmHg, heart rate 98 beats/min and oxygen saturations 99% on air. The woman was positioned in the sitting position and pre-oxygenated with the application of continuous positive airway pressure (CPAP) of 3 cmH_2O for 5 min while a target-controlled infusion (TCI) remifentanil was commenced at 1 ng/mL to aid sedation and control tachycardia. General anaesthesia was induced using a modified rapid-sequence induction with thiopental 275 mg and rocuronium 40 mg. Cricoid pressure was applied. Laryngoscopy revealed a Cormack and Lehane grade 2 view and tracheal intubation was achieved with a 6.5 mm tracheal tube using a stylet. Despite preoxygenation and rapid intubation, oxygen saturations dropped to 65%, but recovered to 99% within 1 min of ventilation. The tracheal tube was secured at 18 cm at the incisors and ventilation with 100% oxygen was commenced. Mean airway pressures were noted to be 30 cmH_2O with tidal volumes of 200 mL. The tracheal tube was pulled back to 15 cm at the incisors. This reduced the airway pressures to 22 cmH_2O. Pressure controlled ventilation of 22 cmH_2O with positive end expiratory pressure (PEEP) of 4 cm delivered a tidal volume of 230 and 280 mL pre- and post-delivery, respectively. The insertion of a right radial arterial line and second intravenous cannula were delayed until post induction due to severe maternal anxiety. The patient was positioned carefully with left lateral tilt and additional support to compensate for spinal deformities. Anaesthesia was maintained using a combination of a TCI remifentanil at 1–3 ng/mL and sevoflurane in an oxygen and air mixture. An infusion of Hartmann's solution was started.

Surgery was commenced through a Pfannenstiel incision 10 min after induction and 4 min later a live male baby weighing 1940 g was delivered: Apgar scores were 4, 7 and 9 at 1, 5 and 10 min, respectively. The umbilical venous blood showed a pH 7.06, pCO_2 12.3 kPa and a base excess of −6.2 mmol/L. The baby was transferred to the neonatal intensive care unit and later required nasal continuous positive airway pressure (CPAP) for several hours. Maternal antimicrobial prophylaxis was given in accordance with local protocol. A bolus dose of oxytocin 2 U was followed by an infusion of 40 U oxytocin in 0.9% saline 500 mL over 8 h. Following delivery, intravenous morphine sulphate 7 mg and rectal diclofenac 50 mg were administered. Throughout the procedure the patient remained cardiovascularly stable. Estimated blood loss was 350 mL (9% of estimated blood volume). At the end of surgery, bilateral transversus abdominis plane (TAP) blocks were performed with 80 mg levobupivacaine in 40 mL volume using a landmark technique. Neuromuscular blockade was reversed with sugammadex 160 mg and the patient was successfully extubated in the sitting position and transferred to the high dependency unit (HDU) on the labour ward for postoperative care. She was prescribed regular oral paracetamol and diclofenac and intravenous morphine via a patient-controlled analgesia device. Pain scores postoperatively were low throughout and only 3 mg of morphine were used within the first 15 h following surgery.

The woman was initially stable in the maternity HDU with a pulse of 115 beats/min, blood pressure of 114/73 mmHg and oxygen saturation of 98% on 4 L/min of oxygen via a Hudson face mask. Postoperative haemoglobin was 10.1 g/dL. However, over the course of the day she struggled to expectorate and chest physiotherapy was initiated. Despite this, she became increasingly tachypnoeic and tachycardic and was transferred to the intensive care unit 15 h postoperatively. Widespread bilateral crepitations were auscultated but a chest X-ray was clear and after a short period of observation she was transferred back to the maternity HDU the following morning. The severity of her symptoms did not warrant further investigation and her transient

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deterioration was thought to be caused by sputum retention and basal atelectasis with tachycardia and tachypnoea exasperated by the patient's anxiety.

After 48 h, the patient was transferred to the postnatal ward. Her observations continued to be stable and she was discharged home on the eighth day postpartum. The baby was discharged with the mother and has not displayed any evidence of achondroplasia.

Discussion

Achondroplasia is the most common type of dwarfism of which 90% of cases are due to a sporadic mutation causing abnormal endochondral ossification.[4] When achondroplasia is inherited, as it was in our case, it is an autosomal dominant trait of the altered FGFR3 gene which codes for a constitutively active fibroblast growth receptor.

Antenatal anaesthetic assessment is essential in women with achondroplasia due to the number of comorbidities that are encountered with dwarfism. Planning should be for caesarean section delivery as this is inevitable due to congenitally small and contracted pelvis.[5,6] In addition to short stature and shortened limbs, those with achondroplasia commonly have significant craniofacial, spinal, skeletal abnormalities with central nervous system, respiratory and cardiac problems. These can all have an impact on the choice and management of anaesthesia (Table 1). Due to the particularly small stature and marked thoracolumbar kyphoscoliosis our patient posed an increased risk of respiratory embarrassment during pregnancy and delivery. This became evident when she was admitted to hospital in early third trimester with worsening respiratory compromise.

The achondroplastic thorax differs from normal and an expansion of its circumference during pregnancy is limited. Additionally, the uterus is an abdominal organ secondary to a small pelvis and with pregnancy this will impact on the small thoracic cage much earlier compared to individuals of normal stature. Functional residual capacity is much reduced and closing capacity encroaches on normal tidal breaths. The existing kyphoscoliosis displayed by our patient reduced lung volumes even further, exhibited by a lung capacity of <1 L early in pregnancy. These features contributed to increasing respiratory distress experienced during third trimester of gestation and the need for early delivery.

The mode of anaesthesia for caesarean section in achondroplastic patients is controversial and has to be decided on an individual basis.[7,8] Previous extensive corrective spinal surgery in this parturient and existent thoracolumbar kyphoscoliosis suggested neuraxial anaesthesia would be technically difficult with risk of neurological complications. In addition, significant supine hypotension and respiratory compromise manifested before delivery. Both phenomena would have been exacerbated by neuraxial anaesthesia. Additionally, anxiety presented as a major issue. Conversely, previous airway management had been straightforward and radiological findings did not suggest risk of cervical spine damage with neck manipulation. Therefore, we preferred a general anaesthetic technique.

The patient was pre-oxygenated in a sitting position and only placed supine after anaesthesia was induced. Nevertheless, there was immediate marked desaturation which could have been avoided by bag-mask ventilation or by intubation in a more upright position. However, intragastric gas insufflation could increase the risk of aspiration and intubation in a sitting position would have been more difficult. The tracheal tube had to be pulled back after high airway pressures were noted initially after intubation. This was partially due to a communication error as tube length at lips was documented as 19 cm from previous anaesthetic charts but in fact had been only 16 cm. This highlights the need for accurate documentation to avoid miscommunication between the teams.

Drug doses were calculated according to the woman's actual body weight of 40 kg as suggested by literature.[9] However, in a recently published case report describing a rapid-sequence induction in a slim achondroplastic patient, intubation doses of thiopental and rocuronium calculated according to body weight needed to be repeated twice before satisfactory anaesthesia was achieved.[10] This suggests that in slim achondroplastic patients, drug doses according to the patients' body weight might cause under-dosing. Our patient's BMI was 41 kg/m^2 at delivery and drug dosages according to actual body weight were successful. Remifentanil TCI was started at 1 ng/mL for sedation and increased to 3 ng/mL, followed by predetermined doses of thiopental 7 mg/kg and rocuronium 1 mg/kg. Anaesthesia and apnoea were achieved promptly and good intubation conditions were found after 45 sec.

We used remifentanil both as a safe[11,12] and titratable way of providing satisfactory sedation and controlling heart rate, in view of anxiety and baseline tachycardia. Remifentanil with an induction agent can provide good intubating conditions alone and prevents the haemodynamic response to intubation. In the event of a failed intubation its effect on spontaneous ventilation is transient.[13] Interestingly there is no known association of tachycardia with achondroplasia but in our patient anxiety might have contributed to her sinus tachycardia.

A higher incidence of difficult intubation is expected in achondroplastic patients due to a large head and tongue, limited neck extension and cervical instability with foramen magnum stenosis. Contrary evidence from the non-pregnant achondroplastic population suggests that this is not the case.[14,15] The previous history of easy intubation and upper airway examination suggested that the risk of difficult intubation was low. However, we

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Table 1 Features of the gravid achondroplastic and anaesthetic implications

Clinical features	Anaesthetic implications
Craniofacial	Airway
Large head	Narrow nasal passages and nasopharynx
Saddle nose	Small trachea
Large tongue	Limited neck extension
Forehead protrusion	Visualisation of larynx usually uncomplicated but consideration of small tracheal tube
Short maxilla	
Large mandible	
Atlanto-axial instability	
Stature	Miscellaneous
Rhizomelic short stature	Appropriately sized equipment (e.g. BP cuff)
Height <148 cm	Weight-related drug doses including fluids
	Care with positioning
Deformities	Neuraxial block
Lumbar hyperlordosis	Narrow epidural space
Rib deformities	Spinal stenosis
Thoracolumbar kyphoskoliosis	Difficult spinal and epidural anaesthesia with unpredictable spread of local anaesthetic
Respiratory	Respiratory
Small narrow chest cage	Restrictive lung disease
Thoracolumbar kyphoskoliosis	Severe reduction of functional residual capacity
Reduced symphysis pubis to xiphoid distance	Central and obstructive sleep apnoea
Cardiac	Cardiac
Cor pulmonale and pulmonary hypertension from restrictive lung disease and apnoea	Avoid hypoxia, hypercarbia, acidosis, hypothermia, nitrous oxide, stress
Obstetrics	Obstetrics
Small contracted pelvis leading to cephalopelvic disproportion	Need for caesarean section
Failure of head to engage into pelvis	
Neurology	Neurology
Foramen magnum stenosis	Cervicomedullary compression
Hydrocephalus	Elevated intracranial pressure; Avoid hyperextension of neck

had to take into consideration the airway changes associated with pregnancy and the increased risk of aspiration. In view of this, we felt it prudent to err on the side of caution and used rocuronium for intubation. This non-depolarising muscle relaxant has a reliable and predictable onset of action within 75 s with minimal cardiovascular effects and in the event of a failed intubation it is readily reversible with sugammadex.[16,17] However, there is no restriction for the use of suxamethonium in achondroplasia unless spinal cord compression with peripheral denervation is present.

After delivery only 2 U of oxytocin was given as slow bolus to minimise cardiovascular side effects; recent dose-finding studies suggest that as little as 0.5–3 U are effective at producing adequate uterine tone after caesarean delivery.[18,19] In addition, 40 U were given over 8 h to prevent delayed uterine atony and postpartum haemorrhage. Intraoperatively Hartmann's solution 500 mL was given through a volumetric pump to replace the estimated blood loss of 350 mL and insensible losses. Postoperatively intravenous fluids were limited to the oxytocin infusion to avoid fluid overload. In addition, a 100-mL bolus of succinylated gelatin (Gelofusine®) was administered to maintain adequate urinary output. In the event of haemorrhage the plan was to restrict resuscitation with crystalloid to one litre and start transfusion with blood products from blood loss of 750 mL, equivalent to 20% of her estimated blood volume.

TAP blocks have been shown to decrease postoperative morphine consumption after caesarean section.[20] We felt this would aid postoperative analgesia and potentially reduce the risk of respiratory complications. We used the landmark technique described by McDonnell et al.[20] introducing local anaesthetics into the transversus abdominis plane via the triangle of Petit as the anaesthetist present was experienced in this technique. This was part of a multimodal analgesia regimen and was effective in keeping morphine requirements to a minimum. A landmark technique for TAP block was standard practice in our department at the time and was effective in our patient. Complications are rare but intraperitoneal placement of the needle tip can have serious consequences.[21] In order to limit risk

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and maximise efficacy with this technique it is advisable to use ultrasound-guidance whenever possible.

The newborn showed a respiratory acidosis with an umbilical venous pH of 7.06. PCO_2 in the umbilical vein is in linear relationship with the mother's $PaCO_2$ and therefore an elevation can be explained by a degree of hypoventilation of the mother during general anaesthesia. Tidal volumes were as small as 230 mL before delivery to minimise maternal barotrauma. In addition, aortocaval compression causing hypoperfusion of the uteroplacental unit most likely contributed to the acidosis. Despite generous left tilt, the disproportion between uterus and abdominal space made a complete negation of vessel compression impossible. The transient hypoxia in the mother following induction is unlikely to have had a significant effect. Nevertheless, the acidosis was mild with a base excess of only −6.2 mmol/L and the baby's neurological development was normal in the immediate postnatal period.

In summary, we present successful management of elective caesarean section in a very short statured woman with achondroplasia with marked thoracolumbar kyphoscoliosis, and a discussion of the risks and challenges presented by such a patient.

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