International Journal of Obstetric Anesthesia (2014) xxx, xxx–xxx
0959-289X/$ - see front matter
http://dx.doi.org/10.1016/j.ijoa.2014.02.002

ELSEVIER
www.obstetanesthesia.com

CASE REPORT

Achondroplasia: anaesthetic challenges for caesarean section

L. Dubiel,[a] G.A. Scott,[a] R. Agaram,[a] E. McGrady,[a] A. Duncan,[b] K.N. Litchfield[a]
[a] *Department of Anaesthesia, Glasgow Royal Infirmary, Glasgow, UK*
[b] *Department of Obstetrics and Gynaecology, Princess Royal Maternity Hospital, Glasgow, UK*

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).

Accepted February 2014
Correspondence to: L. Dubiel, Anaesthetic Department, Ninewells Hospital, Dundee DD19 SY, UK.
E-mail address: l.dubiel@nhs.net

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.

Please cite this article in press as: Dubiel L et al. Achondroplasia: anaesthetic challenges for caesarean section. *Int J Obstet Anesth* (2014), http://dx.doi.org/10.1016/j.ijoa.2014.02.002

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

Please cite this article in press as: Dubiel L et al. Achondroplasia: anaesthetic challenges for caesarean section. *Int J Obstet Anesth* (2014), http://dx.doi.org/10.1016/j.ijoa.2014.02.002