Epidural anaesthesia for Caesarean section in an achondroplastic dwarf

M. J. MORROW AND I. H. BLACK

Summary
We describe the anaesthetic management of a parturient with achondroplasia presenting for Caesarean section under epidural anaesthesia. A block extending from T4 to S4 was established over 25 min using a total of 12 ml of 2.0% lidocaine (lignocaine) with epinephrine (adrenaline) 1:200 000 and fentanyl 37.5 μg. Apart from mild discomfort during peritoneal incision, her perioperative course was uneventful. Achondroplasia is reviewed and the anaesthetic implications of the condition are discussed. (Br. J. Anaesth. 1998; 81: 619–621).

Keywords: anaesthesia, obstetric; anaesthetic techniques, epidural; genetic factors; complications, achondroplasia

Dwarfism is defined as failure to achieve a height of 148 cm by adulthood. More than 100 different types of dwarfism have been described, but the commonest form of this relatively rare condition is achondroplastic dwarfism. The characteristic spinal and craniofacial abnormalities of achondroplasia occur as a consequence of normal periosteal bone deposition coupled with failure of endochondral bone formation. The usual appearance includes a large head, saddle nose, short limbs and a relatively normally proportioned trunk, often associated with marked kyphoscoliosis and lumbar lordosis. Limited neck extension, foramen magnum stenosis, a large tongue, large mandible and atlanto-axial instability are commonly encountered, making general anaesthesia potentially hazardous.

An exaggerated lumbar lordosis, thoracic kyphoscoliosis, generalized spinal stenosis and the unpredictable behaviour of local anaesthetic drugs in the epidural and subarachnoid spaces have, however, led to a reluctance among anaesthetists to consider regional anaesthesia in this group of patients. When combined with the well documented anaesthetic risks encountered during the third trimester of pregnancy of acid aspiration, decreased cardiorespiratory reserve and supine hypotension, the management of the gravid achondroplastic dwarf for Caesarean section presents a significant challenge for the anaesthetist, whether general or regional anaesthesia is chosen.

Case report
A 28-yr-old primagravid achondroplastic dwarf presented for elective Caesarean section at 37 weeks’ gestation. Her only previous general anaesthetic (as a child for tonsillectomy) had apparently been uneventful, but no notes relating to this procedure were available. Her pregnancy had been uneventful and ultrasonic measurements in utero suggested that the fetus was normally developed.

Physical examination revealed a 64-kg, 120-cm healthy female with the typical features of achondroplasia: large head, short limbs and marked kyphoscoliosis of the thoracolumbar spine. She had a short neck, large tongue and full set of teeth; the lower lumbar vertebrae were palpable. Neck extension was limited and it was possible to see only the uvula when the tongue was fully protruded. Examination of the cardiorespiratory system revealed no problems and there were no neurological abnormalities. Preoperative haemoglobin and electrolyte concentrations were within normal limits.

The patient expressed a desire to be awake for birth. Therefore, after discussion of the potential risks and benefits of general compared with regional anaesthesia for Caesarean section, it was decided to perform the operation under epidural anaesthesia. The patient was made aware that a general anaesthetic may be required in the event of failed epidural block.

Premedication comprised ranitidine 150 mg orally on the evening before surgery and again on the morning of the operation. On arrival in the operating room, 30 ml of sodium citrate 0.3 mol litre⁻¹ was administered orally, and invasive radial arterial pressure monitoring established under local anaesthesia. Other monitoring included ECG and pulse oximetry. A 16-gauge i.v. cannula was inserted and 750 ml of compound sodium lactate solution was administered before siting the epidural catheter. Facilities for emergency and fibreoptic intubation were available.

The patient was placed in the sitting position and the epidural space located at the L3–4 interspace using a 16-gauge Tuohy needle and the loss of resistance to saline technique. The needle was inserted at an angle of approximately 40° to the skin in a cephalad direction and directed towards the predicted midline. The epidural space was located with minimal difficulty at a depth of 4.5 cm, and 2.5 cm of catheter were threaded. A continuous flow of blood was obtained on aspiration of the catheter and therefore it was withdrawn. The procedure was repeated in the same interspace but with the needle realigned slightly further towards the anticipated midline. Again the space was located at 4.5 cm, and 2.0 cm of catheter were advanced, this time uneventfully. With the patient remaining in the sitting position, a test dose of 1.5 ml of 2% lidocaine (lignocaine) with...
epinephrine (adrenaline) 1:200 000 was administered without evidence of a subarachnoid block developing after 5 min. Another 6 ml of the same solution was administered in the sitting position in two increments at 5-min intervals. The patient was then placed in the right lateral position and another 3 ml of solution administered. This was followed 5 min later by another 3 ml in the left lateral position. At this stage fentanyl 37.5 μg was administered via the epidural catheter. The patient was then placed in the modified supine position with 15° of left lateral tilt and the extent of the block assessed. After 25 min a sensory block to temperature and fine touch extended from S4 to T4 on the left side and from S4 to T3 on the right. A total of 15 mg of ephedrine i.v. were administered during establishment of the epidural block in response to an anticipated decrease in arterial pressure.

Caesarean section was performed using a midline incision and good muscle relaxation was observed throughout the procedure. The patient experienced a brief episode of discomfort during peritoneal traction which was relieved rapidly by administration of a 50:50 mixture of nitrous oxide in oxygen. A healthy male infant weighing 3545 g was delivered with an Apgar score of 8 at 1 min and 9 at 5 min. No features of achondroplasia were evident. Syntocinon 10 u. i.v. were administered after cord clamping and a further 20 u. added to an i.v. infusion of compound sodium lactate solution 1000 ml.

Maternal heart rate and arterial pressure were well maintained throughout the procedure. Blood loss at the end of operation was estimated at 300 ml. The patient’s partner was present throughout. On completion of the procedure the patient was transferred to the high dependency area, where monitoring was continued until the block had worn off. Analgesia was provided by a suppository of diclofenac 100 mg inserted at the end of the procedure and a patient-controlled analgesia system set to deliver morphine 1-mg boluses at 5-min intervals. Full recovery of neuromuscular function had occurred within 8 h and no complications were reported. Mother and baby were discharged from hospital on day 7 without further incident.

Discussion

Achondroplasia is the commonest form of dwarfism, with an incidence of 0.5–1.5 per 10 000 live births. The condition is transmitted in an autosomal dominant fashion, but only 20% of cases are familial, the remaining 80% representing a spontaneous genetic mutation. Females are affected more frequently than males. Achondroplastic dwarfs characteristically have low fertility rates; however, those that do conceive often require delivery by Caesarean section because the normal-sized fetal head and smaller than normal maternal pelvic diameter results in cephalopelvic disproportion during the later stages of pregnancy.

Anaesthesia, whether general or regional, presents many potential problems during the latter stages of pregnancy. Maintenance of a patent upper airway may be difficult because of the presence of a large tongue and large mandible, usually associated with a full set of teeth. Tracheal intubation may prove difficult as foramen magnum stenosis and cervical spine instability are common, and hyperextension of the neck must be avoided to prevent cervical cord compression. Limited neck extension has been reported as a cause of difficult intubation in two cases, although not all authors have encountered similar difficulties. Mayhew and colleagues reported no difficulty in airway management or direct laryngoscopy in a series of 27 patients undergoing 36 anaesthetic procedures. If tracheal intubation is required in this group of patients, a small tracheal tube should be selected, the most appropriate size being predicted according to weight rather than age.

Cardiorespiratory function may be impaired by several factors specific to achondroplasia and other less common forms of dwarfism. Thoracic kyphoscoliosis, upper airway obstruction, rib deformities and recurrent respiratory tract infections are common. Failure of the normal-sized fetal head to engage in the narrow pelvic inlet results in the uterus remaining an entirely intra-abdominal organ during the latter stages of pregnancy. The subsequent diaphragmatic splinting causes a greater than usual pregnancy-induced reduction in functional residual capacity. By the 16th week, these patients appear to be in the 30th week of pregnancy. Cor pulmonale is a relatively rare occurrence but has been reported as a consequence of these multiple respiratory complications.

Epidural anaesthesia may be technically difficult to perform because of the significant anatomical abnormalities often associated with achondroplasia. Marked lumbar lordosis and thoracic kyphoscoliosis may make identification of bony landmarks difficult. Reduced interpedicular distance, shortened pedicles, osteophyte formation and prolapsed intervertebral discs combined with a narrow epidural space may make identification of the space difficult and increase the risk of dural puncture. The presence of a narrow epidural space may also, as in this case, make insertion of the catheter difficult. Engorged epidural veins increase the risk of venous puncture either by the Tuohy needle or the catheter, and result in an unpredictable spread of local anaesthetic within the space. Spinal stenosis may impair free flow of CSF and make identification of dural puncture more difficult. Aorto-caval compression may be particularly severe during regional anaesthesia as a result of the intra-abdominal position of the uterus. Left uterine displacement, vigorous fluid preloading and vasopressor support have been advocated. It has been suggested that regional anaesthesia should be avoided in patients with achondroplasia because any subsequent neurological abnormalities caused by spinal deformities might be attributed to the anaesthetic technique. But no neurological complications in patients with achondroplasia have yet been reported in those cases performed under regional anaesthesia. As there were no pre-existing neurological problems in our patient, it was felt that there were no specific contraindications to the use of epidural anaesthesia. In common with previous experiences, no neurological sequelae developed.

There are no guidelines on the use of epidural or spinal local anaesthetics in the gravid achondroplastic dwarf. Because of the large inter-individual variation in spinal column anatomy in these patients, it is
unlikely that clear recommendations could be developed. What is clear, however, is that great care must be taken when establishing regional block in these situations, regardless of the local anaesthetic chosen or the technique used.

The most appropriate type and volume of epidural test dose is unclear. Plain 0.5% bupivacaine in doses of 2.0 ml and 3.0 ml have been reported while 1.0 ml of carbonated lidocaine with epinephrine 1:200 000 has also been described. Crawford and Dutton reported the use of a 32-gauge microspinal catheter to allow the establishment of continuous spinal anaesthesia in an achondroplastic dwarf, but these catheters have been withdrawn because of concerns about the incidence of neurological deficits associated with their use. Infusion of 0.5 ml of 0.5% hyperbaric plain bupivacaine intrathecally resulted in a bilateral block to T6 associated with significant hypotension developing within 20 min. Therefore, it would seem prudent to reduce the volume of the test dose in this group of patients; consequently, we chose to use 1.5 ml of lidocaine with epinephrine 1:200 000.

The volume of solution needed to produce surgical block is also unclear. Brimacombe and Caunt reported the development of an epidural block extending from C5 to S4 over a 20-min period using 0.5% plain bupivacaine 12 ml, while Wardall and Frame reported that only 5 ml of 0.5% plain bupivacaine was sufficient to develop a block to T4. Cohen reported adequate block with a total of 9 ml of 3% 2-chloroprocaine, supplemented with another 9 ml after 35 min. Carstoniu, Yee and Halpern achieved adequate block for Caesarean section using 8 ml of 2.0% carbonated lidocaine with epinephrine 1:200 000. The use of epidural fentanyl to supplement surgical block in this group of patients has not been reported previously.

Anaesthesia poses a significant risk to patients with achondroplasia and this must be recognized by all staff involved in the perioperative care of these patients. Early communication with the anaesthetic team is important and a thorough preoperative assessment is mandatory. Facilities for managing the difficult airway and suitably experienced personnel should be available and the use of intra-arterial pressure monitoring should be considered in patients with co-existing cardiorespiratory problems. If opioid supplementation of the block is used, frequent monitoring of vital signs and continuous pulse oximetry should be considered during the postoperative period. If these factors are borne in mind and the procedure is undertaken with adequate expertise and support facilities then the use of epidural anaesthesia is a satisfactory technique for Caesarean section in the gravid achondroplastic dwarf.

References