

Anaesthesia for Caesarean Section in an Achondroplastic Dwarf

Rahul Garg* and Pamela Angle

Division of Obstetrical Anaesthesia, Sunnybrook Health Sciences Centre, University of Toronto, 2075 Bayview Avenue, Toronto, Ontario, Canada

Abstract

Achondroplasia is an inherited skeletal disorder affecting bone growth and development. It presents the most common cause of dwarfism in humans. Associated anatomical and physiological changes present a major challenge to management of both general and regional anaesthesia. This case report describes our anaesthetic management of elective caesarean delivery in an achondroplastic dwarf. In this challenging patient group, we discuss the risks and benefits of different anaesthetic approaches, the safe use of epidural anaesthesia with ultrasound guidance, sacral sparing following successful epidural catheter insertion, and epidural opioids for post-operative analgesia. This is followed by a review of the literature.

Keywords: Achondroplasia; Anaesthesia; Caesarean Section; Epidural; Ultrasound

***Corresponding Author:** Rahul Garg, Division of Obstetrical Anaesthesia, Sunnybrook Health Sciences Centre, University of Toronto, 2075 Bayview Avenue, Toronto, Ontario, Canada; Tel: +1-416-480-6100; E-mail: rahul.garg@doctor.com, Pamela.Angle@sunnybrook.ca

Introduction

Achondroplasia is an inherited skeletal disorder with an incidence of approximately 1 in 30000. It is the most common cause of dwarfism in humans. The underlying pathological process is a decrease in endochondral bone formation but

normal periosteal bone formation, leading to the characteristic skeletal deformities. The disease is inherited in an autosomal dominant pattern; however the majority of cases are thought to arise due to spontaneous genetic mutations.

The anatomical and physiological changes associated with achondroplasia present a major challenge for both general and regional anaesthesia. Patients have short limbs which may increase the difficulty of venous access and non-invasive blood pressure monitoring. Airway management can be challenging, and careful pre-operative assessment is required. Achondroplastic patients may have small maxillas, large mandibles, and large tongues, making direct laryngoscopy difficult. In addition there can be cervical instability and stenosis at the level of the foramen magnum, necessitating care with neck movement. Vertebral and costal abnormalities can be associated with restrictive lung disease, and obstructive sleep apnoea is also common in achondroplastics. The anaesthetist must be aware of the possibility of associated pulmonary hypertension.

Regional anaesthesia, particularly neuraxial techniques, may be challenging due to thoracic kyphoscoliosis, exaggerated lumbar lordosis, and formation of vertebral osteophytes. Spinal stenosis with associated neurological symptoms can be present, increasing the risks associated with neuraxial procedures. The epidural and subarachnoid spaces tend to be reduced in size, leading to increased risk of dural puncture during epidural techniques and difficulties with catheter placement. Calculating appropriate doses for neuraxial anaesthesia and analgesia is

difficult due to short stature, canal stenosis, and inconsistent spread of solution through the epidural space.

Pregnancy can exaggerate many of the above anaesthetic concerns. The gravid uterus can further compromise respiratory compliance, and due to its relatively high abdominal position in dwarfs, cause significant aorto-caval compression. Airway oedema may make laryngoscopy more challenging. Engorgement of epidural veins makes epidural catheterisation more difficult and increases the risk of intravascular placement. Operative delivery is common in this group due to cephalo-pelvic disproportion.

This case report describes our anaesthetic management of elective caesarean delivery in an achondroplastic dwarf. In this challenging patient group, we discuss the risks and benefits of different anaesthetic approaches, the safe use of epidural anaesthesia with ultrasound guidance, sacral sparing following successful epidural catheter insertion, and epidural opioids for post-operative analgesia. This is followed by a review of the literature.

Case Report

A 32 year old achondroplastic primigravida at term gestation presented for elective caesarean delivery of an uncomplicated pregnancy. She had previously undergone assessment in our high risk obstetrical anaesthesia clinic. Her medical history was significant for intermittent pain and paraesthesiae in her lower limbs which was brought on by exertion, consistent with lumbar spinal stenosis. She had previously had a lower limb procedure in childhood under general anaesthetic which she reported as uneventful. She took no medications aside from prenatal vitamins, and reported no drug allergies. There were no other medical co-morbidities.

On physical examination, the patient was 130cm tall and 66kg in weight (BMI 39.1). Frontal bossing, large mandible, short limbs, thoracic kyphoscoliosis, and marked lumbar lordosis were noted. Airway examination revealed a Mallampati class III

airway with restriction of neck extension. Cardio-respiratory examination was unremarkable. Pre-operative full blood count showed a haemoglobin concentration of 119 g/L and a normal platelet count. There was no imaging of her spine available for review. Anaesthetic options were discussed with the patient who expressed a preference for a regional technique.

Following insertion of an 18 gauge intravenous cannula and administration of antibiotic prophylaxis with 2 grams intravenous cephazolin, infusion of Ringer's lactate fluid was commenced. Standard monitoring (pulse oximetry, 3-lead electrocardiography, and non-invasive blood pressure) was instituted. The patient was placed in the sitting position for epidural placement. Due to the spinal deformities and difficult anatomical landmarks, ultrasound of the lumbar spine was used to guide epidural placement (curvilinear probe 2-5MHz). Reasonable images were obtained of the L3-L4 interspace to guide placement (Figure 1).



Figure 1: Transverse view of L3-L4 interspace on ultrasound showing estimated depth of epidural space at approximately 5.5 to 6 cm.

After sterile preparation of the patient's back with 2% chlorhexidine in 70% alcohol and infiltration of the skin with 2% lignocaine, a 17G Tuohy needle was inserted at the L3-L4 interspace. A continuous loss of resistance to saline technique

was used to identify the epidural space which was located approximately 6cm from the skin. On insertion of the epidural catheter, the patient complained of a transient dysaesthesia on the left side, which resolved with retraction of the catheter to 10cm at the skin (4cm within epidural space). The patient was then placed supine with left lateral tilt.

Incremental aliquots (2-3ml) of 2% lignocaine with 1:200000 adrenaline were slowly administered after an initial 2ml test dose of the same solution was negative for signs of intrathecal injection. Sensory levels were followed between injections using ice. The patient complained intermittently of back pain radiating into the left leg during injection of the solution, which resolved when injection ceased. After a total of 8mL of 2% lignocaine given over 15-20 minutes, followed by an epidural dose of 50mcg fentanyl, a sensory block to ice was achieved to the level of T6 bilaterally.

At this point, the obstetric team attempted insertion of a urinary catheter, however the patient complained of pain during the insertion. Sensory testing with ice confirmed sacral sparing of

the block. We positioned the patient into a semi-sitting position and gave a further 4mL of 2% lignocaine over 10-15 minutes (total 12mL). Sacral anaesthesia was achieved, and after urinary catheterisation, the caesarean section was completed uneventfully. No vasopressor was required for hemodynamic support. A live male was delivered weighing 2748 grams, with Apgar scores of 9 and 9 at 1 and 5 minutes. The rest of the procedure was uneventful. Oxytocin 20 units in 1L Ringer's lactate was given after delivery. Estimated blood loss was 500mL. For post-operative analgesia 1.5mg epidural morphine was given prior to epidural catheter removal. The patient also received oral paracetamol 1g every 6 hours and diclofenac 50mg every 8 hours. No supplemental oral opiates were required post-operatively. She remained well and was discharged on day 2 post-operatively.

Discussion

Previous case reports have described various techniques for caesarean section in achondroplastic dwarfs. General, spinal, epidural, and combined spinal-epidural anaesthesia techniques have all been reported (Table 1).

Table 1 - Summary of recent case reports

Authors	Anaesthetic Technique	Issues/Important Points
Dubiel et al; [1]	General anaesthesia (previous spinal surgery, respiratory compromise, patient preference)	No difficulty with rapid sequence induction and standard direct laryngoscopy. Drug doses based on actual body weight. Post-operative respiratory distress due to sputum retention and atelectasis requiring brief HDU admission.
Wight, et al. [2]	Ultrasound guided combined spinal-epidural - 5.5mg bupivacaine + 300mcg diamorphine spinally plus 10mL 0.5% bupivacaine epidurally	Usefulness of ultrasound assistance for neuraxial technique. Safety and efficacy of CSE technique. Dysaesthesia on epidural insertion.
Osorio Rudas et al. [3]	Ultrasound guided combined spinal-epidural - 5mg bupivacaine + 64mcg morphine + 16mcg fentanyl spinally, plus 60mg lignocaine epidurally, plus remifentanyl 48mcg intravenously	Usefulness of ultrasound assistance for neuraxial technique. Need for intravenous analgesia to supplement neuraxial anaesthesia.
Cevik and Colakoglu [4]	General anaesthesia (patient preference)	No difficulty with standard asleep direct laryngoscopy and intubation in this patient.

Authors	Anaesthetic Technique	Issues/Important Points
Huang and Babins [5]	Failed attempt at epidural placement, followed by failed attempt at awake video laryngoscope guided intubation; successful awake fibre-optic intubation	Technical difficulty encountered with neuraxial technique. Efficacy of awake fibre-optic intubation if general anaesthesia chosen.
DeRenzo, et al. [6]	Spinal anaesthesia for emergency case – 10mg bupivacaine + 200mcg morphine; discomfort during procedure (after delivery) requiring 4mg midazolam + 200mcg fentanyl intravenously plus 200mg lignocaine infiltration	Safety of spinal anaesthesia for urgent caesarean section. Unpredictable dosing with single shot approach; inadequate anaesthesia for duration of caesarean section in this case. High dose spinal morphine used without adverse effects.
McGlothlen [7]	Spinal anaesthesia for elective case – 11mg bupivacaine + 25mcg fentanyl + 300mcg morphine	Safety and efficacy of standard dose spinal anaesthetic for elective case. Safety of high dose spinal morphine.
Morrow and Black [8]	Epidural anaesthesia – total 13.5mL 2% lignocaine with 1:200000 adrenaline + 37.5mcg fentanyl; supplemented with 50% nitrous oxide during peritoneal traction	Initial epidural venous cannulation requiring repeat insertion. Need for inhaled analgesia to supplement epidural anaesthesia.
Carstoniu, et al. [9]	Epidural anaesthesia – total 8mL 2% lignocaine with 1:200000 adrenaline + 50mcg fentanyl	Safety and efficacy of low dose epidural anaesthesia for elective case.

Weighing up the risks and benefits of the different anaesthetic options for our case, and also considering the patient's preference to remain awake, we opted for regional anaesthesia over general anaesthesia.

Of particular concern to us was the patient's lumbar canal stenosis. The performance of neuraxial blocks in patients with known spinal canal stenosis carries an increased risk for neurological injury [10], including cauda equina syndrome. The risk is thought to be related to a relatively higher increase in intra-spinal pressures with injection of local anaesthetic solutions, which may lead to ischaemia of nerve roots or the spinal cord. The ischaemic effects may be further enhanced by the potential neurotoxicity of local anaesthetic solutions [10]. Both spinal and epidural anaesthesia are associated with this increased risk of neurological injury. This risk was communicated to our patient. Given the patient's potentially difficult airway, and strong preference to be awake for the procedure, we agreed to a neuraxial anaesthetic approach.

We chose to use epidural anaesthesia which allowed a titrated block for the surgical procedure. We decided against the use of a combined spinal-epidural (CSE) technique in this elective setting as we felt the benefit of speed of block onset did not outweigh the potential increased risks associated with dural puncture and subarachnoid anaesthesia. CSE techniques have been safely and effectively used in previous case reports [2, 3] and would be our method of choice if there were an indication for expedited delivery. Although single shot spinal has been previously used [7], the unpredictable dosing and spread gives advantage to a titratable technique such as epidural or CSE.

We used a solution of 2% lignocaine with 1:200000 adrenaline and fentanyl to slowly achieve adequate epidural anaesthesia. This same solution has been previously used safely and effectively [8, 9]. Our total dose used was relatively small, consistent with the expected reduced requirement of a patient with small stature and reduced neuraxial volume.

Ultrasound provided a useful tool to assist our neuraxial technique in this case, and its successful use in this patient group has been previously reported [2, 3]. Ultrasound is readily available and non-invasive, and appears to be effective in assisting neuraxial anaesthesia in achondroplastics with difficult spinal anatomy. Ultrasound may help avoid the need for general anaesthesia and its associated risks in achondroplastic dwarfs.

The dysaesthesia and pain experienced by our patient during epidural catheter insertion and solution injection is consistent with her known lumbar canal stenosis and reduced neuraxial volume. Vigilance is required during catheter placement as complications are more common, and slow injection of epidural solution is recommended to avoid excessive increases in epidural space pressure.

Spread of local anaesthetic within the epidural space is known to be inconsistent in achondroplastics, and this was seen in our patient with marked initial sacral sparing of the block despite a higher sensory level of T6 bilaterally. We were able to achieve adequate sacral anaesthesia with further dosing and positioning. Again, use of a titratable neuraxial technique is beneficial to overcome this potential problem.

For post-operative analgesia we gave 1.5mg morphine epidurally, half our usual dose (3mg) for most healthy parturients undergoing elective caesarean section under epidural anaesthesia. Previous reports have used high intrathecal morphine doses safely [6, 7], however the appropriate epidural dose is not known. We made a 50% dose reduction given the patient's small stature and reduced neuraxial volume, and achieved excellent post-operative analgesia with no respiratory depression, pruritis, or nausea.

In summary, we have reported the successful use of ultrasound guided epidural anaesthesia for elective caesarean section in an achondroplastic dwarf. The increased risk of neurological injury in these patients who often have spinal canal stenosis must be considered if using a neuraxial approach, and balanced against

the potential risks of a general anaesthetic. We recommend the use of ultrasound to facilitate neuraxial anaesthesia in this patient group, and the use of titrated doses of local anaesthetic solution to achieve effective blockade. In our patient, epidural morphine used at half our usual dose provided safe and effective post-operative analgesia.

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