Clinical Reports

Epidural anaesthesia for Caesarean section in an achondroplastic dwarf

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This report describes the anaesthetic management of an 18-yr-old achondroplastic dwarf who presented for elective Caesarean section. Epidural anaesthesia was performed without technical difficulty using 8 ml carbonated lidocaine 2% with epinephrine 1:200,000. Although the skeletal abnormalities of achondroplasia have been cited as contraindications to the use of epidural anaesthesia, clinical experience does not support this contention. Previous reports have described technical difficulties in these patients, such as dural puncture and inability to advance the catheter into the epidural space, but no serious complications resulted and epidural anaesthesia was successful on subsequent attempts. The existing literature on the anaesthetic management of achondroplasia for Caesarean section is reviewed and considerations are presented concerning the choice of local anaesthetic, the epidural test dose, and dose titration.

Cette observation décrit la conduite anesthésique adoptée pour une césarienne chez une patiente de 18 ans atteinte de nanisme achondroplasique. Une anesthésie épidurale avec 8 ml de lidocaïne carbonatée 2% et adrénaline 1:200,000 est réalisée avec succès et sans difficulté. Les anomalies squelettiques associées à l'achondroplasie ont déjà été considérées parmi les contre-indications de l'anesthésie épidurale. Cependant, l'expérience clinique ne justifie pas cette prise de position. L'anesthésie épidurale chez les achondroplasiques est reconnue pour présenter des difficultés d'ordre technique comme la ponction de la dure-mère et l'impossibilité d'avancer le cathéter dans l'espace épidural. Toutefois des complications majeures associés à la technique n'ont jamais été rapportées et l'anesthésie épidurale est le plus souvent pratiquées avec succès. Cet article présente une revue de la littérature sur le sujet avec des considérations portant sur le choix, la dose-test et la posologie du produit utilisé pour l'épidurale.

Key words
ANAESTHESIA: obstetric, Caesarean section;
ANAESTHETIC TECHNIQUES: epidural;
COMPLICATIONS: achondroplasia.

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There are few detailed accounts of epidural anaesthesia in the achondroplastic dwarf. Some authors have cautioned against the use of epidural anaesthesia in these patients because their abnormal spinal anatomy may subject them to increased risks such as patchy blocks, dural puncture and spinal cord trauma.[1-3] This, combined with the lack of dosage guidelines, has led to the reluctance of clinicians to use regional blockade in cases of achondroplasia.

Although rare (incidence of approximately 4/100,000), achondroplasia is seen more frequently in women.[4] Full term pregnancies in these patients almost invariably lead to Caesarean section due to cephalo-pelvic disproportion and many will request regional anaesthesia. The advantages of regional anaesthesia for Caesarean section are well known and must be weighed against the risks of general anaesthesia in the patient who is both pregnant and achondroplastic.[5-7] This report describes the management of epidural anaesthesia in an achondroplastic dwarf.

Case report

An 18-yr-old, gravida 1, para 0 achondroplastic dwarf presented for elective Caesarean section at 38 wk gestation. Her past medical history was unremarkable except for an allergy to sulpha drugs. She had no neurological or musculoskeletal symptoms. In the latter part of her pregnancy, she had developed symptoms of mild reflux oesophagitis.

On physical examination, she had the typical appearance of an achondroplastic dwarf: a large head with prominent frontal bones and disproportionately short

limbs. Her height was 119 cm and she weighed 61 kg, compared with a pre-pregnancy weight of 48 kg. Her baseline blood pressure was 90/60 mmHg. Examination of her cardiovascular and respiratory systems was unremarkable. She had a marked lumbar lordosis and a moderate thoracic kyphosis without clinical evidence of scoliosis.

Preoperative CBC and urinalysis were within normal limits. Roentgenograms of her cervical spine showed no atlanto-axial instability on flexion and extension views. Her C_2 and C_3 cervical vertebrae appeared to be fused.

After discussing with the patient the potential risks of epidural versus general anaesthesia, it was decided to proceed with lumbar epidural anaesthesia. She received oral premedication consisting of ranitidine 150 mg, metoclopramide 10 mg and 0.3 M sodium citrate 30 ml.

Upon arrival in the operating room the patient received 750 ml of Ringer's lactate *iv* while she was monitored using pulse oximetry, continuous ECG and an automatic blood pressure cuff.

With the patient in the sitting position the epidural space was easily identified at about the T_{11}/T_{12} interspace using a 17-gauge Tuohy needle and loss of resistance to air. Identification of the exact vertebral level was difficult since her hyper-lordosis prevented accurate palpation of discrete lumbar interspaces. The point of insertion marked the lowest spinal interspace that was palpable.

An epidural catheter was easily inserted 5 cm into the epidural space. The patient was then positioned supine with left uterine displacement and given oxygen at 8 $L \cdot min^{-1}$ by mask. A test dose of 1 ml carbonated lidocaine 2% with epinephrine 1:200,000 was administered. As there was no evidence of spinal blockade after five min, a further 6 ml of the same solution was injected via the catheter in 2 ml increments at three min intervals. Just before skin incision, an additional 1 ml of local anaesthetic and 50 μg fentanyl were administered. A T_5 level was reached at the time of incision.

Approximately ten min after skin incision, a 2855 g achondroplastic female infant was delivered with Apgar scores of 8 and 9 at one and five min, respectively. On two occasions after delivery the patient's blood pressure decreased to 60/40 mmHg, eliciting complaints of mild dizziness, but it responded immediately to rapid infusion of Ringer's lactate and 5 mg ephedrine *iv*. Intraoperative blood loss was approximately 400 ml. The postoperative course was uneventful and both mother and baby were discharged six days later.

Discussion

Achondroplasia is characterized by abnormal endochondral bone formation. It is an autosomal dominant genetic condition, but 80% of cases result from a spontaneous mutation.[8] The increased risks of both general and regional anaesthesia resulting from the abnormalities of the musculoskeletal system were recently reviewed.[9] The shape of the head and mandible, the narrower foramen magnum, and the higher incidence of cervical spine instability could theoretically lead to increased difficulty with tracheal intubation.[1] More recent clinical experience, however, has demonstrated few difficulties with intubation in achondroplastics,[6,10] despite earlier warnings to the contrary.[5] Nevertheless, the potential for difficult intubation exists, especially in the presence of pregnancy-induced changes in the upper airway.

Similarly, major regional anaesthesia in achondroplastics could be complicated by the presence of one or more of the following: kyphosis, scoliosis, lumbar lordosis, spinal stenosis, osteophytes, short pedicles, or a small epidural space.[1,4,11] These could lead to difficulties in locating the epidural space, increase the risk of dural puncture, or limit the spread of local anaesthetic. Although the spinal abnormalities in achondroplasia may increase the technical difficulty of regional anaesthesia, one should note that these abnormalities are similar to the degenerative changes seen in the elderly, a population in which regional techniques are frequently used.

A computerized literature search revealed four case reports in the English literature describing epidural anaesthesia for Caesarean section in achondroplastic dwarfs since 1966 (Table).[11–14] Difficulty with catheter insertion was encountered in two cases. Also, in these two cases, one was further complicated by dural puncture and the other by cannulation of an epidural vein. Despite these technical complications, all four cases had successful epidural blocks. An additional report mentioned 12 women with achondroplasia who had Caesarean sections under regional anaesthesia, but this information was based solely on a questionnaire survey of 150 women with chondrodystrophies.[2] No details were provided, nor was it clear whether epidural or spinal anaesthesia was used. The available literature, including this report, supports the feasibility of a continuous epidural technique.

In the present case an epidural catheter was inserted easily although at a higher spinal level than usual because of the patient's pronounced lumbar lordosis. Attempting an epidural puncture above the lordotic lumbar spine may permit easier location and catheterization of the epidural space. A surgical level of anaesthesia for Caesarean section was reached with a very small epidural dose of local anaesthetic. With one exception,[12] the other case reports have also demonstrated a low dosage requirement in achondroplastic dwarfs. This underscores the need for careful epidural titration with small incremental doses of an agent with rapid onset, as

TABLE Epidural anaesthesia in achondroplastic dwarfs – summary of previous case reports

Author	*Patient ht. and wt.*	*Epid. level*	*Local anaesthetic*	*Volume*	*Sensory level*	*Complications*
Cohen[11] 1980	122 cm 57 kg	$L_{2,3}$	3% 2-chloroprocaine	9 ml (+ 9 ml 35 min later)	T_4	1 Epid. catheter could not be advanced on two occasions at $L_{3,4}$ 2 Inadvertent dural puncture at $L_{3,4}$
Waugaman[12] 1986	120 cm 48 kg	$L_{2,3}$	0.75% plain bupivacaine	21 ml	T_3-T_4	
Brimacombe[13] 1990	121 cm 73 kg	$L_{2,3}$ ($?L_{3,4}$)	0.5% plain bupivacaine	12 ml (+ total of 11 ml saline)	C_5	1 Difficult epid. catheter advancement followed by venous cannulation 2 Further difficulty threading epid. catheter on 2nd attempt at same level, but eventually successful
Wardall[14] 1990	111 cm 46 kg	$L_{2,3}$	0.5% plain bupivacaine	5 ml	T_4 on Lt T_6 on Rt	

well as accurate measurement of the sensory level after each dose.

In this patient, carbonated lidocaine was chosen because its fast onset would allow early detection of inadvertent subarachnoid injection. It also allowed rapid titration of dose to the desired level of blockade despite the small increments that were used. In one of the case reports, the slow onset of 0.5% bupivacaine may have contributed to excessive dosing and a very high epidural block.[13]

The optimal epidural test dose in the achondroplastic dwarf is uncertain. The volume of local anaesthetic should be large enough to allow early detection of inadvertent subarachnoid injection, but not so large that high spinal blockade results. The inclusion of epinephrine, and its dose, is also debatable. If, as in our case, one uses a small volume (i.e., 1 ml) of local anaesthetic containing the standard 1:200,000 concentration of epinephrine as a test dose, the small amount of epinephrine (i.e., 5 μg) may not produce signs of intravenous injection. Such uncertainty should not deter one from using epidural anaesthesia in these patients, but it should emphasize the need to observe for signs of catheter misplacement after every epidural injection.

No single test can rule out intravascular injection with absolute certainty. In our patient, the use of a generally accepted test dose of epinephrine (15 μg)[15] would have required preparation of a separate solution (either epinephrine alone in saline or combined with local anaesthetic) in addition to the solution prepared for epidural block. While this is certainly a feasible option, we chose instead to monitor carefully for systemic effects of local anaesthetic rather than rely on epinephrine to indicate intravascular injection.

Spinal anaesthesia is commonly used for Caesarean section, but we could find no detailed case reports of its use in the pregnant achondroplastic patient. Two articles contained only anecdotal references to cases of spinal anaesthesia in the achondroplast. Neither article revealed the type or dose of local anaesthetic that was used. Kalla *et al.*,[16] in their case report of an achondroplastic dwarf who had a Caesarean section under general anaesthesia, also mentioned the successful use of spinal anaesthesia in a non-obstetrical patient undergoing lower extremity surgery. Walts *et al.*[6] in a review of their past experience with anaesthesia for dwarfs, alluded to five cases of spinal anaesthesia in achondroplastic patients. They provided few details, except to say that there were no serious complications and that in the single case of Caesarean section under spinal anaesthesia, "a note on the record indicated that the lumbar puncture was performed with difficulty and the patient became hypotensive following the block." As is the case with epidural anaesthesia, the potential risks imposed by the spinal abnormalities and lack of clinical experience have probably deterred anaesthetists from using spinal anaesthesia. One would expect the narrower spinal canals and exaggerated spinal curvatures of these patients to result in unpredictable spread of a subarachnoid injection.

In summary, this report describes the successful use of epidural anaesthesia for Caesarean section in an achondroplastic dwarf. Although some authors have cautioned against the routine use of regional blockade, this is not supported by the recent clinical experience. Since there

are no dosage guidelines available for spinal anaesthesia in this population, this method should be used with caution. Achondroplastic patients, especially those undergoing Caesarean section, should not be denied the benefits of epidural anaesthesia in the absence of evidence that it is unreliable or unsafe. The risks of general versus epidural anaesthesia must be considered for each patient, as the severity of the spinal abnormalities can vary considerably. Most of these patients have very low epidural dose requirements, so their blocks must be established with smaller than usual increments and sufficient time allowed for each dose to take effect. The chances of successful epidural anaesthesia in the achondroplastic patient will be maximized by careful patient selection and preparation, with discussion of risks and the possibility of technical complications, and by optimal positioning and cautious dose titration.

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