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Anesthetic management of parturients with achondroplasia: a case series

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ABSTRACT

Background: Achondroplasia is the most common form of dwarfism, and cesarean delivery is often required in parturients with achondroplasia due to cephalopelvic disproportion. Given the challenges for both regional and general anesthetic techniques, there is no consensus on the optimal anesthetic management for cesarean delivery in these patients.

Method: A search of our electronic medical records for all female patients who had a diagnosis of achondroplasia and had a delivery in our health system from January 1, 2001 through June 16, 2023 was performed. Institutional review board exemption was obtained.

Results: We identified seven achondroplastic patients with 12 cesarean deliveries and described their anesthetic management during labor and delivery.

Conclusion: Despite the historical preference of general anesthesia in achondroplastic patients due to concerns of unpredictable spinal anatomy and unreliable local anesthetic spread, neuraxial anesthesia was successfully utilized in achondroplastic parturients and is a viable option in carefully selected patients. Reduction of intrathecal local anesthetic dose that minimizes the risk of high spinal and emergent intubation, as well as a titratable neuraxial technique, can be effective in this patient population.

KEYWORDS Achondroplasia; cesarean delivery; cesarean section; dwarfism; neuraxial anesthesia; pregnancy; skeletal dysplasia

Achondroplasia is the most common form of skeletal dysplasia and is categorized by disproportionate short stature, long-bone shortening, and macrocephaly.[1] Parturients with achondroplasia commonly require cesarean delivery due to cephalopelvic disproportion, yet there is no consensus on the best anesthetic technique for cesarean delivery in these patients. Despite the increased risks for general anesthesia, including difficult intubation, altered respiratory mechanics, and sleep apnea in parturients with achondroplasia,[2] general anesthesia was historically thought to be the preferred anesthetic management.[3,4] Concerns for the neuraxial technique in patients with achondroplasia include unpredictable and altered spinal anatomy leading to variable local anesthetic spread, previous lumbar surgeries with potentially difficult neuraxial placement, and residual scarring. Accordingly, to define the peripartum anesthetic implications of achondroplasia, we retrospectively assessed the peripartum anesthetic management and outcomes of patients with achondroplasia at our institution.

METHODS

After obtaining approval from the Mayo Clinic Institutional Review Board, which provided the authors with a waiver for patient authorization of release of protected health information, we searched an institutional database (the Mayo Data Explorer) to identify patients with achondroplasia who delivered at Mayo Clinic in Rochester, Minnesota and in the Mayo Clinic Health System from

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January 1, 2001 through June 16, 2023. The Mayo Data Explorer, a Mayo Clinic–developed self-service web application, aggregates data from various source systems, including prior electronic medical record systems, to provide comprehensive clinical data within a single centralized database. Subsequently, demographic, obstetric, and anesthetic data were manually extracted from the electronic medical record. Immediate neonatal outcomes and any maternal complications were also reviewed. Patients with a form of dwarfism other than achondroplasia were excluded, as well as those with a height >147 cm.

In our institution, for a standard cesarean delivery, we typically administer 12 to 13 mg of 0.75% hyperbaric bupivacaine, along with 15 mcg of fentanyl and 150 mcg of morphine for spinal anesthesia, and 15–20 mL of 2% lidocaine with epinephrine, 100 mcg of fentanyl, and 2 mg of morphine for epidural anesthesia.

RESULTS

We identified seven patients with achondroplasia who had a total of 12 deliveries. All patients delivered via cesarean delivery. Of note, patient 4 had an additional delivery at 19 weeks' gestation for fatal fetal anomalies, for which she had a vaginal delivery without neuraxial analgesia. Patient demographics and obstetric data are summarized in *Table 1*. The parturients' height ranged from 124 to 139 cm. Anesthetic management is summarized in *Table 2*. Seven of the 12 anesthetics were neuraxial (four spinal anesthetics and three epidurals), and five were general anesthetics. Patient 1 received a spinal anesthetic for all three of her pregnancies, patient 2 received general anesthesia for her first pregnancy and an epidural for her second pregnancy, patient 3 received a spinal anesthetic for delivery, patient 4 received an epidural for both of her deliveries, and patients 5, 6, and 7 received general anesthesia for all their pregnancies *(Table 1)*.

Table 1. Characteristics of 12 anesthetics in seven patients with achondroplasia

Patient	Age (years)	Height (cm)	Weight (kg)	Body mass index (kg/m^2)	Gravity/ parity	Gestational age
1A	20	127	47.2	29.3	G1P0	39w
1B	23	127	49.9	30.9	G2P1	39w
1C	25	127	53.5	33.2	G3P2	37w6d
2A	26	128	90	54.9	G1P0	36w
2B	32	128	89	54.3	G2P1	36w4d
3	26	130	67.4	39.9	G1P0	39w1d
4A	22	124	58.3	37.9	G1P0	39w
4B	24	124	55	35.8	G2P1	39w
5A	32	121	57	38.9	G2P0	39w
5B	34	121	52.8	36.1	G3P1	37w1d
6	22	124	52	33.8	G2P0	38w
7	22	139	82.9	42.9	G1P0	39w

Dose reduction for spinal and epidural local anesthetics occurred in all cases in this review. The range of intrathecal bupivacaine dose was 9.75 to 10.5 mg. Neuraxial dosing was adequate in all deliveries except two; patient 1B required local anesthetic infiltration of the skin to augment the spinal during initial skin incision and patient 2B required an additional 10 mL 2% lidocaine in her epidural during the case. Due to a retained sponge, patient 2B received an additional 10 mL of epidural 2% lidocaine for re-exploration and received 325 mcg intravenous fentanyl in divided doses throughout the case. There was no conversion from neuraxial anesthesia to general anesthesia in any case. The parturient who had general anesthesia for her first delivery (2A) required an awake fiberoptic intubation due to concerning airway exam. Direct laryngoscopy was used for intubation of patients 5 and 6, without any reported difficulty and grade 1 views. Lastly, the endotracheal tube was secured using a video laryngoscope for patient 7 without complication.

Reported complications included transient paresthesias that resolved spontaneously (patients 1A, 1B, and 4B), prolonged fundal pressure for infant delivery (4B), significant fetal decelerations to a fetal heart rate of 80 to 90 beats/min at the time of regional anesthesia leading to expeditious delivery (3), and vacuum-assisted delivery with enlargement of uterine hysterotomy due to fetal macrocephaly (5A).

DISCUSSION

Achondroplasia affects approximately 1 in 25,000 births, arising as a sporadic mutation in 80% of cases and, less commonly, as an autosomal dominant mutation in fibroblast growth factor receptor type 3.[5] It is the leading cause of dwarfism.[6] Since this is a rare disorder, there is a paucity of evidence-based care recommendations for patients with achondroplasia, and anesthesia for cesarean delivery is no exception.[5] Despite the risks attributed to general anesthesia in parturients, it was historically the preferred anesthetic management in achondroplastic patients due to concerns of unpredictable spinal anatomy and unreliable local anesthetic spread.[7] We present a review of 12 deliveries in seven patients with achondroplasia, with seven of these deliveries performed using reduced dose neuraxial anesthesia. As evidenced by the success of these neuraxial anesthetics and few complications reported in this review, neuraxial anesthesia can be performed in carefully selected achondroplastic patients with an experienced team.

Anatomic challenges in parturients with achondroplasia complicate achieving a safe and adequate anesthetic technique for cesarean delivery. These patients typically have a small mouth opening, maxillary hypoplasia, flattened nasal bridge, short thyromental distance, macroglossia, temporomandibular

Table 2. Specifics of anesthetic techniques for cesarean delivery and outcomes

Patient	Anesthetic technique	Spinal medications	Epidural medications	Dermatome*	Additional medications	Complications	Neonatal outcomes	Additional notes
1A	Spinal	9.75 mg bupivacaine, 150 mcg morphine	NA	T4-T6	NA	Transient paresthesia	Apgar 8 and 9, infant with achondroplasia	Multiple attempts to identify dura
1B	Spinal	9.75 mg bupivacaine, 150 mcg morphine	NA	NA	NA	NA	Apgar 8 and 9	Local anesthetic infiltration of skin at incision
1C	Spinal	10.5 mg bupivacaine, 15 mcg fentanyl, 100 mcg morphine	NA	T4	NA	Transient paresthesia	Apgar 8 and 9, infant with achondroplasia	Multiple attempts at two different levels
2A	General	NA	NA	NA	*Premedication*: sodium citrate 30 mL, metoclopramide 10 mg, glycopyrrolate 0.4 mg, midazolam 1 mg ×2, lidocaine 4% inhalation ×2, fentanyl 50 mcg. *Intubation*: Awake fiberoptic with a 6.0 ETT, secured at 20 cm. *Postintubation*: Thiopental 250 mg, vecuronium 2 mg, fentanyl 100 mcg. *Maintenance*: Isoflurane and nitrous oxide. *Other*: morphine 2 mg, ondansetron 4 mg, neostigmine 2 mg, glycopyrrolate 0.4 mg.	None	Apgar 7 and 9	Awake fiberoptic intubation. Airway exam: MP I. Short thick neck with diminished mobility, large tongue, small oral aperture.
2B	Epidural	NA	*Test dose:* 3 mL 1.5% lidocaine with epinephrine *Epidural load:* 15 mL 2% lidocaine, 100 mcg fentanyl, 1 mg morphine; additional 10 mL needed during case; 10 mL additional for exploration for retained sponge	NA	325 mcg IV fentanyl in divided doses during procedure after delivery	None	Apgar 8 and 9	NA
3	Spinal	9.75 mg bupivacaine, 15 mcg fentanyl, 100 mcg morphine	NA	NA	NA	Significant, persistent FHR decelerations to 80–90 bpm at time of spinal, leading to expeditious delivery	Apgar 4 and 8	NA
4A	Epidural	NA	*Test dose:* 3 mL 1.5% lidocaine with epinephrine *Epidural load:* 10 mL 2% lidocaine, additional 4 mL needed during case; 4 mg morphine	T6 (L) T7 (R)	NA	None	Apgar scores NA, healthy infant	NA

(*Continued on next page*)

Table 2. Continued

Patient	Anesthetic technique	Spinal medications	Epidural medications	Dermatome*	Additional medications	Complications	Neonatal outcomes	Additional notes
4B	Epidural	NA	*Test dose:* 3 mL 1.5% lidocaine with epinephrine *Epidural load:* 10 mL 2% lidocaine, additional 8 mL needed during case; 4 mg epidural morphine	T5	NA	Transient left-sided paresthesia; 3–4 min of fundal pressure before infant delivery	Apgar 1 and 8, infant with homozygous achondroplasia, NICU admission, passed 9 weeks later	NA
5A	General	NA	NA	NA	*Premedication*: sodium citrate 30 mL *Induction*: Lidocaine 60 mg, fentanyl 50 mcg, propofol 120 mg, succinylcholine 100 mg *Intubation*: Direct laryngoscopy, grade 1 view, 6.0 ETT secured at 19 cm *Maintenance*: sevoflurane and nitrous oxide *Other*: propofol 40 mg, fentanyl 100 mcg, oxymorphone 0.7 mg, granisetron 0.1 mg	Vacuum-assisted delivery and enlargement of uterine hysterotomy due to fetal macrocephaly	Apgar 5 and 9	NA
5B	General	NA	NA	NA	*Premedication*: sodium citrate 15 mL *Induction*: Lidocaine 40 mg, fentanyl 100 mcg, propofol 160 mg, succinylcholine 100 mg *Intubation*: direct laryngoscopy, grade 1 view, 6.0 ETT secured at 17 cm *Maintenance*: sevoflurane and nitrous oxide *Other*: hydromorphone 1 mg, fentanyl 100 mcg, granisetron 0.1 mg	None	Apgar 4 and 9	NA
6	General	NA	NA	NA	*Premedication*: sodium citrate 30 mL, metoclopramide 10 mg *Induction*: fentanyl 150 mcg, thiopental 250 mg, succinylcholine 80 mg *Intubation*: direct laryngoscopy; 6.5 ETT, no difficulty *Maintenance*: isoflurane and nitrous oxide *Other*: morphine 5 mg, fentanyl 25 mcg, ketorolac 15 mg, ondansetron 4 mg	None	Apgar 7 and 9	NA
7	General	NA	NA	NA	*Premedication*: none *Induction*: Lidocaine 20 mg, fentanyl 50 mcg, propofol 140 mg, succinylcholine 120 mg *Intubation*: Video laryngoscope (GlideScope), no mask attempted, size 3 blade, 6.0 ETT, secured at 20 cm *Maintenance*: Sevoflurane and nitrous oxide *Other*: acetaminophen 1000 mg. hydromorphone 1 mg, dexamethasone 4 mg, ondansetron 4 mg, fentanyl 200 mcg	None	Apgar 2, 6, and 9	Arnold-Chiari malformation; ventriculoperitoneal shunt; scoliosis and spinal stenosis

bpm indicates beats per minute; ETT, endotracheal tube; FHR, fetal heart rate; IV, intravenous; MP, Mallampati; NA, not available/not applicable; NICU, neonatal intensive care unit.
*Upper dermatome level documented.

joint rigidity, pharyngeal narrowing, atlanto-occipital instability, and limited neck extension, making intubation for general anesthesia challenging and potentially dangerous.[7,8] The increased incidence of sleep apnea and obesity creates further obstacles for mask-ventilation and intubation; one review of 39 achondroplastic patients found that 54% suffered from sleep apnea.[6,7,9] In addition, patients with achondroplasia have an increased incidence of cardiac and pulmonary abnormalities such as congenital cardiac malformations, cardiomyopathies, restrictive lung disease, and ventilation-perfusion mismatch.[10] All these characteristics combine to increase the risk of difficult intubation and rapid desaturation. Although large-scale research quantifying the incidence of complications with general anesthesia in achondroplastic patients has yet to be published, awake fiberoptic endotracheal intubation and video laryngoscopy have been suggested to reduce risks, particularly in patients with evidence of cervicomedullary compression.[6] In our review, only one patient who received general anesthesia required awake fiberoptic intubation due to concern for altered spinal anatomy and difficult intubation. Given the described anatomic and physiological challenges, it is imperative that anesthesia providers are well prepared and have the necessary equipment readily available for managing difficult airways in these patients, adhering to established difficult airway algorithms.

Despite the numerous anatomical challenges to general anesthesia, this technique has traditionally been preferred for cesarean delivery in achondroplastic patients, as neuraxial anesthesia may also be complicated in this population. Achondroplasia is often characterized by spinal stenosis, kyphoscoliosis, lumbar lordosis, poorly identifiable landmarks, narrow intrathecal and epidural spaces, and by definition, short stature.[6,7,10] Consequences of these abnormalities include increased risk of inadequate anesthesia due to difficult placement and high spinal due to unpredictable spread. Despite these concerns, many cases of successful neuraxial anesthesia in this patient population have been published especially in recent years, with most describing use of reduced doses of local anesthetics.[10–15]

Modifications to the anesthetic plan are necessary for the safety of achondroplastic parturients seeking to avoid general anesthesia for cesarean delivery. It is recommended that parturients with achondroplasia deliver in tertiary centers due to the complexity of these cases and the complicated neonatal resuscitation that is often required.[16] Thorough history-taking is crucial to identify patient-specific risks, and further preparation including spinal imaging, arterial blood gas studies, and cardiac or pulmonary evaluation may be appropriate. Neuraxial ultrasound should also be considered in this population. Preprocedural ultrasound guidance has been shown to reduce the number of needle passes required and increase the first-pass success in parturients with difficult landmarks.[17,18] Moreover, prior case reports have demonstrated the use of neuraxial ultrasound to successfully guide placement of neuraxial blocks in achondroplastic parturients.[12,19] Additionally, reduction of intrathecal bupivacaine or other local anesthetics should be considered in parturients with short stature. Patients with short stature are at increased risk of high spinal if typical doses of hyperbaric bupivacaine are used.[11] High spinal blockade could be precarious in patients with a difficult airway. However, inadequate block can have the same consequences and require emergent conversion to general anesthesia. Inadequate anesthesia with dose reduction of intrathecal bupivacaine occurred in one patient in our case series and a second patient had inadequate anesthesia with an epidural, requiring intravenous fentanyl as an adjunct. Although not reported in our case series, a combined spinal-epidural may be an ideal anesthetic, as one could administer a reduced intrathecal bupivacaine dose and have the reliability, density, and symmetry of spinal anesthesia and subsequently use the epidural catheter for extension or augmentation of surgical anesthesia. Drawing from our experience, we recommend that anesthesia providers consider administering an intrathecal bupivacaine dose of 10.5 mg or less as part of a combined spinal epidural anesthetic.

Finally, these patients should receive additional monitoring consistent with the American Society of Anesthesiologists guidelines for prevention, detection, and management of respiratory depression associated with neuraxial opioids,[20] including monitored pulse oximetry. Due to their predisposition to obstruction, sleep apnea, and pulmonary pathophysiology, they are high risk and not appropriate candidates for the Society for Obstetric Anesthesia and Perinatology modified guidelines for respiratory monitoring after administration of neuraxial morphine.[21]

Our study has the inherent limitations of a retrospective review, including possible incomplete or missing data in medical records and uncertainty in the reason for management decisions. Although this large case series describes anesthetic management in achondroplastic parturients, it is still a small sample size, precluding the ability to provide definite recommendations. Moreover, this review spans 22 years, during which anesthetic and obstetric practice changes invariably evolved. Further research including large-scale, prospective studies are needed to provide a higher level of evidence for which anesthetic techniques are safest and most effective in parturients with achondroplasia.

In summary, this review provides seven cases of successful neuraxial anesthesia in achondroplastic parturients undergoing cesarean delivery. While there are unique challenges to using neuraxial anesthesia in patients with achondroplasia, routine use of general anesthesia introduces the risk of difficult airway management in an already high-risk population and potentially denies the parturient the opportunity to witness the birth of her child. Although this review did not include cases with combined spinal-epidural anesthesia, this method may be ideal for these patients. Neuraxial anesthesia was effective and safe in our series of achondroplastic parturients and should be considered in this patient population.

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