

# Anesthesia And Neuromonitoring For Correction Of Thoracolumbar Deformity In An Achondroplastic Dwarf

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## Abstract

Achondroplastic Dwarfism (AD) is a type of osteochondrodysplasia identifiable at birth caused by defects in the growth of tubular bone and/or spine. Anesthetic challenges include abnormalities of the airway, cervical spine, pulmonary, cardiac and neurologic systems. We describe the anesthetic management of an achondroplastic dwarf presenting for T<sub>6</sub> thru S<sub>1</sub> decompression laminectomies, instrumentation and spinal reconstruction with neurologic monitoring. The patient was a 50-year old, 66 kg, 127 cm tall male with spinal stenosis who suffered from severe back pain and thoraco-lumbar myelopathy. Past medical/surgical history included hypertension, thoracolumbar laminectomy, and shoulder arthroscopies. Airway exam showed macroglossia, short mandible with prognathism, large head, short neck & mallampati class III airway. Pulmonary and cardiac evaluations were unremarkable. Neurological exam revealed severe thoracolumbar kyphosis with lumbar lordosis. Standard ASA and bispectral monitors were applied. Spinal cord and lumbosacral roots were monitored via somatosensory evoked potentials (SSEP), electromyogram (EMG) and transcranial evoked potentials (tcMEP). Only upper extremity SSEPs responses were reliable. TcMEPs and SSEPs showed unreliable lower extremity responses. Continuous electromyogram yielded mixed results. Achondroplastic dwarfism poses significant anesthesia challenges. Airway challenges include sleep apnea resulting from brainstem compression and problems with mask ventilation and laryngoscopy, which we did not experience. Intravenous access can be difficult in AD and in our patient necessitated placement of central line. Through the careful titration of anesthetic infusions, we were able to extubate the patient at the conclusion of surgery to perform a neurologic examination. Intra-operative SSEP, EMG and tcMEP monitoring assisted us in avoiding spinal cord ischemia without resorting to a wake up test. Blood loss was minimized with meticulous surgical technique as well as the use of epsilon-aminocaproic acid which has been shown to reduce transfusion requirements in patients undergoing complex spine procedures. We conclude that with adequate anesthetic management, complex spine procedures can be performed on AD patient in a safe manner.

## INTRODUCTION

Achondroplasia dwarfism (AD), the most common skeletal muscle dysplasia and cause of dwarfism,<sup>1</sup> AD is an autosomal dominant trait with 100% penetrance. Spontaneous mutations are common. Diagnosis is made at birth and is clinically based, with radiographic confirmation.<sup>2</sup> Endochondral ossification is decreased secondary to G-to-A substitution in fibroblast growth factor receptor 3 (FGFR-3) located on nucleotide 1138, adversely affecting the growth plates.<sup>3</sup> Dysplastic features include rhizomelic shortening, spinal kyphosis, lumbar lordosis, and spinal stenosis.<sup>4</sup> Spinal stenosis is attributed to decrease vertebral body height, short and thickened pedicles, and hypertrophy of ligamentous tissue, and can lead to spinal cord ischemia and death.<sup>5</sup>

In addition to trunk and limb skeletal abnormalities, (AD)

have multisystem organ involvement. This case report discusses the unique anesthetic challenges in a 50-year-old male with AD who underwent a two-stage revision thoracolumbar decompression, pedicle subtraction osteotomy and long segment fusion T6- L5. We report the use of transcranial motor evoked potentials (tcMEP), somatosensory evoked potentials (SSEP), and electromyogram (EMG) in AD. To our knowledge this has not been reported in the literature.

## CASE REPORT

A 50-year-old wheel chair bound male achondroplastic dwarf, presented with functionally disabling thoracolumbar pain and progressive loss of ambulation secondary to thoracic myelopathy and neurogenic claudication in association with severe recurrent spinal stenosis and sagittal imbalance. He was 127 cm tall, 66 kg (BMI 40.9). Past medical and surgical history was significant for

hypertension, tonsillectomy, and right knee and shoulder arthroscopy. Medications prior to hospitalization included hydrochlorothiazide. Physical examination of the patient showed severe spinal deformity in both sagittal and coronal planes, forward stooped posture, and bilateral lower extremity motor deficits. Airway examination was remarkable for wide mouth opening, full set of teeth and temporomandibular distance of more than 6 centimeters. Cardiopulmonary examination was normal. Initial laboratory data revealed hemoglobin and hematocrit of 39.1 and 13.7 respectively, and platelet count of 226.

Radiographic studies showed severe recurrent spinal stenosis involving the lower thoracic spine (with spinal cord compression) to the lumbosacral junction. Severe kyphoscoliosis with significant sagittal imbalance was also demonstrated.

The procedure was staged 4 days apart. First stage involved revision thoracolumbar decompression T9-S1 and placement of instrumentation T6-L5. Second stage included pedicle subtraction osteotomy, deformity correction, and completion of instrumentation and fusion T6-L5.

On day one, the patient was premedicated with 2mg of midazolam. Pulse oximetry, electrocardiogram, and non-invasive blood pressure cuff was placed on the patient. The patient was preoxygenated with 100% O<sub>2</sub> for three minutes prior to induction of anesthesia. Anesthesia was induced with 150 mcg of fentanyl, 100 mg of propofol, and 40 mg of rocuronium. Direct laryngoscopy and tracheal intubation using a Macintosh 3 blade was successful on first attempt. After confirmation of ETCO<sub>2</sub>, the patient was maintained with propofol (150-250 micrograms/kg/min) and fentanyl (0.5-1.5 micrograms/kg/hr). Additionally, an aminocaproic acid drip was started and maintained at (1gram/hr). Intravenous access included an 18 gauge peripheral IV, a right internal jugular (RIJ) triple lumen central venous catheter placed under ultrasound guidance, and a 20 gauge right radial arterial line.

Spinal cord function was monitored using transcranial motor evoked potentials (tcMEP), somatosensory evoked potentials (SSEP), and electromyogram (EMG). Post-anesthesia induction SSEP from upper extremities showed good morphology and repeatability.

The sensory pathway was monitored throughout the surgery by means of posterior tibial nerve and median nerve somatosensory evoked potentials recording from C3, C4, Cz,

Cs5, Fpz. Sensory responses were measured at upper brain stem, and sensory cortex. The functional integrity of the motor pathway was monitored throughout the surgery by means of transcranial motor evoked potentials and bipolar high frequency direct cortical stimulation (Bi-DCS). Compound muscle action potentials were recorded referentially from the contralateral trapezius/deltoid, bicep/tricep, extensor carpi radialis/flexor carpi radialis, thenar/hypothenar, vastus lateralis/biceps femoris, tibialis anterior/gastrocnemius, and abductor hallucis muscles. SSEPs were recorded (Cadwell Elite, Cadwell Labs, USA) from C3, C4, Cz, Cs3, Fpz, according to the International 10-20 System, with a band pass of 500-30Hz to alternating stimulation of posterior tibial nerve and median nerve at 4.55Hz, 50-300 averages. TcMEPs were recorded from bilateral thenar, hypothenar, extensor carpi radialis, tibialis anterior, and abductor hallucis muscles using a band pass of 100-1000dB. Stimulation (Digitimer D185, UK or Cadwell TCS-1000, USA) consisted of 3-6 pulses at an inter-stimulus interval of 1-4 milliseconds at C3'-C4' according to the 'Threshold-Level' technique that avoids stronger stimulus intensities.

No responses were seen from lower extremities. TcMEP responses from lower extremities showed unreliable responses as well. Right sided lower extremity threshold was less than or equal to 450 volts while the left sided lower extremity was less than or equal to 350 volts. There were no sustained EMG discharges at baseline. Throughout the procedure intraoperative monitoring of SSEP, tcMEP, and spontaneous EMG revealed no changes. Bispectral Index (BIS) and central venous pressure monitoring were also recorded.

Blood loss was estimated (EBL) at 1700ml, with the patient receiving 3 units of packed red blood cells (prbcs), 500ml of cell saver, and 5000ml crystalloid (3000ml LR, 2000 NS). Vitals signs remained within 20% of baseline. Peak inspiratory pressures remained less than 35 cm H<sub>2</sub>O. Central venous pressures remained less than 18. At the conclusion of 8 hours of surgery, an endotracheal tube leak test was performed, and the patient was extubated following his decompression revision. The patient was transferred to the intensive care unit (ICU) for care and monitoring. The patient returned to the operating room on postoperative day 4 for posterior spinal instrumentation/rodding and wound closure.

Laboratory data prior to the second surgery included a

hemoglobin of 11, hematocrit of 30.9, platelet count of 202, and a PT/INR of 15.2/1.12. IV access included a PICC, and the previous RIJ triple lumen central line, and radial arterial line. The patient was premedicated with 2 mg of midazolam, preoxygenated with 100% O<sub>2</sub> for three minutes, and induced with 150 mcg fentanyl, 100 mg of lidocaine, 150 mg of propofol, 10 mg of rocuronium, and 100 mg of succinylcholine. Direct laryngoscopy, using a Macintosh 3 blade followed by tracheal intubation was successful on first attempt. After confirmation of ETCO<sub>2</sub>, the patient was maintained with propofol (100-250 micrograms/kg/min) and fentanyl (0.25-1 micrograms/kg/hr). Aminocaproic acid drip was started and maintained at (1gram/hr). Estimated blood loss was 2000ml, and was replaced with 5 units prbcs, 225 ml of cell saver, 4000ml crystalloid (2800ml LR, 1200ml NS), and 250 ml albumin.

Vitals signs again remained within 20% of baseline. Peak inspiratory pressures remained less than 37. Central venous pressures remained less than 29. Identical neuromonitoring to the first operative procedure was employed.

At the conclusion of the procedure, the patient remained intubated for the sake of airway protection because there was significant facial edema. The patient was transported to the ICU and subsequently extubated two days postoperatively.

## DISCUSSION

Anesthetic challenges in AD are numerous and involve multiple organ systems. Life expectancy is shortened (on average 10 years) with higher mortality early in life.<sup>6</sup> Careful preoperative anesthetic evaluation aims to document preexisting neurological deficits, comorbidities, and minimize the associated complications.

The neurologic manifestations of achondroplastic dwarfism include upper motor neuron lesions, which manifest as weakness, hyperreflexia and clonus. Cervicomedullary compression in children and young adults can cause respiratory distress and recurrent cyanosis. Spinal deformities are common among AD and can have severe debilitating effects, ultimately leading to mortality secondary to spinal cord and foramen magnum stenosis. Such stenosis causes cord ischemia, which can lead to sudden death.<sup>7</sup> The literature does not describe intraoperative neuromonitoring these patients during multilevel spinal decompression and instrumented posterior spinal fusion.

Pulmonary system should be assessed for signs and

symptoms of airway obstruction and pectus excavatum. Neurogenic effects of brainstem compression lead to reduced vital capacity, which leads to higher incidence of pneumonia, cyanotic spells and apnea. Long term, patients with achondroplasia are prone to obstructive and central sleep apnea, cor pulmonale and pulmonary complications.<sup>8</sup>

Upper airway obstruction is caused by brachycephaly as well as facial hypoplasia.<sup>8</sup> Multiple airway difficulties have been described, including difficulty with a good mask fit and difficulty with mask ventilation. Both are due to macroglossia and a short mandible with prognathism.<sup>9</sup> Airway management (potential difficult airway) must consider a many anatomical abnormalities.<sup>10</sup> Kalla and colleagues suggest mandatory C-spine radiographs to predict difficulty of intubation.<sup>11</sup> Mandibular motion is also reduced.<sup>12</sup> In a review by Sisk, the majority of patients requiring endotracheal intubation utilized a smaller endotracheal tube size.<sup>13</sup> Preoperative scans should be reviewed to evaluate stenosis of foramen magnum and upper cervical spine. If a significant stenosis exists, hyperextension of neck should be minimized to avoid spinal cord compression and ischemia. In this case, we did not experience any problems with mask ventilation or intubation of the trachea. The patient was preoxygenated for 3-5 minutes prior to induction of anesthesia, as concerns with rapid desaturation due to low functional residual capacity and potential difficulty with mask ventilation and tracheal intubation were paramount to us. We inserted an oral airway after induction of anesthesia to help with mask ventilation and avoid airway obstruction due to the patient's large tongue. We did not consider an awake intubation as necessary, but had rescue airway devices available.

Intravenous access was difficult to obtain in our patient secondary to flexion deformities, laxity and excess of skin and subcutaneous tissues common in achondroplastic patients.<sup>11</sup> We inserted a right internal jugular central venous catheter.

Patient neuromonitoring included tcMEP, SSEP and EMG. Our patient had SSEPs that had low amplitude and very long latencies. Spontaneous EMGs of lower extremities. This indicated alterations of sensorimotor pathways prior to surgery. The use of such monitoring obviated an awake test. By using the neuromonitoring techniques used, we were able to compare the baseline with any potential change intraoperatively. Its use demonstrates the successful application of this form of monitoring to a achondroplastic

dwarf. Patient was discharged from the hospital 4 days after surgery.

In conclusion, we report our anesthetic experience with a 50-year-old achondroplastic dwarf undergoing a very complex spinal orthopedic procedure. The absence of changes in SSEP, EMG and tcMEP (albeit limited in the lower extremities) supported lack of surgically induced injury to the spinal cord. With careful planning and collaboration between the anesthesiology and surgical teams, we were able to proceed and complete the procedure without complications.

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