Anesthesia for scoliosis: Dwarfism and congenitally absent odontoid process

WILLIAM ROBERTS, CRNA, MD, PhD
LINDSEY C. HENSON, MD, PhD
Rochester, New York

Scoliosis surgery presents the anesthetist with specific clinical challenges. Since scoliosis is the most common problem for which patients of congenitally short stature present to the operating room, the preoperative evaluation of dwarfs is discussed here in the context of a patient with spondyloepiphyseal dysplasia congenita.

In the case described, many of the deformities associated with dwarfism were present. The significance of these to the preoperative, intraoperative, and postoperative care is discussed.

Consideration is given to cervical spine abnormalities, congenital absence of the odontoid process, pulmonary function abnormalities, and mucopolysaccharidosis (a syndrome which may compromise airway management). The intraoperative monitoring of somatosensory evoked potentials and their significance are also discussed.

Key words: Odontoid hypoplasia, scoliosis, spondyloepiphyseal dysplasia congenita.

Introduction
The anesthetic care of patients with congenitally short stature has been reviewed by Berkowitz and associates. A patient with spondyloepiphyseal dysplasia congenita complicated by absence of the odontoid process and severe scoliosis is presented here. The patient's airway and spinal pathology posed significant challenges illustrative of many which may be seen in this population.

Patient history
This 16-year-old, 25-kg, 96-cm, white female with short extremities was referred to the outpatient department for evaluation of anesthesia risk and recording of preoperative somatosensory evoked potentials. Her spinal curvatures were first noted 6 years earlier and were worsening at an increasing rate. She was able to participate in age-appropriate physical activity. There was no history of sleep apnea or central apnea, daytime somnolence, or shortness of breath suggesting airway obstruction. She took no medications and had no allergies. There was no family history of difficulties with anesthesia. The patient had undergone four previous general anesthetics, and oral intubation had been accomplished without difficulty.

Physical examination
The patient had a short neck, prominent jaw, a relatively high and horizontal sternum, and prominent pectus carinatum. The cervical extension was limited to 20°. Anterior anatomic landmarks of the neck were distorted, with only the thyroid cartilage palpable. The cricoid cartilage was retrosternal. The distance from the thyroid cartilage to the tip of the mentum was 8 cm at maximal cervical extension. Temporomandibular joint range of motion was limited to 5-cm maximal
opening. Her oropharyngeal examination was normal, with ready visualization of the faucial pillars, uvula, and soft palate (Mallampati Class I). There was no evidence of excessive submucosal soft tissue in the pharynx.

Her breath sounds were clear. Despite severe scoliosis, excursion was full bilaterally. There was no evidence of right heart congestion or pulmonary hypertension. Her only other noteworthy physical findings were bilateral cataracts.

**Laboratory evaluation**

Electrolytes, urinalysis, and prothrombin time and partial thromboplastin time were normal. The hematocrit was 41%. Pulmonary function testing revealed a forced vital capacity of 0.96 L (108% of predicted based on height), with a forced expiratory volume in 1 second of 0.96 L (105% of predicted). Room air oxygen saturation was 98%.

**Radiologic evaluation**

Cervical spine films demonstrated congenital absence of the odontoid process (Figure 1). No subluxation of C-1 on C-2 was noted with active flexion and extension. There was fusion of C-1 to the base of the skull. The second and third cervical vertebrae were fused posteriorly (Figures 2a and 2b).

Chest radiographs were remarkable for ab-

---

**Figure 1**

*Open mouth view of the odontoid*

In this view, the odontoid process normally occupies the center of the field. Our patient was known to have a congenitally absent odontoid, discovered in screening radiographs at age 2 years.

**Figures 2a and 2b**

*Lateral flexion-extension films of the cervical spine*

(2a) Left: Lateral view of the cervical spine in full extension. The reduced range of motion is apparent, as is the extremely short neck. The interspace between C-2 and C-3 has been labeled by the radiologist. The distance between the C-1 and C-2 is indicated by dashes in the anterior aspect of the upper spine. (2b) Right: Lateral view of the cervical spine in full flexion. The distance between the C-1 and C-2 is indicated by dashes. There is a slight apparent increase in the distance between C-1 and C-2 in this view, which was felt to be due to differences in magnification between the extension and flexion views. No subluxation of C-1 on C-2 was observed.
sence of the 12th rib bilaterally and normal heart size and lung fields (Figure 3). There was obvious asymmetry of the intercostal spaces with a $50^\circ$ curvature to the right from T-2 to T-7 and a reciprocal curve of $60^\circ$ to the left from T-7 to T-11. The lumbar spine was also affected, with a $50^\circ$ curvature to the right from L-1 to L-5.

**Anesthetic course**

Since prone positioning was required and the potential for blood loss was great, general anesthesia with endotracheal intubation was planned. Due to the risk of difficult intubation and subluxation of the cervical spine in the absence of the odontoid process, consent was obtained for awake fiberoptic intubation. Consent was also obtained for intraarterial monitoring and controlled hypotension for reduction of blood loss. Preoperative autologous blood donation was discussed but refused. Two units of packed red blood cells were crossmatched.

The patient was brought to the preoperative holding area, an 18-gauge intravenous (IV) catheter was inserted and atropine (0.4 mg IV) and midazolam (0.25 mg IV) were administered. In the operating room, routine monitors were established. Topical anesthesia of the airway was accomplished with 4% lidocaine spray, 2% lidocaine jelly, and cetacaine spray. Incremental doses of midazolam (0.25 mg IV) and fentanyl (12.5 $\mu$g IV) were titrated to effect. Because of the extremely short neck and consequent difficulty with location of the landmarks, no attempt at superior laryngeal nerve block or transtracheal injection of local anesthetics was made.

We attempted to insert an intubating oral airway (Ovassapian Fiberoptic Intubating Airway, Kendall Healthcare Products, Mansfield, Massachusetts). None of the airways available to us were of an appropriate size for this patient. Attempts at oral fiberoptic visualization of the trachea using manual traction and elevation of the tongue were unsuccessful. There was abundant soft tissue in the hypopharynx, a problem which was circumvented by topical anesthesia of the left nare and introduction of 5.5-mm internal diameter cuffed endotracheal tube. This tube was passed into the hypopharynx until maximal breath sounds were heard. The fiberoptic bronchoscope (Olympus LF-1) was passed through the endotracheal tube and into the trachea. The tube was advanced, and after confirming intubation by auscultation and capnography, general anesthesia was induced (propofol 2 mg/kg IV). Anesthesia was maintained with fentanyl ($5 \mu$g/kg/hr following a loading dose of 8 $\mu$g/kg) supplemented by 50% nitrous oxide and 0.3% isoflurane. Muscle paralysis was established with vecuronium 60 $\mu$g/kg loading and 1 $\mu$g/kg/min thereafter.

The patient was positioned prone with attention to maintenance of neutral neck position. Operative blood loss was reduced by local infiltration of the incision site with 1% lidocaine with epinephrine 1:100,000 and controlled hypotension (nitroprusside and labetalol for mean arterial pressure of 60 mmHg).

**Evoked potentials**

Loss of right upper extremity evoked potentials was noted at the end of the rod insertion. An Erbs point electrode confirmed arrival of the evoked volley at the level of the brachial plexus. Recording electrodes and the scalp and cervical spine position were checked. Since no mechanical
problems could be identified, an intraoperative wake-up test was carried out. Seven minutes after discontinuing inhalational anesthetics, the patient moved all extremities to command. During the wake-up test, the right arm evoked potentials returned to baseline. Anesthesia was resumed for wound closure.

**Postoperative course**

The patient was admitted to the pediatric intensive care unit. Due to significant facial edema resulting from crystalloid administration and dependent positioning, plans were made to leave the endotracheal tube in place for the night with extubation scheduled for the next day in the operating room. The presence of an ear, nose, and throat surgeon and equipment necessary to accomplish tracheotomy via median sternotomy were arranged. The extubation was uneventful.

**Discussion**

Specific considerations in the planning and management of anesthetics for dwarfs depend on the subtype of dwarfism encountered and the degree of expression of that subtype in the individual patient. Scoliosis surgery is probably the most common major surgical procedure for which dwarfs present to the operating room. Our patient presented for surgical correction of worsening scoliosis as a feature of spondyloepiphyseal dysplasia.

**Pulmonary complications of dwarfism and scoliosis**

- **Restrictive lung disease.** As is true for all patients undergoing correction of scoliosis, careful attention to preoperative evaluation of pulmonary function is indicated for patients of congenitally short stature. In many subtypes of dwarfism, the restrictive disease due to scoliosis can be compounded by rib hypoplasia. Achondroplasia is the most common dwarfism associated with rib hypoplasia, which may cause considerable restrictive disease even in the absence of scoliosis.

Restrictive lung disease caused by scoliosis and/or rib hypoplasia reduces vital capacity and functional residual capacity, predisposing these patients to early airway closure. Resultant ventilation-perfusion mismatching and the intrapulmonary shunts created increase the gradient between alveolar and arterial partial pressures of oxygen. Alveolar hypoventilation and carbon dioxide retention may be seen but only in the most severe cases.

In this patient, pulmonary function tests were normal based on the standards used (adjustment for standing height only). Until recently, there were no published standards of normal spirometry values in dwarfs. Standards are now available for these patients but they were constructed from an achondroplastic subgroup and may not be representative of a true "normal" range for all dwarf subtypes.

- **Obstructive and central apnea.** Thorough questioning is indicated to rule out apnea in all dwarfs evaluated for anesthesia. Apnea may be of obstructive or central origin. Central apnea can occur as the result of direct compression of the cord and midbrain in patients with foremen magnum stenosis. Fortunately, this rare complication of dwarfism is amenable to surgical correction.

Obstructive apnea is more common and usually less difficult to discover. Obstructive disease in dwarfs may be due to mechanisms possible in other populations or may be secondary to less common processes. Subtypes of dwarfism may be subject to obstruction caused by mucopolysaccharidosis (submucosal deposition of mucopolysaccharides in the soft tissues of the airway). This obstruction may be evident only after the onset of sleep or following administration of anesthetics or sedatives. Flow volume loops may demonstrate these defects preoperatively and clarify whether the obstructive process is above or below the glottis.

- **Mucopolysaccharidosis and Morquio syndrome.** Mucopolysaccharidosis Type IV (MPS IV), a deficiency of N-acetylgalactosamine-6-sulfatase, results in Morquio syndrome and shares many of the physical characteristics of spondyloepiphyseal dysplasia congenita. These include short spine, short neck, protruding sternum, cataracts, and odontoid hypoplasia or aplasia with or without atlantoaxial instability. The presence of redundant soft tissue in the hypopharynx, the characteristic body habitus and short neck in our patient might suggest a diagnosis of Morquio syndrome. Our patient was evaluated by the Birth Defects and Genetic Screening Program of our hospital. She carries a diagnosis of spondyloepiphyseal dysplasia as a spontaneous mutation, since neither parent was affected. Furthermore, her dwarfism was diagnosed at birth, while MPS IV usually presents at 2-4 years of age.

In patients who do suffer from Morquio syndrome, the major deposition of mucopolysaccharides is often below the vocal cords. Intubation of the trachea may not resolve obstructive problems precipitated by induction of anesthesia. In this regard, patients with MPS IV may present anesthetic problems reminiscent of those associated with anterior mediastinal masses. In any patient for whom the diagnosis of mucopolysaccharidosis is known or considered, magnetic resonance imaging of the
Airway may be helpful in preoperative evaluation and planning.

**Airway management**

- **Laryngoscopy.** The major causes of anesthetic complications in dwarfs relate to difficult airway management and laryngoscopy. An increase in the rate of scoliotic pathology at the onset of puberty is typical of the natural history of this disease with or without dwarfism. With worsening of scoliosis, our patient had developed increased anterior-posterior diameter of the chest accompanied by marked elevation of the sternum. These changes were accentuated by her extremely short neck and limited cervical range of motion. There is a wealth of literature attesting to increased difficulty of intubation in patients with a short neck. We planned fiberoptic intubation because of the risk of difficult intubation.

- **Airway evaluation and management.** Our efforts at oral fiberoptic intubation were complicated by excessive soft tissues in the hypopharynx. Mallampati scoring failed to identify the risk of difficult fiberoptic intubation. The Mallampati scoring system has been validated for identification of patients in whom direct laryngoscopy will be difficult. Difficult fiberoptic intubation cannot be predicted using this system. Oral fiberoptic intubation was also made more difficult by the lack of fiberoptic intubation airways of an appropriate size. This issue should be considered during planning for all cases involving fiberoptic intubation in patients of short stature or for those who have small mouths.

The size of the endotracheal tube placed in our patient was limited by the size of her nares and our reluctance to use a larger tube. We were concerned with the risk of epistaxis from a larger endotracheal tube, further compromising our already complex airway. Endotracheal tube size selection in dwarfs is a topic which is subject to controversy, but recommendations for size selection based on the patient’s age have been published.

- **Odontoid hypoplasia and cervical instability.** Another indication for fiberoptic intubation in this patient was the history of odontoid hypoplasia, a common bony abnormality associated with dwarfism and trisomy. Preoperative flexion-extension radiographs of the cervical spine failed to demonstrate subluxation of C-1 on C-2, despite absence of the odontoid process. While there is an abundance of literature which describes cervical instability and injury from subluxation in patients with odontoid hypoplasia, we were unable to obtain references reassuring us that stability of the cervical spine during anesthesia with muscle relaxation could be assumed based on active flexion-extension films without subluxation in an awake patient.

**Somatosensory evoked potentials**

The operative monitoring of our patient included somatosensory evoked potentials (SEPs). In our institution it is common practice to monitor SEPs in patients undergoing major reduction of scoliotic deformities.

The value of SEPs in this particular case was twofold. Lower extremity SEPs were used for the usual indications. During the curvature reduction portion of scoliosis operations, vascular compromise of the spinal cord may occur and expresses itself as changes in the latency or amplitude of the lower extremity SEPs prior to permanent injury. Modification in the reduction process can then be undertaken.

In the patient presented, upper extremity SEPs were also very valuable. Concern for risk of cord injury as the result of cervical spine subluxation (due to odontoid hypoplasia) was reduced by continued presence of the upper extremity SEPs. Even in the absence of cervical spine abnormalities, prone positioning puts patients at increased risk of brachial plexus injury secondary to traction and/or pressure on the brachial plexus. Somatosensory evoked potentials have been reported as first indicators of brachial plexus injury intraoperatively. Rapid response to such changes reduces the risk of permanent injury and should be pursued in all cases in which intraoperative evoked potential changes are observed.

It is not clear what caused the transient loss of the right upper extremity evoked potential in our patient. Among the possible causes were improper positioning, unrecognized hypotension, technical difficulty with the electrodes (stimulating or recording), or actual cord injury resulting from subluxation or surgical manipulation.

We chose to awaken the patient and perform a “response to command” test of motor function when attempts at repositioning and adjustment of stimulating and recording electrodes failed to reverse the SEP changes. Although the “wake-up test” can confirm the integrity of the anterior motor tracts intraoperatively, they provide no proof of posterior, sensory tract integrity. The combination of normal motor function in a wake-up test and return of upper extremity evoked potentials during the wake-up test satisfied us that our post-operative goal of intact motor and sensory function was assured.

**Conclusions**

Patients with congenitally short stature may
present a variety of challenges to the anesthetist. Careful preoperative evaluation is indicated prior to anesthesia in this population. Abnormalities of the airway and cervical spine require special consideration and should be thoroughly documented. Preoperative pulmonary function testing must be evaluated and additional studies performed if spirometry or flow-volume loops suggest obstructive or restrictive disease. As is always true, a careful history is the most important tool for guiding the selection of investigative studies. Communication between the anesthesia team and other surgical team members is essential to a satisfactory outcome.

REFERENCES


AUTHORS

William A. Roberts, CRNA, MD, PhD, received his undergraduate degrees in Nursing and Psychology from the University of Vermont, Burlington, Vermont. He received his Nurse Anesthesia training and master of science in Nurse Anesthesiology from the Medical College of Virginia, Richmond, Virginia. He subsequently received his MD training and PhD in Anatomy and Neurobiology from the University of Vermont. He completed his anesthesiology residency at Strong Memorial Hospital, Rochester, New York. His principle area of research has been sensory evoked electrophysiology and neuropharmacology.

Lindsey C. Henson, MD, PhD, received her BS in Nutrition from the University of California, Santa Barbara, and her MS in Nutritional Sciences from Cornell University, Ithaca, New York. She received her PhD in Nutrition from the UCLA School of Public Health. She subsequently received her MD training at the UCLA School of Medicine and completed an Anesthesiology residency at UCLA. Her principle area of research is metabolic and drug effects on the control of breathing.