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Massive Pyramidal Tract Signs after Endotracheal Intubation: A Case Report of Spondyloepiphyseal Dysplasia Congenita

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SURGERY in children with dwarfism is often a challenge to the anesthesiologist because of craniocervical abnormalities, respiratory problems, and behavioral disturbances. I report a case of a young patient with spondyloepiphyseal dysplasia in whom massive pyramidal tract signs developed after surgery and tracheal intubation.

Case Report

An 18-yr-old man with spondyloepiphyseal dysplasia congenita was scheduled for surgery to remove retained hardware after surgical correction of a genu valgus. His height was 120 cm and his weight was 30 kg. Medical history included diabetes mellitus type I treated with insulin. Former surgery of the genu valgus, 3 months previously, occurred uneventfully. Preoperative physical examination revealed thoracic kyphoscoliosis, a short neck, systolic murmur, and respiratory arrhythmia. Laboratory data were within the normal range with the exception of the blood glucose level. Plain radiography of the cervical spine (no flexion/extension series) noticed only "little abnormality

caused by spondylosis." Surgery was planned using a balanced anesthetic technique. After administration of a nondepolarizing muscle relaxant (vecuronium) and controlled ventilation for 3 min by mask, the intubation was performed during direct laryngoscopy with a McIntosh 3 laryngoscope blade (Rüsch, Vienna, Austria). The procedure was performed by an experienced staff anesthesiologist without using a cervical collar. During induction and intubation, the head was maintained in moderate flexion. The anesthesiologist could visualize only the posterior commissure, but nevertheless placed the tube, with a guide wire inside, on the first pass. He described the procedure as "not very difficult." The intraoperative course occurred uneventfully. The patient was extubated, and the early postoperative period in the recovery room and at the ward was uneventful. On the first postoper-



Fig. 1. Computerized tomography of the cervicooccipital region.

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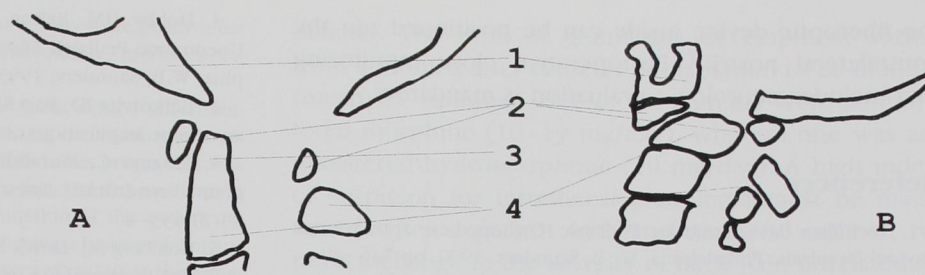
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Fig. 2. (A) Line drawing of a scan of the cervicooccipital region with normal anatomy. (B) Line drawing of the scan of the cervicooccipital region of the patient with spondyloepiphyseal dysplasia and massive pyramidal tract signs. 1 = basilar groove of occipital bone; 2 = atlas; 3 = dens axis; 4 = C2 axis.



active day, a spastic tetraparesis, right more than left, was diagnosed. Neurologic examination revealed a paresis of the upper and lower extremities, right more than left, with positive pyramidal tract signs: increased reflex activity with intact sensibility. Computerized tomography was ordered immediately and showed a massive cord compression to a diameter of 6 mm. Inside the foramen magnum an os odontoid of an 8-mm diameter could be recognized. The anterior arch of the atlas came close to the foramen magnum in a distance of 2 mm. The second vertebra was almost totally inside the atlas (fig. 1). For better understanding, a line drawing of this scan compared to a scan with normal anatomy is provided (fig. 2). The difference in C2 with the dens axis in a healthy patient and the hypoplastic dens axis in a chondrodystrophic patient can be seen. The patient was transferred to the department of neurosurgery. He did not require traction, but a steroid medication was administered. During this therapy the paresis decreased and, after 3 weeks, muscle power returned to 100%. Cranio-cervical instability and stenosis indicated surgical intervention later in the course. The foramen magnum was enlarged and the vertebrae C0 to C3 were stabilized by Compact Cotrell Dufoiset cervical titanium. Postoperatively, symptoms of tetraplegia developed because of stenosis at the level of C1. Paresis persisted despite revised surgery. Two weeks later, drainage of cerebrospinal fluid accumulation in the cervicooccipital region was performed. Later in the course, bronchopulmonary infection and respiratory insufficiency developed. Despite controlled ventilation and intensive medicine care the patient finally died.

Discussion

Spondyloepiphyseal dysplasia is characterized by disproportionate dwarfism with shortening, predominantly of the trunk, and primary progressive involvement of the spine and epiphyses of long bones, particularly of the upper femur. There are two types of spondyloepiphyseal dysplasia: congenital, which can be detected at birth, and tarda, which manifests itself later in childhood. The tarda type has a prevalence of three or four children per one million children in the population of western industrialized countries, the prevalence of the congenita type is one or two per million people. In both clinical types of spondyloepiphyseal dysplasia the vertebrae are flattened throughout and "pear shaped" during early infancy. The odontoid process is dysplastic and delayed in ossification with risk of atlantoaxial instability. Scoliosis and kyphosis develop in late childhood or early adolescence, and

the thorax is deformed with pectus carinatum. Other epiphyses of the long bones show varying degrees of involvement.¹

Atlantoaxial instability is well known in this specific patient population, but rarely leads to neurologic sequelae (in contrast to this case).^{2,3} However, most case reports and articles dealing with anesthesia management of children with dwarfism stress the difficult airway more than the neurologic risk for the patient.^{4,5} A case report with fatal outcome after anesthesia and laryngoscopy in dwarfism could not be found in the literature before.

As a consequence of this case, our regime in children with dwarfism scheduled for surgery is as follows: The exploration is performed very thoroughly while watching for signs of atlantoaxial instability. The first clinical signs without neurologic deficits are history of cyanosis after crying and feeding, respiratory difficulties, poor head control, fatigue, and decreased exercise tolerance. Muscle weakness of the upper or lower extremities is already a sign of cord compression and myelopathy.

Plain radiography of the cervical region provides limited evidence only of the cervicooccipital anatomy, as aforementioned. In this specific patient population we recommend a neutral and extension lateral view of the cervical spine. In the case of suspicion of atlantoaxial instability computerized tomography is added to exclude cord compression. A consultation with a neurosurgical consultant is advisable. Patients with signs of cord compression should undergo surgical correction. Management of children with unstable but asymptomatic upper cervical spine still is a subject of debate because surgical intervention is not without risk.⁶ However, preoperatively, patients with an unstable neck should either be put in traction or otherwise immobilized. The objective is not to flex or extend the head or to move it laterally during the course of intubation. We prefer to approach the airway by controlled, preplanned fiberoptic intubation in deep, general anesthesia and ventilation over a nasopharyngeal tube. The endotracheal tube with

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the fiberoptic device inside can be positioned *via* the contralateral nostril.⁷ Postoperative close monitoring that includes neurologic evaluation is mandatory.

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Illicit Substance Abuse *via* an Implanted Intrathecal Pump

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IMPLANTABLE pumps with intrathecal delivery catheters are increasingly used for management of chronic pain conditions.¹⁻⁴ Originally, these devices were used mostly in patients with cancer-related pain.^{5,6} Now, because of implantation in patients with nonmalignant pain and a longer life expectancy, new complications are seen. These include catheter or pump problems, infections, intrathecal granuloma, lower extremity paralysis, accidental overdose, postdural puncture headache, amenorrhea, arthralgia, decreased libido, chronic headache, and others.⁷ We report a case of self-administration of illicit substances into an implanted intrathecal pump.

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Key words: Chronic pain; complications; illicit substance abuse; intrathecal pump.

Case Report

A 43-yr-old man had a history of lower back pain since age 30 when he underwent the first instance of back surgery—a one-level discectomy and fusion. He underwent a total of five lumbar operations, including decompression with intertransverse process fusions at L5 and S1, pedicle screws with fixation plates, and ultimately hardware removal at age 38. At age 40, he underwent implantation of a Medtronic (Medtronic Neurological, Minneapolis, MN) intrathecal pump for the delivery of morphine to treat chronic back pain. This was implanted at an outside institution, and according to records we obtained, he was psychologically evaluated and thought to be an appropriate candidate for an intrathecal pump device.

The patient was incarcerated in the Texas Criminal Justice System at age 42 and was cared for in the Texas Department of Criminal Justice Hospital System. Our decision was to maintain intrathecal morphine because the pump was already in place. The patient complained of marginal pain relief at every visit, and over the course of 6 months his dose was increased from 2.5 mg/day to 5.0 mg/day. On at least two occasions before the incident described herein, the aspirated amount from the patient's pump was 2 to 4 ml more than the anticipated amount. This led to a suspicion of catheter blockage or pump dysfunction. The patency of the catheter into the intrathecal space was confirmed during fluoroscopy approximately 6 months before this incident. At that same time, pump function was confirmed by filling the pump with saline and using a code supplied by the manufacturer to run the pump at a rapid rate while visually inspecting the pump mechanism during fluoroscopy.

The patient was seen in the pain clinic for a pump refill. A cloudy liquid (8 ml) was aspirated from the pump. The expected volume was less than 2 ml, according to data from the pump-programming com-