was no relationship between lung–thorax compliance and mixed expired oxygen transients, which the predominant effect of cardiac output alterations may explain.

We found that the mixed expired carbon dioxide transients were small and did not correlate with cardiac output or physiologic dead space changes.

Although mixed expired oxygen transients predict directional changes in oxygen transport, we cannot make a quantitative correlation from our data. It should be noted that the magnitude of the mixed expired oxygen changes is quite small, requiring very stable steady-state conditions for detection. At present, the very efficient valving required in our collection of mixed expired gas is quite complex. Thus, we are prevented from using it for early clinical application. We are, however, considering mathematical methods of amplifying the directional changes in the mixed expired gases. One such method is cumulative summation analysis,\(^\text{19}\) in which the deviation of the mixed expired gas tension for each breath is calculated from the initial steady-state value. Figure 2 illustrates a cumulative summation analysis of a transient change in mixed expired oxygen after a step increase in PEEP. The analysis shows that a significant directional change has occurred with the application of PEEP. Such analyses may allow the small changes involved to have clinical relevance.

In conclusion, we have shown that the measurement of mixed expired oxygen transients provides a non-invasive guide to the efficacy of imposed changes in end-expiratory pressure; we have also defined the magnitude of the likely changes. These changes are small and, presently, this form of continuous monitoring by mass spectrometers in the intensive care unit is not readily applicable.

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Anesthesia for Cesarean Section in Achondroplastic Dwarfs

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Achondroplastic dwarfs may encounter problems during both pregnancy and cesarean section delivery. This method of delivery is inevitable, because although birth weights of achondroplastic infants are only slightly less than those of normal infants, the maternal pelvis is invariably small and contracted. This report describes the management of two achondroplastic dwarfs who underwent cesarean section with epidural and general anesthesia, respectively.

REPORT OF TWO CASES

**Patient 1.** A healthy, 25-year-old achondroplastic dwarf was admitted to the hospital for an elective cesarean section following an uneventful pregnancy. Physical examination revealed a 122-cm, 57-kg woman with the habitus of achondroplasia (short limbs, relatively normal trunk size, and a large cranium with prominent frontal bossing and flattened nasal bridge). The spine was slightly

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kyphoscoliotic with a very pronounced lumbar lordosis, which the patient was unable to obliterate by flexion either in the lateral or in the sitting position. Blood pressure was 128/80 torr in the lateral position and 80/50 torr supine. It was assumed that this represented significant aortocaval compression, although the patient complained of no symptom of hypotension. Results of preoperative laboratory tests, including pulmonary function studies, were unremarkable, except for a slightly increased residual volume. Arterial blood-gas analyses were consistent with a slight respiratory alkalosis, which is normal at this stage of pregnancy.

An epidural anesthetic was used because the patient wanted to be awake for delivery. It was attempted at the L3–4 interspace, with the patient in the sitting position. Although the epidural space was located by the loss-of-resistance technique on two occasions, in neither instance could the catheter be advanced beyond the end of an 18-gauge Tuohy needle. On the third attempt, however, cerebrospinal fluid was obtained and the needle was removed.

The procedure was successfully repeated at the next highest interspace and the patient placed in a horizontal position with left uterine displacement. After a 3-ml test dose of 3 per cent chloroprocaine had been administered with no sign of spinal anesthesia, a further 6 ml were administered; this resulted in a T4 level of analgesia within 15 min. Systolic blood pressure fell transiently from 120 torr to 90 torr, but it responded rapidly to the administration of intravenous fluids and ephedrine, 10 mg, iv, and remained stable thereafter. A further 9-ml dose of chloroprocaine, administered 35 min after the initial dose, maintained adequate analgesia during the operation, which lasted 67 min. A 2.4-kg infant with Apgar scores of 8 at 1 min and 10 at 5 min was delivered without difficulty. Although obviously achondroplastic, the infant did not experience the respiratory difficulties that may result from an inadequate thoracic capacity. The mother received two 30-ml epidural injections of saline solution following delivery, and a post-lumbar-puncture headache did not develop. Postoperatively, both mother and baby did well.

**Patient 2.** The patient, a healthy 25-year-old, 117-cm-tall, achondroplastic dwarf weighing 52 kg, was in labor when first seen. She had not sought prenatal care. One previous cesarean section had been performed with general anesthesia, following failure of spinal anesthesia, and the patient again requested general anesthesia. A rapid induction sequence of thiopental, 250 mg, and succinylcholine, 100 mg, was followed by cricoid pressure and endotracheal intubation without difficulty using a 7.0-mm tube. Anesthesia was maintained with 5 per cent oxygen and nitrous oxide, halothane, 0.5 per cent until delivery, and a 0.2 per cent succinylcholine infusion. Left uterine displacement was employed. A 2.2-kg achondroplastic female infant, with Apgar scores of 8 at 1 min and 9 at 5 min, was delivered without difficulty. The postoperative course of the mother and infant was uneventful.

**DISCUSSION**

The incidence of achondroplasia in the United States is about 15 per million births. Approximately 80 per cent of cases result from spontaneous mutation; 20 per cent are familial, with the disease transmitted genetically as an autosomal dominant. The primary defect is caused by underdevelopment and premature ossification of bones forming from cartilage, resulting in characteristic craniofacial and spinal abnormalities. Fertility in achondroplastic dwarfs is low.

Pregnancy in achondroplasia is accompanied by several problems, of which impaired cardiorespiratory function is probably the most hazardous. In normal pregnancy, functional residual capacity (FRC) decreases significantly near term as the enlarging uterus encroaches on the diaphragm. In achondroplasia this is ever more pronounced, as the distance between the symphysis pubis and the xiphoid process is about 25 per cent less than in women of normal height. The small maternal pelvis and severe lumbar lordosis prevent engagement of the fetal head, thus maintaining the uterus as a totally abdominal organ and further decreasing FRC. Kyphoscoliosis may additionally compromise pulmonary function. Severe respiratory embarrassment resulting in respiratory acidosis and inability to assume the recumbent position from the twenty-fourth week of pregnancy onwards have been reported. The anesthesiologist must therefore consider these patients as potentially hypoxic, with severely decreased FRC, resulting in significant shunting of deoxygenated blood.

General anesthesia has been recommended as the technique of choice in achondroplasia, although two case reports have described difficulties with endotracheal intubation. In both reports, extension of the neck was difficult or impossible. Direct laryngoscopy in one patient was not possible because extension was limited at the occipital–atlantic joint. Roentgenographs showed the atlas lying in a concave occipital bone. Premature fusion of the bones at the base of the skull occurs in achondroplasia, resulting in a shortened skull base and small foramen magnum. The latter may result in infantile hydrocephalus or compression of the cervical spine, with death ensuing in the first year of life. Hyperextension of the neck during intubation should probably be avoided to minimize the risk of cervical cord trauma.

Problems are also associated with regional anesthesia. Skeletal abnormalities in achondroplasia may include severe lumbar lordosis, kyphoscoliosis, and a relatively narrow spinal canal containing a normal-sized spinal cord. Neurologic problems may result in later life because of compression of the cord by osteophytes, prolapsed intervertebral discs, or deformed vertebral bodies. Because of these abnormalities technical difficulties in performing epidural or spinal anesthesia have been reported. The narrow subarachnoid space may make it impossible to obtain a free flow of cerebrospinal fluid, and similar narrowing of the epidural space may result in difficulty introducing a catheter and increased likelihood of incurring a dural tap. There are no data regarding the appropriate dose of local anesthetic for spinal or epidural anesthesia in this condition. In the first patient, an adequate sensory level was obtained with a
Anesthetic Considerations for Ehlers-Danlos Syndrome

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Ehlers-Danlos syndrome is an inherited connective tissue disorder characterized by hypermobility of the joints, hyperextensibility of the skin, and a propensity to easy bruising and bleeding.

Many patients with Ehlers-Danlos syndrome never come to medical attention because their symptoms are so slight. The incidence of the disease is, therefore, more common than is generally recognized. Although anesthesia and operation are uneventful in the majority of patients with Ehlers-Danlos syndrome, severely affected individuals can experience life-threatening complications.

The surgical complications of Ehlers-Danlos patients are well documented. These include uncontrollable hemorrhage and wound dehiscence. Anesthesiologists should be familiar with the disorder because it poses several problems in anesthetic management. However, there is presently no information about the condition in the anesthesia-related literature.

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The purpose of this report is to review the clinical features of the syndrome, discuss anesthetic considerations, and make recommendations for perioperative management.

Report of Two Cases

Patient 1. A 29-year-old Caucasian woman died from intra-abdominal hemorrhage during an exploratory laparotomy. She had had a life-long history of easy bruising, intermittent prolonged epistaxis, and excessive bleeding following dental extractions. Since childhood she had had periodic episodes of melena, and after menarche she had suffered unusually heavy menses. Spontaneous hemorrhage from a left retro-ocular aneurysm at the age of 26 years had necessitated an ophthalmologic operation and transfusion of 7 units of blood. At the age of 27 years the patient had a miscarriage at 5 months' gestation. Her family described her as a “free bleeder,” although a coagulation defect had never been identified. Neither her parents nor her siblings had similar problems.

A week prior to her death she was admitted to her community hospital in active labor and gave birth to a normal full-term infant. Coagulation studies were normal. A spontaneous perineal hematoma developed during the second stage of labor. Postpartum bleeding persisted despite good uterine tone and absence of placental fragments on uterine exploration. Hemorrhage continued, necessitating transfusion of 4 units of blood and, eventually, hysterectomy. A further 4-unit transfusion was necessary intraoperatively. Results of all coagulation studies remained normal. The patient was transferred to our hospital for evaluation of the bleeding problem.

Examination on admission revealed lax skin with numerous excoriations over both arms and the lower abdomen and "cigarette-