operations. One should consider either tracheal intubation under topical anesthesia with the patient awake or a rapid intravenous induction-intubation sequence. This latter method can be accomplished as follows: preoxygenation; pretreatment with nondepolarizing muscle relaxants to prevent fasciculations; rapid induction with intravenously administered agents; modest head-up position; a Sellick maneuver; the trachea quickly intubated with a cuffed endotracheal tube.

Since it is unlikely that the problem of aspiration of gastric contents will ever be completely solved, prevention is superior to post hoc treatment.

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Anesthetic Management of a Parturient with Myotonia Atrophica

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Of the three myotonic syndromes, myotonia congenita, paramyotonia congenita, and myotonia atrophica, the last is most feared. It is a hereditary degenerative disease of muscle, especially skeletal muscle, characterized by abnormal delay in relaxation following contraction. As the disease progresses, involvement of skeletal, cardiac, and smooth muscle may lead to impairment of many systems. Abnormal pulmonary function, swallowing disturbances resulting in aspiration, EKG abnormalities, and a variety of arrhythmias have been reported. In women, the disease is associated with ovarian insufficiency, amenorrhea, infertility, and a high incidence of early abortion in those patients who become pregnant. Involvement of the adrenal glands may produce clinical adrenocortical insufficiency in either sex. Evidence for smooth muscle involvement has also been reported. Obstetric patients with myotonia congenita who reach term often have prolonged labor, although too few cases have been described to establish the relationship between myotonia atrophica and abnormal labor patterns. Hydramnios has been reported to occur with myotonia atrophica, perhaps secondary to the inability of the fetus to swallow. Although overt myotonia atrophica in newborn infants has not been reported, neonates born of mothers affected by the disease may initially have difficulty in swallowing and suckling. In view of the disturbed muscle function, and the possibility of multisystemic and smooth muscle involvement, the anesthetic management of patients who have myotonia atrophica, especially obstetric patients, may be difficult.

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REPORT OF A CASE

A 23-year-old primiparous Caucasian woman who had no history of a previous surgical procedure or anesthesia was admitted to the obstetric service in the thirty-sixth week of gestation because of vaginal bleeding. During a previous admission, two weeks earlier, ultrasound scan of the abdomen had suggested placenta praevia; uterine size had been consistent with a 36-week gestation; the estimated size of the fetus was 2 kg.

Ten years previously, the patient had been seen in the neurology clinic and the diagnosis of myotonia atrophica established. She manifested typical symptoms: inability to flex the hand grip, curling of the tongue, myotonic facies (expressionless and unwrinkled), bilateral carotars, and involvement of many muscle groups. Creatine phosphokinase was elevated and electromyelogram showed abnormalities characteristic of myotonia atrophica. There was no mental retardation or cardiopulmonary involvement. There was some atrophy of the muscles of the hands and neck.

The patient had been treated with quinine, but after frequent dosage adjustments, the drug was discontinued because its use resulted in tinnitus and vertigo. The patient had had no drug therapy for myotonia for four years previous to the present admission. Two years before this admission, pulmonary function tests had been normal.

Prenatal evaluation during the pregnancy had disclosed no abnormality. At the time of admission, there was no active uterine bleeding. Results of complete blood count and urinalysis were normal, as were coagulation studies. An electrocardiogram at the time of admission disclosed no abnormality. The uterus was not contracting. Twenty-four hours after admission, vaginal bleeding occurred, with blood loss estimated to be 200 ml. The patient was brought to the operating room, where vaginal speculum examination revealed no visible placenta praepraevia, but profuse bleeding from the cervical os.

Cesarean section was performed using spinal analgesia. A 2.5-kg normal male infant was delivered. The placenta was anterior and low, a marginal placenta praepraevia. Prior to delivery of the infant, the patient's blood pressure and pulse rate had been normal and stable. After delivery, marked uterine atony resulted in rapid blood loss. Within 3-4 minutes after delivery, the blood pressure decreased from 120/70 to 70/40 torr and pulse rate increased to 120/min. Hypotension was treated with intravenous injection of 10 mg ephedrine and transfusion of whole blood. At the conclusion of the operation, blood pressure had risen to 120/70 torr; pulse rate was 110/min.

The patient was taken to the recovery room, and a third unit of whole blood was administered. Estimated blood loss was 1,500 ml. A total of 40 units of oxytocin had been administered during the operation. Two hours postoperatively, after 3 units of blood and 4,000 ml of lactated Ringer's solution, the hematocrit was 25 per cent. Coagulation studies remained normal. Blood pressure decreased to 80/60 torr and pulse rate was 120/min. The uterus was soft and unreactive to manual or oxytocic stimulation, and profuse vaginal bleeding continued.

The patient was returned to the operating room, where she was anesthetized with cyclopropane and the trachea intubated after 30 mg d-tubocurarine, iv. Examination of the uterus revealed it to be markedly flaccid and unresponsive to either manual massage or intravenous and intrauterine oxytocic agents. It was necessary to perform a hysterectomy to control bleeding. During the operation, the blood pressure remained at 70/50 torr, and pulse rates ranged from 110 to 140/min. The patient received an additional 4 units of whole blood, one unit of packed cells, and 3,200 ml of lactated Ringer's solution. At the end of the procedure, the blood pressure was 130/80 torr; pulse rate, 110/min. Ventilation was spontaneous and adequate with an endotracheal tube in place.

The endotracheal tube was removed half an hour later in the recovery room, when the patient became fully reactive, 2½ hours after intubation. Vital signs remained stable after she received a fifth unit of whole blood in the recovery room. The subsequent postoperative course was uneventful and the patient was discharged from the hospital a week later. The infant had Apgar scores of 9 at 1 and 5 minutes, continued to do well, and showed no sign of myotonia.

DISCUSSION

Anesthetic management of patients who have myotonia atrophica includes difficulties associated with complications resulting from multisystemic involvement by the disease. In addition, the myotonic patient may have abnormal responses to specific pharmacologic agents used in anesthesia, notably severe myotonic spasm following administration of depolarizing relaxants.14,15 The mechanism of such abnormal responses to depolarizing relaxants is unclear, but the frequency with which they occur leaves little doubt that use of depolarizing muscle relaxants in the myotonic patient should be avoided.16

Marked respiratory depression following thiopental has been reported to occur in patients with myotonia atrophica.17 It has been suggested that this depression is peripheral as well as central, and may involve an action of thiopental on the neuromuscular junction. It is more likely, however, that thiopental-induced respiratory depression represents a central respiratory depression superimposed upon already atrophic respiratory musculature. For this reason, thiopental...
should be used with extreme caution, if at all, in myotonic patients.  

Nondepolarizing muscle relaxants, such as d-tubocurarine, are not contraindicated, and have been used safely for muscle relaxation during anesthesia. In patients receiving quinidine, the dosages of these compounds should be reduced by approximately half to one third. Nondepolarizing relaxants, however, will not relieve the extreme rigidity associated with myotonic crisis. Reversal of nondepolarizing relaxants with neostigmine may precipitate myotonic crisis and so should be avoided.  

The effects of volatile anesthetic agents on myotonia and the incidence of prolonged sustained generalized muscle contraction are uncertain, but appear to be minimal. The only consistent observation is that post-anesthetic shivering has occasionally been associated with development of so-called "myotonic crisis."  

Regional analgesia does not prevent myotonic spasm. Although myotonic muscle that is infiltrated with a local anesthetic becomes flaccid, brachial plexus block producing sensory and motor paralysis fails to abolish myotonic contraction of the denervated muscle following mechanical stimulation.  

Acute myotonic crisis characterized by generalized sustained skeletal muscle contraction occurring during anesthesia, is, fortunately, rare, but can be a serious problem. There is no agreement on the treatment of this event when it occurs. Intravenous administration of quinidine (300-600 mg) has been used, with varying success. Large doses of steroids (1 g or more) have also been found to be of some value.  

Although myotonia atrophica is one of the degenerative muscle diseases that have been associated with development of malignant hyperpyrexia during general anesthesia, there are no data to prove the existence of a definite relationship between the two diseases.  

The anesthetic management of the present patient was determined by the above-mentioned considerations. The choice of regional analgesia for the cesarean section was based on well-founded obstetric anesthetic principles and represented a convenient method of avoiding the use of muscle relaxants and thiopental. A subarachnoid technique using a low dose of tetracaine (8 mg) was chosen over peridural analgesia because of the lower incidence of local anesthetic toxicity, including shivering, associated with spinal analgesia.  

Although time did not allow a detailed preoperative evaluation of this patient, the electrocardiogram was normal, she had no symptom related to pulmonary dysfunction, and pulmonary function tests had been normal two years earlier. No evidence of endocrine involvement was found in the prenatal evaluation, which included a glucose-tolerance test and protein-bound iodine determination, both of which had normal results. At the time of her second anesthesia, the patient's condition was quite different. A second regional technique was contraindicated because of hemorrhagic hypovolemia. Cyclopropane was chosen as the anesthetic agent because of its speed of induction and the ability to use it with high concentrations of oxygen. Endotracheal intubation was considered necessary because of assumed gastric contents associated with pregnancy. d-Tubocurarine was used because it has not been associated with precipitation of myotonic crisis in such patients. Awake intubation was considered as an alternative but was not done because of the patient's inability to cooperate and because of the necessity for rapid induction of anesthesia in order that the hemorrhage might be stopped. Postoperatively, it was decided that the d-tubocurarine would not be reversed by neostigmine, and that ventilatory support would be provided for as long as necessary.

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Radicular Back Pain Following Lumbar Epidural Blood Patch

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Until recently, management of patients with post-lumbar-puncture headache has been troublesome. In 1960, Gormley described the cases of six patients in whom he injected 2–3 ml of autologous blood into the epidural space at the level of the original lumbar puncture and produced dramatic and lasting relief of the headache. This method received more widespread publicity when DiGiovanni reported a series of 50 patients who received epidural blood patches. Complications following epidural blood patches have been mainly limited to low-back discomfort, transient paresthesias, and tenderness over the injection site. Only one case of a transient neurologic complication has thus far been reported. In the case described below, severe low-back pain with signs of nerve root involvement followed an epidural blood patch.

REPORT OF A CASE

A 36-year-old woman had suffered an anterior septal myocardial infarction in December 1973. In April 1974, she was found to be pregnant, and was scheduled for therapeutic abortion and vaginal tubal ligation. Additional history revealed that she was taking no medication and had never taken anticoagulants, and she had not had a previous epidural or subarachnoid block.

The patient was 162 cm tall and weighed 47 kg. No physical or laboratory abnormality was found. Spinal anesthesia was administered for the operation. Lumbar puncture was performed without difficulty at the L3–4 interspace using a 25-gauge spinal needle. The day after operation, the patient complained of headache, and was instructed to increase her fluid intake. She was discharged from the hospital on the second postoperative day, still complaining of a severe incapacitating headache. On examination, the patient had a continuous headache, the severity of which increased markedly with upright posture. She was...