factor in preventing accurate PetCO2 measurements in patients <8 kg ventilated with an ASV.

In summary, we have shown that PetCO2 measurements sampled from the proximal end of the endotracheal tube do not accurately predict Pao2CO measurements in patients weighing less than 8 kg who are ventilated with a continuous-flow, time-cycled ventilator and a Mapleson D partial rebreathing circuit. By contrast, PetCO2 measurements sampled from proximal sites accurately predict the Pao2CO in patients more than 8 kg in weight who are ventilated with this circuit and in all patients (<8 and ≥8 kg) ventilated with the Siemens-Elema "Servo" 900-C Ventilator. This study indicates the need to develop an accurate technique to sample PetCO2 when continuous flow ventilators and Mapleson D circuits are used in small infants. Meanwhile, the Siemens-Elema "Servo" 900-C remains a very useful ventilator when accurate end-tidal Pco2 monitoring is important in small infants.

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Anesthetic Management for Cesarean Section of a Patient with Charcot-Marie-Tooth Disease

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Charcot-Marie-Tooth disease, a rare degenerative disease of the peripheral nervous system has been recognized as a clinical entity since 1886.** Described separately by Charcot and Marie in France and Tooth in England, the disease usually follows an autosomal dominant mode of inheritance. The hallmark of the disease process is peroneal muscle atrophy, reflecting the tendency for involvement of distal limb musculature. High pedal arches or club feet are common; mildly affected patients may demonstrate only foot deformities. Nerve conduction velocities and sural nerve biopsies permit differentiation into two subtypes. Type I usually has an onset in the first or second decade of life with foot drop and steppe gait. Sensory impairment occurs in a stocking and glove distribution. Later in life, atrophy of intrinsic hand muscles occurs. Tendon reflexes are diminished in affected areas, and foot deformities are common. Type II usually appears in adulthood, with symptoms similar to type I. Either subtype may present at any age, however. Foot deformities may be evident for many years prior to the appearance of muscular atrophy. Progression of type I is slow, and type II, very slow. Incapacitation is very rare, and death usually occurs from other causes.†

We recently encountered a patient with Charcot-Marie-Tooth disease who had experienced a severe exacerbation of her disease process during pregnancy. Such occurrences have been rarely reported.‡,§ Anesthesia management of such a patient has never been described, although anes-
thesis for a non-pregnant patient undergoing hystere-
tomy has.4 Anesthetic management, postoperative course, and
physiologic basis for the underlying disease process are
discussed.

CASE REPORT

A 20-yr-old gravida 1, para 0, abortion 0 female was referred to
the obstetrical service with the diagnosis of Charcot-Marie-Tooth dis-
ease. Motor weakness of her lower extremities had been noted at 10
yr of age and, since then, slow progression of that deficit had occurred.
Prior to pregnancy, no difficulty was encountered with the normal
activities of daily life, and she was fully ambulatory. Initially, pregnancy
seemed to have no effect on her underlying disease state, but, in the
fourth gestational month, she had increasing weakness of the lower
extremities. She also had shortness of breath and orthopnea. The pic-
ture of increasing neuromuscular dysfunction with respiratory com-
promise prompted referral to our center, at which time pregnancy
was at 37 weeks by dates, but 30 weeks by ultrasound. The patient
could walk slowly with assistance, and was unable to raise her arms above
her head. Aid was required with most daily activities, including eating.
Vital signs were remarkable for a resting tachycardia ranging up to
120 bpm and tachypnea of approximately 50 breath/min. A physical
examination revealed an obese patient of 100 kg whose height was
167.6 cm. Arterial blood pressure was 136/92 mmHg. Breath sounds
were decreased bilaterally, and pitting edema was noted in the lower
extremities. The remainder of the physical examination was unre-
markable.

Past history revealed orthopedic operations on both lower extremities
at age 12 yr. Family history revealed Charcot-Marie-Tooth disease in
the patient's mother and maternal grandfather. Additionally, the mother
of the patient reported similar, although much less severe, exacerbations during her pregnancies.

While breathing 100% O2, PaO2 was 49 mmHg, PaCO2 was 79 mmHg,
HCO3 was 29 mmol/L, pH was 7.39, and oxygen saturation was 85.2%.
Other laboratory examinations were unremarkable. A chest radiograph
showed increased vascularity secondary to pregnancy, bilaterally
elevated hemidiaphragms, and a heart size to be within normal limits.

ECG was remarkable only for sinus tachycardia. Forced vital capacity
was 1200 ml (28% of predicted), and forced expiratory volume at 1 s
was 480 ml (24% of predicted).

Neurologic consultation was obtained to further evaluate the nature
and extent of this disease. Subjective evaluation of upper extremity
muscle strength revealed abduction 3+, flexion 7+, extension 7+, wrist
tension 7+, grip 7+. In the lower extremities, knee extension was
5+, plantar flexion 5+, and dorsiflexion 5+. All tendon reflexes were
absent, and response to pinprick and proprioception was decreased in
the lower extremities. Toes were downgoing on plantar stimulation,
and clonus was absent. Neuromuscular strength was scored as follows:
5 = full strength, 4 = slightly diminished strength, 3 = ability to flex
and extend the joint against gravity, 2 = ability to flex and extend the
joint, but not against gravity, 1 = barely detectable movement, and 0
= no movement.

Over the next 3 weeks, the patient remained hospitalized with con-
tinued observation of her pulmonary status. Arterial blood gases
remained unchanged, and daily forced vital capacities were stable in the
range of 1.2–1.3 l. Aminocentesis was performed twice, and revealed
immaturity of the fetal lungs. Because pulmonary function was stable,
she was discharged home for a period of time to allow for further fetal
development.

After a period of home stay for 2 weeks, the patient returned to
the hospital; aminocentesis revealed fetal maturity, and preparations
were made for elective cesarean section the following day. Neurologic
status, arterial blood gases, and pulmonary functions were unchanged
from the previous hospitalization.

On the following day, the patient was taken to the operating room
after premedication with an oral antacid solution. Because a supine
position was intolerable to this patient, she was transferred to the op-
erating table and placed in a sitting position. Intravenous and intra-
arterial catheters were inserted, and glycopyrrolate 0.2 mg was given iv.
While the patient was breathing oxygen, anesthesia was induced by
means of a rapid sequence induction with thiopental and atracurium
iv. Immediately following loss of consciousness, the patient was placed
in a supine position and the trachea intubated. Cricoid pressure was
maintained until tracheal intubation was confirmed by bilateral breath
sounds. Anesthesia was maintained with nitrous oxide, oxygen (50:50),
and enflurane (0.5–1.0%), with atracurium for muscle relaxation.
Pulse oximetry and end-tidal CO2 monitoring were utilized, along with
continuous ECG, precordial stethoscope, peripheral nerve stimulator,
oxygen analyzer, and esophageal temperature. Surgery proceeded un-
eventfully, and a normal female infant was delivered with Apgar scores
of 9 at 1 min and 10 at 5 min. Intraoperative blood gas analysis revealed
a pH of 7.35, PaCO2 of 124 mmHg, PaO2 of 43 mmHg, and HCO3 of
24 mmol/L. Neostigmine and glycopyrrolate were given iv at the end of
the operative procedure for reversal of any residual neuromuscular
blocking. The trachea was extubated, and the patient was taken
to the recovery room where mechanical ventilation was instituted.
Ventilatory support was withdrawn by decreasing the intermittent
mandatory ventilation rate. Six hours later, spontaneous ventilation via
tube revealed a pH of 7.35, PaO2 of 118 mmHg, PaCO2 of 42
mmHg, and HCO3 of 23 mmol/L. The trachea was extubated, and
initially respiratory exchange appeared adequate. However, in the
ensuing 12 h, respiratory function declined as the patient grew progres-
sively weak. PaCO2 increased to 60 mmHg with a respiratory rate of
40 breaths/min. Reintubation of the trachea was performed and me-
chanical support of ventilation resumed. Over the next 4 weeks, mul-
tiple intubations and extubations took place. During this time, slow
improvement in neuromuscular function occurred, allowing improved
respiratory exchange. On the 26th postoperative day, the trachea was
extubated for a final time. Several days later with a PaCO2 of 215, pH
was 7.44, PaO2 was 60 mmHg, PaCO2 was 49 mmHg, and HCO3 was
33 mmol/L. On the 31st postoperative day, the patient was discharged
home. Neuromuscular function had improved markedly in her upper
extremities; however, the patient remained non-ambulatory and are-
flexic. Three months after discharge, the patient had returned to an
ambulatory status. Neurologic function had returned to baseline in
her upper extremities, but some subjective residual weakness remained
in the lower extremities.

DISCUSSION

Exacerbation of Charcot-Marie-Tooth disease by preg-
nancy has rarely been reported.2,3 One patient expe-
rienced multiple pregnancy-associated exacerbations with
intervening remissions.4 The disease state apparently
returned to baseline, and was stable between pregnancies.
None of the pregnancies required operative intervention for
delivery. A second case cited described increasing
weakness and intolerable burning pains in the extremities
secondary to progressive neurologic dysfunction, requir-
ing pregnancy termination at 32 weeks by cesarean sec-
tion.2 Marked symptomatic recovery was reported within
hours following delivery, with return to baseline by 12
weeks. Additional neurologic evaluation in that patient
included nerve biopsies that indicated neural edema as a
causative factor for worsening neurologic function. The anesthetic course of that patient was not described. Anesthetic management of a patient with Charcot-Marie-Tooth disease undergoing abdominal hysterectomy with general anesthesia has been reported; however, that patient was not pregnant, and had no exacerbation of her neurologic disease process. Polyneuropathies associated with pregnancy or oral contraceptives has been reported more frequently.

Elective operative delivery of our patient was undertaken with several aims. First, we believed that termination of pregnancy would reverse the trend of progressive neuromuscular dysfunction. Second, and a greater consideration, was that the patient would have tolerated an active labor poorly. Normal \( P_aCO_2 \) at term is 30–32 mmHg, but our patient's resting \( P_aCO_2 \) was near 50 mmHg, demonstrating little respiratory reserve. To circumvent any progression of her neuromuscular disease and respiratory failure, operative delivery was elected as soon as fetal maturity was established. Operative intervention prevented the marked demand which would have been placed on her respiratory system by active labor. Minute ventilation normally increases to the range of 20–25 l/min with labor. It is unlikely that this patient could have met this increased respiratory demand. Her obesity likely played a secondary, although still significant, role in her respiratory failure. The increased weight of the chest wall and elevated intraabdominal pressure increased the work load of respiration. This placed additional strain on the already failing neuromuscular system. The sudden increase of respiratory exchange secondary to labor could have led to increasing respiratory failure with marked hypoxia, hypercarbia, and acidosis. An urgent situation for intervention and correction of the respiratory failure and delivery of the infant would have been created. Deleterious effects on both the mother and infant would have most likely resulted.

A resting tachycardia was present, suggesting the possibility of myocardial dysfunction resulting from her disease process or, possibly, myocardiopathy of pregnancy. Myocardial involvement has been reported in Charcot-Marie-Tooth disease, but dysfunction was limited to the conducting system of the heart. No evidence of myocardial conduction abnormalities or ventricular enlargement was noted on the electrocardiogram. The chest radiograph did not demonstrate cardiac enlargement or pulmonary edema suggestive of cardiomyopathy.

Regional anesthesia was considered for delivery of the infant, but was felt to be a poor choice in light of the patient's underlying neurologic dysfunction. Furthermore, the inability of this patient to tolerate a supine position would have made operative delivery under regional anesthesia impossible.

 Succinylcholine was avoided in this patient for fear of a hyperkalemic response secondary to her neurologic disease. Although patients with stable Charcot-Marie-Tooth disease might receive succinylcholine without adverse response, this patient's disease process was not stable. The acute change in the patient's neurologic function made us fearful of utilizing depolarizing neuromuscular agents. Hyperkalemia following succinylcholine in patients with lower motor neuron disease has been reported.

In summary, we have presented a patient with Charcot-Marie-Tooth disease who experienced a severe exacerbation of her disease process with pregnancy. Such patients have been rarely reported, and anesthetic management has not. The preoperative evaluation was remarkable only for restrictive lung disease on the basis of neurologic dysfunction. Rapid sequence induction and general anesthesia were accomplished without difficulty. Recovery was much slower than anticipated, and discontinuing ventilatory support took 26 days. The patient was discharged with improving, although still significant, neuromuscular impairment. Follow-up 3 months later revealed full return of function in her upper extremities, with some subjective residual weakness in the lower extremities.

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