

factor in preventing accurate  $P_{etCO_2}$  measurements in patients <8 kg ventilated with an ASV.

In summary, we have shown that  $P_{etCO_2}$  measurements sampled from the proximal end of the endotracheal tube do not accurately predict  $P_{aCO_2}$  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,  $P_{etCO_2}$  measurements sampled from proximal sites accurately predict the  $P_{aCO_2}$  in patients more than 8 kg in weight who are ventilated with this circuit and in all patients (<8 and  $\geq$ 8 kg) ventilated with the Siemens-Elema "Servo" 900-C<sup>®</sup> Ventilator. This study indicates the need to develop an accurate technique to sample  $P_{etCO_2}$  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  $P_{CO_2}$  monitoring is important in small infants.

The authors thank Ms. Terri Cain, Ouidalee Rucker, and Carolyn Barnes for their secretarial assistance, Craig Flinders, Rita Pritchett,

and Jamie Castillo for their technical assistance, and Susan Johnston for her assistance with the artwork. This paper was prepared with the assistance of the Medical Publications Department, The Hospital for Sick Children, Toronto.

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Anesthesiology  
66:410–412, 1987

### 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 per-

oneal 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 steppage 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.<sup>1</sup>

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.<sup>2,3</sup> Anesthesia management of such a patient has never been described, although anes-

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Key words: Anesthesia: obstetric. Nerve: Charcot-Marie-Tooth disease.

¶ Charcot JM, Marie P: Sur une torme particulière d'atrophie musculature progressive souvet familiale débutant par les pieds et les jambes et atteignant plus tard les mains. *Rev Méd Pairs* 6:97–138, 1886

\*\* Tooth HH: The peroneal type of progressive muscular atrophy (M.D. thesis). London: Cambridge University, 1886

thetia for a non-pregnant patient undergoing hysterectomy has.<sup>4</sup> 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 disease. 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 picture of increasing neuromuscular dysfunction with respiratory compromise prompted referral to our center, at which time pregnancy was at 37 weeks by dates, but 30 weeks by ultrasound. The patient could walk only 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 30 breath/min. A physical examination revealed an obese patient of 100 kg whose height was 167.6 cm. Arterial bloodpressure 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 unremarkable.

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 room air,  $P_{aCO_2}$  was 49 mmHg,  $P_{aO_2}$  was 79 mmHg,  $HCO_3$  was 29 mmol/l,  $pH_a$  was 7.39, and oxygen saturation was 83.2%. 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 1100 ml (26% of predicted), and forced expiratory volume at 1 s was 840 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  $\frac{1}{5}$ , flexion  $\frac{2}{5}$ , extension  $\frac{2}{5}$ , wrist extension  $\frac{1}{5}$ , grip  $\frac{1}{5}$ . In the lower extremities, knee extension was  $\frac{3}{5}$ , plantar flexion  $\frac{1}{5}$ , and dorsiflexion  $\frac{1}{5}$ . 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 continued 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. Amniocentesis 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; amniocentesis 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 operating table and placed in a sitting position. Intravenous and intraarterial 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 thiamylal 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  $CO_2$  monitoring were utilized, along with continuous ECG, precordial stethoscope, peripheral nerve stimulator, oxygen analyzer, and esophageal temperature. Surgery proceeded uneventfully, 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  $pH_a$  of 7.35,  $P_{aO_2}$  of 124 mmHg,  $P_{aCO_2}$  of 43 mmHg, and  $HCO_3$  of 24 mmol/l. Neostigmine and glycopyrrolate were given iv at the end of the operative procedure for reversal of any residual neuromuscular blockade. The trachea remained intubated, 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* a T-tube revealed a  $pH_a$  of 7.35,  $P_{aO_2}$  of 118 mmHg,  $P_{aCO_2}$  of 42 mmHg, and  $HCO_3$  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 progressively weak.  $P_{aCO_2}$  increased to 60 mmHg with a respiratory rate of 40 breaths/min. Reintubation of the trachea was performed and mechanical support of ventilation resumed. Over the next 4 weeks, multiple 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  $FI_{O_2}$  of 21%,  $pH_a$  was 7.44,  $P_{aO_2}$  was 60 mmHg,  $P_{aCO_2}$  was 49 mmHg, and  $HCO_3$  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 areflexic. 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 pregnancy has rarely been reported.<sup>2,3</sup> One patient experienced multiple pregnancy-associated exacerbations with intervening remissions.<sup>4</sup> 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, requiring pregnancy termination at 32 weeks by cesarean section.<sup>2</sup> 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.<sup>4</sup> Polyneuropathies associated with pregnancy or oral contraceptives has been reported more frequently.<sup>5-8</sup>

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.<sup>9</sup> 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.<sup>10</sup> 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, cardiomyopathy of pregnancy. Myocardial involvement has been reported in Charcot-Marie-Tooth disease, but dysfunction was limited to the conducting system of the heart.<sup>6</sup> 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.<sup>11</sup>

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.

The authors thank Barbara Pitts for the preparation of this manuscript.

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