cuffed endotracheal tube was passed with relative ease once the
airway opening was identified.
Subsequent surgical exploration revealed anterior displacement of the entire mass of methylmethacrylate out of the vertebral body to an area posterior to and slightly to the left of the larynx. The mass measured approximately 3 × 4 cm, with an attached spine of 3 × 5 cm. Removal of the mass was accomplished and a cricopharyngeal myotomy was performed. No further attempt at stabilization was made. The posterior pharyngeal mass observed during initial laryngoscopy was no longer present upon re-examination at the termination of the surgical procedure.

The patient was awakened with the endotracheal tube in place. The endotracheal tube was removed in the post-anesthesia recovery room. The patient was then admitted to the intensive care unit for observation. The postoperative course was uneventful. Two days after the operation she was able to swallow liquids and semi-solids with ease, and experienced no further dyspnea or dysphagia. Roentgenograms of the chest showed significant clearing.

**DISCUSSION**

The use of methylmethacrylate for stabilization of pathologic fractures is relatively new, and experience with its use for fractures of the cervical spine is limited. In a series of 12 patients in whom methylmethacrylate was used by orthopedic surgeons at our institution, ten have had uneventful courses, with successful stabilization.† One patient had a cardiac arrest in the post-anesthesia recovery room, but it was unrelated to the methylmethacrylate, and the other patient was the one whose case is reported here.

The circumstances leading to displacement of methylmethacrylate in this patient are unclear, although extensive osseous destruction leaving little substance for the methylmethacrylate to key to is a possibility. The displacement probably occurred soon after the original operation, since the patient experienced symptoms immediately. Radiopaque methylmethacrylate is normally used for these procedures, but the dye was inadvertently omitted in the case described. Had it been used, diagnosis of displacement could have been made sooner. With the increasing use of methylmethacrylate for pathologic fractures of the cervical spine, anesthesiologists should be aware of this unusual complication.

**REFERENCES**


Hyperthermia and Ketoacidosis during Anesthesia in a Child with Glycogen-storage Disease

**GERALD EDELSTEIN, M.D.,* AND CAROL A. HIRSHMAN, M.D.*,†**

Hyperthermia and severe non-diabetic ketoacidosis were observed during anesthesia in a child with glycogen-storage disease. This case is being reported because the patient’s initial clinical and laboratory findings resembled those found in malignant hyperthermia. Second, the unusual occurrence of severe ketoacidosis only during anesthesia in a child with glycogen-storage disease has not been described previously.

**REPORT OF A CASE**

A 6-year-old boy was scheduled for surgical correction of an external-auditory-canal stenosis. As an infant he had failed to grow normally and had been found to have hepatomegaly. Examination by light and electron microscopy of a liver biopsy specimen had showed increased glycogen in the liver, and a diagnosis of glycogen-storage disease had been made. Glycogen-storage disease Types I, II, III, IV, VI, and VII had been excluded by enzymatic studies of the biopsy specimen. Studies for Types VIII, IX, and X had not been performed, because these types had not yet been described at the time the biopsy was done. The child never had hypoglycemia, and acidosis never developed during febrile episodes or other stressful periods. Clinically, he was thought to have Type IX glycogen-storage disease (hepatic phosphorylase kinase deficiency). He had undergone general anesthesia with halothane on three previous occasions. At 5 months of age, he had had a hydrocelectomy and inguinal herniorrhaphy. His temperature had been 37°C at the start of the operation. At 20 months of age, the liver biopsy had been performed. Intubation of the trachea had been facilitated by succinylcholine. Thirty-five minutes after induction, the temperature had been 37.4°C, and 70 min after induction, 37.8°C. Anesthesia had lasted 90 min, and the temperature had remained 37.5°C for four hours in the recovery room. At 4 years of age, anesthesia had been administered for tomography of the internal auditory canals. Succinylcholine had again been used to facilitate intubation of the

—* Harrington KD: Verbal communication.

Anesthesiology
52:90–92, 1980

0003-0022/00/0100/0090 $00.60 © The American Society of Anesthesiologists, Inc.
trachea. Temperature had not been monitored during the two-hour procedure.

At the present admission, the child weighed 19 kg, height was 106 cm, and he had bilateral hearing loss. Physical examination revealed bilateral atresia of the external auditory canals and hepatomegaly. Laboratory data showed hemoglobin 13.3 g/dl, hematocrit 38.7 per cent, leukocyte count 9,000/mm$^3$, prothrombin time 12.4 sec (control 9.0 sec), and partial thromboplastin time 27.4 sec (control 32.6 sec).

The patient arrived in the operating room unashed and uncooperative after premedication with meperidine, 30 mg, promethazine, 5 mg, and atropine, 0.2 mg, im. Ketamine, 75 mg, was administered im, followed by enflurane, N$_2$O, and O$_2$ by mask. Succinylcholine, 20 mg, was given iv and the trachea intubated. Rigidity and motting of the skin did not occur. Anesthesia continued with enflurane, N$_2$O, and oxygen. Blood pressure, electrocardiogram, heart tones, and rectal temperature were monitored. Blood sugar was monitored with Dextrostix.$^5$ The patient initially received 5 per cent dextrose in 0.2 physiologic saline solution, iv, and later received two supplementary injections of 50 per cent glucose when Dextrostix indicated blood sugar levels in the range of 90–150 mg/dl.

Rectal temperature was 36.5 C after induction of anesthesia. The patient was lying on a 37-C warm-water mattress and was covered with paper drapes. The operating room temperature was 20 C. An hour after induction, the temperature was 37.5 C, and the warm-mattress heating unit was turned off. Two hours after induction, the temperature was 38.5 C, at which time he was undraped and externally cooled by ice packs. Respiration had been controlled by the anesthesiologist throughout the surgical procedure. A 3-liter total gas flow was used in a semiclosed circle system. Arterial blood gases revealed pH 7.15, P$_{CO_2}$ 52 torr, and P$_{O_2}$ 90 torr; base excess was –12. Electrolyte values were Na$^+$ 135 mEq/l, K$^+$ 4.7 mEq/l. Because of the possibility of malignant hyperthermia, enflurane was discontinued, pancuronium, 1 mg, was given iv, and the patient was deliberately hyperventilated. Cold intravenous fluids were given, with 20 mEq NaHCO$_3$. Blood-gas values in a sample obtained three hours after induction were pH 7.35, P$_{CO_2}$ 33 torr, P$_{O_2}$ 265 torr; base excess was –6. Blood was obtained for determination of lactic acid and creatine phosphokinase (CPK) levels. The temperature gradually decreased, and was 36 C four hours after induction. Blood-gas values at that time revealed ongoing metabolic acidosis, with pH 7.21, P$_{CO_2}$ 44 torr, P$_{O_2}$ 205 torr, and base excess –10. The patient received two subsequent doses of 20 mEq and 10 mEq NaHCO$_3$. The patient had tachycardia (heart rate 120 beats/min) throughout the operation.

Operation ended five hours after induction. Atropine and neostigmine were given for reversal of the action of pancuronium, and spontaneous respiration returned. The patient awakened, the trachea was extubated, and he was taken to the recovery room, where he remained afibrile. CPK was reported to be 295 IU (normal 0–175), and lactic acid was 2.2 mEq/l (normal 0.5–2.2). Urinalysis revealed a very strong reaction for ketones, and detailed urinalysis later that day revealed a large increase in beta-hydroxybutyric acid and a moderate increase in acetacetic acid. Amino acid screen was negative. After his return to his room, blood-gas analysis revealed pH 7.39, P$_{CO_2}$ 90 torr, P$_{O_2}$ 31 torr, base excess –5. Blood glucose was 118 mg/dl and electrolytes were normal. Lactate dehydrogenase (LDH) was 242 IU/l, serum glutamic oxaloacetic transaminase (SGOT) 67 IU/l, CPK 570 IU/l, and alkaline phosphatase 374 IU/l (normal 35–105). Urinalysis later that day showed no ketone bodies. Review of the patient’s previous records revealed a CPK of 29 IU/l at the age of 14 months and a value of 47 IU/l at the age of 2 years.

**Fig. 1. Pathway of glycogen degradation and sites of enzymatic deficiencies in glycogen storage disease, Types VI and IX.**

**DISCUSSION**

A 2-degree C rise in body temperature within a two-hour period of anesthesia suggested the possibility of malignant hyperthermia,$^1$ Moreover, initial blood-gas values, revealing metabolic acidosis and hypercarbia, supported this diagnosis. However, the subsequent finding of normal LDH levels and only slightly elevated CPK levels made the diagnosis of malignant hyperpyrexia unlikely.

The elevation of our patient’s temperature could not easily be explained on the basis of external factors, since the operation was conducted in a cool operating suite, and the patient was loosely covered with paper drapes. Moreover, heating with the warm-water mattress was discontinued when the patient’s temperature was first noticed to be rising, and it continued to rise during the next hour. In addition, he had been noticed to have a rising temperature during a previous 90-min anesthetization. The most likely explanation for the hyperthermia, the persistent tachycardia, and the hypercarbia, which reverted to normocarbia only after marked hyperventilation, was a hypermetabolic state.

Glycogen-storage disease, Type IX, is a deficiency rather than an absence of hepatic phosphorylase kinase. The condition exists in two forms. Type IXa is inherited on an autosomal recessive basis and is characterized mainly by hepatomegaly.$^2$ Type IXb is inherited as a sex-linked recessive. The clinical features are variable and include hepatomegaly, fasting hypoglycemia, and growth retardation, but normal mental development.$^2$ Phosphorylase kinase is an enzyme responsible for converting inactive hepatic phosphorylase b to active phosphorylase a. Active
phosphorylase is necessary for the conversion of glycogen to glucose-1-phosphate and subsequently to glucose\(^8\) (fig. 1).

Ketoacidosis developing during anesthesia in a patient with any type of glycogen-storage disease has not been described previously. Casson found metabolic acidosis in seven patients with glycogen-storage disease (Types I, III, and VI) during anesthesia.\(^4\) The organic acid was assumed to be lactic acid, but was not measured. Acid–base values were recorded for only one of 12 patients with glycogen-storage disease described by Cox.\(^5\) These revealed a profound lactic acidosis. Ketoacids were not measured.

Patients with glycogen-storage disease, Type I (von Gierke's), are prone to develop fasting ketosis.\(^6\) This has not been found in Type IX. The exact mechanism is not understood,\(^7\) but may be related to a defect in the rates of triglyceride and ketone body clearance from blood.\(^2\) This has also been found in Type VI (hepatic phosphorylase deficiency).\(^2\)

We can offer no good explanation for the severe ketoacidosis occurring only during anesthesia in our patient. It is conceivable that anesthesia induced a change in the metabolic pathways involved in glycogen degradation. Changes in hepatic metabolism of free fatty acids are somehow related to a disturbance in hepatic metabolism of carbohydrate.\(^7\)

In summary, hyperthermia and severe ketoacidosis occurred only during anesthesia in a child with glycogen-storage disease, probably of Type IX. The mechanisms involved in the development of a hypermetabolic state with ketoacidosis remain obscure. Biochemical monitoring of patients with glycogen-storage diseases undergoing anesthesia should include measurements of both lactic acid and ketoacids to determine the nature of any intraoperative metabolic acidosis. Additional studies are needed to elucidate the effects of anesthetic agents on hepatic enzymatic activity in patients with glycogen-storage diseases.

References