CLINICAL WORKSHOP

Prophylactic preventive early therapy in response to a high index of suspicion remains, for the present, the only clinically conventional way of decreasing the mortality rate due to malignant hyperthermia. This suggests that all children should routinely have their temperatures monitored and that any increase in temperature or unusual rigidity during anesthesia should lead to its termination as rapidly as possible.

The author gratefully acknowledges the help of Dr. B. Britt and Dr. K. Hellmann in the management of this case.

REFERENCES

Anesthetic Management of a Patient with Epidermolysis Bullosa Dystrophica

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Epidermolysis bullosa, a rare dermatologic condition, constitutes a major anesthetic hazard due to the formation of bullae following pressure or friction. Two varieties of the disease have been described.* The simplex type affects only the skin, especially that of the hands and feet, and healing occurs without scarring. The dystrophic form affects the mucous membranes as well; severe scarring occurs, locomotion is affected, and in some cases the teeth are rudimentary. Cutaneous infe-

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tions are a common complication of both forms. Because of the involvement of mucous membranes in the dystrophic form, maintenance of an adequate airway without the production of facial, intraoral, or tracheal lesions poses a challenge. The following report details the anesthetic management of such a patient.

**REPORT OF A CASE**

A 10-year-old Spanish-American boy whose first lesions had appeared at birth was admitted to the hospital in 1970 for the extraction of five teeth. Previous hospital treatment, mainly for the treatment of cutaneous infections and for supportive care, had been extensive. The patient had spent the three years prior to this admission in a foster home. Although treatment had been more successful since his placement in the foster home, he continued to have persistent bullae and scar formation, especially involving his hands, feet, and pressure points (fig. 1). The disease was chronic, and remissions did not occur.

The patient had been anesthetized for an appendectomy in 1966 in another hospital (unfortunately, the records are unavailable) and for circumcision in 1967. The latter anesthesia was provided with nitrous oxide—halothane—oxygen by mask. The airway was carefully maintained without instrumentation and the procedure was without sequelae.

Extraction of the teeth was first attempted with local anesthesia and sedation, but was terminated unsuccessfully because of lack of patient cooperation. The procedure was rescheduled and general anesthesia was requested. Ketamine was selected because of its ability to provide profound analgesia without impairing spontaneous respiration through an adequate airway. Preoperative medication was withheld since it was felt that such sedation was not required, and because needless bullae might result from the additional injection. Since numerous scars from previous lesions made venous puncture difficult, ketamine was given intramuscularly. The patient weighed 19.5 kg; 100 mg of ketamine were injected into each anterior thigh and 0.4 mg of atropine was administered through one of the needles. Five minutes later, nystagmus and somnolence were present, but stimulation still produced movement. An additional 50 mg of ketamine produced the desired effect. The radial pulse was palpated constantly and respiration was observed. Blood pressure was not taken, to avoid unnecessary skin manipulation. The patient was placed supine in a 15-degree Trendelenberg position, and surgery began 15 minutes after the induction dose had been given.
A mouth gag was inserted against the teeth and suctioning maintained. Ventilation was spontaneous and unobstructed throughout the procedure which lasted 12 minutes; small packs were left in place along the gum lines at its conclusion. Thirty minutes after induction the patient had a lid reflex, was swallowing and moving his extremities. The only evidence of trauma to the skin resulting from the anesthetic technique was a bleb on the left anterior thigh at the injection site. Postoperatively, desquamation of the lips with secondary infection and bullae formation on the palate occurred. These lesions did not interfere with the patient's oral intake of food and fluids, which resumed the afternoon of the operation. There was no evidence of airway obstruction, and the patient was discharged the following day.

**DISCUSSION**

The successful anesthetic management of patients with epidermolysis bullosa requires that contact with skin and mucous membranes be minimized. The absence of difficulties following the patient's two previous operations suggests that extreme care was taken to avoid trauma to the airway. This would have been difficult with an inhalation technique in the present case. However, ketamine proved useful in making these conditions possible. Premedication was avoided; the patient moved himself to the operating table, and undue handling and excessive needle punctures were avoided. Induction was rapid even though the intramuscular route was used, and the hazards of facial trauma from the use of a mask were avoided. Laryngeal reflexes were maintained and, when combined with the head-down position, aspiration was prevented and adequate ventilation maintained without the need for laryngoscopy, tracheal intubation, or intranasal manipulation. Extractions from all four quadrants were accomplished without repositioning the patient or staging the procedure.

**REFERENCES**


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**Surgery**

**CNS DYSFUNCTION** A prospective study of the conditions of 100 open-heart surgery patients elicited information about incidence, characteristics, and causes of central nervous system dysfunction after operation. Neurologic, psychometric, and behavioral observations were made preoperatively and postoperatively. Half of the patients developed neurologic damage which became evident following recovery from anesthesia. Forty-three per cent of survivors developed behavioral abnormalities preceded by focal neurologic damage; intellectual functions were depressed in all such patients. In most patients neurologic signs and mental symptoms disappeared and mental status had normalized by discharge. However, 15 per cent of survivors had signs of cerebral damage at discharge. Cerebral damage was significantly related to increasing age and depression of arterial pressure. In patients with cerebral dysfunction, arterial pressures decreased to levels which did not support adequate cerebral perfusion. Older patients were especially vulnerable to such effects. Neuropathologic findings suggested cerebral ischemia. (Tufo, H. M., and others: *Central Nervous System Dysfunction Following Open-heart Surgery*, *JAMA* 212: 1333 (May) 1970.)