

This study confirms our previous suggestion that fine-screen filters are efficacious, safe, practical, and should be routinely used when transfusion of more than two units is anticipated.

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## Epidermolysis Bullosa Manifested and Treated during Anesthesia

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On occasion, the anesthesiologist, starting an apparently uncomplicated anesthesia, suddenly faces an unexpected life-threatening emergency completely unrelated to the surgical problem. Insufficient information as to the patient's prior medical history is often the cause.

Epidermolysis bullosa was first reported by Koebner in 1886. It is a chronic, non-inflammatory hereditary disease involving the skin and mucous membranes. Lesions are common on the lower extremities, especially the feet, but the hands may also be involved. Least commonly involved is the oral mucous membrane. Lesions consist of bullae, ranging from a few millimeters to several centimeters in size, with or without surrounding erythematous areas. These bullae contain sterile fluid and rupture to form shallow ulcerations.

Epidermolysis bullosa is of two types, simplex and dystrophic. The dystrophic type is classified as dominant or recessive based on autosomal inheritance. Oral lesions are commoner in the dystrophic type, particularly

the recessive form. When present, they are most common in the buccal mucosa and the tongue, followed by the lips, gums, and palate in order of frequency. Oral lesions may manifest as a combination of bullae, infiltrated areas, erosions, or patches of leukoplakia. Impaired motility of the tongue and microstoma has been reported, a result of recurrent lesions that heal with the formation of considerable cicatricial tissue.

These lesions may have their onset soon after birth when the infant starts feeding, or they may be delayed for months or even years. The disease is believed to result from loss of the intercellular bridge, resulting in separation of cells, accumulation of edema fluid, and bulla formation. Eruption is known to occur after stress, trauma, allergic reactions, drug sensitivity, or infection.

#### REPORT OF A CASE

A 60-year-old man weighing 77 kg was admitted the day before a scheduled cataract extraction. His only previous operation, at the age of 40 years, had been an extensive dental reconstruction, completed without complication. Three years prior to this admission, oral bullae that caused difficulty in chewing had developed. A diagnosis of epidermolysis bullosa had been made and the patient treated with steroids for three months. Complete remission occurred, and the patient had since been asymptomatic.

Physical examination revealed that the patient was obese and plethoric, with extensive dental

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work and a healthy oral mucosa. The remaining physical findings and laboratory data were within normal limits.

Premedication was with meperidine, 50 mg, and atropine, 0.4 mg, im, 45 minutes prior to surgery. Anesthesia was induced with thiopental, 300 mg, followed by succinylcholine, 80 mg, iv. After ventilation by mask with 100 per cent O<sub>2</sub>, the trachea was intubated with a no. 8 anode orotracheal tube without difficulty. No oropharyngeal abnormalities or facial lesions were seen in the area of application of the mask. Anesthesia was maintained with N<sub>2</sub>O, O<sub>2</sub> and halothane.

While surgical preparation was in progress, profuse bleeding from the mouth was seen. After suctioning, inspection of the oral cavity disclosed bleeding from ruptured bullae on the right buccal mucosa, the posterior third of the tongue, and the soft palate. Sponges soaked in 40 ml of 1:200,000 epinephrine solution were applied to the bleeding area, providing adequate control so that the operation could be performed. Ventilation was changed from spontaneous to assisted to ensure normocarbia. Following application of epinephrine, increases in pulse rate from 72 to 90/min and blood pressure from 130/70 to 150/100 mg Hg occurred and lasted for 10 minutes. Arrhythmia did not occur. Hydrocortisone sodium succinate, 100 mg, was given iv.

At the end of the cataract extraction, inspection of the oropharynx revealed ulcerated erythematous lesions in the areas described above. The hypopharynx was free of pathologic abnormality. Extubation was performed and the patient was transferred to the recovery room, where he was closely observed for the next two hours. No bleeding or respiratory obstruction occurred.

An additional 100 mg of hydrocortisone sodium succinate was given before the patient's discharge from the recovery room. The following day, the diagnosis of epidermolysis bullosa was confirmed by a dermatologist. The patient was given prednisone, 5 mg, *b.i.d.*, orally. The lesions showed complete healing. He was discharged from the hospital on the fifth postoperative day. He is being followed by the dermatologist and continues steroid therapy.

#### DISCUSSION

Problems that may arise in dealing with a patient with epidermolysis bullosa are: bulla formation, bleeding or ulceration on the face following application of an anesthetic mask, and lesions in the oropharynx or on the epiglottis following dental work, insertion of an airway, or tracheal intubation.

The successful management of these patients requires minimal contact with the skin and mucous membranes. Steroids should be

continued if the patient is being so treated. Avoidance of premedication enables the patient to move from the stretcher to the operating room table under his own power and so avoid unintentional trauma. Barbiturates may be contraindicated in view of the association of epidermolysis bullosa with porphyria.<sup>6</sup> Ketamine has proved useful in four-quadrant dental extraction by providing rapid induction with maintenance of intact laryngeal reflexes; when combined with the head-down position, aspiration has been completely avoided.<sup>7</sup> Epidural anesthesia for prolonged surgical procedures, where feasible, has given satisfactory results.<sup>8</sup>

The use of muslin soaked in a 0.5 per cent solution of hydrocortisone has been reported to decrease the incidence of bulla formation. An insufflation technique combined with the head-down, lateral-tilt position has allowed a clear airway, with prevention of aspiration, and has proved satisfactory for dental work.<sup>9</sup>

In our case, when profuse bleeding from buccal and soft palate areas occurred, application of sponges soaked in a solution of 1:200,000 epinephrine was satisfactory in controlling the bleeding so that the operation could be performed. Epinephrine applied to the buccal mucosa is absorbed at the same rate as an intravenous injection.

The minimal side effects of epinephrine observed, mild tachycardia and hypertension, indicate a plasma level of adrenalin at the upper limits of normal. The 40 ml of epinephrine 1:200,000 solution used was within the range of acceptable maximum dosage for this 77-kg man, considering the losses by retention in sponges and into the saliva.

In our case, it was unnecessary to repeat the application, but according to Katz *et al.*,<sup>10</sup> the dosage can safely be repeated after a 10-minute period.

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## Extrapulmonary Influences on A-aD<sub>O<sub>2</sub></sub><sup>1,0</sup> Following Cardiopulmonary Bypass

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Pulmonary insufficiency is one of many diagnostic and therapeutic challenges posed by postoperative or traumatically injured patients. The alveolar-arterial oxygen tension gradient calculated during inhalation of 100 per cent oxygen (A-aD<sub>O<sub>2</sub></sub><sup>1,0</sup>) has been used widely for assessment of pulmonary dysfunction.<sup>1-5</sup> Changes in A-aD<sub>O<sub>2</sub></sub><sup>1,0</sup> also have been used to dictate oxygen therapy,<sup>1</sup> initiation or withdrawal of mechanical ventilatory support,<sup>2</sup> application of positive end-expiratory pressure (PEEP),<sup>1</sup> administration of diuretic and colloid therapy,<sup>3</sup> and initiation of extracorporeal membrane oxygenation.<sup>6</sup> The purpose of this investigation was to assess the accuracy of A-aD<sub>O<sub>2</sub></sub><sup>1,0</sup> in estimating intrapulmonary right-to-left shunting ( $\dot{Q}_s/\dot{Q}_t$ ) of blood in patients following extracorporeal circulation.

### METHODS AND MATERIALS

Twenty-one consecutive patients admitted to the Surgical Intensive Care Unit following

cardiopulmonary bypass for myocardial revascularization procedures were studied. § Anesthesia was induced with pentothal, followed by nitrous oxide supplemented with morphine (1-3 mg/kg), halothane, or ketamine. All patients had thermistor-tipped, flow-directed pulmonary-artery<sup>6</sup> and radial-artery catheters inserted percutaneously prior to operation. Proper positioning of pulmonary-artery catheters was confirmed by chest roentgenogram.

Following operation, patients were assigned randomly to receive controlled ventilation, intermittent mandatory ventilation (IMV),<sup>7</sup> or IMV with 6 cm H<sub>2</sub>O PEEP, in order to assess the effects of different ventilatory patterns on cardiopulmonary function. One, 4, 8, and 16 hours following initiation of mechanical ventilation, simultaneously drawn samples of blood from the radial and pulmonary arteries were analyzed for hemoglobin concentration (cyanmethemoglobin method),<sup>8</sup> oxygen content by the galvanic cell method,<sup>9</sup> and blood-gas tensions and pH by standard electrode techniques.\*\* Appropriate values

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