Epidermolysis Bullosa in a Newborn

Anthony M.-H. Ho, M.D., F.R.C.P.C., F.C.C.P.,* Cindy S. Tsui, M.B.B.S.
*Department of Anaesthesia and Intensive Care, The Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, New Territories, Hong Kong Special Administrative Region, People’s Republic of China
hoamh@hotmail.com

Epidermolysis bullosa is a rare genetic disease characterized by easy mucocutaneous blistering and subsequent scarring.1 Maintenance of skin integrity is a serious challenge. Applying monitors and facemasks, inserting lines and tubes, moving between crib and bed, positioning, and any minor shearing force can lead to separation of skin layers. No taping with regular tape is allowed.

A 2-day-old, 2-kg, term neonate with epidermolysis bullosa needed a Hickman line and pyloromyotomy. Preoperative examination revealed a contracted submandibular scar that limited neck extension. Mouth opening was 3 cm. There were multiple blisters, including one on the left upper lip where an orogastric tube emerged. During the procedure, we wore latex gloves covered with petroleum jelly, avoided friction on the skin, and covered as much skin with petroleum jelly and gauze as possible. The femoral venous line was sutured to skin and secured with Mepitel® (Mölndlycke Health Care, Norcross, GA). A blood pressure cuff and two oximeters were wrapped around her extremities, which had been wrapped in cotton gauze covered with petroleum jelly. Electrocardiography monitoring was not done.2 Ventilation using a facemask lubricated with petroleum jelly, with gentle head tilt and without an oropharyngeal airway, was uncomplicated. With cricoid pressure, we performed nasotracheal intubation using a laryngoscope covered with petroleum jelly, and the 2.5-mm lubricated uncuffed endotracheal tube was sutured to the nasal septum. By the end of the case, blisters were seen on the incision line and on the ala nasi, where the endotracheal tube emerged. No blister was noted after the procedure at the oximeter and blood pressure cuff sites. No respiratory difficulties ensued upon extubation of the trachea the next day.

References