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Anesthesiology  
56:323-326, 1982

## Airway Management during Anesthesia in Patients with Epidermolysis Bullosa Dystrophica

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Epidermolysis bullosa is a rare hereditary disorder characterized by bullae formation of the skin and of certain mucous membranes. The bullae may arise spontaneously or follow minor mechanical trauma. The disease has been classified on genetic, clinical, and histologic criteria into several distinct categories.<sup>1</sup> Among these are the dominant and recessive forms of epidermolysis bullosa dystrophica in which the bullae heal with scarring. The former is usually mild and the mucous membranes are only occasionally involved. In the severe recessive form the mucous membranes usually are involved and chronic scarring of the oral cavity can result in limited mouth opening and immobility of the tongue,<sup>2</sup> while esophageal bullae may lead to strictures. Concern has been expressed that the larynx may be similarly involved and the possibility of airway obstruction resulting from oropharyngeal and laryngeal bullae has led to avoidance of endotracheal intubation.

This retrospective report reviews the experience from The Hospital For Sick Children in managing the airway during anesthesia for 33 patients with epidermolysis bullosa dystrophica.

### PATIENTS AND METHODS

In the years 1958-1980, 33 patients aged from 3 days to 23 years (19 male, 14 female) with epidermolysis bullosa dystrophica have undergone anesthesia at the Hos-

pital for Sick Children. The anesthetic records and case notes were scrutinized to determine the method of anesthesia and to see whether any complications had arisen.

A total of 309 general anesthetics were given for 329 surgical procedures; dental surgery was combined with another surgical procedure on 20 occasions. Table 1 lists the procedures carried out and reflects the severe scarring and the esophageal and dental lesions commonly seen in these patients. The tracheostomy was carried out because of laryngeal edema in a patient referred for stridor and was not due to bullous formation in the larynx.

Anesthesia generally was induced with 50 per cent cyclopropane in oxygen or intravenous sodium thiopentone (4-6 mg/kg). Maintenance of anesthesia was with nitrous oxide, oxygen, and halothane breathed spontaneously through a modified Ayres T-piece or a Magill circuit (Mapleson A). The youngest patient, aged 3 days, was intubated awake for a gastrojejunostomy and using intermittent suxamethonium chloride, was ventilated with nitrous oxide and oxygen via a T piece.<sup>3</sup>

### RESULTS

#### *Airway Maintenance*

The airway was maintained during anesthesia by endotracheal intubation on 131 occasions, of which 113 were by the oral and 18 by the nasal route (table 2). Twenty-nine of the 33 patients were intubated at least once. An oropharyngeal airway with a Charles adaptor was used four times. A face mask was used 147 times, although it was stated on nine occasions that this was held clear of the face. Table 3 indicates the procedures in which intubation was performed. A gauze throat pack was used 18 times to prevent tracheal soiling during dental procedures.

Thirteen of the 33 patients who had at least one anesthetic had restrictive mouth opening due to scarring and contractures of the corner of the mouth. Six of these

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Received from the Department of Anaesthesia, Hospital for Sick Children, Great Ormond Street, London, WC1, England. Accepted for publication October 9, 1981.

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Key words: Airway: management. Complications: epidermolysis bullosa. Intubation, endotracheal: complications.

TABLE 1. Type of Surgery Performed in the 33 Patients

Operation	Number
Repair syndactyly	87
Change of dressing	151
Dental treatment	42
Esophagoscopy	15
E.U.A. Eyes	17
Sigmoid/Protoscopy	5
Skin biopsy	4
Laryngoscopy and tracheostomy	1
Orthopedic	2
Adenoidectomy	1
Circumcision	1
Gastrojejunostomy	1
Repair umbilical hernia	1
Insertion CVP line	1
<b>TOTAL</b>	<b>329</b>

patients were intubated without difficulty. In one patient with exceedingly limited mouth opening, intubation was not attempted and he was anesthetized twice using a face mask for major plastic surgery to his hands. An unobstructed airway was maintained with little jaw or face manipulation. In the remaining six patients, varying degrees of difficulty with intubation were experienced due to the microstomia. In one of these patients intubation attempts were abandoned and anesthesia for syndactyly repair maintained by means of a face mask. In another patient the laryngoscope blade could not be introduced into the mouth, but blind nasal intubation was performed without difficulty on two occasions, for esophageal dilatation. In another four patients intubation was hindered by loose carious teeth.

#### Bullae Formation

*Facial.* Facial bullae on either the chin, cheek, or bridge of the nose were reported as a complication following nine anesthetics (table 4). On five occasions these were related to the use of a face mask and occurred in the early part of the series. On two of the occasions when facial bullae occurred in patients who had been intubated, one followed esophagoscopy and one followed eye examination. No comment was made as to the exact cause

TABLE 2. Means of Airway Management in the 33 Patients

Method of Maintaining the Airway	Number of Patients
Endotracheal Tube	
oral	113
nasal	18
Charles adaptor	4
Face mask	147
No record	27
<b>TOTAL</b>	<b>309</b>

TABLE 3. Relationship between the Type of Operation and the Number of Patients Who Were Intubated

Operation	Number of Endotracheal Intubations	
	Oral	Nasal
Syndactyly repair	58	
Dental treatment	10	15
Combined plastic/dental	9	1
Esophagoscopy	13	2 (Blind)
E.U.A. Eyes	16	
Orthopedic	2	
Laryngoscopy	1	
Change of dressing	1	
Umbilical hernia	1	
Adenoidectomy	1	
Gastrojejunostomy	1	
<b>TOTAL</b>	<b>113</b>	<b>18</b>

of the bullae. In the remaining two patients the bullae occurred early in the series, when the endotracheal tube strapping was removed. This is not now used.

*Labial.* Blistering of the lips or at the corner of the mouth was reported on six occasions, five of which occurred during dental treatment. These cases involved intubation two times and use of a face mask three times. The sixth occasion was caused by the endotracheal tube connector abrading the angle of the mouth during a repair of syndactyly. Problems occurred on only one occasion, after a patient had undergone dental treatment with anesthesia maintained via a nasal mask. Access for the dentist had been difficult due to limited mouth opening and it had been noted in the operation report that the lips had sloughed slightly. On the journey home, several hours later, the patient found her lips had sealed together. The application of lidocaine ointment allowed the lips to be gently separated.

*Intra-oral.* Bullae were reported on the tongue on six occasions. In three patients the bullae were caused by the surgical procedure and the remainder were caused by the laryngoscope blade. In one of these the anesthetist noted that a large area of mucous membrane peeled off during the intubation. There were no postoperative problems and the patient was intubated a further eight times over a five-year-period without trauma.

TABLE 4. Incidence of Bullae Formation

	Number of Cases	Bullae			
		Facial	Labial	Oral	Laryngeal
Intubation	131	4	3	6*	0
Face Mask	147	5	3	0	0
		9	6	6	0

\* Three cases directly caused by surgical instruments.

On one further occasion a patient had an adenoidectomy performed, but the surgeon declined to proceed to the planned tonsillectomy because "the mucous membrane of the posterior pharynx, uvula and soft palate peeled off on touching." No postoperative problems ensued.

*Laryngeal.* There were no reports of bullae occurring in the larynx in any of these cases, and no patient developed postoperative stridor or airway embarrassment as a result of anesthesia.

Apart from the one case already mentioned, no other postoperative problems resulted from the bullae formation reported, and we were unable to find a case where hospital stay was prolonged by the occurrence of bullae.

### DISCUSSION

The earliest review of anesthesia in epidermolysis bullosa was in 1959,<sup>4</sup> when Wilson commented that although there was no record of bullae occurring within the larynx or trachea, the possibility did exist, and that if these were to arise after extubation, an immediate tracheostomy might be necessary. Though no such problems with bullae have been reported, this anxiety regarding acute airway obstruction following intra-oral and laryngeal manipulations in patients with epidermolysis bullosa has persisted and in an attempt to avoid this problem and that of bullae from face masks, several alternative techniques of anesthesia have been described.<sup>5-20</sup>

However, the successful intubation of patients with epidermolysis bullosa without the development of postoperative airway obstruction has been reported many times.<sup>20-27</sup> The only major problem associated with intubation that has been reported was when profuse bleeding from a ruptured oral bullus occurred.<sup>27</sup> This was controlled by the local application of epinephrine.

In a retrospective analysis such as this, a complete picture of the incidence and pattern of bullous complications of anesthesia cannot be obtained. It is possible that not all instances of minor bullous formation would be recorded. However, it is unlikely that any major airway problem such as acute airway obstruction, or other significant problems would have gone unrecorded.

It is the policy in this hospital when anesthetizing patients with epidermolysis bullosa that in order to avoid prolonged facial manipulations, endotracheal intubation is carried out, where possible, for long operations such as syndactyly repair. Intubation is also carried out, where surgical access requires it, such as for esophagoscopy and in some dental treatment. Oral intubation is the preferred route, using a well-lubricated non-cuffed endotracheal tube, a size smaller than one would normally use, ensuring that the laryngoscope blade is also well-lubricated.

Occasionally the nasal route has to be used. In view of the problem with adhesive tape in both this and other series,<sup>15,24,25</sup> this is avoided. A loose ribbon gauze bandage is preferable. Vaseline gauze is used where the endotracheal tube touches the lips and face.

This series well illustrates the fact that intubation may be difficult in these patients. Seventeen of the 33 patients (51 per cent) had either restricted mouth opening or gross dental problems interfering with intubation, though in only two of these patients could intubation not be carried out.

In short cases, or where intubation is deemed too difficult, anesthesia is maintained by means of a face mask which should have a soft air cushion well-layered with vaseline gauze. One should also protect the under side of the jaw with vaseline gauze, where it is held up by the fingers.

Following this policy of airway maintenance, minor bullae formation during or after anesthesia occurred on only 21 occasions out of 309 anesthetics, and of these, 14 occurred during either dental or esophageal procedures. Intra-oral bullae formation could be directly attributed to endotracheal intubation only three times, and none of these resulted in postoperative problems.

No case of laryngeal or tracheal bullae occurred and there was no instance of postoperative airway obstruction or embarrassment. There have still been no reports in the literature of laryngeal or tracheal bullae occurring during or following anesthesia. It has been suggested that the rarity of laryngeal problems in these patients is related to the fact that the larynx and trachea are lined with ciliated columnar epithelium rather than the squamous epithelium which lines the oropharynx and esophagus.<sup>19</sup> There are, however, two reports of laryngeal stenosis in patients with epidermolysis bullosa,<sup>28,29</sup> though neither of these was related to intubation.

In summary, we report the use of endotracheal intubation in epidermolysis bullosa dystrophica on 131 occasions with no laryngeal or tracheal complications. We feel the hazards of intubation in these patients have been over-emphasized and we would advocate its use more routinely.

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