Epidermolysis bullosa (EB) is a rare genetic disorder resulting in fragility of the skin and mucous membranes (Figure 1). We present a case of neonatal EB for pyloromyotomy with emphasis on the challenges of airway management and secure application of monitors.

CASE REPORT

- 10-day-old, term 2.9kg female with Junctional EB and Pyloric Atresia
- Intubation: Respiratory distress from mucosal sloughing in airway on DOL 9
  - Nasally intubated 3.0 ETT (awake direct laryngoscopy and lubricated laryngoscope)
- Pyloromyotomy:
  - EKG, pulse oximeter, blood pressure cuff, temperature probe placed using materials that avoid excessive friction (Figure 1)
  - Bed lined with non-friction pad, limbs covered with vaseline gauze and soft gauze
  - Saphenous IV placed (tourniquet over soft gauze)
  - Open procedure tolerated well with no new skin lesions
- Multiple failed attempts at extubation in NICU
- Risk of tracheostomy (airway trauma and high rate of infection) was weighed against risk of suspension laryngoscopy and rigid bronchoscopy (airway and skin trauma)
- Suspension Laryngoscopy and Rigid bronchoscopy on DOL 45:
  - Pressure ulcerations of the cricoarytenoid cartilages, no subglottic stenosis (Figure 3)
  - Given glycopyrolate and steroids and extubated the next day
- Extended period on Nasal CPAP (Figure 4)

DISCUSSION

- Junctional EB has an incidence between 2.04-3.59 per million per year, making it a rarely encountered disease in the OR [1]
- Respiratory distress presents many diagnostic and management obstacles
- Differential diagnosis includes sloughing of mucosal debris, ulcerations and blister formation from instrumentation, and sub-glottic stenosis due to prolonged intubation
- Techniques for intubation must include avoidance of pressure on the airway:
  - Lubrication of airway equipment
  - Smaller diameter ETT
  - Possible use of fiberoptic instrumentation of the airway in suspected difficult patients [2]
- In this case of prolonged intubation, rigid bronchoscopy was carried out with minimal trauma, structural abnormalities were ruled out and the patient was successfully extubated

REFERENCES