Introduction

BH is an 8 day old 2kg girl born at 28 weeks gestation with APGAR scores of 5 and 8, at 1 and 5 minutes, respectively. On DOL 1 she developed respiratory failure and hypotension requiring ventilatory support and dopamine infusion for two days. On DOL 6, she presented with signs of an upper GI obstruction, secondary to pyloric obstruction. On preoperative examination, she was noted to have subtle bullae on her skin especially at IV/tape sites, which had not been formally investigated. No work up, or skin biopsy had been performed yet.

Intraoperative Course

On DOL 8, she underwent an abdominal exploration for suspected pyloric obstruction. Given the epidermal blisters (Figure 2) and suspicion of pyloric atresia, epidermolysis bullosa precautions were undertaken perioperatively (Table 1). Adhesives on skin were avoided or minimized and all contact areas were padded with gauze and/or lubrication. EKG pads adhesives were minimized. Gauze was wrapped around the arm under the BP cuff. Eyes were lubricated. Face mask and ETT were also lubricated. She was intubated with a 2.5 mm undersized uncuffed endotracheal tube. Abdominal exploration and resection revealed pyloric atresia and not stenosis. The diagnosis of pyloric atresia in the setting of skin blisters supported the diagnosis of epidermolysis bullosa in this neonate. The intraoperative course was complicated by difficult surgical resection and prolonged operative time. Hence, the decision was made to keep the patient intubated postoperatively. Later, skin biopsies and genetic testing confirmed the diagnosis of epidermolysis bullosa with pyloric atresia (EB-PA.)

Discussion

Epidermolysis bullosa (EB) is a multisystem disorder that poses significant perioperative challenges. EB is caused by a mutation in genes that encode for proteins that provide mechanical adhesion between the epidermis and dermis. EB-PA is a severe form of EB and is often times fatal. EB-PA is an autosomal recessive disorder and is also associated with ureteral/renal anomalies. Its prevalence is unknown, but at least 50 cases have been reported worldwide. EB-PA should be suspected in newborns with blistering of the skin, oral, and mucous membrane with little to no trauma, congenital pyloric atresia, and ureteral and renal anomalies.

Table 1: Perioperative epidermolysis bullosa kit

- Clip-style pulse oximeter probe
- Methylcellulose eye lubricant
- Silicone-based bandage (Mepilex @)
- Silicone-based tape (Mepitac @)
- Silicone-based bandage (Mepitel @)
- Cotton ‘tape’ roll
- Water-based lubricant
- Self-sticking wrap (Corflex @)
- Gauze roll
- Adhesive remover, silicone based (Silcar: @)
- ** Padding for gurney and operating table such as foam ‘egg crates’ should be kept available

References


