Introduction

- Recessive dystrophic epidermolysis bullosa (RDEB) is a rare genetic condition
- Widespread dystrophic scarring and blister formation from cutaneous to mucous membrane
- Perioperative Challenges: reduce new bullae formation, difficult airway, pain control

Case Report

History of Present Illness:
- 16-year-old 36kg male with RDEB
- Left hand reconstruction for complex syndactyly

Perioperative:
- Oral Midazolam 15mg for preoperative anxiety
- Inhaled Oxygen and Nitrous Oxide for PIV insertion
- Standard ASA monitors
- Deep sedation with Propofol (200mcg/kg/min), Ketamine (30mg), and Fentanyl (10mcg)

Regional Anesthesia Technique:
- Insertion of left infraclavicular peripheral nerve catheter
- Pajunk StimuLong NanoLine with tuohy needle (18g x 50mm)
- Ultrasound guidance, brachial plexus identified at 4cm
- Nerve catheter secured at 8cm with bacitracin ointment, 2x2 dressing and Mepitac® tape
- Infiltrated 40mL Ropivicaine 0.2%

Pain Control:
- Required no IV narcotics intraoperatively
- Postoperative pain controlled by left infraclavicular PNC with OnQ Cbloc Pump containing Ropivicaine 0.15% @ 8mL/hr
- Wong-Baker FACES pain scale, self reported pain 0 to 2
- First dose oral narcotic 24 hours postoperatively
- Catheter removed POD 2

Discussion

- RDEB is serious debilitating condition
- Bullae formation leads to widespread scarring, deformity, immobility
- Patients with EB often have chronic pain issues
- Pain management is essential
- Regional anesthesia is effective perioperative pain management option
- Challenges include procedural technical difficulties, catheter dislodgement, and new bullae formation

References