

Care for the Epidermolysis Bullosa Patient Undergoing Cesarean Section

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Background:

- Epidermolysis Bullosa (EB) is a cluster of rare inherited diseases classified by skin fragility with possible multiorgan involvement.
- Blisters develop following minimal injury with the most common being due to mechanical friction
- EB simplex is the most common of the four types

Case Discussion

- A 26 y/o G1 with a history of EB simplex, chronic pancreatitis, epilepsy, depression and chronic opioid use presented in labor at 36 3/7 weeks.
- She elected for a primary c/s due to the risk of skin sloughing with a vaginal delivery
- She did not have an anesthesia consult prior to delivery
- She underwent an uncomplicated spinal anesthetic and primary c-section
- She overall healed well, with minimal skin breakdown



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Teaching Points

- Patients with EB require multidisciplinary care throughout their pregnancy and need specialized planning prior to delivery
- Vaginal delivery, cesarean section and neuraxial anesthesia are generally considered safe with avoidance of instrumental delivery when able
- External cephalic version is not recommended for breech presentation as the shearing force required could cause significant trauma



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Special Considerations:

- Monitors: EKG patches, pulse oximeter, BP Cuff
- Generally, avoid compression stockings as friction can cause injury
- Consider under body grounding pad rather than sticker
- Cervical exams: ensure lubrication of the speculum and/or gloves, avoid fetal scalp electrodes unless baby is known to be unaffected
- Neuraxial anesthesia: consider using towel clamp to hold the drape, EMLA cream for local skin infiltration, padding between epidural catheter and skin under dressing, utilize adhesive remover spray for tape removal
- Airway management: esophageal strictures may increase risk of regurgitation and aspiration, potential for microsomia, scarring and limited neck movement due to contractures, poor dentition and oral blistering.
 - Recommend lubricant on provider's gloves, anything that will touch the patient to minimize skin injury
 - Avoid direct suction where possible and use soft silicone suction devices
 - ETT should be secured with Vaseline gauze padding surface



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