Management of cesarean delivery in a patient with Fanconi Syndrome

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Background

- Fanconi syndrome is a rare genetic or acquired disease
- Defect in the proximal tubule of the renal system
- Malabsorption of multiple electrolytes leading to acidosis, dehydration, rickets, osteomalacia, and growth impairment.
- 1 in 40,000 births; overall incidence is unknown.
- Increased risk of pregnancy complications.
- Multidisciplinary management including nephrology and early anesthesiology consultation is key

Dehydration, Polyuria, Aminoaciduria, Hyperchloremic Acidosis

Short stature, Rickets, Osteomalacia

Muscle Weakness

Electrolyte Disturbances; Hypokalemia, Hypophosphatemia

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Case Presentation

- 31yo G1P0 with genetic Fanconi syndrome complicated by hypophosphatemia-associated rickets presented at 38w6d for induction of labor per pt request in the setting of cholestasis of pregnancy.
- Related clinical findings were notable for short stature (134cm/4'3") and chronic kidney disease(Cr – 2.35).
- Induction of labor was complicated by failure to progress. Given persistent high station of the fetus, concern for cephalopelvic disproportion, and LGA fetus (3.8kg), cesarean section was recommended.
- Ultrasound used to confirm the interspace for combined spinal epidural(CSE) placement
- Reduced spinal dose of only 3 mg of 0.5% bupivacaine, 10 mcg of fentanyl, and 150 mcg of morphine resulted in bilateral T3 sensory and complete motor block
- Intermittent 5ml dosing of 2%Lidocaine+Epi was adequate to maintain anesthesia
- 1.3L crystalloid and 1L of Pitocin, with 125ml urine o/p and QBL of 485ml for the uneventful c-section
- Neonate APGARS of 8 and 9 with nephrology and endocrine consults
- Postop course complicated by hydronephrosis and ATN (Cr-2.9) that improved (2.6) prior to discharge on POD 3.

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Discussion

- Given the rarity of the disorder, limited literature in obstetric patients.
- Optimize electrolyte abnormalities.
- C-section is recommended due to risk of pelvic fracture.
- Neuraxial techniques can be safely performed with the use of ultrasound guidance and appropriate dosing of medications
- For cesarean delivery, a CSE may be a safer choice over a single shot spinal, as appropriate spinal dosing may be difficult to gauge given their short stature and risk of high spinal may be greater.
- We suggest precaution in placement given the risk for damage to osseous structures in the setting of osteomalacia.
- Close post operative monitoring, especially in the setting of neuraxial morphine for analgesia, and follow up with consultants is important

Sources

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