

C-13 The Great Imposters: Metabolic Disorders Disguised as Neonatal Surgical Emergencies

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Objective

After completion of this session, the participant will be able to:

- Formulate the diagnostic paradigm and differential of metabolic acidosis in the neonate.

Case Stem Question

A 2 day old male, born at 35 weeks and 5 days, with a birth weight 1880 grams, presents for emergency exploratory laparotomy. APGAR scores were 9 and 9 after a normal spontaneous vaginal delivery and a pregnancy complicated by maternal type 2 diabetes, maternal positive HIV status, exposure to BIKTARVY-antiretrovirals (bictegravir-emtricitabine-tenofovir alafenamide), intrauterine growth restriction (IUGR) and oligohydramnios. Prematurity and small for gestational age (SGA) status necessitated admission to the NICU where empiric antibiotics and prophylactic antiretrovirals (zidovudine) were initiated.

On day of life (DOL) 2, an episode of bilious emesis with associated apnea and bradycardia necessitated intubation. Subsequent chest x-ray revealed air in a distended stomach with minimal gas in the bowel.

EKG demonstrated findings of ventricular hypertrophy.

Venous blood gas with pH 6.738, pCO₂ 25.3, pO₂ 55.6, Bicarb 5.3, Base excess -28.7, lactic acid 18, glucose 168, Hgb 15.

Guiding Questions for Discussion

1. What is the cognitive framework used in identifying the etiology of metabolic acidosis in a neonate?
2. What surgical neonatal emergencies are associated with neonatal metabolic acidosis and which initial presenting symptoms are the harbingers of probable or impending intestinal obstruction?
3. Please describe the labs and imaging with the highest yield that confirm the anatomic etiology which demand surgical correction.
4. What, if any, significance do the EKG findings have and is any further cardiac work up indicated?
5. If the same patient returned for elective repair of bilateral sizable ilio-inguinal hernias, how would you approach the pre-operative and intra-operative management?

References

1. Ma H, Tang Z, Xiao F, et al. Neonatal Metabolic Acidosis in the Neonatal Intensive Care Unit: What Are the Genetic Causes? *Front Pediatr.* 2021;9:727301. doi: 10.3389/fped.2021.727301. PMID: 34733806; PMCID: PMC8558493.

2. Stacpoole PW, National Organization for Rare Disorders (NORD). The Congenital Lactic Acidosis. <https://rarediseases.org/rare-diseases/congenital-lactic-acidosis>. Published 2009, updated June 6, 2023. Accessed February 6, 2025.
3. Glasser JG, Medscape. Intestinal Obstruction in the Newborn. <https://emedicine.medscape.com/article/2066380-treatment>. Updated February 20, 2021. Accessed February 15, 2025.
4. Unsal Y, Yurdakok M, Yigit S, et al. Organic acidemias in the neonatal period: 30 years of experience in a referral center for inborn errors of metabolism. *J Pediatr Endocrinol Metab.* 2022;35(11):1345-1356. doi: 10.1515/jpem-2021-0780. PMID: 36203204.
5. Merritt JL 2nd, Norris M, Kanungo S. Fatty acid oxidation disorders. *Ann Transl Med.* 2018;6(24):473. doi: 10.21037/atm.2018.10.57. PMID: 30740404; PMCID: PMC6331364.