

Management of Ornithine Transcarbamylase Deficiency in a Parturient with Placenta Accreta

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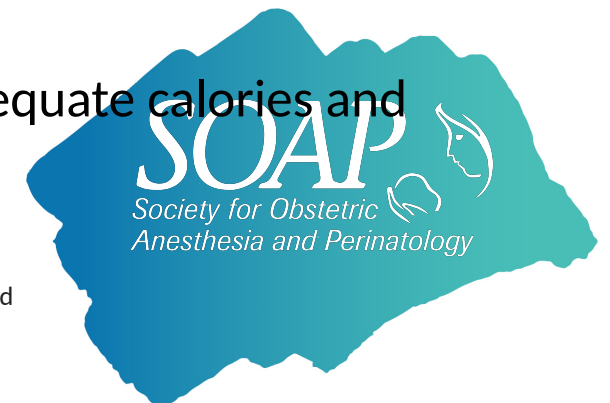
Background:

- Ornithine transcarbamylase deficiency (OTCD) is an X-linked disease and most common urea cycle disorder. Ammonia (NH₃) produced from breakdown of protein cannot be converted into urea —> hyperammonemia —> if untreated can lead to cerebral edema, neurologic damage, and death(1).
- OTCD in pregnancy involves a 47% risk of hemodialysis and 30% mortality with PPD 7-14 of highest risk (2). Maternal protein intake increases to support increased nitrogen demands from placental, uterine, and fetal growth.
- Intra- and post-partum hyperammonemia occurs due to increased physiologic stress and catabolism from uterine involution and maternal weight loss. Lack of fetal nitrogen uptake after birth also contributes(3).
- Management of the parturient involves reducing protein intake, ensuring adequate calories and hydration with dextrose fluids, and providing nitrogen scavengers.

1. Mendez-Figueroa H, et al. Management of ornithine transcarbamylase deficiency in pregnancy. Am J Perinatol. 2010;27(10):775-784

2. Torkzaban, M, et al. Maternal ornithine transcarbamylase deficiency, a genetic condition associated with high maternal neonatal mortality every clinician should know: A systematic review Am J Med Genet. 2019;197A:2091-2100.

3. Ituk U, et al. Peripartum management of two parturients with ornithine transcarbamylase deficiency. Int J Obstet Anesth. 2012;21(1):90-93



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Case events:

- 30 year old G4P1112 w/ history of OTCD and ulcerative colitis admitted for repeat cesarean section @ 37w5d.
- OTCD was diagnosed at 22w1d due to altered mental status from hyperammonemia, requiring prolonged ICU care with intermittent hemodialysis and IV Arg and sodium phenylbutyrate-sodium benzoate nitrogen scavenger infusions to optimize nutritional status and growth.
- Interdisciplinary meeting held. Pre-op NH₃ levels ranged 32-55 umol/L (goal < 75) on IV Arg and nitrogen scavengers with D10NS at 200 ml/h.
- In OR, rapid bleeding and instability ensued due to placenta accreta necessitating conversion to general anesthesia and cesarean hysterectomy. Additional IV access and arterial line placed for resuscitation and monitoring. 2 units PRBCs and 1 unit FFP given for 4100 ml blood loss. Transferred to the MICU post-operatively. NH₃ peaked at 160 umol/L.
- Daily plasma amino acids with q4h NH₃ allowed for rapid weaning off IV scavengers and fluids to a PO regimen without need for hemodialysis.

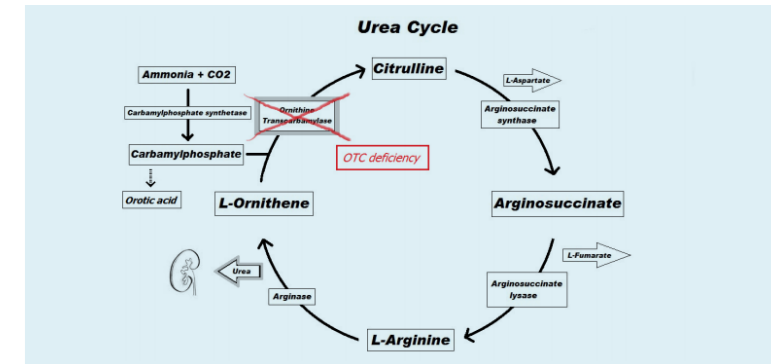


Figure 1: Urea Cycle. Lack of ornithine transcarbamylase prevents breakdown of ammonia.⁴



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Learning points:

- Pregnancy and other catabolic states may unmask OTC deficiency with life-threatening hyperammonemia.
- Presentation can include altered mental status with low BUN and high ammonia levels.
- Our patient did well postpartum with rapid decline in NH₃ despite a prolonged surgery and blood transfusions.
- The continued use of nitrogen scavengers and dextrose containing fluids to avoid protein catabolism was critical in this patient's recovery.
- Her rapid decline in NH₃ to normal levels may be due to her cesarean-hysterectomy, as uterine involution after delivery can lead to hyperammonemia. To our knowledge, this is the first case report of cesarean hysterectomy in a parturient with OTCD.

