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ORNITINE TRANSCARBAMYLASE DEFICIENCY- a case well managed through proper planning and multidisciplinary approach

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Background: OTC is a rare X linked dominant disorder, which results in increased amount of ammonia accumulation in the blood resulting in nervous system toxicity. **Case Report:** We managed a case of this 33 year old para 2 diagnosed as a OTC carrier following the death of her 3 brothers in early twenties. She had uncomplicated pregnancies and postnatal period with her two daughters previously. In current pregnancy the foetus was confirmed to be a male baby, she understood that baby has a 50% risk of having the disease but declined further prenatal diagnosis. The patient had multidisciplinary management in tertiary care hospital involving Adult Metabolic disorders team, anaesthetists and neonatal care unit and detailed plan of management during labour and postpartum period for her and her baby was formulated. Her care was transferred to tertiary care unit and had a normal vaginal delivery with no complications after induction at 38 weeks of gestation in delivery suite where appropriate HDU care and neonatal facilities were available. She stayed in delivery suite HDU for 24 hours after delivery, had serial monitoring of ammonia levels that were reported to be within normal limits. **Discussion:** Subtle cerebral damage may occur in untreated carriers, despite the absence of overt symptoms. Labour and immediate postpartum periods are catabolic states therefore vigilant monitoring is required to prevent hyperammonemic coma which can result in permanent neurological damage and death. **Conclusion:** Pregnancy in women with such rare genetic problems should be managed in a multidisciplinary setting to achieve the best outcome. Close surveillance of male babies is required to prevent any long term sequelae.