INTRODUCTION

Catabolism of amino acid produces free ammonia which is highly toxic to central nervous system; Ammonia is detoxified to urea through series of reaction known as urea cycle.

Hyperammonemia in neonate means elevation of plasma level of ammonia above normal value (normal value < 35 umol/L). It may be a transient phenomenon or an early sign of inborn errors of metabolism.

In neonatal period symptoms and signs are mostly related to brain dysfunction and are similar regardless of the cause of hyperammonemia.

The affected neonate is normal at birth but become symptomatic within a few days after protein feeding, refusal of feeding, vomiting, tachypnea, lethargy quickly progress to coma & convulsions are common. Newborn with hyperammonemia are often misdiagnosed as having sepsis.

Severe transient hyperammoneina in neonate is observed in premature infant with mild Respiratory Distress Syndrome, The cause of this disorder is unknown. Genetic hyperammonemia occur with urea cycle disorders and organic acidopathies.