condition [1]. Usually, no treatment is recommended. cholecystectomy is required in patient who are symptomatic with right upper quadrant pain.

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Thiamine-Responsive Acute Pulmonary Hypertension in an Exclusively Breastfed Infant

Thiamine deficiency presenting as acute severe pulmonary hypertension (PAH) in exclusively breastfed babies has been reported from various parts of India recently [1-3]. Here we report a male infant who presented with acute pulmonary hypertension and responded to thiamine.

A 4-month-old male infant, first born to non-consanguineous parents, belonging to the Chetty community from Mysuru district of Karnataka was referred to our hospital in view of acute onset of cough and respiratory distress. There was a history of approximately ten recent deaths in their community in babies of the same age group, one of them being this baby's first cousin. Most of them presented with bronchiolitis like symptoms, rapidly progressing to acute respiratory distress syndrome (ARDS). This baby was referred to our hospital with the possibility of inborn errors of metabolism.

Baby was delivered at term, was on exclusive breast feeds, thriving well (birth weight 3 kg, present weight 6.5 kg) and had normal development. At presentation, the baby had tachypnea, tachycardia, hypoxia (SpO2 92% in room air) and respiratory distress. Liver was palpable 3 cm below costal margin. Second heart sound was loud and wide split. There was hypotonia and deep tendon reflexes were absent. The arterial blood gas showed mixed disorder with metabolic acidosis and respiratory alkalosis. Tandem mass spectroscopy was normal. Electrocardiogram showed evidence of right atrial (RA) dilatation and right ventricular (RV) hypertrophy (RVH). Echocardiography showed features of severe PAH (RA, RV dilated, moderate tricuspid regurgitation, inter-atrial septum bulging into left atrium) without any evidence of structural heart disease. Contrast enhanced computed tomography (CECT) of lung showed pulmonary plethora with features of PAH without any intrinsic pulmonary pathology.

A detailed dietary history revealed that mother's diet mainly consisted of polished white rice and *Rasam* (a sour soup made with tomato and lentil). There was traditional avoidance of other foods like pulses, vegetables, and non-vegetarian foods during postpartum period. Considering the probability of thiamine deficiency, thiamine was started in a dose of 10 mg/kg, orally. The baby showed clinical improvement, could be weaned off oxygen by 12 hours, and respiratory distress subsided by 48 hours. A repeat echocardiography at 48 hours showed marked improvement with only mild PAH, and subsequently showed complete resolution of PAH at 96 hours. No pulmonary vasodilators other than oxygen were given.

The whole blood thiamine level estimated by HPLC of both baby and mother were low (25 µgm/L and 27.25 µgm/L, respectively, normal range being 28 to 85 µgm/L). There was no history of aphonia, encephalopathy, seizures, or oedema. Serum 25-hydroxy vitamin D3 level was normal. The history, geographic background, clinical features, investigations, and dramatic response to thiamine confirmed the diagnosis of Thiamine responsive acute pulmonary hypertension of early infancy (TRAPHEI). Family was counselled about the dietary modification and both mother and baby were put on thiamine supplementation at 2 mg/kg/day. A repeat echocardiography at one month follow-up visit showed complete recovery. ECG normalized by two months. Muscle tone and deep tendon reflexes normalised after two months of thiamine therapy, suggestive of coexistent neuropathic form of thiamine deficiency. Neuroimaging was not done, as the baby had improved.

Thiamine has an important role in the metabolism of carbohydrates and synthesis of nucleic acid, myelin, and neurotransmitters. A regular dietary supply is must for preventing deficiency because of high turnover rate and low body stores of thiamine. Thiamine causes preferential injury to tissues with heavy metabolic turnover like neurons and cardiac myocytes. The clinical spectrum of thiamine deficiency in children can be acute cardiac form, aphonic form, pseudo meningitic form or neuropathic form [4]. Thiamine deficiency has been documented in those consuming polished cereals and with other dietary restrictions. Infants are at highest risk due to rapid growth rate and high energy needs. In exclusively breastfed babies of thiamine deficient mothers, thiamine levels rapidly decline in the third month of life. As in our case, culturally determined food restrictions during the postpartum period are prevalent in various parts of India [5].

Thiamine deficiency has been identified as a cause of reversible acute PAH attributed to block in glycolysis resulting in ventricular dysfunction and lactic acidosis [1-3,6]. In a relevant geographical and cultural setting, it should be considered in all previously normal babies presenting with acute respiratory distress, tachycardia, and hepatomegaly. Whole blood thiamine estimation is not easily available and may not represent the body thiamine status [4]. So, empirical administration of thiamine is recommended in these infants. Resolution of PAH on thiamine administration is considered as diagnostic. The clinical presentation of this condition is acute, and in untreated cases, fatality is high. Moreover, untreated long-term deficiency can cause intellectual and motor disabilities [4]. For timely diagnosis and treatment, a high index of suspicion among pediatricians is essential. Primary prevention through health education and social reforms is also needs to be addressed.

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