To conclude, optimum management of such cases is still a matter of debate, but prenatal evaluation and management is associated with improved survival. Postnatally we should follow conservative approach for few weeks to give enough time for the lymphatics to heal and develop collaterals [6]. Refractory cases would require additional therapy.

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Stumped by Potassium: A Rare Case of Familial Pseudohyperkalemia

Hyperkalemia is a common electrolyte disturbance requiring emergent intervention to avoid potential fatal arrhythmias. Pseudohyperkalemia should be kept in mind in the absence of symptomatology and other associated laboratory abnormalities. We present a rare case of pseudohyperkalemia detected incidentally during evaluation of a child with acute respiratory infection. Familial pseudohyperkalemia is an asymptomatic condition that is detected incidentally during evaluation if serum is stored below room temperature prior to testing. It is characterized by spuriously high serum potassium levels due to cold-induced 'passive leak' of red blood cell (RBC) potassium ions into plasma [1,2]. *ABCB6* gene (2q36) has been identified as the causative gene of this rare condition [3].

A 2-month-old baby, first born of non-consanguineous parents, presented to a peripheral health care setup with history of cough and fever for 2 days with some lethargy and refusal to feed. On examination, the infant was found to have mild tachypnea and tachycardia and was admitted for monitoring and supportive therapy. Hematological and biochemical parameters were sent to rule out sepsis and dyselectrolytemia. All parameters were within normal limits except for the elevated potassium levels of 6 mEq/L. In view of high potassium levels, repeat sample was sent, which had serum potassium values of 6.8 mEq/L. The ECG did not show signs of hyperkalemia. Echocardiography revealed a structurally normal heart with good biventricular function. However, chest *X*-ray showed diffuse non homogenous opacities suggestive of bronchiolitis and a large homogenous opacity silhouetting the left cardiac border with a linear translucency surrounding it. Tumor lysis syndrome was initially suspected to be the cause of hyperkalemia but computed tomography of chest revealed that this anterior mediastinal mass was an unusually large, hypertrophied thymus gland and not a malignant mass. During PICU stay, baby remained asymptomatic but continued to have hyperkalemia. There were no dysmorphic features or abnormal genitalia suggestive of any recognizable genetic syndrome. Baby was worked up further with plasma renin activity and aldosterone levels for possibility of pseudohypo-aldosteronism. However, all investigations were within normal limits. Common causes of pseudohyperkalemia (cell lysis, extreme leukocytosis or thrombocythemia, or use of EDTA anticoagulant) were ruled out.

Clinical exome sequencing was done to rule out pseudohypoaldosteronism type II; as common causes had been excluded for the cause of hyperkalemia. It

revealed pathogenic heterozygous variation (c.592G>T, p.Gly198Trp) in *ABCB6* gene, which is associated with Autosomal dominant familial pseudohyperkalemia type 2. This is a benign disorder associated with temperature-dependent anomaly in red cell membrane permeability to potassium that leads to high *in vitro* potassium levels in samples stored below 37°C [2]. The diagnosis was confirmed by doing parallel laboratory assessment of serum potassium levels incubated at 37ºC and 4ºC which revealed normal potassium levels at 37ºC (4.8 mEq/L) and hyperkalemia at 4ºC (6.0 mEq/L). Maternal and paternal serum sample did not reveal any abnormality.

Sampling errors including collection and handling may give rise to spuriously high potassium [4,5]. Pseudohype kalemia may also be caused in the presence of leukocytosis and thrombocytosis [6,7].

Inherited defects in RBC membrane structure are rare causes of pseudohyperkalemia. Two common inherited defects in RBC membrane structure that predispose to pseudohyperkalemia include Familial pseudohyper-kalemia and Dehydrated hereditary stomatocytosis.

Familial pseudohyperkalemia is inherited as an autosomal dominant trait caused by heterozygous variant in *ABCB6* gene. This genetic anomaly causes increased *in vitro leak of* potassium from erythrocytes to plasma/serum when blood is exposed (*ex vivo*) to temperatures below normal body temperature (37 °C). It is a benign condition with excellent prognosis and patients reported with this condition remain asymptomatic and this is usually an incidental finding.. No treatment is required for this condition. However, correct diagnosis is important for prognostication and to avoid needless evaluation for more sinister causes. Serum potassium levels in some patients with familial pseudohyperkalemia variants show relatively large abnormalities on storage below room temperatures; therefore affected individuals are not suitable candidate for blood donation.

This case report emphasizes the importance of evaluation for rare causes of hyperkalemia once the common causes have been excluded to ensure early and appropriate management if indicated and avoid unwarranted treatment for benign conditions.

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