
Preoperative adrenal insufficiency in a neonate with congenital heart disease surgery

Sir,

A 12-day-old male baby was brought to hospital with complaints of bluish discoloration of the body, difficulty in breathing, and increased swelling of both the lower limbs for 3 days. He was a full-term vaginal delivery with birth weight of 3.2 kg. The baby had an uneventful course and was discharged after 2 days of hospital stay. The baby was apparently alright for 10 days at home. On the 11th day, he developed respiratory distress with bluish discoloration of the lip and nails. Local pediatrician suspected congenital heart disease (CHD); hence, the baby was referred for further management.

On physical examination, his weight and height were 3.2 kg and 48 cm, respectively. He was tachypneic, irritable, and his oxygen saturation (SpO₂) was 45%–60%. Peripheral pulses were feeble, and his lips were blue. The baby had bilateral pedal edema.

S1 and S2 heart sounds were normal, and ejection systolic murmur was heard at the left sternal border. Along with routine preoperative investigations, blood culture, serum C-reactive protein, and serum cortisol levels were sent. Ultrasonography of the abdomen showed right adrenal mass suggestive of adrenal

hemorrhage (AH) [Figure 1]. His two-dimensional echocardiography revealed d-transposition of the great arteries, small ventricular septal defect (VSD), and small patent foramen ovale. His initial arterial blood gas (ABG) had very high lactates. He was started on infusion prostaglandin E1 (PGE1) and injection dopamine at 5 µg/kg/min. The baby was intubated and ventilated in the pediatric cardiac intensive care unit (PCICU). He had persistent hypotension, which was not improving to fluid boluses and injection dopamine. Hence, intravenous noradrenaline 0.04 mcg/kg/min was started. In spite of all the above measures, his clinical condition deteriorated; hence, emergency balloon atrial septostomy was done. SpO₂ improved to 75%–80% and lactates transiently came down to 8.0, but the effects were not sustained. He started desaturating again and his serum lactates increased to 15. His serum cortisol level came very low (9.6 mcg/dl). In view of the above ultrasonography findings, clinical picture, and serum cortisol level, he was started on injection hydrocortisone 50 mg/m²/day. Gradually, his clinical condition improved, but he underwent emergency arterial switch operation (ASO) + VSD closure on the same day. The procedure was uneventful. In the immediate postoperative period, the baby had hypoglycemia and borderline hemodynamics after cardiopulmonary bypass, which settled in few hours. He was shifted to PCICU with stable hemodynamics at the end of the procedure. Repeat serum cortisol after 48 h of surgery was 4.7 mcg/dl. He was edematous in the postoperative period but remained hemodynamically stable. Injection hydrocortisone was continued, and serum cortisol levels were monitored. Dose of hydrocortisone was adjusted according to serum cortisol levels.

This case was unique as the child was in shock, was on multiple inotropes, and was not responding to treatment. We suspected adrenal insufficiency (AI) because of the above presentation, investigated accordingly, and started on appropriate treatment for AI. The child showed

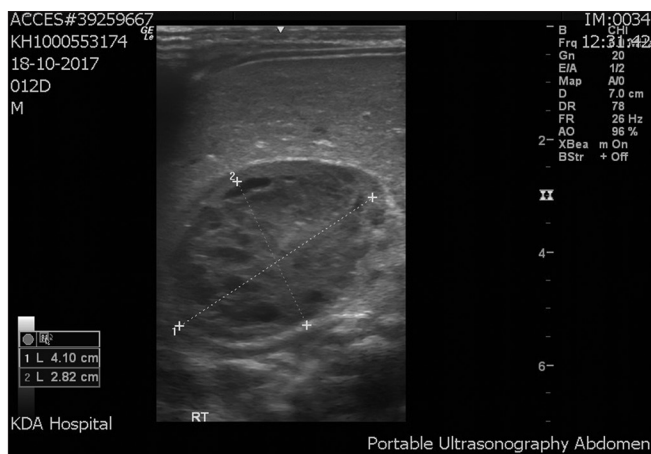


Figure 1: Adrenal hemorrhage on ultrasonography

dramatic improvement after initiating treatment and was sent home after an ASO with a total of 20 days of hospital stay.

Clinical presentations of AH vary from asymptomatic newborn to severe shock. The overall incidence of AH is very low (1:2000) in newborn and infants. AH is generally associated with traumatic vaginal delivery, birth asphyxia, severe sepsis, and prolonged hypotension.^[1] Management of AI in the presence of CHD is very challenging because of the presence of congestive heart failure.

Fever is the most common presentation (50%–70%) with AH, and it may or may not be associated with AI.

Tachycardia has been reported in approximately 40%–50% of patients, and without aggressive therapy, it may progress to shock. Orthostatic hypotension is present in approximately 20% of patients with extensive hemorrhage. Skin hyperpigmentation has been reported, and its presence indicates late recognition of AI in association with AH. Undiagnosed AH is very fatal if not treated appropriately. Even with maximum effort and appropriate treatment, mortality rate is around 15%.^[2]

In our case, the baby had bilateral pedal edema and hypotension, which was not responding to volume replacement, inotropes, and other modes of treatment. This prompted us to investigate for AI. Serum cortisol level along with abdominal ultrasonography helped us confirm our provisional diagnosis. Early diagnosis and treatment of AI are necessary for better outcomes in the presence of varying compounding factors such as CHD and heart failure.

Severe illness, anesthesia, and surgery activate hypothalamopituitary adrenal axis, leading to increased levels of corticotropin and cortisol levels.^[3] This response will be inadequate in the presence of AI. There are many treatment protocols for treatment or replacement of cortisol insufficiency. However, consensus paper by Salem *et al.* is widely accepted for deciding the dose and duration of corticosteroid supplementation.^[4] Dose and duration depend on the level of stress, for example, for minor stress – a total dose of 25 mg, for moderate stress – 50–75 mg, and for major stress – 100–150 mg of hydrocortisone, or its equivalent for 3 days is recommended.

Acknowledgement

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand

that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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