

Congenital Adrenal Hypoplasia Presenting as Severe Respiratory Failure

Congenital adrenal hypoplasia is rare with an incidence of 1 in 12,500 life births. Links between early adrenal insufficiency and adverse respiratory outcome are scarce in the literature.

A term male infant was born by a normal delivery. His birth weight was 3,400 g along the 50th centile. Respiratory symptoms were present at birth, requiring about 30% of incubator oxygen for 3 days. His chest X-ray was normal. He developed mild jaundice and he was discharged home weighing 3,160 g. He was readmitted after 10 days with tachypnea, jaundice, addisonian skin hyperpigmentation, and vomiting. Weight was along the 10th centile. He was alert with periods of drowsiness and bouts of vomiting. His bilirubin was high with normal ALT, AST and elevated GGT: 842 U/L, LDH: 1029 U/L and CK: 1682 U/L, Hypoglycemia (Glucose: 30 mg/dL) was evident only once. His acid balance showed a metabolic alkalosis. Ammonia level was high (ammonia: 113-173 μ mol/L). Urea cycle defects and glycosylation disorders were excluded. Rest of metabolic screen was normal.

A synacten test established the diagnosis of adrenal insufficiency as his clinical condition deteriorated, with convulsions, circulatory collapse, drowsiness and respiratory failure together with progressive hyponatremia, hyperkalemia and low urea. He was intubated and vigorous treatment with hydrocortisone 25-50 mg/m²/d together with 9 α -fluoro-hydrocortisone 0.1mg/d and NaCl supplements were introduced. He showed a dramatic improvement being extubated after 24 hours. Special investigations excluded

congenital adrenal hyperplasia. Plasma renin; 759 pg/mL (NR: 2.4-65.6) was elevated and plasma aldosterone: 9 ng/dL (NR: 5-132) was low. MRI showed normal adrenal and hypothalamo-pituitary glands.

Acute respiratory failure has been described once before(1) as clinical feature of congenital adrenal hypoplasia, although there is reference in the literature of oxygen dependency(2). Our patient although born at term, by normal delivery, developed mild respiratory distress, and by the time of acute illness the respiratory symptoms were prevailing without an obvious underlying lung pathology.

Congenital adrenal hypoplasia although a rare condition should be suspected in case of acute respiratory failure, electrolyte disturbances and/or hypoglycemia in every severely ill neonate, and blind treatment should immediately initiated while awaiting investigations results.

An X-linked mode of inheritance as part of contiguous gene syndrome, associated with Duchenne muscular dystrophy and/or glycerol kinase deficiency(3) is the most probable mode of inheritance in our patient due to his elevated creatine kinase. Other modes of inheritance such as autosomal recessive and X-linked mode have also been described(4).

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Psychogenic Diarrhea

We read with great interest recent communication by Bhatia, *et al.*(1). We greatly appreciate the conclusion drawn by the authors that psychogenic sneezing must be kept in mind in children to avoid unnecessary investigations. We were thus tempted to share our experience in a child with psychogenic diarrhea who responded to behavior therapy within one month.

An 8-year-old male child was brought by her anxious mother with complaints of increased frequency of stools from last 6 months especially after each meal. There was no history of any abdominal distension. The stools were semisolid in consistency but were not bulky or foul smelling. There was no blood or mucus. Physical and systemic examination was non-contributory. His anthropometric data were within normal centiles. Stool microscopy and pH were normal. Stool were also negative for reducing sugars. USG abdomen was also normal. A detailed history revealed other behavior problems in the child such as bruxism, tearing books and beating other classmates in school. He also during his OPD visits dismantled all the articles kept in

the consultation chamber. History revealed the birth of his young brother about 15 months back after which all the attention of the mother has shifted away from him, resulting in sibling jealousy and resulting aggressive behavior as an expression of anger towards her mother. By defecating after each meal, he was able to draw the attention of her mother resulting in secondary gain and persistence of symptoms. The performance of the child in the school also deteriorated further aggravating the symptoms. The mother was counseled about the nature of illness, its onset and perpetuation. A meeting was held with teacher of his school who was also counseled with the problem in the child. He was treated with anxiolytic drugs, aversion therapy and reward on symptomatic improvement. His symptoms improved within a week and disappeared by 3rd week. He performed satisfactorily in yearly exams.

Non-organic recurrent abdominal pain (functional) is a common complaint in children(2). Irritable bowel syndrome is also a well-recognized entity in children presenting commonly as alteration of bowel habits(3). However, psychogenic diarrhea is uncommonly reported in children(4). Early anticipation of the condition and identification of the psychogenic stressor prevented us to