and it becomes an area of anatomical weakness when there is a foreign body inside the abdomen. There are five ridges in the peritoneum that line the lower part of the anterior abdominal wall, which converge in the midline at the umbilical region. Peristaltic activities can thus direct foreign bodies in the peritoneal cavity toward the umbilicus and push it to come out (9). Migration of the lower end of the shunt catheter is an infrequent problem, which occurs without any recognizable cause. The reported incidence of distal shunt migration is 10% (11). Of all the sites of distal end migration reported so far umbilicus is very rare.

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LETTER TO EDITOR (VIEWERS CHOICE)

CONGENITAL ADRENAL HYPERPLASIA IN A MALE CHILD

Manju Mittal, Kumar Ankur, Praveen Kumar, B. Rath

An one and half month old male baby was brought in cardio respiratory arrest with a history of noisy breathing for 3 days and several episodes of vomiting and watery loose motions for 1 day. Baby was immediately resuscitated. His birth weight was 2.5 kg and perinatal course was uneventful. There was one previous hospitalization at day 21 of life with acute gastroenteritis and sepsis. Physical examination revealed an emaciated baby of 1.75 Kg (< 3 rd centile) with normal systemic examination and genitalia. Investigations revealed normal hemogram, hypoglycemia (Blood sugar = 24mg/dl), positive CRP, hyponatremia (serum sodium of 118 mEq/dl) and hyperkalemia (Serum potassium = 6.5 mEq/L). These investigations lead us to suspicion of Congenital Adrenal hyperplasia (CAH). Steroid levels were done which showed Serum Cortisol to be low (25.13, normal range = 28-662 nmol/l) and Serum 17 Hydroxy Progesterone to be high (8800.0 ng/dl, normal: <100 ng/dl) thus confirming the diagnosis of 21 hydroxylase deficiency. The child was managed with antibiotics, Fludrocortisone and hydrocortisone. Child improved, gained a weight of 500 grams over 10 days of hospital stay. Serum electrolytes normalized (Serum sodium of 140mEq/L and serum potassium of 5.5mEq/L) and child was discharged. At 3 months of follow-up he weighs 3.2 kg and electrolytes were normal.

Congenital adrenal hyperplasia is a group of autosomal recessive disorders resulting from the deficiency of one of the five enzymes required for the synthesis of cortisol (1). This disease is easily clinched in female newborns where the ambiguous genitalia are present, but the diagnosis in male newborns is often overlooked as was in our case. He was admitted at 21 days of life and treated as acute gastroenteritis with sepsis and diagnosis of CAH was missed. There are three types of CAH due to 21-hydroxylase deficiency: classical salt wasting disease which is most severe, both cortisol and aldosterone are deficient, classical simple virilizing disease in which adequate levels of aldosterone are synthesized but adrenal androgens are elevated and non-classical disease in which adrenal androgens are mildly elevated leading to signs of androgen excess after birth. Progressive weight loss, anorexia, vomiting,

dehydration, weakness, hypotension, hypoglycemia, hyponatremia and hyperkalemia are the presenting features of classical salt wasting disease (2-3).These symptoms first develop in the affected infant at around 2 weeks of age and if untreated result in shock and death in few days. Prenatal androgen excess in classical disease, leads to development of ambiguous genitalia in affected females while males appear normal at birth Almost all of these symptoms were present in our case. Treatment consists of glucocorticoid (hydrocortisone 10-15 mg/m2/d) and mineralocorticoid (Fludrocortisone 0.05-0.2 mg/d) replacement and salt supplementation (3). Surgical management of ambiguous genitalia is done for significantly virilized females between 2-6 months of age. (3)

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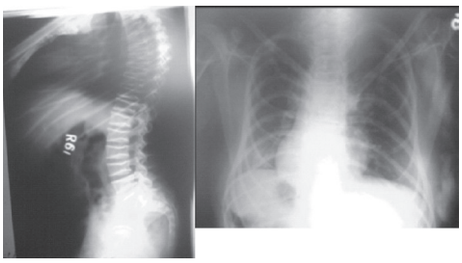
SPOT DIAGNOSIS (IMAGE GALLERY)



Short Stature - A skeletal dysplasia

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A 15 years old girl came to our hospital with short stature. She had a short trunk with upper segment to lower segment ratio of 0.77:1.We worked out on the lines of disproportionate short stature with short trunk. She was normal at birth and first presenting feature was in second decade of life. She had coarse faces, intelligence quotient of 87, bilateral sensorineural deafness, clear cornea, progressive orthopedic anomalies from last 3-4 years, and change in behavior with increase in aggression from last 3 years, disturbed sleep and changing speech from last 2 years. X-rays are depicted in the figure.

What is the diagnosis?

Answer: X-rays are suggestive of Mucopolysaccahroidosis, {MPS}. There was increase in urinary excretion of heparin sulphate. The overall clinical scenario and biochemical markers were suggestive of MPS III {Sanfilippo disease}.

Mucopolysaccahropidosis type III is an autosomal recessive disorder, caused by deficiency in one of the four enzymes involved in the lysosomal degradation of the glycosaminoglycan- heparan sulphate. On the basis of enzyme deficiency there are four different biochemical subtypes, MPS III A, B, C and D with excessive excretion of Heparan sulphate in urine in all these types. Phenotype variations are less common in Sanfilippo than other types of MPS. A milder type may appear totally normal at birth. There is severe progressive central nervous system {CNS} involvement with mild somatic changes. Such disproportionate involvement of the CNS is unique of Sanfilippo. Delay in the diagnosis of MPS III usually occurs because of mild physical features, slow progression of severe CNS involvement and hyperactivity unlike other forms of MPS. Clinical course can be divided into three phases. First phase starts between 1 and 4 years, initially child is normal, later on there is developmental delay. Second phase starts around 3-4 years with severe behavior problems, progressive mental deterioration and dementia. In the third phase behavior problems decreases, there is motor retardation, spasticity and swallowing difficulties. Death usually occurs in 2-3 decade of life, although survival in the fourth decade has been reported {1}.

Presumptive diagnosis is on the basis of clinical and radiological features. Urine screening is done by Berry spot and Acid Turbidity tests. But screening tests may be falsely negative in Sanfillipo. Accurate and confirmed diagnosis is made by 2-dimensional electrophoresis, NMR spectroscopy {2}. Further quantitative estimation of