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Galactosemia

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Galactosemia is an inborn error of metabolism that usually produces clinically recognizable illness within the first few days of life. This disorder is frequently overlooked in the assessment of the sick neonate with features of sepsis despite the fact that a simple urine test for reducing substance can quickly lead to the diagnosis. An example of early neonatal diagnosis of galactosemia is reported here.

Case Report

A five-day-old breast fed male neonate born to non-consanguineous mother was admitted with history of jaundice and vomiting of 3 days duration. Family history revealed that a female infant has died of persistent jaundice at the age of 55 days and the cause was not established. Three other sibs are healthy.

On examination, the neonate was afebrile, lethargic, icteric and had a soft liver palpable just below the costal margin. There was no splenomegaly. Bilirubin level was 14 mg/dl (2 mg/dl direct reacting) and the blood sugar was 120 mg/dl. There was no blood group incompatibility. Sepsis was suspected and cultures of blood and urine were sent. Urinalysis done after intravenous fluids, was positive for reducing substances. After admission, enteral feeding was stopped. With intravenous fluids, antibiotics and phototherapy, he showed clinical improvement. Phototherapy was stopped on the 8th day, when serum bilirubin levels registered a significant drop. Blood and urine cultures were sterile. Antibiotic and intravenous fluids were stopped on 12th day and breast feeding was resumed. A repeat urinalysis done after breast feeding was reinitiated, was positive

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for reducing substances, but negative for glucose.

Based on the family history, clinical and laboratory parameters, a tentative diagnosis of galactosemia was made.

A filter paper blood sample obtained by heel prick was sent to New England Regional Screening Program, Massachusetts where the diagnosis of galactosemia was confirmed. The concentrations of galactose and galactose 1 phosphate (>50 mg/dl) were markedly elevated. The normal newborn level for either of these metabolites is not greater than 6 mg/dl. The absence of erythrocyte galactose phosphate uridyl transferase (GALT) activity by the Beutler spot enzyme assay established the diagnosis.

Breast feeding was stopped on the 14th day of life and the he was started on a commercial milk substitute containing soy protein isolate with dextrimaltose as the carbohydrate source.

The baby is now 6 months of age, is thriving well and is developmentally normal. He has no signs of liver disease and his ocular lenses are clear with no evidence of cataracts.

Discussion

The salient clinical features of this galactosemic neonate included jaundice, hepatomegaly, lethargy, poor feeding, vomiting and continuing weight loss, suggestive of septicemia. By the age of 10 to 14 days, many galactosemic neonates may develop septicemia usually with *E. coli*, unless dietary treatment has been started(1).

Recent observations indicate that the early hyperbilirubinemia of the galactosemic neonates may be predominantly indirect reacting (unconjugated) as it was in this case. This can still be explained by the fact that the bilirubin conjugating mechanism may be inhibited by Galactose-1-phosphate or other toxic metabolites that accumulate in galactosemic neonates(2).

The diagnosis is confirmed initially by urine screening tests and later by specific tests of blood to establish the presence of increased concentration of galactose and the type of enzyme deficiency. Galactose phosphate uridyl transferase activity (GALT) was deficient in the present neonate. The other two entities are galactokinase deficiency and uridine diphosphate galactose-4-epimerase deficiency. Once the diagnosis is established, a non lactose formula must be substituted and the galactosemic neonate must remain on a diet that excludes milk and milk products for the rest of his life. The role of uridine therapy as an adjunct to milk substitutes is currently under investigation.

Galactosemia has been recognized in Indian children, mostly after the establishment of irreversible changes like cataract, cirrhosis and mental retardation(3-5). These reports highlight the pitfalls in the early diagnosis of galactosemia and the need for simple urine screening test to rule out galactosemia in neonates with findings of sepsis and persistent jaundice.

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Hydrops of the Gall Bladder in a Child with Wilson's Disease: A Rare Association

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Hydrops of the gall bladder is an uncommon condition in childhood. It has been described in preterm infants and newborns secondary to parentral hyperalimentation, sepsis as well as in normal newborns(1-3), older infants and children. It has been reported in association with upper respiratory tract infections, gastroenteritis and enteric infections, leptospirosis, scarlet fever, lymphadenitis, and mucocutaneous syndrome, polyarteritis nodosa and familial mediterranean fever(4-8).

We report a case of asymptomatic hydrops of the gall bladder in an 11-year-old Saudi Girl with hepatic cirrhosis secondary to Wilson's disease.

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Case Report

A 12-year-old Saudi girl was admitted to hospital for re-evaluation because of recurrent hematuria, abdominal pain and occasional joint pains and ankle swellings.

She had been evaluated extensively in another hospital in Riyadh an year ago and referred for further evaluation and management in the United Kingdom, with a provisional diagnosis of glomerulonephritis. In the United Kingdom in addition to her suspected renal disease she was found to have a disturbed clotting profile, raised hepatic enzymes and a positive HbsAg screening test. Because of her clotting disturbances, liver and renal biopsies were not done, and a provisional diagnosis of chronic active hepatitis with possible immune complex renal disease was made. An abdominal ultrasound at that time was reported as showing a normal gall bladder.

There was no history of jaundice or neurological symptoms. Her parents are cousins and she has 5 siblings who are living. There was no family history of liver disease or neurological disease.

On physical examination she was pale, without jaundice or obvious Kayser Fleisher (KF) ring. Her weight was 31 kg and height was 128 cm. She had pitting edema