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Glucose-6-Phosphate Dehydrogenase Deficiency with Bilateral Cataract

Various studies from India have shown association between glucose-6-phosphate dehydrogenase (G-6-PD) deficiency in erythrocytes and age-related cataract(1,2). However, its association with childhood or congenital cataract has not been well studied. An 8-year-old boy presented to us with history of bilateral cataract developing at age of 2 years, which was operated at 3.5 years; and pallor requiring 3 blood transfusions at ages 3.5, 4.5 and 8 years. He had received multiple antibiotics including co-trimoxazole for various pharyngeal infections. His mental development was normal, and there was not history of seizures. Family history was negative for early-onset cataract or hemato-logical disorders. On physical examination, patient had bilateral aphakia; mild icterus and pallor. There was no dysmorphism, signifi-cant lymphadenopathy or hepatospleno-megaly.

Laboratory investigations revealed hemoglobin 8.4 g/dL, WBC count 6000/mm^3, platelets 386,000/mm^3, MCV 101.6 fl, MCH 30.4 pg, MCHC 30 g/dL, red blood cell (RBC) count 2.76 million/mm^3 and absolute reticulocyte count 11%. Peripheral smear showed macrocytic erythrocytes with polychromasia. Total bilirubin was 3 mg/dL; direct bilirubin 0.7 mg/dL; serum creatinine 0.6 mg/dL; liver enzymes normal; hemoglobin electrophoresis normal; direct Coombs' test negative; and bone marrow examination revealed erythroid hyperplasia. Methemo-globin reduction test showed defi-ciency of G6PD enzyme in erythrocytes. On quantitative analysis, G6PD was 93.7 mU/10^9 erythrocytes (normal: 118-144 mU/10^9 erythrocytes). Screening for galactosemia in view of early-onset bilateral cataracts was negative with absence of reducing substances in urine.

The patient had congenital nonspherocytic hemolytic anemia due to G-6-PD deficiency with episodic drops in hemoglobin most probably due to cotrimoxazole exposure. G6PD deficiency results in reduced glutathione activity in the lens, making the lens more prone to oxidative damage and resultant aggregation of lens proteins. The development of bilateral cataract at the age of 2 years in a develop-mentally normal child may have been due to the

oxidative stress in the lens from G-6-PD deficiency.

There have been three previous reports of early-onset childhood cataract and G-6-PD deficiency(3-5). It, however, remains unclear as to why certain G-6-PD variants predispose to cataract formation. In view of the epidemiological studies suggesting the association of G-6-PD deficiency and role of oxidative stress in causation of adult human cataract, and some sporadic reports of childhood cataract with G-6-PD deficiency without any other obvious cause, it is reasonable to consider excluding G-6-PD deficiency in children with cataract.

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Urethral Catheter Knotting in Preterm Neonates

Feeding tubes have been universally used as urethral catheters in children for several decades. Though a safe procedure, it presents special risks in infants and children. Here we report a 28 weeks preterm baby who developed a urethral catheter knot and look into measures to avoid this unpleasant complication.

A 28-week preterm baby was admitted to our neonatal intensive care unit (NICU). Due to surfactant deficiency lung disease, he was given surfactant and ventilated. He developed urinary retention and hence was catheterized with a size 4F feeding tube. When his clinical condition improved, it was decided to remove the catheter. While it was being removed, resistance was felt halfway through the procedure. When the catheter tip was out, we were shocked to see a knot in the catheter (*Fig. 1*). Fresh bleeding was noticed at the urethral meatus, which stopped with gentle pressure. His urine output has been adequate since then.

Urethral catheterization is a frequently performed pediatric procedure. In children, Foley's catheter and feeding / nasogastric tubes are used. Urethral trauma and infections are well known complications. Catheter knotting, a rare complication is seen in children 2 years of age or younger following the use of feeding tubes as urethral catheters. The incidence of knotting is reported to be 0.2 per 100,000 catheterizations(1).

The widely accepted hypothesis for