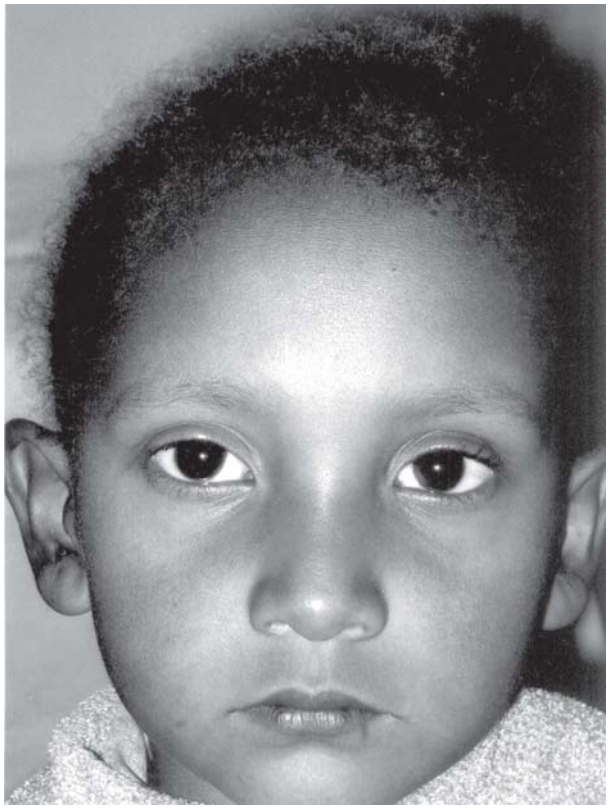

FIG. 1 *Close up of scalp showing sparse, hypopigmented and tightly coiled hair. Also note the sparse eyebrows.*

appears in the first few years of life(1,2). Microscopically the hair shaft exhibits an elliptical cross-section, an axial rotation of 180° on its axis and kinking.

Body hair is generally not affected in autosomal dominant variety but is short, light, relatively sparse and rarefied in autosomal recessive type(3). Eyebrows may also be involved. Woolly hair may be associated with palmoplantar keratoderma (Naxos disease) or cardiomyopathy (Carvajal Syndrome)(4).

Our patient presented with woolly hair after infancy along with sparse body hair, speech delay and learning difficulties. There are very few case reports of woolly hair from India(1,2). No effective treatment is available.

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G6PD Screening: Is it Really Required?

The author recommends a universal neonatal screening program for glucose-6-phosphate dehydro-genase (G6PD) deficiency in India because it is common and causes significant morbidity and mortality due to neonatal jaundice and acute hemolytic crisis(1). Since neonatal jaundice is the commonest presentation of G6PD deficiency, the screening test result and the definitive quantitative test result has to be available and communicated to parents very early to be of any use. Even if G6PD deficiency is detected, the parents will be advised that their newborn baby is at risk for neonatal jaundice, and should be brought early to hospital if he becomes yellow. We should anyway be giving this advice all newborns at discharge. The management of a G6PD deficient newborn with jaundice is the same as that for any other baby with neonatal jaundice.

Acute hemolytic crisis is highly unpredictable and is usually seen after infections or accidental ingestion of chemicals or drugs. Majority of persons with G6PD deficiency never experience a crisis in their whole life. Advice to avoid drugs and chemicals will be of no use in infection induced hemolysis or accidental ingestions, which are more common.

When we communicate news of G6PD deficiency to parents; we might cause a lot of anxiety in the family. The majority of G6PD deficient persons will not have any clinically significant problem, and we will not be offering any preventive or curative therapy. We should analyze what benefit is offered to patients by detecting them to be G6PD deficient before recommending a nationwide G6PD detection program. That it can be done does not imply it should be done.

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REPLY

The purpose of a screening program is to identify individuals not identified by routine physical examination. Presently, approximately half of all childbirths in India occur at home, and a significant proportion of all institutional deliveries are not attended by a pediatrician. Hence the quality of advice at discharge of the newborn is highly variable. In the absence of targeted advice to parents from trained counsellors, the risk of G6PD deficient infants suffering from pathological jaundice and kernicterus remains very high. Other countries in South Asia (Malaysia, Philippines, Taiwan, Hong Kong and Singapore) with a high prevalence of G6PD deficiency have been able to reduce the morbidity and mortality associated with this enzymopathy during the neonatal period and in later life after the introduction of a neonatal screening program(1).

The drugs and chemicals triggering haemolytic crises are used commonly (over the counter or prescription), and the risk of exposure to these triggers later in childhood or adult life is very high. Targeted advice to those identified by neonatal screening, though not completely eliminating the risk of accidental exposure, will substantially reduce the episodes of hemolytic crisis in these individuals. The cost of introducing the screening program can be easily justified by the savings in the medical care as a whole for these patients. Regarding the issue of anxiety and distress to the family and patient, the benefits from being watchful and avoiding all triggers outweigh the psychological harm.

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