



FIG.1 A child with *Hypomelanosis of Ito* showing multiple whorled like hypopigmented lesions present over abdomen, chest and back.

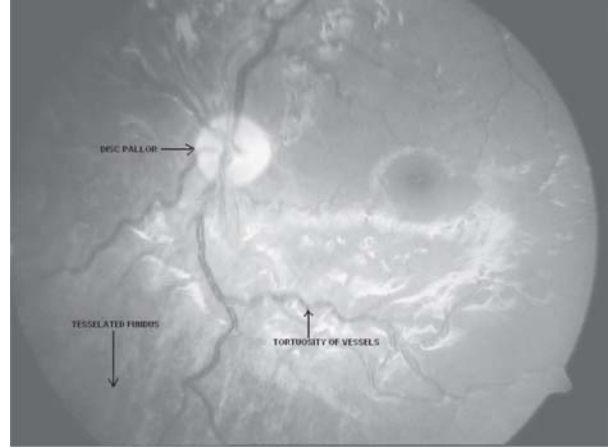


FIG.2 Fundus showing pale disc, tortuous vessels around the disc, radial patchy streaks of hypopigmentation and tessellated background.

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Familial Woolly Hair Disease

A five year old male presented with progressive curling of scalp hair since one year of age. It was a full term product of a non consanguineous marriage with no known or distant African ancestry. Child had a normal motor development; however, speech was delayed and he encountered learning difficulties once he was admitted to school at the age of three years. None of the other family members had similar hair type.

On examination, the hair all over the scalp were found to be tightly coiled (curl diameter being ~ .5

cm), thin, dry, brittle, hypopigmented and sparse. The eyebrows were also sparse (**Fig. 1**). Underlying scalp was normal. Nutritional status was normal. Other ectodermal tissues viz. nails, skin, eye were normal. There was no other apparent congenital abnormality. Systemic examination was normal. Microscopic examination of hair revealed axial rotation of hair shaft. Echocardiography did not reveal any evidence of cardiomyopathy.

Woolly hair syndrome is a rare congenital abnormalities of scalp hair described in Asian and Caucasians. It is characterized by tightly coiled hair involving the entire or part of scalp in an individual of non Negroid origin. It manifests either at birth or



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.