FIG. 1 *Shiny, pearly white papules on chin, forehead, cheeks and nose.*

Epstein pearls); when they occur on the alveolar margins, they are termed Bohn nodules. Main differential diagnoses of milia include sebaceous hyperplasia (more yellow), molluscum contagiosum (dome-shaped papules with central umbilication), miliaria crystallina (skin-colored pin-pint clear vesicles), bacterial and candidal lesions, and transient neonatal pustular melanosis (superficial vesiclopustules that are present at birth, rupture within 24-48 hours and heal with hyperpigmented macules). No treatment is required for neonatal milia as these spontaneously resolve in a few weeks.

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Leprechaunism (Donohue Syndrome)

Born at term with a birth weight of 1700 g to consanguineous parents, this 4½-month-old girl weighed 2900 g with a length of 52 cm and head circumference of 34 cm. She appeared markedly emaciated, and had hirsuitism, thickened skin with patchy hyperpigmentation, coarse facial features, sunken cheeks, pointed chin, flared nostrils, broad open mouth, thick lips, low-set ears, enlarged breasts and clitoromegaly. Her investigations revealed massively enlarged polycystic ovaries, elevated estradiol and testosterone levels, hypoglycemia (blood glucose 27 mg/dL) and hyperinsulinemia (serum insulin of 253 µU/mL). Leprechaunism was diagnosed on the basis of these clinical features and laboratory findings. In view of the poor prognosis and young age, this patient was given symptomatic treatment. She succumbed to pneumonia and septicemia by 6 months of age.

Leprechaunism (Donohue syndrome) is a recessively inherited syndrome of insulin resistance with severe prenatal and postnatal growth retardation, ovarian hyperstimulation, acanthosis nigricans and abnormal facial features, as described above. Over 50 insulin receptor mutations have been described which result in profound insulin resistance and altered glucose homeostasis with fasting hypoglycemia and postprandial hyperglycemia. Almost all affected patients die before two years of age. Other congenital syndromes of insulin resistance are Type A syndrome and Rabson-Mendenhall syndrome in which growth retardation is less severe, and severe symptoms are not seen in early infancy.

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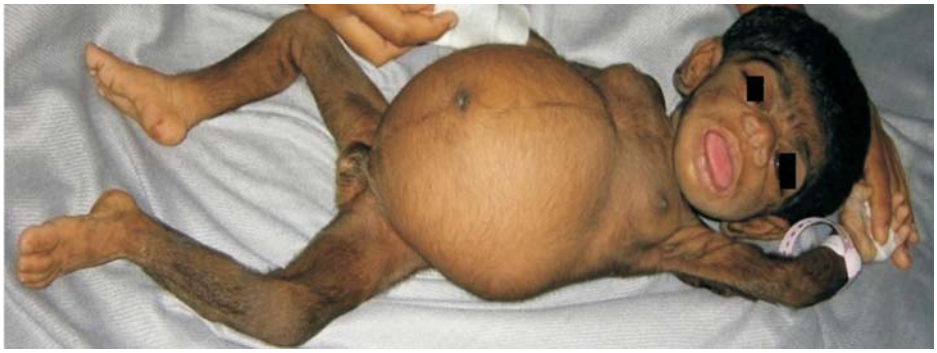

FIG. 1 *Hirsuitism, marked emaciation and coarse facial features in Leprechaunism.*