Monilethrix is a rare autosomal dominant disease of the hair shaft characterized by small nodelike deformities, which result in hair fragility and partial alopecia. Several mutations in the human basic hair keratins hHb1 and hHb6 have recently been reported in this disease.

The morphologic alteration of the disorder is beading of the hair shaft, which causes its breakage at constricted points. Expressivity of the disease appears variable within and between sibships. Some members of an affected family may have very few beaded hairs on their scalp, whereas others have an abundant number of affected hairs. Furthermore, no correlation has been found between the severity of the phenotype and the carried mutation.

The distortion of the hair shaft starts from the hair follicle and is demonstrable along the mature hair shaft. Our observations show that although the distal end of the hair may show no beading, its proximal part will carry the typical anomaly.

Usually the lanugo hair is clinically normal, so an affected newborn will present normal-appearing scalp hair. Only when it is shed during the first few weeks of life does the regrown mature hair show the anomaly. In severe cases the newborn may show a completely bald scalp already at birth and a few remaining hairs are easily “brushed” off. The bald scalp may have the appearance of having been shaved. The differential diagnosis in these cases includes congenital alopecia and ectodermal dysplasia, and there are no hairs available to determine the nature of the hair anomaly.

To establish the presence of severe monilethrix in such cases, we scrape the scalp with a glass slide and collect the epithelial cells with hair rudiments protruding through the epidermis. When the scraped material is examined microscopically, it shows the stratum corneum cells and among them single or paired beads of the typical monilethrix hair.
REFERENCES

This simple procedure makes a scalp biopsy superfluous and permits the diagnosis of monilethrix in a newborn with complete alopecia within minutes.

Fig 3. Scalp scraping shows debris, epithelial cells, and beads of typical monilethrix hair.

Direct all Surgical Pearl submissions to Dr Stuart J. Salasche, 5300 N Montezuma Trail, Tucson, AZ 85750.

Direct all Medical Pearl submissions to Dr Mark G. Lebwohl, Mount Sinai Medical Center, One Gustave L. Levy Place, Box 1048, New York, NY 10029.

What is the castling phenomenon?

Jeffrey D. Bernhard, MD

Answer will appear in the February issue of the Journal.

DECEMBER 2001 IOTA DERMA (#95)

Oblaten is a German word referring to the paper-thin wafers used in religious ceremonies. What dermatologic condition is characterized by Oblaten-like scale, that is, having a thick, sharply defined, single large scale that reveals normal skin when peeled away?

Answer: Oblaten-like scale occurs in pityriasis lichenoides chronica.

REFERENCE