



MONILETHRIX

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Monilethrix is an uncommon disorder that results in breakage of the hair shaft leading to sparse areas of alopecia. The hair appears as "beads on a string" with the breakage points occurring at specific internodes along the hair shaft. Monilethrix develops during infancy and occurs on the back of the scalp, nape of the neck, or the entire scalp. In more severe cases, hair of the eyelashes, eyebrows, axilla, pubic area, and limb hairs may be involved. Patients with monilethrix are also prone to developing **keratosis pilaris** (small bumps on the skin that often appear on the scalp and/or extensor surfaces of arms and thighs) in addition to nonspecific nail deformities such as koilonychia ("spoon nails").

Monilethrix has a strong genetic predisposition and parents with monilethrix will invariably pass on the gene to their children in an autosomal dominant fashion. Family studies have shown that the autosomal dominant form of monilethrix is due to mutations in the type II hair keratin proteins hHb1, hHb3, and Hhb6 that are responsible for the stability and strength of the hair cortex. In some cases, parents without the physical condition may still pass on the gene if the child receives two copies of the defective gene from both parents hence autosomal recessive inheritance. The latter is rare and is associated with defects in a hair protein known as desmoglein 4.

The dermatologist will assess the hair for localized or widespread areas of alopecia and assess the hair shafts with a dermatoscope to localize areas of breakage and constriction points. Hair samples are further evaluated with microscopy to aid in diagnosis.

To date there is no cure for monilethrix. However, there have been reports of spontaneous regressions during puberty and pregnancy. Patients have experienced success and an increase in hair shaft diameter with topical minoxidil 2%. **Steroid preparations, retinoids**, glycolic acids, oral contraceptives, and vitamins can also be used to treat the condition albeit with limited success. Since monilethrix is a lifelong disease, it is important that patients avoid exposing their hair to sunlight, chemicals such as hair dye or bleach, and high heat.

This information has been provided to you compliments of the American Osteopathic College of Dermatology and your physician.

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