

# Treatment of monilethrix with oral minoxidil



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## INTRODUCTION

Monilethrix is an autosomal dominant genodermatosis characterized by hair fragility, keratosis pilaris, and pathognomonic beading of the hair shaft. Hair fragility may lead to hair breakage and the appearance of generalized hair loss.<sup>1</sup>

The gene for autosomal dominant monilethrix has been mapped to the epithelial keratin gene cluster on 12q13,<sup>2</sup> and point mutations have been found in the hair cortex—specific keratin genes *KRT86*, *KRT83*, and *KRT81*.<sup>3,4</sup> Mutations in desmoglein 4 are responsible for an autosomal recessive variant of monilethrix.<sup>5</sup>

Monilethrix has considerable inter- and intrafamilial variations in age of onset, severity, and natural history.<sup>4</sup> Most often, hair is normal at birth and is progressively replaced by short, fragile, brittle hair during the first months of life. Hair breakage secondary to hair fragility may be accompanied by follicular keratosis most commonly on the occiput. Friction from wigs and their adhesives may exacerbate hair loss. Eyebrows, eyelashes, pubic, axillary, and general body hair may be affected along with the scalp.

In most patients, the hair loss persists with little change throughout life. Spontaneous improvement or complete recovery has been reported during pregnancy.<sup>6</sup>

There is no cure for monilethrix. Reduction in hairdressing trauma may diminish weathering and improve severely affected cases.<sup>1</sup> Two women with monilethrix who responded to 6 months of treatment with low-dose oral minoxidil are described.

## CASE SERIES

### Case 1

A 40-year-old woman presented with lifelong fragile, sparse, and thin hair. She had monilethrix



**Fig 1.** Patient 1. Dermoscopic image of the occipital scalp shows numerous broken hairs. Beaded hairs pathognomonic of monilethrix can be seen.

diagnosed in childhood. She reported striking improvement in hair volume and length during each pregnancy but relapsed within 12 months postpartum.

Comorbidities included Crohn's disease and impaired hearing. Her daughter and 2 sons also had monilethrix and suffered from sparse and fragile hair.

On examination, she had brittle, lustreless, fragile, short hair all over the scalp. Her hair turned to powder when rolled between fingers. She was noted to have small keratotic papules on the nape of the neck. There were no nail or tooth abnormalities detected. Dermoscopy (Fig 1) confirmed the diagnosis of monilethrix.

Treatment started with oral minoxidil, 0.25 mg daily. Review after 6 months found significant hair growth with reduced breakage and increased hair volume and length (Fig 2). The patient did not experience any side effects or complication of the treatment. The patient continued on the treatment and is reviewed at 3-month intervals. The initial improvement in hair density has been maintained

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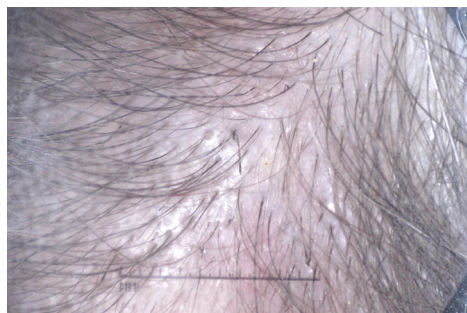
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**Fig 2.** Patient 1. Monilethrix. Lateral and posterior views of the scalp before and after 6 months of treatment with low-dose oral minoxidil.



**Fig 3.** Patient 2. Dermoscopic image of the mid-frontal scalp shows broken hairs. Beaded hairs pathognomonic of monilethrix can be seen.

and no side effects have been noted after 2 years additional follow-up.

### Case 2

A 35-year-old woman with fragile, thin, and sparse hair since childhood secondary to monilethrix presented for assessment and treatment. She had a positive family history of monilethrix affecting her son, daughter, and grandson.

Examination found brittle, lustreless, fragile short hair all over the scalp, keratotic papules on the occipital scalp, and keratosis pilaris. No nail or dental abnormalities were present. Dermoscopy found elliptical nodes separated by narrower internodes (Fig 3).

She started oral minoxidil, 0.25 mg daily. On examination 3 months after the initial consultation, hair shedding decreased significantly. However, the hair density remained unchanged. Minoxidil was increased to 0.5 mg daily. There was a significant improvement in hair density on review at 6 months (Fig 4).

The patient remains on the treatment and is reviewed at 3-month intervals. The initial improvement in hair density has been maintained, and no side effects have emerged after 18 months of additional follow-up.

### DISCUSSION

Monilethrix is a hair shaft fragility genodermatosis. Etretnate is found to reduce hair beading but

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