Primary cicatricial alopecias are a group of rare disorders that lead to permanent hair loss. The first step in managing a patient with a suspected scarring alopecia is to perform a biopsy. The absence of scalp follicular orifices is the clinical sign that suggests a scarring alopecia and the need for a biopsy.

Current treatment selection is based on the predominant cell type (lymphocytic or neutrophilic) seen with light microscopy. It is essential to explain that current treatments relieve symptoms (itching, pain, burning) and signs (erythema, scaling, crusting, pustules), but may not halt progression of hair loss, and hair re-growth is not possible.

Lymphocyte-predominant disorders include lichen planopilaris, frontal fibrosing alopecia, central centrifugal alopecia and pseudopelade (Brocq). Systemic agents are indicated if patients are symptomatic or have clinically active disease, and hair loss is progressive.

First tier treatments include hydroxychloroquine (200mg twice daily) or doxycycline (100mg twice daily). If signs and symptoms persist, then second tier agents include mycophenolate mofetil (0.5gm twice daily for the first month, then 1gm twice daily for five months) or cyclosporine (3 to 5mg/kg per day or 300mg/day for three to five months) or pioglitazone (15 mg daily for three to six months).

Topical treatments include high-potency topical corticosteroids, topical tacrolimus or pimecrolimus, Dermasoothe FS oil, and intralvesional triamcinolone acetonide injections at the active margins of disease.

Neutrophil-predominant disorders include folliculitis decalvans and tufted folliculitis. Repeat culture and sensitivity of pustules or pulled-out hair bulbs or a small scalp biopsy is essential to selecting the optimal oral antibiotic. For staph aureus, we use cephalexin (500mg four times daily for ten weeks) with oral rifampin (600mg daily for ten days). Substitutions include clindamycin (300mg twice daily), trimethoprim-sulfamethoxazole DS (twice daily), ciprofloxacin (750mg twice daily), or doxycycline (100mg twice daily), all given for 10 weeks with rifampin (600mg for ten days).

The above treatments are usually effective in controlling signs and symptoms but may not halt progression of the hair loss since they do not address the underlying pathophysiology. In a recent landmark report, molecular studies of scalp tissue from patients with lichen planopilaris (LPP) revealed a loss of peroxisome proliferator-activated receptor gamma (PPARg), a protein that regulates inflammation and lipid metabolism in the pilosebaceous unit.

This loss leads to a buildup of toxic lipids that generates inflammation and eventual destruction of the hair follicle and sebaceous gland. A similar PPARg deficiency has been found in frontal fibrosing alopecia.

Further study is still needed in central centrifugal cicatricial alopecia and the neutrophilic cicatricial alopecias. The role of PPARg in cicatricial alopecia is supported in mice in which a targeted deletion of PPARg in follicular stem cells causes scarring alopecia.*

These findings suggested that treatment with PPARg agonists may provide a new upstream treatment strategy for patients with LPP and frontal fibrosing alopecia. Pioglitazone and rosiglitazone are glitazones and PPARg agonists that are FDA-approved and used widely to treat type 2 diabetes mellitus as well as atopic dermatitis. Pioglitazone was successful in a 47 year-old male patient with active and symptomatic LPP who had failed all treatments.

The most common side effect of the glitazones is dosage dependent weight gain as a result of fluid retention and edema. Other reported adverse reactions include an increased risk of myocardial infarction with rosiglitazone, CYP P450 drug interactions with pioglitazone.