



# Lichen Planopilaris: Update on Diagnosis and Treatment

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Lichen planopilaris (LPP), a follicular form of lichen planus, is a rare inflammatory lymphocyte mediated disorder. Although the physiopathology is unclear, an autoimmune etiology is generally accepted. Women are affected more than men, and the typical age of onset is between 40 and 60 years. LPP is a primary cicatricial alopecia whose diagnosis is supported in the early stage by both clinical and histopathological findings. Within the margins of the expanding areas of perifollicular violaceous erythema and acuminate keratotic plugs, the histology can show the lichenoid perifollicular inflammation. LPP can be subdivided into 3 variants: classic LPP, frontal fibrosing alopecia (FFA), and Lassueur Graham-Little Piccardi syndrome. With the exception of FFA, the hairless patches of the scalp can be unique or can occur in multiples and can present with a reticular pattern or as large areas of scarring. This condition can have major psychological consequences for the affected patients. The therapeutic management often is quite challenging, as relapses are common after local or systemic treatments. Further research is needed on the pathogenesis, and randomized controlled trials of treatment with scientific evaluation of the results are necessary to appreciate the proposed treatment.  
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Initially described by Pringle in 1895,<sup>1</sup> lichen planopilaris (LPP) is also known as lichen follicularis or follicular lichen planus. Lichen planopilaris is a cutaneous disorder selectively involving hair follicles with a lymphocytic inflammatory process that eventually destroys the follicle; thus, it is a primary cicatricial alopecia. The North American Hair Research Society (NAHRS) classification placed LPP in the lymphocytic group.<sup>2</sup> Usually considered as a rare disease, LPP is the most frequent cause of adult primary scarring alopecia.

LPP has been subdivided into 3 variants<sup>2</sup>: classic LPP, frontal fibrosing alopecia (FFA), and Lassueur Graham-Little Piccardi syndrome. Fibrosing alopecia in a pattern distribution (FAPD) could be added as a fourth variant.<sup>3</sup> Although LPP usually is a clinical diagnosis, it can be a therapeutic challenge and can become a "capillary emergency" as the cicatricial alopecia quickly enlarges.<sup>4</sup> In the early stage, diagnosis is made by both clinical and histopathological findings. In the later (pseudopelagic) stage, no specific sign helps the clinician confirm the diagnosis. LPP is more common in women (60 to 90% of the cases) than in men.<sup>5-7</sup> The age of onset of LPP is frequently between 40 and 60 years. Although LPP was described in a 9-year-old child,<sup>5</sup> it is unusual in children. In India from 1988 to 2000, 3.5% of 87 cases of childhood lichen planus had LPP, and it affected only 0.021% of the total number of new pediatric dermatology outpatients.<sup>8</sup> Caucasian and black patients can be affected, but in Canadians, those of Asian origin seem to be unaffected by this LPP.<sup>7</sup>

## Clinical Diagnosis

The classical form of LPP commonly involves the vertex, but any region of the scalp can be affected. The early classically complete lesions are characterized by a follicular violaceous erythema and acuminate keratotic plugs (Figs. 1 and 2). The lesions are commonly located at the periphery of the bald zone, corresponding to the margin of expanding areas of alopecia. Sometimes perifollicular violaceous erythema can be seen without hair shedding adjacent to the bald areas. Some hair affected by the inflammation process can persist in the center of the bald area (Fig. 3). The affected hairs can sometime be observed within false peripilar casts (migrating tubular flakes corresponding to the parakeratosis). Perifollicular inflammation or hyperkeratosis can be very discreet in some cases, which makes the diagnosis more difficult. Keratotic plugs may be absent. Infrequently, pustules can be observed at the edges of the balding surface, which could mimic folliculitis decalvans. Some tufted hairs may stand witness to previous inflammation. A positive pull test of anagen hairs is commonly present at the margin of alopecia, indicating the disease activity.

After inflammation and hair shedding, atrophic scarring of areas without follicular units replaces all the other lesions. This corresponds to the end-stage of any cicatricial alopecia (Fig. 4). It is, however, important to note that in parallel with the atrophy, there is no dischromy, telangiectasia, or squamous surface. Because it is not the same entity, typical papules of lichen planus are not observed on the scalp. Bald patches can be unique or can occur in multiples. Bald areas can present in a reticular pattern or as large areas of scarring.

Key features that help to make the clinical diagnosis are as follows:

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**Figure 1** Initial little patches of alopecia on the vertex.

- Dermoscopy

In the center of the bald areas (pseudopelagic stage), the cicatricial alopecia lacks follicular orifices. On the margin, the pink-red translucent inflammation is clearly perifollicular, and the keratosis extends along the hair (Fig. 5), with false peripilar casts observed on the hair shaft.<sup>9</sup>

- Functional signs

Most of the patients complain about itching, pain, or burning when inflammation is present.<sup>5,6</sup> This sensation is usually not intense and is not pathognomic for the disease. It is aggravated by heat and also may be associated with seborrheic dermatitis.

- Association

In 17 to 28% of cases,<sup>5-7</sup> to 50% of cases,<sup>10</sup> lichen planus (LP) can be observed on other parts of the body. In our experience, the estimate of LP occurring on other parts of the body in 50% of scalp cases of LPP seems clearly overestimated. We concur with the estimation of scalp involvement in 25% of LP.<sup>11</sup> More rarely, nail involvement with pits or onychorrhexis also may be present,<sup>5,6</sup> and oral or genital mucosa may be involved. If LPP is considered as a pattern of LP, its sex predisposition, evolution, therapeutic options, and effects are extremely different.

LPP also may be present on the face, such as the eyebrows, beard (more rarely in a linear distribution<sup>12</sup>). However, LPP more often



**Figure 2** Peripilar hyperkeratosis.



**Figure 3** Remaining hairs in the cicatricial area.

occurs on the legs or arms,<sup>6</sup> or in axillary and pubic regions. As reported by Mirmirani, et al,<sup>13</sup> 55% of the biopsy samples taken from primary cicatricial alopecia of clinically uninvolved scalp show evidence of the disease. In 28% of cases, seborrheic dermatitis would be associated with LPP.<sup>6</sup>

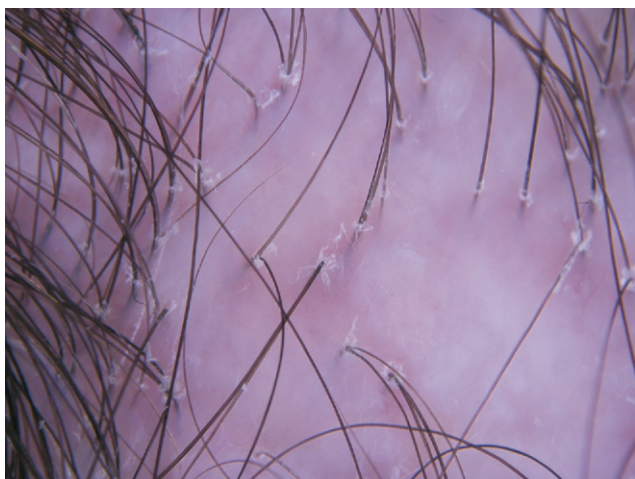
## Evolution

The inflammation tends to resolve spontaneously, leaving whitish atrophic scars of the skin, which lack follicular orifices. The hypothesis is that the inflammation subsides as the putative antigen, the hair follicle, disappeared. Alopecic macules and patches mimic the end-stage of many instances of cicatricial alopecia (pseudopelagic stage). Patches can be of different sizes and shapes, and they are centrifugal and coalescent. This condition can even evolve to affect the entire scalp (Fig. 6).

The course of LPP can be very active and extensive, with a remarkably positive traction test, or slow with a negative traction test. In all cases, the evolution is unpredictable. Both the patient and the doctor ask the vexing question: "When will it end?" There is no long-term study that provides answers about the duration of the disease. LPP sometimes has no apparent ending. LPP is clearly different from LP, which develops, followed by episodes of complete remission.



**Figure 4** A typical patch on the vertex.



**Figure 5** LPP: dermoscopic appearance.

### Clinical Forms

LPP has been categorized into 3 clinical variants.<sup>2</sup>

#### Classical Pattern

Although it affects middle-aged men and women equally, it presents as the centrifugal patches previously described.

#### Frontal Fibrosing Alopecia (FFA)

Initially described in post menopausal women,<sup>14</sup> this particular entity can infrequently appear in premenopausal women (Fig. 7) and more rarely in men (Fig. 8).<sup>15</sup> Found in black or white populations, it seems to be less frequent in Asiatic people. FFA presents as a progressive symmetric bandlike alopecia, affecting the frontal hair line, the preauricular scalp and, less commonly and distinctively, the retroauricular areas. The pale and smooth skin in the affected zones contrasts with the normal skin of the forehead. Even if stabilization seems to occur and progression is relatively slow (average 0.9 mm/month),<sup>16</sup> evolution of this cicatricial alopecia can be widespread, leaving only a band of hair on the top of the scalp, which is described as a “clown alopecia” (Fig. 9). Eyebrows often are affected before the frontal scalp, but clinical inflammation is not observed in the brows. The other vellus or terminal hair of the face can be involved, even the eyelashes. On the face, granular skin caused by the numerous papules that may correspond to sebaceous hyperplasia can



**Figure 6** Progression of classic LPP.



**Figure 7** Typical frontal fibrosing alopecia.

develop. As in the classical pattern, and maybe more often, hair on the other parts of the body can be affected by this inflammatory process.

Although androgenetic alopecia (AGA) is included in the differential diagnosis, it is usually easy to distinguish FFA from AGA. In AGA, vellus are present, and there is no perifollicular inflammation or scale. The preauricular or eyebrow involvement seen in FFA is absent in AGA.

#### Lassueur Graham-Little Piccardi Syndrome

This syndrome consists of scarring patchy alopecia of the scalp, noncicatricial axillary and pubic hair loss, and lichenoid follicular eruption. Most of the patients described are women ages 30 to 60 years. Reported initially by Piccardi in 1913, Graham-Little published an observation of a patient of Lassueur in 1915. The 3 problems may appear simultaneously but scalp alopecia often precedes



**Figure 8** Typical frontal fibrosing alopecia in a man.



**Figure 9** Frontal fibrosing alopecia: slow evolution.

the follicular eruption of horny papules (even years before). Histology of the papules is frequently lichenoid. Histology of the scalp lesions usually shows a pseudopelagic stage of cicatricial alopecia; however, it may not be recognized. The axillary and pubic alopecia is regularly described as noncicatricial, because there is no clinical atrophy but the histology can show destruction of follicles.<sup>17</sup> By these facts, we feel that the diagnosis of Lassueur-Little Piccardi syndrome has been by inappropriately made in a proportion of LPP eruptions,<sup>18,19</sup> because the syndrome can be associated with an eruption of LP or LPP of the body hair (Fig. 10), especially of the pubic and axillary regions.

#### Fibrosing Alopecia in a Pattern Distribution (FAPD)

A fourth variant, fibrosing alopecia in a pattern distribution (FAPD), may be added to the widely accepted 3 clinical variants; however, FAPD is not recognized by all authors as an authentic entity. This entity<sup>3</sup> appears in women as LPP or common baldness or AGA in



**Figure 10** forearm LPP in a LGLP.



**Figure 11** FAPD variant. Clinical appearance.

men (Fig. 11). The hypothesis is that modification of the hair in AGA could be responsible for LPP. Four patients of the original cohort responded to finasteride, suggesting that the lichenoid tissue reaction might be pathologically related to AGA itself at least in those cases.

Body hair involvement is associated with scalp LPP. LPP is frequent, especially on the lower of the legs in centrifugal patches. A rare variant of LPP is the linear form, described on the face, and once on the trunk<sup>20</sup> following the Blaschko lines. In this last case, the absence of scarring in the involved zones has to be noted.

## Differential Diagnosis

Before alopecia appears, seborrheic dermatitis can be seen with scaling inflammation. Clinically, noncicatricial alopecia is eliminated by the presence of follicular ostium. In alopecia areata, exclamation mark hairs, presence of yellow dots on dermoscopy, and white hair can help establish the diagnosis. Differentiation from discoid lupus erythematosus (DLE) is more difficult. In DLE, inflammation is not restricted to surrounding hairs, and the appearance of dyspigmentation, telangiectasias, and scales can help the clinician with the diagnosis. Although LPP and DLE can be seen in the same patient, in our experience this is very rare. Although both DLE and LPP have a lymphocytic infiltrate, the histologic presentation is different. In DLE, there can be an increase in mucin, especially in reticular dermis, the epidermis is focally thinned and the interfollicular epidermis generally not spared, dermal sclerosis is usually less severe than for LPP and telangiectasias are present. The lymphocytic infiltrate is often perivascular and can involve eccrine glands. The direct immunofluorescence (DIF) of DLE is also usually specific with lupus band (Ig G, IgM, C3) having a positive result in the range from 63% to 100%. In LPP the lupus band is negative and, unlike in DLE treatment with hydroxychloroquine is not effective.

Pseudopelade of Brocq (PPB), which can be considered as end-stage LPP, may leave the clinical appearance of "footprints in the snow." If most of the lesions of PPB are old and the LPP is not expanding with perifollicular inflammation and hyperkeratosis, it is impossible to distinguish PPB from LPP.

In the late stages of LPP, it must be differentiated from central centrifugal cicatricial alopecia (CCCA) and folliculitis decalvans. CCCA presents with one symmetric unique centrifugal patch located on the vertex. It occurs most often in black women and can be considered as localized LPP, without definitive histology. When pustules and crusts

are absent, folliculitis decalvans can simulate an LPP. The neutrophilic infiltrate of folliculitis decalvans disappears in advanced stages of the disease. In certain difficult cases, fine observation with dermoscopy, and new biopsies can help the clinician.

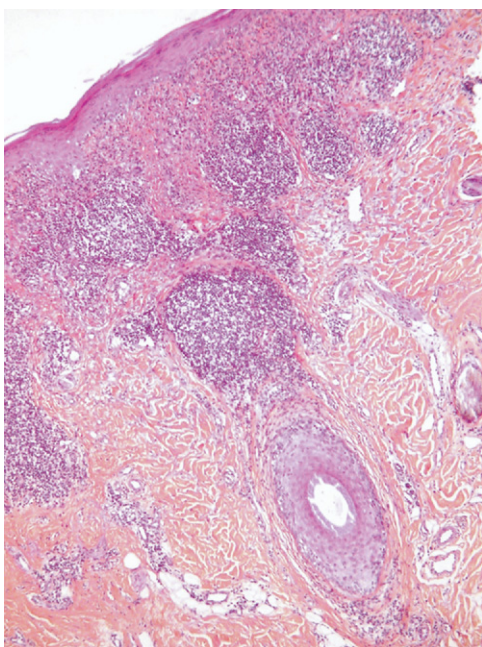
## Histopathology and Other Complementary Examinations

### Histopathology

A good biopsy needs to be done at the margin of alopecia, in an active and inflammatory lesion. A 4-mm punch is the minimal necessary size. Transverse sectioning seems may be much more informative than vertical orientation.<sup>21</sup>

In the active and early stage, the biopsy is a crucial clue for the diagnosis. The histopathology shows a lichenoid interface inflammation with hypergranulosis, hyperkeratosis, hyperacanthosis, degeneration of basal keratinocytes and destruction of the basal layer in half cases (Fig. 12).<sup>5,21,22</sup> A bandlike subepidermal infiltrate of lymphocytes is present, involving (“hugging”) the follicles between the infundibulum and the isthmus, with sparing of the lower portion (different from alopecia areata). In the basal layer are observed colloid bodies which are degenerated keratinocytes that stain pink with eosin.<sup>10</sup>

In late-stage lesions, inflammation can be minimal or absent, with no distinctive lichenoid changes, fibrous tracts take the place of destroyed hair follicles. At this stage, when inflammation is not present, hair transplantation can be considered. As in the end-stage of other primary scarring alopecia, biopsy of the late stage lesion or “pseudopelagic state will not provide any information to assist with the diagnosis.”<sup>23</sup> Histology findings in FFA are similar than LPP.<sup>24</sup> Despite no apparent clinical inflammation, Tosti found a lichenoid infiltrate in 2 of 5 cases on the eyebrows.<sup>25</sup> In 40% of cases, the DIF can show colloid bodies and positive staining for IgM, less commonly IgA or C3 and a linear band of fibrin and/or fibrinogen<sup>5,10</sup> at the dermoepidermal junction. In the remaining cases, DIF is nonspecific.



**Figure 12** Typical appearance of pathology in LPP. (hematoxylin and eosin, zox) credit: B. Cavelier-Balloy.

### Other Complementary Investigations

No blood test seems useful. Antinuclear antibodies are present in 10% cases,<sup>6</sup> and a thyroid disorder in 24% with 7% of these cases having Hashimoto disease. There is no reason to investigate anti-HCV antibodies in this form of lichen planus. A statistically significant association has only been found between erosive LP and infection by HCV.<sup>12</sup> A trichogram would show anagen hairs surrounded by thickened sheets in areas of active disease.

### Physiopathology

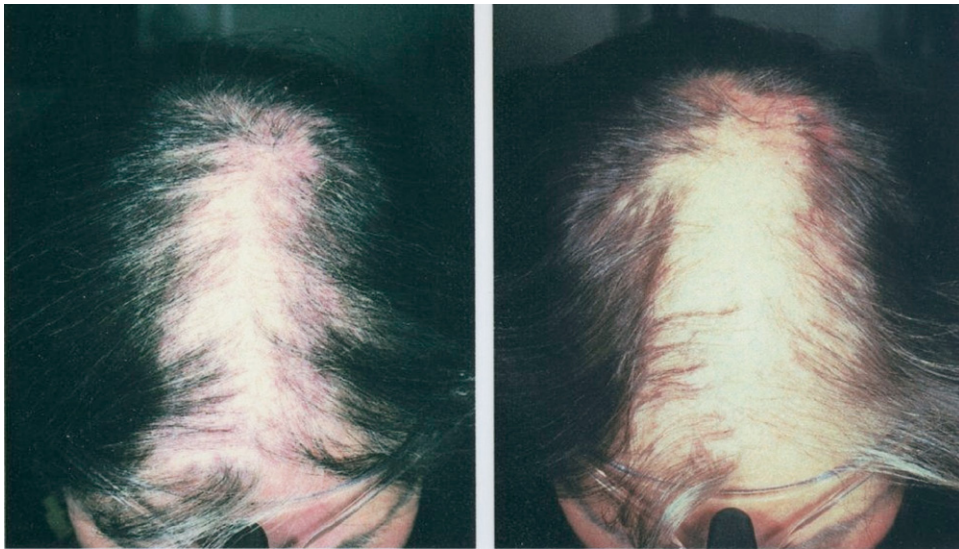
LPP is a disease of unknown etiology, whose pathogenesis is poorly understood, despite a suspected autoimmune origin. This reaction is mediated by T lymphocytes (CD4 and CD8) activated by Langerhans cells that are increased in the dermis and epidermis. The inflammatory cells attack and destroy keratinocytes expressing particular antigens. The inflammation is mostly located around the bulge area, where multipotential cells are present<sup>26</sup> and could be destroyed.<sup>27-29</sup> The target follicular antigens are not known, but the process may be triggered by pharmacologic agents as LPP can be an adverse complication of the therapy. Reactions to pharmacologic treatment are more likely to be LP than true LPP. A contact sensitizer, a virus or another infectious agent could be responsible for triggering the process in predisposed subjects. Altered integrin expression, which has been shown in active LPP lesions,<sup>30</sup> could explain the phenomenon of easy pulling-out of the hair with an abnormal root sheath.

### Treatment

The therapeutic management of LPP depends on the disease activity, patient, and physician variables. Disease variables include the extent of scalp involvement, and rapidity of evolution of the disease, Patient variables are age and comorbidities of the patient, patient motivation quality of life, the psychological consequences of the disease, and patient preference. Also, the dermatologists' experience with the disease and treatment methods both influence the therapeutic recommendations. Patients are distressed by the permanent hair destruction,<sup>31</sup> and treatments often are difficult and disappointing.<sup>32</sup> Treatments are less effective for LPP than for lichen planus. Because the alopecia is cicatricial, hair regrowth is by definition impossible, with the one exception being at the very beginning of the disease.<sup>10</sup> The treatment goals are to block the progression of the illness, prevent further alopecia, and reduce pruritus and pain. Patients must be informed of those goals. Early diagnosis is crucial for early initiation of therapy. As the evaluation of the affected area is clinically very tricky, we highly recommend repeated global photographs (Fig. 13) and hair count assessments (Fig. 14) on selected inflammatory target areas. We use an automated count with the TrichoScan® software<sup>33</sup> and small tattoos. Unfortunately, no treatment has shown a durable response. There is also no publication of any trial with a control group comparing therapeutic effects with biopsies or hair count with long-term follow-up.

### Treatment of Classic LPP: the Therapeutic Ladder

- First-line therapy:
  - At the Centre Sabouraud, we use an ultra-potent topical corticotherapy with propionate clobetasol lotion twice a day for 1 month followed by an application once a day for 3 months, and then every other day for 3 more months. After 12 weeks of



**Figure 13** Example of global photographic assessment at baseline and 6 months.

topical corticotherapy, 66% of Chierigato's patients complete clinical clearing and 13% did not respond to the topical therapy alone.<sup>5</sup> Mehregan obtained similar results with an improvement in 70% of patients.<sup>10</sup> Regrettably, positive results are clearly worse in our experience, and the rate of relapse approaches 80% of cases in a few months. Topical corticosteroid therapy for up to 1 year is recommended for children.<sup>8</sup>

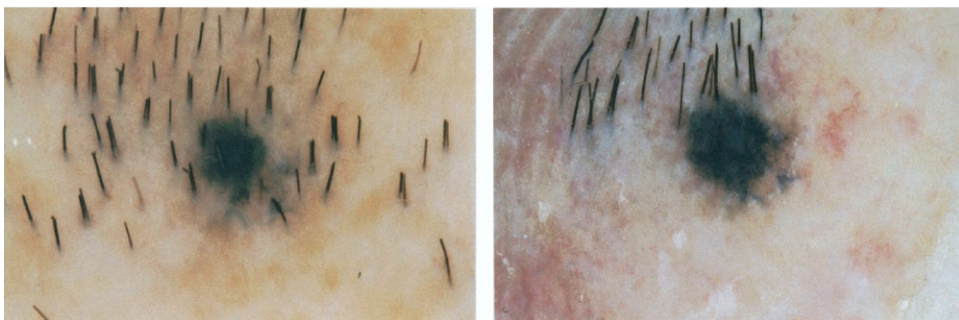
- We add monthly intralesional injections of triamcinolone acetonide (10 mg/mL; up to 3 mL) in the cases of local and severe inflammation.
- Second-line therapy:
  - Systemic oral corticosteroid therapy is started with prednisone, 1 mg/kg<sup>-1</sup>/d<sup>-1</sup> for 15 d, and tapered over 4 months can help stop rapid hair loss. One-half a milligram daily is usually the minimal dose for LP but is inadequate for LPP. A relapse after ceasing treatment is common, occurring in more than 80% of the patients from whom the follow-up is available.<sup>10</sup> Often, an early relapse occurs during the prednisone tapering.
- Third-line therapy:
  - Cyclosporin: in a first report, a short course of oral cyclosporin, 300 mg d<sup>-1</sup> was effective after 3 to 5 months for 3 patients. Remission remained after 12 months for 2 patients.<sup>34</sup> A positive and persistent therapeutic effect with minimal regrowth has been reported with the dosage of 4 mg/kg/d in a case of Lassueur Graham-little Piccardi syndrome (LGLPS).<sup>35</sup> On the basis of those reports, we treated

13 patients with a success rate of 77% (clinical and hair count assessment) without serious side effects and with a little regrowth in a few cases (Fig. 15). The optimal efficient dosage seems to be between 4 mg/kg/d and 5 mg/kg/d and the optimal course about 4 months. The problem remains the high relapse rate between 60% and 80%, respectively 6 months and 12 months after ending the treatment. We do not know whether cyclosporin is more or less effective than systemic therapy with corticoids. They both have potential and different serious side effects. Only a comparative study would be able to define the advantages and disadvantages of each. In the absence of a definitive clinical trial, cyclosporin can be considered a therapeutic option for refractory LPP.

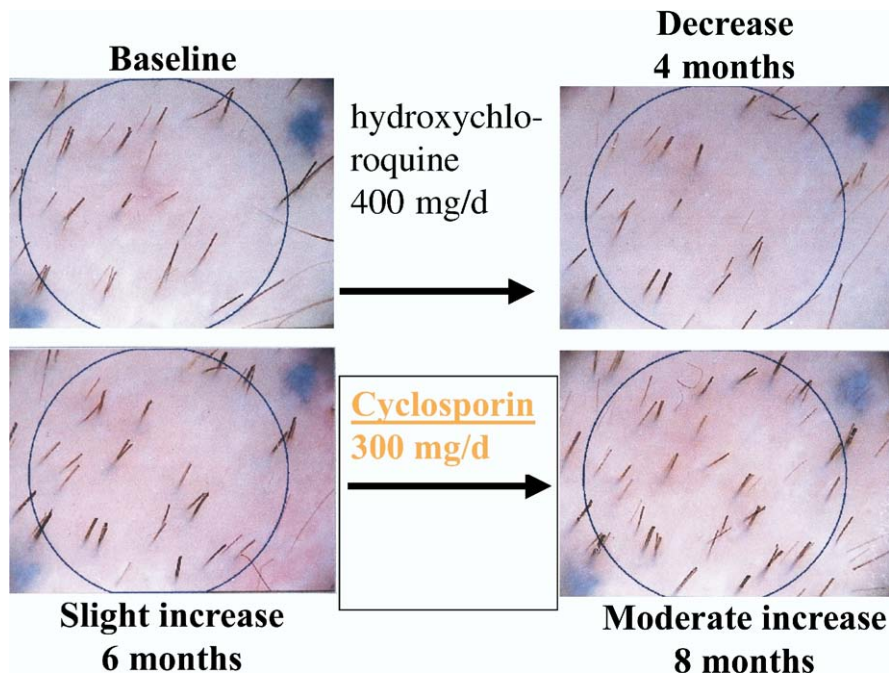
- Mycophenolate mofetil could be a preferable therapeutic choice to cyclosporin because of the safer adverse-effect profile. A first case of successfully treated LPP with mycophenolate mofetil was reported in 2004.<sup>36</sup> Because of this publication, we decided to treat 5 patients at the dosage of 2 g/d for 2 months-8 months with a success rate of 40% (clinical and hair count assessment), without noticeable side effects. To avoid any relapse in case of effective treatment, it can then be continued for many months.

Other treatments:

- Some years ago we showed that hydroxychloroquine is not effective in ending the progression of LPP (Fig. 15). We



**Figure 14** Hair count assessment before and 6 months after hydroxychloroquine therapy.



**Figure 15** Example of a hair count assessment and regrowth with cyclosporin therapy.

treated 12 patients with hydroxychloroquine at 400 mg per day for 6 months without any success. By using a global photographic assessment, we observed a stabilization of the illness in only 3. Unfortunately, we noticed a clear worsening with the hair count for those 3 patients.<sup>37</sup>

- Retinoids have demonstrated their efficiency in the treatment of lichen planus. However, we tried to treat 6 patients with LPP and 2 patients with PPB with etretinate, 25 mg d<sup>-1</sup> without any success. None of the patients showed clinical improvement on global photography, and we observed a decrease of the hair count. Regrettably an important iatrogenic telogen effluvium was observed for all of the patients (Unpublished data, Dr C. Jouanique, 2002). For now no assessment for lower dosage etretinate alone nor associated with hydroxychloroquine, or for isotretinoin has been published.
- Thalidomide, 100 mg d<sup>-1</sup> given for 6 months was not effective for 4 patients presenting with LPP and 2 with PPB. Global photographic assessment and hair counts showed a worsening for all the patients at 6 months.<sup>38</sup>
- Tetracycline, 1 g d<sup>-1</sup> was reported as effective in a retrospective study without photographic and without hair count assessment.<sup>6</sup> These data do not have sufficient evidence to warrant the recommendations of the authors. Efficacy needs to be demonstrated by further investigation with randomized controlled trials. We do not recommend tetracycline as first-line treatment.
- Griseofulvin, 1 g d<sup>-1</sup> has been considered effective in a few anecdotal cases.<sup>10</sup>
- In acute disease, intravenous pulses of methylprednisolone corticotherapy can be a therapeutic option.
- There is as yet no report about methotrexate in LPP, but this drug could be envisaged in a trial either alone or with low dose prednisone in refractory LPP.

- New biological agents targeting T lymphocytes would be a logical avenue to pursue. Alefacept has been employed with little success in treating erosive lichen planus<sup>39</sup> or generalized lichen planus (15 mg/week i.m. for 12 weeks),<sup>40</sup> but at this time there is no report for this treatment in LPP.
- Antitumor necrosis factor alpha treatments do not seem to be a logical option. A case of LPP occurring during a treatment with etanercept has been reported.<sup>41</sup>
- Despite one success reported after a partial relapse after systemic treatment with cyclosporin,<sup>34</sup> topical tacrolimus, 0.1% applied once or twice a day is of low benefit. Furthermore, the ointment is too greasy to use because it is particularly difficult to remove even after several shampoos. Pimecrolimus, 1% cream is effective to treat mucosal oral or genital lichen planus, or lichen striatus,<sup>42</sup> but there is not enough data about pimecrolimus and LPP.
- Topical 2% or 5% minoxidil lotion can help patients by improving regrowth of the remaining hair.

### Treatment of FFA

There is no proven effective treatment for FFA. We tried topical corticotherapy, topical tacrolimus, and systemic therapy with chloroquine or hydroxychloroquine with no satisfying result. A Spanish and an Italian open label series<sup>25,43</sup> suggested the use of a combined therapy: oral finasteride, 2.5 mg/d, topical minoxidil, 2 to 5% twice a day, and intralesional triamcinolone acetonide 20 mg/mL every 3 months.<sup>43</sup> We prefer to employ lower concentration (up to 10 mg/mL) of triamcinolone acetonide to reduce the risk of cutaneous atrophy.

A short course of oral prednisone, 0.5 mg/kg-1 mg/kg, can be proposed in some cases of acute and rapid hair loss, tapered over 3 months, with no expected effect on long term remission.

Most of the time the progression of the alopecia is slow and limited. Topical minoxidil can improve the general appearance of the scalp by stimulating the remaining hairs. When the disease is inactive, observation with photographic documentation is in-

licated. Hair transplants can be proposed when there is no progression of the disease for at least 1 year.

## Treatment of LGLPS

- First-line therapy:

Recommended treatments are similar to those used for management of classical LPP: an ultrapotent topical corticosteroid with propionate clobetasol lotion twice a day for 1 month and then once a day for 3 months, and then every other day for 3 months more. We add monthly intralesional injections of triamcinolone acetonide (10 mg/mL; up to 3 mL) in cases of local and severe inflammation.

- Second-line therapy:

Systemic oral corticotherapy with prednisone, 0.5 mg-1 mg/kg<sup>-1</sup>/d, tapered for over 3 months or cyclosporin, 4 mg/d -5 mg/d for 3 months-6 months.

## Surgery

At the end-stage of cicatricial alopecia, a scalp reduction or hair transplants can be performed. Because surgical procedures could provoke a new episode of a generalized autoimmune disease, this option remains the exception to customary management of the disease. If hair transplantation is embarked upon, it is done after 6 months without active disease, and after a biopsy confirms the clinical impression of the disease being in the noninflammatory stage. A test site with 3 to 5 transplantation punches of hair are first implanted, then a waiting and observation period of 6 months-12 months is necessary. The success of transplanted hair and possible new inflammatory processes are evaluated before a complete hair transplant session is embarked upon.

## Conclusions

LPP is a chronic condition, whose diagnosis needs to be performed as soon as possible. The cicatricial evolution often is visible in areas on the scalp. Clinical characteristics are identified with the help of dermoscopy or videomicroscopy. Histopathology, especially in the early stages, establishes this diagnosis. Photography and/or hair count are necessary for evaluation of the treatment or the evolution without treatment. Therapeutic management is often quite challenging, as relapses are common after any treatment. Considering the psychological repercussions of this illness, a reasonable therapeutic ladder treatment is justified with clear explanation of the problem and the limitations of treatment to the patient. Further research is needed on the pathogenesis, and randomized controlled trials are necessary to appreciate scientifically any treatment.

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## References

- Pringle JJ: Cited by Adamson HG. Lichen pilaris spinulosus. *Br J Dermatol* 17: 78-9, 1905
- Olsen EA, Bergfeld WF, Cotsarelis G, et al: Workshop on cicatricial alopecia. Summary of North American Hair Research Society (NAHRS)-sponsored workshop on cicatricial alopecia, Duke University Medical Center, February 10 and 11, 2001. *J Am Acad Dermatol* 48:103-110, 2003
- Zinkernagel MS, Trüeb RM: Fibrosing alopecia in a pattern distribution: Patterned lichen planopilaris or androgenetic alopecia with a lichenoid tissue reaction pattern? *Arch Dermatol* 136:205-211, 2000
- Otberg N, Wu WY, McElwee KJ, et al: Diagnosis and management of primary cicatricial alopecia: Part I. *Skinmed* 7:19-26, 2008
- Chierigato C, Zini A, Barba A, et al: Lichen planopilaris: Report of 30 cases and review of the literature. *Int J Dermatol* 42:342-345, 2003
- Cevasco NC, Bergfeld WF, Remzi BK, et al: A case-series of 29 patients with lichen planopilaris: The Cleveland Clinic foundation experience on evaluation, diagnosis, and treatment. *J Am Acad Dermatol* 57:47-53, 2007
- Kang H, Alzolibani AA, Otberg N, et al: Lichen planopilaris. *Dermatol Ther* 21:249-256, 2008
- Handa S, Sahoo B: Childhood lichen planus: A study of 87 cases. *Int J Dermatol* 4:423-427, 2002
- Tosti A: *Dermoscopy of Hair and Scalp Disorders*. London, Informa Healthcare 2007
- Mehregan DA, Van Hale HM, Muller SA: Lichen planopilaris: Clinical and pathologic study of forty-five patients. *J Am Acad Dermatol* 27:935-942, 1992
- Rook A, Dawber R (eds): *Hair and Scalp Diseases*. Oxford, Blackwell Scientific Publications, 1982
- Giménez-García R, Lázaro-Cantalejo TE, Sánchez-Ramón S, et al: Linear lichen planopilaris of the face. *J Eur Acad Dermatol Venereol* 19:770-772, 2005
- Mirmirani P, Willey A, Headington JT, et al: Primary cicatricial alopecia: Histopathologic findings do not distinguish clinical variants. *J Am Acad Dermatol* 52:637-643, 2005
- Kossard S, Lee Ms, Wilkinson B: Postmenopausal frontal fibrosing alopecia: A frontal variant of lichen planopilaris. *J Am Acad Dermatol* 36:59-66, 1997
- Vaisse V, Matarad B, Assouly P, et al: 20 CAS. *Ann Dermatol Venereol* 130:607-610, 2003
- Tan KT, Messenger AG: Frontal fibrosing alopecia: Clinical presentations and prognosis. *Br J Dermatol* 160:75-79, 2009
- Ghislain PD, Van Eeckhout P, Ghislain E: Lassueur-Graham little-Piccardi syndrome: A 20-year follow-up. *Dermatology* 206:391, 2003
- Abbas O, Chedraoui A, Ghosn S: Frontal fibrosing alopecia presenting with components of Piccardi-Lassueur-Graham-little syndrome. *J Am Acad Dermatol* 2:S15-S18, 2007 (suppl)
- Crickx B, Blanchet P, Grossin M, et al: Lassueur-Graham-little syndrome. 2 Cases. *Ann Dermatol Venereol* 117:907-909, 1990
- Baker K, Pehr K: Linear lichen planopilaris of the trunk: First report of a case. *J Cutan Med Surg* 10:136-138, 2006
- Sperling LC: Scarring alopecia and the dermatopathologist. *J Cutan Pathol* 28: 333-342, 2001
- Tandon YK, Somani N, Cevasco NC, et al: A histologic review of 27 patients with lichen planopilaris. *J Am Acad Dermatol* 59:91-98, 2008
- Sperling LC, Cowper SE: The histopathology of primary cicatricial alopecia. *Semin Cutan Med Surg* 25:41-50, 2006
- Poblet E, Jiménez F, Pascual A, et al: Frontal fibrosing alopecia versus lichen planopilaris: A clinicopathological study. *Int J Derm* 45:375-380, 2006
- Tosti A, Piraccini BM, Iorizzo M, et al: Frontal fibrosing alopecia in postmenopausal women. *J Am Acad Dermatol* 52:55-60, 2005
- Cotsarelis G, Sun TT, Lavker RM: Label-retaining cells reside in the bulge area of pilosebaceous unit: Implications for follicular stem cells, hair cycle, and skin carcinogenesis. *Cell* 29:1329-1337, 1990
- Horenstein MG, Simon J: Investigation of the hair follicle inner root sheath in scarring and non-scarring alopecia. *J Cutan Pathol* 34:762-768, 2007
- Mobini N, Tam S, Kamino H: Possible role of the bulge region in the pathogenesis of inflammatory scarring alopecia: Lichen planopilaris as the prototype. *J Cutan Pathol* 32:675-679, 2005
- Pozdnyakova O, Mahalingam M: Involvement of the bulge region in primary scarring alopecia. *J Cutan Pathol* 35:922-925, 2008
- D'ovidio R, Sgarra C, Conserva A, et al: Integrin expression in lichen planopilaris. *Head Face Med* 3:11, 2007
- Grace PM: Living with lichen planopilaris. *Dermatol Nurs* 19:184-185, 2007
- Harries MJ, Sinclair RD, Macdonald-Hull S, et al: Management of primary cicatricial alopecias: Options for treatment. *Br J Derm* 159:1-22, 2008
- Hoffman R: Trichoscan: What is new? *Dermatology* 211:54-62, 2005
- Mirmirani P, Willey A, Price VH: Short course of oral cyclosporin in lichen planopilaris. *J Am Acad Derm* 49:667-671, 2003
- Bianchi L, Paro Vidolin A, Piemonte P, et al: Syndrome: Effective treatment with cyclosporin A. *Clin Exp Dermatol* 26:518-520, 2001
- Tursen U, Api H, Kaya T, et al: Treatment of lichen planopilaris with mycophenolate mofetil. *Dermatol Online J* 10:24, 2004
- Jouanique C, et al: IHRS. Tokyo. Poster, 2002
- Jouanique C, Reygagne P, Bachelez H, et al: Thalidomide is ineffective in the treatment of lichen planopilaris. *J Am Acad Dermatol* 51:480-481, 2004
- Chang AL, Badger J, Rehmus W, Kimball AB: Alefacept for erosive lichen planus: A case series. *J Drugs Dermatol* 7:379-383, 2008
- Fiveson DP, Mathes B: Treatment of generalized lichen planus with alefacept. *Arch Dermatol* 142:151-152, 2006
- Garcovich S, Manco S, Zampetti A, et al: Onset of lichen planopilaris during treatment with etanercept. *Br J Dermatol* 158:1161-1163, 2008
- Volz T, Caroli U, Lütke H, et al: Pimecrolimus cream, 1% in erosive oral lichen planus—A prospective randomized double-blind vehicle-controlled study. *Br J Derm* 159:936-941, 2008
- Moreno-Ramírez D, Camacho Martínez F: Frontal fibrosing alopecia: A survey in 16 patients. *J Eur Acad Dermatol Venereol* 19:700-705, 2005