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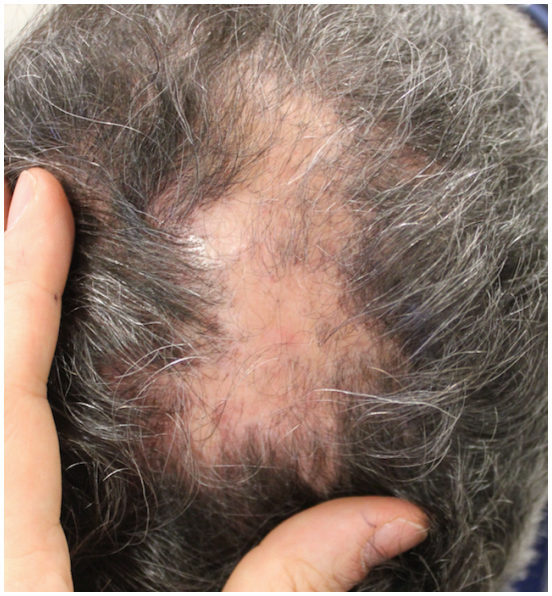
LICHEN PLANOPILARIS



What is lichen planopilaris?

- This is a type of scarring hair loss condition.
- The hair loss in the bald areas is permanent
- It is one of the most common causes of scarring alopecia

Who gets lichen planopilaris?

- Adults of all ethnic backgrounds
- Females more two times more likely than males but both are affected

What are the symptoms of lichen planopilaris?

- In general patients with lichen planopilaris may have itching of the scalp and some may have increased hair shedding.
- They may also have burning, pain and tenderness in the scalp.
- Some patients note that the scalp is tender when the hairs are moved.

- There may be redness in the scalp and some scale.

How does Dr. Donovan arrive at the diagnosis of lichen planopilaris?

- Several features lead to this diagnosis, including areas of scarring hair loss, some redness and scale around the hair follicles, and findings on the biopsy showing inflammation, loss of the fat glands and scarring around the hair follicles

How is lichen planopilaris treated?

- The goal of treatment is to stop the disease. Hair regrowth in the bald areas is not possible in most cases. Even with treatment, some cases to spread, albeit very slowly.

Treatments

- Pills may be used such as Doxycycline, Plaquenil, Isotretinoin (Accutane), Mycophenolate mofetil
- Injection of steroids may be advised
- Use of topical medicines you apply yourself at home may also be advised (Clobetasol, Protopic, Clobex, Elidel)
- Sometimes, hair transplantation can be performed if the disease become quiet and stays quiet

How long will I be on these treatments?

- Your disease will be carefully monitored by Dr Donovan.
- If the disease does not appear to be spreading, the doses of medications will be reduced and possibly stopped.
- However, if there is any evidence the disease is increasing, increased doses or even new medications may be prescribed.

Dr. Donovan – 10 Articles on LPP

ARTICLE 1: How successful are hair transplants in scarring alopecias?

This is an in important question and one that needs good data. I serve as the chair of a committee of the International Society of Hair Restoration Surgery. On Friday we sent out a survey to hair transplant surgeons around the world with the hopes of gathering more information on the successes and failures surgeons have had when transplanting scarring alopecias.

There is no doubt that hair transplantation works wonderfully in some patients with scarring alopecia and does not work well in others. One must always have quiet (inactive) disease for at least 1-2 years before a transplant is attempted.

What are the criteria for transplanting scarring alopecia?
In general, a scarring alopecia must be quiet for 1-2 years before a transplant can be even considered. Several years ago I put forth criteria for determining if an individual with lichen planopilaris is a hair transplant candidate:

Criteria for Hair Transplantation Candidacy in LPP

1. The PATIENT should be off medications.
Ideally the patient should be off all topical, oral and injection medications to truly know that the disease is burnt out and 'inactive'. However, in RARE cases, it may be possible to perform a transplant in someone using medications AND who meets criteria 2, 3 and 4 below. This should only be done on a case by case basis and in rare circumstances as the risk for disease reactivation is high. A patient using medications to suppress disease activity is at high risk for reactivation following hair transplant surgery. It is a last resort in a well-informed patient.

2. **The PATIENT must not report symptoms related to the LPP in the past 12 months, (and ideally 24 months)**.
The patient must have no significant itching, burning or pain. One must always keep in mind that the absence of symptoms does not prove the disease is quiet but the presence of symptoms certainly raises suspicion the disease could be active. Even the periodic development of itching or burning from time to time could indicate the disease has triggers that cause a flare and that the patient is

not a candidate for surgery. The patient who dabs a bit of clobetasol now and then on the scalp to control a bit of itching may also have disease that is not completely quiet.

3. **The PHYSICIAN must make note of no clinical evidence of active LPP in the past 12 months, (and ideally 24 months).**
There must be no scalp clinical evidence of active LPP such as perifollicular erythema, perifollicular scale (follicular hyperkeratosis). In addition, the pull test must be negative.

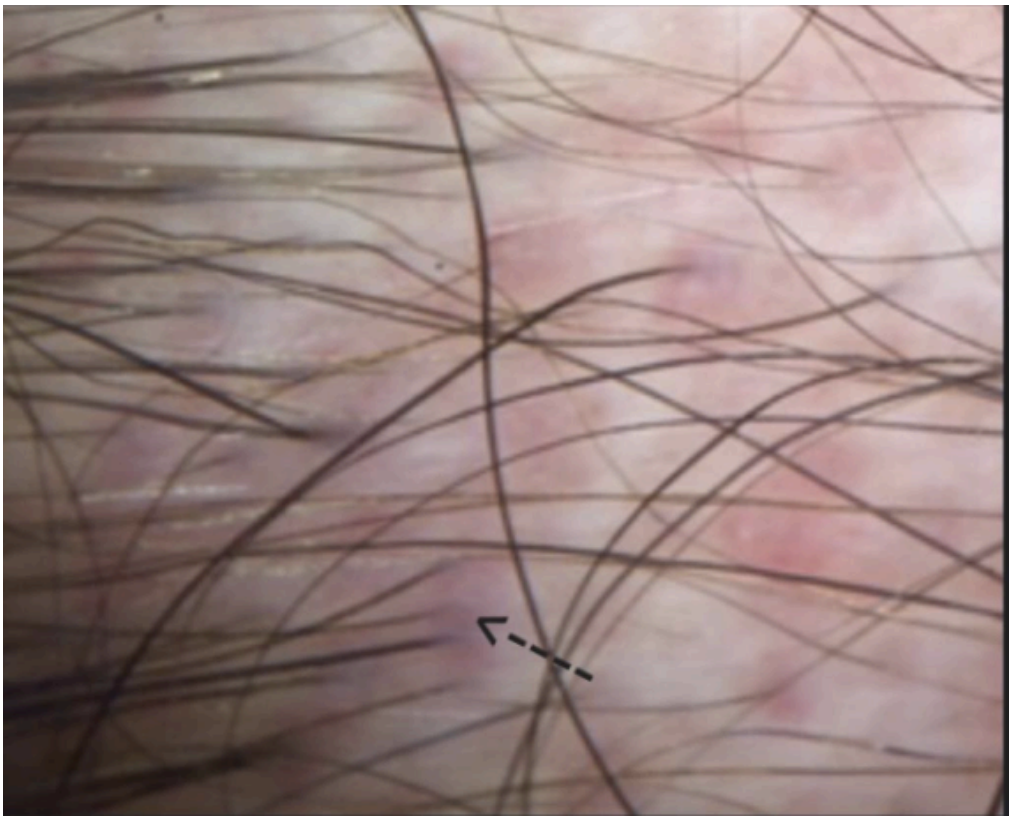
4. **Both the PATIENT and PHYSICIAN must show no evidence of ongoing hair loss over the past 12 months (and ideally 24 months).**
There must be no further hair loss over a period of 24 months of monitoring off the previous hair loss treatment medications. This general includes the patient and physician's perception that there has been no further loss as well as serial photographs every 6-12 months showing no changes. **As discussed above, the 12 month waiting time is the standard of care as an accepted definition for hair transplant candidacy.**

5. The patient must have sufficient donor hair for the transplant.
Not all patients with LPP maintain sufficient donor hair even if the disease has become quiet.

Disease Reactivation Following Surgery
My research has focused on the chances of reactivation of LPP after surgery. It is important to be aware that ANY patient with LPP is at risk for reactivation or a 'flare' of their LPP after surgery. The risk, I estimate, is as follows:

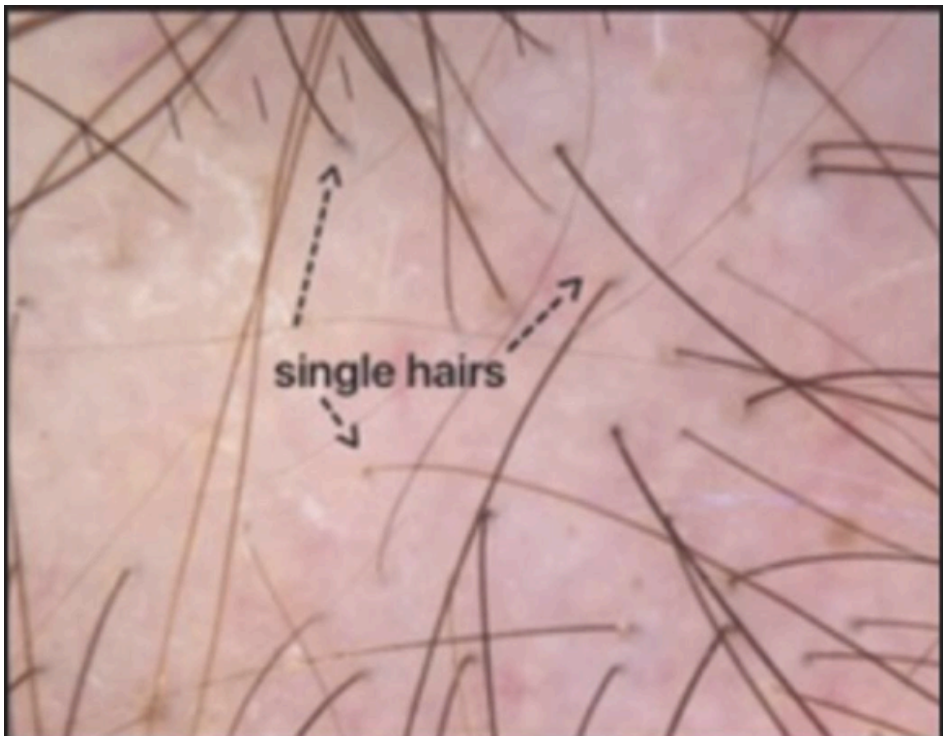
LPP Reactivation Risk (Donovan, J, unpublished data)
i) A patient with active LPP before their transplant is nearly guaranteed to have a flare of his or her LPP if a hair transplant is done. (estimate 90-100 % chance of flare within 2 years post transplant)
ii) A patient with partially active LPP before their transplant is very likely to have a flare if a hair transplant is done. (estimate 70-90 % chance of flare within 2 years post transplant)
iii) A patient with medication induced inactive LPP before their transplant has a moderate chance of a flare if a hair transplant is done (estimate 50-70 % chance of flare within 2 years post transplant)
iv) A patient with inactive LPP off all medications for 1 year before their transplant has a low chance of a flare if a hair transplant is done (estimate 10-25 % chance of flare within 2 years post transplant)
v) A patient with inactive LPP off all medications for 2 years before their transplant has a low but definite chance of a flare if a hair transplant is done (estimate less than 10% chance of flare within 2 years post transplant)

ARTICLE 2: PARTIALLY TREATED LPP



Lichen planopilaris (LPP) is a scarring hair loss condition. The goal of treating LPP is to stop the condition. Successful treatment is associated with a halting of hair loss but also with an improvement in the symptoms and signs of the disease. Patients will notice a reduction in itching and burning and clinically there will be an improvement in scaling and redness around hairs. Sometimes scaling is the first to improve and improvements in redness happen later. This picture shows a patient with partially treated lichen planopilaris. The disease is still active although scaling has improved. The patient's itching has also improved.

ARTICLE 3: SINGLE HAIRS IN LPP



Lichen planopilaris (LPP) is a type of scarring hair loss condition. Patients frequently present with scalp itching, and sometimes scalp burning and tenderness. Increased hair shedding is common in the early stages. Hair loss is generally permanent and treatment helps stop the disease or at least slow down progression.

Clinically, dermoscopy (trichoscopy) of LPP often shows perifollicular erythema and perifollicular scale (follicular keratosis).
These findings are not present in all forms of LPP. A less common presentation of LPP is shown in the photo. Patients have hair loss with scalp itching. However, by dermoscopy they have many single hair follicles growing in a base of redness. This is what I have termed the "sea of singles" (SOS) appearance to describe the numerous single hairs and absence of hair follicle units containing 2 and 3 hairs. This form of LPP is similar to Abbasi's subtype described in 2016 and fibrosing aloepcia in a pattern distribution described by Zinkernagel in 2000. The "SOS" trichoscopic appearance is important to remember and provides a clue that the patient may have a scarring alopecia.

Reference
Zinkernagel MS et al. Arch Dermatol 2000

Abassi A et al. Dermatol Surg. 2016.

ARTICLE 4: Dosing oral immunosuppressants for Lichen planopilaris (LPP)

There are many different immunosuppressants and immune modulators that can be used for treating lichen planopilaris. Examples include doxycycline, hydroxychloroquine, methotrexate, mycophenolate, cyclosporine. I'm often asked what dose a patient should be using?

What dose should a patient be using?

When it comes to immunosuppressant medications, I always try to keep patients on the lowest possible dose that controls their disease. Generally I start at fairly standard doses of immunosuppressants and observe what happens to the patient's hair loss. For example, this might be 200 or 400 mg of hydroxychloroquine (Plaquenil) daily, 15-20 mg of methotrexate weekly, 150-300 mg of cyclosporine, 500-1000 mg of mycophenolate mofetil, 100 mg of doxycycline. If the disease is vastly improved after a few months, we may consider going down on the dose or staying at the same dose for a few more months. If the disease is getting worse, we might consider going up on the dose is their is room to go up or changing the immunosuppressant altogether.

ARTICLE 5 : AGA OR LPP: WHICH IS RIGHT?

In many fields of medicine, the pathology report provides the final answer as to a patient's diagnosis. We're most familiar with this for example with cancer diagnoses. It comes as a surprise for many patients that scalp biopsy reports are sometimes not so definitive.

Differentiating AGA and LPP

A great example is the diagnosis of early androgenetic alopecia (AGA and early lichen planopilaris (LPP). Sometimes it is pretty clear cut - but not always. Sometimes a diagnosis of LPP is made and the patient really has AGA. Sometimes (although much less commonly) a diagnosis of AGA is made and the patient really has LPP.

LPP: Brief Overview

Lichen planopilaris (LPP) is a scarring alopecia that typically starts with scalp symptoms such as itching and burning. Sometimes the scalp is quite tender in areas. Shedding is often present as well. LPP affects similar areas to androgenetic alopecia (female pattern thinning) so it is a close mimicker. In the early stages, some scalp redness may be present and inflammation may be seen around the hairs clinically.

AGA: Brief Overview

Androgenetic alopecia (AGA) also starts with shedding. There can be a hint of itching/tingling but not too often. Usually the front of the scalp is more affected by hair loss than the back.

Biopsies: Helpful or not?

A biopsy can be very helpful provided it is read by an experienced dermatopathologist. Traditionally we have thought of AGA as "non inflammatory" and "non scarring" so one might not think that inflammation and scarring should be present on the biopsy. We know now that's not completely true. Inflammatory infiltrates are present in AGA in the upper hair follicle and so is loose perifollicular

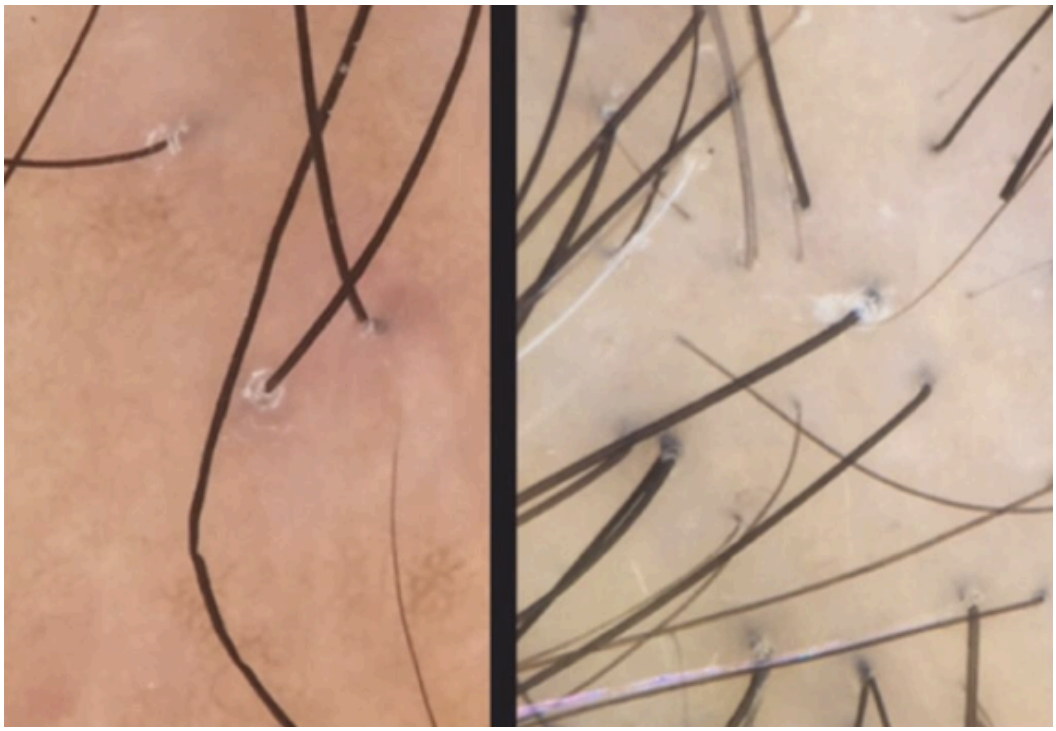
fibrosis. In LPP biopsies, inflammation is also present in the upper hair follicle but it specifically appears to be attacking the hair follicle outer root sheath. (We call this "lichenoid" change). To differentiate AGA and LPP one needs to direct their attention to this specific change in the actual hair follicle. When this specific immune attack is seen, one needs to consider LPP over AGA. Also the amount of perifollicular fibrosis is usually greater as LPP advances. LPP may have other changes in the skin as well that help differentiate it from AGA.

So by biopsy, androgenetic alopecia and LPP can be confused as both can have inflammation (perifollicular inflammation in the isthmus) and both can have scarring (perifollicular fibrosis). An experienced dermatopathologist can sort this out.

So how does one resolve this? Does the patient have AGA or LPP?

One needs to take into account the patient's entire story. If a physician just biopsies every patient that comes into the office, I can guarantee one will make a whole lot more diagnoses of LPP than truly are present. I'm a big believer in this - even though LPP is under diagnosed in the world! But by listening to the patient's entire story, and examining the scalp and reviewing what the biopsy shows (not just the final read out on the bottom line), one can usually get a fairly good sense. However in rare cases - time is the best judge as a missed case of LPP will likely declare itself over time.

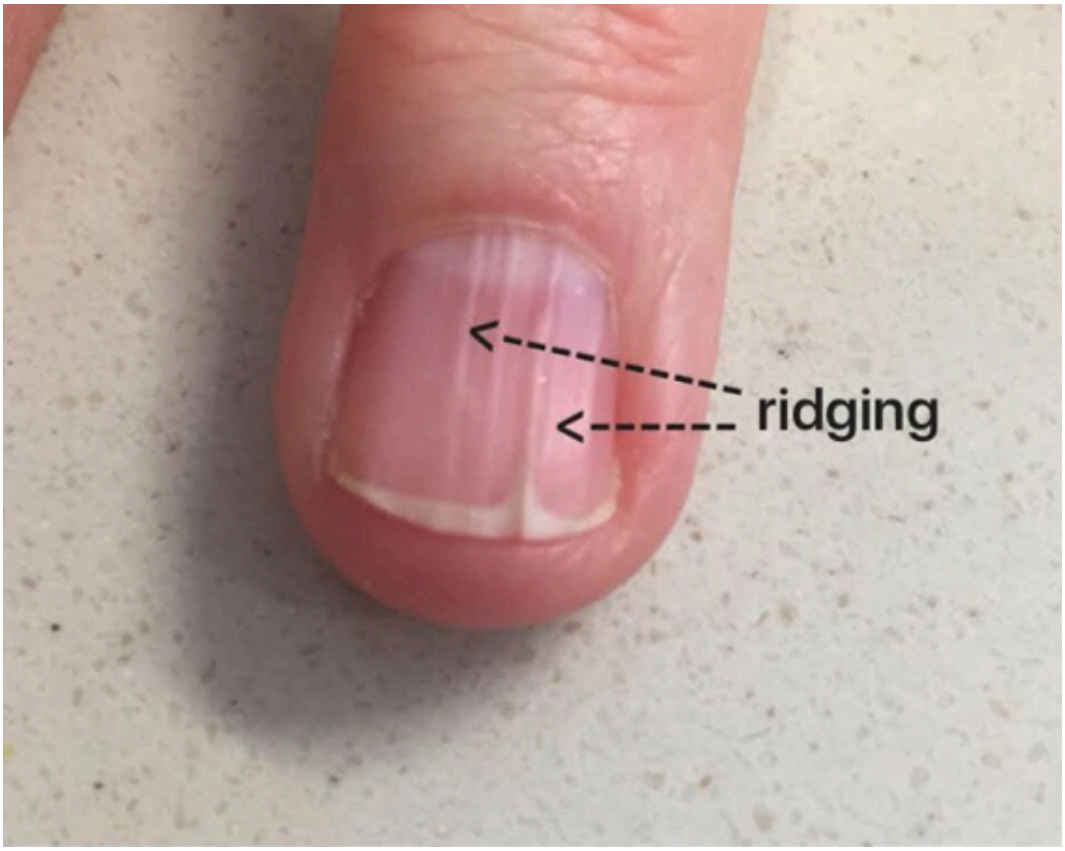
ARTICLE 6: Perifollicular Scale in LPP



The appearance of white colored scale around hair follicles is common. This can either be concerning - or not concerning. The white scale in the right picture is not concerning and represents a mild scale from normal epidermal turnover. The patient also has androgenetic alopecia. There is only one follicle affected and the scale is not tightly adherent to the hair follicle. When I see this, it catches my attention for just a second and then I move on to assess other scalp features.

The picture on the left shows a pattern of scale which is concerning. When I see hair follicles that look like this I am immediately concerned. This scale is tightly adherent to the follicle and forms a circular shape all around the follicle. It is important to note the underlying redness and it is also important to note that all of the follicles in the photo are just single hairs. Scale that tightly encircles the hair follicle in this manner is known as "perifollicular scale." In this left sided picture, the patient has an underlying scarring alopecia known as lichen planopilaris. Perifollicular scale and perifollicular redness are common in lichen planopilaris (as well as frontal fibrosing alopecia). Scale patterns can change if a patient washes his or her hair within 12 hours of their appointment. Sometimes, in order to better appreciate scaling in patients with challenging diagnoses - I will ask them to refrain from washing the scalp for 24-72 hours. I don't commonly do this but it can be helpful.

ARTICLE 7: Nail Lichen Planus



I generally ask about nail changes in most new patients I see in my office. I often describe hair and nails as "cousins" and it should therefore come as no surprise that many conditions that affect the hair also affect the nails. Patients with alopecia areata, lichen planopilaris, telogen effluvium, drug related hair loss, psoriasis may have changes in their nails.

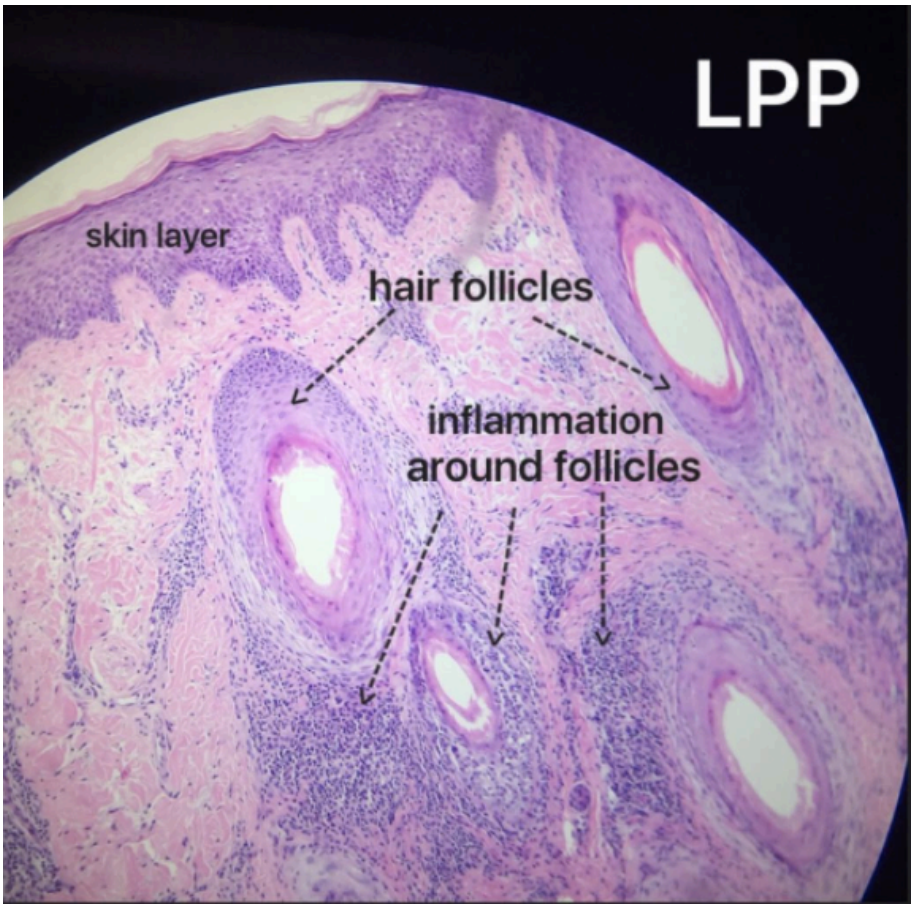
Some patients with scalp lichen planopilaris have nail lichen planus (LP). The clinical features of nail LP depend on where in the nail the disease is attacking (i.e. whether the matrix or nail plate are involved). Longitudinal ridging and splitting are the most commons clinical signs of nail matrix LP. This is shown in the photo. The splitting often extends right to the end as shown in the picture. However, a wide range of additional nail findings are also possible.

Some forms of nail lichen planus lead to rapid scarring and loss of the nail - (very similar to what is seen in the scalp). Other forms only lead to minor changes that may be difficult to differentiate from age related changes. Some patients have

resolution of nail disease even without treatment.

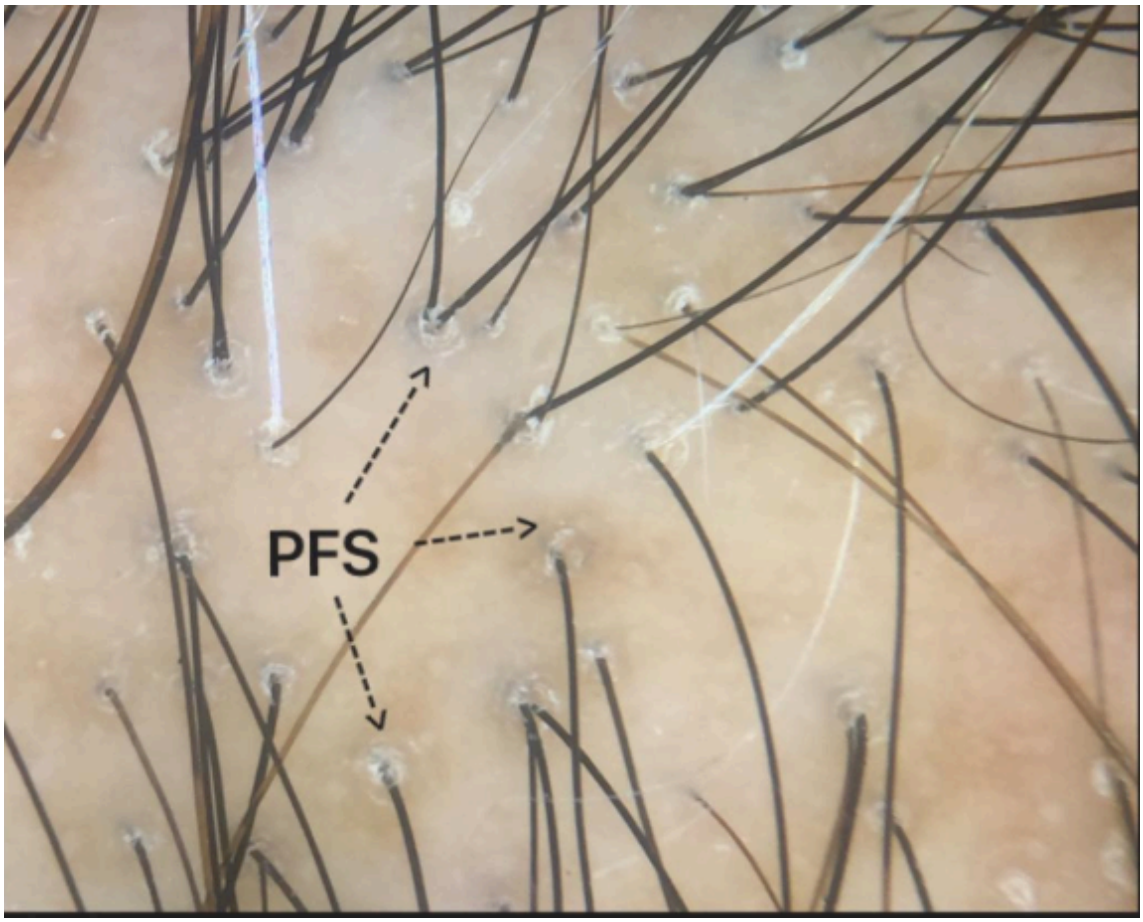
There are a variety of treatments are possible including topical steroids (with occlusion), steroid injections (0.5 to 0.1 mg/nail), intramuscular steroids every 30 days (0.5 mg/kg) and oral steroids for 3 weeks. Antimalarials (i.e. oral hydroxychloroquine), oral retinoids, psoralen, tacrolimus are also used. About 1/2 of patients will not improve despite any type of treatment.

ARTICLE 8: HISTOLOGY OF LPP



Lichen planopilaris (LPP) is a hair loss condition that results in permanent hair loss. Biopsies, as shown here, are associated with inflammation around the hair follicles.

ARTICLE 9 : Scale in Lichen Planopilaris

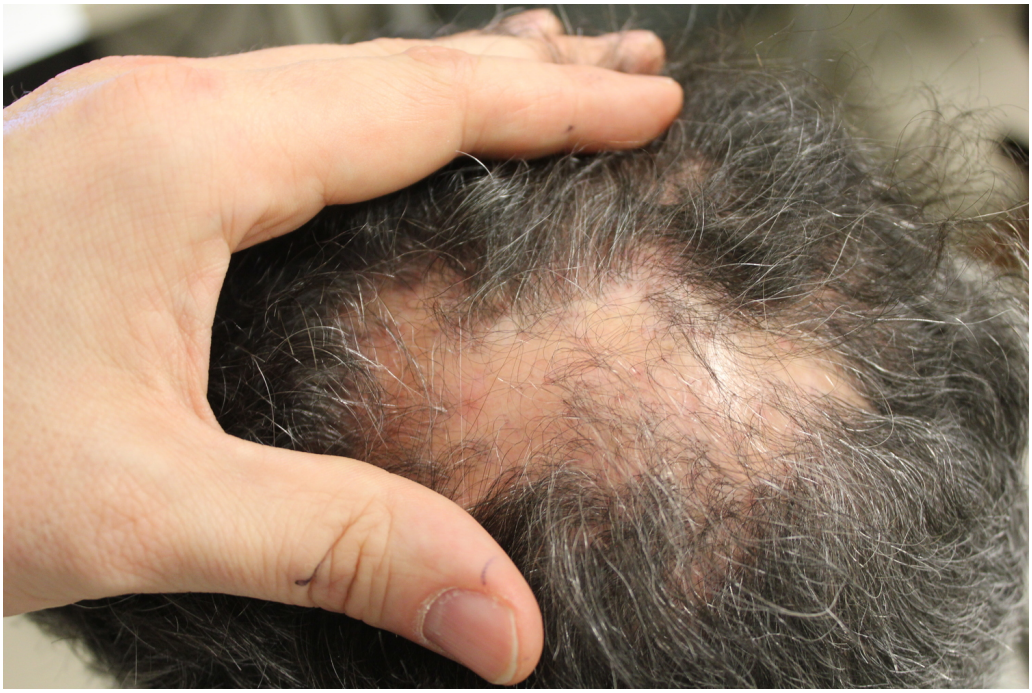


LPP is a type of scarring alopecia where hair loss is permanent. This makes it all the more important to secure the correct diagnosis with minimal delay.

Sometimes that is easier said than done. This picture shows an image of a female patient's scalp. At first look one could easily conclude this female patient has genetic hair loss. However, looking closer it reveals subtle scale around hair follicles (called perifollicular scale, PFS. Most of the hairs that are remaining (and shown in the photo) are the same diameter. There is not much in the way of miniaturization that is expected in pure androgenetic alopecia.

Treatments for LPP include: topical steroids, steroid injections, calcineurin inhibitors, oral pills including prednisone, hydroxychloroquine, doxycycline, methotrexate and others.

ARTICLE 10: LPP treatments: Where does the research point to?



Lichen planopilairis (LPP) is an autoimmune scarring hair loss condition that affected adults between 35 and 60. Patients develop hair loss but also symptoms of itching, burning and pain. The early stages of LPP are accompanied by increased shedding as well. Aggressive and early treatment of LPP is required to stop the hair loss.

What treatments are most effective?
Treatment that block inflammation are most effective. But not any anti-inflammatory can be used. For example, aspirin and ibuprofen don't help. Rather anti-inflammatories belonging to a group of medications known as immunosuppressive and immunomodulatory drugs work best. This includes:

1. Topical steroids (mid to strong potency) and steroid injections
2. Topical tacrolimus (Protopic) and topical pimecrolimus (Elidel)
3. Oral hydroxychloroquine (Plaquenil and generics)
4. Oral tetracyclines (doxycycline, tetracyline, minocycline)
5. Oral cyclosporine (Neoral, prograft, Sandimmune)
6. Oral mycophenolate mofetil (Cellcept, Myfortic)
7. Oral predisone (mainly for flares and early bridging treatment, not long term)

These 7 treatments have the best published evidence for assistance with lichen planopilaris. Any other treatment has less evidence.

Conclusion

Whenever a patient tells me they have tried treatments for lichen planopilaris and it didn't work, I want to know two things. First, I want to know if they truly have lichen planopilaris as there are many many mimickers. Biopsies can be wrong ... yes! and yes! Conditions like pseudopelade of Brocq can mimic LPP and so can a few other scarring alopecias (discoid lupus and folliculitis decalvans). The second thing I want to know is what treatments the patient has tried. I've heard countless treatments - perhaps well over 60 to date. Being on treatment does not count unless it's a potentially beneficial one.