

## What role does a biopsy play in the diagnosis of PRP?

Like many PRPers, Bill McCue's version of pityriasis rubra pilaris was misdiagnosed as seborrheic dermatitis and mistreated with escalating quantities of prednisone. After a week in the hospital and a fifth biopsy, the "official" PRP diagnosis was rendered.

As Bill read the stories of fellow PRPers in the PRP-List daily digest, he recognized a frequent lament:

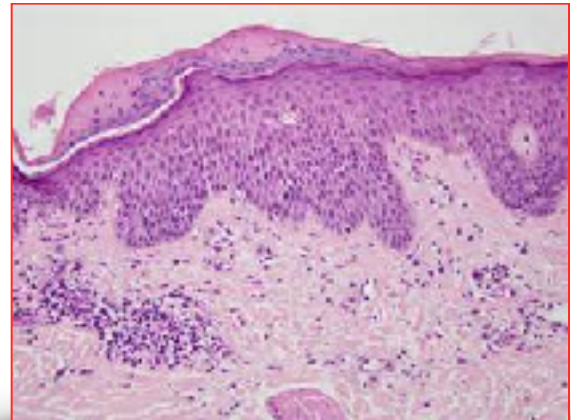
### Why should it take so long to diagnose PRP?

"In retrospect," Bill recalls, "my first dermatologist did not have PRP on her radar screen. The symptoms I presented were consistent with seborrheic dermatitis."

A biopsy in September and two in October did not confirm PRP. Moreover, a fourth biopsy performed at the Medical Center of Plano was also inconclusive. It was only when a new dermatologist ordered the fifth biopsy in late November *and specifically included instructions for the lab to consider PRP* that the results were classified as "a bit more characteristic and compatible with pityriasis rubra pilaris."

The dermatopathology report included an important caveat: "Clinical correlation is recommended." And that is the first answer to the question. We are told repeatedly that there is no "smoking gun" to be found in a skin biopsy. There is no "Gotcha" moment.

This reality begs another question: Are the characteristics of PRP really so elusive that they cannot be seen? Perhaps we should



interview 50 to 100 dermatopathologists as advocates of better PRP biopsies.

### The PRP Biopsy Poll

For several weeks in July 2012, over 29,000 messages maintained in the PRP Support Group archive were mined for two data points:

- \* Occurrences of "biopsy" and "biopsies"
- \* Email addresses of PRPers to be polled

As a result of this effort, a total of 487 PRPers were polled by email and given an opportunity to participate. Remarkably, the 2013 PRP Biopsy Poll reflects information shared by 256 (52.6%) PRPers

The long-term objective of the PRP Biopsy Poll and the resulting report was to initiate an ongoing dialogue within the PRP Community, e.g., dermatologists, dermatopathologists and PRPers.

Quite frankly, this important dialogue has languished — until now. The PRP Community should unite behind the following mission:

*As a diagnostic team, dermatologists and dermatopathologists must find ways to improve their ability to recognize PRP and*

*shorten the time between the onset of symptoms and a confirmed diagnosis.*

For those of us on the journey from onset through remission, a more timely diagnosis of PRP has no impact on us. At this very moment a person with a dime-size blemish on his/her forehead doesn't know whether or not it could signify PRP. How do we shorten the time this person might suffer due to the lack of a proper and timely diagnosis? It is the responsibility of those who are already on the journey. If not us, then who will make it happen?

### **Objectives**

The primary objective of the PRP Biopsy Poll was to better understand the role played by biopsies in the diagnosis of pityriasis rubra pilaris from the perspective of a PRPer. There were only two questions:

- \* How many biopsies have you had that supported a PRP diagnosis?
- \* How many biopsies have you had that failed to confirm PRP?

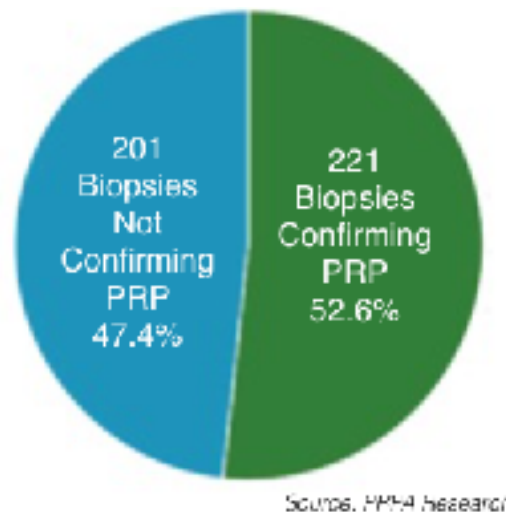
Within a three-week period, the response to the PRP Biopsy Poll reached 256 PRPer—more than double the original goal of 100.

### **Biopsies are a dice roll**

Dermatologists ordered a total of 426 biopsies for 176 individual PRPer. A total of 221 biopsies (52.6%) confirmed PRP while 201 (47.4%) biopsies did not.

The PRP Biopsy Poll also suggests that when a dermatologist instructs a pathologist to consider PRP, the results nearly always confirm PRP.

Clearly, additional polling must be undertaken to identify ways in



which PRP can be "targeted" earlier in the diagnostic process.

It should also be noted that 117 responders provided unsolicited anecdotal commentary about their biopsies which served to underscore the need for additional polling.

### **Biopsy Overview**

The role of a skin biopsy is to aid in the diagnosis of PRP. Responders to the PRP poll fell into one of four general categories:

1. *No biopsies ordered (20 replies — 7.8%).* Diagnoses were made strictly on clinical observations.
2. *Biopsies that support a PRP diagnosis without a contradictory biopsy. (116 replies — 45.3%).* The results of the biopsy provided sufficient information to support a diagnosis.
3. *Biopsies that confirm PRP and contradict other "non-confirming" biopsies. (60 replies — 23.4%).* Non-conclusive biopsies were ignored when PRP was "consistent" with a PRP diagnosis.
4. *Dermatologists who made a PRP diagnosis without the benefit of a confirming biopsy. (60 replies — 23.4%).* The clinical observations of



dermatologists provide sufficient evidence that PRP was the appropriate diagnosis.

### **Biopsies: Just a Tool**

It appears that the most reliable PRP diagnoses — the ones that stand the test of time — either combine clinical observations with supporting biopsies or reflect past experience with PRP.

#### **Editors Note:**

**On January 3, 2015, Becky Schock jump-started a conversation on biopsies. An edited version of the responses is offered here.**

#### **Becky S — Walnutport, PA**

Has anyone had positive biopsy results for PRP? Hubby changed docs and they ran biopsies for PRP and cancer. All inconclusive, but the cancer one leans more to it NOT being cancer.....very confusing

#### **Natalie M — Glasgow, Scotland, UK**

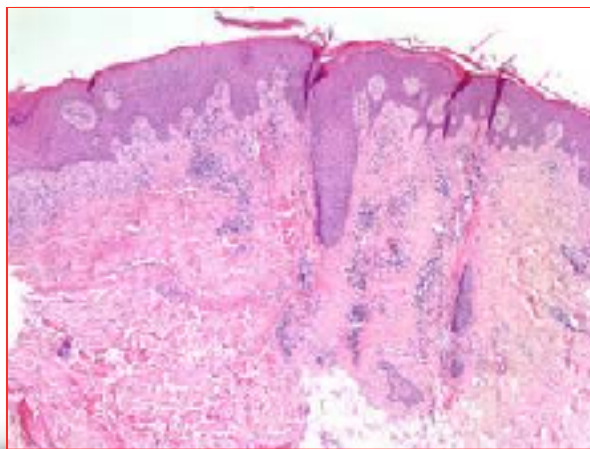
Our doc said a biopsy can't conclusively diagnose PRP; that's done by clinical observation. A biopsy is backup and the one for our wee boy just said the findings were most compatible with PRP. Hope this helps. All very confusing. We're very much still at the learning stage too.

#### **Sharon T — Scottsdale, AZ**

It is confusing, isn't it? My biopsy came back inconclusive. Well, my symptoms and the course of the disease are definitely PRP. And my dermatologist took one look at me and knew exactly what it was.

#### **Estrelita O — Port Elizabeth, South Africa**

I was diagnosed with psoriasis and went for second and third opinions. The dermatologist



who saw me told me the first time, its PRP, did a biopsy and my results came back positive.

#### **Margie D — Queensland, Australia**

My biopsy results definitely showed all the signs of PRP and confirmed with my symptoms etc.

#### **Ginny M — Lexington, SC**

I have had biopsies done at Vanderbilt, Mayo Clinic, the Medical College of Georgia, a private practice in South Carolina as well as Duke. I doubt they use the same lab. They all came back PRP. My first biopsy was done at the age of four and my last was done when I was 30 years old.

#### **Jerri B — Nathalie, VA**

The answer I got from a biopsy sent to the University of Virginia was ... "It looks like it." My doctor said we would get this answer.

#### **Rebecca L — Las Vegas, NV**

If they don't take an appropriate sample size, the biopsy for PRP can come back negative. My husband had to have two or three biopsies run, in order to confirm a PRP diagnosis.

#### **Missy E — Denton, NC**

My doctor said, unless they are specifically looking at the biopsy as PRP, then it often comes back as something else like psoriasis. My first doctor thought psoriasis and that's

what the biopsy initially said. Second doctor said PRP as soon as he saw me. He asked for a second look at the same biopsy and confirmed PRP. I think it takes both an experienced doctor and an experienced lab, to see it since it looks so similar to other things.

**Judy F — Kingsport, TN**

My saga is almost exactly as Missy Miller Ellison's. Though I think even the second doctor's wording was "consistent with PRP..."

**Karen B — North Wales, UK**

My biopsy results were reported to me as confirming the diagnosis of PRP, but when I asked was it type 1 or type 2, my specialist admitted "the results do not provide that information." So, if it is not gone in 3 years, I guess I have type 1. (18 months post diagnosis, and counting )

**Glen M — Glenview, IL**

Two of four were consistent with PRP. For some reason, I think they need to be taken with a hair follicle. The first two were not. Not every dermatologist knows this.

**Pat N — San Diego, CA**

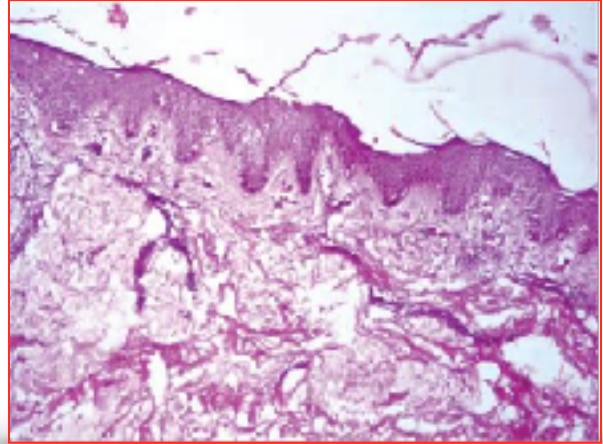
Yes. One of my five biopsies was positive. The rest were not. You are correct Glen M. The biopsy has to include a hair follicle to be diagnostic for PRP

**Laurel A — Kennewick, WA**

My dermatologist said "the biopsy was most consistent with PRP." No mention of which type.

**Lorna R — Eugene, OR**

Basically what a skin biopsy does is rule out other causes.



**Glen M — Glenview, IL**

Interesting discussion. Let's remember that an effective diagnostic tool must be accurate, reliable, produce consistent results and not have false results lead to inappropriate treatments. Finally it must be cost effective. As such, the dermatology community needs to become better able to use this tool in testing for PRP. The survey results reveal a very high proportion of inconclusive results. In most medical specialties this would be unacceptable. Given the number of inconclusive results and then correct results when redone, one must question the skill and knowledge of the dermatologist taking the biopsies. I know that my initial biopsies had to be redone because they did not include hair follicles. This wasted time and money leads to initially inappropriate treatment. Performed correctly, biopsies produce the proper results.

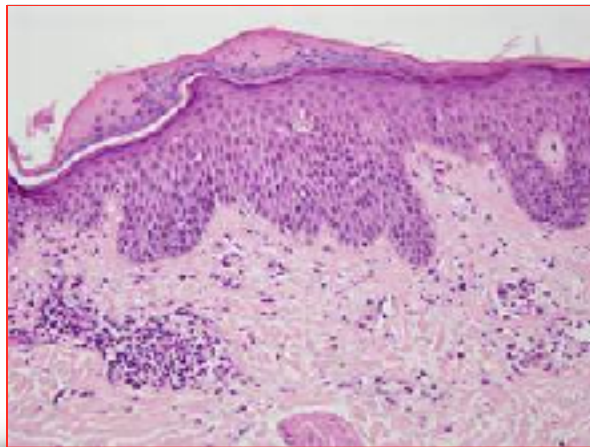
**Bill M — Plano, TX**

Everyone seems to agree that biopsies are a tool that may reinforce the clinical observations of the dermatologist. If the dermatologist has seen PRP "in the flesh", they consider it as a possible diagnosis sooner than a dermatologist who has absolutely no PRP experience.

I have spoken with dermatopathologists and they do look for "indicators", but there doesn't seem to be a "smoking gun" or a "Eureka" moment. I dug up the fifth Dermatopathology Report (Dr. Lydia Essary) and it said: "The findings were compared to the previous biopsy (CT12-204498) and the features in the current biopsy are a bit more characteristic and compatible with pityriasis rubra pilaris. Clinical correlation is recommended. Dr. Clay Cockerell has also reviewed this case and concurs with the diagnosis." The actual diagnosis was rendered by my dermatologist who — earlier in his career — had been Chief Resident of Dermatology at University of Texas Southwestern in Dallas. Bottom line: it's the clinical observation of a dermatologist who doesn't get sucked down the rat hole of psoriasis, seborrheic dermatitis and other skin maladies that seem to mask PRP. I wonder how many people with a psoriasis diagnosis who are NOT responding to the meds appropriate for psoriasis are, in fact, PRPers waiting to be found?

**Lorna R — Eugene , OR**

I often wonder the opposite. How many people who are being treated as PRP patients, actually have a form of psoriasis and or a form of CTCL. I especially wonder that when I see adults here who have had this "PRP" for a protracted period of time and are not improving. I wonder about the people who were diagnosed as having PRP, and their skin responds to the treatment with biologicals, which are designed to treat psoriasis, and other autoimmune disorders. PRP has some classic distinctions, the orange hue to the skin, the build up of Keratin on the palms of the hands, and plantar surface of the feet. These



classic presentations along with the skin biopsies ruling out things like TCL, are usually how the experienced clinicians make the diagnosis of PRP. Just thinking.

**Brenda M — Kent, England, UK**

Lorna, Adult Onset A typical PRP can last 20 years or more. Biopsy's are used to rule out other conditions together with Clinical Observations by the dermatologist. A typical PRP does not usually involve hands and feet.

**Sam D — Sydney, NSW Australia**

I had three biopsies taken at the one time. Left arm, left shoulder and left abdomen. All came back with "Consistent with", and "Other clinical suggestions can be excluded". Doesn't make mention of either positive or negative.

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**OTHER "BIOPSY" COMMENTS**

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**Pat N — San Diego, CA**

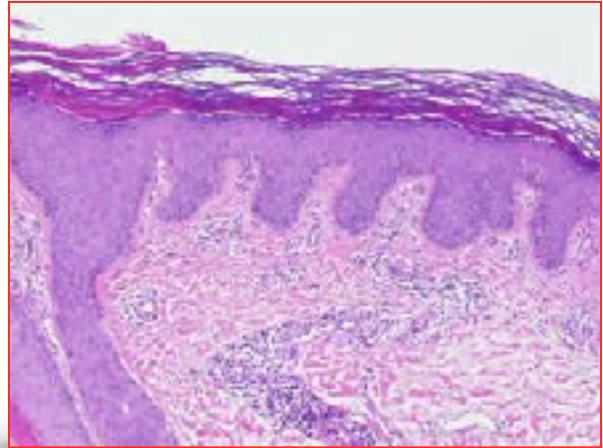
Email dated January 8, 2015

I found the recent Facebook discussion on biopsies very interesting. My own experience with this might shed some light on the topic. I am a retired surgical pathologist and have read my share of biopsies, though my practice

was not in dermatopathology. I am very familiar with the terminology that pathologists use to protect themselves in a pathology report, even when they are 99% certain of the diagnosis. In my own case, after my second biopsy, (the first was inconclusive) I contacted the pathologist reading my slides and spoke to him directly. He is an excellent dermatopathologist who also happens to be board certified in dermatology. I called the pathologist because I was having difficulty reaching my dermatologist, who was not very efficient about returning calls and I was understandably very anxious. I had a thorough discussion with the dermatologist who told me that he was certain that I had PRP on the basis of the biopsy and that he would issue his report accordingly and send me a copy of the report. I received the copy of the report which used the familiar wording of "consistent with PRP.....clinical correlation recommended" I was convinced that I had PRP and that the pathologist was using that kind of wording to protect himself against a possible malpractice claim in the unlikely event that this was something else.

A few days later, I received a call from my dermatologist and she interpreted the same report in a completely different manner. She claimed that I did not have PRP because I did not have the clinical features and it was as if the dermatologist had a completely different report in front of her than the one I had. I was not started on PRP treatment at that point. Needless to say more PRP manifestations erupted very quickly after that and I changed dermatologists and eventually started the correct treatment.

So, in my case, the biopsy results preceded the full blown clinical manifestations of PRP.



#### **Will Sivilli — Tucson, AZ**

Email sent to PRPA dated January 24, 2015

I had two biopsies that "confirmed" PRP.

#### **Karen H — Savannah, GA**

Email sent to PRPA January 25, 2015

I wanted to share my biopsy experience. The first dermatologist said I had a photosensitive reaction to something I was taking. Went off herbs for a period of time and found another, dermatologist who took two biopsy sites.

My report says, "The epidermis appears mildly acanthotic. Within the superficial dermis, there is a lymphocytic infiltrate with rare exocytosis in the epidermis. There are no prominent interface changes. The common "checkerboard" pattern of orthokeratosis and parakeratosis is not seen in this specimen definitively. However there are features suggestive of it. The overall features of this biopsy favor that of PRP over psoriasis. Of note, guttate psoriasis may show similar features. Clinical correlation is recommended.

I'm not really sure the above is a definitive PRP diagnosis. The derm assured me I have the disease. The dermatologist recommended methotrexate immediately and no other course of treatment. After listening to all of you, I decided to not follow his suggestion. I sought alternative medicine.

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**Margie Dillon** (Original Post)

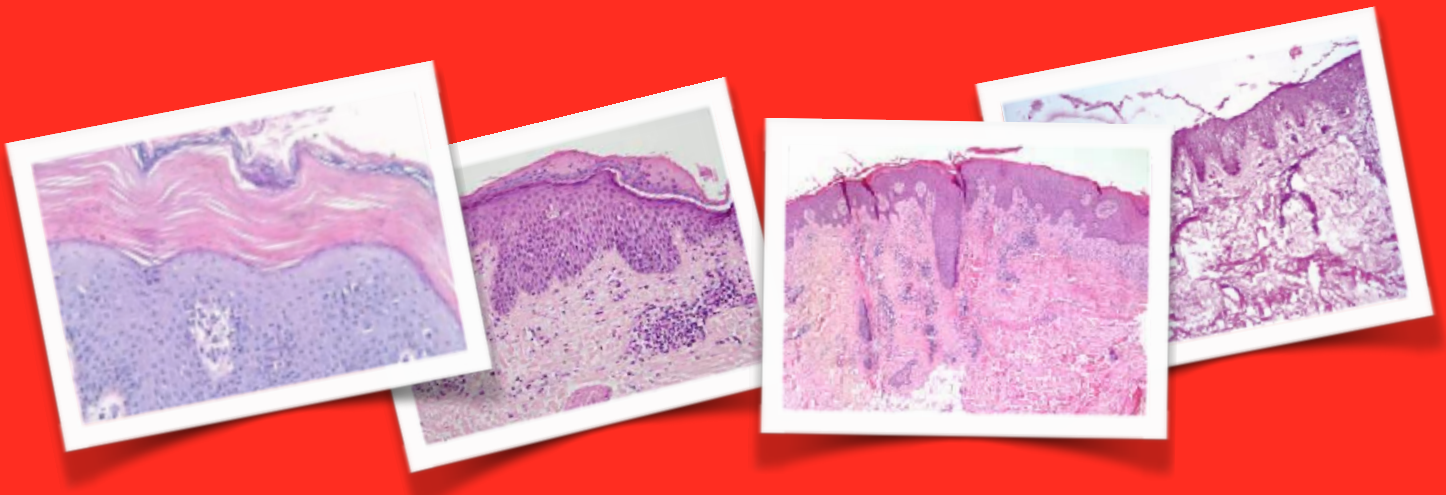
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February 1 at 5:18am

Yesterday I was a guinea pig (lab rat of sorts) at the hospital where I attend dermatology appointments. I had at least 50-60 doctors, consultants and med students check me out, make observations etc. I was asked many questions before they all met and presented our cases. The most interesting person I spoke to was a pathologist who openly admitted in

his latest book that he has never diagnosed PRP with one biopsy. I must be one of the lucky ones as I was diagnosed on the first biopsy. He has recently discovered that the optimum time to do a biopsy is between days 24-28 of the presentation of new lesions. I found that very interesting! I am hoping with all of those experts/minds in one place, I may be given a treatment plan that works! Fingers crossed

## The PRPer Takeaway re: Biopsies



**Biopsies can help a dermatologist diagnose PRP. Unfortunately, biopsies either “rule out” other skin maladies, are “compatible” with PRP or are “inconclusive.” It is always recommended that biopsies be used in concert with clinical observations. Every PRPer should know the role played by biopsies in his or her PRP diagnosis.**

## What role did biopsies play in the diagnosis of your version of PRP?

### Editor's Note:

Every section of the PRP Survival Guide is a work in progress. The preceding article appeared in the February issue of On the Road and "jumpstarts" the discussion.

#### Andy W — Reidsville, NC

I went through that at UNC. It was a little girl from South America that diagnosed me. I had several biopsies.

#### Trine T — Aarhus, Denmark

My prp has never shown in biopsies...but there is no doubts about the diagnosis.

#### Karen B — Rochester, NY

I feel ya Margie,, I did that when I was 17 years old I hated it. Finally the head of Dermatology finally came in the room and new right away what it was. However they did a biopsy just to make sure. I hated it having everyone look at me in awe and feel like a guinea pig as you put it. But it great to know the answer instead of guessing at things it might of ben. Strong lady to put up with that. Hang in there

#### Janice F — Stevens Point, WI

You're helping educate them, too! My husband also did this, part of Grand Rounds at the teaching clinic he went to. Half of the doctors said yes, you have PRP. The other half hadn't seen it before. We thought we would hear feedback after their discussions but we didn't. Hope you get some good information.

#### Margie D — Queensland, Australia

I was diagnosed last year through biopsy after being treated for psoriasis for months. I found it very interesting and there was only one doctor who questioned me having PRP. He asked me how did I know it was PRP? Um.....biopsy?? LOL

#### Mickie C

I was diagnosed 1st with psoriasis with a biopsy, 2nd time plaque psoriasis , 3rd biopsy PRP at Duke hospital

#### Greer C — King George County, VA

What Bill was told at his last Dermatology appointment was that biopsies 'rule out', as opposed to 'diagnosing' a condition. After they ruled out everything else, they came up with PRP. That is what happened in Bill's case. I suppose in some cases the symptoms are so classic that biopsies only back up the diagnosis. Bill had 4 or 5 biopsies. He went to Johns Hopkins for a 2nd opinion and they took one look at him and said he was a classic case of PRP.

#### Mary H — Novato, CA

Righto, PRP cannot be confirmed by biopsy, according to UCSF MDs. A biopsy is generally to make sure it isn't something similar (worse, perhaps). To that extent, PRP can be diagnosed with biopsy because the other diseases are not present.

#### Margie D — Queensland, Australia

I'm in Australia and was diagnosed by biopsy. If we can't go by biopsy, how do we get a diagnosis?