60-SECOND
PRP ROADMAP

THE PREDICTABLY UNPREDICTABLE Journey for Patients Diagnosed with PITYRIASIS RUBRA PILARIS

PEER REVIEW WORKBOOK

Bill McCue

ISBN: 978-1-963764-61-1

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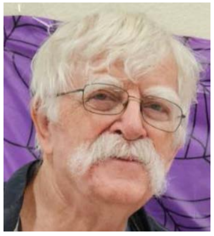
PRP ALLIANCE



DATE: July, 2024

TO: PRP Patients, Caregivers & RP-savvy dermatologists

FROM: Bill McCue, PRP Alliance

SUBJECT: 60-Second PRP Roadmap

No one has ever written a book about pityriasis rubra pilaris from the patient perspective. In fact, no one has ever written about PRP from any perspective. I am asking you to help us make medical history.

My PRP journey began in early August 2012 with the appearance of a red spot on my forehead. It took akmost four months before I was properly diagnosed with PRP. I had no roadmap. I learned about PRP from other PRP patients.

What is the Peer Review Workbook?

- ☐ The Peer Review Workbook is a draft of the 30-Second PRP Roadmap scheduled for publication in March, 2024 and reflects the insights and experiences shared by PRP patients and caregivers since 1997.

Who will participate in the Peer Review?

- ☐ Members of the PRP Support Group.
- ☐ Dermatologists who are currently treating PRP patients or have treated PRP patients in the past.
- ☐ Advocates of patients diagnosed with rare skin disorders will be invited to participate, e.g., the International Organization of Dermatological Patient Organizations (GlobalSkin), the National Organization of Rare Disorders.

What should you be reviewing?

- ☐ What's *missing* or *incomplete*? I can add it.
- ☐ What's *incorrect*? I can fix it.
- ☐ What's *cnfusing?* I can make it make sence.

How to save what you have reviewed?

- ☐ Please take notes as you read.
- ☐ Space has been allocated at the end of each Section and each Appendix for peer reviewers to jot down any observations you might want to share.
- ☐ "Empty box" bullets have been used to make it easier to "tag" a problem or issue needing to be reported.
- ☐ Write notes in the green areas, yellow areas labeled Peer Ponderings,

  and margins. This is a workbook!

How will be share what you have learned?

- ☐ Online survey
- ☐ ZOOM Focus Groups scheduled to accommodate ALL time zones.

Then what?

- ☐ The comments of all peer reviewers will be integrated into the FINAL manuscript and delivered to Amazon Book Publisher.

Thank you for helping the PRP global communuity tell our story.

What's missing, incomplete, incorrect or confusing?

DEDICATION

It was a dark and stormy night. No kiddin'. There are many dark and stormy nights for patients diagnosed with pityriasis rubra pilaris.

The 30-second PRP Roadmap is dedicated to the PRP patients and caregivers who journey together and endure the daily challenges of body, mind and spirit. To those who:

- ☐ ***post*** questions, announce good news, and vent bad news to members of the PRP Support Group.
- ☐ ***comment*** on the sharings of other PRP patients and caregivers.
- ☐ ***react*** with appreciation to the posts and comments of the PRP Support Group with Like, Love, and Care.
- ☐ ***participate*** in PRP research, whether sponsored by the PRP Alliance or third-party researchers, e.g., Oregon Health and Science University, Thomas Jefferson University, GlobalSkin, USC, UCLA, etc.
- ☐ ***go the extra mile* to help** the PRP Alliance meet the financial obligations to GoDaddy, Constant Contact, Claris International, Harmonic Data, ZOOM, Office Depot, and Amazon Book Publisher.

In other words, the 60-Second PRP Roadmap is dedicated to the PRP global community.

INTRODUCTION

The *60-Second PRP Roadmap* was written for three audiences of readers: PRP patients, PRP caregivers and dermatologists. All three face the unique challenges of our ultra-rare skin disorder. However we focus on the patients

PRP patients are the travelers

- ☐ PRP is a unique journey from onset to someplace else.
- ☐ As PRP patients, we have little or no control over the road ahead.
- ☐ We are unwilling riders on a roller coaster that brings terror when we are most vulnerable.

PRP patients need not travel alone

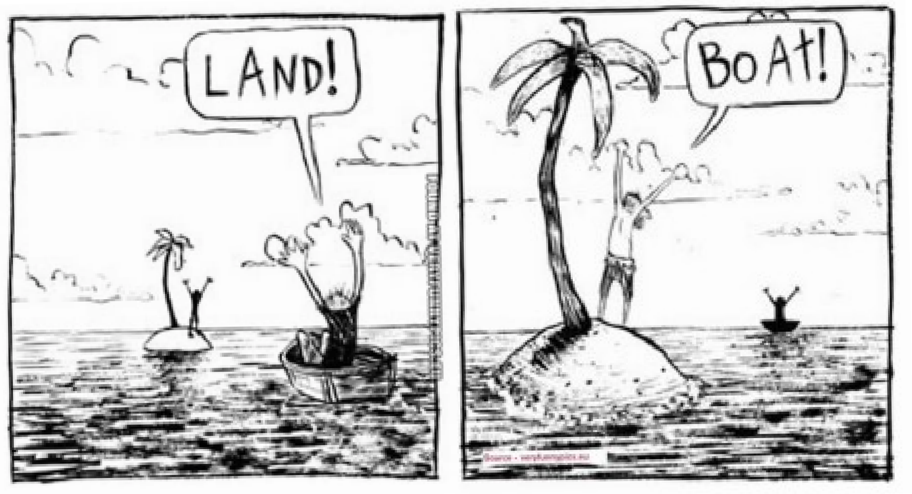
- ☐ We can travel with others who are willing to share what they have learned. We are kindred spirits.
- ☐ The more we share, the more we all learn The more we learn, the less we fear.

PRP patients benefit from the experiences of other PRP patients

- ☐ We lighten the burden of PRP by sharing that burden with the PRP global community.
- ☐ We find emotional support from kindred spirits and minimize loneliness and feelings of isolation.
- ☐ We find strategies with which to cope with pain, anxiety, stress, depression, and frustration.
- ☐ We identify effective treatment options based on what worked for others.

It’s all about the reader — YOU.

The primary target audience for the 60-Second PRP Roadmap is PRP patients and PRP caregivers. The secondary audience are dermatologists.



Patient Perspective

☐ The *PRP Survival Guide* has been a standalone website maintained by the PRP Alliance since May 2015. Hundreds of archived articles reflect the insights and observations of a thousand PRP patients and caregivers. The two questions the 60-Second PRP Roadmap asks:

- What does a PRP patient or caregiver need to know?
- What does a PRP patient or caregiver want to know?

Focus

☐ What does a reader need to know? The 60-Second PRP Roadmap is a broad overview of the basics. The goal is to make the most important information readily accessible and digestible.

Lay language

☐ There is a time and place for technical terms used to explain PRP. However, a newly diagnosed PRP patient needs clarity, not technical terms. An effort has been made to keep the text simple. The Plain Writing Act of 2010 requires federal agencies to write "clear communication that the public can understand and use.” While we are not a federal agency, the 60-Second PRP Roadmap is written to be easily understood and used.

Subheads and Bullets

☐ Subheads are bullets that are like spotlights on a darkened stage that help readers navigate each page. The purpose of the “empty box” bullets is to organize the flow of information and make it more digestible. Patients and caregivers are encouraged to check a box when reading something worthy of note.

60 Seconds Per Page

☐ The average reading speed for learning is 100 to 200 words per minute (WPM).

☐ The 60-second PRP Roadmap represents nearly 50,000 words.

☐ Depending on your readking speed, four to eight hours of information about PRP from the patient perspective seems well worth the effort.

READ THIS BOOK
WITH A PEN IN HAND.

IF YOU CAN'T HOLD A PEN,
RECRUIT A HELPER.

1. PRP patient's name: ____

2. Today's date: (mm/dd/yyyy):______________________

3. PRP patient's onset date (mm/dd/yyyy):______________________

4. PRP patient's onset age? ______________________

THIS IS YOUR PRP ROADMAP!

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> *When my PRP journey began, I was alone and utterly terrified. I saw hopelessness in my mirror. But the flame of hope was ignited when I found the PRP Facebook Support Group. The flame shone brighter as I reached out to my new global family, all on a similar journey. Hope springs eternal.*

— Carol T, Swaffham, England, UK

SECTION ONE
First Questions

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1. What is PRP?

The first definition I saw as a newly diagnosed PRP patient went like this: "… a rare inflammatory papulosquamous disorder of unknown etiology. Includes distinct, well-demarcated plaques of various sizes with characteristic reddish-orange hue." Try these observations!

- ☐ In Latin, PRP stands for scaling (*pityriasis*), redness (*rubra*), and hair follicles (*pilaris*),
- ☐ Every PRP patient and every PRP journey is as unique as a fingerprint or a snowflake, PRP is an ultra-rare skin disorder.
- ☐ "I like to use über-rare because I think that über sounds more rare than ultra. I also use disorder because disorder sounds less contagious."
- ☐ PRP may impact different parts of the body in a variety of ways for unpredictable periods.
- ☐ PRP often appears as a small spot on the face *(or not)* and then spreads elsewhere (usually).
- ☐ PRP inflames the entire body (*or just parts)*, thickens the soles and palms *(or not)*, can affect elbows and knees *(or not)*, causes loss of hair *(or not),* and can
- ☐ make strange things happen to fingernails and toenails *(or not)*.

  PRP may leave clear areas of skin called "islands of sparing" *(or not).*

- ☐ PRP may move slowly or quickly. It may come and go or come and stay. Get the picture?
- ☐ While every PRP journey is predictably unpredictable, we share a common bond: the daily challenges of body, mind and spirit.

2. What Constitutes a Rare Disease?

While not as unique as a unicorn, rare diseases represent a significant medical challenge. Here are a few facts to better understand the uniqueness of PRP.

- ☐ The Orphan Drug Act is a law passed by the US Congress in 1983 that defines a disease or condition as “rare” if it affects less than 200,000 people.
- ☐ The National Institutes of Health, Genetic and Rare Diseases Information Center has identified over 7,000 different types of diseases, disorders and conditions worldwide. A total of 608 are considered rare skin disorders.
- ☐ There are an estimated 30 million people in the United States living with rare diseases.
- ☐ International definitions of rare diseases vary. For example, in the UK, a disease is considered rare if it affects fewer than 50,000 citizens per disease.
- ☐ Approximately 50% of rare diseases do not have a disease-specific foundation supporting or researching their rare disease.
- ☐ According to the National Organization of Rare Disorders, more than 90% of rare diseases are still without an FDA-approved drug treatment.

I loved hearing Kermit the Frog sing, “It’s not easy being green.” It’s also not easy being red and even harder being red and rare.

3. What are the Signs and Symptoms of PRP?

A *sign* indicates a medical condition that others, including your doctors, can observe objectively. The first signs of early onset are most likely seen by the patient or a family member.

"My red dot was nestled into my hairline on my right temple.
I saw it in the mirror while shaving."

SIGN = SEE

In contrast, a *symptom* is information patients share with their healthcare professionals, e.g., answers to questions, unsolicited comments, etc.

"The only way my dermatologist would have known
I could only sleep three hours at night
was for him to ask or for me to tell him! I told him!"

SYMPTOM = SHARE

At some point during early onset, the patient decides that it's time to see a dermatologist.

4. How Does PRP Progress?

After spending over a decade reading or listening to stories shared by thousands of PRP patients and caregivers, I have come to the conclusion that two metaphors will aid in (a) telling your own PRP story to others and (b) understanding your own PRP journey.

PRP IS A JOURNEY

☐ PRP is a journey that starts somewhere with a warning sign of some sort, e.g., spot, rash, etc., and then follows a path all on its own. Every PRP journey is unique. Patients and caregivers are travelers who learn as they go. Sometimes, the road ahead is plagued by hazards and obstacles. There are other times when our road is filled with hope.

PRP IS A FOREST FIRE

☐ PRP is more like a forest fire. Once it starts, it grows. It can get out of control, and, for most, it either is put out with the intervention of firefighters or stops burning.

PRP IS UNPREDICTABLE

☐ Some say PRP will last two to three years; some say three to five years. And some are in it for the “long haul” or six-plus years. A few discover they have a chronic version, and their journey lasts a lifetime. I was one of the lucky ones. My journey lasted only 20 months.

5. Where there's smoke, there's fire (Onset)

The visible signs may have been disguised as dandruff or spots. Nothing special. Indeed, nothing to worry about. In my case, it was a dime-sized spot that appeared on my forehead tucked into my hairline on my right temple. It was in early August 2012 when I saw the smoke.

Some of us know exactly when the Red Dragon appeared, or, at least, we have one or more suspects? But most of us don't have a clue. I had three suspects:

☐ A drop of paint remover hit my forehead while refinishing a bookcase,

☐ stress associated with my efforts to come out of retirement as the director of communications for a national franchise and the stress of an unplanned encounter with wife #2.

Some of us even know when the first sign of PRP first appeared. Most don't. On August 8, 2012, I took a selfie at the top of a hill I had climbed at Marine Corps Base Camp Pendleton in California. Almost a year passed before I took a closer look at the photo. There it was. I saw a dark blemish in the hairline on my right temple.

At some point, we recognize that something is happening with our skin. It could happen quickly or at a snail's pace. But the end result is always the same — we seek the help of a healthcare professional.

6. Out of control (Acute)

Most generally called the Acute Stage, the Red Dragon is out of control, and every body part is fair game. Dry, red, flaking skin may engulf the entire body.

- ☐ Quality of life has reached a new low as PRP patients face the most significant challenge to body, mind, and spirit. The duration is unpredictable, lasting weeks or months.
- ☐ Symptoms become sinister, e.g., the pain of motion, unrelenting itch, heat intolerance, sleep deprivation, loss of energy, overall weakness, and inability to regulate body temperature.
- ☐ Mobility is compromised as the soles of feet become encased in thick, cracked, swollen and bleeding skin.
- ☐ Dexterity is compromised as fingernails either disappear or grow thick and gnarly. Hands may become victims of swelling. Vision may be compromised by tightness and pulling of the eyelids.

PRP patients and caregivers should seek support from kindred spirits. Don't suffer alone or in silence. Look for Healing Milestones that will signal the end of the Acute Stage.

7. Working the Fire (Management)

Getting a fire under control is no small feat. The goal is to stabilize your PRP journey and focus on actions to mitigate and manage signs and symptoms on many fronts. No easy task. This was my to-do list in the Acute Stage.

- ☐ Develop a Nightly Routine (10-11 PM) that included a warm bath, removal of flaking skin, and the slathering of Clobetasol and urea lotion (soles), Desonide (face) and triamcinolone (remaining nook and crannies).
- ☐ Sleep through the night. With the help of my dermatologist, we crafted a cocktail of Tylenol, Ambien, and hydroxyzine. If I didn't drink any liquids after 9 PM, I could sleep until 7 AM.
- ☐ Stay mentally active. Once I had enough energy to sit at my computer (circa March 2013), everything changed for the good. As a member of an email-based support group, I had access to the archive of emails going back to 1997. Over four months, I read over 29,000 emails and built a PRP database.

Make the most out of every clinic visit. Learn more about PRP than the dermatologist. Learn what other PRP patients have experienced.

8. Smoldering embers (Maintenance)

PRP maintenance is all about managing expectations. How can we slow the progress of our own version of PRP? I soon realized that my progress in PRP is best seen in a rear-view mirror. Day to day, I saw no change. Looking back, however, disclosed progress.

- ☐ I prioritized the symptoms that bothered me the most. That was easy: impaired mobility. My feet were swollen, and my soles were encased in thick skin that was cracked and bleeding.
- ☐ With a proper PRP diagnosis (four months into my journey), I was prescribed acitretin and a combination of Clobetasol® and urea lotion for the soles of my feet. Within six weeks, I was wearing sneakers abandoned four months before. I was walking again.
- ☐ Sleep deprivation was the next problem on my list. I eventually had an evening regimen and a pharmaceutical "cocktail" that guaranteed eight hours of sleep.
- ☐ There are no shortcuts. PRP patients have little or no control over what works and what doesn't. I kept working on my list of "issues" and gradually converted them to Healing Milestones.

One by one, my list of active symptoms grew shorter and shorter. I was working the fire!

9. Extinguished (Endgame)

Even when the firestorm is contained, and the last of the burning embers have been extinguished, the remnants of the inferno remain.

- ☐ Neither PRP-savvy dermatologists nor the PRP global community of patients and their caregivers have an agreed-upon standard definition of remission. Sage advice from one of my daughters comes to mind: Whatever floats your boat.
- ☐ Every PRP journey is unique. It has taken me years to understand and fully appreciate that "med-free and symptom-free" is not the endgame for every PRP journey.
- ☐ Remission is absolutely in the eye of the beholder and can include *remission with remnants*.
- ☐ The remission is a decrease in or disappearance of the signs and symptoms of PRP. The caveat is whether or not meds are needed.
- ☐ Here's a six-word gem: "I finally have my life back."
- ☐ For some, "partial" remission means that PRP may still be "lurking," and the phrase "smoldering below the surface" comes to mind.

Over time, flora and fauna return to the forest. So, too, for PRP.

10. Is there a cure for PRP?

According to the Merriam-Webster Dictionary, a CURE usually refers to a complete restoration of health. In contrast, *TREATMENT* refers to a process that improves health but may not include the complete elimination of disease.

☐ The consensus among dermatologists is that there is NO CURE for *pityriasis rubra pilaris*. This is NOT what a PRP patient or caregiver wants to hear.

☐ While there is no "Magic Bullet" to make PRP disappear, there are remedies to relieve specific signs and symptoms. Unfortunately, what works for one doesn't work for all.

☐ We often hear that PRP will "run its course." For many, "a tincture of time" is another unsatisfactory option, given the indeterminate length of a PRP journey.

☐ The best we can hope for and work towards is to be symptom-free, med-free (PRP-specific drugs) and free of relapses.

While "Find a Cure" sounds like a worthy objective, the Federal Drug Administration has yet to approve a treatment for PRP. Simply stated, our patient population in the US is too small to make a traditional drug trial cost-effective. An inconsequential patient population is an insurmountable barrier to commercial investment in the research and development required to show evidence of the safety and efficacy of treatments.

In the US alone, there are 25-30 million patients who, together, suffer from approximately 7,000 rare diseases—some of which affect as few as about a dozen individuals. Advocating FDA-approved treatments for PRP is still a noble cause.

11. What are Healing Milestones?

Healing Milestones are one way for PRP patients and caregivers to measure progress. The PRP global community has adopted a celebratory approach to disease recovery, recognizing various healing milestones.

In my humble opinion, the first healing milestone is the "official" diagnosis of PRP (clinical observation of a dermatologist supported by a biopsy and a dermatopathologist). No more guessing. Here are a few more markers.

The foillowing milestones are examples of symptoms that PRP patients and caregivers *feel* and signs that everyone else *observes*.

☐ Had "jelly beans" (skin and wax) removed by an ENT specialist.

☐ Wearing earrings for the first time in months.

☐ Going back to work.

☐ I was able to button buttons and zipper zippers.

☐ To get eight hours of sleep and to set my alarm for 7:00 AM.

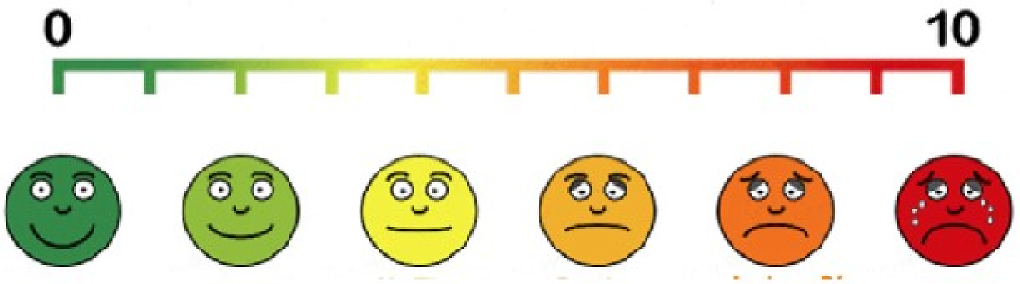
☐ Opening a jar of Vlasic Sweet Relish without a pair of pliers.

☐ Drive myself to Walmart.

- ☐ I kicked a soccer ball around with my daughter.
- ☐ I can flex my toes without them cracking.
- ☐ Not refilling a prescription for acitretin after 12 months.
- ☐ Drive myself to Walmart and NOT use the electric cart for disabled customers.
- ☐ Swept the dark hardwood floors and realized there was more Old English Sheepdog than me.

- ☐ Scheduling an ANNUAL clinic visit rather than a three-month or six-month return.
- ☐ The feet are no longer swollen. Put on sneakers for the first time in three months.
- ☐ Sweating again.
- ☐ No longer need to wear a sauna suit.
- ☐ Taking the first pain-free and nearly tingle-free shower in age.
- ☐ I needed a haircut (I lost hair for six months).
- ☐ Today, I've been able to walk, by which I mean putting one foot in front of the other and going farther from my bed to the bathroom or from my bed to the kitchen. Really walked for the first time in over eight months.

A Healing Milestone is a sign of progress and hope. Perhaps the PRP Support Group should issue merit badges for every Healing Milestone we celebrate!



12. What are the “Quality of Life “Impacts of PRP?

A series of “Quality of Life” focus groups were organized in April and July (2021) involving 40 PRP patients as part of ongoing research at Oregon Health and Science University. The following is the Discussion Guide used to conduct those focus groups. The questions help to frame the issues related to “Quality of Life” as they pertain to PRP.

Physical Impacts

☐ Please tell us about your initial experience with PRP. What was the first sign that something was wrong?

☐ Please discuss your experience with the physical symptoms of PRP.

☐ What symptoms bothered you the most? Did that change throughout your PRP journey?

☐ What symptoms bothered you the least [annoying rather than serious], and did that change throughout your PRP journey?

☐ Are there any symptoms of PRP that you feel were not given adequate attention *by your dermatologist*? If so, what were they?

☐ What was the FIRST SIGN that you were *getting better*?

☐ What other symptoms we haven’t discussed yet should be given adequate attention in a PRP quality of life questionnaire?

Psychological Impacts

☐ If you are comfortable, please share how PRP affected your mental health.

☐ Is/was your mental state particularly affected by specific symptoms or limitations imposed by PRP? If so, what are they?

☐ At what point, if any, during your PRP did you feel you had hit “rock bottom?” What made you think that way?

☐ At what point in the course of your PRP did your mental state start to improve?

☐ What kinds of coping strategies did you use to manage your mental health?

☐ Can you think of any ways that your physician could have helped you improve your mental health while you had PRP?

Daily Living Impacts

☐ Please tell us about how your daily routine is/was impacted by PRP.

☐ What daily activities are made more difficult because of PRP symptoms?

☐ Are there any activities of daily living (personal hygiene, mobility, eating, etc.) you are/were unable to perform without assistance?

☐ How did you adapt your lifestyle to fit your limitations, if any?

☐ How was your sleep affected by PRP?

Social Impacts

☐ Did PRP change your relationships with family, friends, or coworkers? If so, how?

☐ Did PRP change the way you interacted with strangers?

Financial Impacts

☐ Did PRP's physical, psychological, social, or daily living effects affect your finances in a significant way?

☐ Did the cost associated with PRP medical care significantly impact your finances?

13. What are the Metrics Of PRP?

PRP is an über-rare skin disorder. You should know the metrics for where you live. For example, based on the estimated population for 2023, there are 841 PRPers in the USA, 78 in Texas and 16 in the Dallas-Fort Worth area. It does include me because I'm in remission.

PREVALENCE —In March 2003, Dr. Andrew Griffiths, a London-based dermatologist, delivered a "Dowling Oration" to members of the British Association of Dermatology (BAD) assembled in Liverpool, England. Dr. Griffiths reflected on 35 years of diagnosing, treating, and researching *pityriasis rubra pilaris*. He unilaterally set the PRP prevalence rate at one in 400,000. While the methodology used by Dr. Griffiths is subject to debate, dermatologists worldwide have accepted his estimates.

OCCURRENCE BY LOCATION —Based on population estimates for 2023, the following patient populations reflect PRP patient population estimates. PRP Global Database reports 0,000 PRP profiles, of which 0,000 are in the US and 0,000 are international.

OCCURRENCE BY AGE — Based on 0,000 patient profiles reporting onset age, there are 000 patients 18 years and older and 000 less than 18 years of age.

OCCURRENCE BY TYPE — The following is provides a sharper focus on the broader categories of Adult Onset (54%) versus Juvenile Onset (45%).

☐ Type 1 — Classical Adult Onset PRP

- 50% of all PRP cases
- Odds: One in 800,000

☐ *Type 2 — Atypical Adult Onset PRP*

- 5% of all PRP cases
- Odds: One in 8 million

☐ *Type 3 — Classical Juvenile Onset PRP*

- Odds: One in 4 million
- Percentage of patient population: 10% of all PRP cases

☐ *Type 4 — Circumscribed Juvenile Onset PRP*

- Odds: One in 1.6 million
- Patient population percentage of all cases: 25% of all PRP cases

☐ *Type 5 — Atypical Juvenile*

- Odds: One in 8 million
- 5% of all PRP cases

There is a Type 6, which is “HIV Associated.” Type 6 is simply too rare a diagnosis to provide meaningful statistics.

WE ❤ METRICS

14. Who came up with our name?

Storytelling can be a therapeutic part of a PRP journey. Family and friends want to know what's going on.

☐ When James Shooter was admitted to St. Bartholomew's Hospital in London, England, in 1828, he unwittingly became the world's first patient with what the medical community would eventually call *pityriasis rubra pilaris*. In 1828, however, Mr. Shooter's skin disorder did not yet have a name.

☐ Seven years passed before Claudius Tarral, a French dermatologist, wrote about the case in "Traite theorique et pratique des maladies de la peau" (*Treatise on Skin Diseases*) in 1835. Tarral saw it as a variant of psoriasis.

☐ It would take another 21 years (1856) before Marie Guillaume Alphonse Devergie, a dermatologist and forensic doctor at St. Louis Hospital in Paris, published the most complete description of PRP. Devergie's article was considered the "original description" of PRP.

Dr. Marie Devergie

- While Devergie saw the skin disorder as a combination of skin maladies, it would take 21 more years (1877) before another Frenchman and dermatologist named Richaud recognized PRP as a distinct entity.

- In 1889, 12 years after Richaud, 43 years after Devergie and 54 years after Tarral, Ernest Besnier presented nine cases in a 120-page article. He forever fixed the name of the disease as *pityriasis rubra pilaris* from the Latin: *pityriasis* (scalelike skin), *rubra* (red) and *pilaris* (hair follicles).

Dr. Ernest Besnier

- Besnier, too, was a Frenchman and dermatologist. He was also the medical director of the St. Louis Hospital in Paris — the same hospital as Devergie.

Like many rare disease communities, the PRP community laments the snail's pace at which PRP research progresses. What should we expect—it took 61 years and four dermatologists from France just to get the name right.

Remember: in 1828, James Shooter became the first PRP patient when he was admitted to St. Bartholomew's Hospital in London, England. Dr. Ernest Besnier "coined" the name pityriasis rubra pilaris, and I renamed it the "Red Menace" in a 2019 Christmas poem.

St. Louis Hospital, Paris, France

15. Is asking Dr. Internet for answers about PRP a "Fool's Errand?

The spot that appeared on my forehead in August 2012 was initially misdiagnosed as *seborrheic dermatitis*. It took four months, four biopsies, and six days in a hospital before a second dermatologist heard the words: "You have *pityriasis rubra pillaris."*

Like most PRP patients, there was an implicit referral to Dr. Internet, Dr. Google and sometimes Dr. Yahoo.

- ☐ Eleven years ago, information about PRP written for patients was sparse. There were few websites written expressly for PRP patients and caregivers,
- ☐ Today, more information from the patient perspective can be found, e.g., the 5,000-word NORD PRP Report and the *PRP Survival Guide*.
- ☐ Today, a Google search for pityriasis rubra pilaris will return over 200,000 web pages. The deeper you dive into the rabbit hope, the more you may find frustration rather than answers, e.g., research papers written by dermatologists for dermatologists.
- ☐ It also didn't take long to realize that the same questions were being asked and the same answers were being offered.
- ☐ The National Institutes of Health guidelines allow patients and caregivers to find and evaluate health-related information online. Check out Appendix F: Finding health information on the internet and Appendix G: Evaluating health information on the internet.

16. What should I say to family & friends?

It's your story. You are the storeyteller!

☐ When it comes to PRP, you're the subject matter expert. At the very beginning, even when you are befuddled and bewildered by PRP, you are the subject matter expert in the eyes of family and friends. These aren't passersby and gawkers at Walmart.

☐ Choose the right time and place. Quiet. Private. Unrushed. You initiate and control the conversation.

☐ I had my talking points.I had a story to tell. "PRP stands for pityriasis rubra pilaris. PRP is a journey. Mine started in August when I saw a spot the size of a dime on my forehead. PRP is a sporadic skin disorder effecting one in 400,000 people. There are ONLY and estimated 850 in the USA. PRP is predictably unpredictable. It could last one year or five years or longer. I'm being treated at UT Southwestern and joined a PRP Support Group."

☐ When family and friends want more information, refer then to the 60-Second PRP Roadmap and the *PRP Survival Guide*.

☐ Share the challenges you have already faced and the healing milestones. you have celebrated.

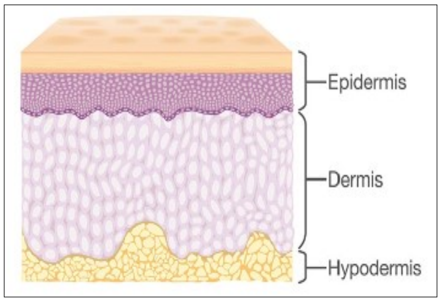
☐ Some PRPers don't want to accept the fact that they have PRP. Once they do, however, family and friends will rally around you.

☐ If you sense that a family member or friend is uncomfortable with the topic, just tell them you just wanted to ensure they know you will be okay and immediately change the conversation.

17. What should we know about skin?

FACTS ABOUT SKIN

Our skin is made up of three layers.



☐ The *epidermis* is the outermost layer that acts as a barrier. A protein in this layer strengthens the skin and makes it waterproof.

☐ The *dermis* is the middle layer comprising sweat glands, hair follicles and connective tissues.

☐ The third and innermost layer is the hypodermis. It helps conserve the body's heat and protect it from injury by acting as a shock absorber.

THE METRICS OF SKIN

☐ Skin is the heaviest organ of the human body. The skin of an average adult weighs around 9.92 lbs (4.5 kgs) and represents about 15% of your body weight.

☐ The average adult has approximately 21 square feet of skin (2 square meters) — the surface area of a twin XL mattress.

☐ Your skin is thickest on your feet (1.4mm) and thinnest on your eyelids (0.2mm).

☐ Your skin typically renews itself every 28 days. PRP patients are more like the snow-making machines used at ski resorts.

☐ The average adult constantly sheds about 30,000 to 40,000 cells *every minute!* I don't want to know how many cells we lose every 60 seconds.

SKIN IS A 24/7 PROTECTION SYSTEM

- ☐ Our skin serves as a protective shield against injury and infection
- ☐ Our skin regulates body temperature.
- ☐ Our skin contains secretions that can kill or prevent bacteria
- ☐ Our skin helps to make vitamin D when exposed to the sun
- ☐ Our skin removes excess waste in the form of sweat

Among its many functions, the skin is an incredible organ that protects the body from external agents.

PAIN & ITCH

- ☐ Skin is a sensory organ. Some of the nerves in your skin are connected to muscles instead of the brain, sending signals through the spinal cord. That’s why we react so quickly to pain and itch.
- ☐ Your skin has at least five different types of receptors that respond to pain and touch. Lucky us.
- ☐ Based on the shared experiences of several thousand PRP patients, the phrase “This too shall pass” is yet another PRP mantra.

You don’t have to be a dermatologist. Still, you can demonstrate your PRP-savviness by knowing more about your skin than your family and friends. Your insights and confidence will establish you as a subject matter expert on your version of PRP.

18. How is PRP pronounced?

Mostly, everyone you know — family, friends, co-workers — will defer to how YOU pronounce pityriasis rubra pilaris, whether your pronunciation is correct. The fact is, even dermatologists don't seem to agree.

☐ pity-RYE-a-sis ROO-bra pill-LAR-is (preferred)

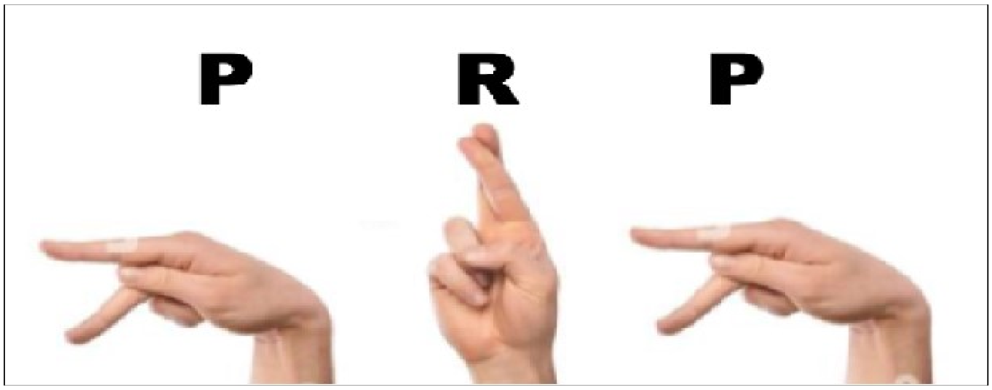
☐ pity-REE-a-sis ROO-bra pill-LAR-is

☐ pity-RYE-a-sis ROO-bra pill-LAIR-is

☐ pity-REE-a-sis ROO-bra pill-LAIR-is

Whatever pronunciation you use, say it with confidence. Say it with authority. Your audience — whether a family member, friend, co-worker, employer, teacher or school administrator — is predisposed to consider you the subject matter expert.

Let's face it — you ARE the subject matter expert of your version of PRP. Moreover, you will become an even more informed subject matter expert daily.



Peer Review Ponderings

> *I was scared when my PRP journey began. Why me? How long will this journey last? I had lost control of my life and was miserable. I knew I had to stay positive and not let the disease control my spirit like it was controlling my body.*
>
> *Staying positive and being hopeful made every day a bit easier.*

— Alan B, Lakeland, Florida, USA

SECTION TWO

DIAGNOSING PRP

19. What are the takeaways from Section One?
20. Quality healthcare requires the timely diagnosis of PRP?
21. What is a PRP diagnostic odyssey?
22. Why is PRP so difficult to diagnose?
23. What if Sherlock Holmes was a dermatologist?

Focus on

What’s Important

19. What are the takeaways from Section One?

In Latin, PRP stands for scaling (*pityriasis*), redness (*rubra*), and hair follicles (*pilaris)*

☐ The proper pronunciation of PRP: pity-RYE-a-sis ROO-bra pill-LAR-is

☐ PRP is predictably unpredictable.

☐ The estimated PRP prevalence rate is one in 400,000.

☐ What are the odds of getting an Adult vs. Juvenile Onset?

☐ A disease or condition is considered “rare” if it affects fewer than 200,000 people. The UK says “less than 50,000”.

☐ There is no cure for PRP.

☐ A “sign” is a medical condition that can be “seen,” while a “symptom" is information that is “shared”.

☐ Communication between the patient and dermatologist is essential.

☐ Healing Milestones are how PRP patients measure progress. One, then another. Some are significant accomplishments, while others are minor.

☐ Our skin comprises three layers: the epidermis, dermis and hypodermis.

In 1828, James Shooter was admitted to St. Bartholomew’s Hospital in London, England, and unwittingly became the world’s first PRP patient. It took four dermatologists from France 61 years just to get the name right. Dr. Ernest Besnier won the PRP name game.

20. Quality healthcare requires the timely diagnosis of PRP?

Communicating an accurate and timely diagnosis to patients is essential to providing high-quality care. Errors in diagnosis are a significant threat to achieving high-quality care.

The Institute of Medicine (IOM) has identified six aims of quality healthcare. The objective is for the dermatologist to avoid an adverse event that results in unintended harm to the patient by an act of commission or omission rather than by the underlying disease or condition of the patient.

☐ safe—avoid injuries to patients from the care that is intended to help them;

☐ effective—provide services based on scientific knowledge to all who could benefit and refrain from providing services to those not likely to benefit;

☐ patient-centered—providing care that is respectful of and responsive to individual preferences, needs, and values and ensuring that patient values guide all clinical decisions;

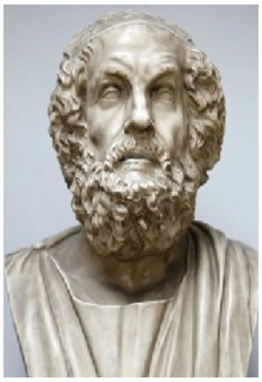
☐ timely—reducing waits and sometimes harmful delays for both those who receive and those who give care;

☐ efficient—avoid waste, including waste of equipment, supplies, ideas, and human resources; and

☐ equitable—providing care that does not vary in quality because of personal characteristics, such as gender, ethnicity, geography, and socio-economic status.”

Communicating accurate and timely diagnoses to patients is essential to providing high-quality care.

21. What is a PRP diagnostic odyssey?



My diagnostic odyssey began on August 12, 2012, with the appearance of a dime-sized spot on my forehead. I saw it in the mirror when I was shaving. It ended 110 days later, on November 28, when my dermatologist confirmed *pityriasis rubra pilaris*.

My first clinic visit was in early September., a month after onset. By then, the dime was now a quarter. My dermatologist thought it was *seborrheic dermatitis*. She prescribed a medication and said to come back in two weeks. Two weeks later, the quarter was now a bar the size of a dollar bill. I was clearly the victim of inflation.

Over the next four weeks, I underwent a series of inconclusive biopsies, an ever-increasing regimen of prednisone, and the mantra: “Come back in two weeks.”

My flame-red skin continued to march down my body like General Sherman to the sea. A few days before the Marine Corps Birthday (November 10), my evening prednisone dosage was increased to 60 mg. The next night, my sleep was interrupted by a parade of brightly colored tropical fish and a yellow rubber duck of biblical proportions.

I woke my wife, told her it was time for the ER, and added, “By the way, we might need a flatbed truck for the duck.“

My six-day visit to the hospital included another inconclusive biopsy. I left without a diagnosis and fired my first dermatologist.

Two weeks later, I arrived for a follow-up appointment with the dermatologist who had been “on call” at the hospital. I was in “full bloom” from head to toe. He instructed the dermatopathologist to *specifically consider* PRP. I must have reached some visual equivalent of PRP Critical Mass.

He did, and I finally got my PRP diagnosis.

22. Why is PRP so difficult to diagnose?

The diagnosis of *pityriasis rubra pilaris is like a mystery to be solved. There are three issues at play here.*

- ☐ PRP is über rare. With a prevalence of one in 400,000, there aren't many opportunities for dermatologists to see PRP beyond a text book in medical school or looking over someone's shoulder.
- ☐ PRP is not easily recognized. PRP mimics other more common skin disorders like dermatitis, eczema and psoriasis.
- ☐ PRP signs and symptoms may not yet be visible.
- ☐ Diagnosing PRP is a process. A dermatologist must physically examine the patient, make clinical observations and document signs and symptoms.
- ☐ Skin disorders mimic one another. A differential diagnosis is a process of eliminating alternative skin disorders to find the" best fit."
- ☐ From a PRP patient's perspective, a differential diagnosis becomes a misdiagnosis when the dermatologist stops weighing the probability of one disease versus that of others. My first dermatologist was convinced seborrheic dermatitis was the culprit. She stopped looking.
- ☐ A biopsy is essential. If PRP is a possible diagnosis, the dermatologist must order a skin biopsy of the affected area with specific instructions to the dermatopathologist to consider PRP.
- ☐ Clinical correlation. When the findings of the biopsy are "compatible with" or "support" the clinical observations of the dermatologist, a PRP diagnosis can be rendered.

23. What if Sherlock Holmes was a dermatologist?

Even Sherlock Holmes needed clues with which to solve a mystery. As a skilled dermatologist, Dr. Holmes starts his investigation by searching for signs and symptoms. When the atient is first seen, there is not enough evidence to confidently identify a suspect — let alone make an arrest.

In fact, PRP is not even in the lineup of probable suspects, e.g., psoriasis, eczema, etc.

As time passes, however, more is learned, and more evidence is collected. The patient is brought back for more questioning. Still, the red and itchy skin mimics the signs of psoriasis, eczema, etc.

Dr Holmes is undaunted. With dogged persistence and deductive reasoning, he revisits the patient for a third interview. He notices a distinct pattern of unaffected skin. He makes a mental note: "Islands of sparing." He orders a biopsy and instructs the dermatopathologist to "consider *pityriasis rubra pilaris*."

It takes a few days to get the results, which confirm what Dr. Holmes had suspected: pityriasis rubra pilaris. To his credit, Dr. Holmes knew that "Islands of sparing" was a signature characteristic of PRP. Of course, he referred to it as "intervening areas of unaffected skin." To each his own.

- ☐ For those of us on the journey from onset through remission, a more timely diagnosis of PRP has no impact on us. However, at this very moment, a person with a dime-size blemish on their forehead doesn't know they have PRP.
- ☐ What can we do as the PRP global community to shorten the time that yet-to-be-diagnosed PRP patients must suffer due to the lack of a proper and timely diagnosis? I submit that it falls to those already embarked on their PRP journey. If not us, then who will make it happen?

Peer Review Ponderings

> *Hope can be elusive. One must chase after it sometimes. But when you grasp it, hold onto it as much as possible each day. Hope is a sustaining attitude that complements, and even improves, our medical treatments. The sharing of information has made my own hope even more robust.*

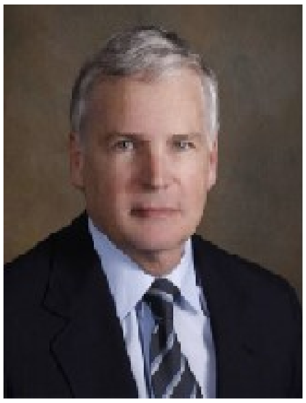
— Richard L, Lansdale, Pennsylvania, USA

SECTION 3

TREATING PRP

24. What if my Dr. Golden was Sherlock Holmes?
25. What is the PRP treatment mantra?
26. What is patient-centered care?
27. Is there a standard PRP treatment protocol?
28. What are the PRP therapies?
29. What are the possile healthcare referrals?
30. How do I find a PRP-savvy dermatologist?
31. What are the impacts of PRP on rare caregivers?
32. Why is measuring coverage important?
33. Why is Self-Advocacy Critical to PRP Patients?
34. What are the strategies for PRP Patient self-advocacy?

24. What if my Dr. Golden was Sherlock Holmes?



I was taken to the ER at the Plano Medical Center at 4:00 a.m. on Thursday, November 9, 2012. My second 60 mg dose of prednisone had launched hallucinations of a 20-foot rubber duck and a bedroom filled with tropical fish.

Later that day Dr. Michael Golden, the on-call dermatologist for the hospital, came to my room, conducted a visual examination, asked a dozen questions, and ordered a biopsy. I had only one question: "What was going on with my skin?"

He rattled off five contenders. It was like listening to the muffled voices of adults in a Charlie Brown cartoon.n*Waaah, Waaah, Waaah, Waaha, or PRP*

When the biopsy results were inconclusive, he scheduled a followup visit in his office after I was discharged.

When Dr. Golden saw me a week later, he performed another visual examination. I asked him again, "What was happening with my skin?" He offered the same list, but PRP had become the Number One contender. He performed another biopsy, which included instructions for the dermatopathologist to consider PRP. Part bloodhound, he had seen something that clicked.

A few days later, my phone rang, and Dr. Golden's office confirmed my diagnosis: *pityriasis rubra pilaris*. They also set an appointment for after Thanksgiving to discuss treatment. .

Luck plays a vital role in my PRP journey. In his last year of training at UTSW, Dr. Golden served as the chief resident in dermatology. His clinic office is also less than 500 feet from my house.

Errors in diagnosis are a significant threat to achieving high-quality care.

25. What is the PRP treatment mantra?

The PRP Alliance has conducted numerous surveys and tallied the comments of hundreds of PRP patients regarding the efficacy of PRP therapies. There are over 17 systemic therapies — a list that keeps growing.

There has been limited research on the efficacy of specific therapies. RCH REGARDING SPECIFIC One of the many frustrating aspects associated with *pityriasis rubra pilaris* is reflected in the following mantra:

"*What works for one doesn't work for all.*"

Over the past few years, however, I have taken the opportunity to amend the treatment mantra to reflect a more positive reality:

> "*What works for one doesn't work for all,*
> *But if it works for ONE, there is hope for YOU.*"

Management of PRP often involves both systemic and topical therapies combined. The efficacy of specific therapies varies from patient to patient. Most PRP patients need systemic therapy to control the condition.

☐ Systemic therapies have potential side effects for the entire body, including a risk of infections. Certain medications may cause nausea or skin reactions at the site of injection.

☐ Topical therapies are associated with side effects in the specific treated areas. Side effects common among PRP patients include thinning of the skin. Topical treatments are usually combined with systemic therapy for PRP that affects a large part of the body.

26. What is patient-centered care?

The driving force behind all patient-centered healthcare decisions is the specific health needs and desired health outcomes.

- ☐ Patient-centered care's primary goal is to improve individual health outcomes.
- ☐ Patients are partners with their healthcare providers.
- ☐ Providers treat patients not only from a clinical perspective but also from an emotional, mental, social and financial perspective.
- ☐ Patient-centered care encourages collaboration and shared decision-making between patients, families, and providers to design and manage a customized and comprehensive care plan.
- ☐ Care is collaborative, coordinated, and accessible. The proper care is provided at the right time and place.
- ☐ Care focuses on physical comfort as well as emotional well-being.
- ☐ Patient and family preferences, values, cultural traditions, and socioeconomic conditions are respected.
- ☐ Patients and their families are an expected part of the care team and play a role in decisions at the patient and system level.
- ☐ The presence of family members in the care setting is encouraged and facilitated.
- ☐ Information is entirely and promptly shared so patients and their family members can make informed decisions.
- ☐ A patient judges a provider based on the accessibility of care, the technical and human components of care delivery, and payment management.

27. Is there a standard PRP treatment protocol?

The simple answer is Yes. It’s called trial and error. From the patient's perspective, there are two primary objectives in the treatment of pityriasis rubra pilaris: (1) relieving symptoms as they present and (2) achieving long-term remission, if possible.

☐ Treatment of PRP is mainly anecdotal and based on case reports, a feature shared by many rare disorders in dermatology.

☐ PRP tends to follow a natural waxing and waning course, with episodes in which there is periodic worsening or cessation of symptoms. Think roller coaster. According to PRP researchers, evaluating the effectiveness of particular therapies is difficult.

☐ The fleeting nature of the large proportion of PRP symptoms also makes it challenging to study in standardized, long-term therapeutic studies. The value of a specific treatment is challenging to assess, as the clinical course is variable for each type of PRP.

☐ As controlled trials are lacking, the effectiveness and safety of treatments is unclear. Thus, there is low-quality evidence supporting the treatment strategies of PRP.

☐ For example, patients with classical adult onset PRP may present with intense and widespread reddening of the entire skin surface (erythroderma). Hospital admission for skin care, fluid replacement and other supportive care may be warranted.

Neither the European Medicines Agency (EMA) nor the US Food and Drug Administration (FDS)have approved any treatments for PRP.

28. What are the PRP therapies?

When PRP therapies work, it seems like magic. Management of PRP often involves systemic and topical therapies combined. Topical therapies can help with the symptoms and may be enough for people with mild PRP. Topical treatments are usually combined with systemic therapy for PRP that affects a large part of the body. Most PRP patients need systemic therapy to control the condition.

RETINOIDS

☐ Oral retinoids are derivatives of vitamin A that slow the growth and shedding of skin cells. Treatment options include

☐ acitretin / Soriatane®

☐ isotretinoin / Accutane®

☐ Oral retinoids (synthetic vitamin A derivatives) are the first-line systemic treatment for PRP. A scientific survey of patients with PRP performed by researchers at Thomas Jefferson University published that oral retinoids were helpful in approximately 60% of patients with PRP.36

IMMUNOSUPPRESSANTS

☐ Immunosuppressants slow down the body's immune system. These can be used in combination when oral retinoids are ineffective.

☐ Treatment options (oral and injection) include methotrexate, cyclosporine, TNF-alpha inhibitors, and IL-12/23 inhibitors.

☐ Methotrexate was reported to be helpful in approximately 50% of patients with PRP.

BIOLOGICALS

Biologicals are injectable or intravenous (IV) medications that target various pathways of inflammation in the body. Biologicals are also immunosuppressants.

With generally fewer side effects, biologicals are targeted to reduce inflammation. The following is a list of biologicals prescribed to or considered by PRP patients. The list is alphabetized by generic name and includes the FIRST APPROVAL DATES by the Federal Drug Administration (FDA)…but NOT for PRP.There are currently 14 treatment options considered for PRP patients by dermatologists. Again — NONE are approved by EMA or FDA.

☐ adalimumab/Humira®

☐ brodalumab/Silliq®

☐ certolizumab/Cimzia®

☐ dupilmab/Dupixent®

☐ etanercept/Enbrel®

☐ golimumab/Simponi®

☐ guselkumab/Tremfya®

☐ infliximab/Remicade®

☐ ixekizumab/Taltz®

☐ risankizumab/Skyrizi®

☐ secukinumab/Cosentyx®

☐ tildrakizumab/Ilamya®

☐ ustekinumab/Stelara®

☐ upadacitinib/Rinvoq®

OTHER PRP THERAPIES

- Topical creams that contain urea or ammonium lactate decrease the scaling and flaking of the skin. Topical corticosteroid creams reduce skin inflammation. These are applied directly to the skin.
- Oral vitamin A. This may be helpful in some people, but only in very high doses that may cause toxicity. Retinoids (synthetic vitamin A derivatives) are safer, more effective, and more commonly used than high-dose vitamin A.
- Traditional Chinese medicine and other alternative medicines have varying degrees of success.

29. What are the possible healthcare referrals?

Depending on the severity, duration and array of signs and symptoms, PRP patients seek the expertise of specialized healthcare professionals:

- ☐ Ophthalmologist: ectropion (eyelids are turned outward) and impaired vision
- ☐ Podiatrist: impaired mobility
- ☐ Otorhinolaryngologist (ENT specialist): impaired hearing, removal of ear wax (cerumen) from the ear canal.
- ☐ Hepatologist: monitor the impact of PRP treatment on the liver.
- ☐ Psychiatrists/Clinical Psychologist: depression and mental wellness

In my humble opinion, every PRP patient should discuss the possible situations when a referral to a specialist by either a dermatologist or a general practitioner. During my PRP journey, I needed vision, morbidity, and hearing impairments. I dealt with my endocrinologist regarding my type 2 diabetes. In retrospect, I should have sought a referral for depression and mental wellness.

30. How do I find a PRP-savvy dermatologist?

While that estimate may be totally bogus, we can say without fear of contradiction that very few dermatologists will consider themselves PRP savvy.

There are at least four reasons to seek out a PRP-savvy dermatologist:

☐ To obtain a second opinion, ask, "Do I really have PRP?" There's no law against finding a dermatologist that

☐ Track down a local dermatologist with prior experience treating PRP. Call and confirm that someone has experience with PRP. There is no guarantee of PRP savviness, but it is a start.

☐ To find a more interested dermatologist who wants to become PRP-savvy. If your dermatologist isn't committed to learning more about PRP, it's time to find one who does.

☐ To find a dermatologist more supportive of your needs as a patient *with PRP* than your current dermatologist. In 1742, English poet Thomas Gray coined the phrase "ignorance is bliss": "In knowing nothing, life is most delightful." Gray didn't have PRP. Ignorance is NOT bliss. Beffudlement is unacceptable.

While the best case scenario is a referral from a fellow PRP patient or caregiver to their PRP-savvy dermatologist, the next best step in the USA is Find-A-Derm.

The American Academy of Dermatology (AAD) maintains a database of individual dermatologists. It is specifically designed to provide easy access to information about dermatologists who are members of the AAD. While there is no guarantee that Find-a-Derm (find-a-derm.aad.org) will find a PRP-savvy dermatologist, it is a reasoned approach.

SEARCH USING VARIABLES

- ☐ Search by location: Enter your city and state. I could have entered either my ZIP CODE (75093) or my city and state (Plano, Texas)
- ☐ Search by conditions: You won't find PRP. I chose "psoriasis," the most common mimicker of PRP on the list of 30 options, rather than "rare skin conditions."
- ☐ Search by procedure: Selecting one of the 23 will limit the results. Search by "ANY Procedure."
- ☐ Search by practice focus: Use "Medical dermatology". If you have a PRP child, select "Pediatric Dermatology."

FIND-A-DERM DELIVERS

- ☐ A list of qualifying dermatologists includes name, address, city, state, and a Google map displaying all locations.

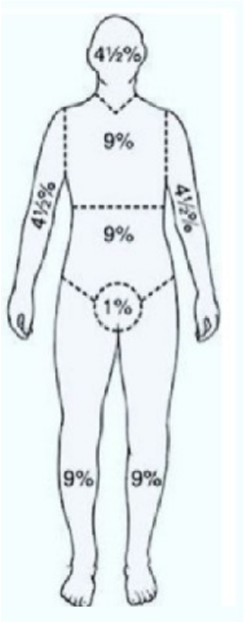
FOLLOW LINKS TO EACH DERMATOLOGIST

- ☐ Find-a-Derm provides additional information for each dermatologist: Website, Location, Office information and Qualifications, e.g., medical school, residency training, hospital affiliations, and certifications.

I have used Find-a-Derm in the past to help other PRP patients but never tested it on myself. A search for a PRP-savvy dermatologist listed Dr. Michael Golden as #4 in Dallas and #1 in Plano.

31. Why is measuring coverage important?

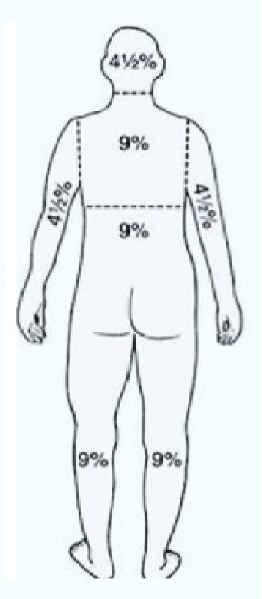
My PRP journey began with a red spot on my forehead. It quickly spread to my face and neck. As the weeks passed, my upper torso became engulfed. Then, my feet. Then, my lower legs. After four weeks, I "guesstimated" my coverage and called it "full bloom."



The Rule of Nines is a technique used to evaluate burn victims. The results help guide treatment decisions, including fluid resuscitation and become part of the guidelines to determine transfer to a burn unit.

Emergency medical responders are some medical workers who use the rule of nines most.

They quickly estimate the burn area to decide on treatments for the hospital.



☐ The front and back of the head and neck equal 9% of the body's surface area.

☐ The front and back of each arm and hand equal 9% of the body's surface area.

☐ The chest equals 9%, and the stomach equals 9% of the body's surface area.

☐ The upper back equals 9%, and the lower back equals 9% of the body's surface area.

☐ Genitals equal 1%

While I might say "I'm in full bloom" to family and friends, I should have used the Rule of Nines when telling my dermatologist.

Peer Review Ponderings

32. What are the impacts of PRP on caregivers?

In writing the 60-Second PRP Roadmap, I read a study commissioned by the National Alliance for Caregiving and Global Genes.

Rare Disease Caregiving in America is an 85-page study of 1,406 caregivers living in the United States who care for a child or adult with a rare disease or condition. Their goal was to paint a picture of occasional caregivers facing emotional, financial, physical, and social strain with little support from outside resources. PRP was omitted. We fall under the über-rare category.

- ☐ Over 80% of rare caregivers rely on medical professionals for information yet report having difficulty in accessing treatments or therapies for symptom management and the underlying disease.
- ☐ Rare caregiving can ultimately impact the finances of the rare caregiver and their family unit, and these financial impacts loom large.
- ☐ The day-to-day impact for rare caregivers is pervasive in all aspects of their lives, with no areas untouched: the effects are seen in the workplace, school, and on their physical and emotional health.

The best message to send to PRP caregivers is that we know your challenges and your critical role in every PRP journey.

A more detailed overview of “Findings” is available in Appendix D: Rare Disease Caregiving in America

It's Up To You!

33. Why is Self-Advocacy Critical to PRP Patients?

A PRP self-advocate makes informed decisions and then takes responsibility to make those decisions a reality.

Because PRP is so rare, there are few "experts" in treating PRP. Each PRP patient must seek PRP-specific resources like the PRP Support Group and the *PRP Survival Guide*. Self-advocacy can help patients and their families be empowered to fully participate in their care. Self-advocacy is an absolute necessity for PRP patients.

Self-Advocacy Can Help You Get High-Quality Health Care

☐ High-quality health care means getting the proper care at the right time, delivered by the right health care professional. As PRP patients, we have a critical role in managing our health. Patients play an important role in determining the need for care and the outcomes of that care.

Self-Advocacy Can Help You Feel in Control

☐ At times, you may feel like it is running your life. In many ways, it is. Standing up for your health can give you back a sense of control. Standing up for your health gives you the information you need to make informed decisions about your medical treatment.

Self-Advocacy Can Help You Get Support

☐ When standing up for your health, you ask for help from family, friends, and others. Their support can help you cope with PRP.

STANDING UP
for your health
SELF-ADVOCACY FOR PATIENTS WITH RARE DISEASES

34. What are the strategies for PRP Patient self-advocacy?

On June 3, 2015, the PRP Alliance participated in a conference call arranged by the National Organization of Rare Disorders (NORD). John Huber, executive director of the plastic Anemia & MDS International Foundation, announced the completion of a 62-page self-advocacy brochure for patients with rare diseases.

I did not expect rank-and-file PRP patients and caregivers to go cover to cover and adopt all of their suggestions. I did recognize, however, that “There’s gold in them dar’ hills.”

For a symbolic licensing fee of $1, the PRP Alliance has been authorized to use the AAMIF brochure without restriction.

I posted a PDF in the *PRP Survival Guide* eight years ago to support STANDING UP for your health. My effort here is to help put their self-advocacy plan on the map.

Whether you are newly diagnosed or have been living with PRP for many years, there are practical strategies designed to help you stand up for your health.

☐ Build a robust PRP Healthcare Team

☐ Learn About PRP and Treatment Options

☐ Make the Most of Every Dermatology Clinic Visit

☐ Retain your PRP Health Information

☐ Build a Strong Personal Support Team

Principle 1: Build a Strong PRP Healthcare Team

Once you have a PRP diagnosis based on the clinical observations of a dermatologist supported by a biopsy.

Find a PRP-savvy dermatologist.

☐ Unfortunately, they are few and far between. Fortunately, mine was an associate professor at the University of Texas Southwestern (Dallas). He taught me what he knew, and I reciprocated by sharing my PRP insights from the patient's perspective.

Identify other healthcare professionals you may need.

☐ In addition to my dermatologist, the following specialists were my PRP healthcare team:

- Endocrinologist (type 2 diabetes),
- Otorhinolaryngologist (impaired hearing),
- Podiatrist (poor mobility)
- Vascular surgeon (two blood clots).
- Family doctor (performed two annual checkups during my PRP journey).

Select a PRP Care Coordinator.

☐ That’s you, your spouse or a partner. Since I was retired and my wife was the primary bread winner, I became my own PRP Care Coordinator)

Do Your Part to Become an Empowered PRP Patient. It won’t happen overnight, but to the victor belongs the spoils.

Principle 2: Learn About PRP and Treatment Options

When the time comes to make a treatment decision, use reliable information—along with input from your dermatologist.

☐ Get up-to-date information about PRP. You can't get more up-to-date information than the insights and experiences of Facebook-based PRP Support Group members.

☐ Get trustworthy information about PRP. *Caveat emptor*… Buyer beware! The National Institutes of Health provides appropriate guidelines for seeking and evaluating medical information online. When you read something on the web that doesn't agree with what your doctor has told you, ask them about it. See Appendix H.

☐ Learn about PRP treatment options under consider. Weigh the pros and cons of each treatment, side effects, and costs. Work with healthcare providers to develop a treatment plan YOU want and feel you can do. Remember, you have the right to choose or refuse treatment.

☐ Keep an eye on your own care. Don't take anything for granted. Ask your dermatologist, "Why?" You aren't challenging your dermatologist when asking why a particular treatment option is favored.

My treatment plan followed the University of Texas Southwestern's PRP protocol, e.g., acitretin (25-50mg). Also, Desonide® (face and ears), Clobetasol®, urea lotion (palms and soles), and triamcinolone in the remaining nooks and crannies. My treatment plan was always subject to change as medications, therapies, or my version of PRP evolved.

Principle 3: Make the Most of Every Dermatology Clinic Visit

I learned early on that my PRP clinic visits at the University of Texas Southwestern would include 10 minutes of intake by a nurse and 15-20 minutes with my dermatologist, Dr. Arturo Dominquez. He would conduct a visual inspection, ask me follow-up questions, and follow up on the follow-ups. But then it was my turn. The proverbial ball was in my court.

☐ I was prepared for each PRP clinic visit. You can manage your dermatologist's expectations if you take the initiative. Exploit every minute of your "derm time."

Dr. Arturo Dominguez

☐ I was prepared to ask five questions. What do you want to know? What do you want your dermatologist to know. Even a PRP-savvy dermatologist appreciates learning something new about PRP.

The benefits of a PRP clinic advocacy strategy accrue over time.

☐ After about five PRP clinic visits, Dr. Dominguez knew what to expect. When he finished "his part," he willingly gave me the floor.

☐ Look for ways to share your PRP insights with the nurse during pre-clinic intake. Don't underestimate the value of a dermatology nurse.

☐ During one PRP clinic visit, Dr. Dominguez asked me to discuss PRP with a visiting dermatologist from Peru.

☐ In 2015, over seven months after my remission, Dr. Dominguez helped coordinate an invitation to speak to 20-plus students and clinicians. For 30 minutes, I had a captive audience.

Take advantage of every opportunity to share your PRP journey with your dermatologist. PRP patients can help a dermatologist to become PRP-savvy.

Principle 4: Retain your PRP Health Information

Keeping all your medical information about PRP organized and in one place makes it easier to track changes in your health and share that information with your dermatologist. Collect and retain information about the PRP Journey.

☐ Start as soon as you can. Better late than never.

☐ Ask your dermatologist for a copy of your medical records during every clinic visit. And do so during every future visit.

☐ Office visit information, e.g., dates of office visits notes on what your dermatologist tells you.

Take notes

☐ Keep a log of changes in how you feel. Some of these changes may be symptoms of your disease. Others may be side effects of treatment. This is critical information that only you can provide.

☐ Keep a running list of questions to ask your dermatologist. And record your dermatologist's answers. Include what they say over the phone, via email, and during office visits.

Find an organizational system that works for you.

☐ What's the best way to organize the information you gather in one place. Reach out to other PRP patients to learn what they do to keep their PRP information organized. Give me a three-ring binder and a three-hole punch, and I'm off to the races!

Principle 5: Build a Strong Personal Support Team

Having a chronic and rare disease can be very difficult. Knowing someone in your corner can help you overcome the ups and downs.

Get Support From Family and Friends.

☐ Some of us have family members or close friends who live nearby and are willing and able to serve as part of our support team.

Get support from the PRP Support Group.

☐ For many of us, asking for help is not easy. Yet, empowered patients know that help from others can help them get the healthcare services and treatment they need. And some of us find joining online groups or forums offers the support we need.

Get support from other PRP Patients.

☐ STANDING UP got this one right. I joined an email-based support group within 24 hours of my PRP diagnosis. A year later, I joined the Facebook-based PRP Support Group, and, as they say, the rest is history.

☐ People living with PRP know more about what you're going through than anyone else. They can let you know you're not alone. Here are some ways to connect with them:

Get support from your dermatologist.

☐ You may be able to get emotional support from a member of your healthcare team. This healthcare provider may be a counselor or social worker. Or it may be a nurse or doctor who goes above and beyond their professional duties. Referral to a specialist.

Peer Review Ponderings

Peer Review Ponderings

> “*Hopeless. Powerless. How would I ever get through this? Nothing to stop the relentless march of PRP. Can’t look in the mirror. With help came hope. With knowledge came power. With community came “We”. Found ways to live with it. Got through it. Got over it. Did not define who I am.*”

— Jan T, Ringwood, New Jersey, USA

SECTION 4

PRP IMPAIRMENTS

35. What is a PRP impairment?
36. What are impairments to the skin?
37. What are impairments to energy?
38. How important is mobility?
39. What are impairments to dexterity?
40. What are impairments to vision?
41. What are impairments to hearing?
42. What are impairments to thermoregulation?
43. What are impairments to sleep?
44. What are impairments to employment?
45. What are impairments to attending school?
46. What are impairments to socialization?
47. What are impairments to financial health?
48. What are impairments to mental wellness?

35. What is a PRP impairment?

As traditionally used, a *medically determinable impairment* refers to a problem with a structure or organ of the body. Skin is our body's largest organ. The World Health Organization (WHO) provided the following definitions in their *International Classification of Impairment, Disability, and Handicap (1980)*:

☐ ***Impairment*** – any loss or abnormality of psychological, physiological or anatomical structure or function.

☐ ***Disability*** – any restriction or lack (resulting from an impairment) of ability to perform an activity in the manner or within the range considered normal for a human being.

☐ ***Handicap*** – the result when an individual with an impairment cannot fulfill a normal life role. Handicap is a disadvantage for a given individual that limits or prevents fulfilling a normal function.

From the perspective of a PRP patient, the technical definition of a PRP impairment is a potentially major problem that impacts lots of big stuff for an indeterminant period. It generally excludes potentially significant problems with topics like baths & showers, flares & setbacks., hair, exercise, handicap parking, clothing, housekeeping, lip care, scalp, weight loss/gain, tattoos, etc.

The focus in Section 4 is 14 impairments that often define the insurmountable mountains PRPers must climb and the indignities we must suffer en route.

36. What are impairments to the skin?

The pain we experience with our PRP skin is time-sensitive. Whenever we move, it's time to feel pain. So, PRPers don't move very much.

While PRP is predictably unpredictable, you can expect itching that is unrelenting flaky skin that ranges from fairy dust that is equally unrelenting to large patches on the palms of hands and the soles of feet.

☐ A PRPer once wrote: "I am not kidding when I say that my skin flakes do not obey the laws of standard physics. They are quantum particles that randomly wink in and out of existence, especially wherever I've just swept or vacuumed. They also have unusual electrostatic effects. If I wipe down the stairs, they creep up the wall and cling to the sides. You may find yourself (or a family member) permanently attached to the vacuum cleaner!

☐ Some PRPers literally leave a trail of dead skin flakes wherever they go. This is not the time to wear dark clothing!

☐ Peeling can be an embarrassment, but remember that the peeling is helping you shed those plaques that makeup PRP, and it is helping you grow new, hopefully clear, skin.

☐ You can do a lot to improve dry skin, including moisturizing.

37. What are impairments to energy?

My “get up and go, got up an went” in October 2012, a few months after the onset of symptoms.

I remember being consumed by fatigue and probably more than my fair share of depression. It was a very lonely time — days filled with nothing. Then, I made an astonishing re-discovery. I re-discovered the “on switch” on the back of my iMac. My computer was my ticket out of the despair of fatigue. It was April 2013. I could sit on my butt at my computer for hours. So I did.

I was a member of an email-based support group with an archive of emails from 1997. I began reading those emails from fellow PRP patients and their caregivers. It took me several months, but I reviewed over 29,000 emails and created over 1,500 PRP patient profiles, including name, onset age and dates, biopsy history, etc.

My PRP-related fatigue was the reason I sought refuge in my computer, and my research turned out to be the therapy for what could have become debilitating depression.

Fatigue and loss of energy is a common symptom of PRP. Your “get up and go” will return.

38. How important is mobility?

The lack of mobility was THE vchallenge during the first six months of my PRP journey.

- ☐ Whether my feet were encased in thick skill, cracked and bleeding, or paper-thin — the pain was debilitating.
- ☐ It was bad enough to be bedridden for a while. Then, one day, I ventured out of the carpeted bedroom and shuffled across dark hardwood floors, leaving white flakes and dust in my wake.
- ☐ Weeks passed before I ventured out of the front door and felt the cold winter air.
- ☐ The first time I was driven to Walmart, I needed one of those scooters. I resolved that the next time, I would ignore the scooter.
- ☐ I did use a handicapped sticker for nearly a year. It was beneficial to get close to the entrance of Walmart or any store, for that matter. I kept looking at the date on the sticker and wondered if I should get it renewed in October 2013 — 14 months after onset.
- ☐ When the time came, I added the retirement of my handicapped sticker to my expanding list of Healing Milestones. If the sticker helps, get one. When you don't need it, get rid of it and do the Happy Dance.

Today, I walk just fine in my Sketchers but hobble barefoot on hardwood floors.

39. What are impairments to dexterity?

There were three manifestations of impaired dexterity during my PRP journey that are common to most PRPers.

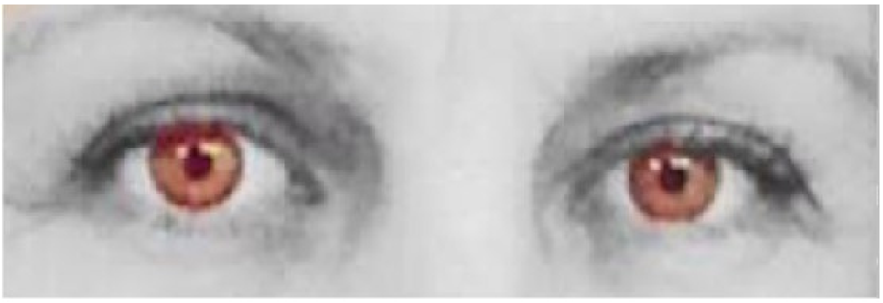
Opening a jar of Vlasic sweet relish.

☐ One day, I tried to open a jar of Vlasic sweet relish that was particularly stubborn. When my grip failed, I used a contraption to remove it. Not too long after that, I couldn't even squeeze the contraption. A growing list of indignities had been amassing in the relish jar, e.g., picking up a credit card on the floor with no fingernails.

☐ Fast forward many months, and I stood over a relish jar on the counter, daring me to open it without a mechanical aid. I focused on the lid. Grasped the lid. Turned the lid. Phoooosh! The sound of victory. I just chalked it up as a significant healing milestone.

Picking up a credit card from a hard surface.

☐ Milestone #5 (date unknown). I picked up a credit card that had fallen on the floor at Walmart. Having lost my nails many months earlier, a dropped credit card meant that some poor, unsuspecting cashier would be forced to learn about pityriasis rubra pilaris as they retrieved the fallen plastic. Dropping things around the house. As my PRP progressed, the skin on my hands thinned, my fingernails dissolved, and my fingertips became sensitive. Have you ever boiled cranberries to make cranberry sauce? The cranberries pop open, and the mini-explosion creates a scar in the shape of an "X." For months, I had 10 fingertips with as many paper cuts.



40. What are impairments to vision?

We need our eyes to read, do our jobs, drive, see our loved ones ’faces, and enjoy the beauty of the world around us…there are countless reasons to guard them from harm. Dry eyes, ectropion, and even the irritation caused by constant exposure to skin flakes leave our eyes vulnerable to damage and severe infections, especially of the cornea.

Patients diagnosed with *PRP* can experience one or more of the following during their journey.

☐ Dry, gritty, irritated eyes with an excessive amount of crusty gunk. We don’t have enough tears, but our eyes get watery to compensate for the dryness, with ineffective non-wetting tears. We have incomplete coverage of tears.

☐ Teary, blurry eyes.

☐ Tight skin on the face that pulls on our eyelids

☐ Flaking skin gets in the eyes, made worse by losing our protective eyelashes and eyebrows. Thankfully, it is usually temporary. However, it should be monitored to prevent severe damage from extreme dryness.

☐ Redness and sensitivity to light.

☐ Symptoms usually improve with treatment and go away with remission.

☐ Thickened and drooping lower eyelids make it hard to look looking into a mirror. That’s life with PRP for many of us. Fortunately, these interrelated

41. What are impairments to hearing?

Sometimes, a debilitating disease like PRP brings out the STUPID in us.

Over nearly three months, my hearing deteriorated. It was such a slow process that I didn't know it was happening. My family was talking louder, so it sounded normal to me. I could hear just OK when I turned up the volume. I didn't hear a disgruntled spouse yelling, "Hey, Bill, check your ears!"

Over the past decade, I have resisted using *otorhinolaryngologists*. I actually had to go to YouTube to listen to someone say it.

OH-toe, RYE-no, lair-in-OL-o-gist

My newly discovered otolaryngologist removed two plugs the size of jelly beans — one out of each ear. The medical term is *cerumen* (pronounced ci-RU-min). It's the brown, orange, red or yellowish waxy substance secreted in the ear to protect the skin of the human ear canal. Flaky skin mixes with the ceruen to create a World Class jelly bean.

In addition to jelly beans, Dr. Hsu also discovered inflammation in both ears. I returned every 30 days for the next nine months, then quarterly for a year. The inflammation finally disappeared.Here are the real takeaways: If I had seen the ENT sooner, I might have avoided a year of inflammation. When speaking with any healthcare professional, say cerumen and not jelly bean.

42. What are impairments to thermoregulation?

Some PRPers realize they aren't sweating until it returns. I went months and months without sweating, including during the summer of 2013 in North Texas (aka Tongues of Fire). Then, one day, my right knee got wet. Very weird. Within a week or two, I was gushing on all cylinders! Like many kindred spirits, I considered the event a major healing milestone and did my Happy Dance.

The return of sweating was also when I added thermoregulation to my PRP vocabulary.

☐ "Stinky armpits," announced with zeal and unbridled enthusiasm.

☐ Perspiration (sweat) keeps the body's temperature regulated. It's how your body cools itself. Not perspiring can make you less heat tolerant.

☐ While the cause is not certain, it is probably tied to the fact that we are already losing much of our body's moisture because of the shedding of our skin.

☐ The amount of skin and "insulation" you lose can make you less tolerant to cold.

☐ With the loss of sweating, PRPers must use appropriate caution when the air temperature is hot.

☐ We are constantly sweating, even though we may not notice it. Sweating is your body's major way of getting rid of excess body heat produced by metabolism or working muscles.

43. What are impairments to sleep?

PRP is such a fickled disease. There are PRP patients who would love to sleep 8 to 10 hours at night but can only manage 4. Here is what worked for me.

PRIOR TO PRP DIAGNOSIS

☐ My first dermatologist was convinced I had *seborrheic dermatitis*. After slinging my body with triamcinolone ointment, I was told to sleep in damp, long johns. There were nights I never fell asleep.

☐ The only benefit of that experience with ointment was to tell my third dermatologist I wanted triamcinolone cream. He told me that the ointment was more effective than the cream. I told him, “I don’t give a rat’s a##. Prescribe cream. NOTE: That was the first time and only time I ever needed to “pull rank” on my dermatologist. I learned that a patient does not have to be a lemming. Articulate a reason and defend your position.

AFTER DIAGNOSIS

☐ It took me a few months to find the proper nighttime regime to guarantee a good night’s sleep from 11 PM to 7 AM.

It began with a relaxing evening shower, followed by a slathering of topical creams, e.g., Desonide© (face and ears), Clobetasol© and urea (palms and soles), and triamcinolone in all the remaining nooks and crannies. My night cap was a nightly cocktail of hydroxyzine, Ambien® and Extra Strength Tylenol® —my recipe with my dermatologist’s approval.

☐ But a fourth rule could not be broken: NO liquids after 9 PM. If you quench your thirst at 11 PM, your bladder will set the alarm in your brain for 3 AM.

44. What are impairments to employment?

While I faced the typical challenges all PRP patients face, one major exception was the workplace. I was retired when the "Red Bastard" knocked on my door.

☐ For most PRP patients, the onset comes when they are employed. The decision to work or not to work is the patient's choice, subject to the realities of their workplace, co-workers and employer. For some, working from home is an option.

☐ To work or not to work — that's the question most PRP patients have to ask. For some, the onset of PRP is so ferocious and relentless that working is simply not a viable option. Others, for whatever reason, usually financially, commit to stay on the job for as long as they can or at least try. The loss of income can have catastrophic results. When the planets are in alignment, however, anything is possible.

☐ Bab P, a graphic designer in a Kansas City advertising agency, had all the impairments of PRP save one — energy. She was a talented dynamo, respected by her co-workers. The only PRP workplace issue she had to deal with was shedding skin.

☐ By her own admission, she was a snowblower. However, neither her employer nor co-workers were grossed out by the fairy dust that accumulated hourly in her private office.

☐ When the client arrived to see Bab, her co-workers would be alerted. A few would intercept and delay the client while others would descend on Bab's office with brooms, dustpans and a Dustbuster. The strategy was successful. Bab kept working, and her clients loved the "Royal Treatment" they received whenever they came on-site.

45. What are impairments to attending school?

Regarding PRP kids, the impairments regarding school are less about the patient and more about the school. Classmates, parents of classmates, teachers and school administrators need a “head up” from PRP parents. The following are insights shared by a PRP mother in Zurich, Switzerland and her daughter, Molly.

☐ Inform the teachers at the start of the school year with an information letter about PRP. Make sure they can answer questions raised by classmates.

☐ Inform the other parents.

☐ Ensure that everyone knows the danger of overheating. The teachers are continually thankful for the extra reminder because they have a lot of topics to think about. As a former teacher myself, I totally understand.

☐ Be sensitive to name-calling. A PRP classmate looks like a “Red Tomato” to some children.

☐ Molly knows she can go to the teacher anytime if she is not feeling well, e.g., dizzy, cold or too warm. And the teacher knows she must take Molly’s call for help seriously.

We know that the teenage times probably won’t be that easy. Still, we firmly believe in Molly’s inner mental strength and support her as much as possible to keep this strength high!

The most significant gift we can provide our daughter is to support her on her PRP journey.

46. What are impairments to socialization?

Young or old, rich or poor, socializing is good for mind and body — even when the body is dealing with PRP. We are naturally social animals and tend to function better in a community of kindred spirits. Too often, however, PRP is not conducive to socialization. We hunker down, slathered with ointments and shedding snow drifts of skin. Socialization is not on our front burner.

As PRP progresses, we spend a lot of time alone. There is an increased risk of depression and lower quality of life. You don't have to be super social to see the benefits of connecting with others.

Socializing staves off feelings of loneliness and helps sharpen memory and cognitive skills, increasing our sense of happiness and well-being. In-person is best, but connecting via technology also works.

Before COVID, the best way to specialize with PRPers was face-to-face Meet and Greets. Now, ZOOM and FaceTime make connecting with kindred spirits easily anywhere in the world. The Facebook-based PRP Support Group is ideal for this.

I participated in two Meet & Greets in Central Florida in 2019. One was in Orlando (August 4), and the other was in Tampa (August 8). It was the largest gathering of PRPers in PRP history. That's our story, and we're sticking to it.

47. What are impairments to financial health?

The financial impact of PRP on a patient can range from virtually no impact to catastrophic. We all know from experience that stress is not a friend of PRP. Financial stress is another unavoidable challenge that must be worked through for many PRP patients and their families.

☐ I was 66 and retired, earning a modest monthly stipend from Social Security. My wife, Heather, was (and still is) the breadwinner. Thankfully, her income as a software designer was not affected by my PRP. She also had excellent health insurance through her employer. UnitedHealthcare covered acitretin, hydroxyzine and topicals with a modest copay. I never had the need to deal with costly biologicals.

☐ At the other end of the financial impact spectrum are countless PRPers whose PRP journeys were plagued by income disruptions. The lack of health insurance worsens the daily challenges of body, mind and spirit. Hope was always in short supply. Stress was not.

☐ Then, there were the PRPers in the middle. Just a wee bit of financial stability. Vulnerable, nonetheless, to any unexpected expense. Always a target for stress.

Members of the PRP Support were always sharing strategies for seeking financial help or finding a work-around for expensive biologicals. They say, “Where there’s a will, there’s a way.”

48. What are impairments to mental wellness?

Our emotional health is equally important as our physical well-being, and you cannot separate the two. When you are dealing with any disease, it is challenging to be your natural optimistic self. When dealing with an appearance-altering and physically debilitating illness like PRP it's many times more difficult.

☐ Find your support system…it may already be in place.

☐ Talk with the friends and family members you can trust to best understand your ordeal.

☐ Watch for signs and symptoms of depression and have others watch for them. Also, seek counsel from your doctor about medications if the feelings of depression cannot be shaken off and become a burden to you.

Always remember that you are not alone. There are many of us out here with PRP, and we are all going through the same experiences that you are. Joining the PRP Facebook Support Group and sharing is of utmost importance. Only someone who has walked in your shoes can truly understand your feelings.

☐ Share what you have learned about PRP as a patient or caregiver.

☐ Share what you have been told by your dermatologist?

☐ Share articles you feel might be worth reading or websites worth visiting.

49. PRP Glossary of Words, Terms & Jargon

I made a New Year's Resolution on January 1, 2020. It took me 20 days to complete the *PRP Glossary of Words and Jargon* a reality.

The *PRP Glossary* represents a journey of sorts — my journey with the PRP global community. Along the way, we have cared, shared, and learned about PRP and each other.

PRP Glossary aims to take some of the mystery and fear out of PRP. Not a lot … just a wee bit. Think back to the day you first heard *pityriasis rubra pilaris*. For me, it was November 28, 2012. A voice on the other end of my phone (not my dermatologist) said something about a "supporting biopsy" as she confirmed the time I was to return to my dermatologist the following day — tone week after Thanksgiving. And that was that.

I hung up the phone. Numb. Scared. Confused. Isolated.

Those first moments have not changed over the past decade for PRP patients worldwide. Newbies still feel numb, scared, confused, and isolated. But, we have something today that we didn't have then — an array of PRP resources to support PRP patients, caregivers and healthcare professionals worldwide.

Peer Review Ponderings

> *Hopeless. Powerless. How would I ever get through this? Nothing to stop the relentless march of PRP. Can't look in the mirror. With help came hope. With knowledge came power. With community came "We". Found ways to live with it. Got through it. Got over it. Did not define who I am.*

— Jan T, Ringwood, New Jersey, USA

SECTION 5

PRP Glossary of Words, Terms & Jargon

A Advocacy, Self — Alcohol Consumption — Anatomy of Skin — Autoinflammatory vs. Autoimmune — **B** Balderdash — Befuddled — Biopsies — **C** Caregivers — Cause of PRP — Census, Global — Challenges, PRP — Clinic visits — Closed Group — Cohort — Contagious — COVID-19 — Cream vs. Ointment — Cup o 'skin — Curable — **D** Delete Button — Depression Dermatologist — Dermatopathologist — Dexterity— Diagnostic odyssey — Diet, Anti-inflammatory — Differential diagnosis— Dowling Oration— Duration — **E** Ear stuf — Ebb & Flow — Efficacy — Energy & Fatigue — Exercise and Physical Activity — Eyes & Vision — **F** Face-to-face — Feedback — Find-a-Derm — Fingernails — Flaking & Shedding — Flares — Freebies — Full Bloom — **G** Good Start — Google, MD — **H** Hair Loss — Happy Dance — Healing Milestones — History of our name — Howling at the Moon — Hijacking a post — **I** In My Humble Opinion — Itch — **L** Land of Chat — Land of Hope — Land of Share — Laundry — Lip care — **M** Managing expectations — Metaphor — Mirror — Mobility — **N** Nitrile gloves — NORD PRP Report — **O** Odds — Onset — the first sighting — **P** Pain — Patient profiles — Phases of PRP — Pillars of Support — Platelet-Rich Plasma — Poking the Meta Bear — — Prednisone and the Duck — Prevalence vs Incidence — Privacy & PRP — Progress & PRP — PRP Awareness Day — PRP Parents & Kids — PRPer, PRP-er — **R** Remission — Repurposing — Roadside Emergency Kit — Rude people — Rule of Nine — Rules of the Road — **S** Sauna Suits — Scalp — Showers & Baths — Sharing is caring — Short end of the stick — Signs & Symptoms — Slathering — Sleep deprivation — Stigmatization — Stress & triggers — Swollen legs & feet — Sunlight — Synonyms — Swimming — **T** Tattoos & Body Art — Take Command — Translation Button — Travel & Vacations — Types of PRP —**U** Unicorns — **V** Vacuum cleaners — Venting — **W** Weight gain/loss — Workplace — Why me? — Why you? —**Z** Zits

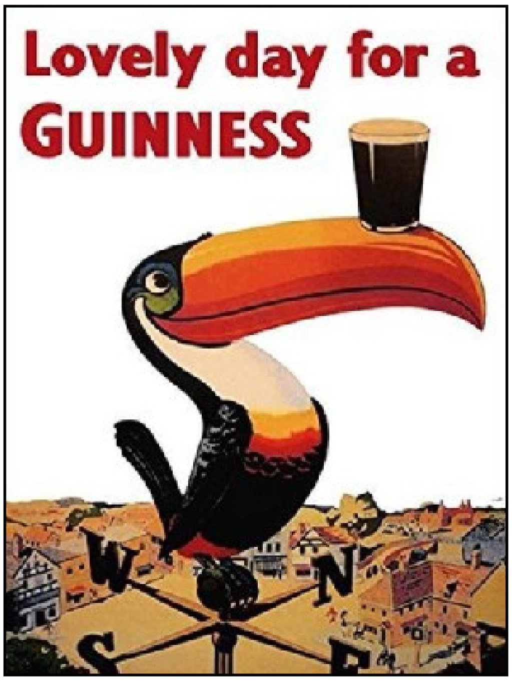
50. Advocacy, Self-

Because PRP is so rare, there are a limited number of "experts" in the treatment of our disease. Self-advocacy is a strategy to help PRP patients and their families fully participate in their healthcare. Self-advocacy is an absolute necessity for patients with a rare disease like PRP.

- ☐ Self-advocacy means standing up for your health—doing everything possible to get high-quality health care.
- ☐ Self-advocacy means regaining a sense of control. At times, you may feel like PRP is running your life. Take charge. Get the information you need to make informed decisions about your PRP treatment options.
- ☐ Self-advocacy enables you to ask for help from family and friends to better manage your version of PRP.

Read about the Five Principles of PRP Self-Advocacy in Appendix E

51. Alcohol Consumption



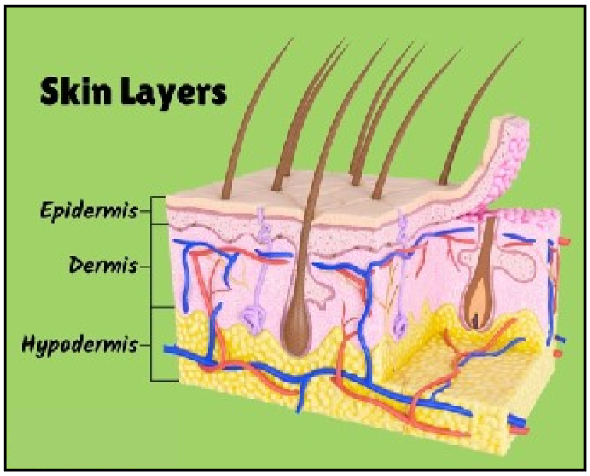
I was prescribed acitretin (25mg- 50mg) for adult-onset PRP during the period from December 2012 to November 2013. I was advised by my dermatologist (University of Texas Southwestern) not to consume alcohol. A liver panel was ordered on the occasion of every clinic visit. Fortunately, my “liver numbers” were all good.

At a point in my PRP journey, the need for a pint of Guinness became an irritant. As luck would have it — the luck of the Irish, you might say — I discovered that Guinness facility in Dublin brewed a non-alcoholic version called Kaliber. Not quite the same, but a non-alcoholic version did soothe the savage beast.

The concensus among PRP patients and caregivers is that a PRP journey is no time to “go on a bender”. As the signs and symptoms of PRP began to wane I found a “ceremonial Guinness” to be therapeutic.

Even today I partake of a Guinness once a month.

52. Anatomy of Skin



PRP patients and caregivers don’t need to become dermatologists. However, we should have a basic understanding of our own skin.

Your skin is the largest organ in your body. It’s made up of several different components.

The skin is primarily made up of three layers.

☐ The *epidermis*, the outermost layer of skin, provides a waterproof barrier and contributes to skin tone.

☐ The *dermis* beneath the epidermis contains connective tissue, hair follicles, blood vessels, lymphatic vessels, and sweat glands.

☐ The deepest layer, the *hypodermis,* is made of fat and connective tissue.

Autoimmune
Diseases
Association

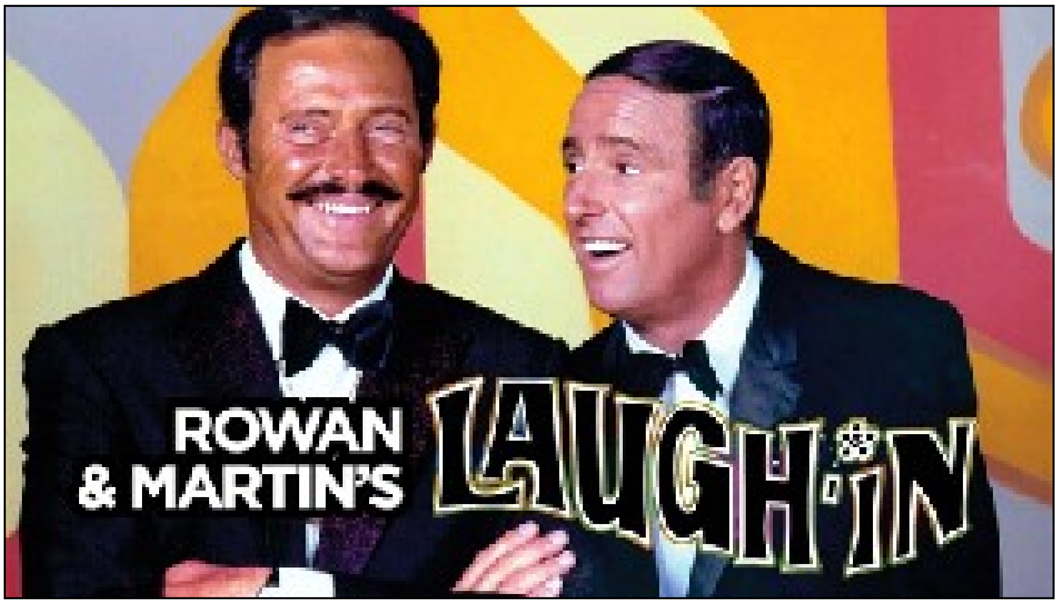
53. Autoinflamatory vs Autoimmune

A frequent misconception among patients and caregivers is that PRP is an autoimmune disease rather than an autoinflammatory disease.

☐ It was on that basis in 2015 that I approached the Autoimmune Diseases Association intent on having the PRP Alliance join. However, AARDA declared that PRP was absolutely NOT an autoimmune disease.

☐ If PRP was an autoimmune disease we would be listed between Pernicious anemia (PA) and POEMS syndrome. We’re not.

☐ That said, I’m sure there will still be PRPers who remain convinced that PRP is an autoimmune disease.



54. Balderdash

As a PRP patient, one of my favorite go-to words as an alternative to “bullsh*t is *balderdash* (Laugh-in, Rowan & Martin).

☐ I used it to respond to a dermatologist who answered “No”: “Are you familiar with the *PRP Survival Guide* and the PRP Support Group?” Always seize the opportunity to enlighten the uninformed.

☐ I lower my voice when I say BALDER-dash to ensure that the person I am balderdashing with knows I am serious.

☐ However, great care must be taken to use Spell Check to avoid the typo “bladderdash, " a non-medical term referring to the consequences of drinking too much water at bedtime, as in, a drank “I had to bladderdash to the bathroom at 3:00AM.

55. Befuddled

Befuddled is another great politically correct term available to PRP patients.

☐ Use it to characterize a healthcare professional's lack of PRP savviness. Rare is the dermatologist who has seen a PRP patient. Even more rare is a dermatologist who has treated a PRP patient. Like us, they are on a learning curve, too.

☐ Since we are on this journey together, we should take every opportunity to share what we know with dermatologists who will listen.

☐ We share a profound desire to enlighten dermatologists with what we have learned. There was a time when we were all befuddled too. But we took time to learn from one another.

56. Biopsies

A biopsy is the missing puzzle piece in the diagnosis of PRP. There are two requirements for a proper diagnosis of PRP:

- ☐ It all starts with the clinical observations of a dermatologist. The more signs and symptoms you present as a patient, the better the odds that a dermatologist will suspect PRP.
- ☐ The dermatologist must also perform a biopsy. The tissue sample is sent to a dermatopathologist for analysis. Based on the rarity of PRP (one in 400,000), the dermatologist should SPECIFICALLY instruct the dermatopathologist to look for PRP.
- ☐ The goal for a yet-to-be-diagnosed PRP patient is a biopsy report that "supports the clinical observations."

57. Caregivers, Rare

While my personal experience is that of a PRP patient, I hold caregivers in high regard. In researching the 60-Second PRP Roadmap, I stumbled upon Rare Disease Caregiving in America. This is a study of 1,406 caregivers living in the United States who provide care to a child or adult with a rare disease or condition. The research was funded by the National Alliance for Caregiving and Global Genes.

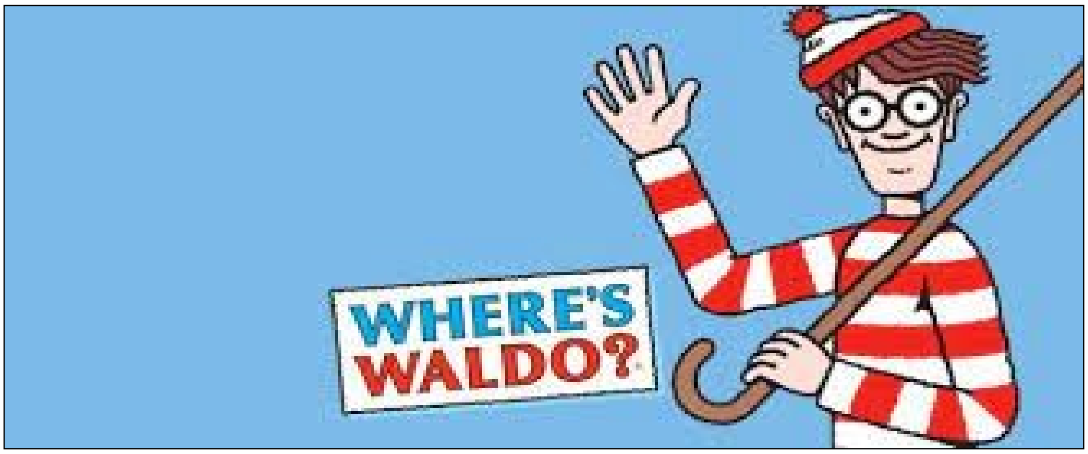
☐ Although understanding the impact of rare diseases on the person with the rare disease is important and worthy of research, this study aims to examine the impact of rare diseases on the caregiver. It is important to note that all data presented herein are from the perspective and experience of the occasional disease caregiver.

☐ A defining feature of rare caregiving seems to be one of expertise. Most "become" teachers, educating healthcare professionals about their care recipient's rare disease or condition (89%). Occasional caregivers can also find themselves researching and accessing clinical trials. Sound familiar? Check out Appendix D —Executive Summary, Rare Disease Caregiving in America,

58. Cause of PRP

What causes PRP? No one knows. However, based on conversations within the PRP community, we can say — with metaphysical certitude — that the onset of *pityriasis rubra pilaris* is neither your failure to return the seat of a toilet (loo in some parts of the world) to its proper “down” position or a punishment for misbehavior.

Over the years, thousands of perfectly wonderful people have — or had — PRP. Conversely, legions of very bad people have dodged the proverial bullet.



59. Census, PRP Global

If Waldo was diagnosed with PRP, we would want to know some basic information about him. In fact, we would like to know the following:

- ☐ Waldo's full name and where he lives (country, not ZIP or Postal Code)
- ☐ Waldo's onset date, onset age, diagnosis date, and current status (active or remission)
- ☐ Waldo's PRP dermatologist and whether Waldo would recommend his dermatologist to another PRPer.

PRP patients and/or their caregivers can use the following link to learn more and access the 2024 PRP Global Census:

https://lp.constantcontactpages.com/sv/wFhTjMr/prpCensus

60. Challenges, PRP

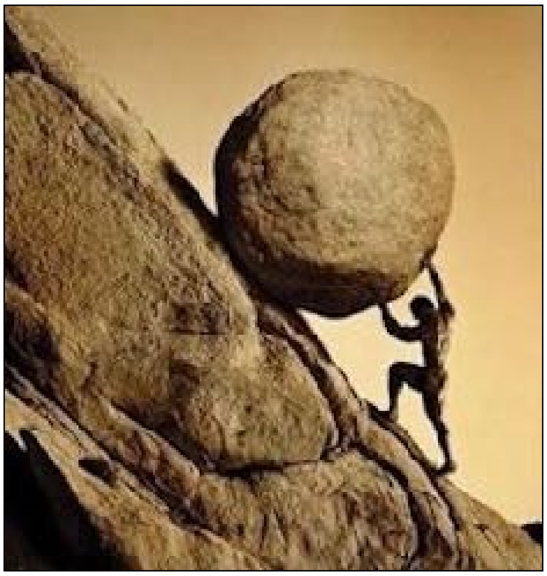
When sharing your PRP journey with others, there is a phrase that helps frame your story into three elements: “the challenges to body, mind, and spirit”.

- ☐ Even though every PRP journey is unique, we all face the same basic challenges: indeterminate severity and duration.

- ☐ Our bodies are the battlefield where pain and itch are too often co-conspirators. Our minds continually calculate whether we can endure endless indignities, impairments, and unexpected losses. A litany of challenges tests our will, e.g., vision, hearing, mobility, dexterity, energy, mental wellness, etc.

- ☐ We seek healing milestones at every turn, and somehow, we must find a previously untested core strength within to fight ferocious demons who are relentless in their attacks. How can we find hope to hold it all together?

- ☐ *I would submit that hope can be found in the PRP global community.*



SCHEDULE AN APPOINTMENT

61. Clinic visits

Take advantage of every clinic or office visit.

☐ **Pre-Clinic: PREPARE**

Don't go to your PRP clinic without questions to ask and information to share.

☐ **During Clinic: PARTICIPATE**

The dermatologist is REQUIRED to do specific tasks. As healthcare providers, they must ask you questions and take notes. Be attentive and responsive (take notes, too).

☐ **During Clinic: TEACH**

YOU may be one of the few PRP patients your dermatologist will ever treat in their career. Take the opportunity to share your unique PRP journey. Focus on three unresolved PRP impairments, e.g., mobility, sleep, and fatigue. Be attentive and take notes. Share the answers with fellow PRPers. Repeat during the next clinic visit, whether in two weeks, one month, 90 days or six months.

62. Closed Group

It's all about your privacy. Facebook offers three types of groups: public, closed, and secret.

- ☐ Public groups are basically general admission. Everyone can find and view the group without needing approval to join. The PRP Facebook Support Group began in 2008 as a Public Group.
- ☐ Closed groups are more exclusive. Non-members cannot view the group's content until they become a member. The PRP Facebook Support Group became a Closed Group in September 2013.
- ☐ Secret groups offer the same level of privacy as closed groups under a cloak of invisibility. The only way to get in is to know the group exists and someone who can invite you.
- ☐ To join the PRP Facebook Support Group, a PRP patient or caregiver must have a Facebook account and search on Facebook for Pityriasis Rubra Pilaris and request to join.

63. Cohort

In medicine, a cohort is a group that is part of a clinical trial or study and is observed over a period of time.

- ☐ The largest cohort of PRP patients ever assembled for third-party research was assembled in late 2018 by the PRP Alliance. 574 PRP patients participated in a survey developed by UCLA David Geffen School of Medicine, USC Keck Medical School, and Kaiser Permanente.
- ☐ The data obtained from this PRP survey was eventually published and made available to dermatologists treating patients diagnosed with PRP.

64. Contagious

While PRP is not contagious, it is essential to include “not contagious” early in explaining why we look the way we look. When dealing with a passerby who stares at your redness and flaking skin in the cereal aisle at Walmart, you might consider the following strategy:

- ☐ Make eye contact. Don’t turn away or look down. Remember, you may have the opportunity to build awareness of PRP with a non-traveler
- ☐ Smile. Combined with eye contact, the “Door of Enlightenment” may swing open.
- ☐ Now say something like: “I have a very rare skin disorder that is NOT contagious. It’s called PRP, which stands for pityriasis rubra pilaris.”

Try not to interpret ignorance for meanness. The best remedy for ignorance is awareness. Whether a PRP patient, PRP caregiver or the friend of a PRP patient/caregiver, YOU are the source of enlightenment.

65. COVID — Assess your own risk

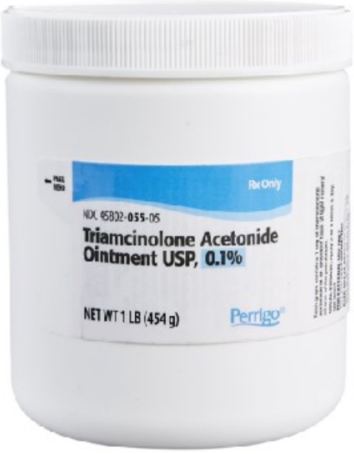
The consensus among those “in the know” is that if you are 70 years or older, your risk is higher than those who are not. Crap. I’m 77.

If you have a chronic medical condition like heart disease, diabetes or lung disease, your risk is even higher. Double crap. I was diagnosed with type 2 diabetes in September 2004. The USS “High Risk” has already sailed for me.

As a PRP patient in remission since April 2014, I depend on my family doctor to help me make the best decisions to help keep me waking up on the right side of the dirt every morning.

66. Cream versus ointment

Before my “official” PRP diagnosis, I was diagnosed with *seborrheic dermatitis* and prescribed triamcinolone *ointment.*



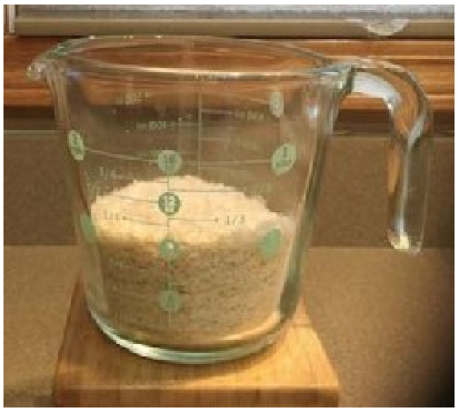
☐ I endured the “slimy feel” for three months. It was an absolutely miserable experience.

☐ When my new dermatologist confirmed I would continue with triamcinolone, I asked him if a “cream version” existed. His response: “Yes, but the ointment is more effective than the cream.”“ How more effective?” I asked.

☐ Then I calculated: ointment feels YUK-kee, and cream = feels YUM-ee.

The decision to “go cream” had an immediate and profound effect on my mental wellness. I slathered with triamcinolone cream for over six months. It was a good decision I did not regret.

67. Cup o ’skin



We all know the adage: “A picture’s worth 1,000 words”. Consider the wife of a PRP patient in Chicago, Illinois, who shared a photo of a measuring cup containing what looks like one cup of uncooked rice.

- ☐ Wrong. At the end of every day, her PRP husband changes his clothes for bed. While shaking his clothes, he collects between 1 and 1-1/2 cups in 24 hours.
- ☐ I’m a solid two-cups-a-day kinda guy! We had an Old English Sheepdog (Guinness) and dark hardwood floors. Before the onset of PRP, I would sweep up a combination of hair and “dog dust” every morning. About two months into my PRP journey, I realized there was more of me on the floor than Guinness. One day, there was more Guinness on the floor than me. It was a healing milestone worthy of a Happy Dance.

68. Curable?

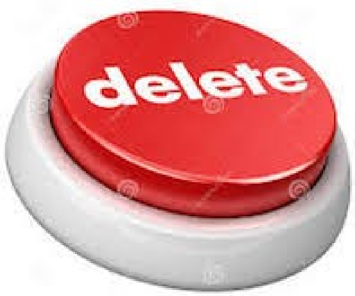
It may be a hard pill to swallow, but there is no cure for *pityriasis rubra pilaris*.

To some, the term cure means the patient no longer has that particular condition after medical treatment. For example, doctors can treat an athlete’s foot using antifungal creams, powders, or sprays. Killing the fungus causing the disease is a cure.

As PRP patients, we depend on our dermatologists to lessen the effects of symptoms and the effects of versions of PRP. Even when I was declared to be in remission (no symptoms and no meds), I did not consider myself cured. The “Red Menace” is still there — lurking in the shadows.

In September 2022, I had a reaction to penicillin — the symptoms of PRP returned. Inflamed skin, head to toe. Rather than lasting 20 months, it lasted less than two weeks. Like I said, lurking in the shadows.

69. Delete Button



Only members of the PRP Facebook Support Group can genuinely appreciate the value and utility of the delete button with which the late Tierney Ratti (Virginia Beach, VA), as lead administrator, dispatched charlatans, the nefarious sellers of Ban-Ray sunglasses, and more than a few evildoers masquerading as us.

The Facebook-based PRP Support Group is a Closed Group with two administrators watching for interlopers. The Tierney Lynch Ratti Memorial Delete Button is primed and ready for action.

At the end of It’s a Wonderful Life. George Bailey’s youngest daughter, Zuzu explains that "every time a bell rings, an angel gets his wings." George looks upward, smiling and says, "Atta boy, Clarence!”

When I push the delete button I look up and say, “Atta girl, Tierney. We deleted another one!”

70. Depression

A PRP patient should not be surprised "down the road" with issues related to mental wellness. Discussing the benefits of a future referral to a mental health professional should be integrated into every treatment plan from Day One.

A PRP patient or caregiver should not be surprised "down the road" with mental wellness issues. The dermatologist can better manage our expectations by being honest with us as part of an over-arching PRP treatment plan.

Our dermatologists should also identify other healthcare professionals we will likely need, e.g., an ear specialist for impaired hearing, an ophthalmologist for impaired vision, and a podiatrist for impaired mobility.

71. Dermatologist, PRP

Excluding the “terminally befuddled, " three types of PRP dermatologists exist.

☐ ***PRP Savvy***

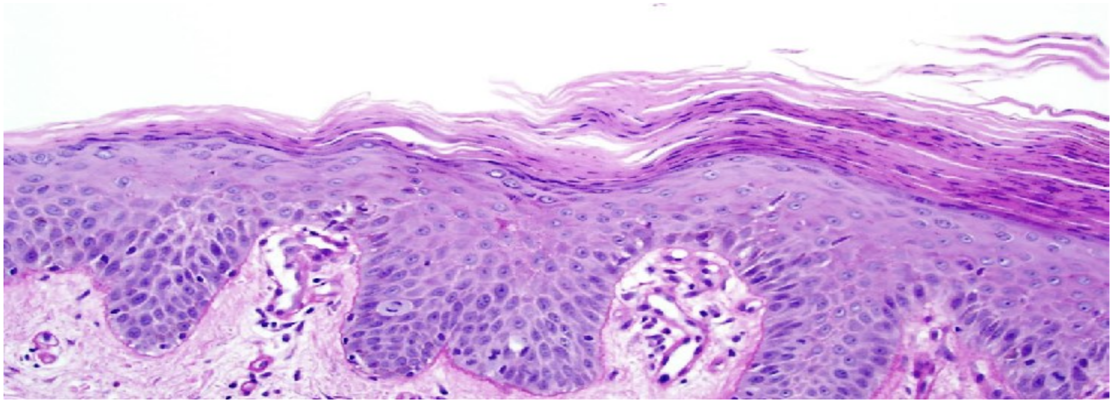
Some get lucky and hook up with a seasoned PRP-savvy dermatologist affiliated with a teaching hospital operating a high-volume dermatology clinic. Having treated more than their fair share of PRP patients.. they know a thing or two because they’ve seen a thing or two.”

☐ ***PRP Experienced***

A dermatologist who has treated one or two PRP patients may be PRP-experienced. Still, that experience does not guarantee familiarity with YOUR version of PRP. They know the PRP mantra:“ What works for one doesn’t work for all.”

☐ ***PRP Curious***

Can’t find PRP savvy or PRP experienced? Find a dermatologist willing to learn; you probably have a keeper.”



72. Dermatopathologist

A proper diagnosis of PRP begins when a Board-certified dermatologist makes clinical observations based on how the patient *presents* in the examination room.

A *dermatologist who suspects* PRP will perform a biopsy and subcontract the evaluation or “reading” of those slides to a dermatopathologist — a pathologist focusing on matters of the skin.

The best outcome for a yet-to-be-diagnosed PRP patient is a finding that “supports the clinical observations” of your dermatologist. That’s all you need to officially start your PRP journey.

73. Dexterity

There may be scientific ways for healthcare professionals to measure and evaluate dexterity and impairments to hands and fingers. However, a 32-ounce jar of R.E. Knudsen Pear Jouice was just as doi=ifficult to open as a jar of Vlasic sweet relish.

Before the onset of PRP, the Knudsen bottle and Vlasic jar were no match for me. In fact, any lid would surrender.

However, as the skin on my fingers thinned, I grew increasingly unable to create the torque required to twist off lids and bottle caps. The loss of dexterity was a painful reminder that PRP was winning the day-to-day battles.

I was routinely thwarted for over nine months. Then, one day, I twisted the lid off the Vlasic and heard the "Poof" of success. The lid turned, and I recorded yet another healing milestone on my 20-month journey.

To this day, opening a jar of Vlasic sweet relish and a bottle of Knudsen pear jiuice brings a smile.

74. Diagnostic Odyssey

The diagnosis of PRP is often delayed when the presenting symptoms of red and itchy skin mimic the signs of psoriasis or eczema. These misdiagnoses can delay proper treatment or provide an opportunity for inappropriate treatments.

A more timely diagnosis of PRP often depends on the dermatologist's ability to consider PRP. Still, the dermatopathologist should also be instructed to consider PRP.

☐ During the summer of 2013, a survey of 487 PRP patients gathered responses from 256 PRP patients who detailed their biopsy experiences.

- PRP diagnosis made with no confirming biopsy: 23.4%
- Biopsy supported PRP diagnosis:: 23.4%
- The diagnosis rendered with no biopsy ordered: 7.8%
- PRP diagnosis made with no contradictory biopsy: 45.3%

75. Diet, PRP

Based on thousands of comments over the past decade, I always invoke the PRP Mantra when the topic of diets is discussed: "What works for one doesn't work for all.

- ☐ Auto-inflammatory diets
- ☐ Autoimmune diets
- ☐ Gluten-free diets
- ☐ Dairy-free
- ☐ Keto
- ☐ More dieting advice: run your dietary plans past your dermatologist, family doctor or nutritionist.

76. Differential Diagnosis

Sometimes, a diagnosis is straightforward. Sometimes it's not. It was easy for an endocrinologist to diagnose my type 2 diabetes with a simple blood test in 2004. It was also easy for the Emergency Room at the Medical City of Plano to diagnose a broken ankle with an X-ray in 2013.

However, when a PRP journey begins, the dermatologist does not have PRP on their radar. Their guess reflects medical training, clinical observations and perhaps a biopsy to make an educated guess. Dermatologists call this educated guess a *differential diagnosis* — not a mistake.

A differential diagnosis is how the medical community defines a diagnosis "other than the one that is finally established." It is more of a "could be." We can't be too hard on them. The fact is, we don't give them much to work with at the beginning.

77. Dowling Oration

The Dowling Oration was delivered by Dr. Andrew Griffiths in Liverpool, England, in March 2003 as he reflected on 35 years as a dermatologist. It is frequently used as a reference in PRP-related research and sets PRP prevalence at one in 400,000.

Griffiths' methodology for calculating the prevalence rate of PRP was a combination of his own patients and the British Association of Dermatologists patients in attendance at meetings where there would be a "show of hands." There are two questions unanswered by Dr. Grifffiths:

☐ What percentage of the PRP diagnoses were supported by a biopsy?

☐ When he asked for a "show of hands," did he deduct the responses of dermatologists who had raised their hands in previous meetings?

78. Duration

“How long will my PRP last?” It’s a pretty common question.

☐ Classical Adult Onset PRP (Type 1): 80%, “spontaneous remission” within three years.

☐ Atypical Adult Onset PRP (Type 2): may persist for 20-plus years.

☐ Classical Juvenile Onset (Type 3) will likely achieve spontaneous remission within one year.

☐ Circumscribed Juvenile Onset PRP (Type 4) where the long-term outcome is unclear with possible improvement in late teens.

☐ Atypical Juvenile Onset PRP (Type 5). Chronic. VERY persistent.

79. Ear Stuff (aka jelly beans)

☐ Sometimes, PRP brings out the STUPID in me. For over three months, I hadn't noticed that my hearing was impaired. I simply turned up the volume on the TV and could hear just fine.

☐ When my wife turned the volume to "normal," I could only hear muffled sounds. She made the appointment with Dr. Kenneth Hsu, an otolaryngologist (ear, nose and throat specialist).

☐ Dr. Hsu removed two plugs the size of jelly beans — one out of each ear. He also discovered inflammation in both ears. I returned every 30 days for the next nine months, then quarterly for a year. The inflammation finally disappeared.

☐ The medical term "Ear Stuff" is cerumen (pronounced ci-RU-min). It is the brown, orange, red or yellowish waxy substance secreted in the ear to protect the skin of the human ear canal.

80. Ebb & Flow

Membership in the PRP Facebook Support Group ebbs and flows. While current members may depart (ebb), new members join (flow). For example, the PRP Facebook Community has grown from 100 in 2013 to over 2,100 in November 2023. What happens to the members who are in remission?

☐ Remissioners who leave (RWL). Some leave when their journey ends. They have had enough and want to distance themselves from anything PRP.

☐ Remissioners who stay. (RWS) Then there are those PRP patients and caregivers who "hang around" when their journey has ended. They need or desire to continue sharing their PRP-related insights and observations with those still facing the challenges of body, mind, and spirit.

81. Efficacy

While many pharmaceuticals used by patients diagnosed with psoriasis or eczema have found relief, there is no universally effective treatment for PRP.

There have been three recent studies on the efficacy of Taltz®, Tremfya® (Oregon Health and Science University), and Cosentyx® (Mayo Clinic).

While the results are promising, the size of the cohorts is too tiny to move the mountain of indifference. However, with a database of nearly 2,000 PRP patients, the PRP Global Community spawns research on the efficacy of treatment options. We must secure the cooperation of PRP patients, dermatologists and pharmaceutical companies.

82. Energy & Fatigue

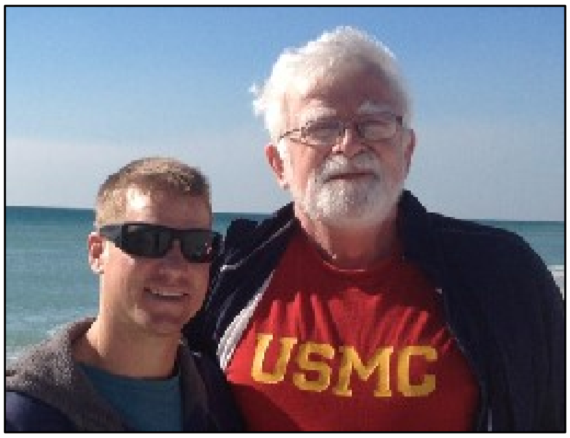
A few months after onset, my“ get up and go, got up and went.” I remember being consumed by fatigue. I was bedridden. My days are filled with nothing.

Then I rediscovered the “on switch” on the back of my iMac. My computer was my ticket out of the despair of fatigue.

It was April 2013 when I turned on my computer and accessed the archives of the now-disbanded email-based PRP support group. It took two months to review 29,000-plus emails going back to November 1997. It was also the beginning of the PRP Global Database.

My PRP-related fatigue was the reason I found refuge with my computer. More importantly, it was the best therapy for what would have become inevitable depression.

83. Exercise & Physical Activity



Before getting mugged by the PRP Red Bastard, I used to walk four miles every few days. Over the past decade, I have marveled how PRP patients who were physically pre-onset generally return to their pre-onset habits.

Walkers walk again. Runners run again. Bike riders ride again. Hikers hike again. It all comes back. And the resumption of physical activity is certainly considered a healing milestone.

I abandoned my sneakers in October 2012, six weeks before my PRP diagnosis, seven weeks before my treatment began and three weeks after putting my sneakers back on.

In mid-January 2013, I walked 5-1/2 miles with my son on Panama City Beach in Florida. But that's another story...

84. Eyes & Vision

A PRP patient can experience one or more symptoms and signs during their journey.

- ☐ Dry, gritty, irritated eyes with an excessive amount of crusty gunk.
- ☐ Teary, blurry eyes. Redness and sensitivity to light.
- ☐ Tight skin on the face that pulls on our eyelids
- ☐ Flaking skin gets in the eyes, made worse by losing our protective eyelashes and eyebrows.
- ☐ Thickened and drooping lower eyelids make looking in the mirror or even seeing it hard.

These interrelated symptoms usually improve with treatment and go away with remission.

85. Face-to-Face

Face-to-face is neither Facetime, ZOOM, Google Meet, nor Skype for PRP patients and their caregivers. It's not about friends and family. Instead, it refers to the number of minutes you spend face-to-face with your dermatologist, e.g., monthly, quarterly, semi-annually or annually.

You should find a new dermatologist if your "Face-to-Face Time" per clinic visit is less than 15 minutes. "Face-to-Face Time".

We are the portal through which our dermatologists can better understand the daily challenges of body, mind, and spirit that we face. Your clinic visit is about to end when your dermatologist steps away from the computer terminal. This is where your FIVE questions are ready to be asked. While your dermatologist may only answer one of the five questions, leave them. If they go unanswered until the next clinic visit, ask them (or revised ones) next time.

86. Feedback

Feedback is the “Coin of the Realm” in the Land of Red & Shed for the PRP global community.

☐ Facebook. A safe environment of kindred spirits — PRP patients and their caregivers sharing insights and experiences.

☐ Posts: Ask questions. Get answers. Reach out to your PRP family.

☐ Comments: Share what you have learned. When you share, we all know.

☐ Reactions. *Love* and *Care* lets PRP patients and caregivers know you appreciate their positions and comments.

☐ In-house surveys: Support the PRP Alliance's efforts to third-party research by teaching hospitals. Stand up and be counted.

☐ Third-party research. Over the past 10 years, we have helped PRP researchers at Oregon Health and Science University, Thomas Jefferson University, UCLA and USC. This is the most credible and impactful research we can and must support,

87. Find-a-Derm

It has always been a needle-in-the-haystack proposition to find a PRP-savvy dermatologist.

☐ Share your location (city, state. province, country) with the PRP Support Group. At least you may get a referral from a kindred spirit.

☐ The next best place is the American Academy of Dermatology’s website at find-a-derm.aad.org.

- Enter your LOCATION,
- Enter PSORIASIS as a specialty, and you might be in the right ballpark. From there, it is a matter of asking questions...

88. Fingernails, Signs & Symptoms

Based on a survey of 300-plus PRP patients, I learned that one-third reported no impact on their fingernails, and another third confirmed the dramatic thinning of their fingernails. The final one-third were devastated, as seen by the words they used to describe their nails/

☐ Appearance: thick, super thick, too thick to cut, ugly, bent upward, curled down, peeled off, gnarly, and detached.

☐ Colors: yellow, yellowish, yellowish-brown

☐ Pain: from pressure, snagging, nail lifting off of the nail bed

☐ Appearance: ridges like razor blades, pits, splitting, dips, lines, cracked, brittle

All PRP patients who experienced fingernail issues quickly reaffirmed that their fingernails returned to their pre-onset appearance.

89. Flaking & Shedding

Skin cells change shape. They start off kind of fat and square. Over time, they move to the top of the epidermis, flattening out. When they get to the top, they flake off. PRP accelerates this process.

☐ There are about 19 million skin cells in every inch of your body.

☐ Your body gets rid of 30,000 to 40,000 old skin cells every day! The skin you see now will be gone in about a month.

☐ The top 18 to 23 layers of your skin are made of dead cells.

☐ New skin cells form at the bottom of the epidermis and move up.

90. Flares, PRP

A flare is NOT the initial onset of PRP. A PRP flare occurs when symptoms of PRP that have been present and manageable suddenly worsen.

While a flare is perceived as a setback, it may be the normal progression of your unique PRP journey. By their very nature, flares are not permanent. Consider it a road hazard as you press forward.

- ☐ It is the collective opinion of a majority of PRP patients that STRESS causes flares. The best stressor I ever heard was from a PRP patient who lamented a flare that began the moment her mother-in-law moved into her house.

91. Freebies

Freebies are hard to come unless you are a PRP patient with a dermatologist who is a problem solver with free samples of biologicals like Cosentyx® or Stelara®, which could be your ticket to the fast lane.

There's another freebie that has been available since 2013. Beiersdorf, makers of Aquaphor® and Eucerin®, offer a program for families in the United States and Canada who use large amounts of these products. The company will send a free case of the product as long as specific instructions are followed.

☐ For more information about ordering a case of Aquaphor or Eucering, please contact Beiersdorf USA at (800) 227-4703 or Beiersdorf Canada at 800-933-0214.

92. Full Bloom

The term “full bloom” is a metaphor and jargon. It describes, with a certain degree of eloquence, how PRP spreads or unfolds. This peaceful metaphor has its limitations. Some might find a more sinister term, e.g., Kudzu, Japanese Barberry, English Ivy, Purple Loosestrife, or Japanese Honeysuckle. If you do use “full bloom,” please do not refer to exfoliation as “harvesting blooms” or bathing in oatmeal as “hydroponics.”

Here, every PRP patient in full bloom likes to go to a grocery store and deal with the stares of judgmental onlookers. “Snow Globe in Aisle #3!”

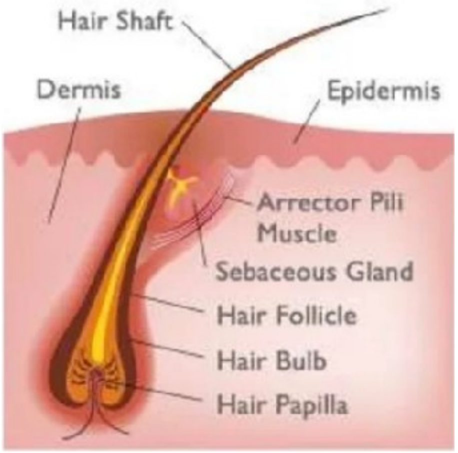
93. Good start on a PRP journey

Whether you are a newcomer or a seasoned traveler, the more you know about *pityriasis rubra pilaris*, the better you will be for the journey ahead. The following information has been developed to enlighten PRP pati

- ☐ Read the 60-Second PRP Roadmap, cover-to-cover… and Appendices
- ☐ Join the PRP Support Group and participate with posts, comments and reactions (Like Love and Care)
- ☐ Complete the PRP Global Census
- ☐ Respond to an in-house survey from the PRP Alliance
- ☐ Participate in third-party PRP research endorsed by the PRP Globa;l Community.

94. Hair Loss

PRP can cause thinning of your hair. All your hair. Anywhere. Everywhere.



- ☐ This is both appearance-altering and depressing.
- ☐ In addition, some medications, such as retinoids and immunosuppresses, can cause hair loss to one degree or another.
- ☐ It differs with each individual. Some only have mild thinning; some lose all their hair, including eyelashes and eyebrows.
- ☐ Just remember, the collective experience of the global PRP community strongly suggests that your hair will grow back.

This brings us to the Happy Dance.

95. Happy Dance

The Happy Dance is the counterbalance to Venting and Howling at the Moon. The Happy Dance is a community celebration. Just as every PRP journey is unique to the patient, so are the healing milestones.

Here are some of mine based on an onset date of August 8, 2012.

☐ HAPPY DANCE #2 (January 2013): Swollen feet returned to normal size. I could retire my “no back” slippers and wear my sneakers again.

☐ HAPPY DANCE #4 (July 2013): One day, I noticed my kneecap was “leaking.” It was as if my sweat had been trying to get out for 11 months and finally found an exit on my knee.

☐ HAPPY DANCE #7 (April 8, 2014). At the end of a clinic visit, Dr. Arturo Dominguez declared I was “in remission,” which I interpreted as “no symptoms and no meds.”

96. Healing Milestones

There should be a healing milestone for every symptom of PRP and every indignity we suffer. Our individual Journeys can be so long and unrelenting that we must recognize markers along the way. Here are a few.

☐ Drive myself to Walmart.

☐ Sleeping through the night (11 PM to 7 AM)

☐ Feet no longer encased in thick skin

☐ No longer needing — or wanting — a handicap placard.

Mark your calendar whenever you recognize ANY healing milestone, do the Happy Dance, and post a message to the PRP global community so we can celebrate with you.

97. History of our name

It took 61 years to get our name right.

☐ 1828, James Shooter was admitted to St. B1835, Dr. Claudius Tarral wrote a *treatise on Skin Diseases*). He saw the never-before-identified skin disorder as a variant of psoriasis.

☐ 1856, Dr. Marie Devergie published the "original description" of PRP. He saw the disorder as a combination of disorders

☐ 1877, Dr. Richaud recognized the skin disorder as a distinct entity.

☐ 1889, Dr. Ernest Besnier fixed the name of the disease as *pityriasis rubra pilaris* from the Latin: *pityriasis* (scalelike skin), *rubra* (red) and *pilaris* (hair follicles)artholomew's Hospital in London, England, with a never-before-identified skin disorder.

Dr. Ernest Besnier

98. Howling at the moon

Unlike venting, howling at the moon does not require a response from kindred spirits. The mere howling brings immediate and long-lasting relief and satisfaction to the howler.

- It is critical, however, that howlers explain why they are howling. Without background, the howl is just noise.
- Some believe that the energized storytelling associated with howling is, in itself, therapeutic to the howler. You be the judge.
- As a PRP advocate, I had conjured up advocacy efforts that sounded great when I was alone at my computer but sounded much different when I howled them. Kindred spirits sometimes caution, “Wait, Papa Bill. Not yet”.
- Sometimes, I howled at the moon, and 250 PRPers would complete a survey. That was serendipity at play. Never howl at the moon and expect anything in return. Just enjoy the echo!

99. Hijacking a post

Hijacking a post was a common occurrence 7-10 years ago. It goes like this:

- ☐ A PRP Facebooker (#1) starts a conversation with a POST.
- ☐ PRP Facebookers (#2-8) comment,
- ☐ PRP Facebooker #8 replies to #3 on an unrelated topic,
- ☐ It doesn’t take long for the train to go off the track.

That was then. Today, the posts and comments flow seamlessly. There is peace in the Lands of Chat, Hope, and Share.

Peer Review Note: This is an opportunity
to discuss other aspects of online ettiquette for
Mmembers of the PRP Support Group (aka PRP Facebookers)

100. In My Humble Opinion (IMHO)

Every PRP patient and/or their PRP caregiver is a subject matter expert of THEIR OWN VERSION of PRP.

Our opinions are hard-earned.

☐ When we share, whether we say it or not, our "sharings" should be prefaced by "In my humble opinion."

☐ Every PRP journey is unique to every PRP patient. While we are in this together, we each have a unique perspective. What works for one doesn't work for all.

☐ While most of us are NOT medically trained, our observations can enlighten one another and the dermatologists and healthcare professionals who serve us.

101. Internet, Dr.

The internet makes finding health information easy and fast.

☐ Much of the information on the internet is valuable; however, the internet also allows rapid and widespread distribution of false and misleading information.

☐ You should carefully consider the source of information you find on the internet and discuss that information with your healthcare provider.

☐ The National Institutes of Health guidelines allow patients and caregivers to find and evaluate health-related information online. Check out Appendix F and G and the end of the 60-Second PRP Roadmap.

- Appendix F: Finding health information on the Internet
- Appendix G: Evaluating health information on the internet

102. Itch

Did you ever think of itching as being anything more than an occasional nuisance? And now you find it quite painful? Itching or pruritus is a common symptom of PRP. The best relief can be found in antihistamines, antidepressants, creams, and lotions containing camphor or other ingredients to soothe the itch. Your best reliefs:

- ☐ Avoid scratching! It can lead to infection! Easier said than done.
- ☐ Avoid long soaks in the tub! They can dry your skin!
- ☐ Enjoy warm to cool showers (if the spraying water isn't too painful)
- ☐ Lavish amounts of creams and lotions. The word is slather!
- ☐ Whatever medications your doctor prescribes. Ask about hydroxyzine.

103. Land of Chat

Our Facebook-based PRP Support Group is the CLOSED GROUP venue for all PRP patients and caregivers to share what we know and learn what we don't...

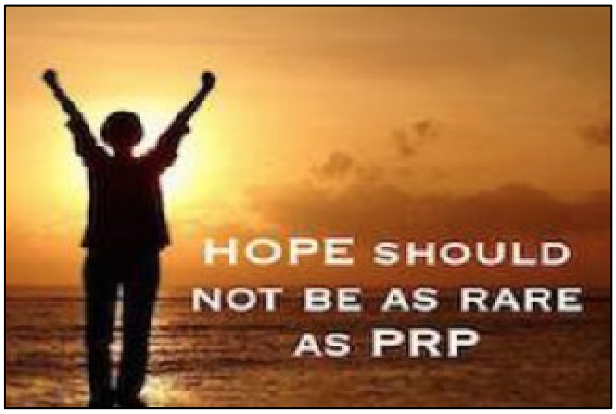
As of November 1, 2023, the PRP Global Database maintains 1,713 PRP patient profiles, including 1,713 PRP Facebookers and 142 non-members.

☐ Posts: 1,031 (86 per month)

☐ Comments: 18,205 (1,517 per month)

☐ Reactions, e.g., Like, Love, etc.: 29,417 (2,451 per month).

There is a lot of chatting going on.



104. Land of Hope

Based on a repurposed meme posted by a PRPer, a new PRP Facebook Support Group homepage was immediately created.

"HOPE should not be as rare as PRP" should be our vision. There are three benefits perceived by PRP patients and PRP caregivers when they become PRP Facebookers:

☐ *I am not alone — a PRP community is on the same journey as me.*

☐ *I have hope — the collective hope of the PRP community*

☐ *I have support — a PRP community has my back*

105. Land of Share

When joins the PRP Support Group, they hear another mantra:

- ☐ People who care, share
- ☐ People who share learn.
- ☐ People who know, share.

Then, human nature takes its course.

- ☐ It doesn’t take a PRP Facebooker long to begin sharing. Newlies posted a question but never had access to another PRPer. They’ve been waiting long and want to jump in with two feet.
- ☐ Some start with a comment. They bond with a newly found family and want to “break the ice.”
- ☐ Others just react with Like, Love or Care to tell the post’s author that you appreciate what they have written.

106. Laundry

Here's something that dermatologists are unlikely to share with a newly diagnosed PRP patient: "The topical ointments, cream, and lotions you will be prescribed and the relentless shedding of skin, will put an extraordinary mental and physical strain on you, caregivers, and push your washing machine to its mechanical limits." For many, the challenges of washing clothes, sheets and blankets are so impactful that Healing Milestones may include

- ☐ You don't have to shake out clothes before you wash them once.
- ☐ Return to when patient and caregiver clothes could be put in the same laundry batch, e.g., one load fits all.

107. Lip Care

Like too many PRP patients, my lips took a beating. I'm unsure what I put on them to reduce the dryness and cracking, but the results were marginal.

One day, a fellow PRP patient described the condition of their lips in a post and recommended Burt's Bees, Beeswax Lip Balm. I had never heard of it. I thought it was one of those products they have in England. I went to my nearby CVS Pharmacy, and there was a GIANT Burt's Bees display.

My lips were never dry and cracked again. Moreover, I still use the product during the winter months.

I'm sure there are other great products for lip care, and I am not endorsing Burt's Bees. PRP patients with dry and/or cracked lips should "pucker up" and find something that works for them.

108. Managing Expectations

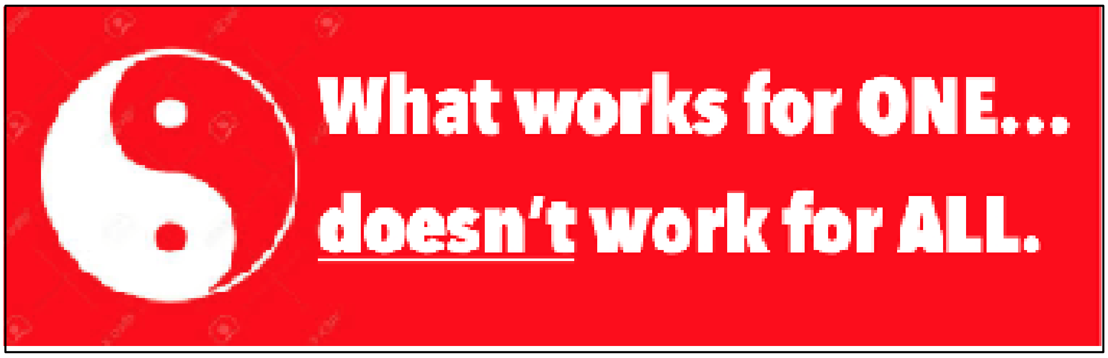
Managing our expectations should be the cornerstone of treating PRP.

☐ Reasonable expectations of PRP patient needs should include the following:

- to be listened to
- to receive clear explanations and instructions about PRP
- to be treated with professionalism, compassion, concern and care, and

☐ Examples of unrealistic patient expectations include:

- I want to discuss several significant problems in one office visit. Pace yourself. Establish priorities. The road we travel is long.
- Wanting your dermatologist to be on speed dial. Getting an email address is a good start.
- I thought that our dermatologist had all the answers. How could they? For too many, is the PRP learning curve flatlined?



109. Mantra, The PRP

The mantra heard within the PRP community is simple, true and loud: “What works for one doesn’t work for all.” From the PRP patient perspective, there are two primary objectives in the treatment of PRP:

- ☐ relieving symptoms as they appear
- ☐ achieving either long-term remission (no symptoms, no meds) or, at the very least, tolerable long-term maintenance.

The challenge is finding a treatment protocol that resolves our symptoms, is available (affordable), and works.

110. Metaphor, The PRP

Very early in my PRP journey, I decided that the road metaphor had lots of potential.

- ☐ In my mind's eye, I could see a series of city limit signs for towns like Onset (population: 1), Diagnosis (a wrong turn will take you to the town of Misdiagnosis) and the twin cities of Hell and Agony (acute stage).
- ☐ The road to Remission has off-ramps like Relapse and Flare-up. I even had the opportunity to take the exit to Blood Clot twice.
- ☐ Some of us travel in the fast lane, others need Roadside Assistance
- ☐ Hopefully, we all get to visit the Land of Hope and visit with kindred spirits. You get the picture.

111. Mirror, rearview

Looking in the rearview mirror can help you see and appreciate where you are today.

- ☐ Some Healing Milestones, like the return of sweating, are in your face immediate and earth-shaking. One day, you don’t sweat. The next day, you flow like a fountain.
- ☐ Other Healing Milestones, like the return of dexterity, take time. It is hard, if not impossible, to see PRP improvements daily or weekly. Then, you can open a vacuum-sealed jar of Vlasic sweet relish one day. The ability to open jars returned much earlier, but I hadn’t noticed until I stood over the new jar.

112. Mobility

There was a time during my PRP journey when I was bedridden. It was a self-imposed banishment from the rest of the house.

☐ Then, one day, I ventured out of the carpeted bedroom and shuffled across dark hardwood floors, leaving a trail of white flakes in my wake.

☐ Weeks passed, and I finally ventured out my front door and felt the cold air of winter. More weeks passed, and I was driven to Walmart, where I reluctantly used an electric scooter.

☐ For nearly a year, I used a handicapped sticker to get a better parking place — closer to a store. I kept looking at the date on the sticker and wondered if I should get it renewed in October 2013. When the time came, I didn't renew the sticker. I added the retirement of my handicapped sticker to my Healing Milestone list.

113. Nitrile gloves

In my humble opinion, based on feedback from PRPers, dermatologists should make their PRP patients aware of nitrile gloves. Forget the white cotton gloves that soak up ointments and creams.

- ☐ Nitrile is made from an allergy-safe compound that feels much like latex but is much stronger, costs less, and is more comfortable to wear. It's a perfect compromise between latex and vinyl.
- ☐ Wear day and night. I Take them off when you shower and apply topicals.
- ☐ Another PRP mantra: without nitrile gloves, my hands are useless.

114. NORD PRP Report

On September 20, 2017, the National Organization of Rare Disorders (NORD) replaced the 2007 PRP Report with the 2017 revision. Here are some not-so-random observations of the revision process:

- ☐ Total word count increased by 4,010 — from 1,274 to 5,284.
- ☐ The first draft was made available to the PRP global community. The revised second draft was made available to dermatologists treating or having treated PRP patients.
- ☐ The final draft was shared with the dermatology departments at Thomas Jefferson University (PA), Oregon Health and Science Univesity (OR), and the Icahn Medical School (NY).

The NORD PRP Report is a must-read for every PRP patient and caregiver. The link should be shared as appropriate with other family and friends, co-workers and employers, teachers and school administrators, and even dermatologists.

115. Odds

The Orphan Drug Act defines a rare disease as a disease or condition that affects less than 200,000 people in the United States. PRP is a very rare skin disorder with a prevalence of 1 in 400,000. For those with a need to know the odds by type:

- ☐ Type 1: 55% — Classic Adult — One in 800,000
- ☐ Type 2: 5% — Atypical Adult — One in 8 million
- ☐ Type 3: 10% — Classic Juvenile — One in 4 million
- ☐ Type 4: 25% — Circumscribed Juvenile — One in 1.6 million.
- ☐ Type 5: 5% —Atypical Juvenile — One in 8 million
- ☐ Type 6: HIV-associated — too rare to calculate meaningful odds.

116. Onset — Words to describe

In 2014, the PRP Global Census collected the words 477 PRP patients used to describe onset symptoms:

☐ rash (201),

☐ spot (97)

☐ itchy (70),

☐ blemish (19).

☐ Other onset terms included sunburn, patches, shedding, dandruff, bumps, and peeling (57).

Words used to describe the location of onset symptoms included: Head (168), which provides for scalp, face, forehead, head, eyes, ears and nose;

☐ upper torso (86), which includes chest, back (36), shoulder and torso,

☐ upper extremities (66): hands, arms and fingers, and

☐ lower extremities (38): feet and legs.

117. Pain

Pain is a passenger on every PRP journey. We each have a pain threshold. My father set the bar for pain for me in the '50s.

We lived in a small two-bedroom house with a cellar. Dad would retreat to his workbench after dinner and build something. Anything.

I heard "Whack! Whack! Whack" through the thin floors as he hammered nails into something. Anything.

Then I inevitably would hear, "Whack! Whack! Thud" as the hammer smashed his thumb. Without hesitation, I would listen to an immediate high-pitched "SWEEEET Jesus!" Almost immediately, the "Whack! Whack! Whack" began anew.

There were plenty of "SWEET Jesus" moments during the first six months of PRP, but then they became "Baby Jesus" moments. When I finally reached remission, it was a "Thank God" moment.

118. Patient profiles

A patient profile is a simple snapshot of a PRP patient based on information maintained in the PRP Global Database. There are 5,000 patient profiles, of which 2.000 have valid email addresses.

- ☐ Name: Never shared. PRP mantra: Numbers, not names
- ☐ **Location**: All we gather is country, state/province. No postal codes. Even the city/town is optional.
- ☐ **Email address**: Never shared with third parties, e.g., researchers.
- ☐ **Onset date**: When did the symptoms of PRP first appear?
- ☐ **Onset age**: At what age did the symptoms of PRP first appear?
- ☐ **Current status**: — Active, Remission or in between

119. Phases of PRP

Over the last decade, I have come to believe there are four phases of PRP.

☐ Phase One – Onset: Includes the arrival of signs and symptoms

☐ Phase Two — Acute. As PRP evolves, the forces of pain and discomfort converge. While the duration is unpredictable, it has an end.

☐ Phase Three — Rest of the Journey. Measured in time. For some, it can be a year or two. Most fall into the 3-5 year bucket. A smaller number are in it for a “longer haul.” x

☐ Phase Four — Managed. This is where the symptoms have become VERY manageable, but medication is still required. A somewhat smoother ride.

Remission is not a phase. Remission was my objective. After 20 months, I had neither symptoms nor meds.

120. Pillars of PRP Support

☐ Pillar #1 — PRP Global Community

- The PRP Global Community is a collection of 2,000 PRP patients and caregivers around the world, including the USA (1,097), United Kingdom (228), EU (176), Australia (142) and Canada (92).

☐ Pillar #2 — PRP Facebook Support Group|

- With over 2,000 members, the PRP Facebook Support Group is a juggernaut for global communication.

☐ Pillar #3 — PRP Survival Guide

- The *PRP Survival Guide* has over 500 web pages organized to help PRP patients and caregivers learn about PRP from the patient's perspective.

☐ Pillar #4 — PRP Global Database

- The PRP Global Database maintains email addresses for over 2,000 PRP patients/caregivers to coordinate third-party research.

121. Platelet-Rich Plasma

When a newly diagnosed PRP patient uses the acronym “PRP" to search for pityriasis rubra pilartis, they retrieve 236 million results in 0.36 seconds for platelet-rich plasma PRP). Platelet-rich plasma therapy uses injections of a concentration of a patient’s own platelets to accelerate the healing of injured tendons, ligaments, muscles and joints.

Try again. A search for *pityriasis rubra pilaris* will retrieve 330,000 results in 034 seconds.

122. Poking the Facebook Bear

In April 2018, seven PRP Facebook Support Group members sent private messages to 660 fellow members. We were asking members to complete a PRP-related survey.

Some of us learned — the hard way — that when you send too many personal messages during a short period, you seriously “Poke the FACEBOOK Bear.”

Some of us were forced into a 24-hour to 48-hour TIME OUT by the Facebook Police.

While we persisted and were not thwarted by Facebook’s growls and snapping jaw, there was universal agreement that the undertaking would not be attempted again.

123. Prednisone and the Duck

In early September 2012, my first dermatologist mistakenly diagnosed my redness as *seborrheic dermatitis*. At the time, the inflammation was limited to my face and neck. She prescribed ever-increasing dosages of prednisone for the next two months with no improvement.

On the evening of November 8, I took my first 60mg dose of prednisone. I retired for the evening only to be summoned by a 20-foot rubber duck at 3:00 AM. Fortunately, the vaunted bedroom ceiling easily accommodated the duck and a school of colorful and equally oversized tropical fish.

While arriving paramedics were unable to confirm the duck, they did admit that none of the first responders on the scene had recently taken 60mg of prednisone.

124. Prevalence vs incidence

According to the National Institutes of Health, I have misused the terms PREVALENCE and INCIDENCE. Specifically, I have been treating them as interchangeable. They are not.

Incidence is a measure of the number of new cases in a population in a specified period, e.g., during 2023 (January-September 2023)

☐ Incidence for PRP during the period January-September 2023: 286

Prevalence is the proportion of a population with a specific characteristic in a given time, regardless of when they first developed the characteristic.

☐ Prevalence equals currently active and not in remission: 1,595

HIPAA
Health Insurance Portability
& Accountability Act

125. Privacy vs PRP

The Health Insurance Portability and Accountability Act of 1996 (HIPAA) is a federal law requiring national standards to protect sensitive patient health information from being disclosed without the patient's consent or knowledge.

- HIPAA only applies to covered entities, health plans, health care clearinghouses, and health care providers who electronically transmit any health information concerning transactions for which the U.S. Department of Health and Human Services adopted standards.
- The PRP Alliance is not obliged to adhere to HIPAA standards of protecting private health information (PHI). We just do.
- The PRP Global Database is on a HIPAA-compliant server with all the bells and whistles a hospital would require.

126. Progress & PRP

Everyone's PRP experience is unique. There are pre-onset signs that include dandruff and a crusty scalp or limited red patches or scaling of the skin.

☐ At some point, the intervention of a healthcare professional is sought. By then, our journeys are well underway.

☐ The progressive inflammation may start on the head and work down, start on the feet and work up, or bounce around with unsettling randomness.

☐ Somewhere along the way, "all hell breaks loose," and the Acute stage kicks in. This is the period where the progression seems to stop. It was a period when I felt like I was hovering over hell.

☐ The duration of this nightmare phase is unknown. For most, it may last a month or two or three. Sometimes longer.

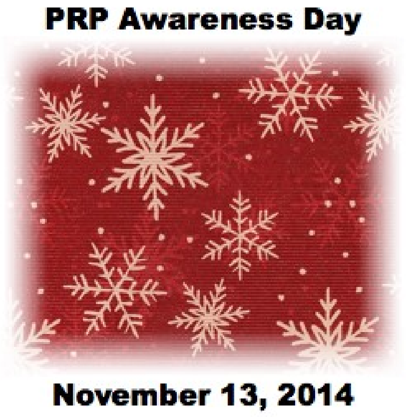
☐ Fortunately, when the acute phase ends, it stays in your rearview mirror. Unfortunately, a longer, more protracted and unpredictable journey goes ahead.

127. PRP Awareness Month

The PRP global community started with a PRP Awareness DAY on November 6, 2014, exactly 17 years after Jean-Luc Deslauriers posted the first message to an audience of 16 PRPers.

PRP Awareness Day was repeated in 2015 on November 6.

PRP Awareness Day was expanded to a month-long event the following year and every year thereafter.



PRP Awareness Month has always been a private, community event with a focus on education. That focus will be expanded n 2024 as advocacy efforts broaden to include dermatologists.

128. PRP Alliance of Parents & Kids

There is a limit to what we know about the PRP Juvenile Onset Community, a subset of the PRP Community Database

- ☐ Every year doe the past eight years, I lament the fact that PRP parents have been unable to establish an active community within the PRP Support Group.

- ☐ The PRP Global Database currently has 1,893 PRP patient profiles of which 201 have an onset age of less than 18. There are, however 759 PRP patient profile where the ONSET AGE datapoint is missing. How many PRPers diagnosed with Juvenile Onset PRP are yet-to-be identified.

- ☐ With over 200 PRP parents and young adults with Juvenile Onset, there are certainly enough souls to make it happen.

- ☐ All it takes is a few PRP parents who want to want to create an alliance of PRP parents, PRP kids and adults who were diagnosed with Juvenile Onset PRP as children.

- ☐ The PRP Global Database maintains PRP patient profiles for over 200 patients diagnosed with Juvenile Onset. For perspective, there were fewer than 120 TOTAL members 120 back in November 2013. If you are a PRP parent, the proverbial ball is n your court. SEE Appendix B for details.

129. PRPer, PRP-er

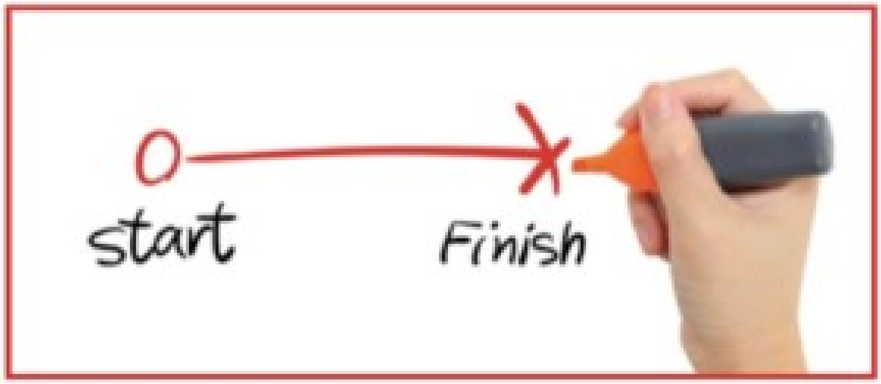
Within the PRP global community, some like the moniker “Pretty Red People.” Many more kindred spirits have repurposed the acronym and call themselves PRP-ers or PRPers. The hyphen is optional.

As jargon among kindred spirits, PRP-er is totally acceptable.

☐ Problem #1 – However, when talking to a dermatologist, using PRPer will earn you a blank stare of profound befuddlement. That’s the problem with jargon. A person with Deep Vein Thrombosis is not a DVT-er. Neither is someone with Attention Deficit Disorder an ADD-er.

☐ Problem #2 — As a retired public relations professional, I was initially taken aback by the use of “PRPers.” I heard PR pee-ers in my Mind’s Eye — or perhaps my Mind’s Ear. That would be a PR professional with a urinary condition. Oh my.

Again, among kindred spirits — PRP-er is fine. Out among civilians, however., please consier “patients with PRP”.



130. Remission

For most PRP patients and caregivers, a diagnosis of pityriasis rubra pilaris is life-altering.

Even before I started treatment, I wanted to know how long my PRP would last. I was told three to five years. My journey nlasted 20 months. I was lucky!

My PRP treatment plan was my roadmap to remission. Yet, neither PRP-savvy dermatologists nor the PRP global community of patients and their caregivers have an agreed-upon definition of remission.

Does remission mean “no symptoms with no PRP meds?" Or “no PRP symptoms.” Or is it something else entirely? There is no consensus at this time.

The terminology used in the PRP Global Datanase offers four options:

☐ Active — Unmanageable

☐ Active — Manageable.

☐ Remission — With remnants.

☐ Remission — Full (no symptoms and no PRP-specific medicatiions)

131. Repurposing drugs

Drug repurposing involves identifying FDA-approved drugs that may be effective for diseases not currently approved.

- ☐ No treatments exist for over 7,000 diseases, of which over 600 are rare skin disorders. Includes PRP.
- ☐ Developing a new drug can cost $2-3 billion and can take 13-15 years before becoming approved for widespread use. Rare disorders provide no incentive for research.
- ☐ We already know the efficacy of drugs approved for psoriasis and atopic dermatitis. Without FDA approval, health insurance doesn't cover costs.
- ☐ By definition, rare diseases do not affect a large patient population and often do not present enough commercial incentive for pharmaceutical companies to develop new treatments. There are an estimated 850 PRP patients in the US.

132. Roadside Emergency Kit

Since pityriasis rubra pilaris is easily described as a journey, we as travelers should prepare for the inevitable emergencies that will befall us en route.

Such a PRP Roadside Emercy Kit should have the following:

- ☐ Paperback copy of the 60-Second PRP Roadmap
- ☐ Circle of Hope Referral card: Offer a card with contact information for the *PRP Survival Guide* and how to join the PRP Support Group.
- ☐ Dermatologist ID: Name and contact information.
- ☐ PRP Patient Journal for note taking. Bring this to every dermatology clinic visit.

133. Rude People (not family or friends)

When you're 6'3", weigh 220 and always wear a US Marine Corps t-shirt, people aren't often rude. But I listen to how my kindred spirits react to rudeness. Forget rude — it's ignorance!

Here's what I would say to a rude passerby:

☐ 1st volley. "It's not contagious. Hurts like hell. Only 850 have what I have in the US, 69 in Texas and 12 in the Dallas-Fort Worth area."

☐ 2nd volley. "Imagine a painful sunburn that covers your entire body for several years. It's hard to walk, and my hands are so sore I can't open a jar of Valsic sweet relish."

☐ 3rd and final volley. It's called pitysiasis rubra pilaris. The cause is unknown, and there is no cure. I just try to get through each day. Thanks for the opportunity to vent.

134. Rule of Nines

As self-advocates, PRP patients and caregivers must communicate accurately with healthcare providers. Accuracy was necessary at the beginning and end of my PRP journeys. Until I discovered the Rule of Nines, my coverage estimates were always incorrect. No guesses now...

The Rule of Nines is a technique used to evaluate burn victims. The results help guide treatment decisions, including fluid resuscitation, and aid in transfer to a burn unit. It also can apply to PRP patients.

- ☐ 9% — The front and back of the head and neck
- ☐ 9% — Front AND back of each arm AND hand (18% total)
- ☐ 9% — Chest AND stomach (18% total)
- ☐ 9% — Upper back AND lower back (18% total)
- ☐ 9% — Front AND back of each leg and foot (18% total)
- ☐ 1% — The groin

135. Rules of the Road

New PRP Facebook Support Group members are asked to follow basic, common sense guidelines.

- ☐ **Posts & Comments**: Any topic is fair game as long as it relates to the PRP experience.
- ☐ **Sharing**: People who care, share. People who share teach. We all win.
- ☐ **Religion**: We are a venue of tolerance. Some of us pray, hug, and think good thoughts. Goodwill comes in a variety of flavors.
- ☐ **Social-Political Issues**: Other than the legalization of marijuana for medical purposes and issues related to the FDA's repurposing of approved drugs for PRP, we steer clear of politics.

136. Sauna Suits

For burn patients, sauna suits help keep moisture and heat contained and enhance the effects of medicated ointments or creams.

☐ The sauna suit is very durable.

☐ Moisten your skin, Take a bath or shower, wash your skin well, and towel dry,

☐ Apply whatever your normal regimen is, including medicated ointment or cream.

☐ Put the sauna suit on, followed by your clothing for the day. At bedtime, sauna suits work as pajamas.

☐ While not an endorsement, DELASCO LLC is a source for sauna suits and is based in Council Bluffs, IA.

137. Scalp

The skin on your scalp is one of the thickest sections of skin on your body. It is thickest at the back of your head and gradually thinner at the front.

The scalp has difficulty maintaining and replenishing hydration compared to the skin found in other places. This vulnerability of the scalp invites all sorts of skin problems compared to other areas of the body.

There are three categories of shampoos used by PRP patients. Each option offers positive benefits as well as disadvantages. All are worthy of consideration. Ask your dermatologist which product is recommended and WHY?

- ☐ Coal Tar Shampoos
- ☐ Salicylic Acid Shampoos
- ☐ Shampoo with steroids

138. Scam Alert

When the text message I received mentioned the “Catalog Of Federal Domestic Assistance,” I knew a kindred spirit had been hacked.

It doesn’t happen often, but when it does, I block the scammer’s email from seeing my posts, tagging me, inviting me to events or groups, messaging me or trying to add me as a friend. The hacker cannot access the Land of Hope or me in seconds.

I also contacted the real PRPer, gave him our link and asked him to rejoin our group. Usually, the PRPer who was hacked is already aware and has warned friends.

Remember — the posts, comments, and images shared with one another are private. As they say, “What happens in the Land of Hope stays in the Land of Hope.”

139. Showers & Baths

Find what works for you. A shower was too painful for me — a combination of feet and the spray. My daily salvation was a daily bath.

☐ Step 1 — Fill the tub with hot water, hot enough to stay submerged in water warm for at least 30 minutes. Set the timer and do nothing but lay there submerged. Listen to the muffled sounds from the TV through the closed door. Sometimes, add a candle.

☐ Step 2 — The sound of my timers signals the start of my nightly "slathering." I start with triamcinolone cream on damp skin — everywhere except my face and ears (Desonide® went there) and my palms and soles (combination of urea lotion and Clobetasol®).

☐ Step 3 — Time for my evening cocktail: hydroxyzine, Ambien® and two Extra Strength Tylenol® capsules. As long as I don't drink water after 9:00 PM, I'm asleep in 15 minutes, from 11 PM to 7:00 AM.

140. Sharing = Engagement

The cornerstone of the PRP Facebook Support Group is engagement. Here are the NUMBERS for January 1 through December 31, 2023:

- ☐ Number of posts: 1,031
- ☐ Number of comments: 18,205
- ☐ Number of reactions, e.g., Like, Love: 29,417
- ☐ Total sharings: 48,653
- ☐ Number of active members: 1.644
- ☐ Average sharings per active member: 30.6

These numbers represet the vital signs of the PRP Support Group

141. Short end of the stick

It doesn’t take long to realize that having a rare disease has consequences beyond the daily challenges of body, mind and spirit. We get the proverbiasl “short end of the stick” on many levels.

With a prevalence of only one in 400,000, few pharma companies are interested in funding PRP-related research.

Kudos to:

☐ Lilly (Taltz) funded trial at Oregon Health & Science University (OHSU)

☐ Janssen (Tremfya) funded trial at OHSU

☐ Novartis Pharmaceuticals (Cosentyx) supported Mayo Clinic

The over-arching strategy for the past six years has been to find a rare disease advocacy organization with a longer stick.

☐ National Organization of Rare Disorders

☐ Genetic and Rare Diseases (GARD) Information Center

☐ International Alliance of Dermatology Patient Organizations

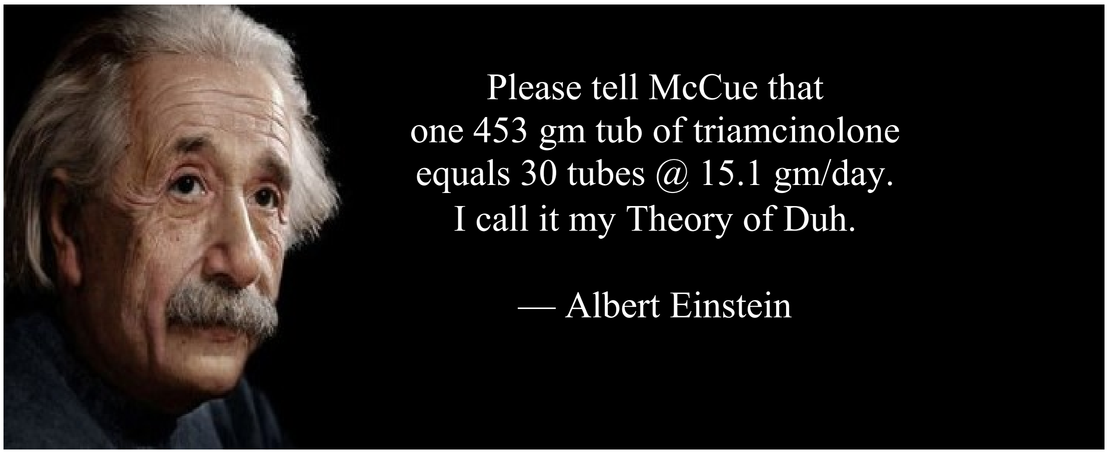
The path forward goal is to stay engaged.

objective
vs.
subjective

142. Signs and Symptoms

The terms “sign” and “symptom” are not redundant.

- ☐ A SIGN indicates a medical condition that others, including healthcare professionals, can observe objectively.
- ☐ In contrast, A SYMPTOM is subjective information that the PRP patient shares with the healthcare provider, such as pain, itching, and fatigue.
- ☐ Signs and symptoms are how we know we are healing or not.



Please tell McCue that
one 453 gm tub of triamcinolone
equals 30 tubes @ 15.1 gm/day.
I call it my Theory of Duh.

— Albert Einstein

143. Slathering

In December 2012, I was prescribed Desonide® for my face and ears, Clobetasol® for my palms and soles, and triamcinolone for everything else. When the nurse handed me a tiny tube of triamcinolone (the size of my little finger), I said, “You’ve got to be kidding. I’m 6’3”, and this tiny tube is not going to cut it.”

After a brief consultation with Dr. Dominguez, the nurse handed me a 453 gm tub. I responded with a “That’s more like it,” and a Thank you.” It was my first act of self-advocacy.

That evening, I used our kitchen scale to extract exactly 15.1 gms from the tub, place it on a cocktail plate and stretch it to every one of my nooks and crannies — every night for six months. I called it slathering.

I didn’t realize a 453 gm tub is the same as 30 tubes. Duh.

144. Sleep Schedule

Getting 7 to 9 hours of sleep each night is essential, but that's quite challenging for a PRP patient. Establishing a consistent sleep schedule seems impossible, but you must try.

- ☐ Most of the PRPers go to bed whenever they are tired. We wake up at all hours. We rarely feel rested. We need to schedule sleep.
- ☐ A fixed schedule can make it easier for us to cope with the daily stress of PRP. It's all about consistency.
- ☐ Make sleep a priority. PRP takes control and wreaks havoc on our sleep. Sometimes, we're too tired to sleep and to wake up.
- ☐ Make a commitment to sleep. I committed to 8 hours (11PM to 7 AM)
- ☐ Create a wind-down routine. Last call for water: 9 PM; take 30-min bath and 30-min of topicals: 10 PM and evening cocktail of Ambien®, Tylenol® and hydroxyzine: 11 PM.
- ☐ Use bedtime reminders and wake-up alarms. Don't oversleep.

And when you sleep through the night, register your "good night sleep" as a success and celebrate tis Healing Milestone.

145. Stigmatization

The stigma of PRP can lead to feelings of low self-esteem, anxiety, and depression. If these feelings linger, think about talking to a mental health professional.

People not understanding your version of PRP and reacting negatively can lead to feelings of low self-esteem, anxiety, and depression. It takes knowledge, effort, and support from others.

- ☐ Learn all you can about PRP.
- ☐ Educate your loved ones.
- ☐ Explain how you suffer,
- ☐ You are the subject matter expert of your version. Tell your story.
- ☐ YOU are the one who ends the stigma of PRP.

146. Stress & Triggers

Ask for a show of hands. How would a PRPer answer this True or False question?

Stress triggered my PRP.

FALSE: If I got a dollar for every True answer, I could buy a printer and a year's supply of ink. The month before my onset, I had three planets in stress.

- ☐ An effort to retire from retirement. I really wanted the job. Lots of stress.
- ☐ Splashed a little stain remover on my face while helping my daughter refinish a bookcase.
- ☐ An encounter ex-wife (#2). My stress registered on the Richter Scale

All I can say as an adult is that sh*t happens. If stress triggers PRP there wouldn't be a rare disease.

Let's try another True or False question?

Stress can trigger a flare or cause a setback

TRUE: We know from the shared experiences of the PRP global community that stress is not suitable for PRPers. *Stress = trigger = flare*

147. Swollen legs & feet

For many, the PRP journey routinely includes complaints about swollen legs and feet. PRPers are eager to share what works for most of us.

- ☐ Put your legs on pillows to raise them above your heart while lying down
- ☐ Exercise your legs. This helps pump fluid from your legs back to your heart.
- ☐ Follow a low-salt diet, which may help reduce fluid buildup and swelling.
- ☐ Wear compression socks
- ☐ When traveling, take breaks often to stand up and move around.
- ☐ Lose weight if you need to.

148. Sunlight

Many PRPers can't tolerate direct sunlight. We hear comments like, "The sun burns right through my clothes" and "Don't do it!"

For others, however, access to sunlight reduces stress and improves quality of life. It's probably what you do outdoors with family in the light of day.

Let's just agree that it's a time for caution. There is general agreement among PRPers to:

☐ Wear long-sleeve UV protective clothes and a wide-brim hat."

☐ Stay in the shade or under an umbrella

☐ Drink lots of liquids.

149. Synonyms

A synonym is a word or phrase that means exactly or nearly the same as another word or phrase in the same language. The following are words provided by PRPers that are decidedly negative, linking with pityriasis rubra pilaris.

- ☐ Alien
- ☐ Atrocious
- ☐ Bizarre
- ☐ Consuming
- ☐ Costly
- ☐ Cruel
- ☐ Debilitating
- ☐ Depressing
- ☐ Despicable
- ☐ Destructive
- ☐ Devastating
- ☐ Disgusting
- ☐ Disruptive
- ☐ Evil
- ☐ Exasperating
- ☐ Exhausting
- ☐ Frustrating
- ☐ Game-changing
- ☐ Gross
- ☐ Gruesome
- ☐ Heartless
- ☐ Horrible
- ☐ Horrific
- ☐ Incessant
- ☐ Itchy
- ☐ Life-sapping
- ☐ Maddening
- ☐ Menacing
- ☐ Merciless
- ☐ Monstrous
- ☐ Mysterious
- ☐ Nightmarish
- ☐ Overwhelming
- ☐ Painful
- ☐ Putrid
- ☐ Punishing
- ☐ Relentless
- ☐ Ruthless
- ☐ Torturous
- ☐ Traumatic
- ☐ Ugly
- ☐ Unpredictable
- ☐ Unrelenting
- ☐ Unwelcome
- ☐ Wretched

150. Swimming

With most of his body affected, a PRPer was unwilling to keep swimming in the chlorinated public pool, thinking the chlorine would irritate his already irritated skin. However, he found that open water swimming in a lake would be fine.

He wore a bright “rash guard” shirt and neon cap and pulled an orange flotation device to have visibility for boaters.

After exiting the water, he applied coconut oil and Vaseline to his skin.

“The past few times I have gone swimming,” he posted, “I have thought about what one person recently stated to our PRP Support Group: ‘I have PRP, but PRP doesn’t have me.’

And he added, “Those are inspiring words for us as we try to live normal lives. The best we can, under the circumstances of our skin disease.”

151. Tattoos & Body Art

Over the years, the subject of tattoos has found its way into the Land of Chat.

- Some PRPers complain about fading, especially red. Some complain that the writing is unreadable. True to form, responses included those who reported that their “tats” were perfectly fine.“
- Some PRPs want to know if getting a tattoo while in the throes of PRP is advisable. That usually brings out the Are-You-Kidding-Me folks. As one member commented, “If I ever got a tattoo, I’d be expecting to trigger the PRP demons just lurking below. “
- While I would never get a tattoo, I enjoyed photos shared by a PRPer in the UK. His body art was tasteful and added his humanity to otherwise flaky arms.

152. Take Command

Once I was officially diagnosed (nearly four months after onset) and Dr. Arturo Dominguez at the University of Texas Southwestern had initiated my treatment plan, I focused on problems I thought could be solved by ME.

I saw my dermatologist as a resource. However, it was up to me to find my way day by day. I knew I had to take charge of my day-to-day recovery.

- ☐ Accept the fact that you are on this journey for the long haul.
- ☐ Identify problems you think YOU can solve and then solve them.
- ☐ Find strength within the PRP Facebook Support Group. The posts and comments of kindred spirits will help you document ways to cope 24/7.

153. Translation Button

The NORD PRP Report was revised in 2017 by PRP patients and reviewed by a team of healthcare professionals, including a past president of the American Academy of Dermatologists, the Chair of the Dermatology Department at Thomas Jefferson University, and current Vice Chair of the Dermatology Department at Oregon Health and Science University.

A“ Translation Version” of the NORD PRP Report was added to the *PRP Survival Guide* to accommodate PRP patients and their caregivers who prefer languages other than English. This “Translation Version” provides easy access to Google Translate.

154. Travel & Vacations

The onset of PRP does not consider vacations booked a year in advance, weddings, and the need for grandparents to see grandchildren. For many, the cancellation of travel can lead to anxiety and disappointment. PRPers want to hear that it is OK to travel. Fortunately, there are countless stories where advance planning made a difference. The mantra is to Manage your expectations.

Build a checklist that includes UV clothing, compression socks, and nitrile gloves. Elevate feet, watch the heat, drink lots of water and moisturize as usual. Depending on location, watch the heat (you may not be sweating). Drink lots of water and moisturize. Take pictures. You will have a story to share.

155. How many types of PRP?

The PRP patient population can be divided into two categories based on the patient's age when the symptoms of PRP first appear.

You’re either adult onset (18 years or older) or juvenile-onset (onset less than 18 years). That’s usually enough to satisfy the curiosity of friends and family. But there’s more.

- ☐ Both adult and juvenile-onset have a classical and atypical version. The latter is considered more long-term or chronic. That’s FOUR more.
- ☐ Juvenile onset PRP has one additional version. Circumscribed juvenile-onset occurs in pre-pubertal children and is usually confined to palms, soles, knees and elbows. That’s another ONE.
- ☐ The last type of PRP is called HIV-Associated. The diagnosis is infrequent and statistically excluded from patient population estimates. And that brings us to another SIX…in my humble opinion.

156. Unicorns

Why should the mythical unicorn be the official mascot of the Global PRP Community? It's all about the horn.

☐ The unicorn is a legendary creature known to possess magical abilities.

☐ European cultures hoped to relieve stomach problems by drinking from its horn.

☐ Moreover, the mythical properties of the horn had served as an antidote to poison."

☐ Given that PRP has no guaranteed cure and what works for one may not work for all, perhaps we should be looking for our unicorn. Unfortunately, the FDA would probably approve UNICORN® for psoriasis but not for PRP.157.

157. Vacuum Cleaners, Basic

Over the years, I have heard tales of woe that describe how vacuum cleaners purchased pre-onset have succumbed to the rigors of “skin drifts.” Some died a hero’s death; others just whimpered.

And don’t forget handheld portables. I recall a PRP family living in a two-story house with a Dyson on the first floor and two Dustbusters® strategically positioned downstairs and upstairs.

PRP families have treated their vacuum cleaners as valued family members. Like Robbie, these “flake-sucking giants” often earned nicknames of endearment and strength.

There once was a vacuum named Robbie,
Who sucked up flakes from my body.
The whole family cried,
The day that he died,
'Cuz my flaking became a tsunami.

158. Venting

Venting is the spontaneous effort by a PRP patient in pain or a caregiver helping a PRP patient in distress to elicit an immediate and empathetic response from one or more kindred spirits in the Land of Chat.

☐ It doesn't matter whether that pain is real or imagined.

☐ While venting is an important part of the healing process for every PRPer, it should not be shared with passersby at Walmart or Target.

☐ When venting to kindred spirits, the outpouring of support is predictable and heartfelt.

☐ Expect cyber hugs and words of encouragement from seasoned travelers.

☐ Venting should not be confused with *Howling,* as in *Howling at the Moon*. While venting is the solicitation of a hug, howling is an effort to resolve a specific problem.

159. Weight gain and loss

During the three months when I was misdiagnosed with seborrheic dermatitis, I gained 35 pounds. Once I was declared a PRPer and a better treatment plan was initiated, I started losing weight very slowly.

Based on feedback from kindred spirits, most PRPers seem to lose 10-20 pounds and then deal with an ebb, flow and weight. The most significant reported weight loss was shared by a Canadian in 2015.

Discuss the topic of weight *and your weight* with your dermatologist as it relates to PRP specifically.

☐ How does PRP impact my weight?

☐ What is it about PRP that causes weight gain or loss?

☐ Should I have a weight goal? Think Healing Milestones!

Remember that PRP is predictably unpredictable. Your version is, too.

160. Workplace

Over the past eight years, I have heard hundreds of unique PRP workplace stories. There are two basic storylines:

- ☐ Patients who no longer have the physical capacity to perform the duties for which they were hired. For some, the onset of PRP is so ferocious and relentless that working is simply not a viable option.
- ☐ Patients who believe they can perform must convince a third party, e.g., employer and/or supervisor. Others, for whatever reason, usually financial, make a commitment to stay on the job. The loss of income can be catastrophic.

Finding kindred spirits who have wrestled with whether to work or not is your best strategy. Sometimes, the answer is “as long as I can.”

161. Why me?

A newly diagnosed PRP patient will ask: *Why me?* It's a defensive question. The simple answer: you were one in 400,000. Regardless of the trigger, you are still the one in 400,000. Don't waste time on Defense. It's time to switch to offense.

☐ When your dermatologist prescribes a drug, ask Why? Asking the question can be as important as the answer you get and write down. Questions don't threaten the authority of any healthcare professional. Instead, they demonstrate that you are engaged in your own treatment.

☐ In fact, you should have five questions ready for your dermatologist before every office visit. Train your dermatologist to expect a Q&A as part of every office visit. When your dermatologist sees that you are serious about the questions you ask about your office visit, their answers and your questions will get better.

162. Zits

(Acne) vs. Pityriasis Rubra Pilaris

Editor’s Note: When I reached the PRP Glossary's end, I realized there was no word, term or jargon beginning with the letter “Z.” Balderdash, I thought. Somehow, the idea of a glossary that goes from A to W wasn’t going to cut it., What follows is a comparison between Zits and PRP.

- ☐ Zits are also known as acne. PRP is also known as the *Red Menance*
- ☐ Zits (acne) is the most common skin condition in the United States, affecting up to 50 million Americans annually. With an estimated patient population 850 in the U.S., I think we qualify nas one of the least common.
- ☐ Every dermatologist will diagnose and treat zits (acne). It has been estimated that only one in 20 dermatologists will ever treat a PRP patient.
- ☐ Zits (acne) begins in adolescence. In contrast, PRP can start as early as birth. The PRP Global Database documents PRP onset at 91 for two PRPers.
- ☐ When someone has zits, the hair and “stuff” stick together inside the pore. This prevents some “stuff” from shedding and keeps other “stuff” from reaching the skin's surface. With PRP, nothing stops the skin (our “stuff”) from shedding like a winter blizzard in the Rockies.
- ☐ Zits can impair skin, socialization and mental wellness. PRP impairs the same trio plus another nine: energy, mobility, dexterity, vision, hearing, sleep, employment, school, and financials
- ☐ Zits (acne) don’t destroy vacuum cleaners. PRP can destroy vacuum cleaners and washing machines.

Peer Review Ponderings

SECTION 6

PATIENT SUPPORT

163. It began with three unanswered questions.

In December 2012, few days after my PRP diagnosis, I became an enthusiastic subscriber to the 800-member, email-based PRP support group. They were the only game in town. I devoured their website and read hundreds of emails from PRPers like me. I no longer felt alone. I had found a community of kindred spirits.

Who lives in Texas?

I asked if any members were from North Texas. I was eager to meet PRPers "in the flesh," even if the flesh was red and shedding like mine. No one responded.

Who has type 2 diabetes?

In early January 2013, I created another "thread" to learn about PRP and diabetes. I had been diagnosed with Type 2 diabetes in 2004, No response.

How long did it take to get your PRP diagnosis?

In mid-January, I wanted to know if my four-month diagnostic odyssey, which included two dermatologists, six days in a hospital, and four biopsies, was typical. The lackluster response was disappointing.

A few weeks later, I received an email from the National Organization of Rare Disorders inviting me to participate in their annual Rare Disease Day 2013. What could I do to support Rare Disease Day? On February 6, 2013 I looked up available domain names and registered prpAlliance.com and became a self-proclaimed PRP Advocate. Another PRP journey had begun.

164. PRP Alliance

The PRP Alliance has served the PRP global community since 2013. Our community has grown from less than 120 to over 2,100 in the past ten years.

- Every PRP patient, young or old, begins their PRP journey at the same starting point — the first milestone: onset. We had no idea what was happening to our skin or the journey we were about to embark on.
- We reached the second milestone, diagnosis, only to discover a lack of PRP savviness among healthcare professionals. For too many of us, we were only told how to spell *pityriasis rubra pilaris* and then referred to the internet (aka Dr. Google).
- Once we had digested our PRP diagnosis, our focus shifted to treatment, where we learned: “What works for one doesn’t work for all.” Arrrgh!
- And then there is the endgame. While most of us set our sights on remission (no meds, no symptoms), others looked “down the road” to a more distant outcome: long-term management.

Along the way, we were continually reminded that *pityriasis rubra pilaris* is an ultra-rare disease, and PRP-savvy dermatologists can be just as rare. The PRP Alliance has functioned as the advocate for the PRP global community.

165. PRP Global Database

Our mission is to advocate for the timely and accurate diagnosis of *pityriasis rubra pilaris* (PRP), implement more effective and accessible treatment options, and increase PRP-specific research. .

The PRP Global Database has two functions: communication and research

Communication — To build awareness

- ☐ We use the database to reach the PRP global community, of which 1,404 (76%) live in the U.S. and 443 (24%) live beyond the U.S. borders. Core data includes name, location, email address, onset date, onset age, and current status. When the need arises, we can get the word out.

Research — To rise above the noise level

The PRP Global Database was created as a tool for PRP research on two fronts: in-house and third-party.

- ☐ In-house research: The PRP Alliance has contracted with Constant Contact since 2013 to support surveys on topics from the role biopsies play, including the diagnosis of PRP, the impact of COVID-19 on PRP, the efficacy of acitretin, methotrexate and biological in the treatment of PRP.
- ☐ Third-party research: Over the years, the PRP Alliance has supported the research efforts undertaken by Oregon Health and Science University, Thomas Jefferson University, UCLA and USC

We will continue to add PRP patients to the PRP Global Database.

166. PRP Global Community

When I was diagnosed in 2012, I became a subscriber to an email list-serve of kindred spirits. But a list-serve does not have a global community.

I already knew about rare disorders. It didn't take me long to find my way back to the National Organization of Rare Disorders.

In 1976 and 1983, two of my children (Marisa and Matt) were diagnosed with a rare, inborn error called phenylketonuria. The diagnosis is based on a simple blood test performed one day after their birth. It's too bad PRP doesn't have one of those.

If untreated, phenylketonuria can lead to severe brain damage, profound intellectual disabilities, behavioral symptoms, or seizures.

Naturally, I joined the PKU network, became an expert in PKU, attended conferences and benefited from a global community of kindred spirits.

There are fewer than 20,000 PKU cases in the U.S. every year versus about 200 PRP cases annually.

I learned with PKU that the support of a community of kindred spirits is essential for successful outcomes. When you are holding a newly diagnosed infant in your arms and see other parents with healthy PKU kids — older kids, you know you can do it, too.

Today, we are a global community of kindred spirits. We are not alone. We are on this journey together.

167. PRP Survival Guide

The *PRP Survival Guide* was designed to be a repository of experiences and insights that PRP patients and their caregivers share. The PRP global community has a wealth of practical knowledge ready for harvest.

BASICS

We begin with the basics. Whether you are a newly diagnosed patient, “seasoned traveler,” or supportive caregiver (spouse, partner, parent, member of the family or friend), the “basic questions” are at the core of understanding pityriasis rubra pilaris and the road ahead.

TREATMENT

From the patient's perspective, there are three observations to be made about treating PRP:

☐ Each PRP journey is unique.
☐ You are not alone. There is a global PRP community.
☐ What works for one doesn’t work for all.

DAILY LIFE

Over 100 topics, ranging from alcohol to the workplace, reflect the collective insights and observations of the PRP global community.

SELF-ADVOCACY

The *PRP Survival Guide* will spotlight self-advocacy in November 2024 and include the PRP version of STANDING UP For Your Health, an adaptation of a brochure developed by the Aplastic Anemia & MDS International Foundation.

168. PRP Support Group

Post a question, and comments flood in. Need a hug — the huggers respond. Need a friend — a community of fellow travelers is ready to lend an ear or a shoulder. Need information — there are over 2,100 members available to share their PRP-related insights and experiences.

A MEMBER of the PRP Support Group can be one of the following:

☐ A patient diagnosed with PRP

☐ Caregiver/supporter of a patient diagnosed with PRP

☐ The patient's caregiver is *awaiting confirmation* of a PRP diagnosis

☐ Dermatologists and other healthcare providers who treat PRPers

Jean-Luc Deslauriers, a Canadian, inspired the original PRP (email-based) support group in 2003. Using list-serve technology, members traded an average of 150 emails each month. Membership has ebbed and flowed for over a decade. Their website was suspended in 2016.

In 2008, another Canadian and PRP sufferer, Jonah Grant-Scarfe, recognized the value of Facebook as an alternative to trading emails. In September 2013, Jonah made a giant leap of faith and converted the PRP (Facebook) Support Group from PUBLIC to PRIVATE. We became MEMBERS ONLY, and comments were measured in the thousands. Today, over 2,100 seasoned "PRP Facebookers" understand the importance of a CLOSED (private) Facebook group.

The late Tierney Ratti (1961-2020) still is the gold standard of caring and sharing. Her journey began in 1972 at the age of 11 with the onset of *pityriasis rubra pilaris*. She was one of the original PRPers who traded emails on AOL in 1997 — You've Got Mail. In 2013, she became the administrator of the Facebook-based PRP Support Group.

169. PRP Research & Advocacy — 2024

The first utterance of “If God is willing and the Creek doesn't rise was made by *Benjamin Hawkins* of North Carolina, a Continental Congress senator (circa 1781-1787) when he was summoned to the nation's capital.

I have learned over the past decade that those words apply to my goals and aspirations as a PRP advocate. That said, here are some research goals for 2024

In-house PRP research

- ☐ *What Works Survey* — We need to quantify the number of PRP patients who have taken or are currently taking retinoids, immunosuppresents and biologcals. What was the efficacy of each? We also need to survey topo
- ☐ *Onset to Diagnosis Survey* — A new crop of PRP patients has joined the PRP Support Group over the past three years and missed the comprehensive Onset to Diagnosis Survey. This is an opportunity for PRPers to document the start of their journey,
- ☐ *Quality of Life Survey* — This will be patterned after research by the International Alliance of Dermatology Patient Organizations and Oregon Health & Science Univesity.

Third-party research

- ☐ An effort will be made during 2024 to support PRP researchers from teaching hospitals in the U.S. to conduct trials of specific biologicals. We will promote the approach made successful by Dr. Teri Greiling, Vice Chair of the Dermatology Department at Oregin Health and Science University is their research of Taltz (Lilly) snd Tremfya (Janssen).

International Alliance of
Dermatology Patient
Organizations

170. PRP and GlobalSkin

In June 2015, I traveled to Vancouver, Canada, to meet with leaders of over 60 global dermatology patient organizations. For several days before the event's official start, I worked closely with seven other patient leaders — two from Canada, two from the UK, and one from France, Nigeria and Pennsylvania. Our goal was to discuss the creation of an international alliance and how best to present the idea to our fellow delegates.

Early on the first day, I remember seeing a chart from the World Health Organization. It was labeled *Mortality and burden of disease* and listed 25 causes of mortality and burden. The first two causes on the list were cardiovascular diseases (#1) and cancers (#2). That sounded reasonable. The last two causes on the list were conflict/terrorism (#24) and natural disasters (#25). OK, I thought.

Then, I finally found the listing for skin diseases (#19).

It was then explained that the DALY chart is based on a formula that allows policymakers and experts to evaluate a particular disease's burden on a given population and recommend appropriate healthcare policy improvements.

Said another way, healthcare policymakers use the DALY analysis to confirm that the relative impact of skin conditions is considerably lower than most other diseases…or at least 18 others.

Fortunately, by week’s end, those in attendance unanimously signed the Vancouver Resolution, set in motion the creation of the International Alliance of Dermatology Patient Organizations (IADPO), 256 patient organizations representing 68 countries and 57 skin disease ares worldwide.

Peer Review Ponderings

EPILOGUE

171. THE ROAD AHEAD

I saw a spot, just a little dime,
I thought would vanish in a little time.
But the spot got bigger instead of less;
The red got redder to my distress.

By the time I saw a medical pro,
The spot had continued to grow and grow.
It was now a bar running down my face,
And heading elsewhere at too fast a pace.

Don't be alarmed," my doctor said,
"I'll give you meds to vanquish the red."
So I took her meds, and to my dismay,
The red got redder with each passing day.

My eyes were all crusty and blurry at times,
My skin was awash with a thousand red dimes.
Small like the first one, but as time surely passed,
They blended together in one solid red mass.

“By the end of six weeks, I was totally red,
And to make matters worse, I started to shed.
No matter where I decided to go,
I always left a lot of skin snow

I wished I was small and not six-foot-three,
Weighing two-forty, there’s plenty of me.
So shedding is quite an embarrassing hell,
‘Cuz there’s a lot of glitter on this big Tinker Bell.

Months had gone by, and I was in a bad way,
Nothing was working; I was itchy all day.
My get up and go had got up and went;
It hurt when I walked, and it hurt when I bent.

The dosage of meds had reached such a height,
Hallucinations appeared in the still of the night.
A humongous duck and tropical fish.
An hour of that prompted only one wish…

But the local ER wasn’t ready for me,
For no one knew anything about PRP.
In fact, I was there for nearly a week,
And the word “PRP” no doctor would speak.

When I was discharged, a new doctor I found,
Well-trained in “skin stuff,” with a little bloodhound
A piece of my skin he sent to a lab,
That confirmed PRP was the problem I had.

On Thanksgiving Day a decade ago,
There were so many answers I wanted to know.
Questions to ask and fears to allay.
And somehow manage
to fight one more day.

‘Twas the night before Christmas, I remember it well.
There was no sign of “Merry” as I drew closer to hell.
My feet were encased with leather-like skin.
That cracked and bled where my real skin had been.

My scalp was a snowstorm, a blizzard at least,
A savage attack by a titanic snow beast.
Some flakes were so big they bounced, and what's more,
They formed giant drifts on my dark hardwood floors.

The previous Christmas, 12 years ago,
The Yuletide Spirit had a radiant glow.
But the following year, a shroud had descended,
For our family reunion, PRP had upended

As their "Papa Bill," I glowed a bright red,
From the end of my toes to the face on my head.
I was "Tinker Bill," to my grandkids 'delight,
As my magical fairy dust spiraled in flight.

The children thought I was a holiday ploy.
To prompt a smile from three girls and a boy.
So I mustered a hearty "Ho Ho Ho,"
To disguise my pain so they would not know.

But to their parents, my kids, I was not at my best,
So they asked lots of questions — A "Papa Bill Test."
By the time I was done, THEY knew PRP,
And the challenges ahead for Heather and me.

At that very moment, I felt terrible despair,
And thought that "next year" I might not even be there.
With a body consumed by a prickly heat,
From the scalp on my head to my swollen red feet.

But two weeks later, my feet were all cleared;
The best they had been since the "Red Menace" appeared.
By the very next Christmas, I could make the admission,
To those who cared: "I'm nearing remission."

As I look back on those challenging years,
I remember the pain and remember the fears.
But how great are the stories that all of us tell,
Of how we survived our own version of hell.

So, whether you are "active" and fighting the fight,
On the "Road to Remission," with some end in sight,
Or in full "Remission" and just looking back;
We all are together — we're on the same track.

My road to remission was remarkably fast,
My meds and my ointments are now in my past.
But I know there's a chance that it may return,
With skin that will flake and skin that will burn.

Some reach remission and bid "Adieu",
Others will ask, "What more can I do?"
To the latter who stay to reach out and care.
It's far better for all that you stay here and share.

But I'm better off than a decade ago,
I've been down that road; I know where to go.
To the people I've met who have journeyed with me,
I was never alone, for I journeyed with 'Thee".


THANK YOU!

ACKNOWLEDGEMENTS

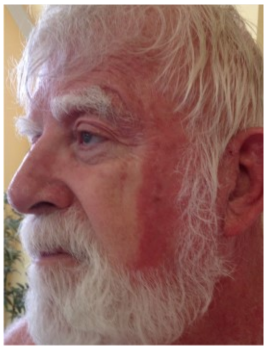
The 30-second Roadmap is dedicated to the PRP global community — past, present and future.

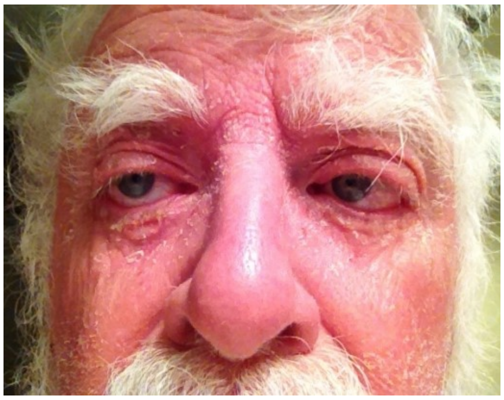
- ☐ **Jean-Luc Deslauriers** (1948-2003) was a Canadian and founder of the email-based PRP Support Group (1997-2016).
- ☐ **Jonah Grant-Scarf** (Canada) recognized the potential of Facebook and created a “public” group on June 10, 2008. Five years later, it became a Closed/member-only group.
- ☐ **Tierney Lynch Ratti** (1959-2020, Virginia) became the PRP Facebook group administrator in November 2013 when membership was less than 120.
- ☐ **Ginny Maxwell** represented the PRP global community meeting members of Congress on Capital Hill in Washington.
- ☐ **Christine Günther** (Switzerland) stepped up as a PRP advocate in Zurich, Switzerland speaking to over 50 dermatologists about Juvenile Onset PRP.
- ☐ **Jan & Malcolm Tennant** (New Jersey) have repeatedly ridden their white horses to help keep the PRP Alliance lights shining brightly.
- ☐ **Carol Terry** (England) for her Tierney-like commitment to caring and sharing.
- ☐ **Murray Rose** (British Columbia, Canada) for his support as a co-administrator of our Facebook Group.
- ☐ **Christopher Canning** (Ontario, Canada) for his support as a co-administrator of our Facebook Group and especially for his weekly New Member Welcome.
- ☐ The 125 PRP patients and their caregivers whose financial support has made the 60-Second PRP Roadmap, Peer Review Workbook possible.

ABOUT THE AUTHOR

- ☐ **Name**: Bill McCue
- ☐ **Residence**: Plano, Texas
- ☐ **Current age**: 77
- ☐ **Height**: 6’ 2”, **Weight**: 218 lbs
- ☐ **Onset date**: August 8, 2012
- ☐ **Onset age**: 66
- ☐ **First sighting**: red spot the size of a dime, left temple
- ☐ **Differential diagnosis**: seborrheic dermatitis (1st dermatologist)
- ☐ **Biopsies**: 3 inconclusive, 1 supported PRP
- ☐ **Diagnosis date**: Novermber 27, 2012
- ☐ **Diagnosing dermatologist**: Michael Golden, MD (2nd dermatologist)
- ☐ **Confirming dermatopathologist**: Cockerell Dermatopathology
- ☐ **Treated by**: Arturo Dominguez, MD, University of Texas Southwestern, Dallas
- ☐ **PRP Treatment protocol**:
  - Acitretin (12-month regimin, 25mg to 50mg daily for 12 months)
  - Desonide (face/ears)
  - Acitretin (soles /palms)
  - Triamcinolone (remaining nooks and crannies)
- ☐ **Comorbidities**: Type 2 diabetes
- ☐ **Allergies**: penicillin
- ☐ **Current status**: Full remission (no symptoms/no meds)
- ☐ **Date of remission**: April 8, 2014
- ☐ **Duration of PRP journey**: 20 months
- ☐ **PRP Advocacy**
  - Founder/president: PRP Alliance
  - Founding Board Member: International Alliance of Dermatology Patient Organizations — GlobalSkin
  - Administrator: Facebook-based PRP Support Group
  - Database manager: PRP Global Database

My father was an optomistic fatalist. He would say, ”Whatever’s gonna happen is gonna happen — but it will turm out good in the end.” At the beginning of my PRP journey there was certainly no “good” to be seen. However, once I had my PRP diagnosis and found my PRP family of kindred spirits, I knew it was going to turn out “good in the end”. And it did.





APPENDICIES

175. Appendix A: 2023 PRP Census
176. Appendix B: Coalition of PRP Parents & Kids
177. Appendix C: New Member Orientation
178. Appendix D: Rare Disease Caregiving
179. Appendix E: STANDING UP for your health: PRP Edition
180. Appendix F: Finding health information on the internet
181. Appendix G: Evaluating health information on the internet
182. Appendix H: Disability Claim Strategies

Appendix A

2023 PRP Global Census

A decade of data

On November 1, 2013, the first PRP Global Community Census was emailed to over 1,500 PRP patients/caregivers. At that time, we were all members of the now-disbanded, email-based PRP Support Group. The list had been harvested from an archive of over 29,000 emails from 1997.

The datapoints included patient/caregiver names, email addresses, onset ages, onset dates, dates of diagnosis, current statuses, and the names of PRP-savvy dermatologists, and the role played by biopsies in the original diagnosis and misdiagnoses.

Almost immediately after I hit the send button, over 500 emails bounced back as undeliverable. While their harvested data provided information, the PRP patients and caregivers with invalid emails were lost to the ages.

On the bright side, over 500 of the original 1,000 PRP patients/caregivers with a valid email address completed the census.

Our problem? We're just too damn rare!

Today, the PRP Global Database has archived nearly 5,000 PRP patient profiles of which 2,250 have valid email addresses—the world's largest PRP database.

The PRP global community is small by any standards. There are an estimated 3,000 skin disorders worldwide, of which 800 are considered rare skin diseases.

Based on a prevalence of one in 400,000 and a population of 340 million, there are an estimated 850 PRP patients currently dealing with the challenges of PRP.

The PRP global community exists in the shadow of psoriasis (8 million U.S. patients) and eczema (16.5 million U.S. patients). Member surveys show approved FDA treatment for psoriasis and eczema works for PRP patients. Without FDA approval for PRP, however, insurance providers will not cover the cost of treatment.

There is a reason only 100 drugs have been developed for treating rare skin diseases in the past 38 years. Our patient population is too small to justify the investment in research.

Repurposing

Collectively, the pharmaceutical companies spend billions to bring FDA-approved drugs to market and then invest additional billions in direct-to-consumer advertising. Pharma advertising in 2022 was estimated to be over $8 billion.

Drug repurposing is a process available to pharmaceutical companies to find new uses for existing FDA-approved drugs. The PRP global community may be positioned to advocate repurposing biologicals used in treating psoriasis and eczema for PRP patients. It is necessary to have a well-organized database to investigate potential treatments through drug repurposing for rare skin diseases.

Why is the PRP Global Database so important?

We can whet the appetites of PRP researchers worldwide who have relationships with pharmaceutical companies. In 2018, the PRP Alliance helped recruit participants for Dr. Teri Greiling, Vice Chair of the dermatology department at Oregon Health and Science University. She was researching the efficacy of Taltz® manufactured by Eli Lilly for treating PRP.

Two years later, we helped OHSU recruit participants in a similar study of Tremfya® (manufactured by Janssen) to treat PRP.

In 2022, we supported her Quality of Life research, which resulted in her publication in the *Clinical Journal of the European Academy of Dermatology and Venereology.*

The PRP Global Database allows us to do the heavy lifting of research, e.g., recruiting participants and helping researchers build cohorts.

What is PRP core data we seek to confirm?

For over 10 years, I have collected core data representing over 4,676 PRP patients. Today, the PRP Global Database maintains 1,674 "reachable" PRP patient profiles. CORE DATA includes the following:

☐ PRP patient's name: Bill McCue

☐ PRP location: Plano, TX, USA

☐ PRP patient's email address: papa.bill@mac.com

☐ PRP patient's onset age: 66

☐ PRP patient's onset date: August 8. 2012

☐ PRP patient's diagnosis date: November 27, 2012

☐ Current status of PRP patient: In remission as of April 2014

☐ PRP dermatologist: Dr. Arturo Dominguez, UT Southwestern, Dallas, TX

Can you imagine counting 6,438 jelly beans?

As of October 31, 2023, the PRP Global database maintains 1,853 PRP patient profiles. Each profile contains eight "core" datapoints. The more datapoints we confirm, the more valuable the data will be to PRP researchers. It's that simple. If PRP datapoints were jelly beans, our database would be missing 6,438.

☐ PRP patient's name: 1,853 datapoints, all confirmed

☐ PRP patient's location: 1,853 datapoints, all confirmed

☐ Valid email address: 1,853 datapoints, all confirmed

☐ PRP patient's onset date: missing 636

☐ PRP patient's age at onset: missing 717

☐ PRP patient's diagnosis date: missing 1,379

☐ PRP patient's current status: confirm 1,853

☐ PRP dermatologist contact info: confirm 1,853

☐ Can you imagine counting 6,438 jelly beans? It's a trick question. I'm counting all the jelly beans — you just need to count your five.

https://lp.constantcontactpages.com/sv/wFhTjMr/prpCensus

The PRP Alliance launched the first PRP Global Census on November 1, 2013. The PRP Global Database has grown steadily over the past 10 years. Today, the database maintains profiles of over 4,900 PRP patients, of whom 1,853 are part of the PRP global community.

If you have been diagnosed with pityriasis rubra pilaris or are the caregiver for a PRP patient, please complete the 2023 PRP Global Community Census. Follow the link below:

Peer Review Ponderings

Peer Review Ponderings

Appendix B

Coalition of Parents & Kids

Coalition of PRP Parents & Kids

Every so often I lament the fact that after (or in spite of) nine years of trying I have been unable to create or inspire a community of PRP parents within the PRP Facebook Community. That doesn't stop me from raising the issue again in hopes that a Coalition of PRP Parents & Kids might evolve. So here I go again.

The PRP Juvenile Onset Community

There is a limit to what we know about the PRP Juvenile Onset Community, a subset of the PRP Community Database.

Over the past ten years the PRP Global Database has been evolving. In the Spring of 2013 I began gathering the methodical review of over 29,000 archived by the email-based PRP Support Group. These were email messages shared by members during the period November 1997 and April 2014. As a subscriber I "harvested" what I considered to be "core data" and entered that "core data" into a secure and confidential database.

The PRP Global Database currently has 1,893 PRP patient profiles of which 201 have an onset age of less than 18. There are, however 759 PRP patient profile where the ONSET AGE datapoint is missing. How many PRPers diagnosed with Juvenile Onset PRP are yet-to-be identified.

A total of 34 PRP patients or parents have completed a PRP Global Census within the past year. Are you or your child on this list? If not, then please complete the PRP Global Census.

GO TO: https://lp.constantcontactpages.com/sv/wFhTjMr/prpCensus

The database has been undated to accommodate the following datapoints:

- ☐ Name of juvenile patient:
- ☐ Name of parent(s):
- ☐ Email address for primary contact:
- ☐ Location (city, state, country)
- ☐ Onset age:
- ☐ Onset date (mm/yyyy)
- ☐ Diagnosis date: (mm/yyyy)
- ☐ Current status (Remission Date if in remission)

JUVENILE ONSET OVERVIEW

The consensus among dermatologists is that 40% of all PRP cases are juvenile onset. Let’s take a closer look at the three types of Juvenile Onset PRP.

Classical Juvenile Onset PRP (Type 3)

- ☐ Usually occurs between the ages of 5 and 10
- ☐ Accounts for about 10 percent of all cases of PRP
- ☐ Remission can occur sooner than Classic Adult Onset, Type 1
- ☐ Average duration of Type 3 is one year
- ☐ Odds: One in 4 million

Circumscribed Juvenile Onset PRP (Type 4)

- ☐ Occurs in pre-pubertal children
- ☐ Usually confined to palms, soles, knees and elbows
- ☐ Accounts for about 25 percent of all cases of PRP
- ☐ Occurs in pre-pubertal children (age less than 14)
- ☐ Not a long-term affliction
- ☐ Odds: One in 1.6 million

Atypical Juvenile Onset PRP (Type 5)

- ☐ Occurs at birth or early in childhood, sometimes inherited
- ☐ Accounts for about five percent of all cases of PRP
- ☐ Most cases of “familial PRP” belong to this group.
- ☐ Runs a chronic, long-term duration
- ☐ Odds: One in 8 million.

WHAT DO WE ACTUALLY KNOW NOW?

When it comes to PRP patients who have been diagnosed with Juvenile Onset PRP (onset age less than 18), the number drops dramatically to 186. However, there is a glimmer of hope in the fact that 1,027 PRP patients in the PRP Community Database have yet to indicate whether they are Adult Onset or Juvenile Onset. There may be more “wee ones” already in the database. and more to be found. Here is some of what we know about PRP Parents and Kids:

In addition to the **ONSET DATE (mm/yyyy)**, we need to know Location and Onset Age

BY LOCATION

- ☐ USA (broken down by states):
- ☐ International (broken down by countries, states (Australia) or provinces (Canada):

ONSET AGE

- ☐ Less than 1: 46
- ☐ 1 to less than 2: 9
- ☐ 2 to less than 3: 8
- ☐ 3 to less than 4: 12
- ☐ 4 to less than 5: 16
- ☐ 5 to less than 6: 8
- ☐ 6 to less than 7: 13
- ☐ 7 to less than 8: 11
- ☐ 8 to less than 9: 4
- ☐ 9 to less than 10: 7
- ☐ !0 to less than 11: 3
- ☐ 11 to less than 12: 8
- ☐ 12 to less than 13: 6
- ☐ 13 to less than 14: 6
- ☐ 14 to less than 15: 5
- ☐ 15 to less than 16: 5
- ☐ 16 to less than 17: 11
- ☐ 17 to less than 18: 11

THE PATH FORWARD FOR PRP PARENTS & KIDS

Your guess is as good as mine. At a minimum, however, PRP parents should consider the following.

☐ Every PRP parent or adult diagnosed with juvenile onset PRP should complete the 2024 PRP Global Census.

☐ Schedule, organize and prepare for a web-based January 2023 Video Conference.

☐ **Establish a PRP Parent Coalition?**

☐ Plan and execute outreach to Society for Pediatric Dermatology (SPD)

☐ Plan and execute outreach to Pediatric Dermatology, the official journal of the SPD that is published six times a yaear.

☐ Plan and execute outreach effort to teaching hospitals with dermatology departments.

☐ Plan and execute separate outreach to dermatology clinics specializing in pediatric dermatology.

☐ Plan and execute separate outreach to the Society for Peciatric Dermatology

THE SOCIETY FOR
pediatric
dermatology
SINCE 1975

Peer Review Ponderings

Appendix C

New Member Orientation

Most of us begin our PRP journey with an “official” diagnosis of pityriasis rubra pilaris based on the clinical observations of a dermatologist supported by a biopsy and the findings of a dermatopathologist. Some members have a diagnosis without a supporting biopsy. And there are still others searching for an “official” diagnosis. ALL are kindred spirits and members our Land of Hope family.

First Things First

☐ Read the 60-Second PRP Roadmap from cover to cover.

☐ Take your time. Think of it more as a 2-Minute PRP Roadmap.

☐ Become familiar with the PRP Survival Guide. Understand the organization, e.g., BASICS, TREATMENT, DAILY LIFE and SELF-ADVOCACY. Know what’s where so you can find answers more quickly.

☐ Read the patient-friendly NORD PRP Report when time permits. While this overview was researched and written by PRP patients, it was reviewed by dermatologists and approved by the National Organization of Rare Disorders.

Scroll the posts

Scroll through the first 20 posts you encounter to “get the feel” for how PRP Facebookers share their respective PRP journeys.

If any topic piques your interest, click the icon for LIKE or LOVE. It’s a simple “Thank You” or a “Tip of the hat” to every author who took the time to post.

If you want to share a PRP-related insight or experience, then comment. Know that other readers will benefit from your sharing.

Author your own post

1. Any PRP-related topic is fair game. However, we avoid politics and topics that are not PRP-related.

2. Introduce yourself. The more you share, the more others care.

Find your “Sharing” comfort zone

1. The PRP Support Group is a CLOSED COMMUNITY. We can share information with members that would otherwise be kept private.

2. Whether you post or comment, the more you share, the more you learn.

3. Share your location. If you want to find kindred spirits near you, share your location. People don’t need your address, a postal code or a telephone number. They do need your country. If you live in the USA or Australia, then share your state. If you live in Canada, share the province. Everyone knows I live in Plano, Texas.

4. Share a photo. PRP patients and caregivers visit the Land of Hope tin search of answers. Sometimes, as the saying goes, “A picture is worth a thousand words.” Don’t be shy.

Searching the Land of Hope

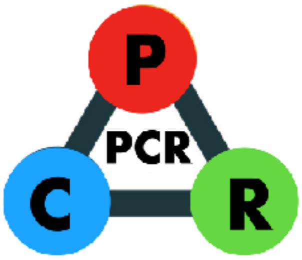
1. Find the search icon in the top right corner of the HOME page. CLICK the icon.
2. Enter a keyword. The inner workings of Facebook will first harvest posts wherever the keyword appears; The second harvest captures posts where the keyword appears as a comment. I'm obviously not shy.

Missing data

1. Why is data missing? As of January 2022, membership in the PRP Support Group required answers to three questions, e.g., role (patient, caregiver, supporter), location (country) and email address.
2. What's missing, e.g., onset date, onset age, and diagnosis date?
3. Please REPLY to this email and share the PRP patient's Onset Date, Onset Age and Diagnosis Date.

Path Forward

1. Jump in with both feet. This is not the time to watch from afar. Each month, 50 to 75 posts will appear. There's always something to learn.
2. GO TO:



   https://www.facebook.com/groups/15865278115

3. Engage! Seek out the insights and experiences of the PRP global community. The mantra is PCR — Post. Comment. React.

Why should I join the PRP Support Group?

Members of the Facebook-based PRP Support Group were asked this question The 11 responses sreflect the collective voice of the PRP global community.

1. To provide — and to receive — hope, encouragement, and emotional support. This is especially important in the absence of an existing support system.
2. To better understand pityriasis rubra pilaris through the collective, first-hand experiences of those afflicted.
3. To minimize loneliness and feelings of isolation. To know that we are not alone. We are a community of kindred spirits.
4. To effectively manage expectations and maximize hope. We want more than our fair share of good news and will work for it.
5. To lighten the burden placed on loving family members by sharing that burden with the PRP community.
6. To recognize that everyone has their own version of PRP and what works for one may not work for another. We can find effective ways to cope with pain, anxiety, stress, depression and frustration.
7. To promote a better understanding of our disease within the PRP community by sharing our personal experiences with treatments, dermatologists and other healthcare professionals.
8. To promote participation in bona fide research projects designed to improve the diagnosis, treatment and understanding of PRP by the medical community. We must find ways to enlighten dermatologists and other healthcare professionals.
9. To maintain a core value of acceptance, understanding and empathy.
10. To vent to each other when our pain is too great, our frustrations unbearable, and our fear overwhelming
11. To meet a fellow PRP face to face, in the flesh. Isn’t it about time we hugged?

Peer Review Ponderings

Appendix D

Rare Disease Caregiving in America

EXECUTIVE SUMMARY

This report outlines findings from the research study Rare Disease Caregiving in America funded by the National Alliance for Caregiving and Global Genes. It is based on 1,406 caregivers ages 18 or older living in the United States who provide care to a child or adult with a rare disease or condition from data collected during fall 2017.

Rare diseases or conditions are those that affect fewer than 200,000 people.[1] This study aims to describe the experiences and challenges of this targeted group of caregivers holistically across many diseases.

Although understanding the impact of rare diseases on the person with the rare disease is important and worthy of research, this study aims to examine the impact of rare diseases on the caregiver. It is important to note that all data presented herein are from the perspective and experience of the rare disease caregiver.

Approximately 25 to 30 million Americans currently have a rare disease or condition.[2] This deep-dive research into the lives of rare caregivers[3] – that is, American adults who provide care to an adult or child with a rare disease or condition – suggests rare disease has a broad and lasting impact on caregivers in daily life and longterm well-being.

Most rare caregivers provide care to a child under 18 (62%), with the average age of the care recipient – that is, the person who had a rare disease, condition, or illness – being 20.9 years old. Most rare caregivers are immediate relatives, with 59% caring for their child under 18, 17% caring for their adult child, and 14%

caring for a spouse or partner. Nearly all rare caregivers live in the same household as their care recipient (89%).

Rare caregivers tend to care for someone whose rare condition or disease is genetic (71%). However, the list of conditions for which care is provided in this study is over 400 unique conditions long (see Appendix B for a complete list of conditions, available online at www.caregiving.org/rare). Only 43% report a treatment available for the underlying disease or disorder, and most (82%) report that cures or therapies are used for symptom management rather than actual disease treatment.

Rare caregivers rely on doctors and medical professionals to help them and their care recipient through the experience of rare diseases. More than eight in ten rely on medical professionals for information (82%). However, adequate medical and support services can be challenging to access. Fewer than two in five rare caregivers feel their care recipient's local hospital can handle the underlying rare disease or condition (38%). Roughly one out of every three rare caregivers report having difficulty in accessing treatments or therapies for symptom management (34%) or treatment(s) for the underlying disease (31%).

Rare caregivers may need more support from the doctors and care professionals they interact with. Fewer than half (48%) have had a doctor, nurse, or social worker ask what was needed to provide care to the recipient, and just one in four have had these discussions about their own care needs.

This lack of outreach about rare caregiver needs may be a missed opportunity in that caregiving can become a role that fundamentally alters daily life for many years to come. Most rare caregivers have been providing care for a significant period, 8.9 years, on average – more than double the caregiving commitment found among caregivers more generally.[4] Rare caregiving is also time-intensive.

Rare caregivers of adults spend about 37 hours a week providing care on average – about 12 more hours a week than general caregivers. [5] Even more striking is the amount of care supplied by rare caregivers of a child: 53 hours a week, on average, compared to 30 hours for general child caregivers. [6]

Rare caregivers spend their time taking on various tasks for their care recipient, which translates into a high-burden[7] caregiving situation. Nearly all rare caregivers help their care recipient with at least one Instrumental Activity of Daily Living (IADL) task, such as housework, shopping, or transportation. Three in five rare caregivers (62%) help with at least one Activity of Daily Living (ADL). Two out of every three rare caregivers have a high caregiver burden on the Burden of Care Index (67%) – far higher than general caregivers. [8]

A defining feature of rare caregiving seems to be one of expertise. Rare caregivers have to function as nurses, performing medical/nursing tasks:[9] 84% help their care recipient with medical/nursing tasks. These tasks are a defining feature of rare caregiving: as rare caregivers face barriers to service support and care access, they are called upon to do these complex medical/nursing tasks for the duration of their role as caregivers. Most "become" teachers, educating health-care professionals about their care recipient's rare disease or condition (89%). Rare caregivers can also find themselves researching and accessing clinical trials. One in four report their care recipient has participated in a clinical trial. When that occurs, the rare caregiver often has to help with things like paperwork (77%), transportation (65%), trial response documentation (62%), and care coordination (59%).

Despite rare caregivers 'higher Burden of Care score versus general caregivers, they are no more likely to rely on paid help, with just one in three reporting the use of paid assistance or aides (33%). Support service use is also uncommon, with just 22% having used respite and 14% relying on an outside transportation service. This day-to-day responsibility means that other immediate and extended family members may be called upon to help provide care, with 59% reporting help from at least one unpaid caregiver. This domino-effect of care can extend to youth in the family, with 24% reporting their care recipient receives outstanding care from at least one youth under age 18.

Rare caregivers note the ripple effect of rare diseases on their family, with 74% struggling with a sense of loss for what their care recipient's life could have been and fewer than half feeling their role has had a positive impact on their family (44%). This ripple effect has some positive results, as most rare caregivers report pride in improving the understanding of their recipient's condition (94%) and feeling close to their care recipient (85%). More than half (56%) feel a sense of purpose due to their caregiving role.

The day-to-day impact for rare caregivers is pervasive in all aspects of their lives, with no areas untouched: the effects are seen in the workplace, school, and on their physical and emotional health. Two out of three rare caregivers have worked while providing care (65%), and nearly all report a bleed over into their work life, more so than general caregivers who work.[10] Almost all have gone in late or left work early to provide care (91%). One in ten rare caregivers is also a student, and, as with work, they often struggle to fulfill their school responsibilities.

Rare caregivers say providing care to their care recipient is emotionally stressful (67%), twice as high as that of general caregivers.[11] Four in ten report having fair or poor emotional or mental health (41%). To a lesser degree, rare caregiving can physically strain caregivers, with 37% reporting high physical strain. Those who do more hands-on care, like medical/nursing tasks and Activities of Daily Living, experience more significant physical stress. Rare caregiving takes a precise toll on the physical health of caregivers, with 30% rating their physical health as fair or poor and 58% finding it difficult to take care of their own health. The ripple effect of rare caregiving can extend past day-to-day tasks of providing care, the need for others in the family to pitch in, and the emotional and physical well-being of the caregiver. Rare caregiving can ultimately impact the finances of the rare caregiver and their family unit, and these financial impacts loom large. Half of rare caregivers face high levels of financial strain (51%), and three out of four worry about the ability of their own family or their care recipient's family to pay for care. Nearly all rare caregivers have experienced financial hardship because of their caregiving role (86%). Common areas of impact include cutting back on household spending (81%), not saving for long-term goals (62%), and using up personal savings (59%). Nearly one in ten have filed for bankruptcy (9%). Future planning is not the norm among rare caregivers: 35% have a contingency plan in place in case they cannot provide care, and only 29% have plans for their care recipient's future care (i.e., financial, health, and living arrangements). Rare caregiving impacts the caregiver's daily life, the family unit, their wellbeing, and their long-term finances, and the role of rare caregiver changes their social lives. About half of rare caregivers have difficulty maintaining friendships (51%), and 53% report feeling alone.

Peer Review Ponderings

Peer Review Ponderings

Appendix E

Self-Advocacy for Patients with PRP

STANDING UP for your health

SELF-ADVOCACY FOR PATIENTS WITH PRP

On June 3, 2015, the PRP Alliance participated in a conference call arranged by the National Organization of Rare Disorders (NORD). John Huber, executive director of the Plastic Anemia & MDS International Foundation, announced the completion of a five-year grant and shared their 62-page self-advocacy brochure written for patients with rare diseases.

The AAMDS self-advocacy guide outlines fundamental principles empowered patients with pityriasis rubra pilaris can use to advocate for their best healthcare.

Truth be told, I did not expect rank-and-file PRP patients and caregivers to read *STANDING UP for your health* from cover to cover and adopt all of their suggestions. However, I did recognize, “There’s gold in them dar’ hills”

STANDING UP has been reimagined to encourage PRP patients and their caregivers to fully participate in the treatment of PRP. It is not meant to be a substitute for the advice of your dermatologist or any other healthcare professional.

For a symbolic licensing fee of $1, the PRP Alliance was authorized that day to use *STANDING UP for your health* without restriction. A PDF was immediately posted online in the PRP Survival Guide, which has remained for over eight years.

It seemed proper and fitting for *STANDING-UP* to be reimagined as a collection of guidelines specifically tailored for PRP patients and their caregivers.

And so it is. Whether you are newly diagnosed or have been living with PRP for many years, there are practical strategies designed to help you stand up for your health

— Bill McCue, PRP Advocate.

What is Standing Up for Your Health?

This guide was initially developed to empower a large, often forgotten and underserved population: the 30 million Americans with rare diseases who face challenges in obtaining an accurate diagnosis and have difficulty finding medical experts with experience managing their rare diseases.

STANDING UP for Your Health means being a strong self-advocate and an empowered patient. It means taking a more active role in your care to ensure you or your loved one receives the best care and treatment. Assigned patients learn all they can about PRP and their treatment options to fully participate in their care.

Initially created by the Aplastic Anemia & MDS International Foundation, this program was developed to discover the steps empowered patients and strong self-advocates take to enable them to receive high-quality care.

To bring you the most up-to-date information, AAMDS consulted with medical experts and interviewed patients with rare diseases about the skills they use as they effectively navigate the healthcare system. They read the latest research and talked with physicians, nurse specialists, patients, and their families to get their perspectives on the skills needed for good health advocacy. Then, they compiled what they had learned into logical steps any rare disease patient can take toward standing up for their health.

This booklet includes tools, learning experiences, materials, and resources for patients with PRP and their families. Our goal is to help PRP patients on their journey to become more effective advocates for their own healthcare and more powerful patients.

Why Self-Advocacy Matters

Why is Self-Advocacy Important to PRP Patients?

Self-advocacy implies that a person makes an informed decision about a matter of importance to them and then takes responsibility for bringing about the change necessary to make that choice a reality. There are approximately 6,800 rare diseases, of which 615 are rare skin disorders. Rare diseases are often chronic.

Because PRP is rare, there are a limited number of “experts” in the treatment of our disease. Self-advocacy can help PRP patients and their families be empowered to fully participate in their care. Self-advocacy is a necessity for patients with a rare disease like PRP.

Self-Advocacy Can Help You Get High-Quality Health Care

High-quality health care means getting the proper care delivered by the right dermatologist at the right time. When standing up for your health, you do everything possible to get high-quality care. Patients can play an important role in determining the need for care and the outcomes of that care.

Self-Advocacy Can Help You Feel in Control

At times, you may feel like PRP is running your life. Standing Up for your health can give you back a sense of control. Standing Up for your health gives you the information you need to make informed decisions about your medical treatment and healthcare providers.

Self-Advocacy Can Help You Get Support

When standing up for your health, you ask for help from family, friends, and others. Their support can help you cope with your disease.

Are You Standing Up for Your Health?

Read about the Five Principles of Self-Advocacy and share it with your family and friends. You’ll find practical tips and other resources you can use to become an empowered patient and strong self-advocate.

Principle 1

Build a Strong PRP Healthcare Team

Be a Strong Player on Your Team

Your dermatologist and specialists will offer you the information you need to make informed health decisions. Your PRP healthcare team can also include the following:

- ☐ Otorhinolaryngologist (impaired hearing),
- ☐ Podiatrist (impaired mobility)
- ☐ Mental health provider (impaired mental wellness)
- ☐ Family doctor or general practitioner

Empowered PRP patients see themselves at the center of their healthcare team. Only you can ensure you get your questions answered and ensure that your goals for treatment are considered.

Only you can:

- ☐ Keep your PRP healthcare team informed about your symptoms, side effects, setbacks, and healing milestones
- ☐ Share your goals related to treating your version of PRP
- ☐ Follow through on treatments as prescribed
- ☐ Make the tough choices about your healthcare

Building a solid healthcare team is important for effective self-advocacy.

- ☐ Find an expert in PRP.
- ☐ Put together a team of skilled healthcare providers you trust and can talk openly with.
- ☐ Make sure you are doing your part.

Step 1: Find a PRP-savvy dermatologist

To receive the best treatment, you'll want to see a dermatologist who either is or wants to be an expert in PRP. This will be someone who has treated similar cases and stays up-to-date on treatments. However, the problem with "experience" is that every PRP can differ. Most dermatologist (19 out of 20) will never treat PRP in their entire career. While ideal, finding a genuinely PRP-savvy expert near you is not always possible.

How can I find a dermatologist with experience in PRP?

Here are some resources that may assist in your search for a dermatologist with PRP experience or a mindset to become PRP savvy with your version of PRP.

- ☐ The best case scenario is to join the PRP Support Group and ask members living in your area for a referral to their PRP-savvy dermatologist.
- ☐ The next best step is the American Academy of Dermatology's Find-a-Derm database. It is specifically designed to provide easy access to information about dermatologists who are members of the AAD. While there is no guarantee that Find-a-Derm will identify a PRP-savvy dermatologist, it is a reasoned approach. SEE 60-Sec PRP Roadmap, Part 3.
- ☐ Contact the Dermatology Department of a teaching hospital. In many cases, the doctors at or affiliated with these teaching hospitals will be familiar with PRP.

You may need to go out of your area to visit an expert recognized as PRP savvy. Ask this expert to keep in touch with your local healthcare team.

Learn about Your Dermatologist's Background

To learn more about your dermatologist's competence and experience, you may want to visit the following:

- ☐ Federation of State Medical Boards
- ☐ This group offers profiles of licensed osteopathic and physician assistants. For a small fee, you can find facts about your doctor's education, disciplinary actions, and certification, among other areas. To order a report, go to www.docinfo.org.

☐ Some information on licensed doctors may also be available online for free through your state medical board. To find a listing of websites for state medical boards, go to

29. http://www.fsmb.org/state-medical-oards/contacts.

☐ American Board of Medical Specialties

This group will tell you for free if your doctor is board-certified in the specialty area that treats your disease. A board-certified doctor has shown great expertise in a specific area of medicine. To find out if your doctor is board-certified, call (866) 275-2267. Or, go to www.abms.org

Interview the Expert

Once you find an expert in treating your disease, you will want to interview them. Sometimes, you can call the office and speak directly to the physician or a nurse manager for a few minutes at no charge. In other cases, you will need to schedule an initial visit.

Before seeing an expert, determine whether the doctor is covered under your insurance. Some insurance plans allow you to see someone outside your program if you get pre-approval. Others require you to pay out of pocket or find another expert covered by your insurance.

Questions to Ask an Expert

☐ In what practice areas are you board-certified?

☐ How many patients with my disease have you treated?

☐ What were the outcomes?

☐ At which hospitals do you have privileges to practice

How to Select a Dermatologist

Suppose you're seeing a dermatologist for the first time, or it's been a while since your last appointment. In that case, the following tips from board-certified dermatologists can help you:

☐ Find a dermatologist that's right for you.

☐ Get the most from your appointment.

☐ Look for the acronym FAAD. These letters stand for “Fellow of the American Academy of Dermatology” and tell you that the dermatologist is board-certified. An excellent place to find out if the dermatologist is a FAAD is to go to https://find-a-derm.aad.org/. If a dermatologist is board-certified, you’ll see FAAD after their name.

☐ A word of caution about board certification. There are many types of boards and certifications. If you don’t see FAAD after your dermatologist’s name, make sure your dermatologist is board-certified by one of these organizations:

- American Board of Dermatology
- American Osteopathic Board of Dermatology
- Royal College of Physicians and Surgeons of Canada

☐ When you see one of these boards listed, you know you’re receiving care from a dermatologist who has passed board exams that thoroughly test knowledge, experience, and skills.

☐ Check what insurance the dermatologist accepts. If you are uncertain about insurance coverage, call your insurance provider. They are the best source for learning whether a dermatologist is in your network and if the visit is covered. They can also tell you whether treatment for a particular condition is covered.

Step 2: Choose a dermatologist who is a good fit for you

What is important to you?

You have seen a lot of doctors, nurses, and other healthcare professionals over the years. Some you have liked better than others. What is most important to you in the members of your healthcare team.

- ☐ My doctor considers me an important part of the team and asks me for my feedback.
- ☐ My doctor will share information freely with me and other healthcare providers as needed.
- ☐ My doctor talks openly with me about what I can expect from my treatment and my prognosis (life expectancy).
- ☐ The office is nearby, and the office hours fit my schedule.
- ☐ The time in the waiting room is not too long.
- ☐ The doctor takes the time to explain the staff members 'roles.
- ☐ My doctor has a backup healthcare provider I can reach during emergencies or non-office hours. Answer my questions and explain things in terms I can understand. They will offer examples or rephrase in lay terms if I don't understand.
- ☐ My healthcare team members refer me to the right source if they don't know the answers to my questions.
- ☐ My healthcare team members gave me articles and brochures about my disease.
- ☐ My healthcare team members return my phone calls and emails on time.

Feel Free to Get a Second Opinion

Even if you're happy with your healthcare team, getting a second opinion is okay.

- ☐ Getting a second opinion will not offend your doctor. In fact, most healthcare providers appreciate and encourage another point of view. And it's your right.

☐ Seeking a second opinion can help you and your family make difficult treatment decisions.

☐ While most insurance companies will cover a second opinion, checking with your insurance company before seeing the new healthcare provider is a good idea.

☐ You should be aware of and keep track of your out-of-pocket expenses.

Scheduling a PRP clinic visit

Dermatologists offer these tips to help you get timely care.

☐ Make your appointment as soon as possible. The sooner you can make your appointment, the better. Schedule several weeks or even months ahead of when you wish to be seen.

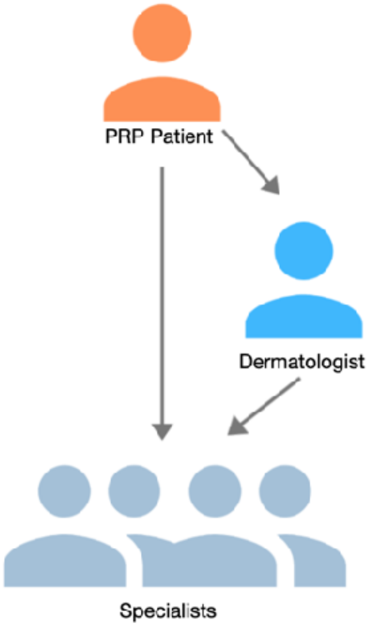
☐ Explain your concerns. If you are worried about a particular symptom or have pain, briefly explain this to the receptionist or put it in the online form. Dermatologists try to work with patients with urgent issues as soon as possible.

☐ Explain your concerns

30. Dermatologists want to keep their patients healthy and happy. They will know if your condition needs urgent care.

☐ Ask if there's a waitlist. Many dermatologists keep a waitlist, allowing patients on the waitlist to see the dermatologist before their scheduled appointment if another patient cancels. If the office doesn't own a waitlist, check with the office frequently to see if an earlier appointment has become available.

Step 3: Select a PRP Care Coordinator



Your care may involve many specialists for various impairments, e.g., vision, hearing, mobility, mental wellness, etc. You may find choosing one healthcare professional responsible for coordinating your PRP-related care helpful. Some patients use their dermatologist as their care coordinator, while others work with their primary care physician.

Your care coordinator can:

- Help arrange consultations with specialty doctors and supportive care providers
- Maintain your medical records in their office (Of course, you'll also want copies of them.)
- Talk with your other healthcare team members when needed
- Of course, your dermatologist may want to dodge this bullet.

Know How to Reach Your Healthcare Providers

You need to know how to reach your healthcare providers, day or night. Find out the best ways to contact them. Know the rules regarding giving and leaving messages.

Self-Advocacy When You Are in the Hospital

While you know about the healthcare providers you work with regularly, it is different when you need to visit the hospital. You'll want to find out what the healthcare providers 'names are and who does what.

Step 4: Do your part to become an empowered PRP patient

Remind yourself that you are at the center of your PRP healthcare team. Once you have your medical care in place, you'll want to concentrate on what you can do to be a strong player.

Share Information

- Share information with your PRP healthcare team. Tell them your symptoms — no matter how minor they seem.
- List all medicines (including over-the-counter medicines), vitamins, and complementary and alternative therapies (including herbal remedies or supplements) you are taking.
- Discuss any emotional symptoms you are having. The more you share, the better your healthcare team can help you.
- Share your goals for treatment. What are you hoping treatments will do for you?

Be a Team Player

- Go to all your scheduled office visits.
- Once you agree to a treatment plan, be sure to follow through. If the treatment is not what you expected, share your concern.

Make the most of PRP office visits

- Think about what you hope to get out of the visit. Bring questions.
- Ask for a copy of lab results or health records at the doctor's office.
- Ask your healthcare providers to give you any written medicine or treatment instructions.

Be politely assertive when asking for what you need

- Try using "I" statements, e.g., "I think," "I feel," or "I don't understand."

Satisfaction Assessment Checklist

Use the checklist below to help you assess what you are looking for when choosing members for your healthcare team. You can also use this checklist to determine how satisfied you are with your current healthcare providers.

- ☐ My doctor considers me an important part of the team and asks me for my feedback. Very important Somewhat important Not very important to me
- ☐ My doctor will share information freely with me and other healthcare providers as needed. Very important Somewhat important Not very important to me
- ☐ My doctor talks openly with me about what I can expect from my treatment and my prognosis (life expectancy). Very important Somewhat important Not very important to me
- ☐ The office is nearby, and the office hours fit my schedule. Very important Somewhat important Not very important to me
- ☐ The time in the waiting room is not too long. Very important Somewhat important Not very important to me
- ☐ The doctor takes the time to explain the staff members ’roles. Very important Somewhat important Not very important to me
- ☐ My doctor has a backup healthcare provider I can reach during emergencies or non-office hours. Very important Somewhat important Not very important to me
- ☐ My healthcare team members answer my questions and explain things in terms I can understand. They will offer examples or rephrase in lay terms if I don't understand. Very important, Somewhat important, Not very important to me.
- ☐ My healthcare team members refer me to the right source if they don’t know the answers to my questions. Very important Somewhat important Not very important to me
- ☐ My healthcare team members gave me articles and brochures about my disease. Very important Somewhat important Not very important to me

☐ My healthcare team members return my phone calls and emails promptly.
Very important Somewhat important Not very important to me

☐ Summary

Building a solid healthcare team you trust and can talk freely to is vital to becoming an empowered patient. Being comfortable with your team can help you get the answers you seek. And remember, you have the right to seek a second opinion.

It can take time to feel at ease when taking the lead in your healthcare. You've done other difficult things and can do this, too. Be patient with yourself and know that with practice, you can develop solid skills as you advocate for your best healthcare.

SKILLS CHECKLIST

Use this checklist to assess your skills in building an effective healthcare team.

☐ I have an expert in my disease on my healthcare team.

☐ I feel comfortable with each of my healthcare providers.

☐ I'm doing everything possible to be an empowered team player. I'm able to reach my crucial healthcare providers whenever I need to.

☐ I feel comfortable getting a second opinion.

☐ I find my healthcare team supportive of my seeking a second opinion.

Peer Review Ponderings

Principle 2

Learn About PRP.

Get Up-To-Date Information

Many rare diseases are complex. You need to learn all you can about PRP and the available treatments.

Staying well-informed helps empower you to be knowledgeable about PRP and actively share in your treatment decisions. It allows you to educate family and friends, get more out of office visits, make choices about your treatment, and monitor your care.

Today, learning about PRP and treatment options is easier than ever. You can get information from books, leaflebrochures, medical journals, medical magazines, and—of course— the Internet.

Tips for Getting Information About PRP

Connect with the PRP Alliance and the PRP Support Group. Access the *PRP Survival Guide* and locate reliable information on the internet. Learn all you can about your treatment options.

Questions to Ask Your Healthcare Team

You may want to take the 60-Second PRP Roadmap with you as you ask the following questions:

Confirm the BASICS about PRP

☐ What constitutes a rare disease?

☐ What is PRP?

☐ What are the signs and symptoms of PRP

☐ How does PRP progress?

☐ Is there a cure for PRP?

☐ What are healing milestones?

☐ What are the metrics of PRP?

☐ What are the quality of life impacts of PRP

☐ What should I know about my skin?

☐ What should we say to family & friends?

☐ **What words are important to know?**

☐ **What is the importance of hope?**

☐ **Does anything good from a PRP journey?**

About Treatment Options

☐ **What are all my treatment options?**

☐ **What treatment option do you recommend for me? Why?**

☐ **How likely am I to get better with the treatment?**

☐ **Has this treatment been used often, or is it new or experimental?**

☐ **How long will the treatment take to work? When and how will I know if it is working? What happens if the treatment doesn't work?**

☐ **What are the potential side effects of treatment?**

☐ **Can my disease return, even after successful treatment?**

About Prescription Drugs

☐ **Has the drug been approved by the U.S. Food and Drug Administration (FDA) for treating my disease?**

☐ **How do I take the medicine? How often do I need to take it?**

☐ **How long will I need to take the medicine? When can I expect the treatment to start working?**

☐ **What are the common side effects of this medicine? What are the most severe side effects? What can be done to control them? Do you have any tips for staying on the treatment plan?**

☐ **What are the long-term side effects of this medicine?**

☐ **How much does it cost? Is it covered by my insurance?**

☐ **Can this prescription drug cause interactions or side effects with any other prescription drugs, over-the-counter medicines, or supplements that I take?**

Find a PRP Patient Advocacy Resources

Patient advocacy is an area of specialization concerned with advocacy for patients, survivors, and caregivers. Patient advocacy organizations exist for many diseases, even rare diseases, including PRP. These organizations may have information about your disease. They may be able to connect you to other patients who understand what it is like to live with your disease. The following organization have patient-friendly content about pityriasis rubra pilaris. focus on your rare disease.

☐ PRP Alliance" See page XXX

☐ PRP Support Group: See page XXX

☐ PRP Global Database: See page XXX

☐ PRP Survival Guide: See page XXX

☐ International Alliance of Dermatology Patient Organizations: See page XXXNational Organization of Rare Disorders See:www.rarediseases.org NORD is a unique federation of voluntary health organizations dedicated to helping people with rare "orphan" diseases and assisting the organizations that serve them. NORD is committed to identifying, treating, and curing rare disorders through education, advocacy, research, and service programs.

☐ Genetic and Rare Diseases (GARD) Information Center:See https://rarediseases.info.nih.gov/

☐ Global Genes: See: www.globalgenes.org

The Global Genes Project's mission is to increase rare disease awareness and public and physician education, build community through social media, and support research initiatives to find treatments and cures for rare and genetic diseases.

☐ Health Hotlines: See: www.healthhotlines.nlm.nih.gov

This is a database of toll-free numbers from the National Library of Medicine (NLM) with descriptions of more than 14,000 biomedical information resources, including organizations, databases, research resources, etc.

☐ National Library of Medicine: Seewww.nlm.nih.gov

This website has reliable and detailed information on many diseases. It has a simple search tool on the main page.

☐ MedlinePlus; See: www.medlineplus.gov

This website offers a wealth of information about health topics, drugs, and supplements. It also provides videos and interactive tutorials. The website is maintained by the National Institutes of Health. It includes a tutorial that teaches you how to evaluate the information you find online.

Finding Trustworthy Information on the Internet

There is a lot of information on the Internet. It can be like trying to sip water from an open fire hydrant. How do you know what to trust? You'll want to find and use only accurate and current information. Reliable health information comes from scientific research conducted in government, university, or private laboratories, and dedicated websites will tell you where the data came from and how it has been reviewed.

Information on a website should not replace your doctor's advice. Your doctor is the best person to answer questions about your personal health. If you read something on the web that doesn't agree with what your doctor has told you, ask them about it. Use these tips for finding good and reliable information on the Internet:

Is it clear who sponsors the website?

☐ The we address can help you learn about the type of site you are visiting:

- A government agency has .gov in the address.
- An educational institution is indicated by .edu in the address.
- A professional organization such as a scientific or research society and patient advocacy organizations will be identified as .org.
- Commercial sites identified by .com will most often recognize the sponsor as a company, such as a pharmaceutical firm.

Is there a way to contact the website sponsor?

☐ Trustworthy websites provide contact information. Many even have toll-free numbers to call.

Is there an editorial board that reviews the information?

☐ Click on a website's "About Us" page to see if an editorial or medical advisory board reviews the information before putting it online. Government websites may not have this page.

Is the information current?

☐ New research findings can make a difference in making intelligent choices regarding your treatment for your rare disease. You'll want to look carefully to determine when the web page was last updated. Older information isn't useless, however. Many websites provide older articles so readers can get a historical view of the information.

Is your privacy protected?

You want to be as specific as possible that your personal information is not shared with other lists or companies. Take time to identify and read the website's policy. If the website includes something like, "We share information with companies that can provide you with products," that's a sign your information isn't private. Never give out your Social Security number. If you asked for personal information, be sure to find out how the information is being used by contacting the website sponsor by phone, mail, or the "Contact Us" feature on the website.

Could the claims be too good to be true?

Be careful of claims that various remedies will offer dramatic cures or that offer to sell you something to treat your disease. Talk with your healthcare team about all treatment claims before buying and trying products you may find on the Internet.

- Drugs@FDA: See: www.FDA.gov
  This website, developed by the U.S. Food and Drug Administration (FDA), provides detailed information about all FDA-approved drug products.
  You can easily search by generic or brand name.

Learn About Your Treatment Options

When making a treatment decision, use reliable information and input from your healthcare providers, family, friends, and others. Weigh the pros and cons of each treatment. Remember, you have the right to choose or refuse treatment.

What is a treatment plan?

A treatment plan is like a road map that a patient will follow on their journey through treatment. It outlines the progress of disease treatment or therapy. A treatment plan may be highly formalized in a medical record or consist of loosely handwritten notes. Remember that the treatment plan is always subject to change as medications, therapies, or your rare disease progresses.

Thinking of Participating in Research?

If you have PRP, you may want to consider participating in research.

☐ Patient-initiated PRP research — PRP Alliance, e.g Onset to Diagnosis, What Works (Efficacy), Quality of Life

☐ Third-Party PRP research — Oregon Health and Science University, GlobalSkin. Thomas Jefferson University, Mayo Clinic, etc.

☐ Third-party rare disease research — Caregiver

☐ Third-party skin disorder research — International Alliance of Dermatiology Patient Organizations (GlobalSkin)

Keep an Eye on Your Care

Healthcare providers are only human—mistakes can happen. As an informed patient, you can check to ensure you get the proper treatment every time—in the office, lab, and hospital.

Tips for Preventing Medical Mistakes

Be an active member of your PRP healthcare team. Take part in every decision about your health. Research shows that patients who are more involved get better care.

- ☐ **Make sure your dermatologist knows all the medicines and supplements you take.**
- ☐ **Make sure your dermatologist knows about any allergies or adverse reactions you have had to medicines.**
- ☐ **Ask for information about medicines and treatments in terms you understand–when your medicines are prescribed and when you receive them. What is this medicine for? How am I supposed to take it? What side effects are likely? What do I do if they occur? Is this medicine safe to take with other medicines I am taking? What food or drink should I avoid while taking this?**
- ☐ **When you pick up a medicine or get a treatment at a medical center, ask: Is this the medicine/treatment my dermatologist prescribed?**
- ☐ **Make sure that all the healthcare providers involved in your care have essential information about you. Don't assume that everyone knows what they need to know.**
- ☐ **Ask a family member or friend to be there with you and be an advocate.**
- ☐ **Know that more is not always better. Ask why a test or treatment is needed and how it can help. You might be better off without it.**
- ☐ **Don't assume that no news is good if you have a test. Find out the results.**

Need help to pay for PRP clinic visits or treatments?

What if your health insurance doesn't cover seeing specialists or certain treatments?

Read below to find out where you can get help.

- ☐ **Patient Advocate Foundation**: www.patientadvocate.org.
  Call (800) 532-5274 to see if they can help with fees. Ask about their Co-Pay Relief program.
- ☐ **Chronic Disease Fund**: www.cdfund.org.
  This non-profit organization provides financial help to seriously ill Americans without insurance who cannot afford their medicines. Call toll-free (877) 968-7233
- ☐ **Department of Health and Human Servic**es (DHHS): www.hhs.gov. DHHS has information on assistance in paying for medical care for low-income families. Go to the "Families & Children" section of the website. Call toll-free (877) 696-6775.
- ☐ **Insure Kids Now**www.insurekidsnow.gov. Insure Kids Now is a resource for free or low-cost health insurance for children and teens.
- ☐ **Find a Health Center**: www.findahealthcenter.hrsa.gov.
  You can find federally funded free or low-cost medical care using the "HRSA Find a Health Center" tool.
- ☐ **National Organization for Rare Disorders (NORD)** www.rarediseases.org.
  The NORD website includes information on medication assistance programs and other online resources. Contact NORD by email at orphan@rarediseases.org. or call (203) 744-0100.
- ☐ **RxAssist**: See: www.rxassist.org
  Website has a complete database of patient assistance programs, practical tools, news, and articles.

Gather and Organize

Once you've gathered all the information on PRP treatment options, read it and organize it. Then, share what you've learned with healthcare providers, family, and friends.

Summary

Take the time to learn about your disease and your PRP treatment options. Remember, knowledge is a powerful part of self-advocacy. The more you know, the better you can make informed choices about your care.

As you gain health advocacy skills, taking this patient guide to your doctor visits may be helpful. Some patients find reading the questions from the list helps them stay on track as they seek answers from their healthcare providers.

SKILLS CHECKLIST

Use this checklist to assess your skills in learning about your disease and available treatment options:

- **I have sought up-to-date information about my disease by talking to specialists and doing my own research.**
- **I seek information only from reliable sources on the Internet.**
- **I asked my healthcare team about treatment options.**
- **I know whom to contact for help paying for medicines or treatment.**
- **I feel able to take the ten steps to help prevent medical mistakes.**

Peer Review Ponderings

Principle 3

Make the Most of PRP Office Visits

Prepare for Each PRP Office Visit

Office visits may be shorter than you hoped. To be a strong advocate for yourself, you must do everything you can to make the most of every visit. You'll want to do some work in advance.

Set an Agenda For the Visit

Ask yourself, "What do I want to get from this visit?" Write down the top three things you want to discuss. Create an agenda.

Ask For the Time You Need

When you make an appointment, let the staff know if you have unique concerns requiring extra time with your doctor.

Do Your Homework

Learn all you can about your condition and the available treatments.

Bring Information With You

Bring your medical information to the visit. This information should include:

☐ A list of questions related to your PRP journey in priority order

☐ A list of prescription and over-the-counter medicines, vitamins, herbs, and supplements you have used

☐ Notes on symptoms and side effects you are having

Form a Good Relationship With Your PRP Healthcare Team

Showing appreciation for your dermatologist and staff is essential. And it enables them to focus on your condition and make the best use of your time with you. Going to the doctor can be overwhelming. Asking a family member or friend to come along can help. This person can help you ask questions, write down answers, and ensure you get answers you understand. A family member or friend can also provide emotional support.

Ask Questions, Get Answers

Clear communication is an integral part of good healthcare. To make intelligent choices about treatment and follow your care plan correctly, you need to fully understand your disease and treatment options. These are the things that empowered patients do to make sure they get the best care and treatment.

Ask the Most Important Questions First

Be sure to bring a list of questions and put them in priority order. You may have just a few minutes to ask your questions, so ask the most important ones first.

Ask for Clarification

Don't worry about offending your healthcare provider. Healthcare providers tend to share more information with patients who pay attention and ask questions. Asking questions shows healthcare providers that you are actively involved in your care and want to stay informed. Let your doctor know if the information is confusing or if there is anything you don't understand.

If you have trouble understanding your dermatologist

☐ Ask the dermatologist to repeat things or use simpler words until you know them.

☐ Ask a family member or friend to listen to you.

☐ Ask for brochures, drawings, or models.

☐ Ask the provider to write down answers.

Take Notes

After the visit, it may be hard to recall exactly what your provider said. Take notes, ask a family member or friend to take notes, or use an audio recorder. Keep these notes with your medical information.

Remember:

- ☐ Your healthcare provider is your paid consultant and part of the team you must organize to obtain the best care possible.
- ☐ Be respectful, but do not be intimidated.
- ☐ Always consult with your doctor before making any medical care changes.
- ☐ You are always entitled to a second opinion.

Summary

Have you ever left your doctor's office unsure of what to do? Not quite sure how to take your medicine? You're not alone. There is a lot of information to take in at each visit. The steps outlined in this section can enable you to get the most from your office visits. While not always easy, asking questions until you get answers you understand is a central part of becoming an empowered patient.

SKILLS CHECKLIST

Use this checklist to assess your skills in preparing for office visits and making sure you get your questions answered in a way you can understand:

- I take steps to prepare for each visit by writing a list of questions beforehand and bringing them along with me.
- I ask my healthcare team questions.
- I make sure to get answers I understand.

Principle 4

Keep Track of Health Information

An Organized Patient is a Powerful Patient

Keeping all your medical information organized and in one place makes it easier to track changes in your health and share information with your healthcare provider. Read this section to learn how to keep track of your health information.

Gather Information

Collect information about your health, office visits, and treatment in one place. Don't feel that you must gather all your health information simultaneously. The next time you visit the dermatologist, simply ask for recent records and do so each time you visit a healthcare provider. Here are some of the items you'll want to hold onto:

Health Information related to your PRP journey

☐ **Progression of signs and symptoms**

☐ **Flares & setbacks**

☐ **Treatments, e.g., what worked and what didn't**

☐ **Healing milestones**

☐ **Test results**

Office Visit Information

☐ **Dates of office visits**

☐ **Notes on what providers tell you**

☐ **Questions to ask healthcare providers**

Treatment Information

☐ **List of medicines and other treatments, vitamins, herbs, and supplements you have used**

☐ **Notes on side effects**

Treatment instructions

☐ **Financial and Legal Information**

☐ **Insurance claims—paid and unpaid (You can appeal claims denied by your insurance company. Call your insurance company for steps to take to appeal.)**

Contact Information

☐ **Family and friends**

☐ **Other patients**

☐ **Pharmacy—note when they are open**

☐ **Healthcare providers—note when they have office hours and how to reach them after hours during an emergency.**

☐ **Insurance—note when they are available to take calls from members**

Ask for Copies of Health Records at Each Visit

Getting your health records weeks or months after a doctor's visit can be difficult. Different states have different rules. Sometimes, you must complete an application; sometimes, there is a fee. Ask for crucial health records at the end of each visit—test results and transfusion records.

A federal law, the HIPAA Privacy Rule (available at www.hhs.gov/ocr/privacy/index.html), gives you the right to see, get a copy of, and correct your medical records. Most states also have laws that provide you with access to your records.

Take Notes

Keep a log of changes in how you feel. Some of these changes may be symptoms of your disease. Others may be side effects of treatment. This is vital information that only you can provide.

Keep a running list of questions to ask providers. And record your healthcare providers 'answers. Include what they say over the phone, via email, and during office visits.

Find an Organization System that Works for You

Now that you have all this information, you must figure out the best way to keep it organized and in one place. Here are some tools other patients have found helpful:

- ☐ **3-ring binder with dividers**
- ☐ **Accordion folder**
- ☐ **Manila file folders**
- ☐ **Notebook or journal**
- ☐ **Electronic files and spreadsheets on a laptop or desktop computer, tablet, flash drive, or smartphone**
- ☐ **Online tools such as a personal electronic health record.**

Remember—there's no right way to compile or organize. Talk to other patients to learn how to keep their health information organized.

Consider Creating Your Own Personal Health Record

If you don't have one, now may be the time to create a complete personal health record or PHR. The PHR is a tool that you can use to collect, track, and share all your past and current health information. A complete PHR can help you share your health history with healthcare providers, avoid unneeded tests and procedures, and lower the chance of medical errors. To start your personal health record, you must request a copy of your health records from all of your healthcare providers, including your primary care doctor, eye doctor, dentist, and any other specialists you have seen.

Remember, you're in charge of making choices about your health. A PHR can help you do that. For detailed instructions on how to create your own PHR, go to www.myPHR.com. This website is a service of the American Health Information Management Association (AHIMA).

Share Information

Share your medical information with your healthcare providers, family, and friends. It will help them stay in the loop—so they can assist and support you better.

Take Medical Records With You When Traveling

Are you going on a long trip? Be sure to take your medical information with you. You never know when you might need it. Having a current medication list in your wallet or purse in an emergency is also a good idea. Make photocopies and leave a set at home if you lose this information.

If you are going on a trip, you may want to locate hospitals, pharmacies, and other medical facilities near your destination in advance. You can include this information in your healthcare files.

Summary

Keeping track of your medical records and health information can be quite a task. Yet, being able to put your hands on the health information you need with a moment's notice can be very helpful as you advocate for your health. While it may not come naturally to you to ask for your health records, gaining this assertiveness skill can help you feel in control of your health.

SKILLS CHECKLIST

Use this checklist to assess your skills in organizing essential health information:

- **I use a system for managing my health information.**
- **I can put my hands on health information when I need it.**
- **I ask for essential health records at the end of each visit.**
- **When I travel, I take my health information and ensure I have a second copy at home.**

Principle 5

Build a Strong PRP Support Team

You Don't Have To Do It Alone!

Having PRP can be a heavy burden. But you don't have to carry it alone. There are people in your life—and people you don't know yet—who would be glad to help you cope with the physical and emotional demands of PRP. With their support, you can become a strong self-advocate.

Get Support from Family and Friends

Family and friends can be a tremendous source of strength. They can help you with daily tasks and give you emotional support. It can be hard to accept that you need support–but it is important for your health. Try these tips:

Make a list of tasks

Keep a list of tasks you could use help with. For instance, you might ask family and friends to:

- Cook, clean, and shop for you
- Give you rides
- Go to PRP clinic visits with you or look after your children
- Organize your medical information
- Research your disease and treatment options
- Talk with you or listen while you talk

Ask for help

Family and friends often want to support you, yet they may not know how. Feel free to tell them what you need. Try these conversation starters:

- ☐ "I could use a hand with …"
- ☐ "It would be really helpful if you would …"
- ☐ "I'm not feeling very well today. Would you mind …"
- ☐ "I sure could use some help with …"
- ☐ "I'd really appreciate it if you …"

Keep friends and family "in the loop"

Family and friends want to know how you're doing. But answering their questions can be tiring. Think about asking one family member or friend to give updates on your behalf. Or post on your behalf to the PRP Support Group.

Know your limits

Having a rare disease takes a lot out of you. Know your limits, and feel free to say "no" to outings and activities planned by family and friends.

Get Support from a PRP Health Advocate

A health advocate is a family member, friend, trusted coworker, or even a paid professional who can help you advocate for your best care. This person can accompany you to doctor visits, ask questions, and write down information. You may also want your health advocate to speak up for you when needed.

Ask a Family or Friend To Be Your PRP Health Advocate

A friend or family member may be able to serve as a patient advocate. You may want to ask a few people to share that role. Each person can take on different tasks. One person may be better able to assist you with a doctor's visit. At the same time, another might be able to help you with insurance issues.

Or choose a professional patient advocate.

The Patient Advocate Foundation has professional case managers who provide various services to help patients settle issues with access to care, medical debt, and job retention related to their illness. Their services may be free. To learn more, call them at (800) 532-5274. Or, go to www.patientadvocate.org. Also, many hospitals, community health centers, and long-term care facilities have professional patient advocates on staff. They go by different titles, including care manager or case manager, health advocate, healthcare or nurse navigator, health advisor or social worker.

Why Should You Use a Health Advocate?

According to Carolyn M. Clancy, MD, former Director of the U.S. Health and Human Services Agency for Healthcare Research and Quality's research shows that quality healthcare means taking an active role in decisions about your care. She writes:

"If you're facing a difficult medical decision, it's a good idea to bring someone with you who can help you take an active role in your care when you're not fully up to it. As a doctor and a patient, I've seen how valuable it is to have 'another set of ears and eyes 'in the exam room.

"Having an advocate at medical appointments or during a hospital stay can ensure you get the information you need to manage your health. Who makes a good health advocate? Someone calm pays attention to details and can clearly ask questions and state information."

Health advocates can:

- Ask questions or voice concerns to your dermatologist for you.
- Ask the “what’s next” questions: “If this test is negative, what does it mean? If it’s positive, will more tests be needed?”
- Compile or update your medicine list.
- File paperwork or assist with insurance matters.
- Help arrange rides.
- Help you follow treatment instructions, including asking questions about your follow-up care.
- Remember your medicine schedule.
- Research treatment options, procedures, doctors, and hospitals.

Get Support From Other PRP Patients

People living with PRP know more about what you’re going through than anyone else. They can let you know you’re not alone. John, the PRP Support Group. Over 2,000 members share their PRP-related insights and recognize that we are on this journey together. One voice!

Get Support From Your PRP Healthcare Team

You may be able to get emotional support from a member of your healthcare team. This healthcare provider may be a counselor or social worker. Or it may be a nurse or doctor who goes above and beyond their professional duties.

Get Support From Patient Advocacy Organizations

The PRP global community is supported by the PRP Alliance, the PRP Support Group and the International Alliance of Dermatology Patient Organizations (also known as Globalskin).

Stand Up for Your Health—Today!

Standing up for your health is a process. It happens over time as you learn more about PRP and gain confidence. Refer to this patient guide to remind yourself of the core skills needed to become an empowered patient. We hope it will help you take steps to become a stronger self-advocate.

Summary

For many of us, asking for help is not easy. Yet, empowered patients know that help from others can help them get the healthcare services and treatment they need. Some of us have family members or close friends who live nearby and are willing to serve as part of our support team. And most of us have joined the PRP Support Group.

Having PRP can be complicated. Knowing someone in your corner can help you overcome the ups and downs.

SKILLS CHECKLIST

Use this checklist to assess your skills in building a strong personal support team:

- I ask friends, family, or trusted coworkers to help when needed.
- I get support by connecting with other patients.
- My healthcare team offers support.

Peer Review Ponderings

Peer Review Ponderings

Helping Those You Love to Stand Up for Their Health

Empowering parents and adult children of patients with PRP.

Editor's Note: Having a child, teenager, or parent with PRP presents the same unique challenges for those who love and care for them as it can for adult patients with PRP. The first five principles described in this guide apply to caregivers managing the care of a loved one. We have added some specific suggestions to empower parents and adult children of patients with PRP.

Standing Up for Your Child's Health

There is nothing more devastating to a parent than to see your child suffer from Juvenile Onset PRP. For every parent who has heard these words, "Your child has pityriasis rubra pilaris," it is a moment frozen in time and a life-changing event. Whether you've just learned your child has PRP or you have been managing your child's PRP for many years, this section provides tips, information, and resources that can help.

Is Your Child Recently Diagnosed with PRP?

If your child was recently diagnosed, it is likely your days have suddenly become filled with worrying. This can be overwhelming. You might feel depressed or even angry and ask, "Why did this happen to us?" While those feelings are normal, knowing you're not alone is essential. The first five principles in this guide can all be used to help you be a firm parent advocate to stand up for your child's health and ensure that they and you get the support you need.

Supporting Students With PRP

Sending your child back to school after treatment for any disease can be scary and overwhelming. Speak with your child's teacher, doctors, and school nurse. Explain any limitations or precautions necessary to have your child safely return to their classroom.

Take Care of Yourself

As any parent of a sick child knows, this is a genuine dilemma. On the one hand, your focus must be on helping your child get healthy. On the other hand, your child needs you to be healthy, strong, and in control. You can only be this model for your child if you care for yourself by eating right, exercising, and getting plenty of sleep. If you don't have time to do this, eliminate other things from your life and make the time. Your mental, physical, and spiritual health is critical to your child's health.

Find Time to Have Fun

Your child's illness is serious. Sometimes, you may feel like every second of your life revolves around medication schedules and trips to the doctor. You need relief by not taking every moment of the day too seriously. Make time to do things you enjoy – even if you need to sandwich fun times between appointments. This could be as simple as walking, playing cards with a friend, doing a craft project or going to the park. List five things you have fun doing – and commit to doing one this week. You will find you feel more refreshed when you do.

Adolescents with PRP Transitioning to Adult Care

As parents, we do our best to prepare our children for adulthood. We teach them right from wrong, social skills, and how to do their laundry and drive a car, but do we prepare them to transition from pediatric medical care to adult medical care?

"Transition" in patient care is moving from pediatric to adult health care. The process can often present challenges for youth with PRP and their families. The transition generally begins once your child is a teenager, but don't wait to get your child involved. The sooner your child learns about their illness, the better prepared they are to begin taking charge and stand up for their own health.

How do parents begin preparing their children to assume responsibility for their own medical care and make the best choices? Below are some suggestions on how to start the process.

☐ Identify a transition team. These key people will help your child transition from pediatric to adult care.

☐ Establish a transition plan to identify your child's specific needs.

☐ Create a transition record, including a notebook summarizing medical information, including the patient's medical insurance and history of hospitalizations, procedures, and tests.

☐ Complete this skills assessment checklist. This will help you and your child identify their strengths and in what areas they need assistance.

☐ Seek recommendations from your child's pediatrician for adult care physicians. Schedule an appointment to meet the physicians so you can help your child in the selection process.

Pediatric to adult healthcare transition can be a scary process for parents, especially when your child has a chronic illness. Ask your child's dermatologist about resources that can help make the move from pediatric care to adult care successful.

Standing Up for Your Parent's Health

Standing up for your health is essential for every patient with PRP. It becomes more and more complicated as parents age. They may have other illnesses or disabilities that must also be considered when advocating for their health. You and/or your parent may need to be more proactive with their healthcare providers.

These first five principles of standing up for your health may be new to your parent and might take some coaching or assistance from you. The goal is to empower your parent to stand up for their health. That includes encouraging them to do all that they are capable of doing.

Suppose you want them to be more engaged in their healthcare decisions. In that case, it is important that you not assume the role of primary advocate for them unless it is clear that they cannot adequately stand up for themselves. Many adult children step in prematurely to "help" their parents without first assessing the parent's true capabilities.

How you answer the questions below will help determine to what degree, if any, you should partner with your parent as their health advocate. Regardless of whether your parent is a strong self-advocate or does so with your assistance, the five principles described in this guide suggest practical ways to ensure that your parent gets the quality healthcare they deserve.

Here are several essential questions you can ask yourself to better assess whether your parent needs you to be their health advocate or if they just need a little support to be able to stand up for their own health:

- ☐ What is your parent's overall health status? Are there vision or hearing impairments that would make communication with healthcare providers difficult or confusing? Does your parent exhibit cognitive difficulties, such as memory loss or confusion?

☐ Is your parent expressing a desire for help?

☐ Is your parent easily overwhelmed by details about their PRP condition or other health issues?

☐ Does your parent typically ask pointed questions of healthcare providers to get the necessary information? Is your parent's information about symptoms, medical history, or other details about PRP and overall health accurate and thorough?

Peer Review Ponderings

Peer Review Ponderings

Appendix F

Online PRP Resources

Online Resources for Empowering PRP Patients

Peer Review Message: When I was diagnosed with PRP in November 2012, I searched the Internet for answers. It would have been helpful if someone had a list of online resources that are truely appropriate for PRP patients and caregivers.

To that end, Peer Review participants will be asked to pre-screen and evaluate online resources,

Federal Government Website Resources

Use the information and organizations in this resource list as you seek information to help you become an empowered patient.

Agency for Healthcare Research and Quality (AHRQ)

www.ahrq.gov (301) 427-1104

The Agency for Healthcare Research and Quality's (AHRQ) mission is to produce evidence to make health care safer, higher quality, more accessible, equitable, and affordable, and to work within the U.S. Department of Health and Human Services and with other partners to make sure that the evidence is understood and used. This branch offers resources for healthcare consumers at www.ahrq.gov/consumer.

Clinical Trials

www.clinicaltrials.gov

This website provides up-to-date information about clinical research being carried out worldwide for various diseases and conditions. It tells each trial's purpose, who may participate, locations, and contact numbers. It also describes the clinical trial process and reports recent results.

Food and Drug Administration (FDA)

www.fda.gov (888) 868-4000

The FDA is the federal agency regulating drugs, medical devices, and biologics. This website contains information on drugs, blood products, recalls, and safety alerts. The FDA also has a website called Drugs@FDA. This site provides detailed information about all FDA-approved drug products. You can easily search by generic or brand name. To find this site, go to www.fda.gov and search for Drugs@FDA.

Genetic and Rare Disease Information Center (GARD)

https://rarediseases.info.nih.gov/

Modernizing GARD means adapting how information is shared for rare diseases identified in a few cases, diseases affecting thousands yearly, and everything in between. We are translating information from significant data sources developed for clinicians and scientists to make it easy for you to use and understand.

Medline Plus

www. medlineplus.gov

This website offers a wealth of information about health topics, drugs, and supplements. It also provides videos, interactive tutorials and information about PRP, including a link to prpAlliance.comNational Institutes of Health Nationbal **Institutes of Health**

www.nih.gov

> The National Institutes of Health (NIH), a part of the U.S. Department of Health and Human Services, is the nation's medical research agency—making important discoveries that improve health and save lives. The NIH is the U.S. Department of Health and Human Services research arm.

PubMed

www.pubmed.gov

> PubMed comprises more than 24 million citations for biomedical literature from MEDLINE, life science journals, and online books. Citations may include links to full-text content from PubMed Central and publisher websites.

Non Profit Websites

Again, use the information and organizations in this resource list as you seek information to help you become an empowered patient.

National Organization for Rare Disorders (NORD)

www.rarediseases.org

> The National Organization for Rare Disorders (NORD) provides advocacy, education and other services to improve the lives of all people affected by rare diseases. NORD maintains a patient organization database to help search for organizations that help people with rare diseases.

Patient Advocate Foundation

www.patientadvocate.org (800) 532-5274

This group can help you find professional case managers who provide various services to help patients settle issues with access to care, medical debt, and job retention related to their illness. This group also provides resources for how to be your own advocate.

Rare Connect

www.rareconnect.org

RareConnect was created by EURORDIS (European Rare Disease Organisation) and NORD (National Organization for Rare Disorders) to provide a safe space where individuals and families affected by rare diseases can connect, share vital experiences, and find helpful information and resources.

RxAssist

www.rxassist.org

This website has a complete database of patient assistance programs run by drug makers. These programs can provide free or low-cost drugs to patients who qualify. They may also be able to help you work with your insurance company to get your medication covered. The website also has practical tools, news, and articles.

Finding Health Information

Finding Health Information on the Internet: How To Start

To find accurate health information, start with one of these organized collections of high-quality resources:

- ☐ MedlinePlus, sponsored by the National Library of Medicine, which is part of the National Institutes of Health (NIH)
- ☐ healthfinder.gov, sponsored by the Office of Disease Prevention and Health Promotion in the U.S. Department of Health and Human Services
- ☐ For information on dietary supplements, visit the NIH Office of Dietary Supplements website. GO TO: https://ods.od.nih.gov/

More To Consider

- If you're considering using a dietary supplement, first get information from reliable sources. Remember that dietary supplements may interact with medications or other supplements and contain ingredients not listed on the label. Your healthcare provider can advise you.
- Take charge of your health—talk with your healthcare providers about any complementary health approaches you use. Together, you can make shared, well-informed decisions.

For More Information

NCCIH Clearinghouse

The NCCIH Clearinghouse provides information on NCCIH and complementary and integrative health approaches, including publications and searches of Federal databases of scientific and medical literature. The Clearinghouse does not provide medical advice, treatment recommendations, or referrals to practitioners.
Website: https://www.nccih.nih.gov

NCCIH and the National Institutes of Health (NIH) provide tools to help you understand the basics and terminology of scientific research to make well-informed health decisions. Know the Science features a variety of materials, including interactive modules, quizzes, videos, and links to informative content from Federal resources designed to help consumers make sense of health information.

Explaining How Research Works (NIH)

Know the Science: 9 Questions To Help You Make Sense of Health Research

Understanding Clinical Studies (NIH)

PubMed®

A service of the National Library of Medicine, PubMed® contains publication information and (in most cases) brief summaries of articles from scientific and medical journals. For guidance from NCCIH on using PubMed, see How To Find Information About Complementary Health Approaches on PubMed.

Office of Dietary Supplements (ODS), National Institutes of Health (NIH)

ODS seeks to strengthen knowledge and understanding of dietary supplements by evaluating scientific information, supporting research, sharing research results, and educating the public. Its resources include publications (such as *Dietary Supplements: What You Need To Know*) and facts on various specific supplement ingredients and products (such as vitamin D and multivitamin/mineral supplements).

Website: https://ods.od.nih.gov
Email: ods@nih.gov

MedlinePlus

To provide resources that help answer health questions, MedlinePlus (a service of the National Library of Medicine) brings together authoritative information from the National Institutes of Health and other Government agencies and health-related organizations.

Website: https://www.medlineplus.gov

U.S. Food and Drug Administration (FDA)

The FDA oversees the safety of many products, such as foods, medicines, dietary supplements, medical devices, and cosmetics. See its webpage on Dietary Supplements.

Website: https://www.fda.gov/

Center for Food Safety and Applied Nutrition (CFSAN)

Part of the FDA, CFSAN oversees the safety and labeling of supplements, foods, and cosmetics. It provides information on dietary supplements. Online consumer resources include *Tips for Dietary Supplement Users: Making Informed Decisions and Evaluating Information.*

LINKS TO OTHER HEALTH ORGANIZATIONS

National Institutes of Health (NIH)

- National Library of Medicine (NLM)
    - Herbs and Supplements, A–Z
    - MedlinePlus Complementary and Integrative Medicine Page
    - MedlinePlus Herbal Medicine Page
- Office of Dietary Supplements (ODS)
    - Frequently Asked Questions About Dietary Supplements
- Agency for Healthcare Research and Quality (AHRQ)
    - Safety of Vaccines Used for Routine Immunization in the United States
- Food and Drug Administration (FDA)
    - Dietary Supplements
    - FoodSafety.gov
    - Quick Tips for Buying Medicines Over the Internet
    - Information for Consumers on Using Dietary Supplements

Peer Review Ponderings

Appendix G

Evaluating Online Health Information

To Evaluate Health Information on the Internet

The internet makes finding health information easy and fast. Much of the information on the internet is valuable; however, the internet also allows rapid and widespread distribution of false and misleading information. You should carefully consider the source of information you find on the internet and discuss that information with your healthcare provider. This fact sheet can help you decide whether the health information you find online or receive via email, text, or social media is reliable.

Who runs the site?

Online resources such as web and social media sites should make it easy to learn who is responsible for the site and its information. For example, on the Office of Dietary Supplements (ODS) website, the ODS is clearly noted on every significant page, along with a link to the site's homepage.

Who pays for the site?

It costs money to run web and social media sites. A website's funding source should be clearly stated or readily apparent. For example, the U.S. government funds websites with addresses ending in ".gov," educational institutes maintain ".edu" sites, noncommercial organizations 'addresses often use ".org," and ".com" denotes a commercial organization. A site's source of funding can affect the content it presents, how it presents that content, and what the owner wants to accomplish on the site.

What is the site's purpose?

The person or organization that runs a website and the site's funding sources determine the site's purpose. Many websites have a link to information about the site, often called "About This Site." This webpage should clearly state the site's purpose and help you evaluate the trustworthiness of the site's information. Although many legitimate websites sell health and medical products, remember that the website owner's desire to promote a product or service can influence the accuracy of their health information. Looking for another source of health information that is independent and unbiased can help you validate the accuracy of the material presented on a site.

What is the original source of the site's information?

Many health and medical websites post information the owner has collected from other websites or sources. Suppose the person or organization in charge of the site did not write the material. In that case, they should clearly identify the original source. For social media sites, it is also essential to consider whether the source of the information is reliable; that is, what is the background and expertise of the person posting the content?

How does the site document its information?

Websites should identify the medical and scientific evidence that supports the material presented on the site. Medical facts and figures should have references (such as citations of articles published in medical journals). Also, opinions or advice should be clearly set apart from "evidence-based" information (based on research results). Testimonials from people who said they have tried a particular product or service are not evidence-based and usually cannot be corroborated.

Who reviewed the information before the owner posted it on the site?

Health-related websites should give information about the medical credentials of the people who prepared or reviewed the material on the website. For example, the ODS website contains fact sheets about vitamins, minerals, and other dietary supplements. These documents undergo extensive scientific review by recognized experts from the academic and research communities.

How current is the information on the site?

Experts should review and update the material on websites regularly. Medical information must be current because medical research is constantly coming up with new information about medical conditions and how best to treat or prevent them. Sites should clearly post the most recent update or review date. Even if the information has not changed in a long time, the site owner should indicate that someone has reviewed it recently to ensure it is still valid.

How does the site owner choose links to other sites?

Owners of reliable websites usually have a policy governing which links to other sites they post. Some medical websites take a conservative approach and do not provide links to other sites; some sites provide links to any site that asks or pays for a link, and others provide links only to sites that have met specific criteria. Checking a website's linking policy can help you understand how they choose links to other sites and what they're trying to accomplish by posting those links.

What information about users does the site collect, and why?

Websites routinely track the path users take through their sites to determine what pages people are viewing. However, many health-related websites also ask users to “subscribe” to or “become a member” of the site. Sites sometimes do this to collect a user fee or select relevant information for the user. The subscription or membership might allow the website owner to collect personal information about the user.

Any web or social media site asking you for personal information should explain exactly what the site will and will not do with the information. Many commercial sites sell “aggregate” data—such as what percent of their users take dietary supplements—about their users to other companies. Sometimes, sites collect and reuse " personally identifiable information,” such as your ZIP code, gender, and birth date. Be sure to read and understand any privacy policy or similar language on the site. Do not sign up for anything you do not fully understand.

How does the site manage interactions with users?

Web and social media sites should always offer a way for users to contact the site owner with problems, feedback, and questions. If the site hosts a chat room or online discussion, it should explain the service's terms. For example, the site should explain whether anyone moderates the forum and, if so, who provides the moderation and what criteria the moderator uses to determine which comments to accept and which to reject. Always read online discussions before participating to ensure you are comfortable with the discussion and what participants say to one another.

How can you verify the accuracy of information you receive via email/text?

Carefully evaluate any email or text messages you receive that provide health-related information. Consider the message’s origin and purpose. Some companies or organizations use email or text to advertise products or attract people to their websites. A critical eye is warranted if an individual or company promotes a particular medical product or service in an email or text without supporting medical evidence.

Peer Review Ponderings

Peer Review Ponderings

Appendix H

Preparing for a Disability Claim

SOCIAL SECURITY DISABILITY CLAIM

Failure to complete this form in its entirety may result in a delay in processing this claim.

APPROVED

Disability Evaluation Under Social Security Section 8.00 Skin Disorders - Adult

Author's Note: I stumbled upon the following document on the Social Security Administration website and thought it might be a good place for PRP paatients, whether they live in the US start pondering a disability claim. For non-residents, the process by which a claim is made and justified would be useful in dealing withn any international jurisdiction.

The most critical evaluators are those from whom you want financial support.

Social Security provides financial protection for our nation's people, supporting Americans throughout all of life's journeys.

They administer retirement, disability, survivor, and family benefits, and enroll individuals in Medicare. They also determine eligibility for certain government services, e.g., disability claims for U.S. citizens diagnosed with pityriasis rubra pilaris. At least that's what we hope.

Section 8.00 Skin Disorders Adult

A. *What skin disorders do we evaluate with these listings?*

We use these listings to evaluate skin disorders that may result from hereditary, congenital, or acquired pathological processes. The kinds of impairments covered by these listings are: Ichthyosis, bullous diseases, chronic infections of the skin or mucous membranes, dermatitis, hidradenitis suppurativa, genetic photosensitivity disorders, and burns.

B. *What documentation do we need?*

When we evaluate the existence and severity of your skin disorder, we generally need information about the onset, duration, frequency of flare-ups, and prognosis of your skin disorder; the location, size, and appearance of lesions; and, when applicable, history of exposure to toxins, allergens, or irritants, familial incidence, seasonal variation, stress factors, and your ability to function outside of a highly protective environment.

To confirm the diagnosis, we may need laboratory findings (for example, results of a biopsy obtained independently of Social Security disability evaluation or blood tests) or evidence from other medically acceptable methods consistent with the prevailing state of medical knowledge and clinical practice.

C. How do we assess the severity of your skin disorder(s)?

We generally base our assessment of severity on the extent of your skin lesions, the frequency of flare-ups of your skin lesions, how your symptoms (including pain) limit you, the extent of your treatment, and how your treatment affects you.

1. *Extensive skin lesions.*

a) Extensive skin lesions are those that involve multiple body sites or critical body areas, and result in a very serious limitation. Examples of extensive skin lesions that result in a very serious limitation include but are not limited to:

b) Skin lesions that interfere with the motion of your joints and that very seriously limit your use of more than one extremity; that is, two upper extremities, two lower extremities, or one upper and one lower extremity.

c) Skin lesions on the palms of both hands that very seriously limit your ability to do fine and gross motor movements.

d) Skin lesions on the soles of both feet, the perineum, or both inguinal areas that very seriously limit your ability to ambulate.

2. *Frequency of flare-ups.*

a) If you have skin lesions, but they do not meet the requirements of any of the listings in this body system, you may still have an impairment that prevents you from doing any gainful activity when we consider your condition over time, especially if your flare-ups result in extensive skin lesions, as defined in C1 of this section.

b) Therefore, if you have frequent flare-ups, we may find that your impairment(s) is medically equal to one of these listings even though you have some periods during which your condition is in remission.

c) We will consider how frequent and serious your flare-ups are, how quickly they resolve, and how you function between flare-ups to determine whether you have been unable to do any gainful activity for a continuous period of at least 12 months or can be expected to be unable to do any gainful activity for a continuous period of at least 12 months.

d) We will also consider the frequency of your flare-ups when we determine whether you have a severe impairment and when we need to assess your residual functional capacity.

3. *Symptoms (including pain).*

Symptoms (including pain) may be important factors contributing to the severity of your skin disorder(s). We assess the impact of symptoms as explained in §§ 404.1521, 404.1529, 416.921, and 416.929 of this chapter.

4. *Treatment.*

We assess the effects of medication, therapy, surgery, and any other form of treatment you receive when we determine the severity and duration of your impairment(s). Skin disorders frequently respond to treatment; however, response to treatment can vary widely, with some impairments becoming resistant to treatment. Some treatments can have side effects that can in themselves result in limitations

a. We assess the effects of continuing treatment as prescribed by determining if there is improvement in the symptoms, signs, and laboratory findings of your disorder, and if you experience side effects that result in functional limitations. To assess the effects of your treatment, we may need information about:

i. The treatment you have been prescribed (for example, the type, dosage, method, and frequency of administration of medication or therapy);

ii. Your response to the treatment;

iii. Any adverse effects of the treatment; and

iv. The expected duration of the treatment.

b. Because treatment itself or the effects of treatment may be temporary, in most cases sufficient time must elapse to allow us to evaluate the impact and expected duration of treatment and its side effects. Except under 8.07 and 8.08, you must follow continuing treatment as prescribed for at least 3 months before your impairment can be determined to meet the requirements of a skin disorder listing. (See 8.00H if you are not undergoing treatment or did not have treatment for 3 months.) We consider your specific response to treatment when we evaluate the overall severity of your impairment.

D. *How do we assess impairments that may affect the skin and other body systems?*

When your impairment affects your skin and has effects in other body systems, we first evaluate the predominant feature of your impairment under the appropriate body system. Examples include, but are not limited to the following.

1. *Tuberous sclerosis* primarily affects the brain. The predominant features are seizures, which we evaluate under the neurological listings in 11.00, and developmental delays or other mental disorders, which we evaluate under the mental disorders listings in 12.00.
2. *Malignant tumors of the skin* (for example, malignant melanomas) are cancers, or neoplastic diseases, which we evaluate under the listings in 13.00.
3. *Autoimmune disorders* and other immune system disorders (for example, systemic lupus erythematosus (SLE), scleroderma, human immunodeficiency virus (HIV) infection, and Sjögren's syndrome) often involve more than one body system. We first evaluate these disorders under the immune system disorders listings in 14.00. We evaluate SLE under 14.02, scleroderma under 14.04, Sjögren’s syndrome under 14.10, and HIV infection under 14.11.
4. *Disfigurement or deformity* resulting from skin lesions may result in loss of sight, hearing, speech, and the ability to chew (mastication). We evaluate these impairments and their effects under the special senses and speech listings in 2.00 and the digestive system listings in 5.00. Facial disfigurement or other physical deformities may also have effects we evaluate under the mental disorders listings in 12.00, such as when they affect mood or social functioning.

E. *How do we evaluate genetic photosensitivity disorders?*

1. Xeroderma pigmentosum (XP). When you have XP, your impairment meets the requirements of 8.07A if you have clinical and laboratory findings showing that you have the disorder. (See 8.00E3.) People who have XP have a lifelong hypersensitivity to all forms of ultraviolet light and generally lead extremely restricted lives in highly protective environments in order to prevent skin cancers from developing. Some people with XP also experience problems with their eyes, neurological problems, mental disorders, and problems in other body systems.

2. *Other genetic photosensitivity disorders.*

Other genetic photosensitivity disorders may vary in their effects on different people, and may not result in an inability to engage in any gainful activity for a continuous period of at least 12 months. Therefore, if you have a genetic photosensitivity disorder other than XP (established by clinical and laboratory findings as described in 8.00E3), you must show that you have either extensive skin lesions or an inability to function outside of a highly protective environment to meet the requirements of 8.07B.

You must also show that your impairment meets the duration requirement. By inability to function outside of a highly protective environment we mean that you must avoid exposure to ultraviolet light (including sunlight passing through windows and light from unshielded fluorescent bulbs), wear protective clothing and eyeglasses, and use opaque broad spectrum sunscreens in order to avoid skin cancer or other serious effects. Some genetic photosensitivity disorders can have very serious effects in other body systems, especially special senses and speech (2.00), neurological (11.00), mental (12.00), and neoplastic (13.00). We will evaluate the predominant feature of your impairment under the appropriate body system, as explained in 8.00D.

3. Clinical and laboratory findings.

a. *General*. We need documentation from an acceptable medical source to establish that you have a medically determinable impairment. In general, we must have evidence of appropriate laboratory testing showing that you have XP or another genetic photosensitivity disorder. We will find that you have XP or another genetic photosensitivity disorder based on a report from an acceptable medical source indicating that you have the impairment, supported by definitive genetic laboratory studies documenting appropriate chromosomal changes, including abnormal DNA repair or another DNA or genetic abnormality specific to your type of photosensitivity disorder.

b. *What we will accept as medical evidence instead of the actual laboratory report*. When we do not have the actual laboratory report, we need evidence from an acceptable medical source that includes appropriate clinical findings for your impairment and that is persuasive that a positive diagnosis has been confirmed by appropriate laboratory testing at some time prior to our evaluation. To be persuasive, the report must state that the appropriate definitive genetic laboratory study was conducted and that the results confirmed the diagnosis. The report must be consistent with other evidence in your case record.

F. *How do we evaluate burns?*

Electrical, chemical, or thermal burns frequently affect other body systems; for example, musculoskeletal, special senses and speech, respiratory, cardiovascular, renal, neurological, or mental. Consequently, we evaluate burns the way we evaluate other disorders that can affect the skin and other body systems, using the listing for the predominant feature of your impairment. For example, if your soft tissue injuries are under continuing surgical management (as defined in 1.00M), we will evaluate your impairment under 1.08. However, if your burns do not meet the requirements of 1.08 and you have extensive skin lesions that result in a very serious limitation (as defined in 8.00C1) that has lasted or can be expected to last for a continuous period of at least 12 months, we will evaluate them under 8.08.

G. *How do we determine if your skin disorder(s) will continue at a disabling level of severity in order to meet the duration requirement?*

For all of these skin disorder listings except 8.07 and 8.08, we will find that your impairment meets the duration requirement if your skin disorder results in extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed. By persist, we mean that the longitudinal clinical record shows that, with few exceptions, your lesions have been at the level of severity specified in the listing. For 8.07A, we will presume that you meet the duration requirement. For 8.07B and 8.08, we will consider all of the relevant medical and other information in your case record to determine whether your skin disorder meets the duration requirement.

H. *How do we assess your skin disorder(s) if your impairment does not meet the requirements of one of these listings?*

1. These listings are only examples of common skin disorders that we consider severe enough to prevent you from engaging in any gainful activity. For most of these listings, if you do not have continuing treatment as prescribed, if your treatment has not lasted for at least 3 months, or if you do not have extensive skin lesions that have persisted for at least 3 months, your impairment cannot meet the requirements of these skin disorder listings. (This provision does not apply to 8.07 and 8.08.) However, we may still find that you are disabled because your impairment(s) meets the requirements of a listing in another body system or medically equals the severity of a listing. (See §§ 404.1526 and 416.926 of this chapter.) We may also find you disabled at the last step of the sequential evaluation process.

2. If you have not received ongoing treatment or do not have an ongoing relationship with the medical community despite the existence of a severe impairment(s), or if your skin lesions have not persisted for at least 3 months

but you are undergoing continuing treatment as prescribed, you may still have an impairment(s) that meets a listing in another body system or that medically equals a listing. If you do not have an impairment(s) that meets or medically equals a listing, we will assess your residual functional capacity and proceed to the fourth and, if necessary, the fifth step of the sequential evaluation process in §§ 404.1520 and 416.920 of this chapter. When we decide whether you continue to be disabled, we use the rules in §§ 404.1594 and 416.994 of this chapter.

8.01 Category of Impairments, Skin Disorders

8.02 *Ichthyosis,* with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.03 *Bullous disease* (for example, pemphigus, erythema multiforme bullosum, epidermolysis bullosa, bullous pemphigoid, dermatitis herpetiformis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed. .

8.04 *Chronic infections of the skin or mucous membranes,* with extensive fungating or extensive ulcerating skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.05 *Dermatitis* (for example, psoriasis, dyshidrosis, atopic dermatitis, exfoliative dermatitis, allergic contact dermatitis), with extensive skin lesions that persist for at least 3 months despite continuing treatment as prescribed.

8.06 *Hidradenitis suppurativa,* with extensive skin lesions involving both axillae, both inguinal areas or the perineum that persist for at least 3 months despite continuing treatment as prescribed.

Peer Review Ponderings