In Primary Periodic Paralysis (PPP)

BE YOUR FAMILY FACILITATOR

Talking about PPP with your family can be hard.
This guide can help you start the conversation.

STRONGBRIDGE BIOPHARMA®

Sharing your experience can empower your family

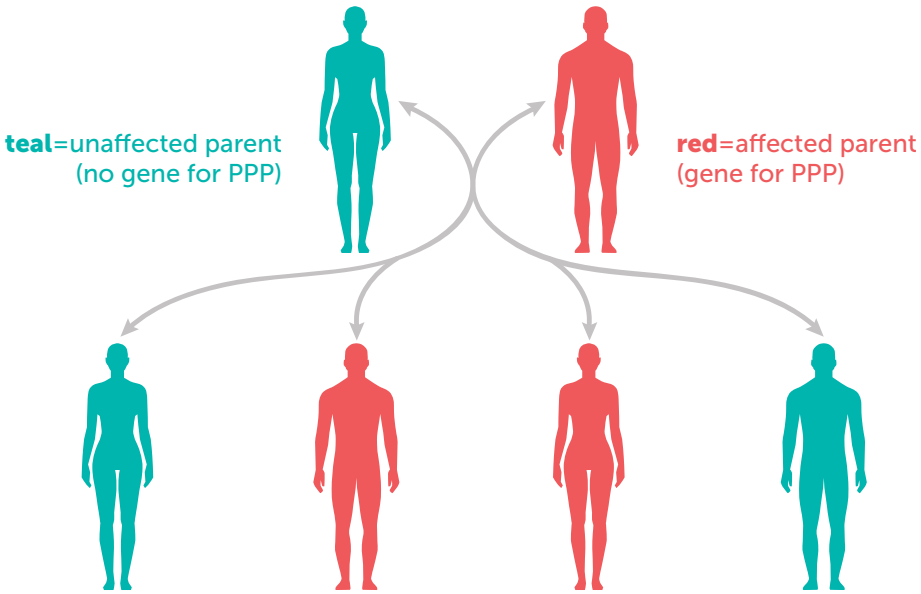
PPP is often an inherited condition.[1] Your family members may not know they have PPP, or, if they are experiencing symptoms, may not realize they could be caused by PPP.[1,2]

Think back to how you felt when you were finally diagnosed with PPP. At last, you had an answer for why you were feeling the way you were feeling—and a path forward.

As your Family Facilitator, you can help guide your family members' paths by encouraging them to discuss the possibility of PPP with their doctors.

What causes PPP?

PPP is a rare condition caused by a change, called a *mutation*, in your genes. If someone has a gene that causes PPP, there is a 50% chance that each of his or her children will inherit that gene, too.[3]



It affects your muscles and can lead to episodes of muscle weakness and/or temporary paralysis.[1]

Some people with PPP develop Permanent Muscle Weakness (PMW), meaning that weakness does not go away in between episodes.[4] PMW is most common in people with PPP who are between the ages of 40 and 50.[2,4]

Talking to a family member about PPP

Since you know that PPP is often a genetic condition, you might be worried that some of your family members may also have this condition. This is a sensitive topic, and while you don't want to frighten anyone, you do want to pass along facts and useful information about PPP.

Talking openly about PPP is a brave decision. By sharing your own knowledge and experience, you can act as a source of information and inspiration. You can also help remove the stigma from this rare, often-misunderstood condition.

Planning the conversation: know what you want to say and how to say it

Think about what you want to say. Imagine if you didn't know you had PPP, but there was a chance you could be living with it, or could pass it on to your children. What would you wish someone had told you?

Also, consider your family member's preferences—would he or she prefer to have the conversation in person or over the phone? Alone or with the support of other family members?

Starting the conversation: keep it nice and simple

Start off on a positive note by letting your family member know why you wanted to have this conversation in the first place: ***to empower them with knowledge.***

> ***"I'd like to talk with you about a genetic condition I have—is that okay with you? I think this information will be important for you and the rest of our family, too."***

Tips for a productive conversation

The information in this brochure, along with the family worksheet, can help you have a conversation about PPP. And, to help make the conversation go more smoothly, remember to stick to the **FACTS**:

F

Family connection and history

Discuss the genetic nature of PPP.[1] Your family member should know that he or she, along with other family members, may have also inherited a gene that can cause PPP.

A

Answering questions

This may be the first time your family member has heard of PPP, and he or she will probably have questions. Do your best to answer honestly, sharing both your challenges and your triumphs.

C

Completing the family worksheet

After you fill out the family worksheet, you can share it with your family member, or even complete it with them. You may notice patterns in your family, which could lead to a more productive conversation with a doctor.

› *See page 5 for the family worksheet.*

T

Talking to a doctor

Encourage your family member to talk to a doctor who is familiar with PPP about any symptoms he or she may be experiencing, so that his or her doctor can consider a diagnosis of (and testing for) PPP, and begin treatment, if appropriate.

› *Find a doctor who diagnoses and treats PPP at* **pppdocfinder.com**.

S

Support resources

Set your family up to educate themselves by pointing them to resources they can explore to learn more about PPP.

› *Join in on social media:*

@PavingMyPPPath **@pavingmypppath** **@PavingMyPPPath**

› *Visit* **inspire.com/groups/primary-periodic-paralysis** *to join a community of people with PPP.*

PPP family history worksheet

To complete the worksheet, think about if any of your relatives had a confirmed diagnosis of PPP or if they have experienced episodes of paralysis or weakness after exposure to a known trigger of PPP, such as[1]:

Foods or beverages high in salt, carbohydrates, or potassium

Stress or tiredness

Becoming cold

Periods of inactivity

Resting after exercise

Also, write down any additional information you feel is relevant. If you're unsure of what to write, the full worksheet has thought starters for you to consider.

Once you're finished, you can bring the worksheet along to the doctor's office to discuss your family history at appointments.

Examples

Grandfather

- [x] Confirmed PPP diagnosis
- [x] Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Started using a cane around age 40, later moved to a wheelchair after several injuries from falls

Child

- [ ] Confirmed PPP diagnosis
- [x] Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Experienced episodes of muscle weakness after soccer games

PPP family history worksheet

This worksheet can help you and your family members track your family history, which is important information for a doctor to have when diagnosing PPP.[3]

"In 2010, I finally began connecting more dots. A wellness physician ...was thinking Parkinson's or multiple sclerosis, so when asked about my family history, I told her no one in my family had anything like that. Later, I would piece together through ancestry and asking questions that my mother, aunt, and cousin all seemed to have PPP."

– **Teresa,** Strongbridge Patient Ambassador

Notes

Use this space to keep track of any additional notes and information from conversations with your family members.

Consider the following when filling out the additional information section of this worksheet:

- Have any of your family members experienced injuries in their 40s and 50s from falls due to symptoms of weakness or paralysis?
- Have any of your family members experienced permanent weakness in their 40s and 50s, which resulted in use of a cane, wheelchair, scooter, or walker?

These symptoms, in combination with others, could be signs of **Permanent Muscle Weakness**.[4]

Also, consider:

- Do you know how old these family members were when they started experiencing episodes of weakness or paralysis?

"Although none of my children have been officially diagnosed yet, knowing they may end up having Primary Periodic Paralysis weighs heavy on my heart. Thankfully, however, they will be armed with information and management options I didn't have."

– **Stefanie,**
Strongbridge Patient Ambassador

Grandmother

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Grandfather

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Aunt/Uncle

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Mother/Father

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Aunt/Uncle

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Sister/Brother

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

You

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Sister/Brother

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Child

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Child

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

Child

- ☐ Confirmed PPP diagnosis
- ☐ Had episodes of muscle weakness or paralysis after exposure to triggers

Additional information:

How is PPP diagnosed?

A doctor can review family history, complete physical examinations, or perform other tests to confirm a diagnosis of PPP.[1,5] These tests include:

- Electrocardiogram (ECG/EKG)[1]
- Electromyography (EMG)[1]
- Compound muscle action potential (CMAP)[5]

Doctors may also order genetic testing.[5] If the test comes back positive, a diagnosis of PPP can be confirmed.[5] A negative genetic test result does not always rule out a diagnosis of PPP, though. About **30%** of people with PPP do not have one of the common genetic mutations associated with this condition.[1]

Family history is very important when considering a diagnosis of PPP.[2]

Meet **Kim**—who was diagnosed because of a family history of PPP

Kim's father was diagnosed with PPP when he was only 12 years old, so he immediately suspected PPP when he found Kim lying motionless in her crib at 2 years old. Although he took her to the hospital right away, Kim's doctors weren't able to diagnose her at the time.

She went without a diagnosis until she was 17, when she went to another doctor's appointment. Because of her father's experience with PPP, Kim was able to identify her triggers and start a conversation with the doctor that ultimately led to her being diagnosed with PPP and getting started on a treatment.

"It wasn't until I started hearing other people's stories—just like mine—that I realized we aren't alone in this. I learned the basics from my doctor, and that's important, but through other people, I've learned how to live a better life. Truly, we're in this together."

– Kim, Strongbridge Patient Ambassador

You can make a difference by being your Family Facilitator.

Talk to your family about PPP.

Support and educational resources are available

Sign up for more information about PPP at **PavingMyPath.com.**

"You may not feel like you can walk the difficult path of Primary Periodic Paralysis by yourself. Know that you don't have to. We can walk together until you feel ready to walk forward on your own. Then it will be your turn to help others as well."

– **John,** Strongbridge Patient Ambassador

References: 1. Statland JM, Fontaine B, Hanna MG, et al. Review of the diagnosis and treatment of periodic paralysis. *Muscle Nerve*. 2018;57:522-530. **2.** Charles G, Zheng C, Lehmann-Horn F, Jurkat-Rott K, Levitt J. Characterization of hyperkalemic periodic paralysis: a survey of genetically diagnosed individuals. *J Neurol*. 2013;260:2606-2613. **3.** Fontaine B, Phillips LH, II. A newly approved drug for a rare group of diseases: dichlorphenamide for periodic paralysis. *Neurology*. 2016;86:1366-1367. **4.** Cavel-Greant D, Lehmann-Horn F, Jurkat-Rott K. The impact of permanent muscle weakness on quality of life in periodic paralysis: a survey of 66 patients. *Acta Myol*. 2012;31:126-133. **5.** Weber F, Jurkat-Rott K, Lehmann-Horn F. Hyperkalemic Periodic Paralysis. GeneReviews®. NCBI Bookshelf. A service of the National Library of Medicine, National Institutes of Health.

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