

Talking with your doctor



Being prepared can be a source of strength as you manage pyruvate kinase (PK) deficiency. Keeping track of goals, obstacles, and questions can help make sure you and your healthcare team come up with the right plan.

STEP 1: Get organized

If you haven't been keeping track of your condition in a journal, a good place to start is to gather any information you may have, such as:

- Surgeries
- Transfusion history
- Blood or other lab tests
- Any other medical conditions
- Medicines or supplements you take on a regular basis

Consider keeping track of your symptoms on a weekly basis. These data can help your healthcare team understand which symptoms you have and how they affect you. If you undergo regular transfusions, for example, it may help to show what kind of effect they have, and how long those effects last.

STEP 2: Talk about daily life

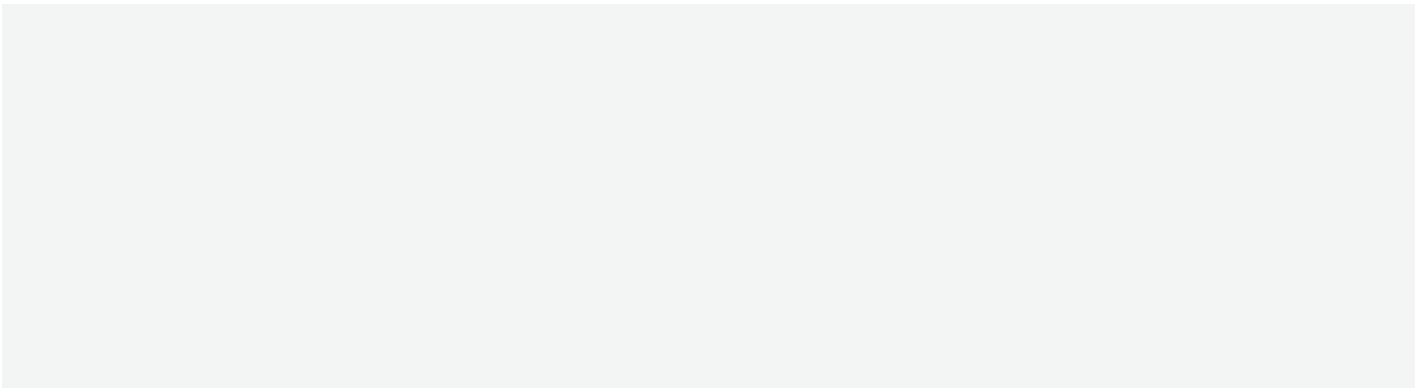
Talking in detail about your life with PK deficiency can help your healthcare team get a better idea of how you feel.

To help you be specific at your next appointment, try completing these prompts.

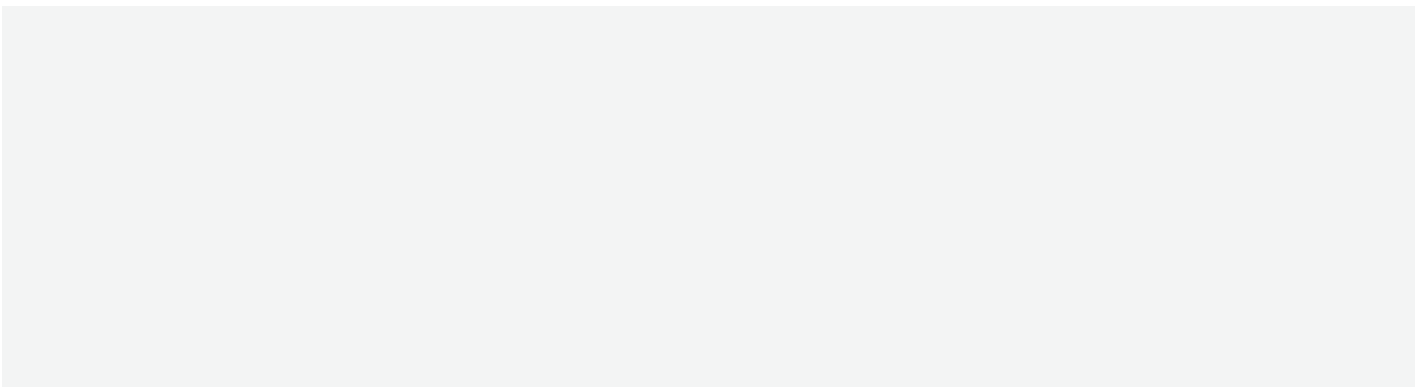
- I enjoy the following activities/hobbies, but I can't do as much as I want to

These steps and examples of discussion questions can help you prepare for your next appointment. Symptoms can affect people’s lives differently, so speaking from your own experiences will be best to help your doctor understand how PK deficiency affects you.

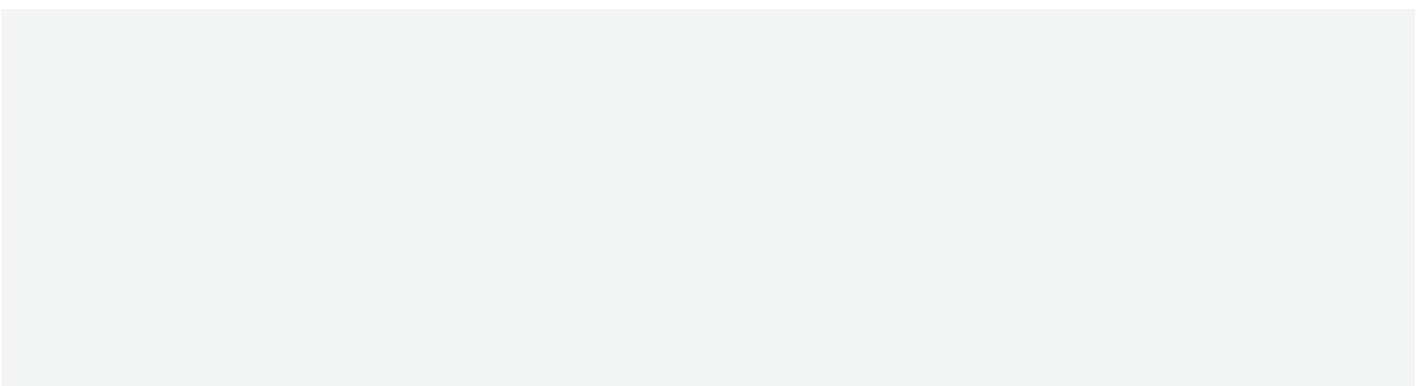
- The symptoms that most interfere with my daily life are



- I notice my symptoms are worse during this part of my day

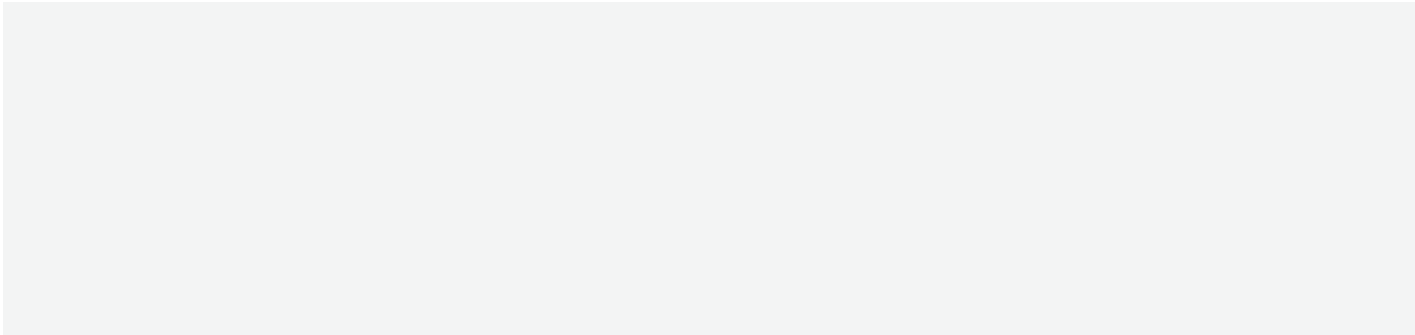


- The symptom that bothers me the most and why is



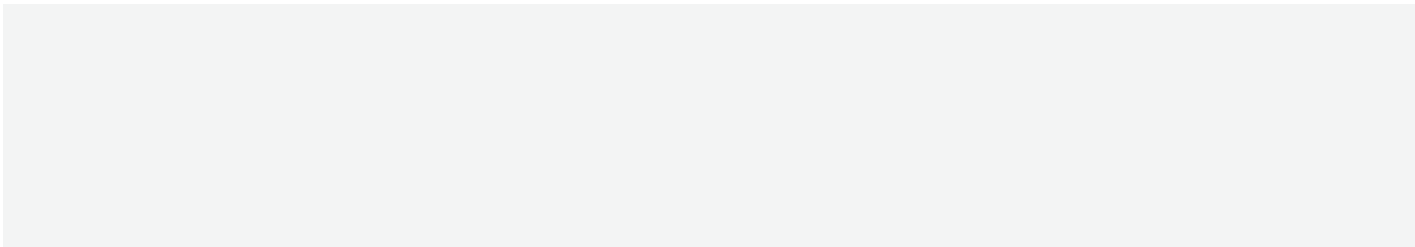
STEP 2 continued

It may be helpful to describe how the symptoms affect your life directly; for example:

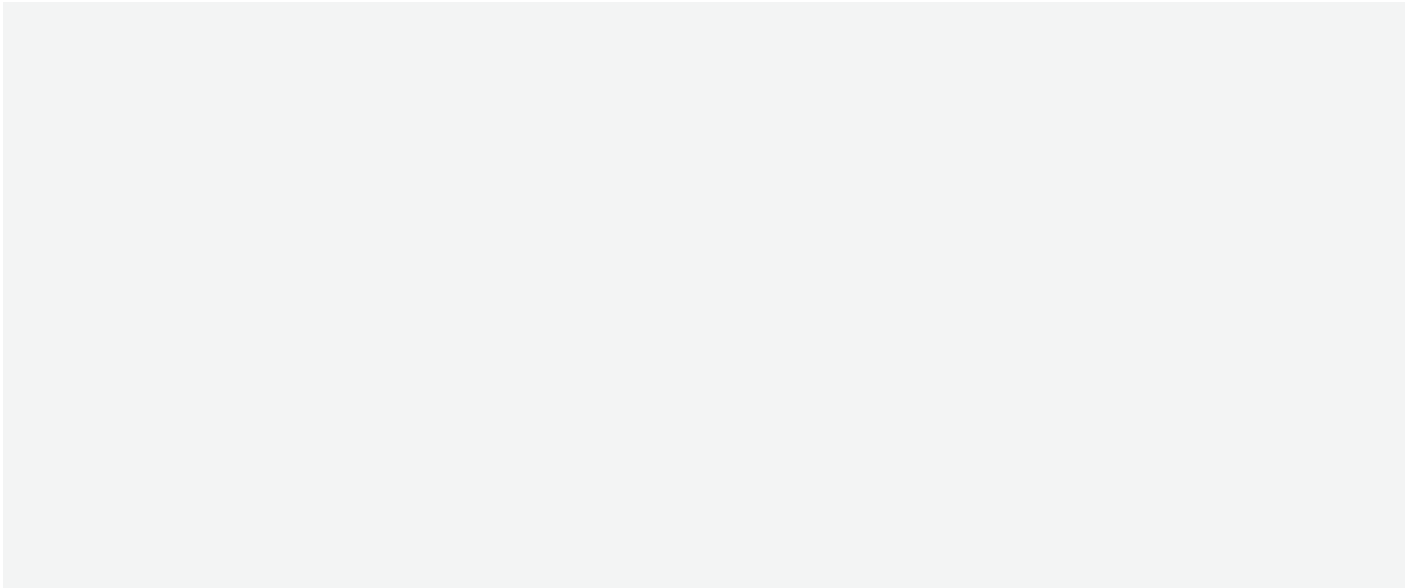
- I always get a headache in the late afternoon so, instead of spending time in my garden, I have to lie down
 - I want to be able to attend my child's school events, but can't find the energy to leave the house after 5 PM
 - Sometimes I can't finish my homework because my brain gets foggy and reading my textbook becomes a struggle
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STEP 3: Make connections

Talk to your hematologist about how frequently your PK deficiency needs to be monitored. Think about these conversation starters and make note of other topics you want to discuss:

- I've read that tests to monitor for PK deficiency complications can vary in frequency depending on transfusions, the need for other therapies, and discoveries from previous assessments. Based on my history, can we discuss how often we should be testing?
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- Some assessments become more relevant at a certain age, such as a dual-energy X-ray absorbitometry (DXA) scan to monitor osteoporosis and an echo to monitor pulmonary hypertension. When should we start thinking about these tests for me?



Iron overload can occur in people with PK deficiency regardless of age, transfusion history, or degree of anemia. It may be a topic worth discussing on its own. Some prompts to start a conversation include:

- I came across research that suggests people with PK deficiency should be monitored regularly for iron overload and its complications. Recent research shows that iron overload can be a concern when ferritin levels are high (greater than 500 nanograms per milliliter). How are my ferritin levels?
- I also learned that people who undergo regular transfusions, or who are on chelation therapy, may need to have their ferritin levels tested more often. What do you think about testing my ferritin levels more frequently, like every 3 or 6 months instead of every year?

STEP 4: Advocate for yourself

When discussing support options, it can be helpful to mention the methods you've learned about in your own research. You may even find resources that are new to your physician. Consider sharing a resource and any questions you have about its content.

Talking about your specific situation along with the hematologist's recommendations can help you both create an effective plan.

See some examples of discussion points below and add in your own.

- I want to be proactive about my PK deficiency. Can we talk about managing my condition and build a plan together?

- Based on my symptoms, what else can we do to manage my anemia?
(You can also use our [symptom tool](#) on KnowPKdeficiency.com to create a list.)

- Some of my symptoms are affecting my daily life. What changes in my management plan can we consider based on what I'm still feeling?

- I've read that transfusions or a splenectomy could help my anemia and fatigue. It would be helpful to talk through the added risks and benefits of these options together, as well as what they might mean for me.

- I find my condition to be overwhelming sometimes, and think I may benefit from more support. Are there any groups recommended for people living with a rare condition?

- My yellow skin has started to take a toll on my self-esteem. What else can we do to reduce my bilirubin levels?

STEP 5: Set goals

Be open about the plans you have for the future. Think about what you want to do next, and ask your healthcare team if they can help you get there. Plan ahead by considering these prompts:

- My school and career plans include

- I have events with family/friends that I want to attend

- I want to be able to participate in the following activities or hobbies



The [Resources](#) section on [KnowPKDeficiency.com](#) includes additional information, including [Pyruvate Kinase Deficiency Through the Decade](#). This resource includes summaries of journal articles that may help support conversations with your doctor. Consider sharing the title and author of the article, or the journal article itself.

Discussion topics might include:

- I found an article focusing on the burden of PK deficiency, and it mentioned some common symptoms that I've been noticing in myself, like memory loss and difficulty concentrating. I don't want this to further affect my career/schoolwork/hobbies/family life. What else can we consider to help?
- I read a study that talked about bone fragility, and a key finding was that long-term complications should be monitored regularly, since there are few early predictors. What can we do to proactively support my bone health?



Focus on your future with PK deficiency

Moving forward, with the right information

Strong communication and a partnership with your hematologist can ensure you're getting the most out of every appointment and management method.

- Organize information to share with your doctor based on what you've learned in your own research
- Keep track of your symptoms over time, so you can give your doctor a clear picture of your health
- Make a list of any questions ahead of time, so you don't forget to ask
- Take notes during discussions with your doctor



You have tools for a productive and effective conversation about moving forward with PK deficiency. Managing PK deficiency is a process that will evolve over time. Building a strong relationship with the right hematologist can make sure you're on track, every step of the way.