What is the most important information I should know about Palynziq?

Palynziq can cause a severe allergic reaction (anaphylaxis) that may be life-threatening and can happen any time during treatment with Palynziq. Severe allergic reactions are a serious but common side effect of Palynziq.

- You will receive your first injection of Palynziq in a healthcare setting where you will be closely watched for at least 1 hour after your injection for a severe allergic reaction.
- If you have a severe allergic reaction during treatment with Palynziq, you will need to receive an auto-injection of epinephrine immediately and get emergency medical help right away.
- Your healthcare provider will decide if you (or your caregiver) are able to give the Palynziq injections, recognize the signs and symptoms of a severe allergic reaction, give an injection of epinephrine and call for emergency medical help, if needed.
- Your healthcare provider may recommend that an adult observer (or your caregiver) be with you when you give your Palynziq injection, and for at least 1 hour after your injection to watch you for signs and symptoms of a severe allergic reaction and, if needed, give you an injection of epinephrine and call for emergency medical help.

Stop injecting Palynziq and get emergency medical care right away if you have any of the following symptoms of a severe allergic reaction during treatment with Palynziq:

- fainting (passing out)
- dizziness or lightheadedness
- sudden confusion
- trouble breathing or wheezing
- chest discomfort or chest tightness
- fast heart rate
- swelling of your face, lips, eyes, or tongue
- throat tightness
- flushed skin
- skin rash, itching, or raised bumps on skin
- nausea, vomiting, or diarrhea
- losing control of urine or stools

Your healthcare provider will prescribed an auto-injectable epinephrine for you and will teach you (or your caregiver) and your observer, if needed, when and how to use it if you have a severe allergic reaction. Keep the auto-injectable epinephrine with you at all times during treatment with Palynziq. Read the Patient Information that comes with the auto-injectable epinephrine that your healthcare provider prescribes for you for more information.

If you have a severe allergic reaction, do not continue to take Palynziq until you talk with your healthcare provider. Tell your healthcare provider that you had a severe allergic reaction. Your healthcare provider will tell you if you can continue treatment with Palynziq.

Your healthcare provider may prescribe other medicines to take before your Palynziq injection that may help reduce the symptoms of an allergic reaction.

If your healthcare provider decides that you can continue treatment with Palynziq after a severe allergic reaction, you will receive your next injection of Palynziq in a healthcare setting where you will be closely watched for at least 1 hour after your injection for a severe allergic reaction.

Your healthcare provider will give you a Palynziq Patient Wallet Card that describes symptoms that you (or your caregiver), or your observer, should know that require you to get emergency medical care right away. Carry this card with you at all times during treatment with Palynziq. It is important to show your Palynziq Patient Wallet Card to any other healthcare provider who treats you.

Palynziq is only available through a restricted program called the Palynziq Risk Evaluation and Mitigation Strategy (REMS) Program. Before you can receive Palynziq, you must:

- enroll in this program.
- receive education about the risk of a severe allergic reaction (anaphylaxis) by a healthcare provider certified in the Palynziq REMS to be sure that you understand the risks and benefits of treatment with Palynziq.
- fill the prescription your healthcare provider gives you for the auto-injectable epinephrine and carry it with you at all times during treatment with Palynziq.
- carry the Palynziq Patient Wallet Card with you at all times.

Talk to your healthcare provider for more information about the Palynziq REMS and how to enroll.

What is Palynziq?

Palynziq is a prescription medicine used to lower blood levels of phenylalanine in adults with PKU (phenylketonuria) who have uncontrolled blood phenylalanine levels above 600 micromol/L (10 mg/dL) on their current treatment.

It is not known if Palynziq is safe and effective in children.
The most common side effects of Palynziq include:

- injection site reactions, such as redness, itching, pain, bruising, rash, swelling, tenderness
- joint pain
- headache
- skin reactions that spread on your skin and last at least 14 days, such as itching, rash, redness
- itching
- nausea
- stomach pain
- mouth and throat pain
- vomiting
- cough
- diarrhea
- feel very tired
- low levels of phenylalanine in your blood

These are not all the possible side effects of Palynziq. Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088.

How should I store Palynziq?

- Store Palynziq in the refrigerator at 36°F to 46°F (2°C to 8°C).
- If needed, you may store Palynziq at room temperature between 68°F to 77°F (20°C to 25°C) for up to 30 days.
  - Write the date that you remove Palynziq from the refrigerator on the carton.
  - If stored at room temperature, do not put Palynziq back in the refrigerator.
- Keep Palynziq in the original carton to protect from light.
- Do not freeze or shake Palynziq.
- Throw away Palynziq if it has been kept at room temperature for 30 days and has not been used, or after the expiration date on the carton, whichever comes first.

Keep Palynziq and all medicines out of the reach of children.

General Information about the safe and effective use of Palynziq.
Medicines are sometimes prescribed for purposes other than those listed in a Medication Guide. Do not use Palynziq for a condition for which it was not prescribed. Do not give Palynziq to other people, even if they have the same symptoms that you have. It may harm them. You can ask your pharmacist or healthcare provider for information about Palynziq that is written for health professionals.

<table>
<thead>
<tr>
<th>What are the ingredients in Palynziq?</th>
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<tbody>
<tr>
<td><strong>Active ingredients:</strong> pegvaliase-pqpz</td>
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<tr>
<td><strong>Inactive ingredients:</strong> sodium chloride, \textit{trans}-cinnamic acid, tromethamine, and tromethamine hydrochloride</td>
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Manufactured by: BioMarin Pharmaceutical Inc. Novato, CA 94949 US License No. 1649
For more information, go to http://www.Palynziq.com/ or call 1-866-906-6100.

This Medication Guide has been approved by the U.S. Food and Drug Administration. Issued: May 2018