

Only for patients prescribed  
Zokinvy™ (lonafarnib) and their caregivers

## ZOKINVY™▼(lonafarnib) Guide for Patients and Caregivers

A guide to Hutchinson-Gilford progeria  
syndrome (Progeria) and processing-  
deficient progeroid laminopathies (PL),  
and your medicine, Zokinvy.

Photo courtesy of The Progeria Research Foundation

### Reporting of side effects

▼ This medicine is subject to additional monitoring. This will allow quick identification of new safety information. You can help by reporting any side effects you may get.

If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in the patient leaflet. You can also report side effects directly via 0800 4565992. By reporting side effects, you can help provide more information on the safety of this medicine.

This medicinal product has been authorised under 'exceptional circumstances'. This means that due to the rarity of the disease it has not been possible to obtain complete information on this medicinal product.








*This booklet is for patients and parents/ caregivers of young people and children who have been prescribed Zokinvy. It must have been recommended by a healthcare professional and examined after reading the patient information leaflet provided with Zokinvy.*

Please see Important Safety Information throughout and on pages 3 and 6, and full Prescribing Information at [Zokinvy.eu](https://www.zokinvy.eu) or <https://www.ema.europa.eu/en/medicines/human/EPAR/zokinvy>

 **Zokinvy** ▼  
(lonafarnib)  
hard capsules 50 mg/75 mg

Job code: PRG-UK-2022000029. Date of preparation: March 2023.

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## Welcome to the Zokinvy™ (lonafarnib) guide for patients and caregivers

In this guide, you will find detailed information on Progeria (also known as Hutchinson-Gilford progeria syndrome, or HGPS) and processing-deficient progeroid laminopathies (PL), and their treatment with Zokinvy.



HGPS and PL, collectively known as progeria, are diseases people are born with.<sup>1-3</sup>



In both diseases, genetic mutations cause harmful proteins to build up within the cells of the body. This results in premature aging.<sup>3</sup>



Your medicine, Zokinvy, is used to treat children and young adults aged 12 months and older with progeria.



Zokinvy capsules are taken twice a day with meals, with flexible options to fit your family member's needs.<sup>4</sup>



The most commonly reported side effects are mild-to-moderate vomiting, diarrhoea, infection and nausea.<sup>4</sup>



The Eiger One Patient Support programme is a dedicated programme created with the help of the European progeria support group, Progeria Family Circle, to support people with progeria and their families and caregivers.

### IMPORTANT SAFETY INFORMATION

Do not take Zokinvy if you are taking:

- a strong or moderate CYP3A inhibitor or inducer
- midazolam
- lovastatin, simvastatin, and atorvastatin

Tell your healthcare provider about all the medicines you take, including prescription and over-the-counter medicines, vitamins, and herbal supplements. Zokinvy and other medicines may affect each other, causing serious side effects. Talk with your healthcare provider before taking any new medicines.

Please see Important Safety Information throughout and on pages 3 and 6, and full Prescribing Information at [Zokinvy.eu](https://www.zokinvy.eu) or <https://www.ema.europa.eu/en/medicines/human/EPAR/zokinvy>

## About Progeria or PL

Hutchinson-Gilford progeria syndrome (HGPS) and processing-deficient progeroid laminopathies (PL) are devastating, ultra-rare, and fatal pediatric diseases that cause dramatically accelerated aging and premature death.<sup>1,2</sup>

HGPS and PL, collectively known as progeria, are caused when the body makes harmful proteins called progerin or progerin-like proteins.<sup>1,2</sup>

These harmful proteins lead to cell damage that resembles the effects of aging.<sup>1,2</sup>

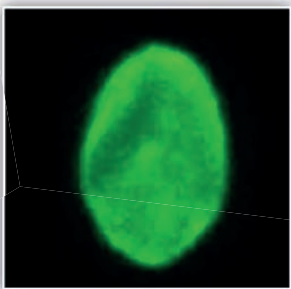
In progeria, these proteins also build up in the walls of large arteries, which can cause them to inflame and thicken. This can cause high blood pressure and heart disease, and can lead to heart attacks.<sup>1,2</sup>

Untreated, most young people and children with progeria die of cardiovascular complications, primarily of heart failure, at an average age of 14.5 years.<sup>5</sup>

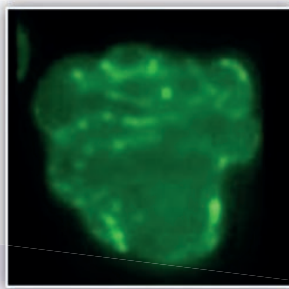
## How your medicine works

Zokinvy (zoh-KIN-vee) is a medicine used to treat young people and children with progeria aged 12 months and older.<sup>4</sup>

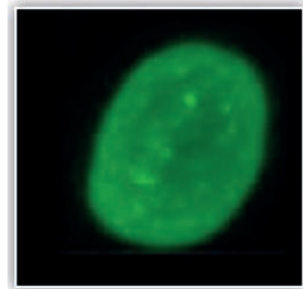
The active drug in Zokinvy is called lonafarnib. It is a disease modifying agent that works by helping to reduce the build-up of harmful progerin or progerin-like proteins.



Healthy cell nucleus.\*



Progeria cell nucleus containing abnormal protein.



Healthy cell nucleus treated with Zokinvy.

\*The cell nucleus is found in the middle of most cells of the human body. It stores the DNA and carries out several jobs to keep cells working normally.

Photo courtesy of The Progeria Research Foundation

As a result, treatment with Zokinvy can increase the life span of young people and children with Progeria.<sup>4,6</sup>

### IMPORTANT SAFETY INFORMATION

**Before taking Zokinvy, tell your healthcare provider about all of your medical conditions, including if you:**

- have kidney problems.
- have eye problems.
- are pregnant or plan to become pregnant. Zokinvy can harm your unborn baby. Women who are able to become pregnant should use effective birth control (contraception) during treatment with Zokinvy.
- are breastfeeding or plan to breastfeed. It is not known if Zokinvy can pass into your breast milk.

Talk to your healthcare provider about the best way to feed your baby during treatment with Zokinvy.

Do not give Zokinvy to children under the age of 12 months because it has not been studied in this age group.

Please see Important Safety Information throughout and on pages 3 and 6, and full Prescribing Information at [Zokinvy.eu](https://www.zokinvy.eu) or <https://www.ema.europa.eu/en/medicines/human/EPAR/zokinvy>



## Taking your medicine

**Always take this medicine exactly as your doctor or pharmacist has told you. Check with your doctor or pharmacist if you are not sure.**

Zokinvy is taken as 1 or 2 capsules twice a day about 12 hours apart (morning and evening). Your doctor will work out the right starting dosage of Zokinvy for you, which is based on your height and weight.<sup>4</sup>

This may mean taking capsules of different strengths to make up the right amount. After 4 months of treatment with Zokinvy, your doctor may increase your dosage.<sup>4</sup>

Make sure you know how many capsules you need to take and the strength of each capsule. Ask your doctor, pharmacist or nurse to write it down (including the colour of the capsule(s) to be taken for each dose).

Take the capsules with food, washing them down with enough water to help you swallow them. Taking Zokinvy with food may help to reduce gastrointestinal side effects.<sup>4</sup>

### If you cannot swallow a Zokinvy capsule whole:

- You can mix the capsule content with orange juice.<sup>4</sup>
- Do not use other drinks to mix Zokinvy.<sup>4</sup>

### Drink lots of water while taking Zokinvy

It is important to drink lots of water and other liquids while taking Zokinvy. This may help to reduce potential dehydration associated with diarrhea or vomiting. Do not eat food or drink juices that contain grapefruit, cranberries, pomegranates or Seville oranges (known as sour or bitter oranges).<sup>4</sup>

### If you take more Zokinvy than you should

If you take more capsules than you should, stop taking the medicine and contact your doctor.<sup>4</sup>

### If you forget to take Zokinvy

If you forget to take a dose and 8 hours or more remains until your next planned dose, take the missed dose as soon as possible with some food. If less than 8 hours remains before the next scheduled dose, skip the missed dose and resume taking Zokinvy at the next scheduled dose.<sup>4</sup>

### If you stop taking Zokinvy

Do not stop taking Zokinvy without talking to your doctor.<sup>4</sup>

*Please carefully read the section of the patient leaflet that discusses how to take your medicine (Section 3: How to take Zokinvy). If you have any concerns, please contact your doctor or pharmacist.*

## Possible side effects

**Zokinvy can cause side effects, although not everybody gets them.**

Some side effects are very common, which means that they may affect more than 1 in 10 people. The most common are vomiting and diarrhoea, although these tend to be mild or moderate and reduce over time.<sup>4</sup>

Other side effects are common, which means they may affect up to 1 in 10 people. They include headache, dehydration and infections.<sup>4</sup>

These are not all of the possible side effects. For a full list of possible side effects, please read Section 4: Possible side effects in the patient leaflet.

If you get any side effects, talk to your doctor, pharmacist or nurse. This includes any possible side effects not listed in the leaflet. If you have any concerns, please contact your doctor or pharmacist.

Zokinvy can cause serious side effects and you should contact your doctor immediately if you:<sup>4</sup>

- Develop symptoms that could be signs of a severe allergic reaction, such as weakness, feeling dizzy, difficulty breathing or a rash or swelling of the face, lips, mouth, tongue or throat.
- Have persistent nausea, vomiting or diarrhoea that leads to loss of appetite, weight loss or dehydration. Vomiting or diarrhoea are very common and may lead to electrolyte deficiencies requiring supportive care.
- Develop any symptoms of kidney problems such as passing less urine, swelling of the legs, feet or ankles, shortness of breath, irregular heartbeat or confusion.
- Have changes in your vision.



Photo courtesy of The Progeria Research Foundation

**Zokinvy dosage can be adjusted to manage side effects.**

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## You can get information and support about progeria via the EigerOne Patient Support programme. Healthcare professionals will enrol eligible patients on the programme.

This dedicated programme has been co-developed with the European progeria support group, Progeria Family Circle, to meet the specific needs of people with progeria and their families and caregivers.



Our team of knowledgeable, specialist nurses can support you with a wide range of information and support, including:

- The disease and its symptoms
- The treatment and potential side-effects
- How it affects you as a patient or family member
- Tips and support to help you cope with daily living more easily

Please ask your prescriber for more details about the EigerOne Patient Support programme and how to enrol.

1. Gordon LB, Shappell H, Massaro J, *et al.* Association of lonafarnib treatment vs no treatment with mortality rate in patients with Hutchinson-Gilford progeria syndrome. *JAMA*. 2018;319(16):1687-1695. doi:10.1001/jama.2018.3264.
2. Marcelot A, Worman HJ, and Zinn-Justin S. Protein structural and mechanistic basis of progeroid laminopathies. *FEBS Journal*. 2021;288:2757-2772. Doi:10.1111/febs.15526.
3. Devi AS, Thokchom S, Devi AM. Children living with progeria. *Nurs Care Open Acces J*. 2017;3(4):275-278. doi:10.15406/ncoaj.2017.03.00077.
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6. Gordon BL, *et al.* Clinical trial of a farnesyltransferase inhibitor in children with Hutchinson-Gilford progeria syndrome. *PNAS*. October 9, 2012. vol. 109. no. 41. www.pnas.org/cgi/doi/10.1073/pnas.1202529109.



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