

**A BROCHURE FOR
PATIENTS LIVING
WITH ADA-SCID AND
THEIR FAMILIES
AND CAREGIVERS**

Revcovi[®]
elapegademase-lvlr
injection 1.6 mg/mL

**There for the journey
with ADA-SCID**

Josh, actual
Revcovi patient

INDICATION

Revcovi[®] (elapegademase-lvlr) is indicated for the treatment of adenosine deaminase severe combined immune deficiency (ADA-SCID) in pediatric and adult patients.

IMPORTANT SAFETY INFORMATION

WARNINGS AND PRECAUTIONS

- If you have lower than normal platelet counts, you may be at increased risk of bleeding at the injection site. Your doctor should not prescribe Revcovi if you have severely low blood platelet counts.

- There may be a delay in improvement of your immune system's function after starting Revcovi treatment. During this delay, it is important to be protected from exposure to infections until your immune system improves.

Please see Important Safety Information throughout and accompanying full [Prescribing Information](#).

WHAT IS ADA-SCID?

ADA: 3 Important letters for your body

People with ADA-SCID have a deficiency in the ADA enzyme. ADA stands for adenosine deaminase, and SCID stands for severe combined immunodeficiency. Those are the medical terms—but what do they actually mean?

If left untreated, ADA-SCID becomes life-threatening before the age of 2 years.

It's crucial for doctors to diagnose ADA-SCID and start treatment as early as possible.

Here's what goes on inside the body of someone with ADA-SCID:



Every day, cells divide in your body. This is a normal part of life for all people.



The dividing process creates leftover material that your body doesn't need. ADA helps to remove this leftover material.



But in ADA-SCID, ADA doesn't work correctly and the leftover materials can't be removed. The materials, including deoxyadenosine nucleotides (dAXP), build up in cells.



dAXP damages the development of immune cells called lymphocytes—key tools that your body needs to fight off infections.

When ADA can't do its job, neither can the immune system.

This leads to **common signs of ADA-SCID**, which are usually diagnosed in infants:

- Slowed growth or failure to thrive
- Pneumonia
- Serious infections in the body, including the mouth
- Skin rashes
- Chronic diarrhea



HOW DO DOCTORS DIAGNOSE ADA-SCID?

Identifying ADA-SCID

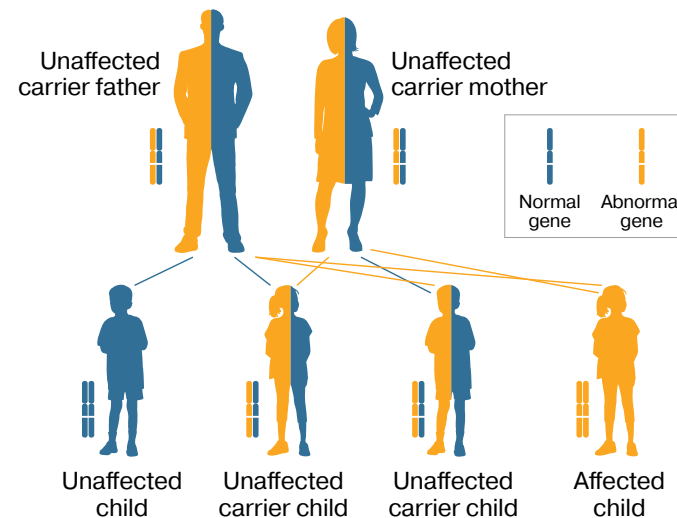
ADA-SCID is a very rare disease. By some estimates, for every million babies born in the world, only 1 has ADA-SCID. This can make it difficult to find.

SCID is usually diagnosed in infants via newborn screening, which is available in all 50 states, the District of Columbia, and Puerto Rico. When doctors notice common signs of ADA-SCID, they may order a blood test. Genetic testing can also confirm ADA-SCID.

Your genes have the clues

ADA-SCID is a genetic disorder, meaning it's passed down from parents to their child. In most cases, for someone to have ADA-SCID, both of their parents must have a copy of a gene that can cause ADA-SCID.

If you or someone in your family has ADA-SCID, other family members may be at risk. Your doctor can help you with how to talk to your family about getting tested.



Two parents, each with 1 defective gene, have a:

- 25% chance of having a child with 2 normal ADA genes (bottom left)
- 50% chance of having a child who has 1 ADA gene but is not affected by ADA-SCID (bottom middle two)
- 25% chance of having a child with 2 defective ADA genes who may be affected by ADA-SCID (bottom right)

Knowing a person's family history can help doctors decide to screen before birth. For example, unexplained infant death or infant death due to infection could be a sign of ADA-SCID within the family.

Your doctor may recommend Revcovi if you used to be on a different ERT

You may have heard of, or previously received, an ERT for ADA-SCID called Adagen[®] (pegademase bovine). Adagen is no longer available, but Revcovi was designed to replace it.



Ashley, actual
Revcovi patient

REVCОВI: THERE FOR THE JOURNEY WITH ADA-SCID

The only FDA-approved enzyme replacement therapy (ERT) for ADA-SCID

Revcovi replaces the ADA enzyme in people with ADA-SCID—infants, children, and adults.

Here's what goes on inside the body of someone with ADA-SCID:



Revcovi helps do the job of the ADA enzyme: removing the leftover material from the dividing of the cells.



If the leftover material is removed, dAXP can't build up. And without dAXP, the body can produce more lymphocytes to fight off infections.

IMPORTANT SAFETY INFORMATION

WHAT ARE THE POSSIBLE SIDE EFFECTS OF REVCОВI?

- Cough and vomiting were the most common side effects reported by patients receiving Revcovi.
- Based on reports of side effects of other enzyme replacement therapies used to treat ADA-SCID, which are similar to Revcovi, patients taking Revcovi may also experience:
 - Blood and blood cell-related problems: red blood cells being destroyed faster than they can be made, resulting in anemia; and higher or lower than normal number of platelets in the blood

Please see Important Safety Information throughout and accompanying full Prescribing Information.

Revcovi

Revcovi works to help keep your immune system functioning normally.

Most patients have improved immune system function by 24 weeks. It's important to stay on treatment so that Revcovi can help your immune system continue doing what it's supposed to do: fighting off the infections that ADA-SCID may cause.

TAKING REVCОВI

2x
a week

Intramuscular
injection

At home
or at the doctor's office

You can take Revcovi at home



If you'd like to take Revcovi at home, your doctor can show you how. You can also have your doctor administer it to you in their office.

How to store Revcovi:

- Store in the refrigerator between 2°C and 8°C (36°F to 46°F) in the original carton to protect from light
- Do not freeze or shake. Do not use if any indication that vial may have been frozen
- Do not reuse the vial. Throw away unused portions
- Throw away if solution is discolored, cloudy, or contains particles

How will you know when Revcovi is working?

Revcovi is designed to help replace the ADA enzyme. It's meant to decrease levels of dAXP in your blood, improve ADA activity throughout your body, and increase lymphocyte numbers in your blood.



dAXP going down
dAXP can damage lymphocytes.



Lymphocytes going up
Lymphocytes help your body fight infection.



ADA activity going up
The ADA enzyme helps your immune system work normally.

To see if Revcovi is working for you, your doctor will keep track of these levels in your blood.

SUPPORTING PATIENTS DURING TREATMENT WITH REVCОВI

Chiesi offers one-stop patient support services through Chiesi Total CareSM.



Assistance with coverage and reimbursement questions.



Case managers who provide patient education, access solutions, and liaising with insurance companies.



Some patients may be eligible for copay assistance, patient assistance, or other patient support programs.*

For more information, including the full terms and conditions for each patient support program, contact the **Chiesi Total Care Team at 1-866-272-7078.**

Advocacy and support organizations

The following organizations offer support and services for people with ADA-SCID and their families:



Immune Deficiency Foundation
primaryimmune.org



Jeffrey Modell Foundation
info4pi.org



SCID Angels for Life Foundation
scidangelsforlife.com



National Organization for Rare Disorders
rarediseases.org



Global Genes
globalgenes.org



World PI Week
worldpiweek.org

The Revcovi copay program

You may pay as little as \$0 for your prescription.*

Program eligibility:

- You must be enrolled in Chiesi Total Care.
(Enrollment form can be found at ChiesiTotalCare.com)
- You have commercial insurance and a valid prescription for a US Food and Drug Administration (FDA)–approved indication for Revcovi
- You must be a resident of the United States or one of its territories



IMPORTANT SAFETY INFORMATION WHAT ARE THE POSSIBLE SIDE EFFECTS OF REVCОВI? (CONT'D)

- Based on reports of side effects of other enzyme replacement therapies used to treat ADA-SCID, which are similar to Revcovi, patients taking Revcovi may also experience:
 - Skin-related problems: redness and itching at the injection site
 - Cancers of the immune system called lymphomas

* Please refer to the full accompanying [Terms and Conditions](#) for additional eligibility requirements.

Lesa, Chiesi Total Care Pharmacist

For more information,
download *Tips
for Effective
Communication
With Your Healthcare
Providers* available at
[Revcovi.com](https://www.revcovi.com).

FAQS AND GLOSSARY

Questions to ask your doctor about living with ADA-SCID

Treatment

- How long will I need treatment, and why is it important not to miss any treatments?
- What are the potential side effects?
- How often will I need to have my blood tested?

Healthcare team

- Who will be part of my healthcare team?
- Do I need to see any other specialists?

Medical information

- What can I do to help prevent getting infections?
- Are there any restrictions on my activity level or diet?
- Can I get all the recommended vaccinations?
- What are the long-term health effects of ADA-SCID?

Glossary

- **ADA:** Stands for adenosine deaminase, an enzyme that removes buildup of toxic materials in your cells.
- **ADA-SCID:** Stands for adenosine deaminase severe combined immune deficiency, which is a rare genetic disorder that prevents the body from making enough ADA.
- **Enzyme:** A protein that performs a specific job. Your body has many enzymes.
- **Enzyme replacement therapy:** A treatment that replaces a missing enzyme in the body.
- **dAXP:** Stands for deoxyadenosine nucleotide, which is a toxic buildup that can happen in the cells of people with ADA-SCID. dAXP can harm the immune system by damaging and removing lymphocytes.
- **Genetic:** Something that can be passed down from parent to child.
- **Lymphocyte:** An immune cell that helps the body fight infections.

Please see Important Safety Information throughout and accompanying full [Prescribing Information](#).

Josh, actual
Revcovi patient
and partner

There for the journey with ADA-SCID

For information about getting your Revcovi
prescription, please contact the Chiesi Total CareSM
team at **1-866-272-7078**.

Learn more at
[Revcovi.com](https://www.Revcovi.com) or by
talking to your doctor.
Find out more about
support services
available to patients at
[ChiesiTotalCare.com](https://www.ChiesiTotalCare.com)

IMPORTANT SAFETY INFORMATION WHAT TYPES OF MONITORING WILL BE NEEDED WHILE I AM TAKING REVCovi?

- Continuous therapy and adherence to the recommended drug schedule is important for the success of the treatment.
- While you are being treated with Revcovi, your doctor will need to do blood tests to monitor the levels of ADA activity and metabolites and overall immune function.
- This monitoring will ensure you are maintaining appropriate levels for therapeutic benefit. If an ongoing decline in your blood's measured adenosine deaminase. (ADA) activity occurs, your

immune function and clinical status will be monitored closely and precautions will be taken to help reduce your risk of infection.

WHAT IS KNOWN ABOUT TAKING REVCovi DURING PREGNANCY?

- If you are pregnant or planning to become pregnant, or breastfeeding or planning to breastfeed, please talk to your doctor.

You are encouraged to report negative side effects of prescription drugs to the FDA. Visit **www.fda.gov/medwatch** or call **1-800-FDA-1088**.

**Please see Important Safety
Information throughout and
accompanying full [Prescribing
Information](#).**

Where applicable, personal information shared and processed for adverse events reporting, product orders, reimbursement support, and patient assistance is subject to Chiesi's Privacy Policy available at **<https://www.chiesiusa.com/privacy-policy/>**.

References available upon request.

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