

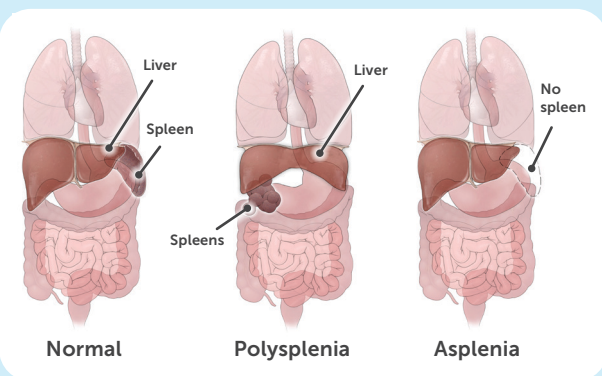
What is heterotaxy syndrome?

Heterotaxy syndrome is a rare condition in which some of the body's organs may be in an abnormal place or formed abnormally. They may not work the right way. The organ systems most commonly affected are the heart, lungs, immune system and the gastrointestinal (GI) tract.

How does heterotaxy syndrome affect the immune system?

The spleen is often affected in children with heterotaxy. The spleen does several jobs. One is to hold white blood cells that fight infections. When blood filters through the spleen, white blood cells can detect and fight bacteria that can cause an infection.

In children with heterotaxy, the spleen may not form at all (**asplenia**) or may be divided into multiple parts (**poly-splenia**). Even if the spleen looks normal, it may not work normally. This is called **hyposplenia**.




What else should I know?


Emergency antibiotic: Your child's immunology provider may prescribe an emergency antibiotic in case your child gets a fever or systemic infection. If your child has signs or symptoms of a systemic infection, follow the recommendations for emergency care.

Travel planning: If you're planning to travel to another country, we recommend you visit a travel clinic 4–12 weeks before you plan to travel.

- Malaria can cause a severe infection in children with this condition. Malaria prophylaxis and avoiding mosquitos are recommended for children travelling to tropical regions or Africa.
- Outbreaks of meningitis (meningococcal disease) happen more often in parts of the world where fewer people are vaccinated. Children with this condition are at increased risk for severe infections with meningococcus. Talk about this with your child's immunologist before making travel plans. They may need additional meningococcal vaccines.

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<https://childrenshospital.org/conditions/heterotaxy>

Heterotaxy syndrome

and your child's immune system

How can I know if my child's spleen is working?

Your child's immunologist will probably recommend several tests.

Sometimes, the tests can tell for sure the spleen isn't working normally. Other times, the tests don't tell for sure.

Your child's immunologist will explain the results and make a plan with you and your family.

All children with heterotaxy need to be seen by an immunologist. Most children will need regular visits with an immunologist. If you can't see an immunologist near you, please let us know and your child can be seen by an immunologist here at Boston Children's.

What are some specific immune system tests my child might have?



Abdominal ultrasound – an ultrasound of the abdomen to look at the abdominal organs



Nuclear medicine spleen scan – a test that sees how well the spleen is working



Special blood tests – special blood tests can be used to provide information about whether the immune system and spleen are working normally

What is the risk of infection?

If your child's spleen isn't working normally, they're at higher risk of developing a severe infection compared to someone with a normal spleen. These infections are serious and can be life-threatening. Your child's immunologist will explain how to protect your child against infections. They'll also talk to you about signs of a serious infection and what to do if you see them.

Still, most children can participate in all normal activities with good preventive care from an immunologist.

How can I protect my child against infections?

Immunizations: Vaccines are **extremely** important in reducing the risk of serious infections in all children, but especially in children with heterotaxy.

- We recommend that your child follow a slightly different vaccine schedule than most children.
- This allows them to be protected earlier and have extra protection against bacteria and other germs that are life-threatening to children with heterotaxy.
- In addition to the regular vaccine series recommended by the CDC, we also recommend:
 - » Pneumococcal boosters (PCV20 or PPSV23) every 5-7 years. This protects against more strains of **pneumococcus** than the primary series alone.
 - » The **meningococcal A** series by age 2 and the **meningococcal B** series starting at age 10.
 - » Boosters are also recommended for **meningococcal** vaccines. Please check with your immunologist for specific recommendations
 - » Annual **influenza** vaccination and **COVID-19** vaccination for children 6 months and older
- If you or your child's primary care provider have questions or concerns about the vaccination schedule, please talk about it with your child's immunologist.

How do I know if an infection is an emergency?

Even with the best preventive care, infections can happen. Infections can spread quickly and become severe.

Share the information below with all of your child's caregivers:

- **Fever:** If your child gets a fever (higher than 101°F/38.3°C) or other signs of a severe infection (chills, shivering, difficulty breathing, vomiting (throwing up), diarrhea, a headache or isn't alert and responsive), take them to the nearest emergency department right away.

Antibiotic prophylaxis: Antibiotics are often used to treat infections, but they can also be taken regularly at a lower dose to prevent infections. This is called **prophylaxis**.

- Using prophylactic antibiotics reduces the risk of getting an infection, especially in young children.
- The most typical antibiotic used for prophylaxis is amoxicillin. If your child has an allergy or can't take amoxicillin, they can take a different antibiotic.
- Prophylactic antibiotics are very safe. There's a concern in the general population for overusing antibiotics, but the benefits often far outweigh any downside for children with heterotaxy.
- Some children take antibiotics only until age 5. Others benefit from long-term (even life-long) antibiotics. Your child's immunologist will discuss their recommendations and make a joint decision with you.

Prophylaxis for procedures: Your child may need a higher dose of antibiotic or a different antibiotic before certain procedures involving the **sinuses**, the **respiratory tract (lungs)** and the **teeth (dental work)**.

This can be especially important for some children with certain heart conditions. Talk to your child's cardiologist, immunologist and pediatrician about whether they need to adjust antibiotics before any procedure.

- » Generally, children with symptoms of a viral upper respiratory illnesses (runny nose, nasal congestion, mild cough) who don't have a fever can be cared for at home. If you aren't sure, talk to your child's pediatrician.

- **Animal bites:** Your child needs medical care right away for any animal bite. Dog bites, in particular, can be deadly in people with spleen conditions.



Consider having your child carry an alert card or wear an alert bracelet with information about their condition and the risk of serious infections.