



How well do you know the liver?



The role of the liver

Over 500 jobs to help keep you healthy

Some of the most important functions of the liver:

- Converts food into energy
- Cleans the blood
- Removes waste from the body



Other important liver functions

- Cleans the blood of bacteria to prevent infections
- Produces, breaks down, and regulates hormones in the body
- Helps the body easily and effectively use some medicines
- Maintaining the body's energy



Bile production: An important job of the liver

- The liver produces a thick, yellow-green fluid called bile, which carries cholesterol, waste, and other toxins away from the body
- Bile helps break down and absorb fats and nutrients from the food we eat
- Rids the body of excess cholesterol, bilirubin (an orange-yellow pigment created when blood cells are broken down), and toxins



How PBD-ZSD affects the liver

- Liver damage is almost universal in PBD-ZSD patients
- Peroxisomes found in liver cells (responsible for producing bile acids) only partially work or do not work at all
- Loss of peroxisome function interferes with the liver's ability to produce bile acids, leading to the production of toxic substances in their place
- Toxic build-up can lead to scarring of the liver (fibrosis) and liver failure over time

Is your child's liver impacted?

With PBD-ZSD, it is important to recognize symptoms early on. **Use this checklist to help you identify symptoms of liver involvement in your child.**

- Yellowing skin or white parts of eyes (jaundice)
- Pale, foul-smelling stools
- Dark urine
- Difficulties with feeding
- Poor growth (not meeting weight or height milestones for age)
- Reports of enlarged liver (hepatomegaly) from previous doctor visits

The importance of monitoring liver function

- To find new symptoms
- To know if damage has been done to the liver or if it is at risk of damage
- To adjust how your child's liver health is managed

Monitoring the liver might not seem urgent in the face of noticeably serious issues, like seizures or hearing loss, but a healthy liver is so important that we can't live without it. Your doctor can assess your child's liver health with these tests.

Measuring liver chemistries

- A common series of blood tests that look at levels of different enzymes and proteins in the liver to check if the liver is working properly and if there is any damage
- Routinely performed by your child's hepatologist

Atypical bile acid test

- A less routine test that detects high levels of abnormal/atypical bile acids (DHCA and THCA) in the blood serum that may indicate the liver is at risk of damage
 - DHCA, dihydroxycholestanoic acid (pronounced di-hi-drox-ee-kohl-esta-no-ik)
 - THCA, trihydroxycholestanoic acid (pronounced try-hi-drox-ee-kohl-esta-no-ik)
- This test is uncommon, so you may have to ask your child's hepatologist or gastroenterologist to run this test at your next visit

Understanding your test results

- Elevated levels of liver enzymes or proteins (with the exception of albumin) indicate potential liver involvement
- Due to the complexity of PBD-ZSD, detection of harmful bile acids, such as DHCA and THCA, with an atypical bile acid test can help indicate liver involvement in your child*

PBD-ZSD liver symptoms may be managed if identified early. Talk to your child's hepatologist or gastroenterologist about a free atypical bile acid test and treatment options that can help protect the liver.

Ask your doctor to request free atypical bile acid testing at ZSDLiverTest.com.

*This test is offered for free to patients through a program sponsored by Cincinnati Children's Hospital and Mirum Pharmaceuticals, Inc. If left undetected, liver injury caused by PBD-ZSD could lead to long-term liver damage.



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