

ZELLWEGER SPECTRUM DISORDER

What is it?

Zellweger Spectrum Disorder (ZSD) was recently viewed as 3 separate diseases but today is categorized as set of disorders that form a spectrum, or continuum, of 1 disease. This spectrum can range from mild Infantile Refsum Disease (IRD), to moderate Neonatal Adrenoleukodystrophy (NALD), to severe Zellweger Syndrome (ZS)

ZSD is also known as peroxisome biogenesis disorders (PBDs). These disorders are caused by a loss of function in important parts of your cells called "peroxisomes," which are responsible for breaking down fats and chemicals and getting rid of waste so that your body can function properly. This disorder can affect many parts of the body from the eyes to the liver. The various body systems and functions as described below.

Nutrition	Malabsorption of nutrients can lead to poor growth, feeding problems, and deficiency in fat-soluble vitamins.
Hearing	Varying degree of hearing loss requires the child receive a yearly evaluation .
Vision	Vision loss is the most common problem.
Neurological	Improper development of the white matter in the brain (leukodystrophy) results in nerve damage and can potentially affect their development. Damage to the nerves that send information from the brain to the rest of the body (peripheral neuropathy) can often cause numbness or weakness in the hands and/or feet. Walking abnormality is the main neurological complication in adults with ZSDs. In people with mild forms of ZSDs, nerve damage to the muscles, skin, and internal organs usually begins during adolescence.
Kidney	Kidney issues occur in children 4 years and older and include kidney stones, kidney cyst, and kidney failure.
Liver	Jaundice and liver damage can also result.
Bone	Children are more prone to bone fractures that can occur without an identified cause.
Teeth	Teeth develop slowly and poorly. They are often small in size and have poor enamel and can be yellow or orange color.
Motor	Low muscle tone and poor strength causes poor movement and the main complication of walking abnormality.
Cognitive	Intellectual delay can be present.

What is the treatment?

There is no cure for the disease or a way to stop the disease progression. Treatment is focused on managing the signs and symptoms of the disorder. A multidisciplinary team could include physical, occupational, and speech therapy.



Kennedy Krieger Institute

The Specialized Health Needs Interagency Collaboration (SHNIC) program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

As a spectrum disorder each student with ZSD will present differently and student specific signs and symptoms could impact the student's ability to access their education. Supporting students with these conditions in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504/IEP could include:

- PT/OT/SLP evaluation
- Safety concerns related to mobility and low muscle tone
- Monitor for anxiety and fatigue
- Peer support
- Modified school day
- Wrap around services
- Plan for absences and make-up work
- Staff education and training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- Diagnosis and complete history
- Current medication list, including PRN medications
- Nutrition orders if applicable (including use of feeding tube and/or tube replacement policy)
- Toileting schedule
- Orthotics/splints, consistent skin checks
- Equipment troubleshooting
- Communicate with school staff, parents/guardian, and provider any changes or concerns about the disease
- Emergency Care Plan(s) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

NIH- Zellweger spectrum disorder

<https://medlineplus.gov/genetics/condition/zellweger-spectrum-disorder/#:~:text=Zellweger%20spectrum%20disorder%20is%20a,%2C%20and%20infantile%20Refsum%20disease>

NORD- Zellweger Spectrum Disorders

<https://rarediseases.org/rare-diseases/zellweger-spectrum-disorders/>

Understanding ZSD

<https://www.understandingzsd.com/resources>