

Bubble baby syndrome

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A girl from Karnataka, suffering from **Bubble baby syndrome**, recently underwent a groundbreaking **bone marrow transplant**, becoming the **youngest recipient** of such a procedure from an **unrelated donor**.



[ref-WTAP]

About the Bubble baby syndrome:

- The Bubble baby syndrome is a **rare genetic disorder** where babies are born without an **immune system**, leaving them susceptible to life-threatening infections.
- Genetic defects resulted in a deficiency of T cells and B cells, crucial for the immune system.
 - T cells are a part of the immune system that focuses on specific foreign particles.
 - **B cells** produce **antibodies** that are used to attack invading bacteria, viruses, and toxins.
- It is also called **Severe Combined Immunodeficiency** (SCID), living in the bubble syndrome and **Swiss-type agammaglobulinemia**.
- Children with SCID are **highly susceptible** to infections and often must live in a sterile or "**bubble-like**" environment to protect them from **pathogens**.
- It is usually inherited in an **autosomal recessive manner**, meaning that the condition occurs when a child inherits **2 defective copies** of a gene (usually IL2RG, ADA, or JAK3), one from each parent.
- Affected individuals are unable to mount effective immune responses against infections, making them vulnerable to a wide range of pathogens.
- Children with SCID become very sick with infections such as pneumonia, meningitis, and chickenpox.
- Many children die before they reach their first birthday.
- Among several types of SCID, the 2 most common types are classical X-linked and ADA deficiency.

Symptoms:

- Pneumonia
- Repeated ear infections
- Meningitis
- Blood infection
- · Chronic skin infections
- Yeast infections in the mouth and diaper area



- Diarrhea
- Liver infection (hepatitis)

Treatment:

• Stem cell transplant or bone marrow transplant or gene therapy can be effective treatments, providing a functional immune system.

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