



### Hyperimmunoglobulin E syndrome

Hyperimmunoglobulin E syndrome is a suspected genetic defect that produces high levels of the antibody immunoglobulin (IgE). It causes serious skin and lung infections as well as eczema

## Causes

Hyperimmunoglobulin E syndrome is also known as Job syndrome, after the biblical character Job whose faithfulness was tested by a lifelong affliction of draining skin sores and pustules. People with this condition have chronic and severe skin infections.

It is an extremely rare disease, and the cause is unknown. However, the bacteria *Staphylococcus aureus* is believed to most commonly trigger the infections related to the disease. Other common triggers include *Streptococcus pneumoniae*, *Haemophilus influenzae*

,  
*Candida albicans*  
, and the herpes virus.

**Hyper IgE syndrome** is, in most cases, inherited in a dominant fashion, meaning that it can run in families with high frequency.

## Symptoms

- Persistent skin abscesses and infection
- Recurrent pus in the sinus
- Eczema
- Bone and tooth defects, including fractures and late shedding of baby teeth

## Exams and Tests

An eye exam may reveal signs of dry eye syndrome. A physical exam may show signs of osteomyelitis, curving of the spine (kyphoscoliosis), and recurrent sinus infections.

A chest x-ray may reveal lung abscesses.

Tests used to confirm a diagnosis include:

- Absolute eosinophil count
- CBC with blood differential
- Serum globulin electrophoresis to look for high blood IgE levels

Other tests that may be done:

- X-ray of the sinuses

- X-ray of the bones
- CT scan of the chest
- Cultures of infected site

## Treatment

There is no known cure for the condition. The goal of treatment is to control the recurrent infections. Medications include antibiotic treatment. Antifungal agents and antiviral agents are prescribed when appropriate.

Sometimes, surgical drainage of abscesses is needed.

IV gamma globulin may help to build up the immune system temporarily when there are severe infections.

## Outlook (Prognosis)

Job syndrome is a lifelong chronic condition. Each new infection requires treatment.

## Possible Complications

- Repeated infections
- Sepsis

## Prevention

There is no proven prevention. Good general hygiene is helpful. Some doctors may recommend preventive antibiotics for people with many infections, particularly with *Staphylococcus aureus*. This does not change the underlying disorder but rather its consequences.