

Autoimmune hemolytic anemia in childhood

What is autoimmune hemolytic anemia (AIHA)?

AIHA is a condition where there is a drop in the number of red blood cells (anemia), due to destruction of red blood cells (hemolysis) by the body's immune system.

What are the symptoms of AIHA?

- Dark urine
- Fatigue
- Pale color (pallor)
- Rapid heartbeat
- Shortness of breath
- Yellow skin color (jaundice)

What causes AIHA?

AIHA is an acquired disease that occurs when antibodies form against a person's own red blood cells. In the idiopathic form of this disease (which accounts for one-half of all AIHA) the cause is unknown. It is not inherited, and other children cannot catch it from your child. There are other types of AIHA which result from an underlying disease or medication. In children it is uncommon to find an underlying cause.

How is AIHA diagnosed?

Physical examination reveals an enlarged spleen and liver with pallor and jaundice, but any hemolytic anemia can give rise to this. It is very important to be sure what the underlying cause is and the following tests are helpful:

- Direct and Indirect Coombs test (which looks for antibody)
- Hemoglobin in the urine
- Red blood cell count and serum hemoglobin
- Reticulocyte count
- Serum bilirubin levels
- Serum haptoglobin

What treatment will my child receive?

The first therapy tried is usually a steroid, such as prednisone. Intravenous immune globulin (IVIG) may also be given. Removal of the spleen (splenectomy) may be considered in older patients. Medications to suppress the immune system are considered in patients not responding to initial treatment, such as azathioprine (Imuran™), cyclophosphamide (Cytoxan™), and rituximab (Rituxan™).

Blood transfusions are given with caution to keep the hemoglobin above 6-8 g/dL, because the blood that is given may also end up being destroyed (hemolysis). It is common to have difficulty finding blood that is properly matched in this situation, and your doctor will discuss the risks and benefits of blood transfusion in this setting.

Will my child be cured?

Complete remission is obtained at the end of one month of treatment in almost two-thirds of patients, almost all of whom will end up in long-term remission without treatment. A quarter of patients need long-term treatment. Death is rare due to AIHA.