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Review Article

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A Comprehensive Review of the Pathological Mechanism of Evans Syndrome

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drome is defined as the association between AIHA and ITP. Neutro-
penia occurs less often. In some cases, autoimmune destruction of these blood cells occurs simultaneously. In most cases, one disease

Evans syndrome was first described in the medical literature in 1951 by Dr. Robert Evans and colleagues. For many years this disorder was considered a random event of AIHA with thrombocy-

Other people may initially present with low platelet levels, known as thrombocytopenia. Thrombocytopenia can cause tiny reddish or purple spots on the skin (petechiae), a broader purplish discolor-

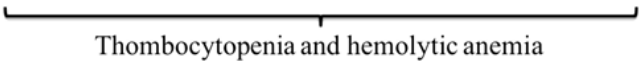

Figure 1: *Schematic of the physiological mechanism of the immune system in Evans syndrome. [1]*

Some people with Evans syndrome may initially destroy red blood cells faster than the body can replace them. Low levels of circulating red blood cells, known as anemia, can cause a variety of symptoms, including fatigue, pale skin (pallor), dizziness, shortness of breath, dark urine, and palpitations. Some people may experience yellowing of the skin and especially the whites of the eyes (jaundice). [1,2]

stances that mark them for destruction by white blood cells. When antibodies target healthy tissue, they may be called autoantibodies. Researchers believe that a trigger event (such as an infection or underlying disorder) may cause the immune system to produce autoantibodies in Evans syndrome. [1,3]

Evans syndrome can occur with another disorder as a secondary disease. Secondary Evans syndrome may be associated with other disorders such as autoimmune lymphoproliferative syndrome (ALPS), lupus, antiphospholipid syndrome, Sjogren's syndrome, common variable immunodeficiency, IgA deficiency, specific lymphoma, and chronic lymphocytic leukemia. [1,3]

with ALPS have a mutation in the tumor necrosis factor receptor superfamily member gene (TNFRSF6), also called CD95 or Fas.

mal nocturnal hemoglobinuria (PNH), acquired thrombotic thrombocytopenic purpura, hemolytic uremic syndrome, and Kasabach-Merritt syndrome. [1,4]

Autoimmune lymphoproliferative syndrome (ALPS) is a rare genetic disorder that overlaps with Evans syndrome. A characteristic finding of ALPS is the presence of abnormally high numbers of white blood cells called lymphocytes, which can accumulate in the lymph nodes, liver, and spleen and cause enlargement of these organs. ALPS can lead to symptoms similar to Evans syndrome, particularly anemia, thrombocytopenia, and neutropenia. Most people

Treatment Paths for Evans Syndrome

There is no cure for Evans syndrome, and treatment is often challenging. Treatment is directed at specific symptoms evident in each individual. Treatment may require the coordinated effort of a team of specialists. Pediatricians, surgeons, hematologists, pediatric hematologists, immunologists, rheumatologists and other healthcare professionals may need to systematically and comprehensively plan an effective treatment for a child. [1,5]

Most affected individuals require treatment, although spontaneous recovery has been reported in rare cases. Several different types of treatments have been used to treat people with Evans syndrome,

and their effectiveness has varied greatly among affected people. Some people have long-term recovery from this disorder. Others

of Evans syndrome caused by underlying ALPS, hypogammaglobulinemia appears to develop which may persist in patients treated with rituximab. Hypogammaglobulinemia is a condition in which the body's immune system does not produce enough antibodies and potentially makes those affected susceptible to bacterial infections and, to some extent, some viral infections. [1,7]

Additional drugs have been studied in a small number of people with Evans syndrome. The second factor that appears to be relatively effective in this disorder is mycophenolate mofetil. These drugs can be used alone or in combination (multi-agent therapy)

as second-line treatments for people with Evans syndrome who do not respond to corticosteroids or IVIg therapy. More research is

problems, including l heart failure. Some doctors recommend that children with Evans syndrome be screened for ALPS because the prevalence of these two disorders is high. Screening includes testing for the presence of double negative T cells (DNT) by flow cytometry, the presence of which indicates ALPS. New treatments for Evans syndrome are being studied. Rituximab appears to be a very effective treatment for patients with Evans syndrome. Rituximab is classified as a monoclonal antibody or biologic therapy, drugs that act like antibodies but are created artificially in a laboratory. Preliminary studies have shown this drug to be generally safe and

effective. The advantages of rituximab are to avoid severe immunosuppression and side effects associated with other immunosuppressive agents. The disadvantage is that in cases of Evans syndrome caused by underlying ALPS, hypogammaglobulinemia appears to develop which may persist in patients treated with rituximab. Hypogammaglobulinemia is a condition in which the body's immune system does not produce enough antibodies and potentially makes those affected susceptible to bacterial infections and, to some extent, some viral infections. [1-7]

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