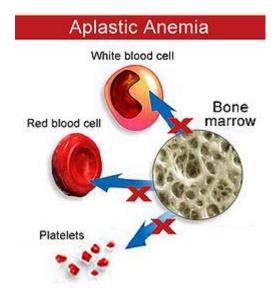
Aplastic anemia

"Those who think they have not time for bodily exercise will sooner or later have to find time for illness." — Edward Stanley

From Health and Wellness

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Aplastic anemia is a relatively rare but potentially fatal blood disorder.



Aplastic Anemia (Uploaded by <u>troyshado</u> on 6 January 2009)

Aplastic anemia is a relatively rare blood disease where the bone marrow cannot produce enough new blood cells to replace dying and damaged ones. Red blood cells usually live for about 120 days, platelets for about 6 days, while white blood cells do not last more than 24 hours. As its name suggests, "aplastic anemia" involves both aplasia and anemia.

Symptoms of aplastic anemia

The symptoms of aplastic anemia are:

- Anemia with malaise, pallor, and associated symptoms such as palpitations;
- Low platelet counts (thrombocytopenia), leading to increased risk of hemorrhage, bruising, and petechiae (small purplish spot on a body surface, caused by a minute hemorrhage);
- Low white blood cell counts (leukopenia), leading to increased risk of infection; and/or
- Low reticulocyte counts (reticulocytopenia).

Causes of aplastic anemia

- 1. In many cases, the cause of aplastic anemia cannot be determined, but one known cause is an autoimmune disorder in which white blood cells attack the bone marrow.
- 2. Aplastic anemia is also sometimes associated with:
 - o exposure to toxins such as benzene;
 - o the use of certain drugs, including quinine;
 - exposure to ionizing radiation from radioactive materials or radiationproducing devices.

Diagnosis of aplastic anemia

- 1. The diagnosis of aplastic anemia can only be confirmed by a bone marrow examination because this disease has to be differentiated from anemia and pure red cell aplasia. Typically, anemia refers to low red blood cell counts, whereas pure red cell aplasia is a type of anemia affecting the precursors to red blood cells. Unlike pure red cell aplasia which is characterized by a reduction in only red cells, aplastic anemia patients have pancytopenia (i.e., anemia, neutropenia and thrombocytopenia), resulting in a decrease of all formed elements.
- 2. Before bone marrow examination is undertaken, a patient will generally have undergone <u>blood tests</u> to find diagnostic clues, including a complete blood count (CBC).

Treatment for aplastic anemia

- 1. Treatment of aplastic anemia involves suppression of the immune system, an effect achieved by daily intake of medicine, with more severe cases necessitating a bone marrow transplant. The stem cells in the transplanted bone marrow reconstitute all three blood cell lines, giving the patient a new immune system. Besides the risk of graft failure, however, there is also a risk that the newly-created white blood cells may attack the rest of the body (known as "graft-versus-host disease").
- 2. Medical therapy of aplastic anemia often includes a short course of anti-thymocyte globulin (ATG) or anti-lymphocyte globulin (ALG), and several months of treatment with ciclosporin to modulate the immune system. Mild chemotherapy with agents such as cyclophosphamide and vincristine may also be effective, while antibody therapy, such as ATG, targets T-cells which are believed to attack the bone marrow. Steroids are generally ineffective, though they are often used to combat serum sickness caused by ATG use.

Follow-up treatment

- 1. Regular full blood counts are required to determine whether the patient is still in a state of remission.
- 2. Flow cytometry testing is performed regularly in people with previous aplastic anemia to monitor for the development of paroxysmal nocturnal hemoglobinuria (PNH, i.e. anemia with thrombopenia and/or thrombosis). PNH is a rare disease which develops in 10-33% of all aplastic anemia patients, and is caused by the bone marrow trying to escape from being destroyed by the immune system.

Prognosis

- 1. Untreated aplastic anemia is an illness that leads to rapid death, typically within 6 months. If treatment is given promptly, the survival rate for the next 5 10 years is substantially improved, and many patients live well beyond that length of time.
- 2. Occasionally, milder cases of the disease resolve on their own. Relapses of previously controlled disease are, however, much more common. Relapse following ATG/ciclosporin use can sometimes be treated with a repeated course of therapy.
- 3. Well-matched bone marrow transplants from siblings have been successful in young, otherwise healthy people, with a long-term survival rate of 80-90%. Most successful bone marrow transplant recipients eventually reach a point where they consider themselves cured for all practical purposes, although they need to be compliant with follow-up care permanently.
- 4. Older people who are either too frail to undergo bone marrow transplants, or are unable to find a good bone marrow match, usually undergo immune suppression which has a 5-year survival rate of up to 75%.