

# Overview of Aplastic Anemia

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## ABSTRACT

Aplastic anemia (AA) is characterized by abnormalities in stem and progenitor cells, bone marrow cytopenia, and peripheral blood pancytopenia. This condition mainly affects children, teenagers, and people aged 60 or above. Bone marrow is a porous substance found in the medullary cavity of the skeletal elements, primarily in the vertebrae, pelvis, and lower extremities. Bone marrow contains stem cells. Stem cells can self-renew, create more stem cells, and develop into different cell types like erythrocytes, leukocytes, and platelets. In acquired aplastic anemia, there is a severe deficiency of blood elements and subsequent reduction in levels of red blood cells, white blood cells and platelets.

### Introduction:

Aplastic anemia is a seldom encountered and grave medical condition. In 1888, Paul Ehrlich became the pioneer in documenting the initial case of this particular ailment. Vaux and Obertin are believed to have originally introduced the term "aprasco" in their analysis of the bone marrow in other patients (Marmont, 1995). Nonetheless, the term pernicious anemia was employed in the title of their scholarly article, which was published in 1904. In Ferrata's seminal work on hematology, "Le Emopatie" (1934), the author astutely emphasizes the need to distinguish AA from pernicious anemia, thus asserting its significance as a fundamental textbook in the field. Currently, the expression appears somewhat intricate due to the fact that pernicious anemia exhibits a substantial level of manageability, whereas AA continues to pose a perilous menace. Indeed, bone marrow failure (BMF) is frequently employed as an interchangeable descriptor for aplastic anemia (AA) due to its hematopoietic inadequacy in regenerating surplus blood cells [2].

### Initial assessment

The primary assessment of pancytopenia seeks to ascertain the diagnosis, evaluate the degree of severity, discern any causative factors, and establish its existence.

### Symptoms

Symptoms encompassing fatigue, dyspnea, tachycardia, pallor, unexplained susceptibility to recurrent or protracted infections, epistaxis, and gingival hemorrhage may manifest if present. Aplastic anemia can manifest as either a transient or a persistent condition. This condition bears severe consequences and may result in mortality. Aplastic anemia manifests through the presence of anemia, hemorrhaging, and susceptibility to infections. Bone marrow failure can be a consequence of various underlying conditions, although the majority of cases of aplastic anemia stem from a misdirection of the immune system in the bone marrow, known as autoimmunity. In reality, the majority of patients exhibit favorable responses to therapeutic interventions aimed at immune system suppression.

### Diagnosis

The diagnostic procedures utilized encompass comprehensive blood count analysis, the extraction of bone marrow through aspiration and trephine biopsy methods, examination of liver function indicators, as well as the screening for hereditary anomalies.

### Cause of aplastic anemia

The etiology most frequently associated with aplastic anemia entails an auto-immune response whereby the immune system targets and assaults the stem cells housed within the bone marrow. Several other factors have the potential to cause harm to the bone marrow and disrupt the process of blood cell production.

### Radiation and chemotherapy treatments

Anti-tumor therapies exert cytotoxic effects on cancer cells, albeit posing potential harm to normal cellular components, specifically stem cells located within the bone marrow. Aplastic anemia could potentially manifest as a transient consequence of the administration of this therapeutic intervention [1].

### Exposure to toxic chemicals

Certain noxious substances present in pesticides and herbicides along with benzene, a constituent of gasoline, have been implicated in the association with aplastic anemia. The potential for amelioration of this form of anemia

exists if individuals refrain from encountering the hazardous substances responsible for inducing this condition.

### Use of certain drugs

Some medications, including those indicated for the treatment of rheumatoid arthritis and select antibiotics, have the potential to induce the development of aplastic anemia.

### Autoimmune disorders

Bone marrow stem cells have been proposed to play a role in the pathogenesis of autoimmune diseases, characterized by aberrant immune activity targeting otherwise healthy cells.

### Risk factors of aplastic anemia

Aplastic anemia harbors risk factors that may emanate from either environmental or genetic origins. One illustrative instance involves individuals who become exposed to elevated levels of nuclear radiation or organic solvents, such as toluene, in the absence of adequate protection. It is noteworthy that these individuals face potential consequences in the form of aplastic anemia development. The ingestion of specific medications, such as sulphonamides and anti-seizure drugs, can have varying impacts on health. Exposure to hazardous substances frequently employed in agricultural settings, including those utilized for eradicating coca plants used in the production of cocaine in Colombia.

### Prevention

Anemia is frequently avoidable and can be effectively remedied through sufficient consumption of iron. Anemia is a medical condition that arises as a result of an inadequate presence of robust red blood cells within the human body. There exists the possibility of experiencing hematological conditions characterized by either insufficient red blood cell count or deficiency in hemoglobin, an iron-rich protein.

### Clinical manifestations

Patients with AA tend to be diagnosed at earlier stages of the disease. An individual with progressive anemia may potentially consider pursuing medical care to address symptoms such as fatigue and other related manifestations. Common symptoms often observed include recurring infections caused by severe neutropenia, as well as mucosal bleeding resulting from thrombocytopenia. Microbial infections are predominantly of bacterial origin. Invasive fungal infections represent a prevailing etiological factor contributing to mortality rates. Particularly in individuals exhibiting prolonged, profound neutropenia, Menorrhagia is a prevalent manifestation among females experiencing menopause.

### Treatment

Several medical interventions are commonly employed in clinical settings. These interventions encompass the utilization of red blood cell transfusions for the purpose of rectifying anemia, platelet transfusions to prevent or treat instances of excessive bleeding, as well as the administration of antibiotics to manage or guard against infections. Bone marrow transplantation is the preferred therapeutic approach, particularly in pediatric and young adult populations, particularly when employing allogeneic grafts.

### Immunosuppressive therapy

Anti-thymocyte globulin (ATG), an Immunoglobulin G (IgG), derived from equine sources, demonstrates reactivity towards human antigens on T-lymphocytes. This leads to the elimination of T lymphocytes present in the peripheral blood or alteration of their functionality. This specific

immunoglobulin G (IgG) has the capacity to initiate a comprehensive or partial restoration of hematopoiesis in individuals diagnosed with aplastic anemia. The half-life of this substance ranges from 1.5 to 12 days. Its administration should be limited to physicians with expertise in immunosuppressive therapy, and patients should exclusively receive this medication in facilities that possess sufficient laboratory and ancillary medical resources.

#### **Hematopoietic stem cell transplantation (HSCT)**

The curative potential of allogeneic hematopoietic cell transplantation (HCT) is constrained by the scarcity of an HLA-matched sibling. In contrast to hematologic neoplasms, the bone marrow is the favored source of stem cells in aplastic anemia (AA), as opposed to peripheral blood. Unrelated donor transplantation, administered to patients who have encountered an unsuccessful primary cycle of immunosuppressive therapy (IST), merits exclusive consideration, especially among pediatric and young adult populations.

#### **Prognosis and survival**

Without therapeutic intervention, individuals diagnosed with aplastic anemia exhibit a significantly elevated fatality rate reaching approximately 70 percent within a span of one year. Typically, the clinical trajectory exhibits heterogeneity, marked by complications arising from pancytopenia, namely infection and bleeding, as well as relapse and clonal evolution. However, the rising accessibility of hematopoietic stem cell transplantation and effective immunosuppressive therapy has concurrently led to a significant surge in survival rates, reaching as high as 80 percent.

#### **Epidemiology**

The occurrence of aplastic anemia displayed geographical variation, as evidenced by the incidence observed in patients with pancytopenia, which ranged from 10% to 57.14% the age distribution manifested as biphasic, with a prominent peak occurring between the ages of 15 and 25, and a minor peak in incidence observed in individuals aged 60 years and above. Furthermore, a noteworthy disparity in incidence rates was observed between males and females. Aplastic anemia impacts individuals across diverse age groups and ethnicities. The fluctuations in prevalence rates in developing countries remain uncertain, lacking clarity. Nevertheless, potential influences on an individual's state are likely to stem from various environmental factors, including but not limited to viruses, drugs, chemicals, genetic background, diagnostic criteria, and study design. Numerous studies conducted across diverse regions have consistently reported an average age of 8 years. The prevalence of Fanconi anemia in Pakistan, as recently reported, was found to be 16.6% this higher prevalence can be attributed to the increased degree of familial consanguinity within the population. These figures surpass those reported in the Western literature however, they align more closely with studies conducted in India [3].

#### **Conclusion**

The management of severe aplastic anemia has witnessed significant advancements in both allogeneic stem cell transplantation and immunosuppression approaches over the course of time, resulting in a remarkable enhancement of outcomes. With either therapeutic modality, it is now feasible to anticipate long-term survival rates exceeding 75% among patients.

#### **References**

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