Disorders of the Blood

This section is designed to provide a very brief over-view of some blood disorders and dyscrasias which may be encountered, but it is by no means an exhaustive, nor in-depth list. It is suggested that you carry out further reading to learn more about these disorders, and how their manifestations may present in, and affect, your patient.

Polycythaemia

Polycythaemia is characterised by an overabundance of erythrocytes, resulting in increased blood viscosity, reduced flow rates, and, if severe, plugging of the capillaries. Polycythaemia vera is a chronic form of polycythaemia of unknown cause. Secondary polycythaemia results from a decreased oxygen supply, such as in high altitudes, chronic obstructive pulmonary disease, or congestive heart failure. The resulting decrease in oxygen delivery to the kidneys stimulates erythropoietin secretion, causing an increase in erythrocyte production.

Anaemia

This is a deficiency of haemoglobin in the blood. It can result from a decrease in the number of erythrocytes, a decrease in the amount of haemoglobin in each erythrocyte, or both. The decreased haemoglobin reduces the ability of the blood to transport oxygen. Anaemic patients suffer from a lack of energy and feel excessively tired and listless. They can appear pale and quickly become short of breath with only slight exertion.

One general cause of anaemia is insufficient production of erythrocytes. Aplastic anaemia is caused by an inability of the red bone marrow to produce erythrocytes. It is usually acquired as a result of damage to the red marrow by chemicals (e.g., benzene), drugs (e.g., certain antibiotics and sedatives), or radiation.

Erythrocyte production can also be reduced because of nutritional deficiencies. Iron-deficiency anaemia results from a deficient intake or absorption of iron or from excessive iron loss. Consequently, not enough haemoglobin is produced, and the erythrocytes are smaller than normal. Folate deficiency can also cause anaemia. Inadequate amounts of folate in the diet is the usual cause of folate deficiency, with the disorder developing most often in the poor, in pregnant women, and in chronic alcoholics. Because folate helps in the synthesis of DNA, a folate deficiency results in fewer cell divisions and, therefore, decreased erythrocyte production. Another type of nutritional anaemia is pernicious anaemia, which is caused by inadequate amounts of vitamin B12. Because vitamin B12 is important for folate synthesis, inadequate amounts of vitamin B12 can also result in decreased erythrocyte production.

Although inadequate levels of vitamin B12, in the diet can cause pernicious anaemia, the usual cause is insufficient absorption of the vitamin. Normally the stomach produces intrinsic factor, a protein that binds to vitamin B12. The combined molecules pass into the lower intestine, where intrinsic factor facilitates the absorption of the vitamin. Without adequate levels of intrinsic factor, insufficient vitamin B12, is absorbed, and pernicious anaemia develops. Present evidence suggests that the inability to produce intrinsic factor is an autoimmune disease in which the body's immune system damages the cells in the stomach that produce intrinsic factor.

Another general cause of anaemia is loss or destruction of erythrocytes. Haemorrhagic anaemia results from a loss of blood, such as can result from trauma, ulcers, or excessive menstrual bleeding. Chronic blood loss, in which small amounts of blood are lost over time, can result in iron-deficiency anaemia. Haemolytic anaemia is a disorder in which erythrocytes rupture or are destroyed at an excessive rate. It can be caused by inherited defects within the erythrocytes. For example, one kind of inherited haemolytic anaemia results from a defect in the cell membrane that causes erythrocytes to rupture easily. Many kinds of haemolytic anaemia result from

unusual damage to the erythrocytes by drugs, snake venom, artificial heart valves, autoimmune disease, or haemolytic disease of the newborn.

Some anaemias result from inadequate or defective haemoglobin production. Thalassaemia is a hereditary disease found predominately in people of Mediterranean, Asian, and African ancestry. It is caused by insufficient production of the globin part of the haemoglobin molecule. The major form of the disease results in death by age 20, and the minor form in a mild anaemia. Sickle-cell anaemia is a hereditary disease found mostly in people of African ancestry but also occasionally among people of Mediterranean heritage. It results in the formation of an abnormal haemoglobin, in which the erythrocytes assume a rigid sickle shape and plug up small blood vessels. They are also more fragile than normal erythrocytes. In its severe form, sickle cell anaemia is usually fatal before the person is 30 years of age, whereas in its minor form, sickle-cell trait, there are usually no symptoms.

Von Willebrand's Disease

Von Willebrand's disease is the most common inherited bleeding disorder, occurring as frequently as 1 in 1000 individuals. Von Willebrand factor (vWF) facilitates platelet adhesion and is the plasma carrier for factor VIII. One treatment for von Willebrand's disease involves injections of vWF or concentrates of factor VIII to which vWF is attached. Another therapeutic approach is to administer a drug that increases vWF levels in the blood.

Haemophilia

Haemophilia is a genetic disorder in which clotting is abnormal or absent. It is most often found in people from northern Europe and their descendants. Because haemophilia is an X-linked trait, it occurs almost exclusively in males. Haemophilia A (classic haemophilia) results from a deficiency of plasma coagulation factor VIII, and haemophilia B is caused by a deficiency in plasma factor IX. Haemophilia A occurs in approximately 1 in 10,000 male births, and haemophilia B occurs in

approximately 1 in 100,000 male births. Treatment of haemophilia involves injection of the missing clotting factor taken from donated blood.

Thrombocytopenia

Thrombocytopenia is a condition in which the platelet number is greatly reduced, resulting in chronic bleeding through small vessels and capillaries. There are several causes of thrombocytopenia, including increased platelet destruction, caused by autoimmune disease or infections, or decreased platelet production, resulting from hereditary disorders, pernicious anaemia, drug therapy, or radiation therapy.

Leukaemia

Leukaemia is a type of cancer in which abnormal production of one or more of the leukocyte types occurs. Because these cells are usually immature or abnormal and lack their normal immunologic functions, patients are very susceptible to infections. The excess production of leukocytes in the red marrow can also interfere with erythrocyte and platelet formation and thus lead to anaemia and bleeding.

Infectious Diseases of the Blood

Microorganisms do not normally survive in the blood. Blood can transport microorganisms, however, and they can multiply in the blood. Microorganisms can enter the body and be transported by the blood to the tissues they infect. For example, the poliomyelitis virus enters through the gastrointestinal tract and is carried to nervous tissue.

After microorganisms are established at a site of infection, some can enter the blood. They can then be transported to other locations in the body, multiply within the blood, or be eliminated by the body's immune system.

Septicaemia, or blood poisoning, is the spread of microorganisms and their toxins by the blood. Often septicaemia results from the introduction of microorganisms by a medical procedure such as the insertion of an intravenous tube into a blood vessel.

The release of toxins by microorganisms can cause septic shock, which is a decrease in blood pressure that can result in death.

In a few diseases, microorganisms actually multiply within blood cells. Malaria is caused by a protozoan (Plasmodium) that is introduced into the blood by the bite of the Anopheles mosquito. Part of the development of the protozoan occurs inside erythrocytes. The symptoms of chills and fever in malaria are produced by toxins released when the protozoan causes the erythrocytes to rupture. Infectious mononucleosis (glandular fever) is caused by a virus (Epstein-Barr virus) that infects the salivary glands and lymphocytes. The lymphocytes are altered by the virus, and the immune system attacks and destroys the lymphocytes. The immune system response is believed to produce the symptoms of fever, sore throat, and swollen lymph nodes. The acquired immune deficiency syndrome (AIDS) is caused by the human immunodeficiency virus (HIV, which infects lymphocytes and suppresses the immune system.

The presence of microorganisms in the blood is a concern when transfusions are made, because it is possible to infect the blood recipient. Blood is routinely tested in an effort to eliminate this risk, especially for AIDS and hepatitis. Hepatitis is an infection of the liver caused by several different kinds of viruses. After recovering, hepatitis victims can become carriers. Although they show no signs of the disease, they release the virus into their blood or bile. To prevent infection of others, anyone who has had hepatitis is asked not to donate blood products.