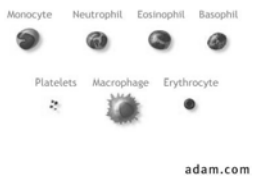
RCS 6080
Medical and Psychosocial Aspects of Rehabilitation Counseling

Hematological Disorders

Hematological Disorders

- Normal blood cells are produced in the bone marrow
- Types of blood cells
  - Platelets: blood clotting cells
  - Red blood cells: carry oxygen to the tissue
  - White blood cells: help fight infection

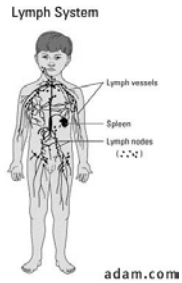


Lymphoma

- Lymphomas are a malignant proliferation of lymphocytes – either B or T
- 3% of all cancers in the US result from lymphomas
- The lymphomas are classified by the appearance of malignant lymphocytes on biopsy of tumor
- 3 categories
  - Low-grade
  - Intermediate-grade
  - High-grade

Functional Presentation of Lymphoma

- People present with swollen, growing lymph glands (nodal disease) or tumors in other organs (extramodal disease)
- Person can be asymptomatic
- Common B symptoms include fever, drenching night sweats, loss of 10% of body weight, and pruritis (severe itching)



Staging of Lymphoma

- Stage I – involvement of a single lymph node region or single extranodal organ or site
- Stage II – involvement limited to one side of the diaphragm with 2 or more lymph node regions
- Stage III – involvement of lymph node regions on both sides of the diaphragm
- Stage IV – diffuse or disseminated involvement of one or more extralymphatic organs

Treatment and Prognosis of Lymphoma

- Since the majority of lymphomas present in multiple areas of the body, localized surgery or radiation is rarely curative
- Primarily, treatment is chemotherapy
- Prognosis is dependent on the grade and stage
- For people who do not respond to primary treatment, bone marrow transplantation is increasingly used
- Biological agents and vaccine therapy were being tested

Leukemia

- Acute leukemia is characterized by an abnormal proliferation of immature white blood cells, called blasts or progenitor cells
- Two main forms of acute leukemia
  - Acute lymphoblastic leukemia
    - A cancer at the earliest stages of lymphocyte maturation
    - Occurs more often in the young
  - Acute nonlymphoblastic leukemia
    - Usually a malignancy of the myeloblast
    - More common in adults

Functional Presentation of Leukemia

- People with leukemia present with signs and symptoms of low red blood cell count (anemia), decreased white blood cells (granulocytopenia) with infection and fever, and a low platelet count (thrombocytopenia) with bleeding
- People will usually present critically

Treatment of Leukemia

- The course of treatment includes red blood cell transfusions to correct the anemia, treatments for infections caused by the lack of mature white blood cells, platelet transfusions to stop any bleeding, and starting chemotherapy to kill the leukemia cells
- Once chemotherapy stops, tumor cells die, the normal stem cells in the marrow that are resistant to chemotherapy divide, and their progeny cells mature and repopulate the marrow over the next 3 weeks

Vocational Implications of Lymphoma and Leukemia

- Depression, sleep disorder, and anxiety over personal appearance are common
- Long term survivors also have persistent problems including decreased energy level, negative body image, depression, employment problems, and marital problems
- Vocational implications and accommodations are similar to other cancers and are based on symptoms and side effects of treatment

Hemophilia

- Hemophilia is a sex linked hereditary blood disease characterized by greatly prolonged coagulation time
  - Hemophilia A is due to a deficiency of blood coagulation Factor VIII
    - Accounts for 75% of hemophilia
    - Incidence is 1 in 10,000 male births
  - Hemophilia B is due to a deficiency of blood coagulation Factor IX
    - Incidence is 1 in 75,000 male births
    - Is clinically indistinguishable from hemophilia A

Functional Presentation of Hemophilia

- The person can present with mild, moderate, or severe hemorrhagic disease, depending on the amount of active protein produced
  - People with mild hemophilia rarely bleed spontaneously and usually are discovered after excessive bleeding secondary to trauma or surgery
  - People with moderate hemophilia have rare episodes of spontaneous bleeding, but can hemorrhage with any trauma
  - People with severe hemophilia have frequent spontaneous hemorrhage from early childhood

Functional Presentation of Hemophilia

- People with hemophilia can bleed anywhere, but bleeding into joints (hemarthrosis), soft tissue (such as muscle), urine (hematuria), and the brain are common
- Chronic bleeding into joints or an acute bleed into the brain or spinal canal can lead to chronic disabilities, both functional and psychological

Treatment and Prognosis of Hemophilia

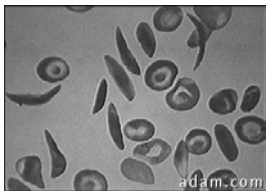
- The general principle of treatment of hemophilia is, first, to avoid drugs than can interfere with clotting, particularly aspirin and other NSAIDS that inhibit platelet function
- Second, early recognition of bleeding episodes or potential trauma and treatment with replacement Factor VIII or IX is imperative
- Prognosis has improved with the advent of factor concentrate treatment in the 1960s, with fewer severe bleeds, less crippling arthritis from hemarthrosis, and less intracranial bleeding

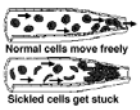
Vocational Implications of Hemophilia

- Vocational training should stress jobs that limit potentially hazardous situations
- People with hemophilia who are on effective replacement therapy can compete equally for most jobs
- For people with severe hemophilia, the ability to self- infuse agents effectively reduces morbidity and loss of work time

Sickle Cell Disease

- Sickle cell disease causes the red cell to assume a nonpliable sickle shape
- The resultant cellular defect leads to the main manifestations of the disease, which include:
  - premature death of the cells (hemolytic anemia)
  - vascular occlusion of vessels and subsequent tissue infarction
  - increased susceptibility to infection





Sickle Cell Disease

- A person with sickle cell is homozygous for the abnormal gene, therefore, both parents must be heterozygous for the abnormal gene
- The frequency of one abnormal gene is the African-American population is 1 in 12 and the incidence of sickle cell anemia is 1 in 650
- The frequency of the gene is also high in Mediterranean and African populations

Functional Presentation of Sickle Cell Disease

- People with sickle cell disease usually present in the first decade of life with complications of the three main characteristics of the disorder
  - Anemia
  - Vascular occlusion
  - Increase susceptability to infections, particularly pneumococcal pneumonia

Complications of Sickle Cell Disease

- pain episodes
- strokes
- increased infections
- leg ulcers
- bone damage
- yellow eyes or jaundice
- early gallstones
- lung blockage
- kidney damage and loss of body water in urine
- painful erections in men (priapism)
- blood blockage in the spleen or liver (sequestration)
- eye damage
- anemia
- delayed growth

Treatment of Sickle Cell Disease

- There is no specific treatment for sickle cell disease, therefore, most therapy is supportive in treatment of the complications
- Early recognition of infection, administration of prophylactic antibiotics, and vaccination may forestall or prevent other complications
- If a painful crisis persists or there is infection of a major organ (brain, lung, or heart), exchange transfusion is performed to remove some of the sickle red cells - the effect is temporary

Treatment of Sickle Cell Disease

- General guidelines
  - Taking the vitamin folic acid (folate) daily to help make new red cells
  - Daily penicillin until age six to prevent serious infection
  - Drinking plenty of water daily (8-10 glasses for adults)
  - Avoiding too hot or too cold temperatures
  - Avoiding over exertion and stress
  - Getting plenty of rest
  - Getting regular check-ups from knowledgeable health care providers

Prognosis of Sickle Cell Disease

- Prognosis has improved with good supportive care, and many people with sickle cell disease survive into middle age
- However, frequent admissions for painful crises, the complication of sickle cell disease, narcotic use and abuse due to chronic pain, and absence from school and work lead to significant psychological and vocational problems

Vocational Implications of Sickle Cell Disease

- The greatest dysfunction was found in the areas of employment, finances, sleep habits, and performance of daily activities
- The implications of these findings suggest a strong need for vocational rehabilitation services, training in areas of communication and self-esteem, medical treatment, and psychological help for depression and drug dependence
- Advisable that these individuals stay away from jobs that cannot be interrupted to take fluids, jobs that involve extreme temperature changes, and jobs with lower O_2 concentration

Additional Resources and Information from the Web

- Leukemia and Lymphoma Society (www.leukemia.org)
- Lymphoma Information Network (www.lymphomainfo.net)
- Lymphoma Forum and Lymphoma Association (www.lymphoma.org.uk)
- Children's Leukemia Research Association (www.charities.org/member/clra.html)
- World Federation of Hemophilia (www.wfh.org)
- National Hemophilia Foundation (www.hemophilia.org)
- Sickle Cell Society (www.sicklecellsociety.org)
- Sickle Cell Information Center (www.emory.edu/PEDS/SICKLE)