MEDAVIE HealthEd ÉduSanté

# Hematology and Oncological Emergencies

## Advanced Care Paramedicine

Module: 04

Section: 07b

# Introduction

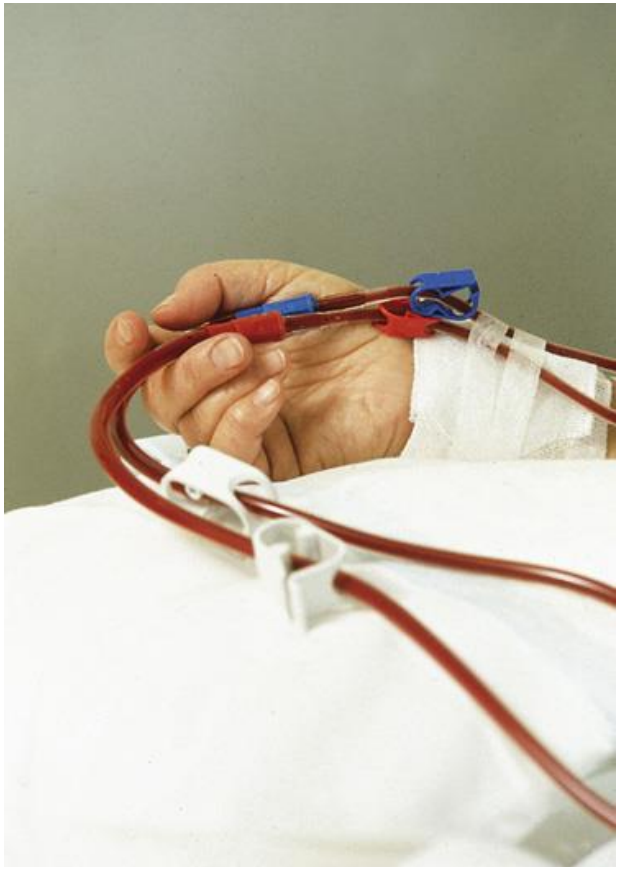
- Hematology
  - Study of blood and blood forming organs
- Disorders common
  - Rarely the primary problem
  - Genetic or acquired
  - Presentations tend not to point to a specific disease process

# Blood Products and Typing

- Transfusion
  - Transplantation of blood or a component of blood
- Antigen
  - Surface proteins that distinguish self from not self
  - Must be compatible for a transfusion to be successful
- Cross-matching
  - Checking donor and recipient samples for compatibility

# Antigens

- A (39% of population)
- B (11% of population)
- AB (5% of population)
  - Universal recipient
- O (45% of population)
  - Universal donor
- Rh factor
  - Positive or negative



# Blood Products and Blood Typing

**Table 35-1 Types of Transfusions**

| Type of Transfusion | Contents | Use |
|---|---|---|
| Whole blood | All cells, platelets, clotting factors, and plasma | Replace blood loss from hemorrhage |
| Packed red blood cells (PRBCs) | Red blood cells and some plasma | Replace red blood cells in anemic patients |
| Platelets | Thrombocytes and some plasma | Replace platelets in a patient with thrombocytopenia |
| Fresh frozen plasma (FFP) | Plasma, a combination of fluids, clotting factors, and proteins | Replace volume in a burn patient or in hypovolemia secondary to low oncotic pressure |
| Clotting factors | Specific clotting factors needed for coagulation | Replace factors missing due to inadequate production as in hemophilia |

# Transfusion Reactions

- Hemolytic reactions
- Febrile nonhemolytic reactions
- Circulatory overload

# Hemolytic Reactions

- Blood types not compatible
  - Donor blood trigger antibody response in recipient
  - Hemolysis occurs
- Presentation
  - Facial flushing, hyperventilation
  - Tachycardia, chest pain
  - Wheezing
  - Hives fever, chills, and cyanosis

# Febrile Nonhemolytic Reactions

- Most common transfusion reaction
- Sensitization of antigens on
  - White blood cells
  - Platelets
  - Plasma proteins
- Presentation
  - Headache, fever, chills

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# Identification and Management

| Type of Reaction | Incidence/Treatment | Signs and Symptoms | Etiology |
|---|---|---|---|
| Febrile (Non-Hemolytic) | • Most common type of reaction (1:200-1:100)<br>• Usually occurs in patients with a history of previous transfusions or pregnancies<br>• Premedication with acetaminophen | • Rise in temperature 1C above baseline, rigors<br>• Headache, malaise, vomiting | • Antibody to donor leukocytes<br>• Accumulated cytokines in component bag |
| Acute Hemolytic Reaction (Immediate) | • ·Treatment is aimed primarily at prevention of renal failure (IV fluids and diuretics)<br>• ·Incidence is 1:33,000-1:12,000 | • ·Acute onset of symptoms(often within the first 15 minutes)<br>• ·Fever, chills<br>• ·Hemoglobinuria, renal failure, hypotension, DIC, oliguria, oozing from IV site, back pain, pain along infusion vein | Due to administration of incompatible blood:<br>1. Crossmatch error.<br>2. Wrong identification of blood specimen.<br>3. Blood administered to wrong patient. |
| Delayed Hemolytic Reaction | • Incidence is 1:11,000-1:5,000<br>• Occur more often in patients with a history of multiple transfusions or pregnancies.<br>• Transfuse with antigen negative rbc's as necessary | • Weakness, unexplained fall in posttransfusion hemoglobin, elevated serum bilirubin | Immune response to rbc antigens |
| Circulatory Overload | • Incidence is 1:10,000-1:100<br>• Head of bed should be kept elevated.<br>• Oxygen and diuretics may be ordered. | • Dyspnea, orthopnea, productive cough with pink, frothy sputum, tachycardia, hypertension, headache. | Volume overload. |
| Bacterial contamination | • Incidence is 1:1700 in pooled platelet units and 1:500,000 in red cell components<br>• Treatment of shock, renal failure and DIC<br>• Antibiotics | • Fever, shock, disseminated intravascular coagulation (DIC) | Bacteria originating from:<br>1. The donor venipuncture site or<br>2. Donor with an undiagnosed bacteremia<br>3. Proliferation during storage |
| Allergic Reaction | • Incidence is 1:100-1:33<br>• Antihistamines | • Pruritis, rash, urticaria, flushing | Antibody to donor plasma proteins. |
| Anaphylactic | • Incidence is 1:170,000-1:18,000<br>• Epinephrine<br>• Antihistamines | • Urticaria,erythema, anxiety, respiratory distress, hypotension, laryngeal /pharyngeal edema, bronchospasm | Antibody to donor plasma proteins. |
| Transfusion Related Acute Lung Injury (TRALI) | • Incidence unknown but thought to occur as often as 1:5000<br>• Treat ARDS - acute respiratory distress syndrome (oxygen therapy/ventilatory support steroids) | • Shortness of breath, hypoxemia, chills, fever, cyanosis, and hypotension<br>• Xray findings consistent with pulmonary edema but with no evidence of cardiac failure | Transfused antibodies to HLA or white cell antigens which may react with recipients leukocytes. |
| Hypotensive Reaction | • Unknown/Administer saline bolus | • Hypotension, erythema, brachycardia, nausea | • Hypothesized as generation of brachykinins +/- ACE Inhibitor use |

# Management

- Support ABCs
- Stop trxansfusion
- Change all IV tubing
- Initiate IV therapy
  - Normal saline or lactated Ringer's
- Consider medications
  - Furosemide
  - Dopamine
  - Diphenhydramine
- Differentiate between hemolytic and nonhemolytic reactions

# Assessment

- History
  - Hematological disorders are rarely the chief complaint
- Physical assessment
  - Nervous system
  - Skin signs
  - Lymphatic signs
  - Gastrointestinal signs
  - Musculoskeletal signs
  - Cardiorespiratory signs
  - Genitourinary signs

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# Specific Patient Problems

- Diseases of the Red Blood Cells
- Diseases of the White Blood Cells
- Diseases of the Platelets/Blood Clotting Abnormalities
- Other Hematopoietic Disorders

# Diseases of Red Blood Cells

- Polycythemia
  - Excess of red blood cells
  - Uncommon but several diseases can cause it
- Anemia
  - Lack of red blood cells or inadequate hemoglobin
- Impairment of red cell function
  - Problems with hemoglobin structure and function
  - Problems with red blood cell membrane

# Polycythemia

- Overproduction of erythrocytes.
  - Occurs in patients > 50 years old or with secondary dehydration.
- Results in bleeding abnormalities:
  - Epistaxis, spontaneous bruising, GI bleeding.
- Management:
  - Follow general treatment guidelines

# Anemia

- Result of a lower than normal level of erythrocytes
- Can be caused by
  - Blood loss (as in surgery)
  - Diseases (cancer, kidney disease or HIV)
  - Treatment (chemotherapy, AZT)
  - Deficiencies (Fe, folate and/or vitamin B12)
- Not a separate disease process
  - Signs and symptoms may not be present until the body is stressed
  - Differentiate chronic anemia from acute episode

# Anemia

- Treatments
  - Maximize oxygenation and limit blood loss
  - Establish IV therapy if indicated
  - Based on cause
    - Increase nutrient intake
      - Vitamins, or iron via pills or injections
    - Correct blood loss
    - Medication to improve hemoglobin
      - related to chronic disease
      - recombinant erythropoietin
    - Blood transfusion.
      - For some types of anemia (such as thalassemia major, aplastic anemia, or sickle cell disease) regular blood transfusions may be required to manage anemia and to avoid serious complications.
    - Bone marrow transplant

# Bleeding Disorders

- Hemophilia
- Von Willebrand Disease
- Clotting Factor deficiencies
- Platelet function disorders

# Hemophilia

- Blood does not clot normally
- Does not bleed more profusely or more quickly than other people
- External wounds are usually not serious.
  - Internal hemorrhaging
    - joints (especially knees, ankles and elbows)
    - tissues and muscles
    - When bleeding occurs in a vital organ, especially the brain, can be life threatening

- Hemophilia A
  - Classical hemophilia
  - Factor VIII deficiency hemophilia
    - Lack of the Factor VIII protein
- Hemophilia B
  - Christmas Disease, named after Steven Christmas, a Canadian who in 1952 was the first person to be diagnosed with this distinct form of hemophilia
  - Factor IX deficiency hemophilia

# Von Willebrand Disease

- Most common bleeding disorder
- Is a family of related diseases (Classed in 3 types)
- All the different types are caused by a problem with the Von Willebrand Factor (VWF).
  - A protein in blood which is necessary for proper blood coagulation, or clotting. The genes that make VWF are "turned on" in two cell types in the body:
    - the lining cells of blood vessels (endothelial cells) and platelets
  - When there is not enough VWF in the blood, or when it does not work the way it should, the blood takes longer to clot

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# Clotting Factors

| Factor | Other Name | Bleeding Severity |
|---|---|---|
| I | Fibrinogen | Usually mild, except with complete absence of fibrinogen |
| II | Prothrombin | Usually mild |
| V | Parahemophilia | Usually mild |
| V and VIII | | Usually mild |
| VII | Alexander's | Severe when Factor VII levels are low |
| VIII | Hemophilia | Severe when Factor VIII levels are below 1% |
| IX | Hemophilia B | Severe when Factor IX levels are below 1% |
| X | Stuart-Prower | Moderate to severe when Factor X levels are below 10% |
| XI | Hemophilia C | Mild to moderate when Factor XI levels are below 15% |
| XIII | | Severe |

# Hemolytic Disease

- $Rh^-$ Mother and $Rh^+$ Father
- Baby inherits father's with $Rh^+$
- Mother develops anti-Rh agglutinins
- Next pregnancy with $Rh^+$ baby, mother's anti-Rh agglutinins will be activated and cause erythroblastosis fetalis
  - Hemolysis of baby's RBC's occurs

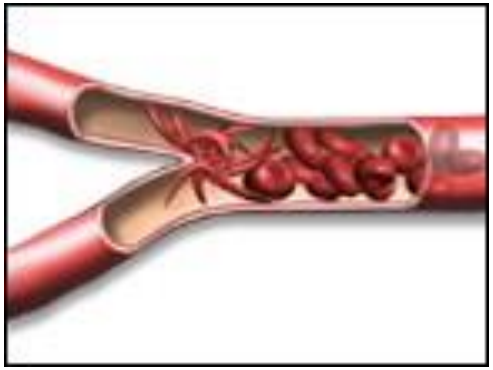
# Hemolytic Disease

- This decrease $O_2$ carrying capability and increase bilirubin levels
- This may lead to brain damage
- $Rh^-$ Mothers with $Rh^+$ babies are given medication to keep her system from forming the anti-Rh antibodies

# Hemolytic Disease

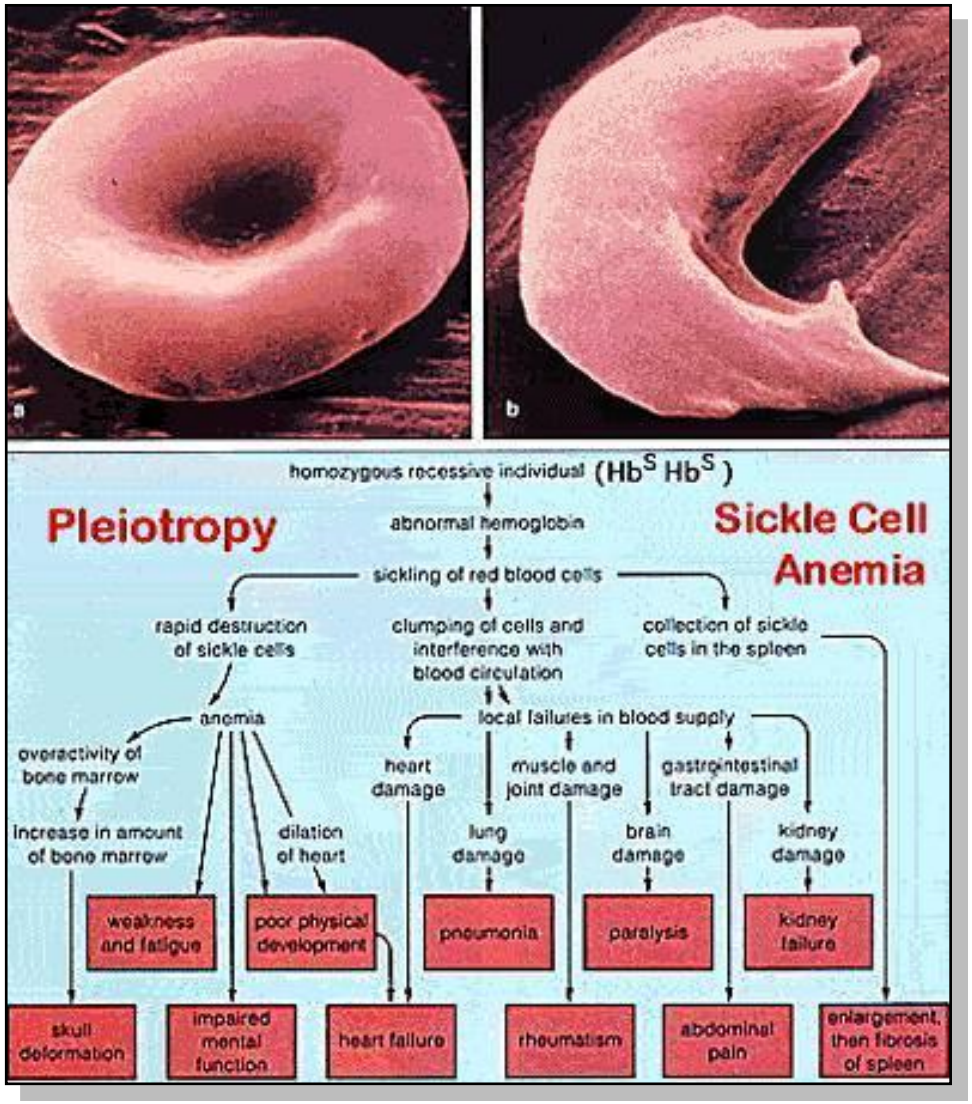
- Baby will soon replace $Rh^+$ with $Rh^-$ if hemolytic disease develops
- $Rh^-$ depletes through normal life span and as the $Rh^-$ transfusion is depleted $Rh^+$ is reintroduced through the bone marrow

# Sickle Cell Disease



- An inherited disease of RBC's which can cause attacks of pain and damage to vital organs and can lead to early death.
- Affects hemoglobin

- Disease occurs when a person inherits two sickle cell genes or a combination of one sickle cell gene plus any one of several other abnormal hemoglobin genes.
- Sickle cells tend to become trapped and to be destroyed in the liver and in the spleen.
  - Results in anemia


a
b
homozygous recessive individual ($Hb^S Hb^S$)
Pleiotropy
abnormal hemoglobin
Sickle Cell Anemia
sickling of red blood cells
rapid destruction of sickle cells
clumping of cells and interference with blood circulation
collection of sickle cells in the spleen
anemia
local failures in blood supply
overactivity of bone marrow
heart damage
muscle and joint damage
gastrointestinal tract damage
increase in amount of bone marrow
dilation of heart
lung damage
brain damage
kidney damage
weakness and fatigue
poor physical development
pneumonia
paralysis
kidney failure
skull deformation
impaired mental function
heart failure
rheumatism
abdominal pain
enlargement, then fibrosis of spleen

# Platelet Function Disorders

- Common symptoms of platelet function disorders are:
  - Frequent nose bleeds
  - Easy bruising
  - Bleeding from the gums when baby teeth fall out or after tooth extractions
  - Heavy menstrual bleeding (menorrhagia)
  - Menstrual bleeding that lasts longer than 7 days
  - Bleeding into the stomach or intestine
  - Heavy or prolonged bleeding after serious injury or at the time of surgery
- These symptoms may be so mild that many people do not notice them
- Realize only when they have a serious injury or surgery

# Diseases of the Platelets

- Affect platelets or clotting
- Hereditary or acquired
- Thrombocytosis
  - Increased platelets
- Thrombocytopenia
  - Reduced platelets

# Diseases of White Blood Cells

- Body's principle defence system
- Leukopenia/neutropenia
- Leukocytosis

# Diseases of White Blood Cells

- Leukopenia/Neutropenia
  - Too few white blood cells or neutrophils.
  - Bacterial infections are a real risk
  - Follow general treatment guidelines and provide supportive care.
- Leukocytosis
  - An increase in the number of circulating white blood cells, often due to infection.
  - Leukemoid reaction

# Cancer

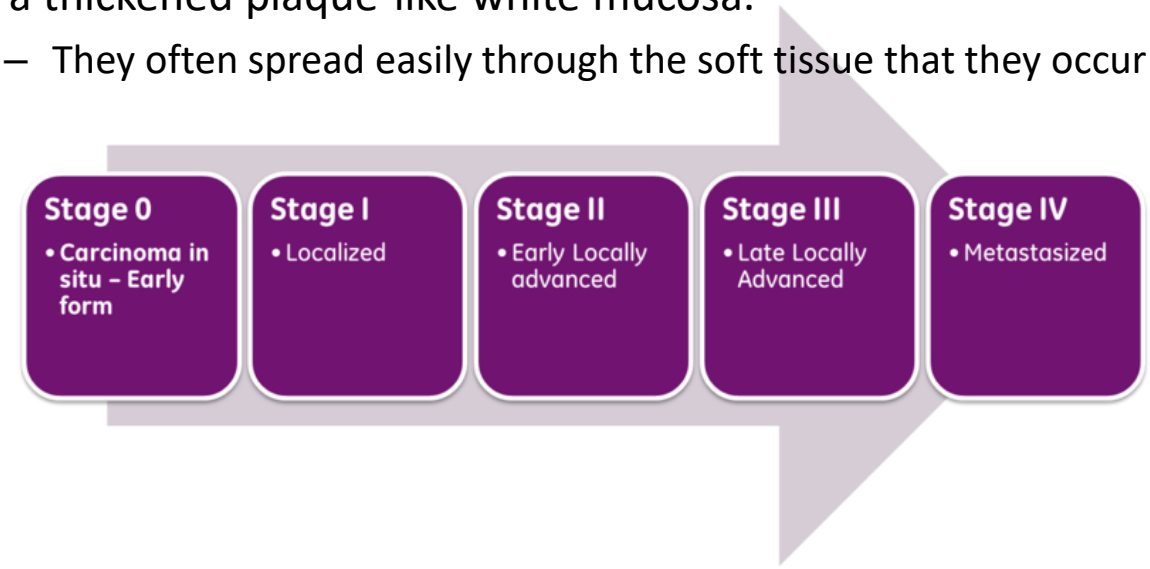
- Occurs when body cells become abnormal and divide without control or order.
- Tumours can be either:
  - Benign (non-cancerous)
    - cells stay in one place in the body and are not usually life-threatening
  - Malignant (cancerous)
    - Can invade and damage tissues and organs near the tumor.
    - Can break away and enter the bloodstream or lymphatic system.
    - This is how cancer spreads from the original (primary) tumor to form new tumors in other parts of the body.
- The spread of cancer is called metastasis.
  - Cancers are named after the part of the body where they start.
    - Cancer that starts in the colon but spreads to the liver is called colon cancer with liver metastases.

# Cancer

- Classifications
  - Carcinoma
  - Sarcomas
  - Leukemia
  - Lymphoma
  - Multiple Cell Myeloma

# Cancer

- Carcinoma
  - A cancerous growth that develops in the skin, or in glandular tissue ex. tissue of the breast or prostate.
  - Two types:
    - Squamous cell carcinomas generally occur in areas that have chronic exposure to sunlight.
    - Adenocarcinomas generally occur on mucus membranes and are first seen as a thickened plaque-like white mucosa.
      - They often spread easily through the soft tissue that they occur in



# Cancer

- Sarcomas
  - Sarcomas grow in supportive and connective tissues such as bone, blood vessels, and muscle.
  - In young adults a sarcoma often develops as a painful mass on the bone.
  - Sarcoma tumors usually resemble the tissue in which they grow. That is why the lump on the bone is so painful.

# Cancer

- Leukemia
  - Mostly develops in the bone marrow.
  - Leukemia is a Greek word that translates to "white blood“
  - Often associated with the overproduction of immature white blood cells.
    - These immature white cells do not function normally therefore the patient is often prone to infection.
    - Leukemia also effects red blood cells and can therefore cause poor blood clotting and tiredness due to anemia.

# Leukemia

- Cancer of hematopoietic cells
  - Precursors of white blood cells
  - Produces abnormal and ineffective cells
  - Proliferate and spread
- Classified by cells involved
  - Acute lymphocytic leukemia (ALL)
  - Acute myelogenous leukemia (AML)
  - Chronic lymphocytic leukemia (CLL)
  - Chronic myelogenous leukemia (CML)

# Leukemia

- Initial presentation
  - Acutely ill, fatigued, febrile and weak, anemic.
  - Often have a secondary infection.
- Management
  - Follow general treatment guidelines.
  - Utilize isolation techniques to limit risk of infection.

# Cancer

- Lymphoma
  - Lymphoma generally occurs in the lymph nodes and the spleen
    - There are over 29 different types of lymphoma. The type of cells that grow in the tumor distinguishes each type
  - Swelling of the lymph nodes
  - Fever, night sweats, anorexia, weight loss, fatigue, and pruritis
  - Two main forms
    - Hodgkin's disease (chance for long term survival better)
    - Non-Hodgkin's lymphoma

# Cancer

- Multiple Cell Myeloma
  - Cancer of plasma cells
  - Collections of abnormal plasma cells accumulate in the bone marrow where they interfere with the production of normal blood cells
  - Most cases of myeloma also feature the production of a paraprotein (an abnormal antibody which can cause kidney problems)
  - Bone lesions and hypercalcemia are also often encountered

# Cancer

- Locations
  - May be found in most tissue of the body

# Cancer

- Treatments
  - Chemotherapy
  - Radiation
  - Surgery
  - Complementary and alternative therapy
  - Side effect management

# Chemotherapy

- Chemotherapy is a method of treating disease with the use of drugs or medications
- Treats the whole body rather than just a specific area
- Often used to destroy cancers of the blood and bone marrow, as well as cancers of the lymphatic system
- Also used to destroy metastasized cells
  - the idea is to wipe out these cancerous cells before they have a chance to multiply and form a new tumour
- May be given IV, PO, intraperitoneal, intra-arterial and CNS delivery

# Chemotherapy

- Side effects
  - N/V (Most immediate)
    - Occur because of the effect chemotherapy drugs can have on the vomit center centre of the brain and the GI tract, the stomach and the bowels
  - Late (delayed) side effects
    - hair loss
    - sore mouth
    - digestive problems such as nausea, diarrhea or constipation
    - Infections (due to low blood counts)
    - Usually start during the first few weeks of treatment

# Radiation

- About 1/3 require no other type of treatment
  - Remainder receive radiation combined with chemotherapy and/or surgery
- In general, radiation therapy works best on a fixed tumour, or group of tumours, that are in an area at which x-ray beams can be directly aimed.
    - Hodgkin's disease
    - Cervical cancer
  - If the tumour is in an area where healthy organs would be harmed by the radiation, then radiation therapy cannot be used.
- Sometimes used to shrink a tumour so that it becomes small enough to be removed by surgery.
- Can be used after surgery to destroy any cancer cells that may have been missed.
- May destroy healthy cells

# Alternative Therapies

- Therapies are not usually provided by doctors or other conventionally trained healthcare providers
- Complementary therapies are used along with conventional therapies.
- Alternative therapies are used instead of them
  - herbal preparations
  - reflexology
  - acupuncture
  - Traditional Chinese medicine
- Complementary and alternative therapies have not been scientifically proven to be safe or effective in the treatment of cancer.

# Side Effects

- Fatigue and Flu
- Eating and Appetite
  - N/V
  - Loss of appetite
  - Changes in taste or smell
  - Swallowing difficulties
  - Heartburn and reflux
  - Gas, bloating or cramping
  - Diarrhea
  - Constipation
- Fertility
- Hair Loss
- Sore and/or dry mouth, thick saliva

# Oncological Emergencies

- Broadly defined as any complication related to cancer or anticancer therapy that requires immediate intervention
  - Acute Bleeding
  - Brain metastases, Increased ICP & Seizures
  - Febrile neutropenia
  - Hyperviscosity Syndrome
  - Malignancy associated hypercalcemia
  - Malignant airway Obstruction
  - Malignant epidural spinal cord compression
  - Superior vena cava obstruction
  - Syndrome of inappropriate ADH Secretion
  - Tumor lysis syndrome

# Acute Bleeding

- DIC
  - Most commonly seen with acute myelocytic leukemia, adenocarcinoma, septicemia or transfusion reactions
- GI bleeding
  - Upper GI bleeding can be caused by primary upper GI malignancies, a number of non-malignancy related causes (peptic ulcer disease, esophageal and gastric varices, hemorrhagic gastritis, etc.)
  - Lower GI bleeding can be caused by primary upper and lower GI malignancies, non-malignancy related causes (diverticular disease, ischemic colitis, inflammatory bowel disease, hemorrhoids, etc.) and various cancer therapies (e.g., graft-versus-host disease following stem cell transplantation, radiation-induced proctosigmoiditis, etc.).
- Hematuria
  - Bleeding anywhere along the urinary tract secondary to drug or radiation-induced damage, infection or progression of cancer
  - Drug-induced cystitis is most commonly seen in patients receiving cyclophosphamide or ifosfamide
  - Radiation-induced cystitis results from damage to the vascular endothelium and endarteritis, resulting in progressive ischemia, inflammation, fibrosis and tissue necrosis
- Hemoptysis
  - Is the most immediate life-threatening symptom of progressive intrathoracic disease
  - Massive hemoptysis can lead to asphyxiation or exsanguination
  - The primary causes are malignancy, infection and hemostatic abnormalities

# Brain Metastases, Increased ICP and Seizures

- Represent the most common type of brain malignancy (occur in 10 – 30 % all adult patients with cancer)
- Any primary tumour can metastasize to the brain
  - lung cancer, breast cancer, and melanoma are the most common (70 – 90% of all cases)
  - Multiple brain metastases
    - Melanoma and lung cancer are most frequently
  - Solitary metastases
    - Breast, colorectal, and renal cancers are more likely
- Can lead to:
  - Increased ICP
  - Status epilepticus

# Febrile Neutropenia

- The development of fever, often with other signs of infection, in a patient with neutropenia (an abnormally low number of neutrophil granulocytes)
- Most common complications related to cancer therapy
- Considered a potentially life-threatening (2 – 20% mortality)
- Risk for development is 25 to 40% in adult patients with cancer
- Associated Risks
  - Older age
  - Prior treatment with chemotherapy
  - Compromised hepatic, cardiovascular, or renal function
  - Abnormal bone marrow
  - Low baseline white blood cell counts
  - Concomitant use of immunosuppressive medications

# Hyperviscosity Syndrome

- A group of clinical symptoms related to increases in blood viscosity leading to adverse effects on tissue perfusion
- Causes include:
  - Increased protein content and large molecular size
  - Abnormal polymerization
  - Abnormal shape of immunoglobulin molecules
- Can occur secondary to a variety of hematologic malignancies
  - Most common being Waldenström macroglobulinemia (80%)
  - Less frequent with multiple myeloma, leukemia, polycythemia
- Suspect in pt's of known hematological malignancy that present with symptom triad:
  - Bleeding, visual disturbances, and/or focal neurologic signs

# Malignancy Associated Hypercalcemia

- Defined as a ***corrected*** serum calcium > 2.6 mmol/L
- Occurs in up to 30% of patients with cancer
  - Most commonly breast, lung and head/neck tumours, multiple myeloma and adult T-cell leukemia/lymphoma
    - Humoral hypercalcemia accounts for majority of cases (80%)
    - Osteolytic bone metastases account for 20%

# Malignant Airway Obstruction

- Most commonly caused adjacent tumour (lung cancer, thymoma) or a primary tumour of the head and neck

# Malignant Epidural Spinal Cord Compression

- Common neurological complication that affects up to 5% of adult patients with cancer
- Most common in widespread metastatic disease
- A true oncologic emergency that needs rapid diagnosis and treatment
  - Left untreated it can lead to progressive pain, sensory loss, incontinence, and irreversible paralysis
  - S/S
    - Back pain (earliest and most common – 90%)
    - Motor weakness and sensory impairment
    - Autonomic dysfunction (Late and poor prognosis)

# Superior Vena Cava Obstruction

- Partial or complete obstruction of blood flow through the superior vena cava to the right atrium
  - May be caused by compression, invasion, thrombosis, or fibrosis
  - Lung cancer (small cell and non-small cell) and non-Hodgkin lymphoma account for 85%

# Syndrome of Inappropriate ADH Secretion (SIADHS)

- Results from the inappropriate production and secretion of antidiuretic hormone which leads to:
  - Water retention/intoxication
  - Hyponatremia
  - Hyposmolality
  - In cancer patients can be caused by the ectopic production of ADH by tumour tissue
  - Small cell lung cancer most common cause

# Tumor Lysis Syndrome

- Massive cellular breakdown in tumours and subsequent release of intracellular contents into the bloodstream
- Characterized by
  - Hyperuricemia
  - Major electrolyte disturbances (hyperkalemia, hyperphosphatemia and hypocalcemia)
- Can quickly lead to oliguric renal failure, seizures, cardiac arrhythmias and death