Erythrocytosis

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Today's Discussion

- Introduction
- Absolute Erythrocytosis
  - Secondary Erythrocytosis
- Relative Erythrocytosis

Erythrocytosis

- Condition in which the red blood cells, hemoglobin, and hematocrit values are elevated

Separated into Two groups based on the determination of Red Cell Mass:
• Absolute Erythrocytosis • Relative Erythrocytosis

Erythrocytosis

Absolute Erythrocytosis

- Red cell mass is elevated → true increase in the number of circulating erythrocytes

Relative Erythrocytosis

- Increase in Hct with the absence of the elevation in red cell volume
  - Plasma level does not increase as the RBC level increases

Absolute Erythrocytosis

Absolute Erythrocytosis

- Red cell mass (or red cell volume) is elevated
  - True increase in number of circulating erythrocytes
- Divided into 3 distinct groups:
  - Polycythemia Vera
    - Chronic myeloproliferative disorder arising as a clonal hematologic malignancy of the BM
  - **Secondary Erythrocytosis**
    - Physiologic response to abnormal stimulus
      - Tissue hypoxia, increased erthryopoietic activity
  - Idiopathic group
    - Neither myeloproliferative nor secondary cause of sustained erythrocytosis can be implicated

Secondary Erythrocytosis

- Diagnosed by an increased secretion of erythropoietin
  - Increase production of RBCs by the BM

- Separated into 3 groups:
  - Appropriate, compensatory increase in EPO
  - Inappropriate or pathologic secretion of EPO
  - Increase from defective oxygen transport

Secondary Erythrocytosis

Appropriate, compensatory increase in EPO

- Release of erythropoietin from a compensatory effect to minimize impending tissue hypoxia
- Abnormal hemoglobin
  - High-affinity variants with abnormal oxygen affinity
  - AA substitutions in variants interferes with release of oxygen to the tissue
    - Prevent normal conformational changes → deoxygenation → tissue hypoxia
      - Kidney senses hypoxia and respond by increasing the release of EPO
  - Characterized by Left shift oxygen-dissociation curve
  - Seen in β-thalassemia minor and other rare hemoglobinopathies
  - Most individuals are asymptomatic and have no physical symptoms except ruddy complexion

Secondary Erythrocytosis

Appropriate, compensatory increase in EPO

- Methemoglobinemia
  - Caused from hereditary deficiency of the enzyme NADH-methemoglobin reductase
    - Due to hemoglobin M disease, various drugs, or toxic substance exposure
  - Methemoglobin (ferric state) is incapable of carrying oxygen and cyanosis is observed
  - Mild, associated erythrocytosis
    - Result of a shift to the left of the oxygen-dissociation curve (increased oxygen affinity)

Secondary Erythrocytosis

Appropriate, compensatory increase in EPO

- Cardiac or respiratory diseases that lead to significant arterial oxygen desaturation
  - Most common
  - Most common lung disease is COPD
- Caused by ascent to high altitudes due to low atmospheric pressure
  - Most normal individuals experience no symptoms
  - Acute mountain sickness- travel to high altitude too quickly
- Sleep apnea

Secondary Erythrocytosis

Inappropriate Increase in EPO

- Increase in EPO without tissue hypoxia
- Associated with a wide range of disorders
  - Renal disease
  - Benign and malignant tumors (liver, cerebellum, and uterus)
- No physiologic advantage and causes the clinical features observed with these associated disorders
  - Hct and Red cell mass are increased, no WBC or platelet count increase

Secondary Erythrocytosis

Defective Oxygen Transport

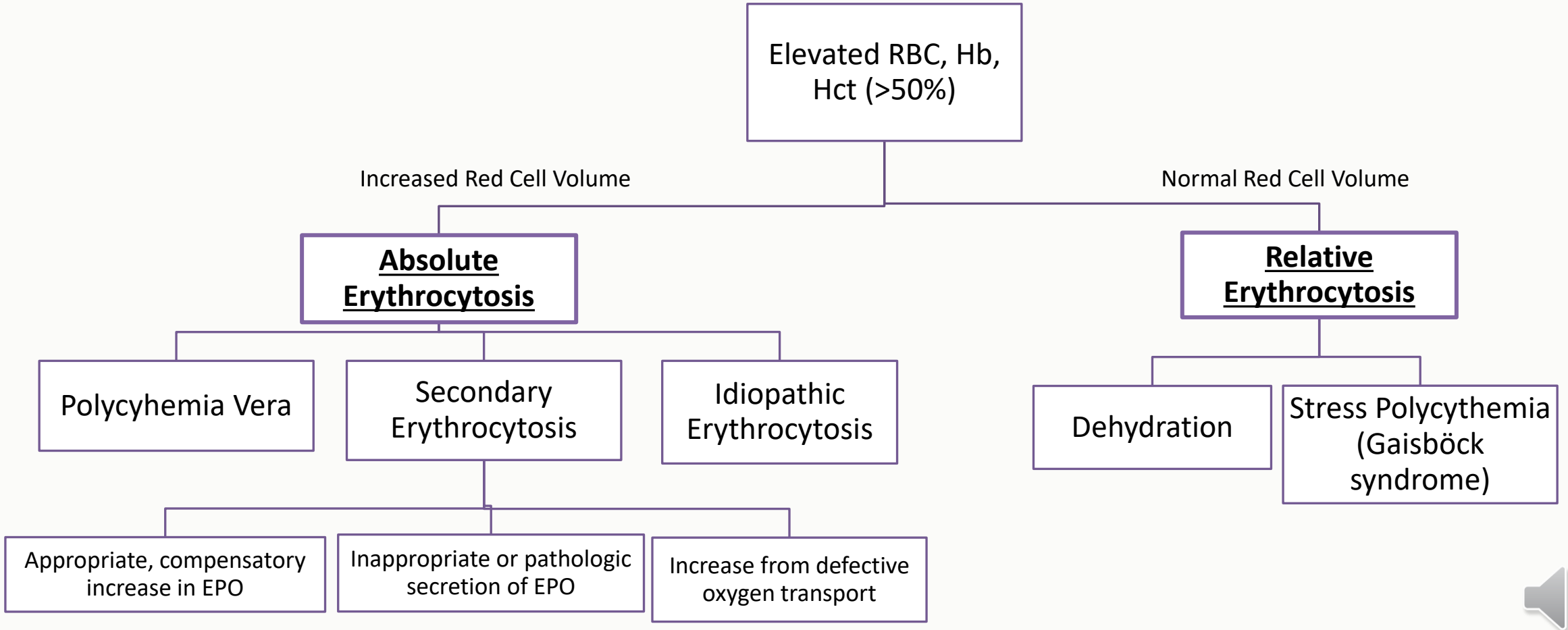
- Seen with heavy cigarette smoking due to chronic carbon monoxide intoxication
  - Hemoglobin is bound to carbon monoxide, the carboxyhemoglobin loses its capacity to carry oxygen
    - Results in tissue hypoxia
  - Left shift in the oxygen-dissociation curve
  - Increase in EPO will cause mild erythrocytosis
  - Approximately seen in 600 to 1,000 per 100,000 population
- Environmental pollution
  - Increase levels of carboxyhemoglobin
  - Mild erythrocytosis

Relative Erythrocytosis

Relative Erythrocytosis

- Elevated Hct, normal red cell mass, and decreased plasma volume
  - Hct between 50-60%
- Caused by:
  - Dehydration
    - Loss of body fluids causes the decrease in plasma volume
    - Ex. Burns, diuretics, vomiting, diarrhea
  - Stress erythrocytosis
    - Gaisböck syndrome
      - Asymptomatic middle-aged white men who are hypertensive, obese, and long history of smoking
      - Low plasma volume possibly associated with changes in hormone level
      - Usually benign

Classification of the Polycythemias



References

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