Essential Learning

High Altitude Cerebral Edema (HACE)

- **Differential Diagnosis**
  - The differential diagnosis for headache, weakness, nausea and vomiting in a patient at high altitude should include
    - acute mountain sickness
    - HACE
    - traumatic injury
    - Infection
    - alcohol or drug intoxication
    - CO poisoning
    - intracranial bleeding.
    - For any patient with respiratory symptoms at high altitude, HAPE (high altitude pulmonary edema) should also be considered

- **Difference between HACE and Acute Mountain Sickness (AMS)?**
  - AMS is characterized by flu-like symptoms including headache with a combination of nausea, vomiting, dizziness, anorexia, weakness, decreased urination, fatigue or sleep disturbance with recent ascent to greater than 2000m (6560 ft).
    - Symptoms typically abate between 15 - 94 hours with symptomatic treatment.
    - Appropriate treatment includes oxygen, NSAIDs, acetazolamide, +/- steroids and descent.
  - HACE occurs in those who recently ascended to > 8200 ft (2500m) who have progressive neurologic deterioration.
    - This typically starts with symptoms of AMS, then the patient begins to exhibit ataxia or altered mental status. Any neurological deficit should be concerning for HACE.
  - How often does AMS progress to HACE?
    - AMS progresses to HACE in 1-5% of cases.
  - The mainstays of treatment of HACE:
    - rapid descent and steroids.
    - Hyperbaric oxygen (Gamow bag) may be used as a temporizing measure.
    - Acetazolamide may be given for suspected Acute Mountain Sickness but offers limited benefit in HACE.
  - Death from HACE has been reported at as low as 8200 ft. HACE is most common at >12,000 ft.

- **Why is treatment of HACE so important?**

- Long-term neurologic sequelae of HACE including ataxia and cognitive impairment have been reported after recovery
- Once coma occurs, mortality exceeds 60%.
- Oxygen decreases intracranial blood flow at high altitude and steroids are likely to reduce long-term deficits.

- **What is considered high altitude?**
  - High altitude is a hypoxic environment with an elevation of >2440 m or >8000 ft
  - Although partial pressure of oxygen remains the same at altitude, the air pressure decreases.
    - In Denver at 5280 ft, the air pressure is 17% less than at sea level.
    - In Aspen at 8600 ft, the air has 26% less oxygen.
    - This is a common altitude in Colorado and several mountainous areas of the western United States.

- **How can one acclimatize and prevent HAPE/HACE?**
  - Natural acclimatization occurs when people hyperventilate causing respiratory alkalosis and bicarb diuresis.
  - Prophylactic treatment with Acetazolamide (125 mg PO q12 hours, started 48 hr before ascent and continued for 48 hr after ascent) works by causing bicarb diuresis and metabolic acidosis, thus triggering hyperventilation and speeding acclimatization.
  - Steroids may also be used for treatment (Decadron 8-10 mg IV, IM, or PO x 1, then 4 mg IV, IM, or PO q6 hours) to help reduce vasogenic cerebral edema.

- **What is the normal range of O2 saturation at high altitude? Does this change with acclimatization?**
  - At Sea level SaO2 96%
  - 5000 ft (1520 m) SaO2 95%
  - 7500 ft (2290 m) SaO2 92-93%
  - 15000 ft (4570 m) SaO2 86%
  - 20000 ft (7000 m) SaO2 76%
  - This changes minimally with acclimatization

- **What is the Pathophysiology of HACE?**
  - High-altitude exposure → hypoxemia → fluid retention and cerebral hypoxemia (mediator-induced permeability) → sympathetic discharge/peripheral vasoconstriction and increased pulmonary/cerebral blood volume → altered cerebral hemodynamics and increased cerebral blood flow and volume → capillary pressure increase → vasogenic edema → brain swelling → increased ICP = HACE

- **What is HAPE? How often are HAPE and HACE associated?**
  - High Altitude Pulmonary Edema (HAPE) is the most common lethal altitude illness.
  - This may occur at altitude >3000m (9500ft).

- Patients present with pneumonia-like symptoms and shortness of breath at rest, cough, hypoxia and fever. Clinical progression towards ARDS is likely to occur.
- Treatment of HAPE should include oxygen, rapid descent, nifedipine/PDEIs (for pulmonary HTN), and hyperbaric oxygen.
- HAPE is concomitant in approximately 50% of cases of HACE.

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