Essential Learning
Botulism

- **Case Teaching Points**
    - The differential for respiratory failure should include:
        - Infection- RSV, pertussis, sepsis, meningitis, botulism
        - Trauma- intracranial bleeding, shaken baby, c-spine trauma
        - Electrolyte imbalance- hypoglycemia, hypo/hypernatremia
        - Neuromuscular disease (usually exacerbated by an illness)- spinal muscular atrophy, myasthenia gravis, botulism, Guillain-Barre, tick paralysis
        - Metabolic disease- hyperammonemia
        - Intoxication- sedative-hypnotics (alcohol, barbiturates, opioids, benzodiazepines)
    - In the case of a child with respiratory failure but lack of distress, especially with poor tone, the differential is narrowed to:
        - Severe infection/sepsis
        - Intracranial/C-spine pathology
        - Acute neuromuscular disease (likely botulism)
        - Note that electrolyte imbalance, hyperammonemia, meningitis, or trauma leading to respiratory failure is unlikely to present without true lethargy when severe enough to cause respiratory failure.
        - Clinical evaluation should include workup with this broader differential.
    - Clues to botulism in this case include:
        - Symmetric motor deficits, most notably hypotonia
        - Bulbar symptoms: poor gag, disconjugate gaze, ptosis, poor suck/latch
        - Decreased stooling
        - Normal mental status
        - Lack of fever and otherwise normal vitals signs

- **What is the differential for bradypnea and low tone in an infant?**
    - STENTSS mnemonic
        - Sepsis/meningitis
        - Trauma (head/neck)
        - Electrolyte imbalance: hypo/hypernatremia, hypoglycemia
        - Neuromuscular disease (spinal muscular atrophy, infantile myasthenia, Guillain-Barre, botulism)
        - Toxins: opioids or other sedative-hypnotics
        - Seizure/postictal phase

        - Stroke or intracranial hemorrhage

- **What causes botulism?**
    - *Clostridium botulinum* bacteria can produce 7 subtypes of botulinum neurotoxin, known as botulinum neurotoxin (BoNT) A through G.
    - Serotypes A (40%) and B (59%) cause almost all cases of infant botulism.
    - Botulinum toxin is one of the most potent toxins known to humans.
    - No vaccine is available.
    - Botulism may occur in one of four forms:
        - Adult botulism (also known as “food-borne” botulism)- ingestion of preformed toxin leads to rapid onset and deterioration
            - Typically presents as small outbreaks involving home-canned foods and fish.
            - Outbreaks in prisons can occur from “prison brew” (bootleg alcohol)
        - Infant botulism (also known as “intestinal toxemia botulism”)- ingestion of spores from environmental dust or soil, which then germinate into mature *C. botulinum* bacteria in the GI tract and slowly elute toxin leading to a more insidious presentation than food-borne botulism.
            - Highest spore counts are in CA and PA.
            - Raw honey is a minor reservoir.
        - Wound botulism- associated with IV drug use (particularly with “black tar” heroin), skin popping, puncture wounds, abscesses, deep space infections
        - Inhalational botulism (inhalation of spores)- main concern is potential use for bioterrorism but technical complexities make successful use unlikely.

- **How does infant botulism differ from adult botulism?**
    - Infant botulism is caused by infection from the spores which release toxin in the GI tract.
        - Breastfed infants are at increased risk compared to those who are formula fed.
        - After about a year, exposure to the spore is no longer a risk as GI flora will prevent toxin release.
    - Adult botulism is caused by ingestion of preformed toxin and thus is of quicker onset.

- **How does botulism typically present?**
    - Acute onset of bilateral cranial neuropathies and symmetric descending weakness.
    - Other key features:
        - Progressive respiratory failure (low RR, dec Sat) but normal Temp, HR, BP (unless peri-arrest)
        - Symmetric neurologic deficits, spares sensory deficits (except blurred vision)
        - Normal mental status
    - Clinical symptoms evolve over a few hours to days and last for months. Progressive respiratory failure may occur rapidly.

- **How is botulism treated?**

- Supportive care and antitoxin are the mainstays of treatment.
    - Monitor closely for respiratory failure and intubate as needed (facial paralysis may obscure typical signs of respiratory failure/distress)
    - Consider intubation for vital capacity < 30% of expected.
- Confirmatory tests (toxin detection or *C. botulinum* growth in relevant specimens) take a long time to result; treatment should started based on presumptive clinical diagnosis.
- As soon as botulism is suspected, public health authorities should be contacted to assist with diagnostic testing and treatment.
    - A heptavalent (active against serotypes A through G) antitoxin for food-borne botulism is available from the CDC.
    - For infant botulism, the bivalent (active against serotypes A and B only) antitoxin known as BabyBIG® (human-derived IV botulism immune globulin) is available from the California Department of Public Health (cost around $80,000).
- Antitoxin neutralizes only unbound, circulating toxin so will only prevent further deterioration and does not reverse paralysis that has already occurred.
- The longer the delay in delivering antitoxin, the poorer the prognosis and the longer the need for ventilatory support.
- Antibiotics should be avoided (except for some cases of wound botulism) unless they are necessary to treat comorbid conditions. Theoretically, lysis of *C. botulinum* with antibiotic therapy could release additional toxin.
- Consider the source of exposure and need to monitor family/close contacts for evolving symptoms.

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