Hemorrhagic Shock and Encephalopathy Syndrome (HSES)

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**Etiologic Theories**
- Overwrapping of infants who have febrile illness.
- Reaction to intestinal or environmental toxins, pancreatic release of trypsin, or unidentified virus or bacterium.

**Clinical Features**
- Occurs predominantly in 3-8 mo infants (but was reported in 15-yr-old).
- Encephalopathy (seizures, coma, and hypotonia) with focal hemorrhages and infarcts, diffuse cerebral edema, herniation.
- Hyperpyrexia (up to 43.9° C rectally)
- DIC
- Hepatic dysfunction
- Renal dysfunction
- Rhabdomyolysis, myoglobinuria
- Lungs and myocardium are not primarily involved!

**Diagnosis**
- By exclusion.
- ABG (metabolic acidosis), CBC (thrombocytopenia, falling Hct, leukocytosis), hyperkalemia, BUN↑, creatinine↑, PT/PTT, liver function tests (liver transaminases↑), hypoglycemia.
- Blood and urine bacteriologic and viral cultures are negative.

**Differential**
- Sepsis
- Reye's syndrome
- Hemolytic-uremic syndrome (HUS).

**Treatment**
- Entirely supportive:
  1. Large volumes of isotonic solutions and blood products (fresh frozen plasma, albumin, whole blood, packed RBCs)
  2. Inotropic support (dopamine, epinephrine)
  3. Hyperpyrexia requires external cooling.
  4. ICP↑ → hyperventilation, etc.

**Prognosis**
- > 60% patients die, ≥ 70% survivors have severe neurologic sequelae.

**Bibliography** for ch. “Metabolic Disorders” → follow this LINK >>

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