Hemorrhagic Shock and Encephalopathy Syndrome (HSES)

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HSES - extremely rare acute disease in previously healthy children, resulting in death or catastrophic neurologic outcome.

- occurs predominantly in 3-8 mo infants (but was reported in 15-yr-old).

ETIOLOGIC THEORIES

a) *overwrapping of* infants who have febrile illness.
b) reaction to intestinal or environmental toxins, pancreatic release of trypsin, or unidentified virus or bacterium.

CLINICAL FEATURES

- resembles heatstroke, with extremely high temperature and multiple organ dysfunction.
- prodrome - fever, upper respiratory tract symptoms, or vomiting and bloody / watery diarrhea.

1. Severe shock
2. Encephalopathy (seizures, coma, and hypotonia) with focal hemorrhages and infarcts, diffuse cerebral edema, herniation.
3. Hyperpyrexia (up to 43.9° C rectally)
4. DIC
5. Hepatic dysfunction
6. Renal dysfunction
7. 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

1) sepsis
2) Reye's syndrome
3) 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 >>