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

* 1. *overwrapping of* infants who have febrile illness.
	2. 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 >>](http://www.neurosurgeryresident.net/Mus.%20Muscular%2C%20Neuromuscular%20disorders%5CMus.%20Bibliography.pdf)

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