

# Hemorrhagic Shock and Encephalopathy Syndrome (HSES)

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ETIOLOGIC THEORIES .....	1
CLINICAL FEATURES .....	1
DIAGNOSIS .....	1
DIFFERENTIAL .....	1
TREATMENT .....	1
PROGNOSIS.....	1

**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

- overwrapping of infants who have febrile illness.
- 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.

- Severe shock
- 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:

- large volumes of isotonic solutions and blood products (fresh frozen plasma, albumin, whole blood, packed RBCs)
- inotropic support (dopamine, epinephrine)
- hyperpyrexia requires external cooling.
- ICP↑ → hyperventilation, etc.

## PROGNOSIS

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

BIBLIOGRAPHY for ch. "Metabolic Disorders" → follow this [LINK >>](#)