



Merlin disease code: 42000 Hemolytic Uremic Syndrome (HUS)

No paper case report form
No Merlin extended data

Background

Hemolytic uremic syndrome (HUS) is characterized by the acute onset of microangiopathic hemolytic anemia, renal injury, and low platelet count. Thrombotic thrombocytopenic purpura (TTP) is also characterized by these features but can include central nervous system (CNS) involvement and fever and may have a more gradual onset. Most cases of HUS (but few cases of TTP) occur after an acute gastrointestinal illness (usually diarrheal).

Clinical criteria for case classification

Confirmatory:

An acute illness diagnosed as HUS or TTP within three weeks after onset of an episode of acute or bloody diarrhea.

Presumptive:

An acute illness diagnosed as HUS or TTP.

Laboratory criteria for case classification

Confirmatory:

All of the following:

- Anemia (acute onset),
- **And** microangiopathic changes (i.e., presence of schistocytes, keratocytes, helmet cells, echinocytes, or burr cells) on peripheral blood smear,
- **And** renal injury (acute onset) evidenced by either hematuria, proteinuria, or elevated creatinine level (i.e., ≥ 1.0 mg/dL in a child aged <13 years or ≥ 1.5 mg/dL in a person aged ≥ 13 years, or $\geq 50\%$ increase over baseline).

Presumptive:

Both of the following:

- Anemia (acute onset)
- **And** renal injury (acute onset) evidenced by either hematuria, proteinuria, or elevated creatinine level (i.e., ≥ 1.0 mg/dL in a child aged <13 years or ≥ 1.5 mg/dL in a person aged ≥ 13 years, or $\geq 50\%$ increase over baseline).

Epidemiological criteria for case classification

Not applicable.

Case classification

Confirmed:

A person with confirmatory clinical criteria and confirmatory laboratory criteria.

Probable:

Either of the following:

- A person with presumptive clinical criteria and confirmatory laboratory criteria
- **Or** a person with confirmatory clinical criteria and presumptive laboratory criteria.

Criteria to distinguish a new case from previous reports

Not applicable.

Comments

A low platelet count can usually, but not always, be detected early in the illness, but it may then become normal or even high. If a platelet count obtained within 7 days after onset of the acute gastrointestinal illness is not $<150,000/\text{mm}^3$, other diagnoses should be considered.

Some investigators consider HUS and TTP to be part of a continuum of disease. Therefore, criteria for diagnosing TTP on the basis of CNS involvement and fever are not provided because cases diagnosed clinically as postdiarrheal TTP also should meet the criteria for HUS. These cases are reported as postdiarrheal HUS.

Most diarrhea-associated HUS is caused by Shiga toxin-producing *Escherichia coli* (STEC), most commonly *E. coli* O157.

If a person meets the case definition for both Shiga toxin-producing *E. coli* (STEC) (Merlin code=00800) and HUS (Merlin code=4200), a case should be created and reported for each condition in Merlin.