

A 6-year-old girl is brought to the physician due to pallor, decreased energy, bloody diarrhea, and "spots" over her arms and legs that developed over the past 2 days. Her immunizations are up to date. The patient's past medical history is unremarkable and she has been generally healthy. The girl has no fever, vomiting, or joint pain. She has had no travel or animal exposures. The patient's temperature is 37.1 C (98.8 F). On examination, she appears lethargic and weak; her face and extremities are swollen and she has poor skin turgor. The patient has scattered petechiae and a few purpura on her arms. She also has moderate, non-localized abdominal tenderness. Laboratory studies are as follows:

Hemoglobin	6.4 g/dL
Platelets	45,000/ μ L
Creatinine	2.3 mg/dL
Total bilirubin	3 mg/dL
Direct bilirubin	0.2 mg/dL

Urinalysis

Specific gravity	1.025
pH	5
Protein	+2
Blood	moderate
Leukocyte esterase	negative
Nitrites	negative
Bacteria	none
White blood cells	50+/hpf
Red blood cells	20–30/hpf
Casts	hyaline casts

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Which of the following is the most likely diagnosis?

- ☐ A. Hemolytic-uremic syndrome
- ☐ B. Henoch-Schönlein purpura
- ☐ C. Immune thrombocytopenic purpura
- ☐ D. Post-streptococcal glomerulonephritis
- ☐ E. Rocky Mountain spotted fever

Submit

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Casts hyaline casts

Which of the following is the most likely diagnosis?

- ☒ A. Hemolytic-uremic syndrome [71%]
- ☐ B. Henoch-Schönlein purpura [21%]
- ☐ C. Immune thrombocytopenic purpura [7%]
- ☐ D. Post-streptococcal glomerulonephritis [1%]
- ☐ E. Rocky Mountain spotted fever [0%]

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Explanation:

User Id: [REDACTED]

Hemolytic-uremic syndrome	
Etiology	Enterocolitis from Shiga toxin-producing bacteria (<i>E coli</i> O157:H7, <i>Shigella</i>) most common
Presentation	<ul style="list-style-type: none"> • Diarrhea (often bloody) • Lethargy, irritability, pallor • Bruising or petechiae • Oliguria, edema
Laboratory Findings	<ul style="list-style-type: none"> • Hemolytic anemia • Thrombocytopenia • ↑ Creatinine, hematuria, proteinuria, casts (due to glomerular hemolysis) • ↑ Bilirubin (due to hemolysis)
Treatment	<ul style="list-style-type: none"> • Fluid & electrolyte management • Blood transfusions • Dialysis

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Hemolytic-uremic syndrome (HUS) is a clinical syndrome consisting of **hemolytic anemia, thrombocytopenia, and acute renal failure**. It is most commonly seen in toddlers. More than 90% of HUS occurs due to shiga-toxin producing **diarrheogenic** pathogens such as *Escherichia coli* O157:H7 or *Shigella*. Approximately 10% of HUS cases are due to infection with *Streptococcus pneumoniae*; these patients have pneumonia or meningitis instead of diarrhea. In both settings, the toxins enter the systemic circulation and injure the endothelial cells in the kidney.

Children with HUS generally have fatigue/lethargy and pallor due to the hemolysis. In addition, the hemolysis often leads to some degree of hyperbilirubinemia. If the central nervous system is affected, irritability or headache may be present. Progressive renal involvement leads to decreased urine output and gross or microscopic hematuria. As the kidney function worsens, fluid overload develops (eg, pulmonary edema, congestive heart failure).

Once HUS is diagnosed, the management is supportive. Fluid and electrolyte balance must be maintained, and anemia and thrombocytopenia should be managed with conservative transfusions. Approximately half of all children with HUS will require **dialysis** for their acute renal failure. With supportive care, the mortality of HUS is <5%.

(Choice B) Henoch-Schönlein purpura presents with purpura on the legs and buttocks in the setting of a normal platelet count. In this scenario, the thrombocytopenia, lack of joint pain, distribution of the purpura in the upper extremities, and the history of preceding diarrhea are more suggestive of HUS.

(Choice C) Immune thrombocytopenic purpura can present with thrombocytopenia and purpura. However, anemia and renal failure are not seen in immune thrombocytopenic purpura.

(Choice D) Post-streptococcal glomerulonephritis (PSGN) can present with hematuria and signs of volume overload after infection with group A *Streptococcus*. If present, thrombocytopenia and anemia are generally mild, and renal failure is unusual in PSGN.

(Choice E) Rocky Mountain spotted fever (RMSF) can cause a vasculitis, including petechiae, anemia, thrombocytopenia, and hematuria. Children with RMSF are almost invariably febrile. In addition, this patient does not have an exposure history that would place her at risk for RMSF or other tick-borne infections.

Educational objective:

Hemolytic-uremic syndrome is a clinical syndrome of hemolytic anemia, thrombocytopenia, and acute renal failure. It is most often caused by Shiga-toxin-mediated endothelial cell damage, after infection with *Escherichia coli*

involvement leads to decreased urine output and gross or microscopic hematuria. As the kidney function worsens, fluid overload develops (eg, pulmonary edema, congestive heart failure).

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Educational objective:

Hemolytic-uremic syndrome is a clinical syndrome of hemolytic anemia, thrombocytopenia, and acute renal failure. It is most often caused by Shiga-toxin-mediated endothelial cell damage, after infection with *Escherichia coli* O157:H7 or *Shigella*. Treatment is supportive; approximately 50% of children with hemolytic-uremic syndrome require dialysis.

References:

1. [HUS and TTP in children](#)
2. [Update on *Streptococcus pneumoniae* associated hemolytic uremic syndrome](#)
3. [Shiga toxins and the pathophysiology of hemolytic uremic syndrome in humans and animals.](#)