

A 6-year-old boy is brought to the clinic by his father for fatigue and pallor. The patient was seen in another clinic a week ago for abdominal pain and diarrhea, and according to his father, "some medication" was prescribed to help slow the diarrhea. The diarrhea has improved, but the patient's activity level is still far below baseline. Temperature is 36.7 C (98 F), blood pressure is 100/60 mm Hg, pulse is 104/min, and respirations are 20/min. Physical examination shows a pale, uncomfortable-appearing child with multiple petechiae. Chest and cardiac examinations disclose no abnormalities. The abdomen is soft and nontender, and the liver and spleen are not palpable. Laboratory values are as follows:

Complete blood count

Hemoglobin	7.8 g/dL
Mean corpuscular volume	84 fL
Platelets	50,000/mm ³
Leukocytes	13,000/mm ³

Serum chemistry

Sodium	136 mEq/L
Potassium	5 mEq/L
Blood urea nitrogen	38 mg/dL
Creatinine	2.5 mg/dL

Liver function studies

Total bilirubin	3.3 mg/dL
Direct bilirubin	0.4 mg/dL

Coagulation studies

Prothrombin time	12 sec
Activated partial thromboplastin time	35 sec

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A peripheral blood smear reveals marked red cell fragmentation and helmet cells. What is the most likely underlying pathophysiology for this patient's pallor?

- ☐ A. Antibody-mediated destruction of red cells
- ☐ B. Decreased erythropoietin production
- ☐ C. Decreased globin chain synthesis
- ☐ D. Defective globin chain synthesis
- ☐ E. Microangiopathic hemolytic anemia
- ☐ F. Systemic activation of coagulation system

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- ☐ A. Antibody-mediated destruction of red cells [10%]
- ☐ B. Decreased erythropoietin production [1%]
- ☐ C. Decreased globin chain synthesis [1%]
- ☐ D. Defective globin chain synthesis [2%]
- ☒ E. Microangiopathic hemolytic anemia [82%]
- ☐ F. Systemic activation of coagulation system [4%]

[Proceed to Next Item](#)

Explanation:

User Id: [REDACTED]

Hemolytic uremic syndrome	
Pathogenesis	<ul style="list-style-type: none">Initial insult from Shiga toxin (<i>Escherichia coli</i> serotype O157:H7)Vascular damage & microthrombi formation
Clinical features	<ul style="list-style-type: none">Preceding bloody diarrheaFatigue, pallorBruising, petechiaeOliguria, edema
Laboratory findings	<ul style="list-style-type: none">Hemolytic anemia (schistocytes, ↑ bilirubin)ThrombocytopeniaAcute kidney injury (↑ BUN, ↑ Cr)
Treatment	<ul style="list-style-type: none">Fluid & electrolyte managementBlood transfusionsDialysis

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Treatment	<ul style="list-style-type: none">• Fluid & electrolyte management• Blood transfusions• Dialysis

BUN = blood urea nitrogen; Cr = creatinine.

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This patient has normocytic anemia, **thrombocytopenia**, and **acute renal injury** following a diarrheal illness, a triad that suggests **hemolytic uremic syndrome** (HUS). HUS most commonly occurs after acute diarrheal illness caused by Shiga toxin-producing *Escherichia coli* serotype O157:H7. A prodrome of abdominal pain, vomiting, and **bloody diarrhea** occurs in the majority of patients 5-10 days prior to the onset of HUS.

The Shiga toxin induces endothelial damage in renal glomeruli, leading to platelet activation, microthrombi formation, and **microangiopathic hemolytic anemia**. The mechanical destruction of red blood cells results in **schistocytes** (ie, fragmented red blood cells) on peripheral smear. HUS laboratory findings include thrombocytopenia as well as evidence of hemolytic anemia (low hemoglobin, normal mean corpuscular volume, elevated indirect bilirubin, elevated reticulocyte count) and renal injury (elevated blood urea nitrogen and creatinine).

Diagnosis is based on clinical and laboratory findings. Treatment is supportive and includes fluid and electrolyte management, blood transfusions, and dialysis. Antibiotics

Treatment

- Blood transfusions
- Dialysis

BUN = blood urea nitrogen; Cr = creatinine.

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Diagnosis is based on clinical and laboratory findings. Treatment is supportive and includes fluid and electrolyte management, blood transfusions, and dialysis. Antibiotics are not effective in preventing or treating HUS.

(Choice A) Autoimmune hemolytic anemia results from antibody-mediated destruction of red blood cells and usually presents after a respiratory illness or in the setting of systemic disease (eg, systemic lupus erythematosus). The platelet count is typically normal, and **spherocytes** are seen on peripheral smear.

(Choice B) Erythropoietin, a hormone produced by the kidney and involved in red blood cell regulation, is decreased in patients with chronic kidney disease. This patient's illness is acute, with an elevated bilirubin indicative of hemolytic anemia. **Burr cells (echinocytes)** would be seen on peripheral smear.

(Choice C) An imbalance between α - and β -globin chain production results in thalassemia with clinical features ranging from asymptomatic microcytic anemia to transfusion-dependent anemia. The peripheral smear reveals **target cells** and hypochromic microcytes.

(Choice D) Sickle cell anemia is a hemolytic anemia due to a missense mutation in the β -globin gene. Most patients have painful vaso-occlusive crises in early childhood, and peripheral smear demonstrates **"sickled" red blood cells**.

(Choice F) Disseminated intravascular coagulation results from systemic activation of

well as evidence of hemolytic anemia (low hemoglobin, normal mean corpuscular volume, elevated indirect bilirubin, elevated reticulocyte count) and renal injury (elevated blood urea nitrogen and creatinine).

Diagnosis is based on clinical and laboratory findings. Treatment is supportive and includes fluid and electrolyte management, blood transfusions, and dialysis. Antibiotics are not effective in preventing or treating HUS.

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(Choice D) Sickle cell anemia is a hemolytic anemia due to a missense mutation in the β -globin gene. Most patients have painful vaso-occlusive crises in early childhood, and peripheral smear demonstrates **"sickled" red blood cells**.

(Choice F) Disseminated intravascular coagulation results from systemic activation of the coagulation system from an underlying disorder (eg, sepsis, trauma). It presents with thrombocytopenia and anemia (due to bleeding) but also with prolonged prothrombin and activated partial thromboplastin times.

Educational objective:

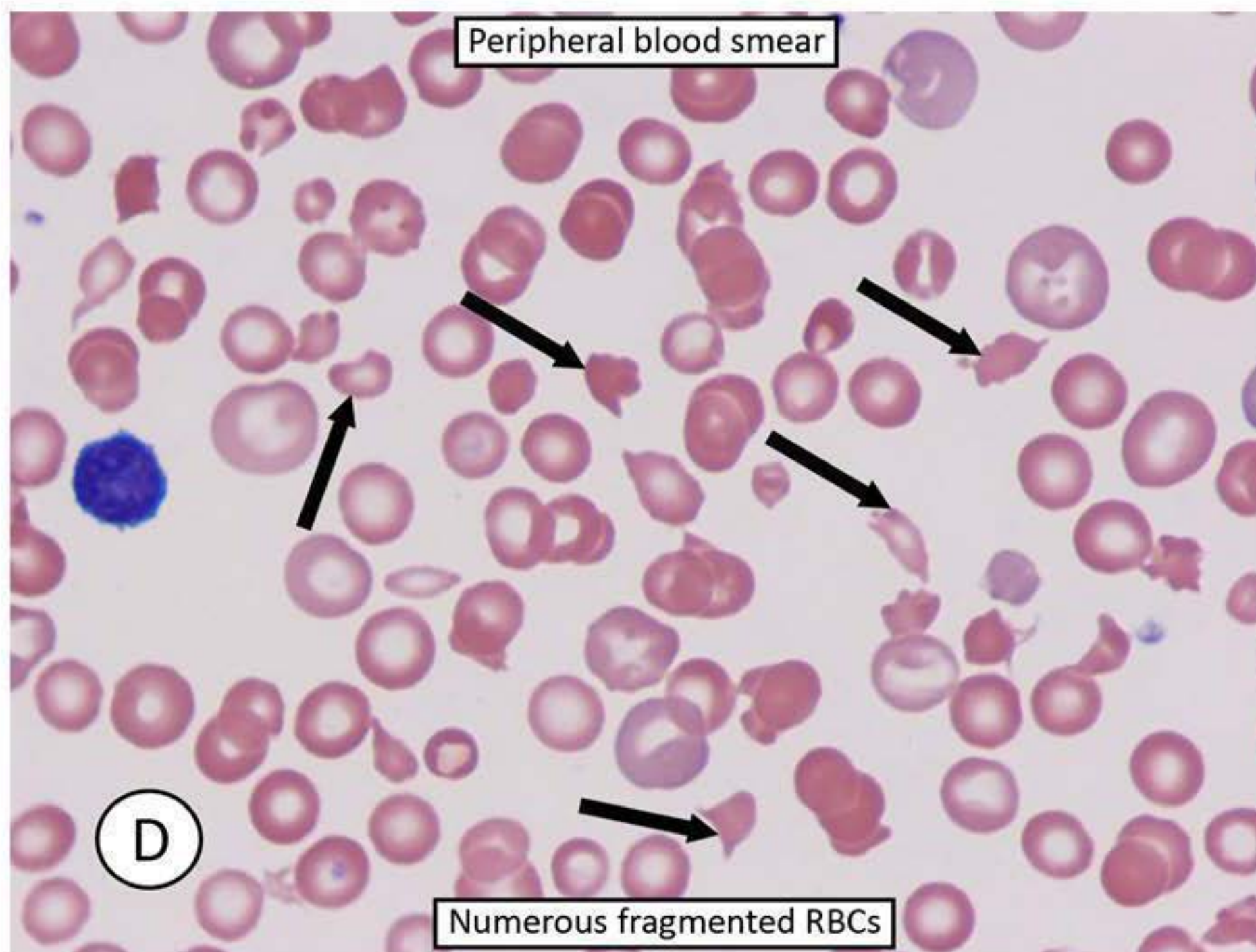
Hemolytic uremic syndrome typically occurs in a child who has recently recovered from a diarrheal illness and has acute renal injury, thrombocytopenia, and microangiopathic hemolytic anemia with schistocytes on peripheral smear.

References:

1. **Hemolytic uremic syndrome.**
2. **Hemolytic uremic syndrome.**

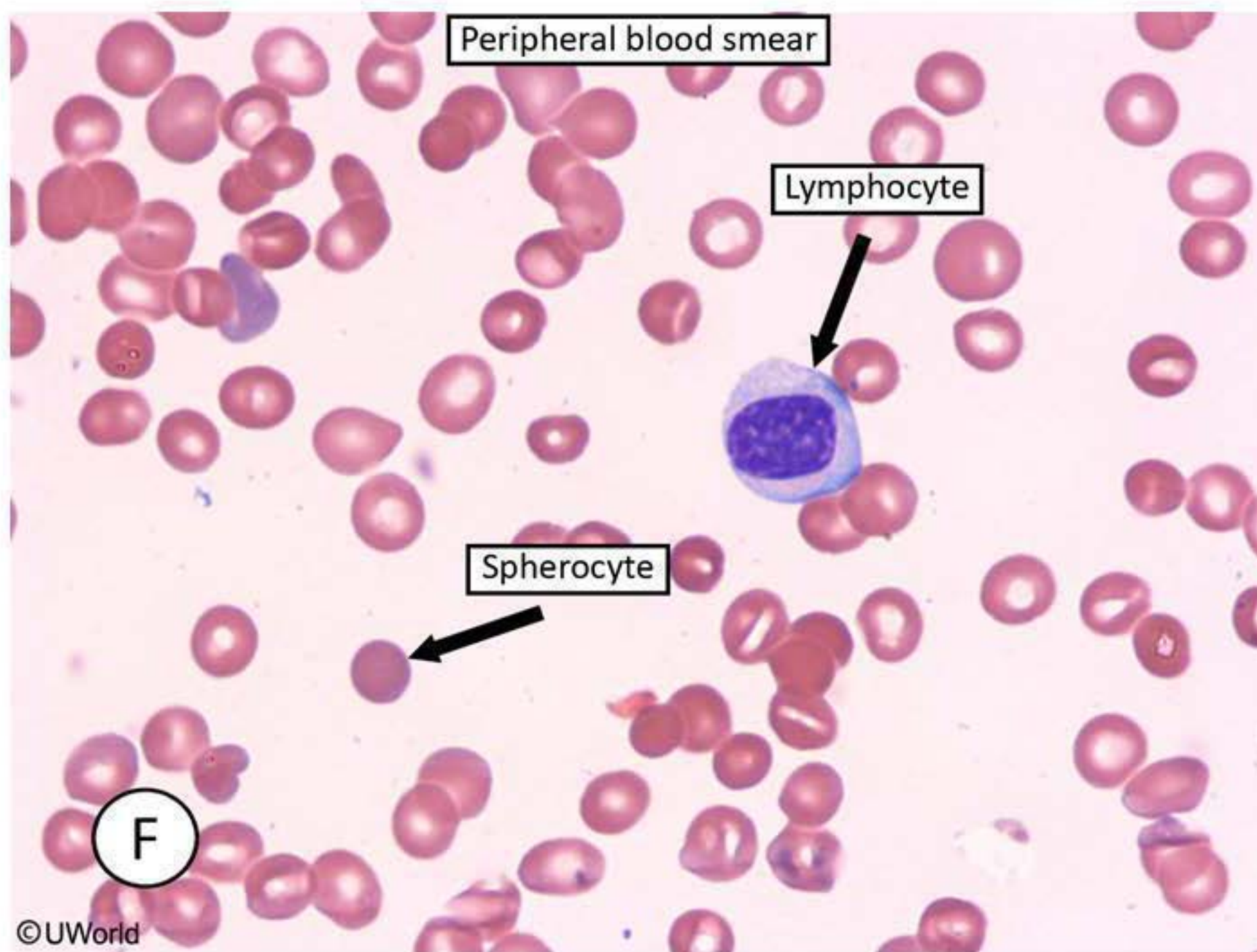
Media Exhibit

angiopathic hemolytic anemia: MAHA



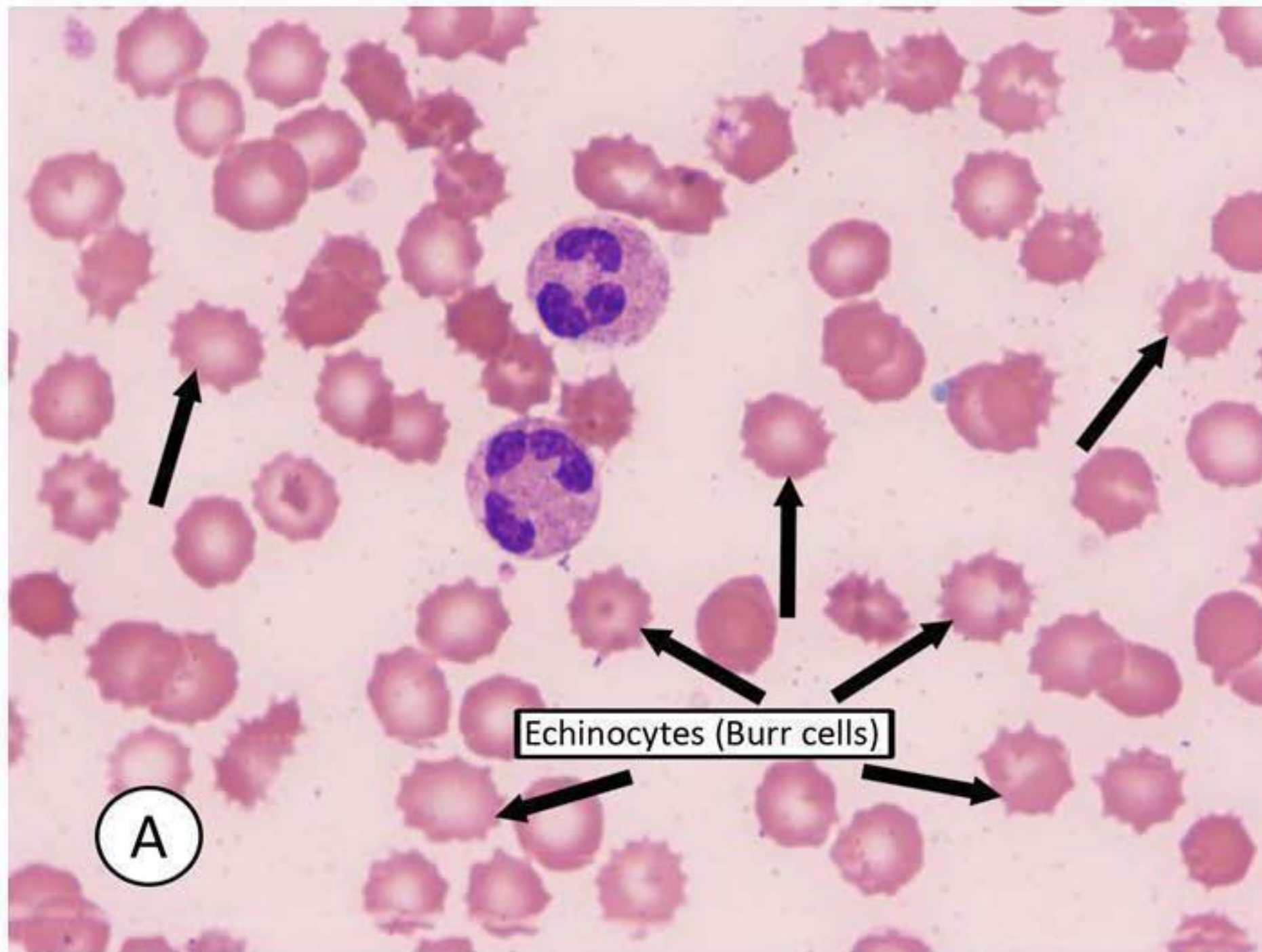
Media Exhibit

immune hemolytic anemia



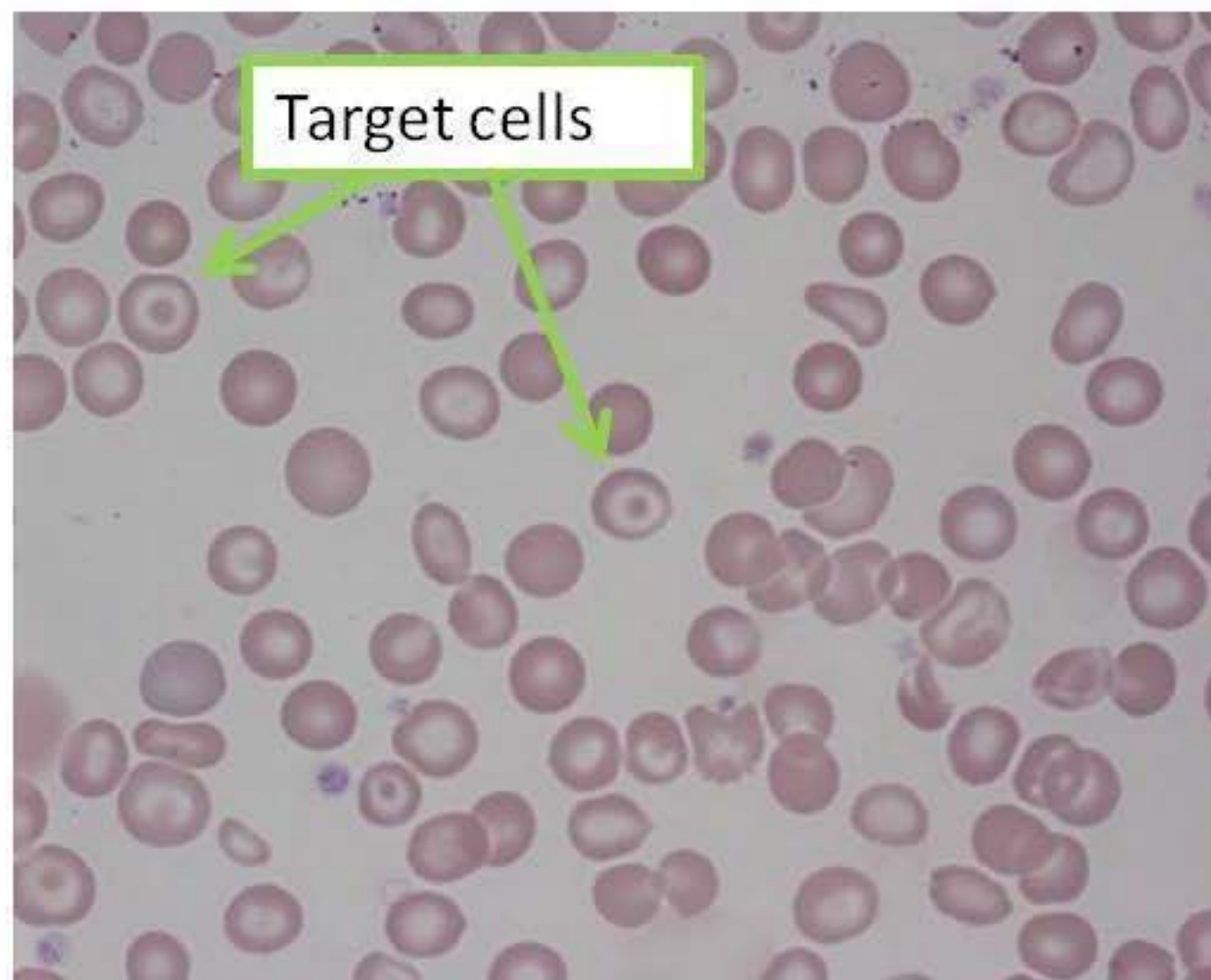
Media Exhibit

ytes: Burr cells



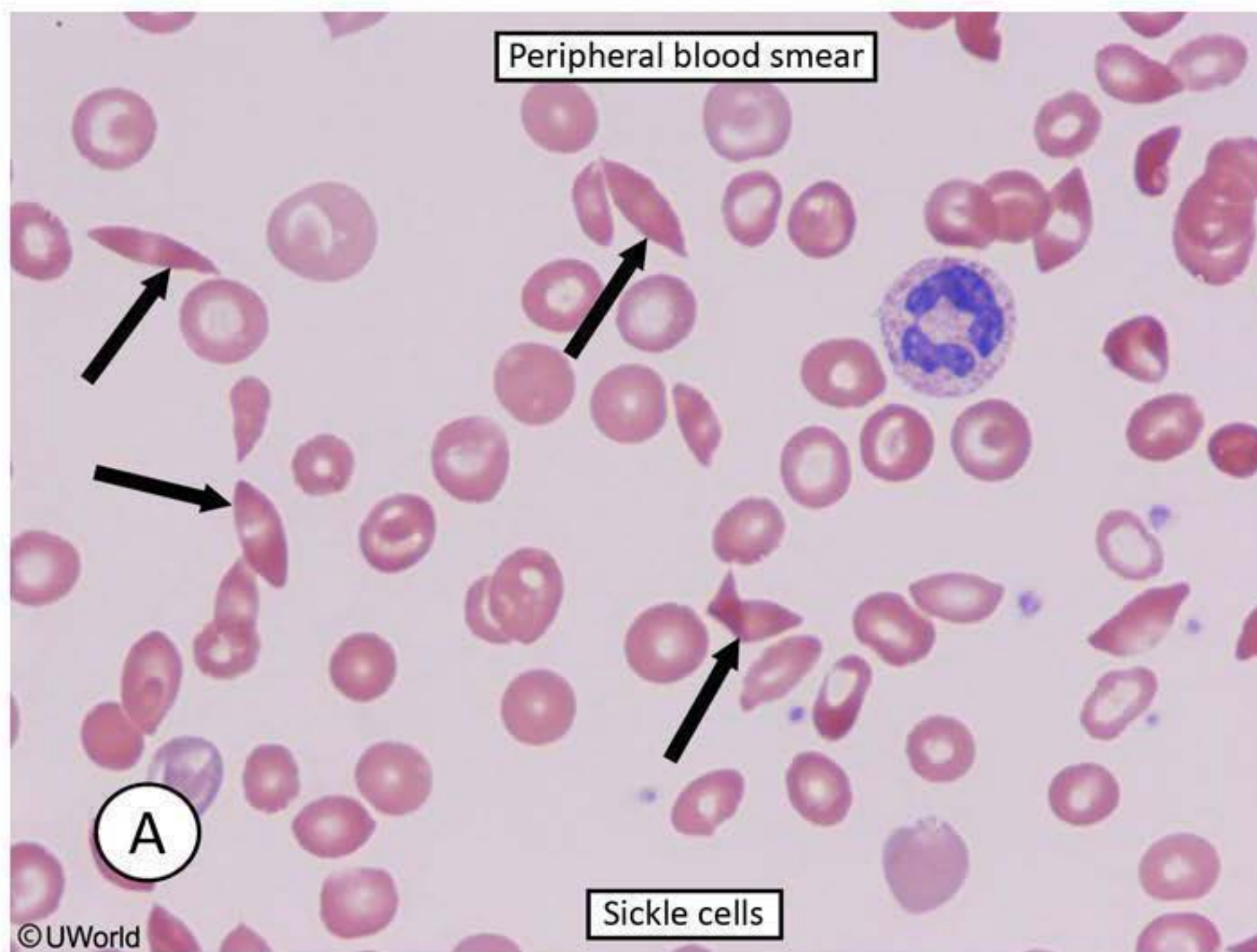
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