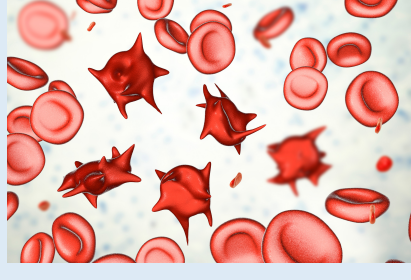


SPUR CELL ANEMIA

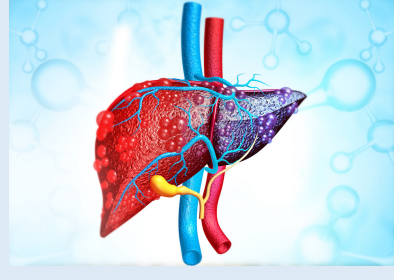
TERM DEFINITION

Acquired Coombs-negative nonimmune hemolytic anemia characterized by increased numbers of acanthocytes (spur cells) in patients with end stage (typically alcoholic) liver disease.

Acanthocytes are red cells with irregular surface and spike-like projections that vary in width, length and distribution.



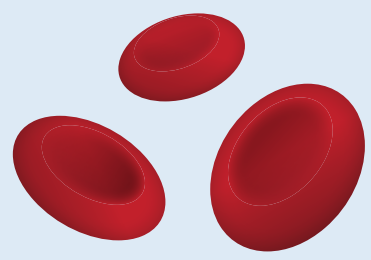
Seen exclusively in **end stage liver disease**, particularly in the setting of alcoholic cirrhosis.



There is no universal acceptance of the number of acanthocytes required to diagnose spur cell anemia, but most studies call for **at least 20%** of the red cells seen on the peripheral smear.



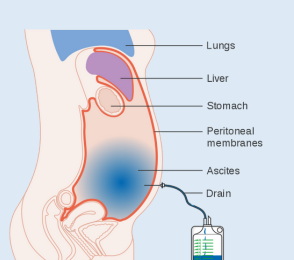
CLINICAL PEARLS



Spur cells or acanthocytes also found in post-splenectomy state, McCleod syndrome and abetalipoproteinemia.



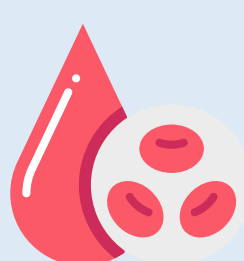
Spur cell anemia occurs primarily in alcoholic liver disease but has been reported in many types of chronic liver disease.



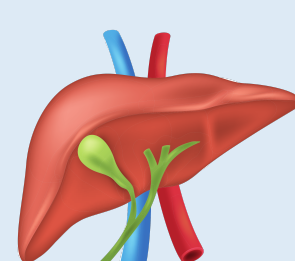
Almost all patients have scleral icterus, ascites and splenomegaly.



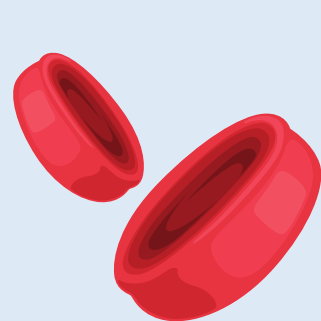
Spur cell anemia carries a poor prognosis; median survival of 1.9 months (marker of severe liver disease).



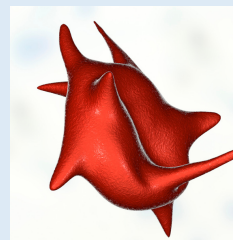
Spur cell anemia is a type of extracorporeal hemolytic anemia; the defect is extrinsic to the red cell.



Spur cell anemia is cured by liver transplantation.



Normal plasma lacks spur cell-forming factors from **Cirrhotic plasma**



PRESENTATION

SYMPTOMS

ANEMIA:

- Fatigue
- Shortness of breath
- Headache
- Palpitations

LIVER DISEASE:

- Jaundice
- Abdominal fullness
- Confusion
- Loss of appetite
- Itchy skin

SIGNS

ANEMIA:

- Pallor
- Tachycardia

LIVER DISEASE:

- Ascites
- Spider nevi
- Palmar erythema
- Scleral icterus

LABS

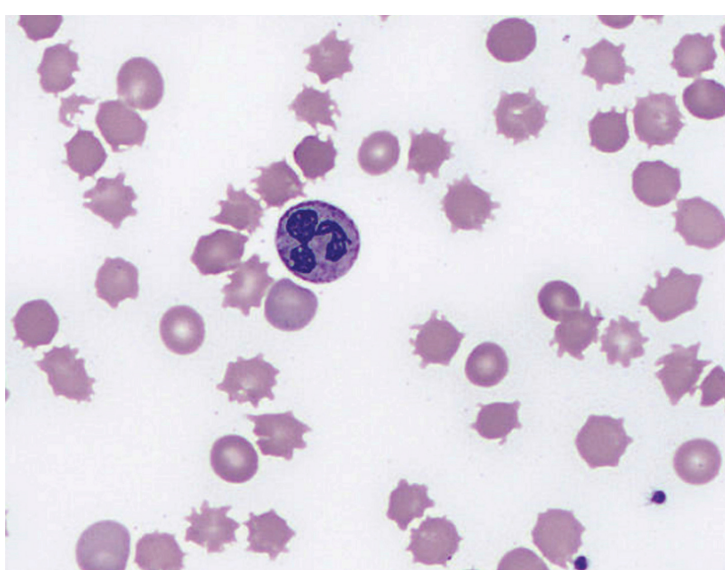
HEMATOLOGY LABS:

- Anemia, often macrocytic
- Thrombocytopenia from liver disease
- Leukopenia from liver disease
- Elevated reticulocyte count
- > 20% acanthocytes on peripheral smear (right)

HEMOLYTIC LABS:

- High LDH
- High AST
- Low haptoglobin

LABS ASSOCIATED WITH CIRRHOSIS



DIAGNOSIS

Hemolytic anemia + >20% acanthocytes on peripheral smear

Often transfusion dependent

DIFFERENTIAL DIAGNOSIS

There are many causes of anemia in chronic liver disease including blood loss and iron deficiency. Conversely, acanthocytes are not restricted to liver disease.

TREATMENT PRINCIPLES

- Red cell transfusion
- Folic acid
- Abstinence from alcohol
- Transplantation (curative)

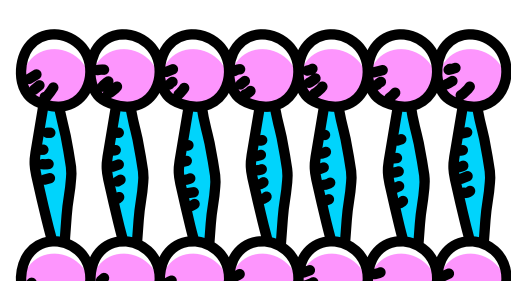
Transient alcohol-related hemolytic anemia was first described by Dr. Leslie Zieve in 1958* that manifests as the triad of:

- Jaundice, from hemolysis & cholestasis due to alcohol-induced liver injury
- Hemolytic anemia, typically with spur cells
- Transient hyperlipidemia (serum cholesterol concentration can exceed 1000 mg/dl, normalizes 1-2 weeks after insult).

It is considered by some to be a subtype of spur cell anemia; by others a mimic. Recovery can be expected within 4 to 6 weeks after alcohol withdrawal.

*Ann Intern Med 1958;48:471-96

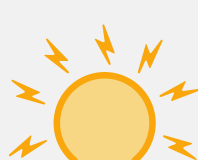
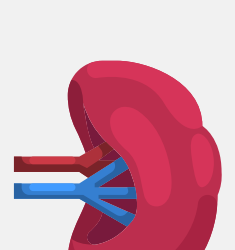
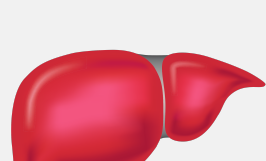
ZIEVE'S SYNDROME



PROXIMATE MECHANISMS

Spur cell formation has been described as a two phase process:

- 1 The first involves liver-dependent incorporation of excess cholesterol in the red cell membrane.
- 2 The second stage involves splenic conditioning of the cholesterol-rich red cells resulting in loss of red cell surface area and transformation to a spiculated cell contour.



The spur cells demonstrate increased rigidity and decreased deformability, resulting in their entrapment and destruction in the spleen

EVOLUTIONARY MECHANISMS



Spur cell anemia is an example of an **evolutionary mismatch**, whereby disease risks can be altered for organisms living in environments that differ from those in which their ancestors evolved. Our ancestors who lived 10,000 years ago did not consume alcohol. Their genes (and our genes) were not selected for the ability to attenuate alcohol-induced liver damage and any associated spur cell anemia. This of course explains why spur cell anemia is not found outside human populations.



It is no clear whether cholesterol loading of red cell membranes in spur cell anemia is simply an exaggeration of a normal process, or whether it involves a disease-specific pathway.

DID YOU KNOW?



HISTORY OF MEDICINE

Spur cell anemia was first described in 1964. The following excerpt constitutes the entire Introduction. The patient reported had alcoholic liver disease.

THE syndrome in the case presented below has been designated "spur-cell anemia" because of the curious projections on the surfaces of the red blood cells. These bizarre erythrocytes were associated with intense hemolytic anemia in a young man with hepatic cirrhosis.

N Engl J Med 1964;271:396

NOTES

ATTRIBUTIONS

Aurhor Dr. William Aird
Graphic design Janie Vu



The Blood Project
ENCYCLOPEDIA OF BLOOD