ACANTHOCYTE

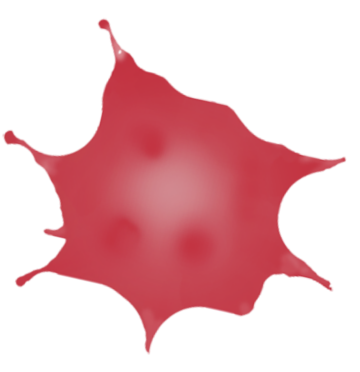
TERM DEFINITION

An acanthocyte is an irregularly shaped erythrocyte with multiple spiculated projections that are typically distributed unevenly over the cell surface.

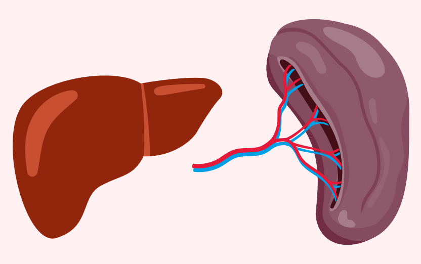
They are associated with a variety of inherited and acquired disorders.

DESCRIPTION

- Spheroidal in shape.
- Smaller than normal red cell.
- Lacks central pallor.
- Has 3 to 20 spikes.
- Spikes are irregularly distributed over the surface.
- Most spikes are sharp-tipped, though may be knobby.

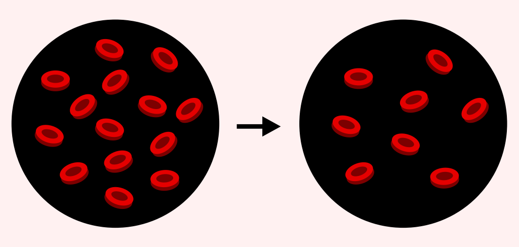


CLINICAL PEARLS



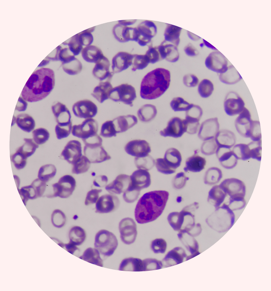
Small numbers of acanthocytes are seen **post-splenectomy**; larger numbers in **liver disease** and abetalipoproteinemia.

Acanthocytes and echinocytes may form part of a morphologic **spectrum**, and transitional forms between the two can occur.

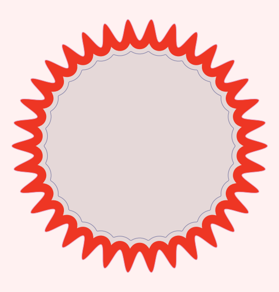


Acanthocytosis is often accompanied by **severe anemia** in patients with liver disease, but not in those with neuroacanthocytosis.

Mean corpuscular hemoglobin concentration (**MCHC**) may be elevated in patients with high numbers of acanthocytes.



True acanthocytes are not seen on normal peripheral smears, though they may be confused with **crenated forms** (i.e., burr like cells), of which there may be up to 3%.



Acanthocytes are also called **spur cells**, which is a term often reserved for patients with liver disease.

DIFFERENTIAL DIAGNOSIS

ACANTHOCYTOSIS

Neuro-acanthocytosis

Liver disease

Post-splenectomy

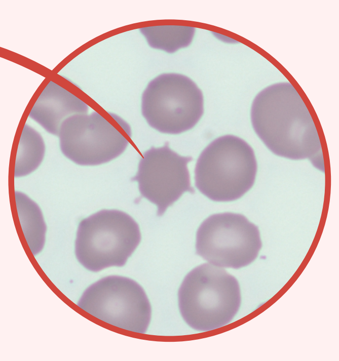
> 10%-20% ACANTHOCYTES	< 10% ACANTHOCYTES
Neuroacanthocytosis (NA) syndromes* ◦ Chorea-Acanthocytosis ◦ McLeod syndrome ◦ Huntington disease-like 2 (HDL 2) ◦ Pantothenate kinase-associated neurodegeneration (PKAN) ◦ Abeta- & hypobeta-lipoproteinemia ◦ Aceruloplasminemia	**Postsplenectomy state**
Abetalipoproteinemia	**Myeloproliferative disorders**
Advanced liver disease	**Microangiopathic hemolytic anemia**

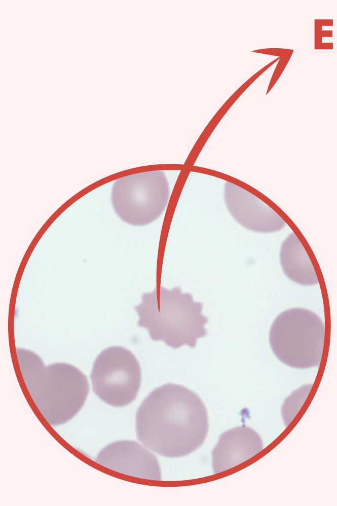
** NA syndromes are exceedingly rare hereditary neurodegenerative diseases defined by neurological abnormalities in combination with misshaped acanthocytic red blood cells.*

ACANTHOCYTE VS. ECHINOCYTE

ACANTHOCYTE

- Smaller than normal red cells
- Loss of central pallor
- Projections (spicules):
  - Fewer
  - Unevenly distributed around red cells
  - Variable thickness & width
  - Often with knobby ends





ECHINOCYTE (BURR CELL)

- Same size as normal red cells
- Typically with central pallor
- Projections (spicules):
  - More numerous
  - Evenly distributed around red cells
  - Equal thickness and width
  - Blunter tips

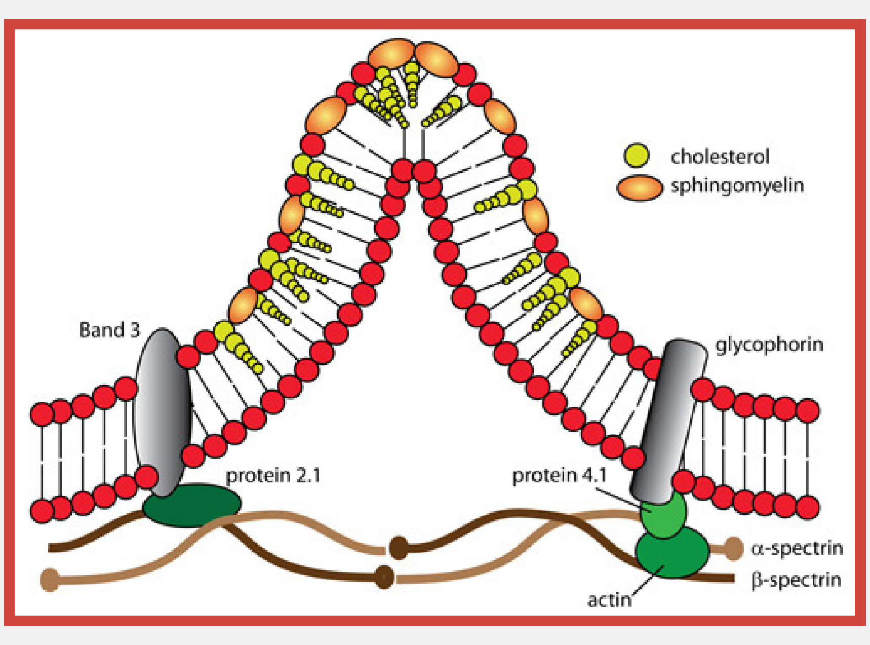
PROXIMATE MECHANISMS

Abetalipoproteinemia:
- Blood contains 50%-90% acanthocytes.
- Arise from marked increase in red cell ***membrane sphingomyelin*** and a decrease in lecithin.
- Sphingomyelin, which is more rigid than lecithin, accumulates on outer half of the lipid bilayer, resulting in selective expansion and formation of irregular projections.
- Red cells have normal lifespan.

In **McLeod syndrome**, acanthocytosis is caused by the lack of a structural protein.

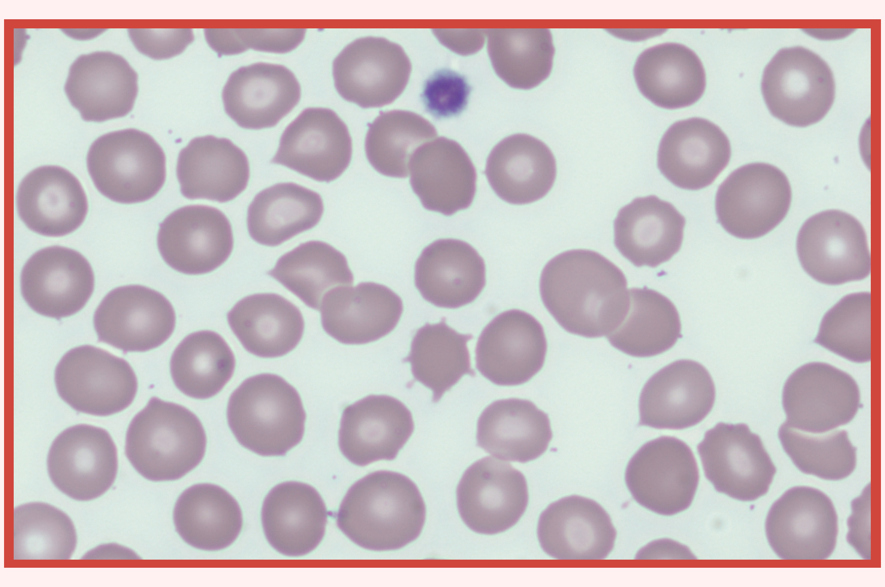
Acanthocytes are readily found in **postsplenectomy** states because of diminished removal of such poikilocytes.

Liver disease (usually alcoholic):
- Associated with increased ***membrane cholesterol*** and increased cholesterol to phospholipid ratio.
- Increased cholesterol leads to increased surface area, manifesting first as target cells then as acanthocytes.
- Poorly deformable and readily destroyed in the spleen, resulting in a shorter life span.

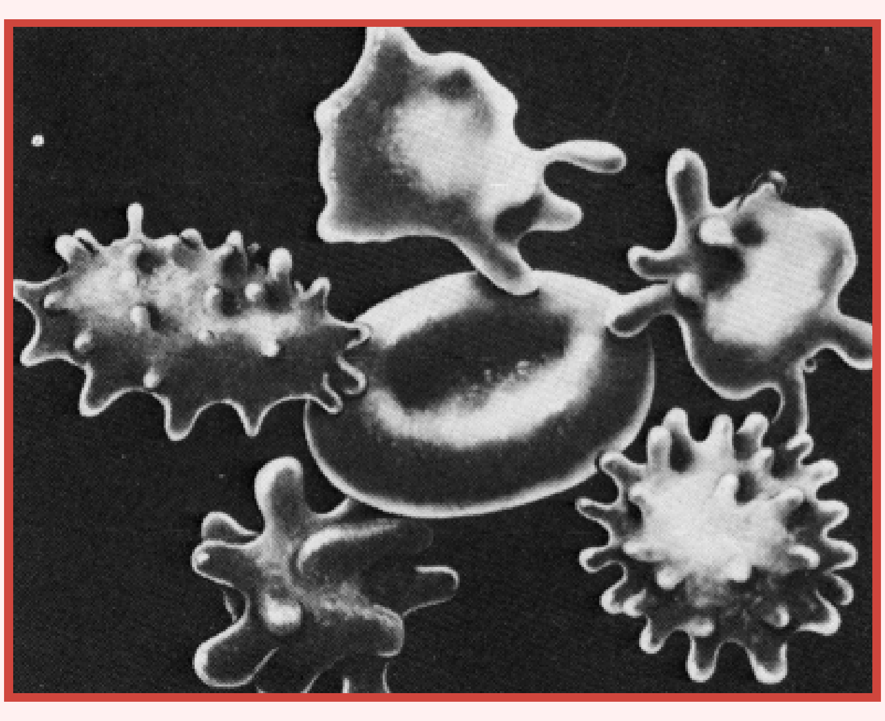


Formation of an acanthocyte spicule.

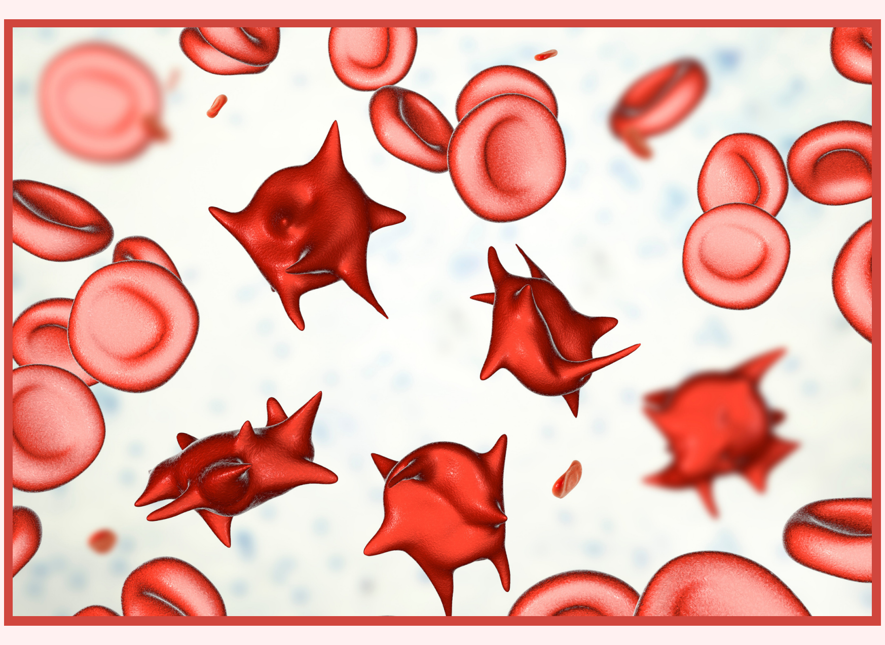
ACANTHOCYTE PERSPECTIVES



Wright-Giemsa stained peripheral smear showing an acanthocyte in the middle of the field (50x).



Artistic composite illustrating a centrally placed discoyte (normal red cell) with acanthocytes at the 12-, 2-, and 7-o'clock positions and echinocytes at 4- and 9-o'clock.



3D schematic of acanthocytes.

DID YOU KNOW?

HISTORY OF MEDICINE

1950: The first description of acanthocytes was in a young patient with diffuse, progressive neurologic disease. "The strangely distorted red cells were noted during the course of a routine blood count . . . erythrocytes of this type have never previously been described." They were noted to have the appearance of small crabs, beetles of stars. *Blood. 1950;5:381-87.*

1952: A case report was published of another young patient with diffuse, progressive neurologic disease and similar red cell changes. The authors introduced the term *acanthocyte*. "Since the most conspicuous feature of these abnormal erythrocytes is their distorted 'thorny' appearance in wet preparations and in the film, we have called them acanthocytes (akantha, thorn in Greek)." *Blood. 1952;7:577-591.*

1968: Acanthocytes first described in postsplenectomy state. *New Eng J Med. 1968:279:948.*

1970: "To distinguish the acanthocytes of abetalipoproteinemia from morphologically similar but chemically dissimilar spiky red cells associated with liver disease the term "spur cell" has been used to describe the latter." *Br Med J 1970;2(5701):68.*

NOTES

ATTRIBUTIONS
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The Blood Project
ENCYCLOPEDIA OF BLOOD