VALVE HEMOLYSIS

TERM DEFINITION

Mechanical destruction of red cells caused by prosthetic valve dysfunction and mediated by hydrodynamic shearing of the erythrocytes by turbulent flow.

- **Subclinical hemolysis** is hemolysis without anemia
- **Hemolytic anemia** is hemolysis with anemia
- **Paravalvular leak** refers to blood flowing through a channel between the structure of the implanted valve and cardiac tissue as a result of a lack of appropriate sealing

INCIDENCE

SURGICAL

AV REPLACEMENT

- Subclinical hemolysis in 20%-50% mechanical valves, 3%-5% biosynthetic valves
- <1% hemolytic anemia

**AV = aortic value*

MV REPLACEMENT

- Subclinical hemolysis in 20%-50% mechanical valves, 3%-5% biosynthetic valves
- <1% hemolytic anemia

**MV = mitral value*

TRANSCATHETER

TAVR

- 15% subclinical hemolysis
- < 1% hemolytic anemia

**TAVR = transcatheter aortic valve replacement*

TMVR

- 4%-40% subclinical hemolysis
- 3% hemolytic anemia

**TMVRV = transcatheter aortic valve repair*

DIAGNOSIS

BLOOD

Completed blood count
- Anemia
- Macrocytosis (from elevated reticulocytes)

Products of red cell lysis
- LDH
- AST
- Free Hb

Sequala of free Hb scavenging by haptoglobin (Hp)
- Decreased haptoglobin

Sequala of macrophage uptake of Hb-Hp complex
- Increased indirect bilirubin

ECHOCARDIOGRAM* +/- CARDIAC CTA

- Typically shows paravalvular leak

URINE

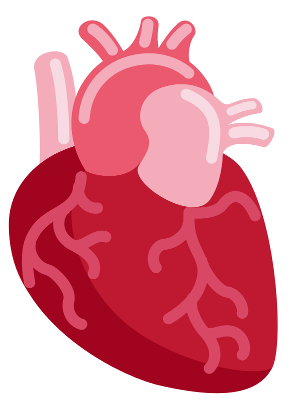
- Hemoglobinuria
- Increased urobilinogen
- Urine hemosiderin

** transesophageal echocardiography (TEE) preferred if peri-mitral leak is suspected*

RISK FACTORS

INCREASED RISK OF HEMOLYSIS

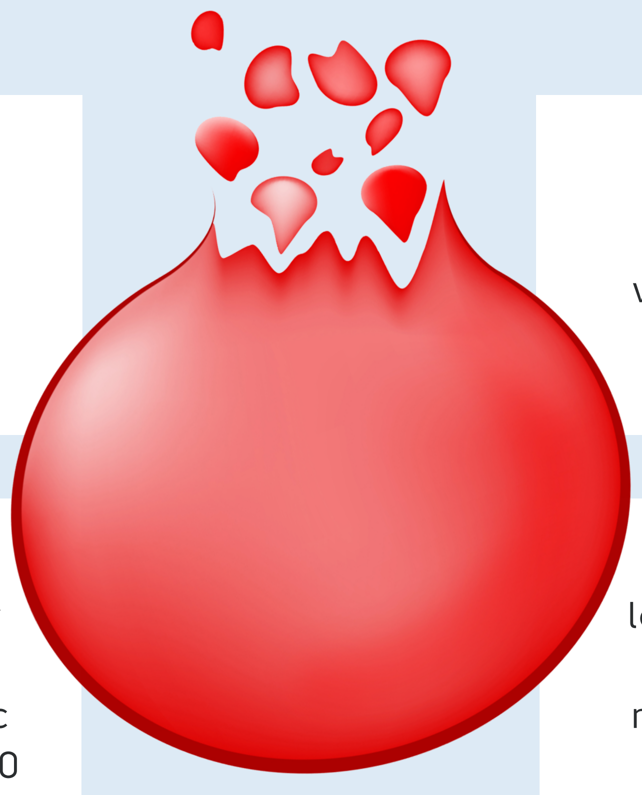
- Double-valve versus single-valve replacement
- Mechanical vs. bioprosthetic valve
- Mitral valve vs. aortic valve
- Smaller defects and high-velocity jets
- Pre-existing anemia



CLINICAL PEARLS

Patient typically presents insidiously with new onset fatigue and pallor +/- jaundice and dark-colored urine.

Hemolytic anemia has been described in native valves, especially severely stenotic aortic valves.



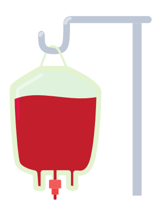
With valve replacement, hemolytic anemia can occur acutely or years later; with mitral valve repair, hemolytic anemia occurs about mean 90 days post procedure.

A paravalvular leak may lead to development of new or changed regurgitant murmur, unexplained heart failure and pulmonary hypertension.

TREATMENT PRINCIPLES

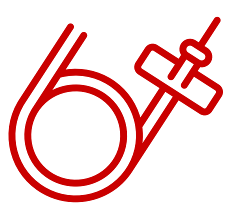
MEDICAL

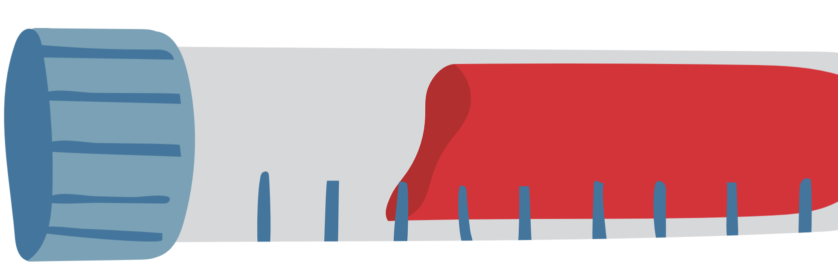
- Iron supplementation
- Folate supplementation
- Pentoxifylline
- Beta blockers
- Erythropoietin
- Transfusion



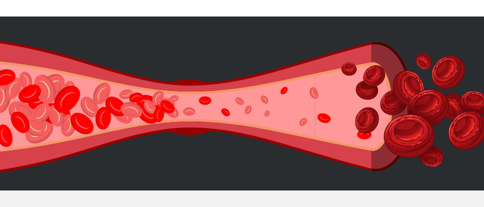
SURGICAL

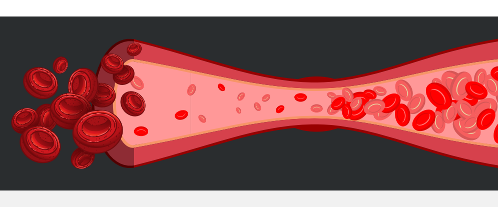
Percutaneous closure of paravalvular leak; if contraindicated or unsuccessful, open heart surgery





PROXIMATE MECHANISMS

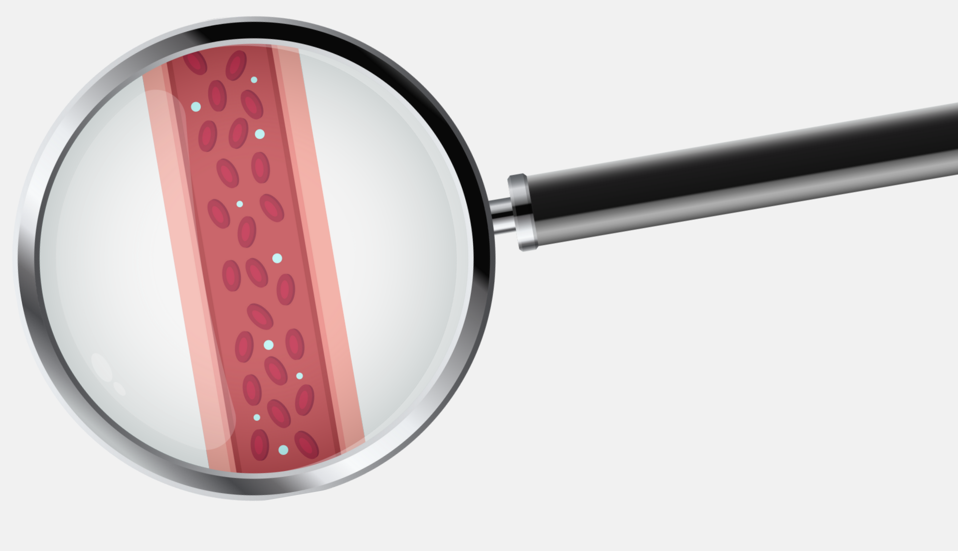




Valve hemolysis is usually caused by **paravalvular leak**. Less commonly it is associated with structural deterioration of the valve. Mechanism of hemolysis is a turbulent flow through the valve or between the sewing ring and the native ring.

USUALLY CAUSED BY PARAVALVULAR LEAK

- Occurs when the prosthetic valve's ring fails to seal adequately or adhere to the native cardiac tissue, resulting in results in regurgitation of blood from downstream to upstream chamber
- Increased red blood cell shear stress due to turbulent flow through the defect can cause mechanical trauma, fragility and fragmentation of red blood cells



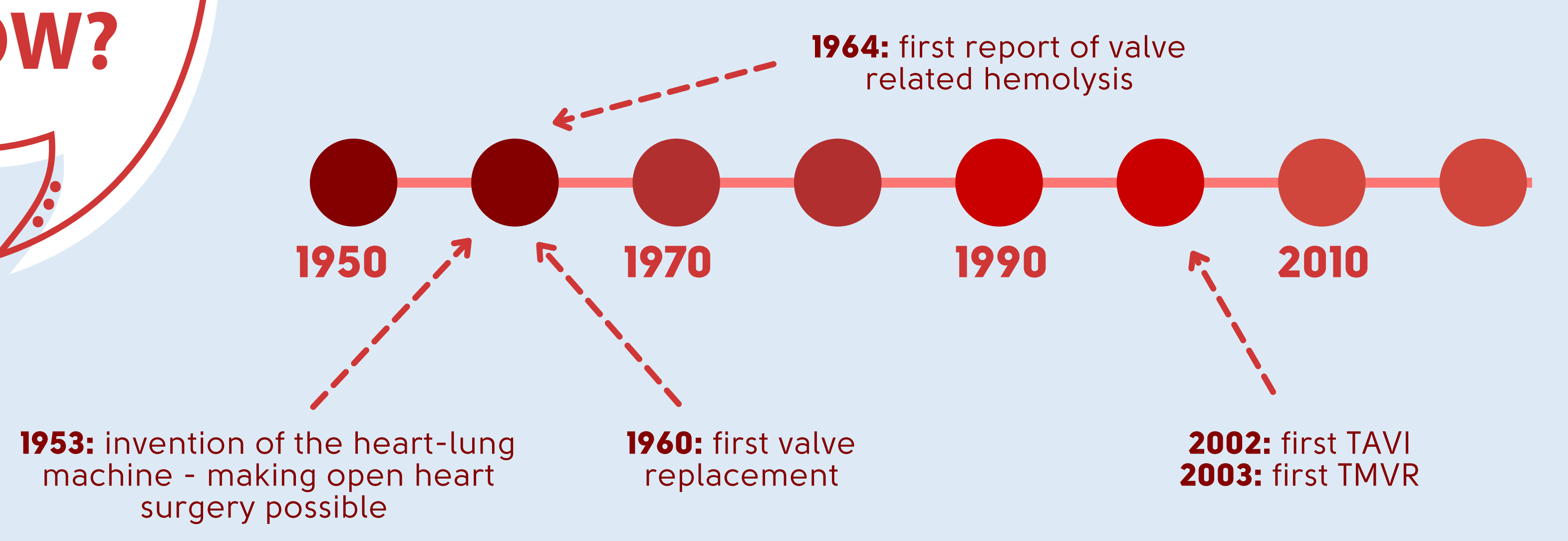
Less commonly occurs from structural deterioration of prosthetic valve

Rarely, native aortic stenosis can lead to hemolysis due to flow acceleration across the stenotic valve

DID YOU KNOW?

HISTORY OF MEDICINE

In early reports of valve-associated hemolysis, there was no mention of performing a peripheral smear and observing schistocytes! Hemolytic anemia was reported in 5%-15% of prosthetic valves in the 1960s and 1970s (vs. < 1% today).



1964: first report of valve related hemolysis

1953: invention of the heart-lung machine - making open heart surgery possible

1960: first valve replacement

2002: first TAVI
2003: first TMVR

NOTES

ATTRIBUTIONS

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The Blood Project
ENCYCLOPEDIA OF BLOOD