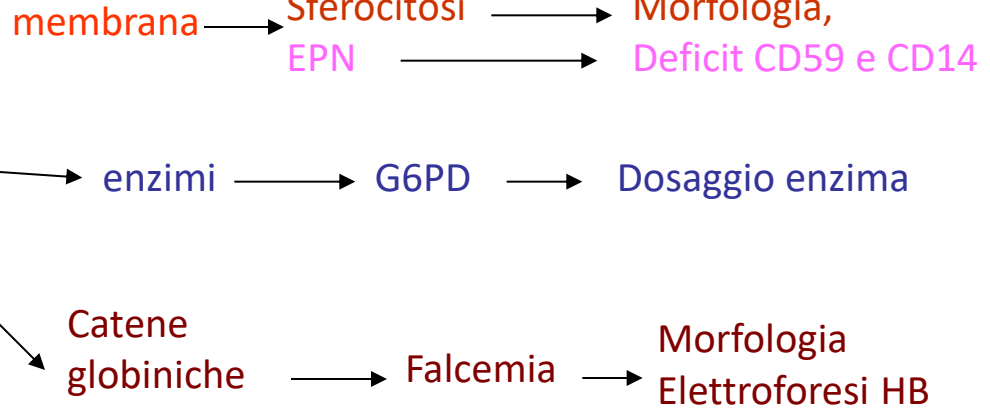


# Ridotta sopravvivenza GR

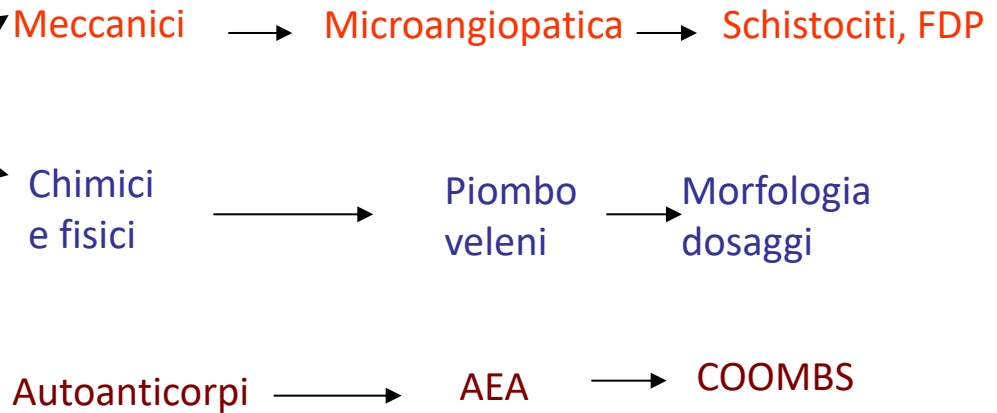
Indici emolisi

- Reticolociti
- Bilirubina indiretta
- LDH, sideremia
- Aptoglobina

intraglobulari



extraglobulari



DIAGNOSI

clinica

Test  
specifici



# Serologic classification of AIHA

**Table 41-1** Characteristics of Autoimmune Hemolytic Anemia (AIHA)

Characteristic	(~80%) Warm Reactive	Type of AIHA	(~20%) Cold Agglutinin Disease	rare Paroxysmal Cold Hemoglobinuria
Antibody isotype	IgG, Rare IgA, IgM		IgM	IgG
Direct antiglobulin test (DAT) result	IgG, Rare C3		C3	C3
Antigen specificity	Multiple, primarily Rh		i/I, Pr	P
Hemolysis	Primarily extravascular		Primarily extravascular	Intravascular
Common disease associations	B-cell neoplasia lymphoproliferative, collagen-vascular		Viral, neoplasia	Syphilis, viral

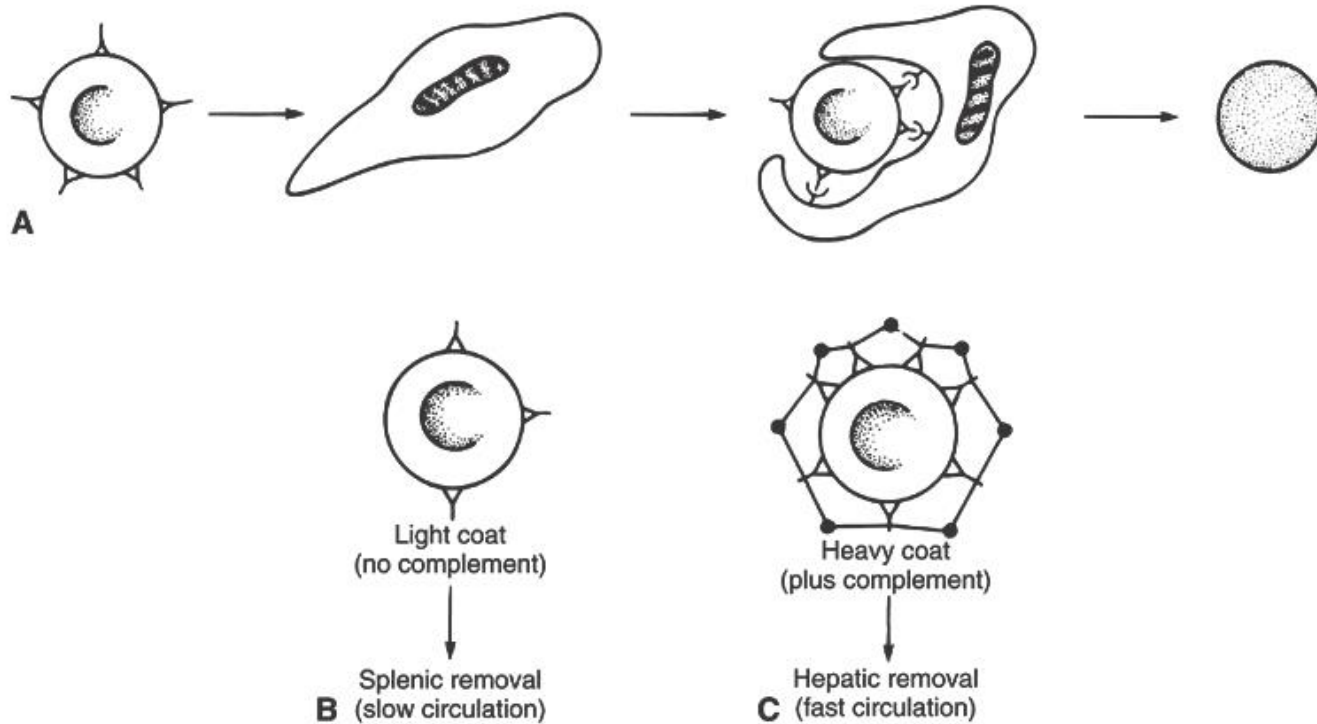
Copyright © 2005 Elsevier Inc. (USA) All rights reserved.

↓  
Acute onset  
Gradual onset

↓  
Acute form (mycoplasma pneumoniae)  
Chronic form (LNH)

Thermic range IgM (0-37 °C)  
Thermic range complement (25-40 °C)  
Overlap around 25°C

# Mechanism of extravascular hemolysis in autoimmune hemolytic anemia



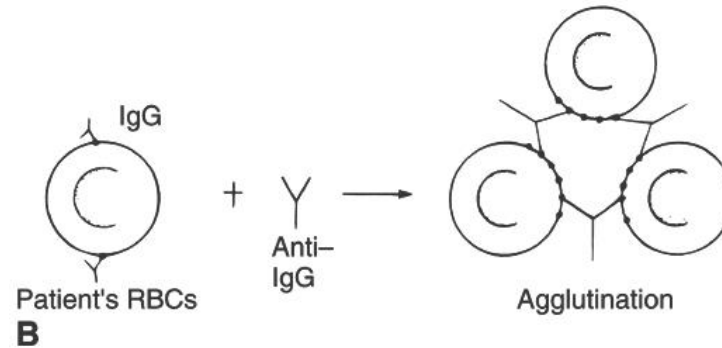
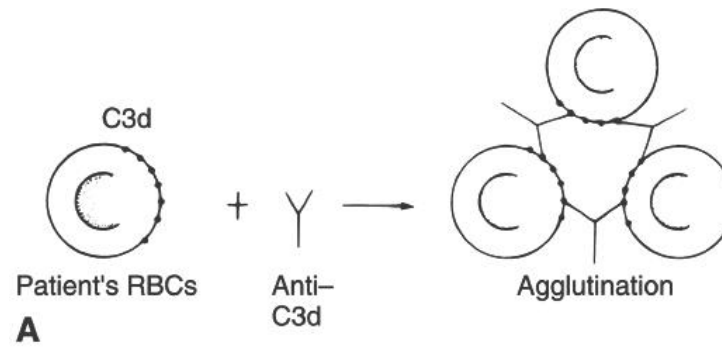
Copyright © 2005 Elsevier Inc. (USA) All rights reserved.

- (A) Macrophage encounters an IgG-coated erythrocyte and binds to it via its Fc receptors. Thus entrapped, the RBC loses bits of its membrane as a result of digestion by the macrophage's ectoenzymes. The discoid erythrocyte transforms into a sphere.
- (B) RBC lightly coated with IgG (and therefore incapable of activating the complement cascade) is preferentially removed in the sluggish circulation of the spleen.
- (C) RBC with a heavy coat of IgG; C3b (black circles) can be removed both by the spleen and the liver.

## Table 41-2 Diseases Rarely Associated with Autoimmune Hemolytic Anemia

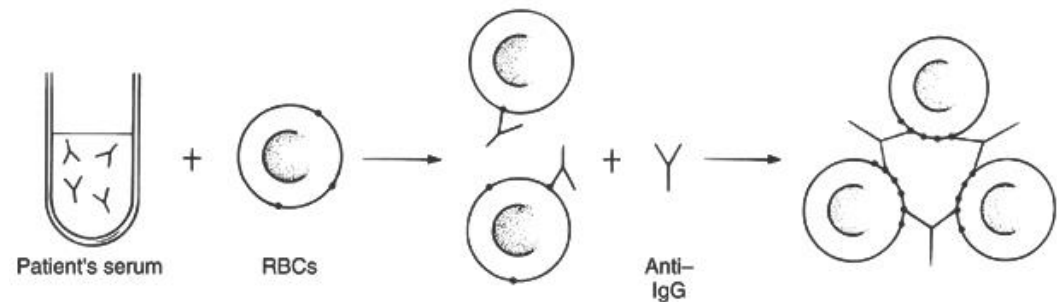
- Collagen vascular disease
  - Rheumatoid arthritis
  - Scleroderma
  - Polyarteritis nodosa
  - Serum sickness
  - Sjögren's syndrome
- Lymphoreticular malignancy
  - Macroglobulinemia
  - Hodgkin's disease
  - Multiple myeloma
  - Mycosis fungoides
- Other malignancy
  - Acute leukemia
  - Thymoma
  - Carcinoma: colon, kidney, lung, ovary
- Miscellaneous diseases
  - Myelofibrosis with myeloid metaplasia
  - Ulcerative colitis
  - Pernicious anemia
  - Thyroid disease
  - Ovarian cysts
  - Mucocutaneous lymph node syndrome (Kawasaki disease)
  - Evans' syndrome (thrombocytopenia and hemolytic anemia)
  - Congenital immunodeficiency syndromes
  - Guillain-Barré syndrome
  - Primary biliary cirrhosis
  - Multiply transfused patients with hemoglobinopathies

## Test di Coombs diretto



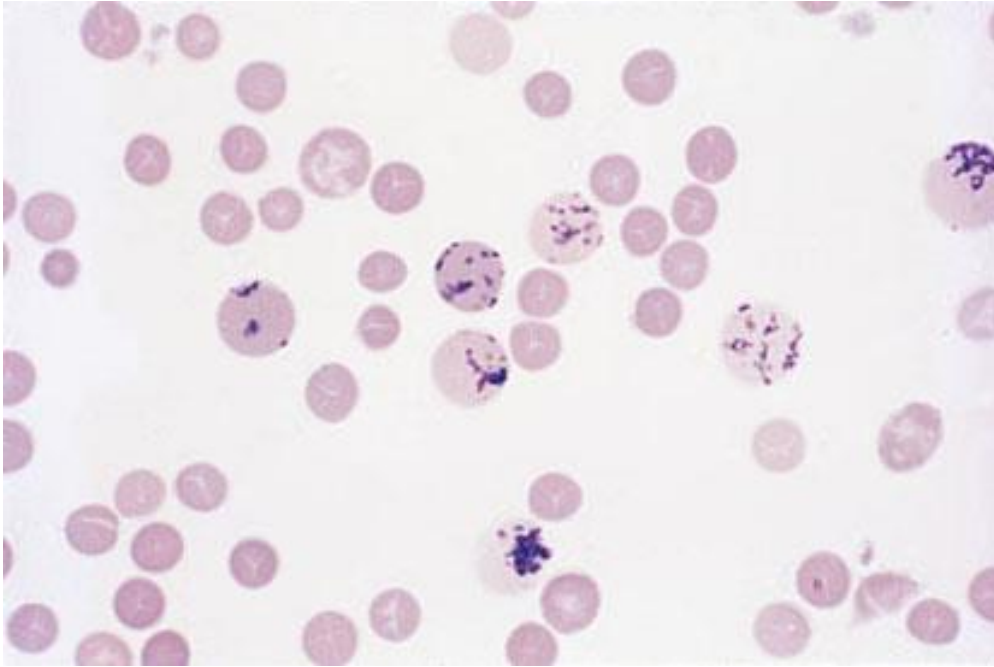
Copyright © 2005 Elsevier Inc. (USA) All rights reserved.

## Test di Coombs indiretto



Copyright © 2005 Elsevier Inc. (USA) All rights reserved.

## DIAGNOSI

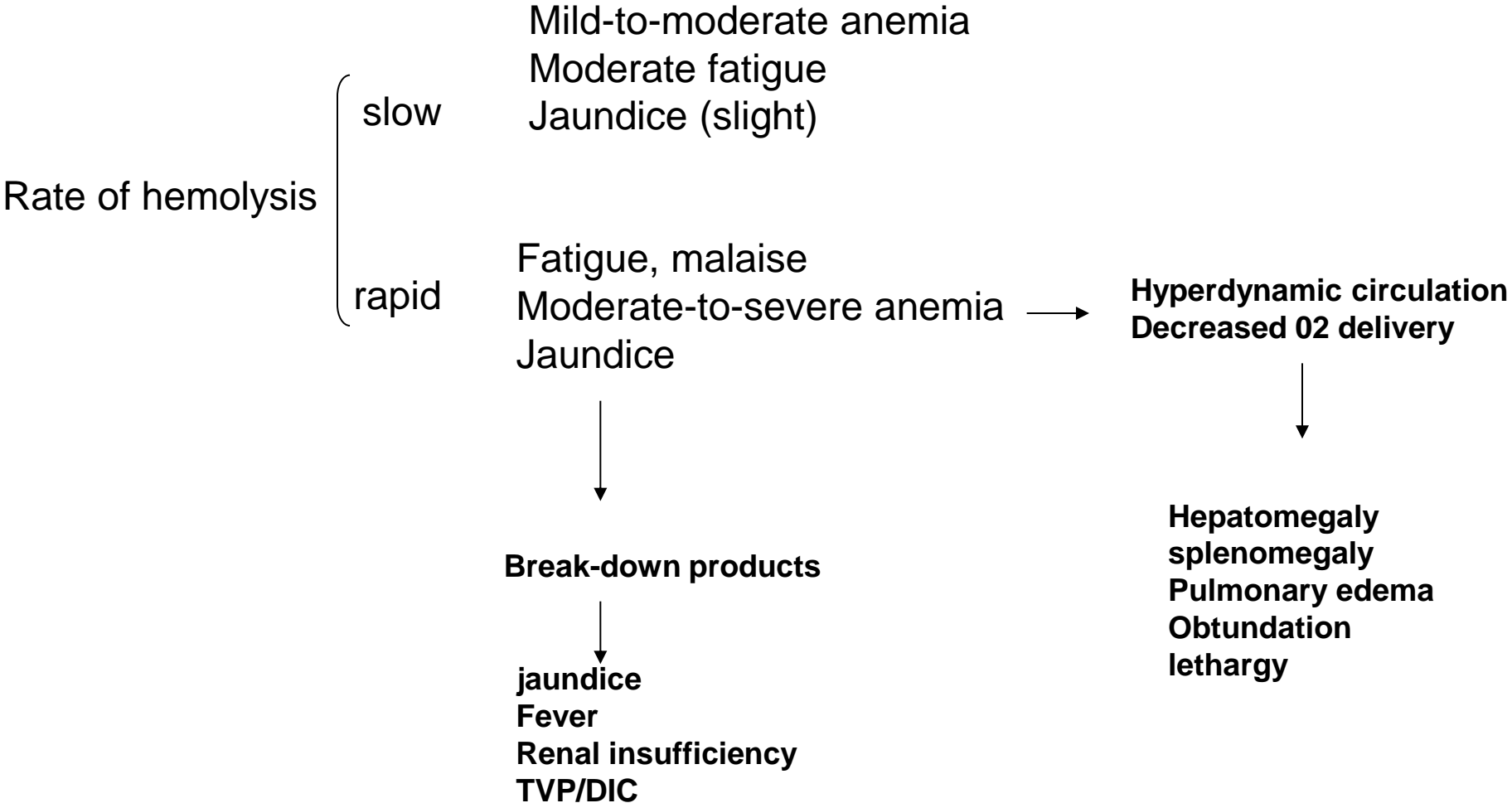


Anemia normocromica con sferociti  
Bilirubina  
Sideremia  
aptoglobina  
LDH  
Urobilinuria  
Bilirubinuria  
Feci ipercromiche  
Coombs (IgG / IgM / C3b, C3d)

**Anemia emolitica: sferociti, macrociti e reticolocitosi.**  
**Nelle cellule più grandi si vede materiale reticolare**  
**(RNA precipitato).**  
**Colorazione con blu di metilene.**  
**(Controcolorazione con Giemsa)**

Fegato  
Milza  
linfonodi  
Pallore  
ittero

# SYMPTOMS



## Terapia

Steroidi pdn 1mg/Kg/die per 4 settimane a scalare lungo 4 settimane  
(inibizione fagocitosi da parte dei macrofagi + immunosoppressione)

Immunoglobuline (400 mg/kg/ die per 3 gg)  
(saturazione recettore Fc macrofagico)

## Splenectomia

### Immunosoppressori

Alchilanti (azatioprina, ciclofosfamide, anti CD20, ciclosporina)

Trasfusioni (anche se non perfettamente compatibili per gli autoanticorpi)