# AUTOIMMUNE HEMOLYTIC ANEMIA

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## Acquired haemolytic anaemia

**Immune**
Autoimmune haemolytic anaemias
Drug-induced immune haemolytic anaemia
Isoimmune:
- haemolytic transfusion reaction
- haemolytic disease of the newborn

**Red cell fragmentation syndromes**

**Hypersplenism**

**Paroxysmal nocturnal haemoglobinuria**

**Secondary**
Renal disease, liver disease, etc.

**Miscellaneous**
Chemicals
Drugs
Infections
Toxins
Wilson's disease

**Fig. 4.36** Acquired haemolytic anaemia: causes.

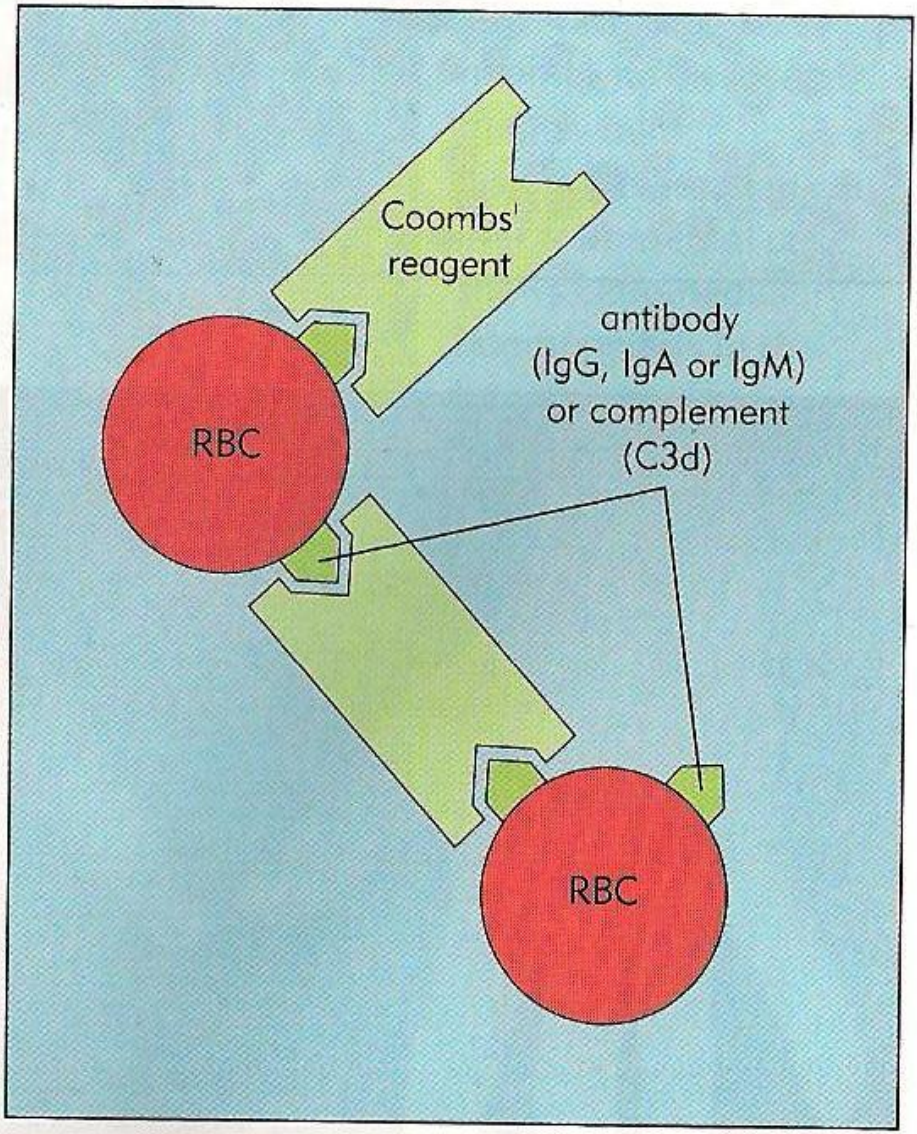



**Fig. 4.37** Direct antiglobulin (Coombs') test: the Coombs' reagent may be broad spectrum or specifically directed against IgG, IgM, IgA or complement (C3d). The test is positive if the red cells agglutinate.

| Autoimmune haemolytic anaemia | |
|---|---|
| **Warm type** | **Cold type** |
| Idiopathic | Idiopathic |
| Secondary | Secondary |
| Systemic lupus erythematosus, other connective tissue disorders | Mycoplasma pneumonia |
| Chronic lymphocytic leukaemia | Infectious mononucleosis |
| Malignant lymphoma | Malignant lymphoma |
| Ovarian teratoma | Ulcerative colitis |
| Drugs (e.g. methyldopa, fludarabine) | Paroxysmal cold haemoglobinuria: rare; may be primary or associated with infection |

**Fig. 4.38** Autoimmune haemolytic anaemia: causes.

# EPIDEMIOLOGY

- Incidence: 10:1000000 population
- Women>men
- Usually midlife, can occur at any age
- 50% idiopathic
- Can be associated with autoimmune diseases, drugs, B-lymphoproliferative disorders – CLL, NHL

# CLINICAL FINDINGS

- Jaundice, usually mild
- Signs and symptoms of anemia – acute or chronic
- 30% splenomegaly
- Lymphadenopathy, fever, renal falure, rash, petechiae or echymoses – alert of other underlying disease
- Evan's syndrome – AIHA and Imuune Thrombocytopenia

# LABORATORY EVALUATION

- Anemia with enhanced erythropoesis
- Reticulocytosis
- Blood smear: spherocytes, occasional fragments, nucleated RBC
- Bone marrow – erythroid hyperplasia, megaloblastosis with folate deficiency

# LABORATORY EVALUATION

- Unconjugated bilirubinemia, increased LDH, low haptoglobin
- Intravascular hemolysis – free Hb in plasma, hemosiderin in urine
- DAT + IgG or Complement on patient's RBC - in 80% of AIHA positive

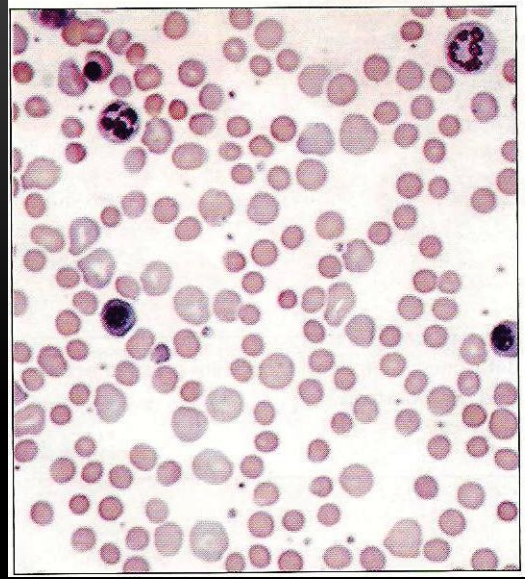


**Fig. 4.39** Autoimmun haemolytic anaemia: peripheral blood film showing erythroblasts, polychromatic macrocytes and marke spherocytosis.

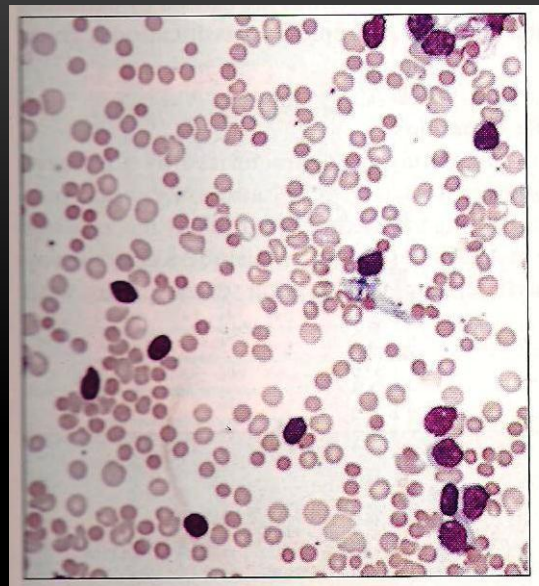


**Fig. 4.40** Autoimmune haemolytic anaemia with associated chronic lymphocytic leukaemia: peripheral blood film showing red cell polychromasia spherocytosis and increased numbers of lymphocytes.

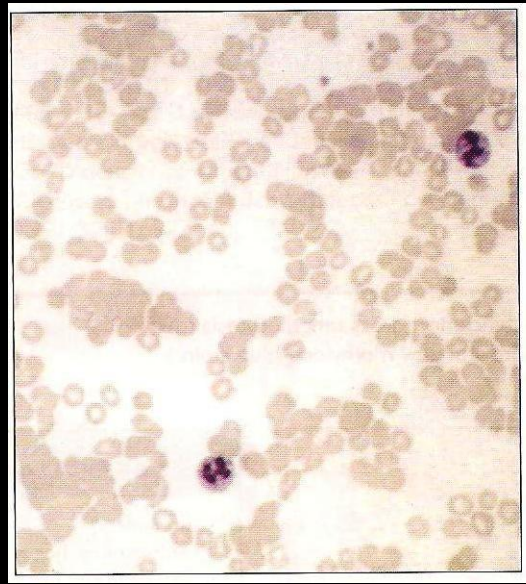


**Fig. 4.41** Autoimmun haemolytic anaemia (cold type): peripheral blood film showing autoagglutination of r cells.

**Fig. 4.43** Autoimmune haemolytic anaemia: peripheral blood film show

# TREATMENT

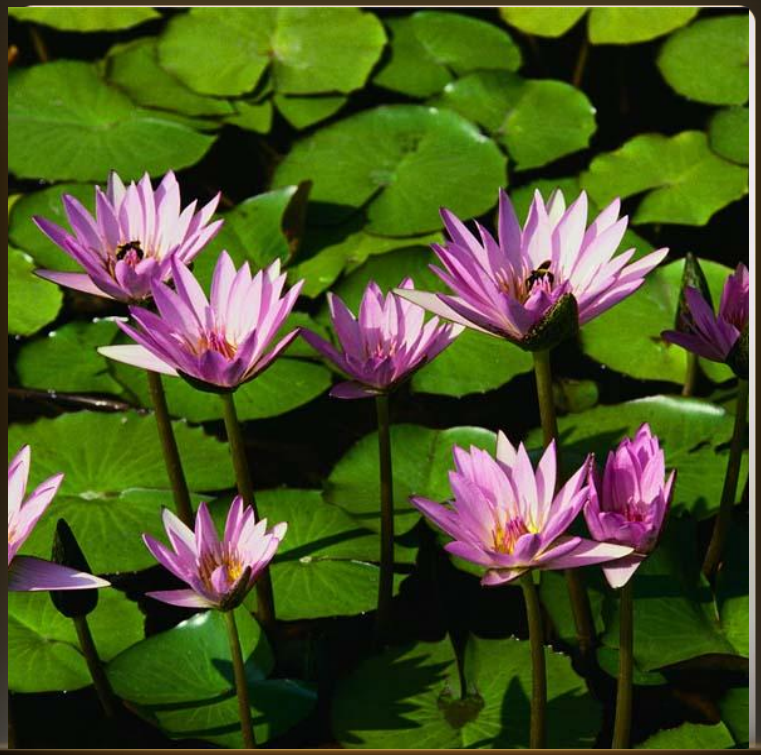
- Transfusion, if severe symptomatic anemia, with steroids, close follow up and monitoring
- Corticosteroids – prednisone 1-2 mg/kg/day in two divided doses, continue until Hb≥10, than slow tapering down
- Splenectomy in steroid refractory or dependent cases, 50-60% response
- IVIG 0.4 gr/kg/day for 5 days
- Cytotoxic: azathioprine, cytoxane, vincristine
- Danazol

# COLD AGGLUTININ DISEASE

- Antibodies that bind RBC at cold temperature (5-18°C), usually IgM
- Chronic – idiopathic or associated with B cell lymphoma
- Transient – post infectious Mycoplasma Pneumonia, EBV, HIV, collagen vascular disease

# THERAPY

- Warming, warmed blood transfusion
- Prednisone, splenectomy - mostly non beneficial
- Plasma exchange - temporal relief
- Chemotherapy – azathioprine, CVP
- Immune suppression – Ciclosporin A, etc.



# THANK YOU