Autoimmune hemolytic anemia (AIHAs):

Disorders caused by antibody production by the body against its own red cells.
They are characterized by a **positive direct antiglobulin test** (DAT) also known as **Coombs test.**
The antibodies may be IgG or M or more rarly IgE or A.
It can be divided into **warm** and **cold** types according to whether the antibodies react more strongly with the red cells at 37 C or 4 C

Classification:

Warm type	**Cold type**
-Idiopathic -Secondary SLE, Other autoimmune diseases, CLL, lymphomas, Drugs.	Idiopathic Secondary: **- Infection (mycoplasma pneumonia and infectious mononucleosis syndrome)** **- Lymphoma.** **- Paroxysmal cold haemoglobinuria**

1. Warm autoimmune hemolytic anemia:

Here the red cells are usually coated with immunoglobulin (Ig), usually immunoglobulin G (IgG) alone or with complement and therefore takes by RE macrophages which have receptors to the Ig Fc fragment. Part of the coated membrane is lost so the cells become progressively more spherical to maintain the same volume and is ultimately prematurely destroyed predominantly in the spleen.

Clinical features:

- The disease may occur at any age in either sex but more common in middle ages and in females.
- No underlying cause is identified in up to 50 % of cases .The reminder are secondary to a wide variety of other condition .
- The patients presents as a hemolytic anemia of varying severity.
- The spleen is often enlarged.
- The disease tends to remit and relapse.
- When it's associated with idiopathic thrombocytopenic Purpura (ITP), which is similar condition affecting the platelets, it's known as **"Evans syndrome".**

Laboratory Finding:

The hematological finding and biochemical finding are typical of an extra-vascular hemolytic anemia with spherocytosis prominent in the peripheral blood.
The diagnosis is confirm by The DAT (coombs test) is positive .
The antibodies both in the cell surface or free in the serum are best detected at 37 C .

Treatment:

1. Treat or Remove the underlying cause (e.g. drugs) if the cause of anemia is secondary.
2. **Corticosteroid**, Prednisolone is the usual first line of treatment. The initial dose should be Prednisolone 1-2 mg per kg per day.

High doses of corticosteroid should be continued for 10-14 days, according to response.

A response is seen in 70-80% of cases but may take up to 4 weeks. A rise in hemoglobin will be matched by a fall in bilirubin, LDH and reticulocytes level.

Once the hemoglobin has normalized and Reticulocytosis resolved , the corticosteroid dose can be reduced slowly over about 10 weeks.

The dose can be give once daily if tolerated.

3. **Splenectomy**: May be of value in those who fail to response well or fail to maintain a satisfactory hemoglobin level on an acceptably small steroid dosage.

If splenectomy is not appropriates , other line of treatment should be considered.

4. **Immunosuppressant**: May be tried after other measures have been failed

These include Azathioprine, cyclophosphamide, and cyclosporine.

5. **Folic acid** is given to sever cases.
6. **Blood transfusion** may be needed if anemia is sever and causing symptoms.
7. **High dose immunoglobulin** has been used but with less success than in ITP.

2. Cold autoimmune hemolytic anemia:

In these syndrome the antibody whether, monoclonal attaches to red cells mainly in the peripheral circulation where the blood temperature is cooled.

The antibody is usually IgM and binds to red cells best at 4 C.

IgM antibodies are highly efficient at fixing complement and both intravascular and extra vascular hemolysis can occur.

Clinical features:

The patients have a chronic hemolytic anemia aggregated by the cold and often - associated with intravascular hemolysis.

- Mild jaundice and Splenomegaly may be present. -
- The patient may develop acrocynosis (purplish skin discoloration) at the tip of the - nose, ears, fingers and toes caused by the agglutination of the red cells in small vessels.

Laboratory Finding:

Are similar to those of warm AIHA except that spherocytosis is less marked, red cells agglutinate in cold.

Treatment:

- Consist of keeping patients warm**.**
- Treating the underlying cause, if present.
- Alkylating agents such as chloambucil may be helpful in chronic varieties.
- Splenectomy dose not usually help unless massive Splenomegaly is present.
- Underlying lymphoma should be excluded in idiopathic cases.