

# CASE REPORT

## AUTOIMMUNE HEMOLYTIC ANAEMIA IN CHILDHOOD: CASE REPORT

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**ABSTRACT:** The hemolytic anemia brought about by autoimmune antibodies against red cells resulting in shortening of red cell survival. And excessive erythropoiesis as a compensatory response of the marrow is autoimmune hemolytic anemia. There are two types due to warm antibodies and cold antibodies. Warm antibodies mediated anemia has hemolysis in spleen mediated by IgG antibody. Macrophage Fc receptor can detect IgG coated red cells in absence of C 3b. It acts in the range of 37-42deg c.<sup>(1)</sup> Cold antibody mediated anemia is mediated by IgM antibody, they bind avidly at 4 deg c. carry hemolysis at low temp. Liver is the major site of hemolysis. Autoimmune hemolytic anemia presents as sudden onset of pallor, jaundice, dark coloured urine, and hepato splenomegaly. In 5 cases cold antibodies were positive, 3 cases warm were positive too. 4 of the 5 cases were females and steroids were effective in management of cold auto immune hemolytic anemia.

**KEYWORDS:** Cold antibody, Hemolytic, Anaemia.

**INTRODUCTION:** The Hemolytic Anaemia brought about by autoimmune antibodies against red cells resulting in shortened red cell survival and increased erythropoiesis as a compensatory response of the marrow is called Autoimmune Hemolytic Anaemia.<sup>(2,3)</sup>

The incidence of autoimmune hemolytic anaemia varies from 1 per 80,000 to 2.6 per 1,00,000 population world wide in India, in spite of much literature search, we couldn't get case reports or studies mentioning incidence.<sup>(4,5)</sup>

There are two types of autoimmune hemolytic anaemia - due to either 1. Warm Antibodies or 2. Cold Antibodies.

The anaemia due to Warm antibodies may be Primary (Idiopathic) or Secondary to Lymph proliferative disorders, connective tissue disorders (especially SLE) Non-lymphoid neoplasms (Eg: Ovarian Tumors), chronic inflammatory diseases (eg. Ulcerative colitis) or drug induced (Methyldopa, Levodopa).

The anaemia due to Cold antibodies may be Primary (Idiopathic) or Secondary to lymph proliferative disorders, infections like Mycoplasma Pneumonia, Epstein Barr virus, Congenital or Tertiary Syphilis.

Warm Antibody mediated Anaemia, It is mediated by IgG antibody which act in range of 37 C - 42 C. The Major Site of hemolysis is Spleen. Macrophage FC receptor can detect IgG coated red cells even in the absence of C3b.<sup>(6)</sup>

Cold Antibody mediated Anaemia. This is mediated by IgM antibody. They bind avidly at 40C and carry hemolysis at low temperature. Liver is major site of hemolysis. IgM antibodies interact with red cell membrane and activate complement system on exposure to cold. A Single

# CASE REPORT

molecule of IgM antibody activates complement C1, which further activates fourth and second component by enzymatic cleavage to form a complex C142 or C3 convertase and fragments. Red cells coated by C3b molecules are cleared by macrophages. Macrophages have no receptors to detect IgM coated cells in absence of complement. Infections like Mycoplasma Pneumonia, Infections mononucleosis target oligosaccharide antigens I&I and form anti I and anti-antibodies respectively.

Autoimmune hemolytic anaemia common presents with sudden onset of pallor, jaundice, dark colored urine and Hepatosplenomegaly. Children may appear acutely ill with tachycardia, signs of hypoxia and even cardiovascular collapse. Two types of Presentation, acute arising in age group 2-12 Years, seen in 70-80% of cases and the other i.e. Chronic Presentation is seen in infancy and >12 years of age with high mortality rate of 10%.<sup>(7,8)</sup>

At Niloufer Hospital, a teaching hospital in Telangana, we have seen 5 case of autoimmune hemolytic anaemia from February to June. These cases are studies from clinical etiological pathological and therapeutic aspects.

**SUMMARY OF THE CASE:** Total of 5 Cases were studies all the 5 cases showed cold antibodies out of which 3 had warm antibodies too. In addition 2 cases were known cases of Thalassemia and one case revealed antibodies for collagen vascular disease. Other details of the cases are given in the following tables.

Clinical Features	Case No.	1	2	3	4	5
Age (In Yrs)/ Sex		12/F	3/F	9/M	10/ 12 M	7/F
Pallor		Present	Present	Present	Present	Present
Icterus		Present	Present	Absent	Absent	Present
Liver		1.5 Cm below RCM	4cm below RCM	4cm below RCM	2.5cm below RCM	1cm below RCM
Spleen		4 cm below LCm	3 cm below LC	2 cm below LC	3 cm below LC	Tip Palpable

Table 1

M: Male F: Female RCM: Right Costal Margin, LCM: Left Costal Margin.

Investigation	1	2	3	4	5
Haemoglobin	9g%	8.2g%	3.6g%	8g%	10.6g%
Platelets	3 lakh/mm3	1.2 lakh/mm3	1.2 lakh/mm3	1.4 lakh/mm3	1.8 lakh/mm3
Reticulocyte Count	4%	5%	5%	8%	3%
Hb Electrophoresis	Normal Adult HbA	Thalassemia major	Normal Adult HbA	Thalassemia Major	Normal Adult HbA

# CASE REPORT

Direct Coombs Test	+	+	+	+	+
Indirect Coombs test	-	-	+	-	+
Warm Antibodies	+	-	-	+	+
Cold Antibodies	+	+	+	+	+

Apart from features mentioned in tables above, our cases had no generalized lymphadenopathy, rash or any other feature suggestive of infection. Malaria was excluded and C - reactive protein was uniformly negative. X-Ray chest was normal in all of them. There were no atypical lymphocytes. WBC Count was adequate, Polychromasia was present and bone marrow aspiration reports didn't given any evidence of lymph proliferative disorder. The collagen vascular disease profile was carried out and was negative in all of them except case No. 5 in which Anticardiolipin antibodies were positive. Paul Bunnell test to detect EBV infection has been done, but as it does not rule out infection in 10-20% of cases, cases <5 Years of age, and the non-availability of PCR Test, it was not possible to rule out EBV Infection. There was no h/o. drug intake in any of the cases. Case No. 4 presented with Acute Diarrhoea is incidental since etiologically organisms producing acute diarrhoea are not known to cause immune hemolytic anaemias. All the cases were treated with Prednisolone 1mg/kg/day for 2-4 weeks except Case No. 5 who was switched over to alternate day steroid after Haemoglobin reached 13 g%.

**RESULTS:** 4 cases were in age group of 2-12 years. I was 10 month old.

4 cases out of 5 were females. All were acute presentations, except case No. 5 which was chronic. The etiology of first four cases was idiopathic, whereas for case No. 5 it was connective tissue disorder. All of them had cold antibodies positive with 3 cases having warm antibodies. Steroids were effective in the Management of cold autoimmune hemolytic anaemia.

**DISCUSSION:** As per the Literature available, boys are said to be predominantly affected by auto immune hemolytic anaemia, but in our 5 cases 4 of them were girls.<sup>(8)</sup> First four cases were acute and idiopathic, a common mode of presentation. Case No. 5 was chronic, had connective tissue disorder. The features of connective tissue disease were not present, perhaps may manifest in due course of time as majority of them do. Case No. 4 who is an infant presented acutely which is an exception. In case No. 2 who is the thalassemic and cold antibodies positive, the infective focus causing any hemolytic anaemia is more common. But available resources say that warm auto immune hemolytic anaemia is more common.<sup>(5,6)</sup> But all the cases had cold antibodies with only 3 of them being positive for warm antibodies.

Contrary to the general opinion our studies in above 5 cases proved that steroids are effective in the management of cold autoimmune hemolytic anaemia. The cases started responding from 7-10 days and by 4 weeks they were normal. The Patients required no blood transfusions after 10-12 days of onset of therapy and hepatosplenomegaly slowly regressed. The

# CASE REPORT

most probable explanation is that cold agglutinins may have high thermal amplitude as far as 37°C and steroids avoid interaction of macrophage with C3 coated erythrocytes.

Our experience with these cases shows that steroids are not to be labeled as ineffective in the management of cold autoimmune hemolytic anaemia and they should be tried at earliest as first line of management.

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