Abstract: Evans syndrome is an idiopathic direct Coombs positive autoimmune haemolytic anaemia with immune mediated thrombocytopenia. It presents with thrombocytopenia, anemia, neutropenia and pancytopenia in the decreasing order of frequency. The mainstay of management is medical therapy which includes stabilisation of respiratory and cardiovascular dysfunction, transfusion of blood products and pharmacological therapy. It is rare to come across an antenatal mother with Evans syndrome who goes on to complete term and deliver the baby. A 28 year old antenatal mother who came to our hospital as an emergency was a case of diagnosed Evans syndrome on Prednisolone 40mg BD. She was referred from a local hospital to our tertiary care centre. She had gone into labour at 38 weeks of gestation. She was treated medically for thrombocytopenia and underwent emergency LSCS (Lower segment Caesarean section). Both baby and mother were normal at the time of discharge.

Keyword: Evans syndrome, pregnancy, thrombocytopenia, autoimmune haemolytic anaemia

Introduction: Evans syndrome is an idiopathic direct Coombs positive autoimmune haemolytic anaemia with immune mediated thrombocytopenia. It is more common in the female population. It presents with thrombocytopenia, anemia, neutropenia and pancytopenia in the decreasing order of frequency. Lefkou et al from the Department of Hematology in London did a thorough search of electronic databases till 2008 and found 14 cases and they reported 2 cases totaling to 16 cases. I would like to highlight that this is the next case after that and can be called the 17th case. Diagnostic armamentarium includes complete hemogram, reticulocyte count, direct Coombs test, lupus antibody and ANA (Anti Nuclear Antibody). The mainstay of management is medical therapy which includes stabilisation of respiratory and cardiovascular dysfunction, transfusion of blood products and pharmacological therapy. It is rare to come across an antenatal mother who goes on to complete term and deliver the baby.

Case Report
History - A 28 year old antenatal mother who came to our hospital as an emergency was a case of diagnosed Evans syndrome on Prednisolone 40mg BD. She was diagnosed at the age of 16yrs when she was evaluated for abnormal uterine bleeding. Her lab reports showed anemia and thrombocytopenia. During pregnancy her platelet count was normal and the dose of prednisolone was reduced to 20mg bd. She was referred from a local hospital to our tertiary care centre. She had gone into labour at 38 weeks of gestation. She was a primi gravida with breech presentation and her Hb was 6.9g/dl, platelets 20000 cu mm and hematocrit was PCV 20g/dl. Treatment - She was taken for emergency lower segment caesarean section and was transfused 3 units of packed cells and 2 doses of intravenous Immunglobulin because of the low platelet count despite treatment with prednisolone. Outcome - She delivered a healthy baby and she received intravenous methyl prednisolone for 3 days followed by oral prednisolone. On discharge her Hb was 10g/dl, platelets 80000 cu mm and PCV was 30g/dl. The newborn baby was handed over to the neonatologist. Baby was on close follow up and the investigations revealed no evidence of thrombocytopenia immediate after birth, one week after birth and on follow up at 1 month, 6 months and 12 months after birth.

Discussion: Idiopathic Auto Immune Hemolytic Anemia and Immune -mediated thrombocytopenia coexisting together is called as Evans syndrome. A few cases may be associated with SLE (Systemic Lupus Erythematosus) and other autoimmune diseases, chronic lymphadenopathy or hypogammaglobulinemia. There are limited articles discussing Evans syndrome in literature and even fewer articles on pregnancy associated with Evans syndrome. Among the two components of Evans syndrome, there is an article by Webert et al published in 2003 discussing only the presence of ITP and pregnancy and the outcomes. They did a retrospective study for 11 years and studied 92 women with Evans syndrome and on follow up at 1 month, 6 months and 12 months after birth.
against erythrocytes, platelets and neutrophils. Patients show decreased serum levels of immunoglobulins (IgG, IgM and IgA). The cytopenias are related to decreased helper T cell functions and increased suppressor T cell function. Symptomatic ranges from features of thrombocytopenia (purpura, petechia and ecchymoses), anemia (pallor, fatigue and dizziness), hemolysis (jaundice) to life threatening complications like hemorrhage with severe thrombocytopenia and serious infections with neutropenia. Diagnosis of Evans includes an extensive work up comprising Complete hemogram, Reticulocyte count, Coombs test (direct), Tests for anterythrocyte, antineutrophil and anti-platelet Antibody, Lupus antibody and ANA (Anti Nuclear Antibody) tests, Measurement of serum immunoglobulins, Flow cytometry of blood samples, Gene mutation studies. Treatment includes primarily medical management. In patients with severe anemia or thrombocytopenia, symptomatic management needs to be given by replacement of the deficient blood products. Pharmacological therapy commonly used includes Prednisolone (most common first line agent), IVIgintravenous Immunoglobulin( for those with persistent immune cytopenias or requiring high dose Steroids) and Other newer generation of drugs like rituximab, danazol, cyclosporine, azathioprine, cyclophosphamide, and vincristine. Severe cases may need splenectomy as the last resort.

**Conclusion**

We present this case in view of the rarity and the emergency management required in mothers with hemolytic disorders and thrombocytopenia co-existing. It also emphasizes the need for a good Obstetric team and a neonatologist who can take care of the baby immediately after delivery. As stated in the study by Lefkou et al in 2010, successful pregnancies can be seen in mothers with Evans syndrome with good response to conventional treatment.

**Reference:**
