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Review Article

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Sickle Cell Anaemia: Haemolysis and Anemia

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Abstract

The role of inflammation cannot be over emphasized in the pathophysiology of Sickle cell disease. Adhesive sickle reticulocytes initiate vaso occlusion Sickle cell disease is characterized by chronic intravascular and extravascular haemolysis. Membrane damage also leads to extravascular haemolysis through entrapment of poorly deformable cells or uptake by macrophages. This paper discussed haemolysis and anaemia in sickle cell anaemia. Haemolysis should be prevented in sickle cell anaemia patients to prevent anaemia.

Keywords: sickle cell anaemia, Haemolysis, inflammation.

Haemolysis

The role of inflammation cannot be over emphasized in the pathophysiology of Sickle cell disease (Hagar *et al.,* 2008), studies have shown increased levels of inflammation markers in sickle cell patients even in steady state (Akohoue *et al .,* 2007). Adhesive sickle reticulocytes initiate vaso occlusion Sickle cell disease is characterized by chronic intravascular and extravascular haemolysis. Sickling- induced membrane fragmentation and complement-mediated lysis causing intravascular destruction of red cells. Membrane damage also leads to extravascular haemolysis through entrapment of poorly deformable cells or uptake by macrophages. The red cell survival measured by 51Cr assay is 4–25 days, with dense cells surviving for a considerably shorter time than red cells containing some HbF (F cells). Patients have greatly expanded bone marrow space, but the serum erythropoietin level is lower than expected for the extent of anaemia owing to decreased oxygen affinity of HbS. Individuals with concomitant deletion of one or two α-globin genes, or the Senegal or Arab– India haplotypes, have higher baseline haemoglobin levels. Co-inheritance of one or two α-gene deletions also modifies the clinical picture. The high HbF level observed in hereditary persistence of foetal haemoglobin (HPFH) is associated with very mild disease. However, for poorly recognized reasons, the disease severity varies enormously even within the subgroup of patients with HbSS.

In countries with inadequate healthcare, SCD is associated with high mortality in the first 3 years of life as a result of sepsis and splenic sequestration. In the developed world, the typical patient with SCD has moderately severe anaemia, leads a relatively normal life interrupted by ‘crises’ as a result of vaso occlusion, and has a life expectancy of over 45 years (Hoffbrand *et al*., 2010).

Anaemia

The underlying globin genotype primarily determines the baseline haemoglobin value in SCD, but exacerbation of anaemia can occur for numerous reasons. Patients with more severe anaemia at baseline have a greater probability of developing

stroke and most often renal dysfunction. Severe anaemia is the consequence of the shortened lifespan of red cells and the course is similar to other chronic haemolytic anaemia (Obeagu, 2018). Aplastic crisis is typically preceded by fever and upper respiratory or gastrointestinal symptoms, and several family members may fall ill over a period of days. The reticulocytopeniabegins 5 days after exposure, lasts for 7–10 days and is followed by recovery with reticulocytosis and normoblasts in peripheral blood. Blood transfusion is often required in the short term. Parvovirus B19 infection is followed by development of lifelong protective immunity. Splenic sequestration is a serious complication in young children whose spleen has not yet undergone fibrosis due to recurrent vaso-occlusion. The peak incidence of first episode of sequestration is between 6 and 12 months of age and it affects 30% of all patients. Approximately 15% of patients die during the acute episode and the condition reoccurs in one-half of the survivors. Episodes may be triggered by a viral illness and the rapid acute enlargement of the spleen traps a significant proportion of the blood volume. Clinically, the child presents with acutely worsening anaemia (2 g/dL fall in haemoglobin), reticulocytosis, enlarging spleen and hypovolaemic shock. Prompt restoration of the blood volume and correction of anaemia is required. Splenectomy is recommended following a sequestration crisis due to the risk of recurrences. Chronic transfusion therapy or partial splenectomy is sometimes used in infants with sickle cell disease (Obeagu *et al.,*2015; Swem *et al.,* 2018).

Conclusion

The role of inflammation cannot be over emphasized in the pathophysiology of Sickle cell disease. Adhesive sickle reticulocytes initiate vaso occlusion Sickle cell disease is characterized by chronic intravascular and extravascular haemolysis. Membrane damage also leads to extravascular haemolysis through entrapment of poorly deformable cells or uptake by macrophages. Inlammation is one of the mechanisms used to cause haemolysis in sickle cell and due to red cell membrane integrity.It is one of the ways that leads to crisis in the patients. Haemolysis should be prevented in sickle cell anaemia patients to prevent anaemia.

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