Sickle cell disease is one of the most common genetic disorders in the world, affecting over 75,000 North Americans. The causative defect, a point mutation in the beta globin gene resulting in a glutamic acid to valine switch at position six, was first described by Vernon Ingram in 1957. It is responsible for the multisystem complications that characterize the disorder including chronic hemolysis, stroke, recurrent pain crises and pulmonary hypertension.

Since Ingram’s discovery, significant advances in supportive care – the adoption of penicillin prophylaxis, newborn screening, stroke prevention and hydroxyurea therapy – have been made. However, disease-modifying and curative treatments remain elusive.

We will review the pathogenesis of this complex disorder and the current management options, focusing primarily on the role of transfusion therapy.