

'Race', Health and Social Care

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NEW

# THE POLITICS OF SICKLE CELL AND THALASSAEMIA

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**"...a most interesting, eye-opening and often challenging book...it should be read by anyone involved in dealing with haemoglobinopathies, in the health or social care setting. Many others who are interested in the broader issues around chronic and particularly genetic disease will also find it stimulating reading."** - Dr Anne Yardumian, Consultant Haematologist, North Middlesex Hospital, London

Sickle cell disorders and thalassaemia are inherited blood disorders. Sickle cell disorder alters the shape of the red blood cells from their usual round appearance to something which resembles a sickle, or half moon. Those born with thalassaemia major are unable to make a sufficient amount of haemoglobin. They will develop a fatal anaemia in early childhood if not treated with blood transfusion every four to six weeks, for life. Sickle cell disorders and thalassaemia are found mainly in families that come from Africa, South Asia, the Caribbean, the Eastern Mediterranean and the Middle East. There are estimated to be over 10,000 people with a sickle cell disorder (SCD) and around 600 cases of thalassaemia in the UK. Despite this high incidence, it is still an under researched topic, and a subject about which health professionals and policy makers know very little. After years of neglect, it is now attracting policy interest and there are new moves to improve existing provision.

This timely book examines the politics of sickle cell and thalassaemia and offers a detailed evaluation of the services available. It is unusual in placing patients and their families at the centre of the study, allowing their views to be heard, and relating them to the delivery and organization of services. The problems that emerge range from inadequate language support, inappropriate generalizations, poor quality care, as well as institutional and individual racism. The book also identifies models of good practice and suggests ways in which we can learn from these. General policy and practice issues are highlighted throughout, and the need for a more systematic approach to planning and providing culturally sensitive services is addressed.

**Contents:** The politics of sickle cell disorders and thalassaemia - Origins, geographic distribution, genetics and laboratory investigations - Clinical features and management - Genetic screening and counselling: ethics, politics and practice - The experience of sickle cell and thalassaemia - Health and social care provision - Past achievements and future strategies - Bibliography - Index.

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Key Difference - Sickle Cell Anemia vs Thalassemia Thalassemia is a heterogeneous group of disorders caused by inherited mutations that decrease the synt. Presence of sickle cells and target cells in the blood film. Screening tests for sickling with chemicals such as dithionate are positive when the blood is deoxygenated. In HPLC, HbA is not detected. Sickle cell syndromes are hereditary hemoglobinopathies. Homozygous sickle cell anemia (HbSS, autosomal recessive) is the most common variant of the sickle cell. Other rare variants of sickle cell syndrome occur in individuals with one HbS allele and one other allele (HbC or Hb- $\beta^0$  thalassemia). A point mutation in the beta chain of hemoglobin leads to substitution of glutamic acid by valine, thus changing the structure (and properties) of hemoglobin. Abnormal hemoglobin polymerizes when deoxygenated, resulting in sickle-shaped erythrocytes, which cause vascular occlusion and ischemia. Sickle cell anemia manifests in early childhood with symptoms associated with vascular occlusion and hemolytic anemia. Start by marking "The Politics of Sickle Cell and Thalassemia" as Want to Read: Want to Read saving| Want to Read. Sickle cell disorder is the name for several related but different inherited disorders associated with the sickling of the red blood cell. Sickle cell disorder alters the shape of the red blood cells from their usual round appearance to something which resembles a sickle, or half moon. The different kinds of disorder are found mainly in people whose families come from Sickle cell disorder is the name for several related but different inherited disorders associated with the sickling of the red blood cell. Sickle cell disorder alters the shape of the red blood cells from their usual round appear