STANDARD TREATMENT WORKFLOW (STW)

Sickle Cell Disease

Tulika Seth, Dipti Jain, Manisha Madkaikar, N S Chatterjee, P K Patra, Roshan Colah

1 All India Institute of Medical Science, New Delhi; 2 Govt Medical College, Nagpur; 3 Indian Council of Medical Research-National Institute of Immunohaematology Mumbai; 4 Indian Council of Medical Research, New Delhi; 5 Chandulal Chandrakar Memorial Government Medical College; 6 Indian Council of Medical Research-National Institute of Immunohaematology Mumbai

CORRESPONDING AUTHOR
Dr Tulika Seth, Department of Haemolytic Anaemia, All India Institute of Medical Science, New Delhi
Email: drtulikaseth@gmail.com

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This family tree shows through mendelian transmission - the risk of having affected children across generations in parents with SCD - heterozygous and homozygous.

- Hemolytic anemia, where RBCs sickle under hypoxia or stress. Sickling and inflammation lead to vaso-occlusive crisis (VOC) and organ damage
- Autosomal recessive - mutations in the β-globin gene
  - ~86% of sickle homozygous cases in Asia are Indians
  - Carriers/ heterozygous (HbAS)
  - Have only one disease allele, usually asymptomatic

- Disease/ sufferers/ homozygous (HbSS)
- Have both defective alleles, usually symptomatic

- Other symptomatic genetic variants
  - HbS-β thalassemia, HbS-D Punjab disease, HbS-E disease etc.

- Manifestations of VOC
  - Experienced as pain, or swelling
  - Each VOC can lead to long lasting problems and end-organ damage
  - Typical sites - hands and feet, limbs, abdominal viscera, ribs, sternum etc.
  - The crisis is usually precipitated by fever, strenuous exercise, dehydration, drenching in rain, surgery, infection etc.