**Title of the Abstract:**
Clinical and demographic profile of patients with sickle cell disease: A single Centre experience from India

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**Aims and Objectives:**
The aim of this study was to analyze the demographic, clinical and laboratory profile of patients (adults and children) with sickle cell disease, and to assess the use and response to hydroxyurea in these patients.

**Patients and Methods:**
This study involved a retrospective analysis of sickle cell disease patients from January 2005 to December 2013 who were diagnosed and treated in Department of Haematology. The outpatient and inpatient records of the patients were analyzed in detail for their clinical presentation, demographic data, treatment details and their response rates to hydroxyurea were assessed. The change in frequency of painful crisis and need for blood transfusion with use hydroxyurea was documented. The surgical procedures done and complications due to surgery, if any, which happened during the study period were recorded. In laboratory parameters, the
change in mean corpuscular volume and HbF percentage with use of hydroxyurea was recorded.

Descriptive statistics were calculated for all variables

**Results:**

230 patients of sickle cell disease (148 homozygous sickle cell disease and 82 sickle beta thalassemia) were included in present study. The median age at first symptom was 4 years (range: 1-54) and majority of patients were from East India (57.3%). The bone pains and symptomatic anemia were most common manifestations present in 187 (81.3%) and 125 (54.5%) respectively. The disease related complications included acute chest syndrome in 14 (6.1%), avascular necrosis in 32 (13.9%), cholelithiasis in 14 (6.1%), leg ulcer 3 (1.3%), stroke in 2 (0.8%) and seizures in 2 (0.8%). Out of 230 patients, Hydroxyurea was used in 222 patients in median dose of 15mg/kg/day (range: 10-35mg/kg/day). Long term response assessment was done after median follow up of 36.25 months revealed complete resolution of symptoms in 57.4% of patients while 37.4% had at least 50% reduction in their presenting symptom.

**Conclusion:**

Bone pain and anemia are the most common manifestation in patients with sickle cell disease. Low dose hydroxyurea is safe and efficacious in ameliorating the disease manifestations in majority of the patients with sickle cell disease