CONCLUSION
Sickle Cell Disease is recognized as a major health problem among the young patients in this part of the country and is found in all castes and communities. The present study highlights the co-inheritance of α and β-thalassemia with βS gene and their influence on various clinical features and hematological indices in this disease. About 1/10th of this disorder are found to be Sβ+ thalassemia in Western Orissa. The molecular basis of Sβ-thalassemia in this population is IVS1-5 G→C mutation. In Sβ+ thalassemia the commonest clinical presentation is painful crisis followed by splenomegaly, anemia and multiple infections. Certain complications like avascular necrosis, leg ulcer and priapism are rare in our patients. The βS gene in Sβ+ thalassemia is linked to Asian haplotype whereas the βthal gene is linked to multiple haplotypes. This finding highlights the spread of β-thalassemia mutation from various parts of India. Influence of βthal haplotype upon clinical features of Sβ+ thalassemia necessitates further research in a larger sample size. The Sβ+ thalassemia type-I is observed to be severe than homozygous Sickle cell Disease. Like sickle gene the α-thalassemia deletion is equally prevalent in both tribal and non-tribal Hindus of Western Orissa. Both heterozygous and homozygous 3.7 α-thalassemia reduce the frequency of painful crisis in homozygous Sickle Cell Disease. Except the frequency of hospitalization there is no influence of α-thalassemia on the clinical features of Sβ+ thalassemia. In view of small no. of cases in this study the influence of α-thalassemia on Sβ+ thalassemia need to be studied in a larger population.