SUMMARY
1. Sixty cases of Sβ+ thalassemia confirmed by ARMS PCR were studied in detail.

2. The incidence of Sβ+ thalassemia was 9.6% amongst SCD patients. All cases had one β thalassemia mutation i.e. IVS1→5(G→C).

3. Sβ+ thalassemia cases were divided into two sub-groups based on HbA% level: type-I (HbA 3-5%) and type-II (HbA >5%).

4. Incidence of painful crisis was significantly higher in the Sβ+ thalassemia type-I than type-II (p<0.05) whereas other clinical features were found to be similar. All the hematological indices (total Hb, RBC count, MCV, MCH, MCHC, HbF%, and HbS%) were found to be similar in both groups except HbA2% i.e. significantly more in type-II in comparison to type-I (p<0.05).

5. The clinical features and hematological indices of the above 60 Sβ+ thalassemia cases were compared with 180 cases of age and sex matched SCD (homozygous) patients. Sβ+ thal type-I cases had severe clinical presentation whereas the type-II cases were comparable to SCD. Both the group Sβ+ thalassemia type-I and type-II had significantly less MCV, MCH and more HbA2 (p<0.05) in comparison to SCD. However the HbF % was significantly more (p<0.05) in SCD in comparison to Sβ+ thalassemia type-I and comparable to type-II.

6. In the Sβ+ thalassemia cases 96% βS chromosomes were found to be linked with Asian haplotype (++-+++-); whereas 4% were atypical. βTh chromosomes were linked to multiple haplotype (42% (+------), 36% (--------) and 12% (--------)).

7. Influence of different βTh haplotype (i.e. type-II, type-III) upon phenotypic manifestation was not observed in Sβ+ thalassemia. However significantly more HbA%, HbA2% and lower MCH, MCHC, was observed in the βTh haplotype type-II group (p<0.05) in comparison to βTh haplotype type-III.

8. In Sβ+ thalassemia type-I 11(26.8%) cases had heterozygous 3.7 α-thalassemia (αα/-α3.7) and a lone case (2.2%) had heterozygous 4.2 α-thalassemia (αα/-α4.2). In the Sβ+ thalassemia type-II 2 cases had (13.3%) heterozygous 3.7 α-thalassemia (αα/-α3.7) and a lone case (6%) had homozygous 3.7 α-thalassemia (-α 3.7/-α 3.7).
9. The clinical severity was similar in the Sβ+ thalassemia patients with and without heterozygous 3.7 α-thalassemia (αα/-α3.7).

10. Sβ+ thalassemia type-I with α 3.7 heterozygous α-thalassemia (αα/-α3.7) had significant increase in HbA2, Hbf and decrease of HbS (p<0.05) in comparison to Sβ+ thalassemia type-I without α-thalassemia (αα/αα).

11. Study of influence of α-thalassemia on SCD revealed that of the 267 SCD cases 199 (74.5%) had a normal α genotype (αα/αα), whereas 68 (25.5 %) had α-thal. Out of 68 cases of SCD with α-thalassemia, 57 (21%) had heterozygous 3.7 α-thalassemia, 3 (1%) had heterozygous 4.2 α-thalassemia and 8 (3%) had homozygous 3.7 α-thalassemia.

12. The clinical presentation in all the three groups of SCD with α-thalassemia (heterozygous 3.7 α-thalassemia, homozygous 3.7 α-thalassemia and heterozygous 4.2 α thalassemia) and without α-thalassemia were similar. However, the painful crisis (VOC > 2 ep/Yr) was significantly less ($\chi^2$-12.5, p< 0.006) in SCD with both heterozygous and homozygous 3.7 α-thalassemia (αα/-α 3.7 and -α 3.7 /-α 3.7) in comparison to SCD without α-thalassemia.

13. SCD patients with α thalassemia (heterozygous 3.7 α-thalassemia, homozygous 3.7 α-thalassemia and heterozygous 4.2 α thalassemia) had significantly lower MCV, MCH and higher RBC count (p < 0.05) in comparison to SCD without α thalassemia.