

Ali S., Kutty S., Kutty S. Recent observations on osteomyelitis in sickle-cell disease. *International Orthopaedics* 1985;9(2)


Ballas S.K., Gay R.N., Chehab F.F. Is HbA2 elevated in adults with sickle-$\alpha$-thalassemia($\beta^s$/ $\beta^s$;-$\alpha$/$\alpha$)?. *Hemoglobin.* 1997,21(5)405-20.


Diggs L.W. Siderofibrosis of the spleen in sickle cell anemia. *J. Am. Med Ass.* 1932;104:538-41


Hirst C., Owusu-Ofori S. Prophylactic antibiotics for preventing pneumococcal infection in children with sickle cell disease. Cochrane Database of Systematic Reviews 2002; 3.


Keclard L., Ollendorf V., Berchel C., Loret H., Merault G. \( \beta ^8 \) Haplotypes, \( \alpha \)-globin gene status, and hematological data of sickle cell disease patients in Guadeloupe (F.W.I). *Hemoglobin.* 1996;20:63-74.


Lewis S.M., Bain J.B. and Bates I. Dacie and Lewis Practical Haematology 2006, 10th edn, Churchill Livingstone Elesvier publications.


Livingstone, F.B. Abnormal Hemoglobin in Human populations. 1967, Aldine, Chicago.


Natta C. Failure of the α-thalassemia gene to decrease the severity of sickle cell anemia. *Blood* 1978; 51: 1163-8


Silverstroni E. & Bianco I. The distribution of the microcythaemias (or thalassemia) in Italy. Some aspects of the hematological and hemoglonobic picture in these hemopathies. In:


Syndenstricker, V.P. Further observations on sickle cell anemia. J. Am. Med. Ass. 1924;83:12-15.


Tuberculosis in adult patients with sickle cell disease. Journal of Infection, 2007;55(5): 439-44


Thesis Query by Dr. Ranjan Bhadra

primer for the FSC-8/9(+G) mutation in the study?

The said mutation FSC-8/9(+G) is studied in the thesis work named as cd 8/9(+G).

Hematological indices of type-I Sβ+ thal was comparatively same with type-II α2 content higher in type-II but the painful crisis was higher in type-I. What is the basis for such observation?

In the associated β-thal mutation, there are many other hematological and genetic Cmn-I polymorphism, FCP locus and underlined SNPs which could be the cause for variations but not evaluated. However the no. of cases were also less which could lead to technical findings.

Presence of α-thalassemia on Sβ+ thalassemia remains inconclusive.

Inheritance of α and β thalassemia with β8 gene leads to modification of α/β chain ratio influence the hematological indices and clinical features. Present study has shown a raised in the quantitative parameters HbF and HbA2 in the Sβ+ thal group having α chain deletion. But it couldn’t establish any role in the field of clinical features; it be due to small no. of patients and some undefined factors like FCP locus, SNPs which has not been taken into consideration.
1. **Standard error of mean (SEM) is not presented in the graph? Why?**

**Ans:** SEM gives an idea of accuracy of the mean whereas SD gives an idea of the variability of a single observation. SEM shown to indicate how accurate the mean is but in fact for descriptive statistics of subjects SD is needed to give the reader an idea of the spread between subjects instead of showing SEM. To indicate the superiority of SD over SEM, I am giving an example as follows.

HbCode 1: S Beta + (Hb 0-5%)

HbCode 2: S Beta + (Hb >5%)

<table>
<thead>
<tr>
<th>Group Statistics</th>
<th>HBCODE</th>
<th>N</th>
<th>Mean</th>
<th>Std. Deviation</th>
<th>Std. Error Mean</th>
<th>t-value</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hb</td>
<td>1</td>
<td>45</td>
<td>8.693</td>
<td>2.184</td>
<td>.326</td>
<td>0.06</td>
<td>0.9</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>15</td>
<td>8.567</td>
<td>2.935</td>
<td>.758</td>
<td></td>
<td></td>
</tr>
<tr>
<td>MCV</td>
<td>1</td>
<td>45</td>
<td>66.504</td>
<td>5.487</td>
<td>.818</td>
<td>0.7</td>
<td>0.3</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>15</td>
<td>69.340</td>
<td>4.286</td>
<td>1.107</td>
<td></td>
<td></td>
</tr>
<tr>
<td>MCH</td>
<td>1</td>
<td>45</td>
<td>22.847</td>
<td>2.934</td>
<td>.437</td>
<td>0.8</td>
<td>0.4</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>15</td>
<td>23.387</td>
<td>1.968</td>
<td>.508</td>
<td></td>
<td></td>
</tr>
<tr>
<td>MCHC</td>
<td>1</td>
<td>45</td>
<td>34.176</td>
<td>2.497</td>
<td>.372</td>
<td>0.1</td>
<td>0.8</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>15</td>
<td>33.673</td>
<td>3.116</td>
<td>.805</td>
<td></td>
<td></td>
</tr>
<tr>
<td>HBA2</td>
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<td>45</td>
<td>4.800</td>
<td>1.104</td>
<td>.165</td>
<td>2.485</td>
<td>0.01*</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>15</td>
<td>5.573</td>
<td>.815</td>
<td>.211</td>
<td></td>
<td></td>
</tr>
<tr>
<td>HBF</td>
<td>1</td>
<td>45</td>
<td>18.553</td>
<td>8.285</td>
<td>1.233</td>
<td>0.9</td>
<td>0.09</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>15</td>
<td>19.087</td>
<td>8.567</td>
<td>2.212</td>
<td></td>
<td></td>
</tr>
<tr>
<td>HBS</td>
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<td>45</td>
<td>72.171</td>
<td>7.330</td>
<td>1.093</td>
<td>0.7</td>
<td>0.4</td>
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<tr>
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<td>1.964</td>
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</tr>
</tbody>
</table>
It was observed from the above table (comparison of hematological indices in Sβ+ thalassemia type-I and type-II) that although there are comparatively more differences in SEM of HbF, HbS than HbA2, but we didn’t get any significant p value. Our study is a comparative study and statistical analysis centralized around value of significance; so we have accounted the SD and p-value.

2. The references are not properly and uniformly written?

Ans: The references are arranged alphabetically. So, there is discontinuity in the subsequent years.


3. More detail work in genetic pattern of inheritance in Sickle Cell Disease is needed. So, in this context what will be the future scope of the study?

Ans: More detail work is needed for

i) associated rare β-thalassemia
ii) haplotype of β-thal chromosome
iii) δβ-thalassemia
iv) associated recent genetic loci (SNPs polymorphism)

Future scope:

i) Prenatal diagnosis of pregnant mothers having hemoglobinopathies and genetic counseling
ii) Better management and prognosis of patients
iii) Drug designing