

The Effect of Hydroxyurea in Sickle Cell Disease Patients

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Abstract

Aim: To know the effect of hydroxyurea in sickle cell disease patients by measuring the HbF levels.

Materials and Methods: 50 patients suffering from sickle cell disease were taken from Prince Saud Bin Jalawy Hospital, Alahsa. The HbF levels were monitored in these patients before and after 4 months of administration of hydroxyurea. The HbF were monitored using Hb Electrophoresis.

Observation: The mean difference was – 0.3979 with SD of 0.0663, standard error 0.0094 and t value 0f-42.32 and p value of p<0.0001

Conclusion: In all patients there was 20-25% of improvement of fetal Hb after 4 months of treatment with hydroxyurea.

Keywords: Sickle cell disease HbF, Hydroxyurea

I. Introduction

Sickle cell disease is a chronic condition and a sinister haemoglobinopathy with serious consequences and high mortality¹.The median age survival of 14.3 yrs,with 20%deaths occurring in first 2 yrs of life,one third occurring before the fifth year of life,half between 5 and 30 yrs age and one sixth after the age of 30²It is caused due to substitution of glutamic acid by valine in the sixth position³. This results in the cell having sickle shaped cells with subsequent consequences of polymerization of RBC and finally sickle cell crises(pain,thrombosis,infection)^{4,5}.HbF is one of the types of hemoglobin seen in late foetal life⁶.Hydroxyurea is an anticancer drug which acts by neutralizing the tyrosyl free radicals and there by inhibiting the enzyme ribonucleotide reductase⁷.Electrophoresis is the separation of charged particles by administration of electric current⁸.

II. Materials And Methods

Fifty patients suffering from sickle cell disease from Prince Saud Bin Jalawy Hospital were taken.HbF levels were monitored before and after administration of hydroxyurea.The HbF levels were monitored using Hb Electrophoresis.All necessary norms were taken in informing the Ethics Committee.

Inclusion Criteria-

- 1.Age group 15-45 yrs including both male and female patients.
- 2.Sickle Cell Anaemia patients showing electrophoretic patterns of of HbS,F,A₂,and A only.

Exclusion Criteria-

- 1.AST >100 IU/L
- 2.Albumin<3gms/dl
- 3.Sr.Creatinine>2mg/dl

The HbF levels were monitored using Hb Electrophoresis.

Statistical Analysis-Statistical Analysis was done using paired 't' Test.

III. Results

D) Parameters of paired 't' test:

1. Sample size (n) = 50
2. Mean difference = -0.3979
3. Standard deviation = 0.0663
4. Standard Error = 0.0094
5. t value = -42.32
6. P value = P < 0.0001
7. Degrees of freedom = 49
8. 95 % confidence intervals = -0.416790 to - 0.379010

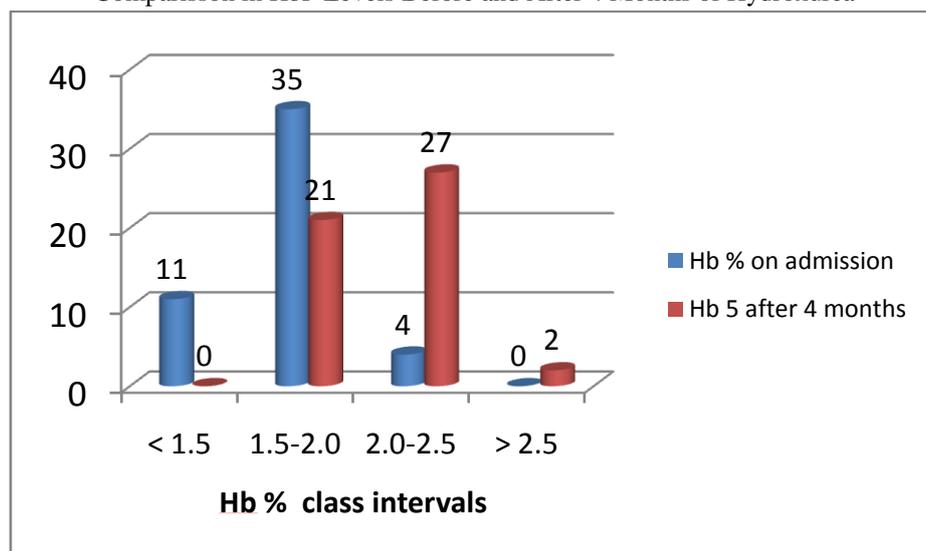
Table No. 1 Showing Hb % levels of Subjects on admission and after 4 months: (n=50)

Hb % Class Interval	Hb % On admission	Hb % After 4 months
1. 1.5	11 (22.00)	00 (0.00)
2. 1.5-2.0	35 (70.00)	21 (42.00)
3. 2.0-2.5	04 (08.00)	27 (54.00)
4. > 2.5	00 (0.00)	02 (04.00)
Total	50 (100.0)	50 (100.0)

- It is observed from the data that 48 % of beneficiary has shown increase of Hb % level 2.0-2.5 range and 4 % of them also shown increase of Hb % level of > 2.5.

Fig-1

Comparison in HbF Levels Before and After 4 Months of Hydroxurea



II) Parameters of paired 't' test: (Male beneficiary)

1. Sample size (n) = 30
2. Mean difference = -0.4087
3. Standard deviation = 0.075
4. Standard Error = 0.0137
5. t value = -29.73
6. P value = P < 0.0001
7. Degrees of freedom = 29
8. 95 % confidence intervals = -0.436720 to -0.380680

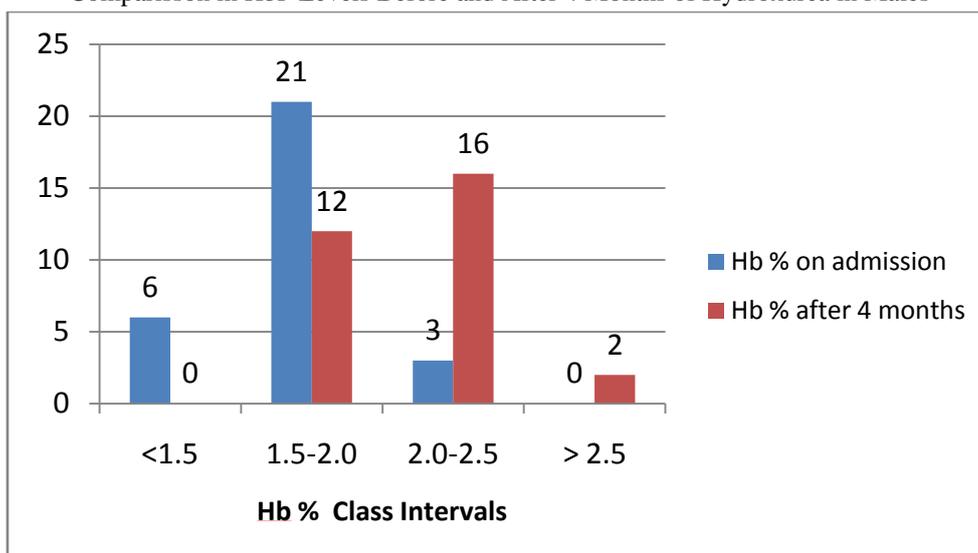
No 2 Distribution Hb % levels in male subjects (n = 30)

Hb % Class Interval	Hb % On admission	Hb % After 4 months
1. 1.5	06 (20.00)	00 (0.00)
2. 1.5-2.0	21 (70.00)	12 (40.00)
3. 2.0-2.5	03 (10.00)	16 (53.33)
4. > 2.5	00 (0.00)	02 (6.67)
Total	30 (100.0)	30 (100.0)

- It is observed that 43.33 % of benefices had shown increase in Hb % 2.0 – 2.5 group followed by 6.67 % increase in > 2.5 group.

Fig-2

Comparisson in HbF Levels Before and After 4 Months of Hydroxurea in Males



III) Parameters of paired 't' test: (Female beneficiary)

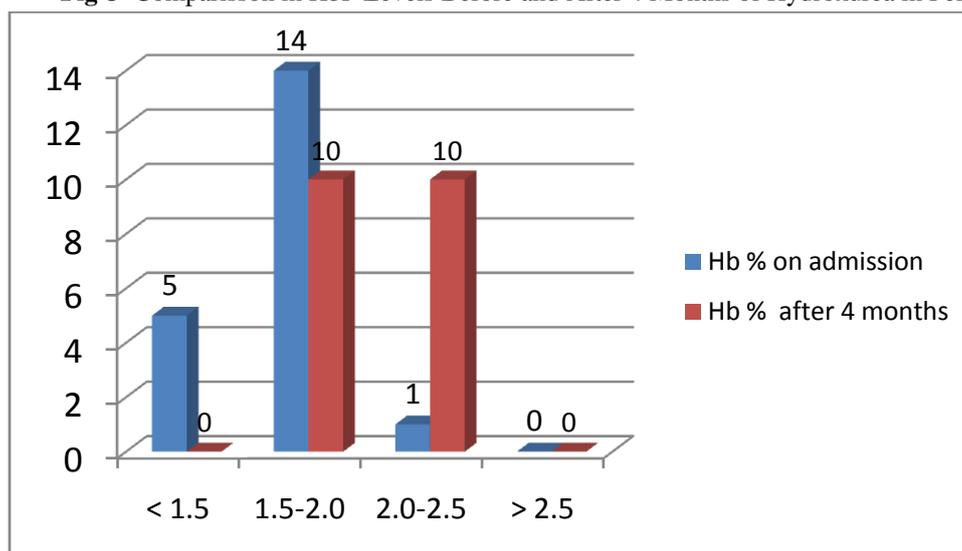
- Sample size (n) = 20
- Mean difference = -0.3825
- Standard deviation = 0.05199
- Standard Error = 0.011
- t value = -32.8806
- P value = P < 0.0001
- Degrees of freedom = 19
- 95 % confidence intervals = -0.406779 to -0.358221

Table No 3 Distribution Hb % levels in female subjects (n = 20)

Hb % Class Interval	Hb % On admission	Hb % After 4 months
5. 1.5	05 (25.00)	00 (0.00)
6. 1.5-2.0	14 (70.00)	10 (50.00)
7. 2.0-2.5	01 (05.00)	10 (50.00)
8. > 2.5	00 (0.00)	00 (0.00)
Total	20 (100.0)	20 (100.0)

- It is observed that 45 % of benefices had shown increase in Hb % 2.0 –2.5.

Fig-3 Comparison in HbF Levels Before and After 4 Months of Hydroxyurea in Females



IV. Observation And Discussion

The Mean difference in HbF was -0.3979 with SD of 0.0663 and p value ,0.0001. In males (n=30) the mean difference was -0.4087 with SD of 0.025 and p value <0.0001. In females the mean difference was -0.3825 with SD 0.05199 and p value <0.0001. There was a previous study by Veith et al⁹. A study by Timson revealed similar findings¹⁰. Charache S, Dover GJ et al published similar findings in their study¹¹. Rodgers et al in his study Hematologic responses of patients with sickle cell disease to treatment with hydroxyurea showed similar findings¹²

V. Conclusion

There is significant increase (20-25%) of HbF levels after administration of hydroxyurea in Sickle Cell Disease patients. There is significant decrease in mortality after usage of hydroxyurea in Sickle cell disease patients. There is overall improvement in well being of patients of sickle cell disease after usage of hydroxyurea.

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