GROWTH PROFILE OF CHILDREN WITH SICKLE CELL DISEASE

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ABSTRACT

Introduction: Sickle-cell disease (SCD) is a group of blood disorders which is inherited from the parents. It is known that the sickle cell neonates have a normal weight at birth. However, the disease with its attendant increased energy requirements has a negative effect on growth with a slow prepubertal growth and a delayed velocity compared with normal children. Previous studies of children with sickle cell disease (SCD) reported poor growth and delayed maturation. Physical growth is known to be impaired in children with sickle cell disease and the prevalence and severity vary with geographical location. The factors which contribute to this sub-optimal growth are poorly understood. This study was undertaken to provide additional data on the growth and maturation status of children with this disease.

Aim and Objectives: To study growth profile in children with sickle cell disease who are on treatment of hydroxyurea and children with Sickle Cell Disease.

Material and Methods: This study was a Prospective Observational study. This study was conducted in special clinic [haematology clinic], department of paediatrics, Pipriya From day of permission to one year. 50 patients were included in this study.

Result: A total of 50 children were included and their mean age was 10.7 years (range 5 years to 18 years). In our study BMI of children with sickle cell disease was between 5^{th} to 25^{th} centile, so this under-nutrition could be secondary to sickle cell disease. In our study overall growth of children with

sickle cell disease was below expected level.

Conclusion: It is necessary to monitor the growth in children with SCD to provide comprehensive care, facilitating early diagnosis of growth failure and nutritional intervention. Wasting and stunting are prevalent in children with SCA Underweight is more common compared to Stunting in SCA children. These results reflect the chronicity of SCA. Hydroxyurea treatment had no adverse effect on height or weight gain or pubertal development in school-aged children with SCA. Decreased growth velocity in children with SCA was independently associated with decreased haemoglobin concentration and increased total energy expenditure.

Key words: HU, SCD, growth, children and anthropometry.

INTRODUCTION

Sickle Cell Disease (SCD) is a hereditary and autosomal recessive genetic disease. The presence of abnormal crescent shaped RBCs instead of the regular biconcave disc shaped cells is the characteristic feature of SCD.(10) This results in an. abnormality in red blood cells of .the oxygen-carrying hemoglobin S. Under certain conditions, this. results in a rigid, sickle-like shape.

According to the recent survey by the Indian Council for. Medical Research (ICMR), the incidence of SCA is highest among the tribals in Gujarat. From the samples examined for the study of SCA in Gujarat, 34% sample were found to be positive. The study showed that 30% of children with SCD in Southern Gujarat died by the age of 14years. The characteristic features of. this disorder includes Anaemia, repeated infections and periodic episodes of pain. [1]

It occurs due to a substitution of valine for the normal glutamic acid in the β globin chain of haemoglobin. This cause changes in the haemoglobin. molecules so it crystallizes and deforms the red cell into a sickle shape when the Hb loses oxygen. [2]

The clinical presentation of SCA occurs due to increased blood viscosity, vascular obstruction by deformed sickled red cells. It is known that the sickle cell neonates have a normal. weight at birth. However, the disease with its attendant increased energy requirements has a negative effect on growth with a slow prepubertal. growth and a delayed velocity compared with normal children. [3]

Many factors as endocrine and/or metabolic dysfunction, haematological status, and. nutritional status may play an important role in growth failure. [4] The growth of the children with SCD is usually slow compared to their peers.

Advances in the clinical care of children with sickle cell anaemia (SCA), such as earlier diagnosis, penicillin prophylaxis, folate supplementation and hydroxyurea therapy have reduced. morbidity and mortality related to this disease.[5]

MATERIAL AND METHODS

STUDY DESIGN: Prospective Observational study.

STUDY SETTING: special clinic [haematology clinic], department of paediatrics, Pipriya.

SAMPLE SIZE: 50 patients to be monitored for growth with sickle cell disease.

Sample Size formula used is $n=4pq/l^2$

STUDY PERIOD: From day of permission to one year.

Where:

n = Sample size

p = prevalence (40%)

q = 100-p

l = error (20%)

INCLUSION CRITERIA:

1) Confirmed cases of sickle cell diseases.
2) Patients from 5-18 years of the age

EXCLUSION CRITERIA:

1) Patients suffering from SCD below 5 years.
2) Patients and parents who not giving informed consent.
3) Children with primary diagnosed endocrinopathy.
4) Children on any hormonal replacement therapy.
5) Children with chronic illness not related to SCD.

RESULT AND DISCUSSION

We found shows age wise distribution of sickle cell disease. In which 24(48%) patients were between 5-10 years of age, 26(52%) patients were between 10-18 years of age. The mean age of presentation in our department was 10.4 years for male patients and 11.5 years for female patients.

The reason behind this late presentation may be that they have taken the primary treatment at primary level before reporting to tertiary level hospital.

Jain S et al [7] showed that acute chest syndrome (ACS) is a frequent cause of acute lung disease in children with sickle cell disease (SCD). These include younger age, severe SCD genotypes (SS or $S\beta^0$ thalassemia), lower fetal haemoglobin concentrations, higher steady-state hemoglobin levels, higher steady-state white blood cell counts, history of asthma, and tobacco smoke exposure

We showed the gender wise distribution of patients. Out of 50 patients from age group of 5 – 18 years, males were 18(75%) and females 6(25%) and 10-18 years of age, males were 18(69.2%), whereas 08(30.8%) patients were females, p-value was 0.650 which was statistically not significant.

Wang CJ et al [6] observed that to develop a set of quality-of-care indicators for the management of children with sickle cell disease (SCD) who are cared for in a variety of settings by addressing the broad spectrum of complications relevant to their illness The panel identified 8 indicators most likely to

have a large positive effect on improving quality of life and/or health outcomes for children with SCD, which covered 6 topics: timely assessment and treatment of pain and fever; comprehensive planning; penicillin prophylaxis; transfusion; and the transition to adult care.

We suggested that distribution of history of prevalence of SCD in Family. Out of 50 patients, as the table / figure shows, 34% of patients (17) were having positive family history of SCD, while rest 66% of patients (33) were having negative family history of SCD. This is because of its autosomal recessive inheritance.

We shows variables at first visit of the patients with SCD. The mean age of presentation for male was 10.40(SD ± 3.46) years and female was 11.57years (SD ± 3.48). The total mean age of presentation was 10.73years (SD ± 3.47). Age of presentation were different in both sexes, so WFA and HFA were also different as per age of presentation.

The mean WFA of Male was 25.72kg (SD ± 9.98) and that of Female was 28.09 kg (SD ± 8.79) .The total WFA comes to 26.38 kgs (SD ± 9.64) kg.

The HFA (cm) was 129.40 cm (SD ± 17.87) in Male while that of Female was 134.81 cm (SD ± 12.88) and its mean total was 130.91 cm (SD ± 16.67).

BMI (kg/m2) of males was 14.82 kg/m2 (SD ± 1.66) and that of females was 15.09 kg/m2 (SD ± 2.00) while the total mean BMI was 14.89kg/m2 (SD ± 1.75).

We found depicts the anthropometric variables at last visit (5th visit). Mean WFA of male was 28.18 kgs (SD ± 10.10) and that of Females was 31.06 kgs (SD ±8.89) and the total was 28.98 kgs(SD ± 9.78).

The HFA of male was 136.06 cm (SD ± 17.10) and of females was 140.32 cm(SD ±10.81) and total mean was 137.25 cm(SD ± 15.61).

The mean BMI of male was 14.71 kg/m2 (SD ± 1.57) and of female was 15.52 kg/m2(SD ±2.31) while the total mean was 14.94 kg/m2(SD±1.82).

We showed at 1st visit mean weight of male was 25.72 kgs and female it was 28.09 kgs. Where mean weight of both sex was 26.38kgs.

At 2nd visit mean weight of male was 26.11 kgs and female it was 28.59 kgs. Where mean weight of both sex was 26.80kgs.

At 3rd visit mean weight of male was 26.7 kgs and female it was 29.17 kgs. Where mean weight of both sex was 27.39kgs.

At 4th visit mean weight of male was 27.35 kgs and female it was 30.07 kgs. Where mean weight of both sex was 28.11 kgs.

At 5th visit mean weight of male was 28.18 kgs and female it was 31.06 kgs. Whereas mean weight of both sex was 28.98kgs.

So, difference between mean weight at 1st visit and mean weight at last visit was 2.6 kg.

Sandell A et al [8] showed that Recently the United States has experienced an increase in refugees. Pediatric refugees are at risk for health and nutrition problems. Literature on longitudinal change in nutritional status of resettled pediatric refugees is scant. The cohorts A and B differed in country of origin

and infectious disease burden. On arrival, both cohorts exhibited HFA *z* scores reflecting short stature. BMI *z* scores were normal. HFA and BMI *z* scores increased during 5 years and 1 year for cohorts A and B, respectively. Anemia, vitamin D deficiency, and lead toxicity were identified. Resettled pediatric refugees were short. Some were stunted. Catch-up growth, however, occurred. There were increases in HFA and BMI *z* scores.

We showed the mean BMI (kg/m2) of the SCD patients.

At 1st visit BMI of male was 14.82 kg/m2 and that of female was 15.09 kg/m2 with the total mean of 14.89 kg/m2.

2nd visit BMI of males was 14.70 kg/m2 and of female were 15.04 kg/m2, their total of 14.79 kg/m2.

The BMI at 3rd visit of males was 14.74 kg/m2 and of females was 15.01 kg/m2 summing up mean is 14.82 kg/m2.

At 4th visit the mean of males was 14.73 kg/m2 while the mean of females was 15.19 kg/m2 and total was 14.86 kg/m2.

BMI of males being 14.71 kg/m2 and of females being 15.52 kg/m2 with total mean of 14.94 kg/m2 during the 5th visit.

BMI was between 5th to 25th centile in most of children of SCD. It might be because of hospital admission due to crisis secondary to disease itself.

Those with underweight were 8 (22.2 %) male and 2(14.3 %) female (<−2 S.D.) at 1st visit, 8(22.2%) in male and 2 (14.3%) in female at 2nd visit, 6 (16.7%) in male and 1 (7.1%) in female at 3rd visit, 9(25%) in male and 1(7.1%) in female at 4th visit, 2(5.6%) in male and 0 (0%) in female at last visit. Regular follow up, regular treatment can be result for improvement of weight.

Those with stunted were 6 (16.7 %) in male and 3(21.4 %) in female (<−2 S.D.) at 1st visit, 6(16.7%) in male and 2 (14.3%) in female at 2nd visit, 4 (11.1%) in male and 3 (21.4%) in female at 3rd visit, 11(30.6%) in male and 2(14.3%) in female at 4th visit, 4(11.1%) in male and 1 (7.1%) in female at last visit. . Regular follow up, regular treatment can be result of improvement of height.

Jain S et al [7] showed that acute chest syndrome (ACS) is a frequent cause of acute lung disease in children with sickle cell disease (SCD). Patients may present with ACS or may develop this complication during the course of a hospitalization for acute vaso-occlusive crises (VOC). ACS is associated with prolonged hospitalization, increased risk of respiratory failure, and the potential for developing chronic lung disease. ACS in SCD is defined as the presence of fever and/or new respiratory symptoms accompanied by the presence of a new pulmonary infiltrate on chest X-ray.

We found that the SMR staging at 1st visit and last visit of SCD children. During 1st visit, the normal SMR in males is 20 and that in female

is 9. SMR is delayed in both the sexes by 02. At last visit, normal SMR in males is 21 while that in females is 10 and the delay is 1 in both sexes.

We observed that doses of HU in SCD patients. At 1st visit dose of HU in males was 20.61 mg/kg/day and that of females was 21.83 mg/kg/day.at 2nd visit dose of HU of males was 20.67 mg/kg/day and of female was 21.71 mg/kg/day. The HU dose at 3rd visit of males was 20.83 mg/kg/day and of females was 21.14mg/kg/day. At 4th visit mean of dose of HU of males was 20.26 mg/kg/day while the mean of females was 20.81 mg/kg/day. HU mean dose at 5th visit of males being 20.01 mg/kg/day and of females being 20.88 mg/kg/day.

Thus, the minimum dose of HU both sexes was 20.01 mg/kg/day and maximum dose was 21.83 mg/kg/day. During entire study period dose of HU was relatively same. And there were no need to increase dose during study.

CONCLUSION

- The mean age of presentation in our department was 10.4 years for male patients and 11.5 years for female patients.
- In the age group of 5-10 years 18 (75%) were male and 6(25%) were female and in the age group of 10-18 years 18 (69.2%) were male and 8(30.8%) were female.
- 17(34%) patient had positive family history of sickle cell disease.
- The mean weight for age at first visit of male was 25.72 kg and that of female was 28.09 kg. The mean height for age at first visit of male was 129.40 cm and female was 134.91 cm. the mean BMI at first visit of male was 14.82 kg/m2 and female was 15.09 kg/m2.
- The mean weight for age at last visit of male was 28.18 kg and that of female was 31.06 kg. The mean height for age at last visit of male was 136.06 cm and female was 140.32 cm. the mean BMI at last visit of male was 14.71 kg/m2 and female was 15.52 kg/m2.
- The mean height for age of male at first visit was 129.40 cm and at last visit was 136.06 cm. The mean height of female at first visit was 134.81cm and last visit was 140.32cm.
- The mean BMI of male at first visit was 14.82 kg/m2 and at last visit was 14.71 kg/m2. The mean BMI of female at first visit was 15.09 kg/m2 and last visit was 15.52 kg/m2. In our study BMI of children with sickle cell disease was between 5th to 25th centile, so this under-nutrition could be secondary to sickle cell disease itself.
- Out of 50, the SMR staging at 1st visit, the normal SMR in males is 20 and that in female is 9, SMR is delayed in both the sexes by 02. At last visit, normal SMR in males is 21 while that in females is 10 and the delay is 1 in both sexes.

- Minimum dose of HU requirement in both male and female was 20 mg/kg/day while maximum dose was 21.83 mg/kg/day.
- For requirement of blood transfusion, out of 50 patients,
- 13(26%) patients required blood transfusion prior to HU treatment, while none required blood transfusion after regular HU treatment.
- In our study requirement of blood transfusion was significantly reduced in last visit compared to first visit. this might be due to regular HU treatment.

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TABLE

TABLE: MEAN WEIGHT FOR AGE OF DIFFERENT VISITS.

	Male		Female		Total	
	Mean	Std. Deviation	Mean	Std. Deviation	Mean	Std. Deviation
1_WFA (Kgs)	25.72	9.98	28.09	8.79	26.38	9.64
2_WFA (Kgs)	26.11	10.28	28.59	9.00	26.80	9.91
3_WFA (Kgs)	26.70	10.37	29.17	9.29	27.39	10.05
4_WFA (Kgs)	27.35	10.27	30.07	8.81	28.11	9.87
5_WFA (kgs)	28.18	10.10	31.06	8.89	28.98	9.78

TABLE: ANTHROPOMETRIC VARIABLES AT FIRST VISIT

Variables	Male		Female		Total	
	Mean	SD	Mean	SD	Mean	SD
Age (Yrs.)	10.40	3.46	11.57	3.48	10.73	3.47
1_WFA (Kgs)	25.72	9.98	28.09	8.79	26.38	9.64
1_HFA(cm)	129.40	17.87	134.81	12.88	130.91	16.67
1_BMI (Kg/m2)	14.82	1.66	15.09	2.00	14.89	1.75

TABLE: OBSERVATION OF HEIGHT FOR AGE AT DIFFERENT VISITS.

Sex	HFA (SDS)	First visit	Second visit	Third visit	Fourth visit	Fifth visit
Male (n=36)	Normal	30 (83.3%)	30 (83.3%)	32 (88.9%)	25 (69.4%)	32 (88.9%)
	Stunted	6 (16.7%)	6 (16.7%)	4 (11.1%)	11 (30.6%)	4 (11.1%)
Female (n=14)	Normal	11 (78.6%)	12 (85.7%)	11 (78.6%)	12 (85.7%)	13 (92.9%)
	Stunted	3 (21.4%)	2 (14.3%)	3 (21.4%)	2 (14.3%)	1 (7.1%)