JIHS The Journal of Integrated Health Sciences

Available online at www.jihs.in

Original Article

Sickle cell disorders in females: Screening of sickle hemoglobinopathy be part of Antenatal and Intensive care?

Jitendra D. Lakhani*, Varsha Shah, Dulari Gandhi, Niraj Pandit, Deep Mehta, Bakul Leuva, , Jasmin Jasani, Pawan Toshiniwal, Sucheta Lakhani, A Muley, Hemal Dave, Paresh Golwala, Dimple Thakore

Smt. B. K. Shah Medical Institute and Research Center & Dhiraj Hospital, Sumandeep Vidyapeeth, Piparia, Waghodia, Vadodara, Gujarat, India

ABSTRACT

Introduction: Women with sickle cell anemia may have complications especially during pregnancy. It may contribute to anemia in pregnancy.

Methodology: As a part of Sumandeep Vidyapeeth's Sickle Project, this observational study of profile of 125 female patients was done.

Observations: 125 female patients with sickle hemoglobinopathy who came either for antenatal checkup or for other health services in last 2 months were studied. 119 had sickle cell trait (SCT) while 6 (4.8%) had sickle cell disease (SCD). Majority of them were young, average age being 22.26 years. 14 (11+3, SCT and SCD) patients required indoor admissions. 3(SCT) patients were admitted in Critical Care Units, 7(6+1) in Obstetrics, 2(1+ 1) in medical and 2 (1+1) were admitted in pediatrics ward. Mean Hb of these 125 patients was 10.79 gm/dL, TC= 10702±3922/cu.mm, PCV of 32.70±7.98%, RBC count of 4.62±1.04 mil/uL, MCV=76.16±66.08fl, MCH=21.74±5.18pg, MCHC=28.35±8.74%, RDW-CV was 16.53±4.09. HPLC in percentage of A1a, A1b, A1c, LA1c and P3 were 0.96±0.72, 0.76±0.56, 4.90±0.81, 0.56±0.17 and 3.70±1.05 respectively. HBF=1.55±2.56, A0=57.34±11.69, A2=3.67±0.49 and S WINDOW was of 28.12±10.82.3. Out of 6 SCD patients,3 were admitted, 2 for severe Anemia and 1 for Crises. Mean Hb of SCD was 8.3±1.99 gm/dL, TC 15438±10562/cumm, PCV 27±5.75%, MCV 76.32±7.95fl, MCH=23.15±2.23pg, MCHC=30.37±1.75% RBC count of 3.60±1.04 Mil/uL, RDW-CV=21.73±6.99, HbS and HbF was 68.11±22.21% and 9.95 ± 6.3%.

Conclusions: Female patients coming from tribal area may need screening for sickling during antenatal checkup. SCT patients may also have morbidity and may require hospital admission.

Key Words: Sickle cell trait (SCT), Sickle cell disease (SCD), HPLC. (High Performance liquid chromatography). Tribal population

INTRODUCTION

Women with sickle cell anemia needs special attention as their profile of presentation and complications may be different. They may need different type of care in comparison with males. They pose special complications especially during pregnancy. It may result into multisystem

***Correspondence:**
E-mail: jitendralakhani@doctor.com

problems which may cause misery. About one third of pregnant women with sickle cell disorder have complications in form of neonatal death, abortion or stillbirth.[1] these complications may be related to Sickle cell Disease (SCD) (homozygous) patients. It is believed that Sickle cell trait (SCT) (heterozygous) pregnant women do not have complications during pregnancy however reports suggests that fetal loss may occur in trait patients also.[2] Urinary tract infections are also common in pregnant women having SCT.[3]

Neonatal screening is considered as a part of governmental health program in many countries, [4,5] however it has limitations.[6] To know natural history of local Sickle cell disorder, neonatal screening may prove beneficial.[7] It is also imperative for timely intervention with hydroxyurea therapy, vaccines and blood transfusions. Prevention as well as treatment of crises and multi organ failure is vital and they may need services for the same in form of indoor admission. Many of such patients need admission in critical care wards for infections, crises and multiorgan dysfunction.

To know what problem this sickle cell disorders female patients have and how useful screening in tribal population of our health seeking patients needs, this study was undertaken.

METHODOLOGY

As a part of Sumandeep Vidyapeeth's Sickle Project, this observational study aimed to find out profile of 125 female patients having sickle cell disorder (Sickle cell Disease or Sickle cell Trait) was done. These were the patients who came for getting health services from our institute. We also had objective to offer them need based and adequate health services including counseling and support services. We also had intention to follow them for pregnancy outcome and to go for neonatal screening. All female patients who came to seek health services in previous 2 months either for antenatal checkup or for other health services at Dhiraj Hospital (attached to SBKS Medical Institute and Research Centre) and who were either found to have sickle cell trait (SCT) or disease (SCD) were included. Screening was done by sickling solubility test and was confirmed by HPLC (High Performance liquid chromatography).

All hematological investigations like CBC, Blood Indices, RDW-CV, RBC count, Mentzer index and Srivastava Index[8,9] was taken. Discriminate Indices like Mentzer Index was calculated by MCV/RBC count and Srivastava Index by MCH/RBC count. Relevant biochemical, microbiological, imaging and pathological ancillary investigations needed by clinicians to treat/offer the services were also done.

RESULTS

Of 125 female patients, 119 had sickle cell trait (SCT) while 6 (4.8%) had sickle cell disease (SCD). Average age was 22.26±4.77 years. Majority (101 of 125) were in age group of 18 to 25, 18 were of 26-35 years while 6 were below 18 years. Of total 125 female patients, 14(11.2%) patients required indoor admission. Of this14 patients, 11 had SCT (9.24% of all trait) and 3 had SCD (50% of total 6). 7(6 SCT,1SCD) patients were admitted in Obstetrics ,3(all SCT) in CCUs, , 2(1 SCD,1 SCT) in medical and 2 (1 SCD,1 SCT) were admitted in pediatrics ward. (Figure 1) 3 out of 6 SCD were admitted, 2 for severe Anemia and 1 for Crises, while 3 patients of Sickle cell disease could be treated on outdoor basis.

Mean Hb of these 125 patients was 10.79 gm/dL, TC= 10702±3922, PCV of 32.70±7.98%, RBC count of 4.62±1.04Mil/uL, MCV=76.16±66.08fl, MCH=21.74±5.18pg, MCHC=28.35±8.74%, RDW-CV was 16.53±4.09.HPLC in percentage of A1a, A1b, A1c, LA1c and P3 were 0.96±0.72, 0.76±0.56, 4.90±0.81, 0.56±0.17 and 3.70±1.05

respectively.HBF=1.55±2.56, A0=57.34±11.69, A2=3.67±0.49 and S WINDOW was of 28.12±10.82.3 out of 6 SCD were admitted, 2 for severe Anemia and 1 for Crises. Mean Hb of SCD was 8.3±1.99 gm/dL, TC, 15438±10562, PCV 27±5.75%, MCV 76.32±7.95fl, MCH=23.15±2.23pg, MCHC=30.37±1.75% RBC count of 3.60±1.04 Mil/uL, RDW-CV=21.73±6.99, HbS and HbF was 68.11±22.21% and 9.95 ± 6.3%.

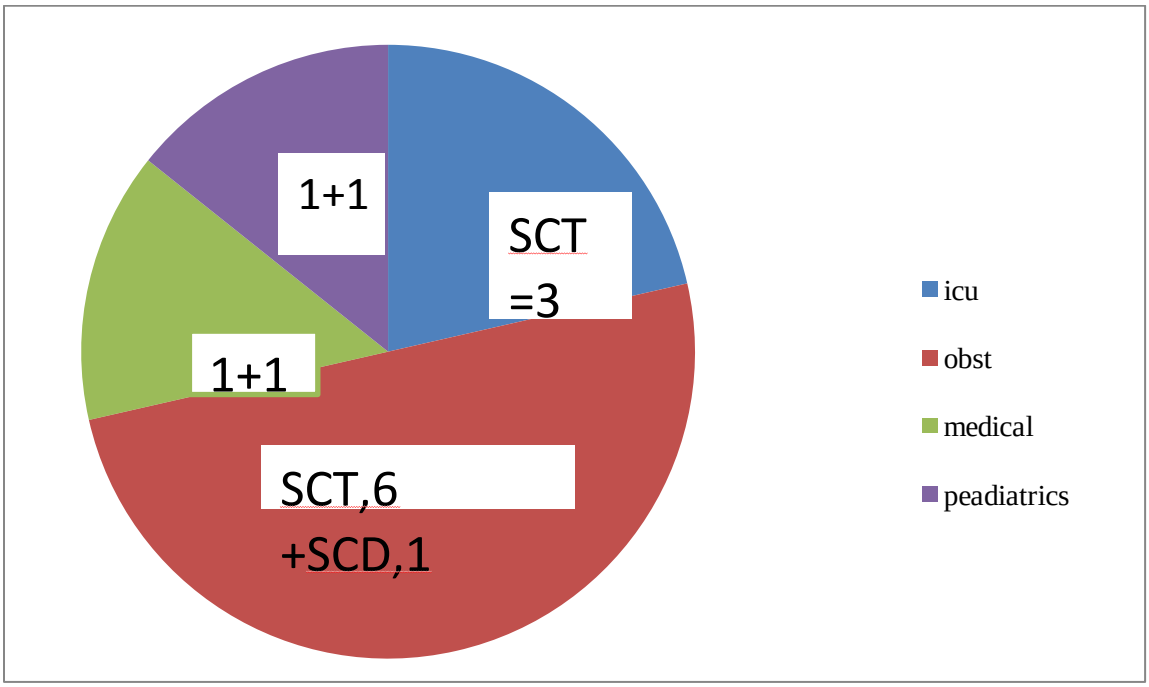

Figure 1: Indoor Admission (N=14) (11.2%)

Table 1: Hematological and HPLC profile of all female (N=125 having SCD & SCT) and of only Sickle cell Disease patients (n=06)

Lab. Parameter (first column)	Values of both SCD & SCT patients (N=125) (Second column)	Sickle cell disease patients (n=06) (Third column)
MEAN HB	10.79 ±6.66 gm/dl	8.3±1.99 gm/dl
TC	10702±3922	15438±10562 mm3
PCV	32.70±7.98%	27±5.75
RBC	4.62±1.04	3.60±1.04 millions/uL,
MCV	76.16±66.08	76.32±7.95fl
MCH	21.74±5.18	23.15±2.23pg
MCHC	28.35±8.74%	30.37±1.75%
RDW-CV	16.53±4.09	21.73±6.99
HbS	28.12±10.82	68.11±22.21