

Aawareness On Sickle Cell Disease (SCD) And Prevention Of Sickle Cell Crisis In Patients Of Sickle Cell Anemia. A Questionnaire Based Study.

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Abstract:

Aim:

To assess the knowledge and attitude of sickle cell disease patients and their practice in preventing sickle cell crisis.

Objective:

1.To create awareness, motivate, and impart health education and educate them about genetic/marriage counseling to affected families.

2. To evaluate outcome of periodic follow up, clinical management and intervention through local PHCs/Hospitals.

Materials and method:

It was a cross sectional study , conducted on 50 sickle cell disease patients attending the sickle cell clinic or admitted in AVBRH with sickle cell crisis, by giving them a self-administered questionnaire in local language. Study included patients from pediatrics, gynecology and medicine department. Thus, covering an age group from 3yrs – 33yrs.

Results:

A total of 50 sickle cell disease patients were surveyed in this study and only 32 patients (64%) were aware of being sickle cell positive, 14 (28%) of them knew how it is caused and 13 (26%) of them knew it is a hereditary disease, even though 70% of the patients were counseled about the disease. Only 8% of the patients underwent pre-marital counseling. Approximately 34% are availing treatment and adapting lifestyle modifications. Negative attitudes towards sickle cell disease were observed among 70-88% of patients.

Conclusions:

Despite fair knowledge of sickle cell anemia and disease in Vidarbha region, there is a need for improving interventional health educational programs regarding sickle cell, since negative attitudes and risky practices to sickle cell disease was documented. It was noticed that on creating awareness among the patients through this survey, a positive response was received from the patients, educating them about their disease, steps such as family screening, pre-marital screening and counseling and certain lifestyle modifications have been adapted by the patients.

Key words: *Sickle cell disease, sickle cell crisis, prevention, intervention, anemia*

Introduction

Hereditary hemoglobinopathies such as sickle cell disease (SCD) is wide spread among tribal communities in India. There have been difficulties for the prevention and control of hereditary hemolytic disorders in the high risk communities of India. The high prevalence of genetic and hemolytic defects and their cause of related morbidity, mortality and fetal wastage drastically affect the reproductive outcome⁽¹⁾. The cumulative gene frequency of haemoglobinopathies in India is 4.2%. With a population of over 1 billion and a birth rate of 28 per 1000, there are over

42 million carriers and over 12,000 infants are born each year with a major and clinical significant haemoglobinopathy⁽²⁻⁵⁾.

Diagnosis and management of these disorders both in adults and in newborns using appropriate approaches and uniform technology are important in different regions in India. For guidelines whom to screen, cost effectiveness technologies that need to be used as well as prenatal diagnosis should not be only given to medical fraternity professionals but those suffering should be counseled with regard to this. Apart from this there is an important need for the general

awareness of the community where hemoglobinopathies are endemic. Sickle cell anemia is endemic in vidharba region of central india. This study was carried out to assess the knowledge and attitude of sickle cell disease patients and their practices in preventing sickle cell crisis.

Aim: To assess the knowledge and attitude of sickle cell disease patients and their practices in preventing sickle cell crisis.

Objectives:

- To create awareness, motivate, and impart health education and educate them about genetic/marriage counseling to affected families.
- To evaluate outcome of periodic follow up, clinical management and intervention through local PHCs/hospitals.

Methodology:

Study setting: This prospective, cross sectional study was conducted in Jawaharlal Nehru Medical College, Acharya Vinobha Bhave Rural Hospital (A.V.B.R.H), a 900 bedded tertiary teaching hospital of Datta Meghe Institute of Medical Sciences University. Study was conducted over a period of 2 months, with due clearance of institutional ethical committee.

Participants and Sample size: The participants were 50 sickle cell disease patients attending the sickle cell clinic or admitted in AVBRH with sickle cell crisis, by giving them a self-administered questionnaire in local language.

प्रश्नावली/Questionnaire

नं.	वर्णन	होय	नाही
१.	काय तुम्हाला माहिती आहे कि तुम्हाला सिकलसेल अनेमिया आहे? Do you know you suffer from SCD?		
२.	तुम्हाला माहिती आहे कि सिकलसेल अनेमिया कश्यामुळे होतो Do you know the etiology of SCD?		
३.	तुम्हाला कळल्यानंतर डॉक्टर किंवा नर्सनि तुम्हाला सिकलसेल अनेमियाबद्दल काहीसांगितले का? After the diagnosis of the disease were you told or counselled about it from the nurse or the doctor?		
४.	सिकलसेल अनेमिया मुळे अंगात काय-काय होते हे तुम्हाला माहिती आहे काय? Do you know the different clinical presentations and which organs are affected in SCD ?		
५.	तुमच्याघरी कुणाला सिकलसेल अनेमिया आहे काय? आणि तुम्हाला माहिती होते काय कि सिकलसेल अनेमिया आईवडिलांना असलातर, मुला-मुलींना होऊ शकतो? Does anyone in your family have SCD? Do you know that children can get scd if		

Inclusion criteria:

- Sickle cell patients who came for regular follow up and /or admitted for sickle cell crisis.
- Patients willing to participate.

Exclusion criteria:

- Patients unwilling to participate.
- Patients with sickle cell trait, sickle cell thalassemia and other genetic hemoglobinopathies other than sickle cell disease.

Procedure/proposed intervention:

- All patients of SCD confirmed by hemoglobin (Hb) electrophoresis and those only willing to participate were included in this study.
- Patients were given a questionnaire in their local language, those unable to read or understand the questions were assisted by the nurse.
- For patients from pediatric department, those under the age of 10 years were assisted by their parents.
- Questions were direct, closed ended, framed in the simplest manner possible.

Statistical analysis

Once the target sample size was obtained, all the filled questionnaire were analyzed. Descriptive statistical analysis was done using frequency and percentages.

	their parents have it?		
६.	तुम्ही लग्नाच्याआधी सिकलसेलअनेमियाची टेस्ट केली होती काय? जरहो, तर तुम्हाला सि कल सेल अनेमियाबद्दल कुणी आणि कधी सांगितले होते? Did you do sickle cell anaemia test before marriage? if yes, who and when told you about SCD?		
७.	जरनाही, तर तुम्हाला माहिती होते, कितुमच्या नवरा/बायकोला सिकलसेल अनेमिया आहे? If no, did you know your spouse had SCD?		
८.	तुम्ही सिकल सेल अनेमियाकरता औषध घेत आहेका? जर नाही, तरकानाही,कत्यांचीकिंमतजास्तआहेका? Are you taking any medicines for SCD ? If no why not? Are they expensive?		
९.	डॉक्टरनि दिलेल्या औषधालासोडून, तुम्ही ठीक व्हयला अजूनकाहीकाळजीघेतआहेका? Apart from medicines are you taking any other care for health?		
१०.	जर हो, तर तुमची यापैकी काय-काय करता? If yes, what do you do out of following- जास्तपाणी/चहा-कॉफीघेणे. More of tea coffee कमीमीठखाणे. Less salt intake रोजफॉलिकअसिडच्यागोळ्याघेणे. Daily folic acid medicines intake जास्तव्यायामकरणे. Daily more exercise रोजनीटझोपणे. Daily adequate sleep सिगारेट/बिडीनपिणे. Smoking Cigarette /beedi रोजजास्तकामनकरणे. Avoiding excess work जास्तगरमवथंडवातावरणातनराहणे. Avoiding too cold and warm weather आजारपनापासूनवाचणे. Prevent diseases डोळ्यांचीतपासणीदरथोड्यादिवसांनीकरूनघेणे. Regular eye check up		
११.	जर तुम्ही बघितलेकि तुमच्या गावात किंवा घरात कोणाला तुमच्यासारखा त्रास होतो, तर तुम्ही त्यांना सिकलसेल अनेमियाबद्दल सांगून, तपासणी करून घ्याला सांगाल का? If you find someone having similar illness in your village will you counsel them for checking for SCD?		
१२.	तुमची सिकलसेल अनेमिया चीलस घेतली आहे का? Have you taken vaccines for SCD?		
१३.	तुम्हाला तुमच्या सिकलसेलअनेमिया बद्दल कळल्यानंतर, तुमची तुमच्या डॉक्टर/नर्सबरोबर सिकलसेल अनेमियाबद्दल बोलता का? After knowing that you are SCD do you ask for information about scd to your doctor or nurse?		
१४.	तुमची सिकलसेल अनेमियाच्या क्लिनिक/दवाखान्याला जाता का? Do you attend SCD clinic?		
१५.	तुमच्याकडे सिकलसेल/अनेमियाचा कार्ड आहे का? Do you have SCD card?		

१६.	तुम्ही स्वतः कधी सिकलसेल अनेमियाच्या क्लिनिकबद्दल कुणाला विचारल आहे का? आणि तुम्हाला माहिती आहे कि हे क्लिनिक कधी-कधी असतात? Have you ever enquired about SCD clinic? Do you know on which dates it run?		
१७.	तुम्हालामाहितीआहेकाकिसरकारनेसिकलसेलअनेमियाच्यारुग्णांच्यामदतीकरिताकाय-कायकेलेआहे? तुम्हालासरकारच्यासिकलसेलअनेमियाच्यायोजनाबद्दलमाहितीआहेका? Do you know what government has done for SCD patients? Do you know about national SCD programme?		

Observations and Results:

Figure 1.

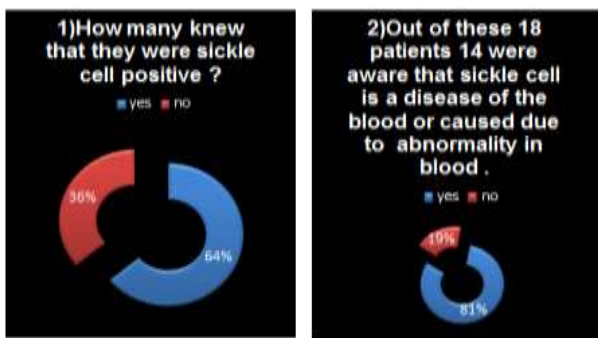


Figure 2: Knowledge of Sickle Cell Patients counseling of disease and Pre marital counselling.

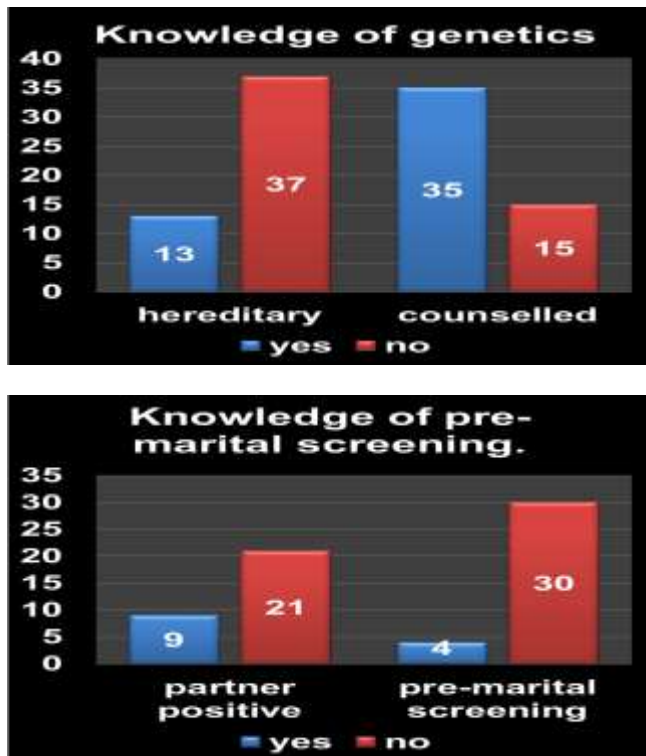


Figure 3: Compliance of Sickle Cell patients towards medical and conservative management.

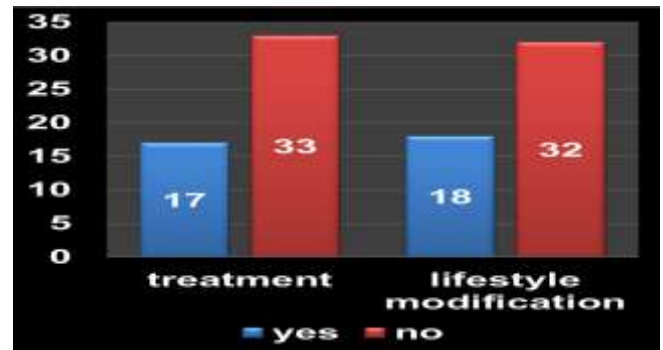
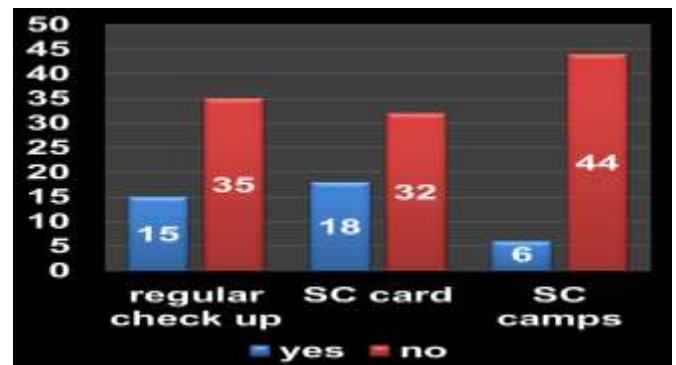


Figure 4: Attitude of Sickle Cell patients



Discussion:

Sickle cell disease (SCD) is one of the most common genetic diseases worldwide. It's a disease of national concern. The disease burden is enormous to the patient, family and community. One potential weapon in its prevention is health information and awareness among the populace.

In a study conducted about the awareness in SCD in Bahrain revealed that only 18 out of 50 patients knew that they were sickle cell positive, out of these 18, only 14 patients knew that SC is an anemic disorder or it is an abnormality in blood. It was seen that only 26% knew that SC is a hereditary disease; even though 70% of the patients were counseled after diagnosis.^[6]

In the year 2005-2006, Department of Health & Family Welfare of Government of Gujarat passed a resolution to initiate Sickle Cell Anaemia Control Program in the 4

districts of south Gujarat. As a part of strategy of Sickle Cell Anaemia Control Programme, mass Sickle Cell Screening was taken up by Government of Gujarat. ^[7]

Another study conducted in Gujarat state of India concluded that only 16% of the study participants knew correct symptoms of Sickle cell anaemia out of them only 30 % patients were availing the treatment of some kind. Females were more active as compared to males in taking medication for the same. It was found that 96 % of the study participants had received Color coded cards after testing showing the high accomplishment of mass screening programs in identifying Sickle Cell Status by simple means. Approximately 90% didn't know the cause of disease and only 18 % were counseled about this disease. More than 95% of the participants were unaware regarding their hemoglobin status. The study concluded that , all the strategies for the prevention of SCD will be effective only if they are utilized to its maximal extent by creating more awareness to the population affected. ^[8] A study had recommended that ideally a person suffering from SCD should test the hemoglobin levels checked every 3 months. ^[9]

In our study only 8% underwent pre-marital screening (even after having wide prevalence in their residential area) and 18% were aware of their partner being SC +ve.) Sixty six (66%) of the patients did not take the prescribed medicines on daily basis. Only 36% of the patients practiced lifestyle modifications and 22% had taken appropriate vaccinations.

Thirty six 36% of cases were registered- with a sickle cell card. After diagnosis and counseling only 28% came regularly for follow up. Tewelve 12% took an initiative to attend sickle cell camps and government initiated sickle cell programmes.

Awareness and health education SCD is the need of the time, as every 5th person in this population in endemic area turns out to be SC +ve, but unfortunately there is no initiative regarding screening procedure for the same. While educating the population regarding medical and conservative management, it should include lifestyle modification, since rural population is not able to keep up with the follow up medical therapy. As the disease entity has genetic predisposition, genetic and pre-marital counseling should be considered and initiated at primary care level. This will help in early identification of carriers of hemoglobinopathies before marriage, before conception or during the early antenatal period. Both outpatient and inpatient management for acute illness and complications should be available. A comprehensive health care program in schools for children and adolescents with SCD should include all systematic examination, hemoglobin analysis and

immunization as per recommended schedule. Facilities that should be made mandatory are;

Primary health care level: Screening HbS by Solubility test .

Rural Hospital Level: Complete blood count, Solubility test and Hb electrophoresis .

District Hospital Level: HPLC analysis, serum ferritin estimation, screening for HIV, G6PD deficiency, HCV and HBsAg, , Xray, blood transfusion and vaccination for pneumococcal, typhoid, Hepatitis B.

In tertiary level: Chorionic villous sampling and Aminocentesis.

Referral Level: for DNA sequencing and capillary electrophoresis.

While the basic genetic and diagnostic facilities should be available to deal with all aspects of prevention and care, the establishment of such facilities, if they do not already exist, may not require the sophistication and high costs that many people think. Action is, therefore, required to initiate activities to control genetic disorders in the country. The nature and sophistication of such activities will vary from one state to another, but the national programs should be established to provide basic services covering prevention, health promotion and case management activities.

To initiate interventions for the control of genetic disorders at the national level, the establishment of a vertical program for medical genetics is necessary. The strategies and public health approaches can be incorporated into the existing health care system. Integration into reproductive health program is probably the most appropriate way to achieve this objective.

Conclusions:

All the strategies for intervention will be effective only after creating awareness by health education, specific protection among the affected and vulnerable population. This also will make a facilitatory channel for early diagnosis and treatment and finally disability limitation that is preventing the patients from sickle cell crisis by educating them simple measures of prevention.

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