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Prevalence of Sickle Cell Anemia: A Study on Bastar Region

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Abstract

Sickle Cell Anemia is a complex inherited autosomal recessive Hemoglobin disease/disorder. It is not only generate the illness but also responsible for stigma in the society. Through the stigma the patients and their family members become more sufferers throughout their life. The mismanagement causes death before medication of the patients. So, there is a need of awareness about sickle cell anemia for various groups of the people like medical personal, researcher, social workers, psychologists, family members and patients. This paper reflects the sickle cell anemia history, origin and distribution in Bastar district of Chhattisgarh. This paper also highlights the knowledge and attitude of the patients with Sickle Cell Disease. The patients' level to policy level recommendations would be the vital for the need of the patients and their families as well as will create an approach to medical attention.

Keywords: Sickle Cell Anemia; Autosomal Recessive Disease/Disorder; Stigma.

Introduction

Sickle cell anemia is an inherited autosomal recessive disorder characterized primarily by chronic anemia and periodic episodes of pain. Sickle cell disease is a group of inherited conditions that affect hemoglobin, a protein that allows red blood cells to carry oxygen to all parts of the body. People with ancestry from parts of Africa, Asia and the Mediterranean are among the most likely to be born with these conditions. Healthy red blood cells are round, and they move through small blood vessels to carry oxygen to all parts of the body. In sickle cell disease, the red blood cells become hard and sticky and look like a C shaped farm tool called a "sickle." It is a tetramer consisting of two pairs of non-identical globin polypeptide chains, each chain being associated with one heme group. It changes its structure during human development. In the fetus, the major hemoglobin component is HbF ($\alpha 2\gamma 2$) while HbA ($\alpha 2\beta 2$) is a minor component. In adults, two components viz. HbA and HbA2 ($\alpha 2\delta 2$) exist. HbF is also present in small quantities. In all Hb components two a chains combine with two non a chains. Each of these chains is controlled by distinct genes. Genes for a chains are present on chromosome 16 while genes for all the non a chains are present on chromosome 11 in a specific manner collectively known as 'β globin cluster' (Weatherall & Clegg 2001).

Sickle gene is confined mainly to dravidians and pre-dravidians tribes inhabiting malaria endemic regions. It is also seen among certain caste groups from coastal areas of Orrisa and Andhra Pradesh. Sickle hemoglobin was first detected by Lehman and Cutbush in 1952 among the tribals from Nilgiris. More

than 7% of the world's population is carriers of some form of hemoglobin disorder. There are about 270 million carriers of sickle cell anemia and/or thalassemia (WHO, 1994). In India, sickle cell gene is mainly restricted to tribal and scheduled caste population where carrier frequencies range between 5-40% or more with three focal points (Bhatia and Rao, 1987). Based on the 1981 census and prevalence of Hb S in various populations studied, Rao (1988) estimated the expected number of sickle homozygotes as 1,31,375 in our country while expected number of sickle cell heterozygotes was 24,34,170. At that time the sickle cell gene was detected in 75 districts form various states. Kate (2000) compiled the data generated by various groups by screening various tribal populations from Maharashtra. This revealed that average prevalence of sickle cell carrier among the tribal population was 10% and that of homozygotes was 0.5%. Considering the tribal population of the state as 90 lakhs (Census, 1991), the expected carriers of sickle cell would be 9 lakhs and expected number of homozygotes would be 45000. Sickle gene is not seen frequently among the tribals of North East India with an exception to Panika from Mirajpur, Himalayan region as reported by Negi (1967). Sickle gene has also been reported among other caste groups (Balgir, 2005). In our country, tribals with sickle gene are mainly concentrated in Madhya Pradesh, Orissa, Chattisgarh, Jharkhand, Gujarat, Andhra Pradesh etc. Therefore as mentioned earlier, there are three focal points for sickle gene where this gene reached very high frequency.

History Sickle Cell Anaemia

The first report of sickle cell anaemia may have been in 1846 where the autopsy of an executed runaway slave was discussed. In 1910 a Chicago physician, James B. Herrick, reported the presence of sickle cells in the blood of an anaemic dental student, Walter Clement Noel. These cells had first been observed by his intern Ernest Irons while they were treating Noel in 1904.

An association with pigmented gall stones was noted in 1911 by Washborn. A genetic basis for this disease was proposed in 1915 by Cook and Meyer. The disease was named sickle cell anaemia in 1922 by Verne Mason after several additional cases were reported. All the known cases had been reported in blacks and he concluded that this disease was confined to those of black African descent. The heterozygous condition was independently recognised in 1923 by Huck and Syndestrickler. Syndestrickler also was the first to note the splenic atrophy that occurs in this condition. It was recognised as a Mendelian autosomal characteristic by Taliaffero and Huck also in 1923. A predisposition to pneumonia was noted in 1924 by Graham. The concept of progressive splenic atrophy was proposed by Hahn and Gilespie in 1927. Pneumococcal meningitis in this condition was first reported in 1928 by Wollstein and Kriedel but it was not until 1966 that the association between splenic atrophy and infection was made by Robinson and Watson.

Blood Transfusions Sickle Cell Disease (SCD) and Sickle Cell Trait (SCT)

Haemoglobin may fall 1–2 g/dl in an uncomplicated painful crisis, but blood transfusion is not routinely indicated. Blood transfusions should be used if the patient develops signs or symptoms which may be due to anaemia, including gunexplained tachycardia, tachypnoea, dyspnoea andfatigue. A low reticulocyte count (<100 * 109/l) and afalling haemoglobin make transfusion more appropriate. Typically, blood transfusion will not be necessary unless the haemoglobin has fallen more than 2 g/dl and is below 5 g/dl, and should aim to return the haemoglobin to the steady state level. Blood should be leucocyte depleted an

Environmental Factors

The importance of environmental factors in the outcome of sickle cell disease is undoubted, whether it relates to climatic factors or skin cooling precipitating bone pain crises or apparently random events such as contact with *Streptococcus pneumonia* or parvovirus *B19.* Malaria continues to be a major determinant of morbidity and mortality in affected areas such as sub-Saharan Africa and central India. Susceptibility to other potentially serious infections by *Salmonella* spp. may reflect carriage rates in the general population. Other important factors are nutrition, access to public health measures such as immunization, and socio-economic status, which may determine the families' access to communication, transport, and medical care. The importance of environmental influences and their mechanisms hold great potential for increasing the understanding and further amelioration of sickle cell disease.

Genetic versus Environmental Factors Modifying SS Disease

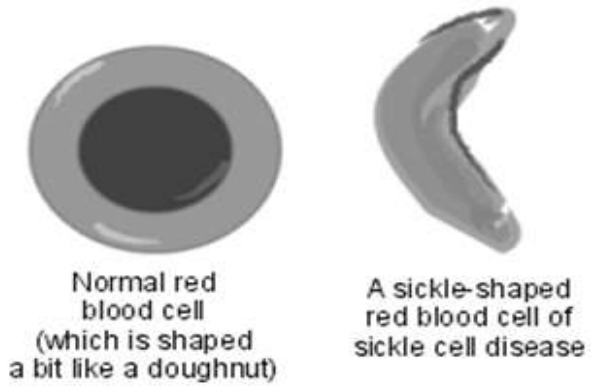
One of the classic methods for distinguishing genetic from environmental influences is the study of identical twins. Identical or monozygotic twins

occur in 0.3 percent –0.4 percent of births, but to be useful for studies of hematology and clinical features, both twins must survive long enough to provide adequate data. These conditions are not frequently met and the largest available series contained six pairs of identical twins in Jamaica (Weatherall, et al, 2005). A comparison of twin pairs showed that genetic factors influenced growth and hematological indices but those clinical events in the twin pairs were frequently discordant, suggesting the importance of nongenetic influences.

Methodology

Need for the Study

Methodology in major sense refers to the methods techniques or tools employed to the collection and the processing of data. Verbal and mechanical procedures are used in the process of data collection and analysis. Methodology is used again to designate the conception and procedure complied in the analysis of data however, collected to arrive at conclusions. Sickle cell disease (SCD) is a serious, inherited condition affecting the blood and various organs in the body. It affects the red blood cells, causing episodes of sickling, which produce episodes of pain and other symptoms. Sickle cell disease (SCD) is a serious group of conditions which are genetic (inherited). It affects the red blood cells in the blood. The following picture depicts what sickle cell is; red blood cells are flexible so that they can move through the smallest blood vessels. In sickle cell anemia, the hemoglobin is abnormal, causing the red blood cells to be rigid and shaped like a "C" or sickle, the shape from which the disease takes its name. Sickle cells can get stuck and block blood flow, causing pain and infections. Complications of sickle cell anemia are a result of sickle cells blocking blood flow to specific organs, and include stroke, acute chest syndrome (a condition that lowers the level of oxygen in the blood), organ damage, other disabilities, and in some cases premature death. Most of the tribal people genetically inherit this disease and suffers very hard.



Sample Design

For the purpose of the study the investigator had chosen the Government Maharani Hospital, Jagdalpur and with the support of the medical officer the data was collected. For the purpose of the study the investigator collected data from 133 patients with Sickle Cell Disease (SCD).

Objectives of the Study

1. To find out the knowledge and attitude of the patients with Sickle Cell Disease (SCD).
2. To assess the problems facing by the patients
3. To explore how the Sickle Cell Disease (SCD) is affecting their income
4. To study the stigma in and around the family
5. To study the health services for Sickle Cell Disease (SCD) and lastly,
6. To give suggestions for policy making.

Tools of Data Collection

The study is designed to explore the situation of the Sickle Cell Disease (SCD) patient in different angle like socio-economic, stigma and other physiological problems. The data was collected from the Government Maharani Hospital, Jagdalpur with the support of the Medical Officer. The following tools are used for the data collection from the patients with Sickle Cell Disease. A structured interview schedule was prepared for the respondents. Through this tool the respondents were elucidated their feelings about the Sickle Cell Disease, affects and other determinants were uttered by the respondents in a systematic and scientific manner. The investigator conducted focused group discussion with the Sickle Cell Disease patients in the hospital premises. The patients shared their physiological problems, social problems and how the disease effecting their economic conditions flatly. The responses that were given by the respondents were observed and their body language, family environment and the surroundings were observed to interlink the information to that of actuality. The investigator had to spend 1 month to collect and study the review of literature. Construction of schedule and data collection took 2 month. Finally the study period has consumed 6 months. After collection of data through the primary

source it has been coded and a code book was prepared. The data were entered into a master chart very meticulously. Thereafter, it was processed into the computer through MS Excel package. Later, the computerized data was taken in print form and the same was cross checked with the master chart to find out error(s), if any.

Scientific Test of Sickle Cell Anemia

Collection of Blood Sample

2 ml. Intravenous bloods were collected through disposable syringes in EDTA bulb. 2 ml. bloods were utilized for haemoglobin determination, sickling and for electrophoresis. The researcher was adequately traned by the staff at Maharani Hospital who is routinely undertaking test for sickle cell hemoglobin in Bastar district. A total of 133 blood samples comprising 78 males and 55 females were collected for the analysis for sickle cell hemoglobin and haemoglobin determination.

Solubility Test for the Presence of HbS

Principle

This is a rapid method for detection of the presence of HbS. HbS has decreased solubility in a deoxygenated state in hypertonic buffer, which produces turbidity.

Reagents

Phosphate Buffer (pH7.1)

Stock solution – Potassium dehydrogenate phosphate (KH_2PO_4) - 125 gms

Dipotassium hydrogen phosphate (K_2HPO_4) - 217 gms

Saponin - 2.5 gms

Distilled water - 1 lit.

To this is added a pinch of benzoic acid.

Working Solution

To 10.0 ml. of this stock solution, 0.1 gm of sodium dithionate is added immediately prier to use.

Procedure

20 µl of red cells washed with normal saline are added in 2 ml of the working phosphate buffer and mixed well to give a light pinkish violet colour. The results are taken after 10 minutes.

The test read as positive if the turbidity impaired the visibility of a dark black line on a white paper held against a bright source of light at a distance of few inches. A negative test is indicated by a clear pinkish violet solution through which the dark line easily seen.

Out of 133 individuals on whom solubility test was conducted, 35 were found to be positive i.e.35 individuals were found to be ticklers. Thus, 26.31 percent of the total individuals tested were ticklers.

Cellulose Acetate Electrophoresis

Blood sample of these 61 individuals were then subjected to further analysis on electrophoresis to identify different types of hemoglobin, which separate due to charge differences caused by structural variations. In this regard cellulose acetate electrophoresis procedure was undertaken to identify the different pattern types. At pH 8.9 hemoglobin is a negatively charged protein and migrates towards anode in an electrical field. Was used to determine the presence of AA, AS, and SS in the samples and to confirm the results generated by the above methods. The principle of this method was based on the fact that proteins normally have either positive or negative charge that is determined by the charged amino acid they contain. When electric field is applied to a solution containing protein molecules, positively charged proteins will move to the cathode and negatively charged proteins will migrate to the anode. Depending on their charges, size and shape, different haemoglobins will separate and migrate at different rates. They are then stained and their bands compared with the known controls.

Reagents 1

1. Tries EDTA borate buffer pH - 8.9
   - Tries - 14.4 gm
   - EDTA (disodium) - 1.5 gm
   - Boric acid - 0.9 gm
   - Distilled water - 1 lit.
2. Cellulose acetate strips

Procedure

- Soak the strips in TEB buffer for 30 minutes.
- Remove excess buffer by blotting between what man no. 1 filter papers.
- Place the strips across the bridge of the horizontal electrophoresis chamber.

- Place the strips using a double layer of what man no. 1 filter paper as wicks. These wicks dip in the anodal and cathode buffer compartments of the chamber.
- Heamolysate (is prepared by using CCI_4) is applied at the cathode end of the strip using a fine tipped paint brush.

Electrophoresis is run at a constant voltage of 200 to 250 volts (depend on no. of sample) for about 30-40 minutes or until one gets a fine separation of bands.

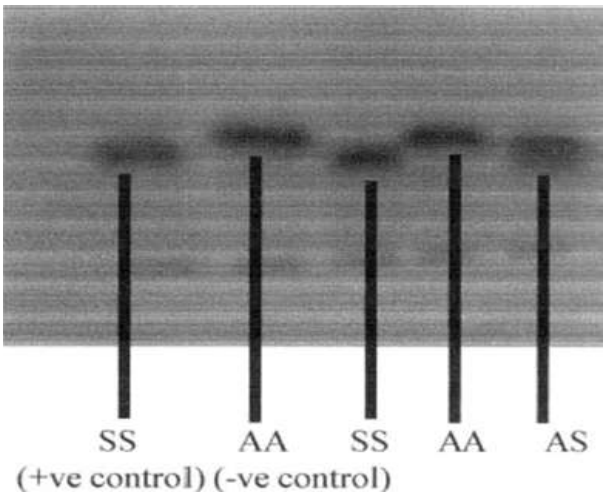


Fig. 1: Hb electrophoresis bands

- Bands are cut and eluted in distilled water.
- The eluted haemoglobin is allowed to stand at room temperature for 15-20 minutes and the bands are read using distilled water as blank.
- Normal hemoglobin has a single band.
- Heterozygote's (HbAS) have two bands.
- Homozygous (HbSS) has a single band.

Limitations of the Study

The research had some difficulties in getting support by patients with Sickle Cell Disease during collection of the data. The patients usually face psychosocial problems apart from physiological problems. Their attitude towards the disease cannot let them open their mouth. However, the investigator used different types of skills to collect the data. This challenge was overcome by changing the data collection approach. In addition individual efforts were made to collect extra information apart from the questionnaire. The data were collected with pre-tested schedule/questionnaires. The study has some limitations such as-

- Data collected is based on patient's past and present memory. This can lead to data inaccuracy. Efforts were made by the researcher to crosscheck to make data reliable and accurate.
- Since the study pertaining to a particular location, it cannot be generalized and implied to other locations.
- A majority of the respondents were illiterate it affected the data collection process.
- The respondents may have poor understanding on the Sickle Cell Disease.

Result and Discussion

If $f(AA)$, $f(Aa)$ $f(aa)$ and are the frequencies of the three genotypes at a locus with two alleles, then the frequency p of the A-allele and the frequency q of the a-allele are obtained by counting alleles. Because each homozygote AA consists only of A-alleles, and because half of the alleles of each heterozygote Aa are A-alleles, the total frequency p of A-alleles in the population is calculated as

$$p = f(AA) + \frac{1}{2} f(Aa) = frequency\ of\ A$$

Similarly, the frequency q of the a-allele is given by

$$q = f(aa) + \frac{1}{2} f(Aa) = frequency\ of\ a$$

It would be expected that p and q sum to 1, since they are the frequencies of the only two alleles present. Indeed they do:

$$p + q = f(AA) + f(\text{aa}) + f(\text{Aa}) = 1$$

and from this we get:

q = 1-p and p = 1-q

If there are more than two different allelic forms, the frequency for each allele is simply the frequency of its homozygote plus half the sum of the frequencies for all the heterozygotes in which it appears. Allele frequency can always be calculated from genotype frequency, whereas the reverse requires that the Hardy Weinberg conditions of random mating apply. This is partly due to the *three* genotype frequencies and the *two* allele frequencies. It is easier to reduce from three to two.

An example population

Consider a population of ten individuals and a given locus with two possible alleles, *A* and *a*. Suppose that the genotypes of the individuals are as follows:

AA, *Aa*, *AA*, *aa*, *Aa*, *AA*, *AA*, *Aa*, *Aa*, and *AA*

Then the allele frequencies of allele *A* and allele *a* are:

$$p = prob_A = \frac{2+1+2+0+1+2+2+1+1+2}{2\ 10} = 0.7$$

$$q = prob_a = \frac{0+1+0+2+1+0+0+1+1+0}{2\ 10} = 0.3$$

so if a locus is chosen at random there is a 70% chance it will be the *A* allele, and a 30% chance it will be the *a* allele

Results

$p^2 + 2pq + q^2 = 1$ *and* $p + q = 1$

p= frequency of the dominant allele in the population

q = frequency of the recessive allele in the population

p^2= percentage of homozygous dominant individuals

q^2= percentage of homozygous recessive individuals

2pq = percentage of heterozygous individuals

The frequency of AA is equal to p^2

The frequency of SS is q^2

AS = 2pq

p^2 = 73.68

73.68/ 100 =0.7368

p^2= 0.7368

p = 0.8583

q^2 = 3.01

3.01/100 = 0.0301

q^2= 0.0301

q = 0. 1735 (1.000 – 0.858 = 0.142 = q)

Out of 35 samples of ticklers only 3 were found to be homozygous SS and rest AS heterozygous. The results are presented in the table 6.1.

It can be seen that gene S occurs with a frequency of 73 percent and which is appreciably high. Presence of sickle cell anemia elevates the suffering, who suffers from Jaundice and diseases of blood and blood forming organs in varying proportion.

Caste Wise Suspected Cases Identification

Table 2 depicted that there was highly percentage of SC population (31.58 percent) who were suffering from sickling. Its observe that 29.82 percent of OBC population in study village was suspected and 27.82 percent in ST, 11.28 percent in General category were suffering from sickle cell anemia. It's clear from the table that where there is marriage found in close relationship in where the genetic disorder as like sickle is frequency distributed.

Identification of Sickle Cell Anemia

It reveals from table 3 that identification of sickle cell anemia was done by blood investigation (69.17 percent) and clinical checkup (18.80 percent) and both type of investigation was 12.03 percent.

Table 1: Analysis of Hemoglobin Types

Genotype	Number	Percentage	Allele	Allele frequency
AA	98	73.68	A	0.858
AS	31	23.31		
SS	4	3.01	S	0.142
Total	133	100.00	Total	1.00

Table 2: Caste Wise Suspected Cases Identification

Caste	Number	Percentage
SC	42	31.58
ST	37	27.82
OBC	39	29.32
General	15	11.28
Total	133	100.00

Table 3: Identification of Sickle Cell Anemia

Diagnosis	Number	Percentage
Blood Investigation	92	69.17
Clinical Checkup	25	18.80
Both	16	12.03
Total	133	100

Family Member Suffer by Sickle Cell Anemia

Table 4 shows that out of 133 respondents 73.68 percent family members were suffering from sickling and 26.32 percent respondents were not suffering.

Diagnosis of Sickle Cell Anemia

It is observed from the table 5 that highly percentage of respondents (60.00 percent) was diagnosed by doctor and 11.43 percent of patients diagnosed by laboratory technicians. Its important observation that 17.14 percent respondents were observed by both doctor and traditional header. Only 2.86 percent and 5.71 percent of patients diagnosed by health worker + others and traditional headers respectively.

Status of Sickle Cell Anemia

Table 6 shows that status of sickle cell anemia in study area where 72 percent patients were in all age group, 16.00 percent were in child group, 8.00 percent adult and 4.00 percent were in older group.

Medicine Used to Prevention and Treatment of Sickle Cell Anemia

Its observe from table 7 that 56.00 percent respondents were using allopathic medicine, 24.00 percent using ayurvedic and 8.00 percent respondents were using herbal medicine. A very few percentage of respondents were using 4.00 percent homeopathic, 4.00 percent unani and 4.00 percent by others kind of treatments.

Status of Blood Transfusion among Sickle Cell Anemia Patients

Table 8 shows the status of blood transfusion among sickle cell anemia patients i.e. more than two of 24.00 percent and 76.00 percent not received blood transfusion.There were absent of highly risk of blood crises among patients.

Table 4: Family Member Suffer by Sickle Cell Anemia

Responses	Number	Percentage
Yes	98	73.68
No	35	26.32
Total	133	100.00

Table 5: Diagnosis of Sickle Cell Anemia

Diagnosis	Number	Percentage
Doctor	21	60.00
Laboratory Technician	4	11.43
Health Worker	1	2.86
Traditional Healers	2	5.71
Doctor and Traditional Healar	6	17.14
Others	1	2.86
Total	35	100

Table 6: Status of Sickle Cell Anemia

Status	Number	Percentage
Child	4	16.00
Adult	2	8.00
Older	1	4.00
All age group	18	72.00
Total	25	100.00

Table 7: Medicine Used to Prevention and Treatment of Sickle Cell Anemia

Consulted	Number	Percentage
Allopathic	14	56.00
Homeopathic	1	4.00
Ayurvedic	6	24.00
Unanani	1	4.00
Herbal Medicine	2	8.00
Other	1	4.00
Total	25	100.00

Knowledge about Sickle Cell Anemia

Table 9 depict that 58.65 percent respondents have knowledge about sickle cell anemia but out of 133 respondents 41.35 percent have no knowledge about this disease.

Sources of Information

Table 10 shows that information regarding sources is maximum 39.1 percent received from health workers and minimum 3.01 percent from radio. Information received from TV is 6.02 percent, News paper is 9.02 percent, Medical doctor is 28.57 percent, pamphlet, leaflet, poster is 10.53 percent and others are 3.76 percent.

Perception of Respondents on the Occurrence of Sickle Cell Ddiaease

Table 11 shows that the perception of respondents on the occurrence of sickle cell disease are from God gifted is 15.79 percent, evil effect is 13.53 percent, Nutritional deficiency is 22.56 percent deed of last laif is 7.52 percent, genetic is 28.57 percent and others is 12.03 percent.

Perception of Respondents about Reason of Sickle Cell Anemia

Table 12 shows that the perception of respondents about reason of sickle cell anemia among patients, high percentage of respondents (48.00 percent) believe that they affected by FF & FM. Only 12.00 percent and 24.00 percent respondents agreed that they affected by Father and Mother respectively and rest 12.00 percent affected by bith Mother and Father.

Perception about Prevention and Treatment of Sickle Cell Anemia

According to table 13, perception accepted about prevention and treatment of sickle cell anemia by respondents is 76.69 percent and 23.31 percent are not agreed.

Table 8: Status of Blood Transfusion Among Sickle Cell Anemia Patients

Status of Blood Transfusion	Number	Percentage
15 days	0	0.00
One months	0	0.00
Two months	0	0.00
More than two	6	24.00
None	19	76.00
Total	25	100.00

Table 9: Knowledge About Sickle Cell Anemia

Responses	Number	Percentage
Yes	78	58.65
No	55	41.35
Total	133	100.00

Table 10: Sources of Information

Source	Number	Percentage
Redio	4	3.01
TV	8	6.02
Newspaper	12	9.02
Medical Doctor	38	28.57
Health Worker	52	39.10
Pamplet, Leaflet, Poster	14	10.53
Other	5	3.76
Total	133	100.00

Table 11: Perception of Respondents on the Occurance of Sickle Cell Ddiaease

Causes	Number	Percentage
God Gifted	21	15.79
Evil Effect	18	13.53
Nutritional deficiency	30	22.56
Deed of last laif	10	7.52
Genetic	38	28.57
Other	16	12.03
Total	133	100.00

Table 12: Perception of Respondents about Reason of Sickle Cell Anemia

Variables	Number	Percentage
Father Affected	3	12.00
Mother Affected	6	24.00
Both FM affected	3	12.00
FF affected	1	4.00
FM affected	0	0.00
Both FF & FM affected	0	0.00
MF affected	0	0.00
MM affected	0	0.00
Both MF & MM affected	0	0.00
None of these	0	0.00
FF & FM affected	12	48.00
Total	25	100.00

Table 13: Perception about Prevention and Treatment of Sickle Cell Anemia

Responses	Number	Percentage
Yes	102	76.69
No	31	23.31
Total	133	100.00

Table 14: Perception of Respondents on Curable of Sickle Cell Anemia

Responses	Number	Percentage
Curable	38	28.57
Non-curable	95	71.43
Total	133	100

Table 15: Knowledge about Sickle Cell Information cum Counseling Centre located in Maharani Hospital

Responses	Number	Percentage
Yes	44	33.08
No	89	66.92
Total	133	100.00

Table 16: Opinion about Availability, Accessibility and Affordability for Prevention and Treatment of Sickle Cell Anemia

Responses	Number	Percentage
Yes	32	24.06
No	101	75.94
Total	133	100.00

Perception of Respondents on Curable of Sickle Cell Anemia

It has observed according to table 14 that 28.57 percent respondents perceived that sickle cell anemia is curable and 71.43 percent are not agreed.

Knowledge about Sickle Cell Information cum Counseling Centre located in Maharani Hospital

Table 15 shows that only 33.08 percent respondents have the knowledge of sickle cell information cum counseling centre located in Maharani Hospital other 66.92 percent were not aware.

Openion about Availability, Accessibility and Affordability for Prevention and Treatment of Sickle Cell Anemia

According to table 16, it has been observed that only 24.06 percent respondents were capable for availability, accessibility and affordability for prevention and treatment of sickle cell anemia and rest 75.94 percent were found incapable.

Stigma toward of Sickle Cell Anemia

There's quite a lot of stigma toward the whole subject of sickle cell anemia. People can feel guilty because they carry a gene and they choose not to talk about it. So they need to talk about it to start breaking down the barriers and the stigma. People are a bit sensitive about screening but it now can be enrolled on a program and start to care for baby with sickle cell anemia. Sickle cell anemia can no longer be overlooked upon as a largely black disorder. There has been the crossing of racial boundaries with sickle cell.

Sickle cell anemia has not been highlighted because it is a black disorder so it has not received

any spotlight with interracial mixing. People still have a stigma about sickle cell anemia they think it is a "curse of the devil". Many physicians and scientists both black and white have complained that restrictions against blacks with the sickle cell trait was a senseless stigma and unscientific suggestion that their genes were somehow inferior in addition of its use in barring blacks. From the air force academy the trait has also been cited by the navy in keeping blacks out of the submarine service and by the army although they will not allow the sickle cell trait carriers to become aircrew members. This policy persists in the air force itself despite today's change in admissions policy but it is under review. Blacks have also been charged more money for insurance policies when it was learned that they had the trait. Sickle cell trait screening has not been limited to the military or to the insurance companies in the chemical industry theories have been expounded for years that sickle cell trait carriers were at special risk in the chemical work place.

Today the law would be condemned as racial profiling. The stigma was made worse by a misunderstanding of the inheritance of the condition contrary to report of premature deaths carriers of the sickle cell gene were in almost all cases, healthy genetic screening and public immunization programs have also raised suspicions among blacks and sickle cell anemia. Screening programs of the 1970's created misinformation confusion and feared inadequate planning and preparation on the part of the medical profession and public health officials and a disease and having it resulted in unnecessary stigma and discrimination as a result.

Health related stigma is increasingly becoming a major public health issue that is receiving more attention. Young adults with sickle cell disease (SCD) are at risk for healthrelated stigmatization due to the many challenges of the disease. SCD includes the lifelong challenges of managing the chronic illness while accessing and navigating the health care system. The burdens of the disease can affect all aspects of the lives of individuals with SCD to include physiological, psychological and social well being. Although others may be involved in the process of stigmatization, the purpose of this paper was to support the need to develop patient oriented interventions to prevent and treat health related stigma in young adults with SCD, as these individuals may face health related stigma throughout their lives, but especially immediately after transitioning from pediatric to adult care. Additionally, the Revised Theory of Self Care Management for Sickle Cell Disease is offered as a framework from which theory based interventions can be derived.

Recommendations

People with sickle cell disease can live full lives and enjoy most of the activities that other people do. There are things that people with sickle cell disease can do to stay as healthy as possible (Creary, M., Williamson, D., Kulkarni, R.J., 2007; Treadwell, M.J., McClough, L., Vichinsky, E.,2006; Pack Mabien, A., Haynes, J. Jr, 2009).

- Regular health checkups with a primary care doctor can help prevent some serious problems.
- Common illnesses, like the flu, can quickly become dangerous for a child with sickle cell disease. The best defense is to take simple steps to help prevent infections.
- People with sickle cell disease should drink 8 to 10 glasses of water every day and eat healthy food. They also should try not to get too hot, too cold, or too tired.
- New clinical research studies are happening all the time to find better treatments and hopefully, a cure for sickle cell disease. People who participate in these studies might have access to new medicines and treatment options.
- Find a patient support group or community based organization in your area that can provide information, assistance, and support.
- A training programme about the haemoglobin disorders and appropriate management is provided for key staff, specifying the objectives, volume, methods and evaluation of training.

Conclusion

Although there has been extensive clinical and basic science research in SCD, many public health issues, such as blood safety surveillance, compliance with immunizations, follow up of newborns with positive screening tests, stroke prevention, pregnancy complications, pain prevention, quality of life and thrombosis, in people with SCT remain unaddressed. Currently, efforts are under way to strengthen SCD related activities. In all states and the district of Bastar have universal newborn screening (NBS) programs for sickle cell disease (SCD), which also identify sickle cell trait.

With the global scope of sickle cell disease, knowledge of the countless clinical presentations

and treatment of this disorder need to be familiar to generalists, haematologists, internists and paediatricians alike. Additionally, an underlying grasp of sickle cell pathophysiology, which has rapidly accrued new knowledge in areas related to erythrocyte and extra erythrocyte events, is crucial to an understanding of the complexity of this molecular disease with protean manifestations. We highlight studies from past decades related to such translational research as the use of hydroxyurea in treatment, as well as the therapeutic promise of red cell ion channel blockers, and antiadhesion and anti-inflammatory therapy. The novel role of nitric oxide in sickle cell pathophysiology and the range of its potential use in treatment are also reviewed. Understanding of disease as the result of a continuing interaction between basic scientists and clinical researchers is best exemplified by this entity.

Apart from some factors such as malaria, endogamy marriage, ethnicity and imbreeding are responsible for this variability. Hence, a systematic study in a small geographic area like a district is required to understand the population structure and natural history of sickleing. This should be followed by creating awareness, genetic counseling prenatal diagnosis to control the birth of sicklers.

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