International Journal of Science and Research (IJSR)
ISSN: 2319-7064
ResearchGate Impact Factor (2018): 0.28 | SJIF (2018): 7.426

A Study Assessing Health System's Preparedness against Sickle Cell Anemia in Nashik, Maharashtra

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Abstract: ***Prevalence rate of 4% is present in Kokana tribe for sickle cell anemia, which is quite high and this tribe in indigenous to Nasik district constituting around 1/4th of the tribal population. The sickle cell anaemia is concentrated in tribal population in Nashik similar to general trend. ST population of 15,64,369 forms 25.6% of total population and is substantially high [census, 2011]. Out of total 15 blocks in a district 9 are tribal. Since these communities are closed ones following local customs of endogamy, consanguineal marriages, endogamy, during my field visits to District Hospital (DH), Nasik, I observed there were many carriers who have got married to carriers or trait patients which will lead to further inheritance of the disease. Also, lack of provision of new-born screening leads to missing cases and late diagnosis. Thus it important to have active response from health system against SCD in Nasik. Also, since Haematology program and day care centre was launched in Nasik in 2013, the treatment program for sickle cell anaemia was integrated with thalassemia and haemophilia. The governance of Sickle Cell Disease Control Programme launched under NRHM on 17 January 2008, which was till 2015 done with a partner NGO (Gauri samajik Kalyankari Sanstha) was withdrawn from it in July 2015. Since then program has been sidelined. (Observation of available data and anecdotal learning during visit to DH). My study aims to provide evaluation of effectiveness of systems' response to SCD and give policy options for reviving the health system's response to SCD and further improvement of existing initiatives.***

Keywords: sickle cell anemia

1. Introduction and Importance of Study

Sickle-cell anaemia is an inheritable blood related disorder that affects the haemoglobin molecule, and causes the entire blood cell to change shape under stressed conditions. In sickle cell anaemia, the haemoglobin molecule is defective. After haemoglobin molecules give up their oxygen, some may cluster together and form long, rod-like structures which become stiff and assume sickle shape. Unlike healthy red blood cells, which are usually smooth, sickled red blood cells cannot squeeze through small blood vessels. Instead, they stack up and cause blockages that deprive organs and tissues of oxygen-carrying blood. This process produces periodic episodes of pain and ultimately can damage tissues and vital organs and lead to other serious medical problems. Because they cannot be replaced fast enough, the blood is chronically short of red blood cells, leading to a condition commonly referred to as anemia. . [Genes and human diseases, WHO, 2017]

Prevalence: Sickle cell disease is a genetic disorder found in one in every four hundred individuals. Sickle cell anaemia is a genetic disorder with single gene defect which affect over two million people globally[TOI, 2013]. Globally it is particularly common among people whose ancestors come from Sub-Saharan Africa, South America, Cuba, Central America, Saudi Arabia, India, and Mediterranean countries [Genes and human diseases, WHO, 2017]. Taking into our huge population size, more than 50 % of the world's sickle cell anaemia cases are in India. It is estimated that most of the cases are in the Central and South India [Kate, Sudam, Lingojwar et al, 2002].SCD affects mostly the socioeconomically underprivileged communities, living in the three clusters of 10 neighbouring states in India. Most of this endogamous population in India, including, scheduled caste (SC), scheduled tribe (ST), nomadic tribe (NT) and other communities (also known as OBC) are confined to locations, where SCD co-exists with endemic malaria. SCD in tribal population is well known in Central India. [Lingojwar, Devendra, Jadhav et al, 2016]. The overall prevalence amongst SC, ST and OBC is 10%. [Kate, Sudam , Lingojwar et al, 2002].The overall prevalence among tribal populations is about 10% for carriers of the disease and 0.5% for sufferers in India [Colah, Mukherjee, Martin et al, 2015].

About the prevalence of the disease in Maharashtra, the report published in Times of India, counts 10.5% of the tribal population, approximately 9,45,000 people, as affected by the disease. While the overall incidence of the disease in the state is less than 0.1%, it is very high among the tribal population groups from Nandurbar and Gadchiroli districts. [TOI, 2013].

During last 50 years, because of simple, reliable and inexpensive laboratory methods available, the large number of population genetic surveys conducted by different scientific groups and data on geographical distribution, clinical manifestation along with its variations, large no. of cases data is available from the state of Maharashtra.

Tribe	District	Prevalence
Otkar	Gadchiroli	35
Pardhan	Gadchiroli	32
Pawara	Chadrapur, yewatmal	25
Madia	Nandurbar, Jalgaon	20
Bhill	Gadchiroli	13
Haibi	Gadchiroli	11
Rajgond	Gadchiroli	10
Korku	Amravati	09
Kolam	Yewatmal	09
Warli	Thane	07
Katkari	Pune, Raigad	04
Kokana	Kokana, Nasik	02
Andha	Nanded	01
Mahadev Koli	Pune, Nasik	01
Thakar	Pune, Raigad	01
Paradhi	Solapur	00

Source: [Colah, Mukherjee, Martin et al, 2015]

As we see in the figure, Prevalence rate of 4% is present in Kokana tribe which is quite high and this tribe in indigenous to Nasik district constituting around 1/4th of the tribal population. The sickle cell anaemia is concentrated in tribal population in Nashik similar to general trend. ST population of 15,64,369 forms 25.6% of total population and is substantially high [census, 2011]. Out of total 15 blocks in a district 9 are tribal. Since these communities are closed ones following local customs of endogamy, consanguineal marriages, endogamy, during my field visits to District Hospital (DH), Nasik, I observed there were many carriers who have got married to carriers or trait patients which will lead to further inheritance of the disease. Also, lack of provision of new-born screening leads to missing cases and late diagnosis. Thus it important to have active response from health system against SCD in Nasik.

Also, since Haematology program and day care centre was launched in Nasik in 2013, the treatment program for sickle cell anaemia was integrated with thalassemia and haemophilia. The governance of Sickle Cell Disease Control Programme launched under NRHM on 17 January 2008, which was till 2015 done with a partner NGO (Gauri samajik Kalyankari Sanstha) was withdrawn from it in July 2015. Since then program has been sidelined. (Observation of available data and anecdotal learning during visit to DH).

My study aims to provide evaluation of effectiveness of systems' response to SCD and give policy options for reviving the health system's response to SCD and further improvement of existing initiatives.

2. Literature Review

Many population groups have been screened and the sickle cell gene has shown to be prevalent among three socio-economically disadvantaged ethnic groups, the scheduled tribes, scheduled castes and other backward classes in India. In Maharashtra, the sickle gene is widespread in all the eastern districts, also known as the Vidarbha region, in the Satpura ranges in the north and in some parts of Marathawada. The prevalence of sickle cell carriers in different tribes varies from 0 to 35 per cent. The tribal groups with a high prevalence of HbS (20-35 %) include the *Bhils*, *Madias*, *Pawaras*, *Pardhans* and *Otkars* [Colah, Mukherjee, Martin et al, 2015].

The pathophysiology of sickle cell anemia (SCA) arises from hemolytic anemia and acute vaso-occlusion; organ damage develops from recurrent erythrocyte sickling, chronic hemolysis, and progressive endothelial vasculopathy [Ware, Russell, 2010]. The painful vaso-occlusive event (often referred to as pain "crisis") is the hallmark acute clinical manifestation of SCA, characterized by the sudden onset of discomfort. Often occurring without a specific trigger, pain typically presents in the chest, back, or extremities and requires hydration, analgesia, and frequent treatment, including hospitalization. Other acute vaso-occlusive events include splenic sequestration, acute chest syndrome (ACS), and stroke, whereas others derive more from hemolytic anaemia, such as pigmented bilirubin gallstones and jaundice. Organ damage begins early in life and worsens over time and culminates in early mortality. SCA is recognized, therefore, as a chronic and progressively debilitating medical condition with considerable morbidity from insidious but inexorable organ damage. Accordingly, early diagnosis and treatment in young patients who have not yet developed serious or irreversible organ damage is a worthy goal [Ware, Russell, 2010].

Most of the early studies on epidemiology of sickle haemoglobin in different parts of the country found that the sickling or the solubility test is used and this was followed by Hb electrophoresis to determine the phenotypes. However, in recent years, high performance liquid chromatography (HPLC) analysis has been used in many large programmes to identify carriers of both sickle haemoglobin as well as β-thalassaemia. Capillary electrophoresis has also now been introduced at some centres. Nonetheless, even the simple and cost-effective solubility test has been shown to have a sensitivity and specificity of 97.4 and 100 per cent, respectively in comparison to HPLC and could still serve as a good first line screen for sickle haemoglobin in remote areas where other facilities are not available [Colah, Mukherjee, Martin et al, 2015].In high-income countries that provide neonatal diagnosis and care for patients, most survive well into adult life and, because there is limited use of prenatal diagnosis, numbers of patients are rising steadily. Most affected children born in low-income countries still die undiagnosed, usually from malaria [Modell , Darliso, 2008].

At the current time, curative therapy with stem cell transplantation remains an unavailable option to most patients Until something better becomes available that has a similar wide spectrum of efficacy and safety, hydroxyurea appears to be the best available treatment option for children and adolescents with SCA. Based on currently available data, hydroxyurea treatment fulfills these criteria for SCA and should be offered much more frequently, especially to young patients before the development of chronic complications and end-organ damage[Ware, Russell, 2010].While penicillin prophylaxis is the mainstay, pneumococcal vaccination is also important in the fight against pneumococcal infection [King, Madden, 2015].

Delivering health care to tribal populations is a challenge and a village based model has been described in Bardoli in Gujarat where an outreach programme is being undertaken with the help of a mobile clinical unit and a local villager has also been given basic health care training to regularly visit and monitor sickle cell disease patients and send those with significant complications to the hospital coordinating the programme[Colah, Mukherjee, Martin et al, 2015].

State Intervention for Sickle Cell Disease To reduce and control the high prevalence rate and to reduce incidence of sickle cell disease in the state, Maharashtra government started the Sickle Cell Disease Control Programme under NRHM on 17 January 2008, with aims such as to create awareness in the community about the disease, provides diagnostic services, management counselling etc of SCD The NRHM have following objective in the sickle cell disease control program:

1) Create awareness about the sickle cell disease.
2) Screening the general population for sickle cell disease (target age group 1 to 30 years and with special focus on adolescent age and pregnant women).
3) Provision to the hospital with electrophoresis machines.
4) Providing the diagnostic testing facilities at primary health centre, Rural hospital, Sub district hospital and District hospital.
5) Identification of the carrier and sufferer.
6) Issuing the red cards and yellow cards to the carrier and sufferer.
7) Counselling the carrier and sufferer for marriage and importance of regular treatments.
8) Providing prophylactic and symptomatic treatment at primary health centre, Rural hospital and District hospital level.
9) Specialty treatment and care at district and medical colleges.

3. Objectives and Research Questions

Objectives- 1
To study epidemiological context of sickle cell anaemia in Nashik district
Research question
1) What is burden of Sickle cell Anemia in Nashik?
2) What is age/sex/caste/area/year wise distribution of a disease?

Objective-2
To analyse health systems context
1) How is the physical performance under district sickle cell control program?
2) How is health providers' perspective in managing sickle cell anaemia?
3) What are morbidities present in SCD cases?
4) What is mortality scenario for SCD in district?

Objective-3
To understand social context of sickle cell anemia in Nashik district
1) Has the health seeking behaviour of the patients improved?
2) How well is the availability, accessibility, affordability of the treatment according to the patients?

Objective -4
To examine policy options for combating sickle cell anemia
1) What are areas which need to be improved in implementation of strategy to combat SCD?
2) Are these solutions feasible in the district?

4. Methodology

Type of study: Cross sectional, Mixed method, descriptive study
Sampling: Simple random sampling is used to select patients from record register at Civil Hospital, Nashik

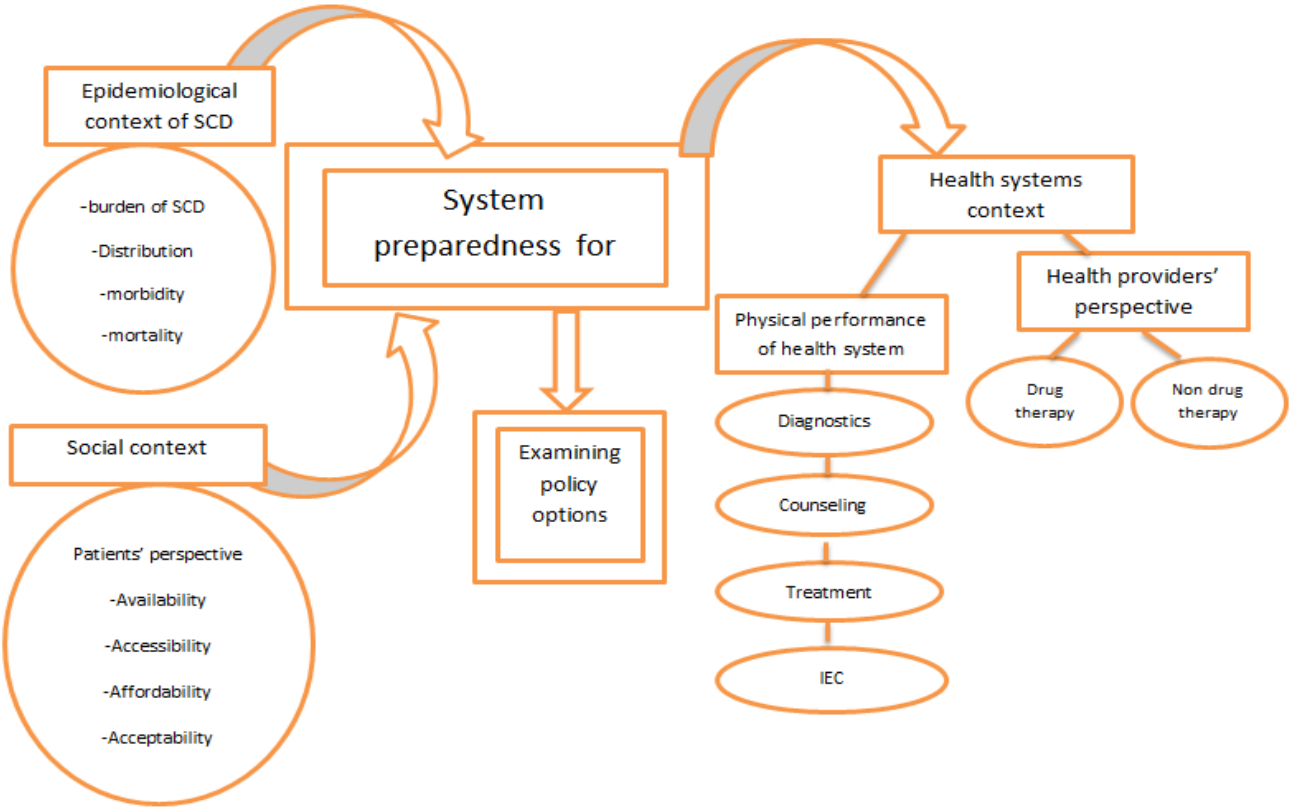
Purposive sampling is used to interview health providers. (Health providers working under sickle cell district control program)
Convenience sampling is used to interview patients (Patients attending care at DH, Nasik)

Secondary data is obtained from district office, Sickle cell disease control program and patients 'records in DH, Nasik as well from reports of Gauri Samajik Kalyankari Sanstha

Area of study:
This study is carried out in Nasik District in Maharashtra.

Tools of data collection:
Secondary data for 175 sufferers of SCD and under sickle cell control program, NRHM from district office (from 2009 till 2014)
Secondary data from reports of Gauri Samajik Kalyankari Sanstha (from 2009 till 2014)
Secondary data from patients 'records in DH, Nasik
Face to face / telephonic interviews of 20 sickle cell anaemia sufferers
Face to face interviews with 10 health providers (1 MO, 2 Counsellors, 1 physiotherapist, 1 program co-ordinator, 1MO at DH, Hematology Dpt. , 2 ASHAs, 2ANMs)

5. Conceptual Framework



Analysis:
The secondary data analysis is done using Excel and SPSS software. The manual qualitative as well as quantitative analysis of interviews of all the respondents is done. The data were obtained mainly in Marathi language. All the text was translated into English language for analysis purposes.

Ethical consideration:
The study participants were given full information about the research, researcher and about the purpose of the study. Each participant`s informed consent about their participation in the study was obtained orally. Participant`s name was not mentioned anywhere in the study and they were assured about confidentiality of the information provided by them. Participants were told they can have right to decline or refuse to answer a particular question during the interview and free to make an independent statement in the study.

6. Findings

1) Epidemiological context

Epidemiology of SCD
Prevalence of SCD
There are 175 sufferers and 2840 carriers (till august 2017) which means prevalence of SCD for sufferers is 0.003% in district and prevalence for Carriers is 0.05%.

There are total 131 sufferers belonging to the ST category making prevalence rate for sufferers 0.008%.

Year wise case detection (sufferers)

Year	Frequency	Percentage increase over last year
2008-09	4	-
2009-10	8	100
2010-11	8	0
2011-12	7	-12.5
2012-13	10	42.9
2013-14	28	180
2014-15	50	78.6
2015-16	32	-36
2016-17	20	-37.5
2017-18	5	-75
Total	175	-

This table shows year wise detected cases from different public facilities as well as screening camps. The case detection is seen to decrease in last 3 years. This could mean either the disease has been controlled or that screening rate of population could be declining.

However analysis in the further part shows that the screening rate has been drastically declining.

Talukawise distribution of sufferers (According to the place of detection)

Name of block	Frequency
Deola	2
Dindori	4
Igatpuri	1
Kalwan	13
Nandgaon	1
Nasik	36
Civil hosp	08
Niphad	1
Peth	12

Satana	4
Surgana	4
Triambak	4
Vani	3

(From 2009 till date)

High no. of patients detected in Nashik district could be due the fact that they are referred to DH for testing as well as treatment (from the primary data collected it was evident that most of the times the patients were diagnosed for SCD only when they were seriously ill and were referred to DH by other health facilities. This could be also due to high level of migration in Nashik block especially by tribal communities for seeking work (anecdotal finding).The data has some faults since for few patients area of detection centre was noted as area of residence.

Distribution of SCD

Age wise distribution of sufferers

Age group	Frequency	Percentage
0-5	13	7.4
6-20	114	65.1
21- 35	41	23.42
36-50	6	3.4
50 and above	1	0.6
Total	175	100

Highest no. of sufferers are in the age group 6 to 20 followed by the age group of 21 to 35. This could be due to the fact that 0 to 30 is the target age group under sickle cell disease control program and screening in this age group has been more focussed on. Also sickle cell symptoms start manifestation during early ages of life and patients tend to seek care on such acute episodes of disease and get diagnosed thereafter (As told by one of the medical officer interviewed)

Sex wise distribution of sufferers

Male	87	49.7
Female	88	50.3
Total	175	100

There is no difference in prevalence among both sexes in Nasik.

Category wise distribution of sufferers:

Sr. no.	Category	frequency	%
1	ST	131	74.9
2	SC	35	20
3	OBC	2	0.01
4	Open	7	0.04
Total		175	100

As we see almost 75% of the case load is from ST category.

Caste wise distribution

Caste	Frequency	Percentage
Kokana	115	65.7
Warli	19	10.9
Mahadev koli	13	7.4
Mahar	10	5.7
Maratha	7	4
Bhil	5	2.9
Mali	3	1.7
Baudha	1	0.6
Chambhar	1	0.6
Muslim	1	0.6
Total	175	100

Among tribal communities Kokna tribe is most affected. In Nasik this community constitutes around 1/4th of the tribal population thus there is increased risk of continued transmission. (Anecdotal learning from interview one of the ANM and ASHA)

Disease Morbidity

Pain: Frequent episodes of pain were reported by all the patients. They also said that pain strikes almost anywhere in the body but the pain often occurs in the lower back, Legs, Arms, Abdomen (on left side). According to the doctor interviewed, People described this pain as sharp, intense, or throbbing.

Anaemia: All the patients had mild to moderate anaemia. All the patients interviewed had evidently pale skin and reported frequent fatigue and feeling of tiredness. From the reports in the patients records 80% of the patients had Hbs level in the electrophoresis reports above 65%. Also all the patients had Haemolytic Anemia according to the reports as well as doctor's interview.

Sickle cell crisis: 45% of the patients reported they had episodes of severe joint pain which was unbearable, fever, severe weakness, loss of apetite etc. later, they were told they were in the crisis phase of SCD. Shockingly 45% of the patients were diagnosed for SCD after going into crisis phase.

Other complications: Coughing, chest pain, shortness of breathing and fever were reported by patients among other symptoms. According to the doctor such symptoms can be suspected to be caused by a serious complication of sickle cell disease called **acute chest syndrome.**

Increased number and frequency of **infections, Jaundice, splenomegaly, leg edema and ulcer**s are other complications in all the patients seeking treatment at DH (records).

Mental Health: 15 out of 20 patients reported they feel sad and frustrated at times. The limitations that SCD imposes on their daily activities causes them to feel isolated from others. Sometimes they become depressed. According to the counsellor interviewed, People with SCD have trouble coping with pain and fatigue, as well as with frequent medical visits and hospitalizations.

Mortality

There are total 9 deaths reported among sickle cell sufferers in a district from the period of 2009 to this year. Out of 9, 5 deaths were reported due to Multiple organ failure whereas 1 was due to kidney failure and for others cause of death was not reported. Also other specific data regarding the death cases was not available.

2) Health systems context

a) Physical performance under sickle cell disease control program

Sickle cell awareness and control program was launched in 2007 under NRHM. In Nashik district it was implemented from November 2008. Initially it was launched in 9 tribal bloks only, covering 52 PHCs and 13 RHs.
The objectives of the mission are-

- Create a public awareness Screening general population (Target age 1 to 30 years)
- Training of medical officers and other paramedical staff PHC, RH &
- DH for testing, counseling and treatment.
- Equipping hospitals with electrophoresis machine.
- Providing testing facilities at Primary Health center, Rural health, Sub-district hospital, District hospital.
- Identify carrier and sufferer
- Issuing yellow card to carrier and red card to diseased individual.
- Counseling carrier and sufferer for marriage and importance of regular treatment.
- Special treatment at District Hospital and medical college.

b) Sickle cell control program physical status as on Aug, 2017

- *Availability of infrastructure*

Sr. No.	Name of District	Total no. of PHCs implementing SCD program	Total no. of electrophoresis machines available in RH	No. of machines in working condition	No. of PHC where Solubility test performed	Medicine available in (No. of PHC)	No. of Electro - phoresis kits available in Centers	Consumables (EDTA Blub, syringe, Lancet, needles, etc.) available (No. of PHC)
1	Nashik	104	**11**	11	104	52	15	52
TOTAL		**104**	**11**	**11**	**104**	**52**	**15**	**52**

All the PHCs in the district are centres for implementing District Sickle Cell Disease control program (SCDCP) under NRHM. Also, solubility testing is available at all the centres. At 50% of the PHCs medicines like folvite, Zinc, MVBC, calcium is available. (Note: hydroxyurea is available only in the DH. All the PHCs have solubility testing available. There are total 11 electrophoresis testing centres (details given in further report). However none of the PHC has availability of electrophoresis testing.

- *Solubility test performance against target*

Year	Target	Achievement	Percentage achieved (%)
2014-15	200000	197639	98.81
2015-16	200000	40847	20.42
2016-17	200000	5368	2.68

We see that the solubility test performance is drastically declining with more than 78% fall from 2014-15 to 2015-16 and further dropped by 8% in 2016-17.

- *Testing report*

Sr. no.	Year	Solubility Testing	Carriers	Sufferers
1	2008-09	1892	74	4
2	2009-10	31649	387	11
3	2010-11	79069	449	9
4	2011-12	222338	312	10
5	2012-13	114253	308	19
6	2013-14	203039	385	32
7	2014-15	197639	375	49
8	2015-16	40847	Data unavailable	32
9	2016-17	5368	71	5

- *Validity of solubility testing*

No of Taluka	*Total tested for solubility Test*	*No. of samples Found positive in solubility Test*	*No. of positive samples sent for electrophoresis*	*Result of Electrophoresis*			
				Carriers	Sufferers	Total (C+S)	Negative
15	5368	164	164	71	**5**	76	87

Only 76 out of 164 solubility test positive samples were found to be positive during confirmation with electrophoresis which means only46% of the samples tested with solubility test were actually carriers or sufferers.

Positive predictive value=true positive/ true positive+ false positive =76/87+76=76/163=46.62%

So, the chance that a person with a positive test truly has the disease is 46.62%

However Positive and negative predictive values are influenced by the prevalence of disease in the population that is being tested. In a high prevalence setting, it is more likely that persons who test positive truly have disease than if the test is performed in a population with low prevalence.

- *Coverage of target population by solubility testing*

No of Talukas	Total Population (census 2011)	Target Population of 1 to 30 Age group	Total tested for solubility Test
15	**3638580**	**2001219**	5368

Even if we assume all 5368 testing during 2016-17 were from the target age group it is only 2.68%

- *Electrophoresis testing*

No. of Electrophoresis centers	Total Electrophoresis machine supplied from state level	Name of Electrophoresis centre	Total Electrophoresis machine available in centres	Total Electrophoresis test (April 2015 to March 2016)	Total Electrophoresis test (2016-17)
4	11	RH Vani	2	344	64
		RH Triambak	2	101	9
		SDH Kalwan	3	287	20
		DH Nasik	4	159	29
Total			11	891	122

However tribal blocks like Peth, Igatpuri, Surgana, Deola, Dindori still lack electrophoresis testing centres. For some of them nearest electrophoresis testing centre is as far as 40-50 kms. Also except Nashik block none of the non tribal blocks have provision of electrophoresis testing in public facilities in district.

There is however high migration among tribals in Nasik and they need to come to DH Nasik for testing and thus need for testing centres in other blocks as well.

- *HPLC testing*

Name of Electrophoresis centre	No. of HPLC Machine Supplied	No of HPLC test kits Available	Total HPLC test done in 13-14	Total HPLC test done in 14-15	Total HPLC test done in 15-16	Total HPLC test done From 16-17	Total progressive
DH, Nasik	1	2	33	Data unavailable			101

There is only one HPLC centre in a district at DH, Nasik.

- *Card distribution*

The data shows that all sufferers as well carriers have been given red and yellow cards respectively and also patient tested negative have been given white cards.

- *Awareness*

The data on important component of the program i.e. awareness is missing after 2015.
For 2014-15 the blockwise awareness data is available for all the tribal blocks.
For awareness following activities are carried out-

Organizing seminar in colleges, school levels & also at Grampanchayat level through Anganwadi workers, Asha teachers & local people. With the help of print media awareness is created in society though Brochures ,pamphlets, hand bills, stickers, wall paintings & various competition also through street-play, slogan , rally, cultural plays information to people about sickle cell anemia. All these activities used to be done with the help of volunteers of Gauri Samajik Kalyankari Santha, Nasik (the NGO which was partner of the program from 2008 to 2015) [reports].

Also if we see screening camping conducted, most of them are concentrated in tribal blocks with very low emphasis on non-tribal blocks. As mentioned above migration among tribal population is evident thus leading to chance of missing out of cases.

Counselling

Counselling is an important component of program and data regarding same is available under SCDCP, district office.

Total Carrier	Total Sufferer	No of carriers					No of sufferers				
		Total Married	Counseled from married	Total unmarried	counseled from unmarried	Total Counseled Carriers	Total Married	Counseled from married	Total unmarried	counseled from unmarried	Total Counseled Sufferer
2832	171	1045	1045	1787	1787	2832	50	50	121	121	171

From 2009-till now

The table shows that all sufferers as well as carriers had undergone genetic counselling from the certified counsellors under program. Around 37% of the sufferers and 30% of the sufferers are married. Which means these many persons did not receive premarital genetic counselling for SCD.

Impact of counselling:

Total Carrier	Total Sufferer	Total Married couples		No. Married couple tested		No. married to healthy partner		Total % of couples tested		% married to non SCD partner	
		Carrier	Sufferer	Carrier	Sufferer	Carrier	Sufferer	Carrier %	Sufferer %	carriers	sufferers
2832	171	1002	50	927	50	762	42	93%	100%	76%	84%

At least one of the individuals among these couples had received counselling and were recommended to get spouse testing done, however from the table we see that 7% of the couples still have not done testing for SCD.

24% of the carriers and 16% of the sufferers are married to either carrier or sufferer. This is a cause of concern as it indicated chances of further continuance of inheritance of disease in the closely knit tribal communities. Thus arises

need for active prenatal/neonatal screening to avoid further spread by early detections.

Treatment:

No. of Suffers HPLC test done	Total Health File Distributed to Sufferers at DH	Prophylactic treatment (Folic Acid & Vit C	Symptomatic treatment (Pain killer Antibiotic etc.)	Hydroxyurea	Blood Transfusion	No. of patients given treatment in Day Care Centre	Referred through Telemedicine Specialist
170	150	170	8	150	54	82	112

We see that all the patients at DH are on Hydroxyurea treatment. There is a telemedicine centre at DH where patients can contact during emergency to avoiding travelling long distances.

Impact of treatment:

From the records at DH,

No. of Patients having Pain crisis before regular treatment	No. of Pain Crisis reduced due to reguler treatment	No. Patients Needed Regular Blood Transfusion before tt	No. of Blood transfusions reduced due to regular treatment	No. of patients Hb gm % increased due to reguler treatment	No. of Children not able to attend School due to pains before regular treatment	No. of School Children attending school due to reguler treatment
15	6	9	44	5	0	2

We see that there is obvious positive impact for those following regular treatment.

IEC

The IEC material was seen to be available in RHs, SDHs and was seen to be available and displayed only in DH, Nashik. However though abundant IEC material has been available in local language on sickle cell anaemia, its availability in sub centres and PHCs was very low or completely lacking.

The awareness and testing camps were held in orphanages, primary schools, health centres in tribal blocks. Earlier the partner NGO use to take active part in these camps with their volunteers taking group meetings for teachers, parents, displaying posters, distributing pamphlets one day prior to testing camp followed by testing of people by lab technicians, ANMs, MPWs the next day. According to the founder of this NGO, the response was overwhelming in all the tribal blocks. Also according to few providers the response to the camps is better in tribal parts as compared to non tribal parts.

3) Health providers' perspective on management of sickle cell anemia:

At grass root levels

All the ASHAs interviewed were aware that they are supposed to give information on SCA in Gramsabhas at least 4 times in an year and take adolescents meetings and meetings with women self help groups. They were also aware about meanings of white, yellow and red cards and about their responsibility of distribution and records. They were also aware about home visits and counselling to be done of marriageable age group carriers, newly married carrier couples or carrier- sufferer couple or sufferer-sufferer couple.

All the health providers were aware of provisions under beneficial schemes and programs for sickle cell patients – Sickle cell disease control program under NRHM

Sanjay Gandhi Niradhar Anudan Yojna under state government

Rashtriya bal arogya karyakram, 2013

Disability status to be given to sickle cell patients according recent disability act

Day care centre is available in district hospital with separate wards for females, males and children

Diagnostics

All the health providers reported that newborn screening is not done in Nashik district. All of them explained that since prenatal screening is done in all ANC mothers new-born screening was not required as such. However given substantially low level of ANC coverage (26% only have got full ANC checkups done- NFHS 4)and no. of non institutional deliveries in Nasik district (15% total and 27% rura- NFHS) there are high chances of missing out of cases in prenatal screening.

Under NRHM the target age group was 0 to 30 years of age and there is no selective adolescents screening done in Nashik district.

The solubility tests are done in all PHCs where as electrophoresis is done only at 4 centres at Triambak, Vani, Kalwan and Nashik blocks. HPLC test is available only at the district hospital, nasik.

Few providers reported need for integration of genetic medical services at primary health care level since the people do not understand seriousness of the condition and go for further confirmatory tests late or do not go at all. The patients who turn out to be electrophoresis test positive are advised for spouse testing as well as family screening. However compliance is low and regular reminding is needed.

Prevention of infections:

All the sickle cell anemia patients are told by health providers that they are prone to recurrent infections. The parents are insisted to take routine immunization for their

children but there is no provision of pneumococcal infections. (though under 5 mortality due to Pneuminia in the year 2016-17 is 13% of the total U5 mortality ie 44 out of 344)

Drug therapy
Folvite, Multivitamin B complex and Hydroxyurea is given to all the sufferers visiting district hospital.

Few patients are also given Zinc and Calcium tablets.

Hydroxyurea is available only in District hospital, Nashik other drugs are also available at RH level.

The stock outs of hydroxyura were not rare in district hospital according to few staff interviewed and also during my visits to department the drug was not available.

Non drug therapy
According to the interview with the physiopherapist in District hospital, **physiotherapy is of very little use in case of Sickle cell anemia patients.** ''they are intolerant to exercise and tend to be fatigued easily, sternous activities tend to worsen their situation and may aggravate crisis phase thus these patients are advised to avoid physical exertion as far as possible and adequate rest. **Diet counselling and advice to drink plenty of water are only effective solutions**. Even children are advised not to play outdoor games and avoid physical activity''.

Diet counselling and advice for drinking plenty of water are most important non drug routine care strategies for managing sickle cell anemia according to all the health providers.

Genetic counselling

According to the counsellors at the district hospital the genetic counselling has its limitations especially in case of tribal population. Consanguineous marriages ie the marriages within the family are common in tribal communities. Asha respondent provided the information about the consanguineous marriage practices existed in the community. Doctors of the hospital provide counselling to the patients.

During pre-marriage counselling doctors explain to the patient or relatives of the patient the importance of avoiding marriage between sickle cell disease patients or carriers. **Many times they explain to the patient with the help of diagrammatic representation.** It is easy to understand for the illiterate person. According to the doctors the community people accept the counselling provided by them.

'For the prevention of this disease we provide the marriage counselling to the patient. In the marriage counselling we tell to the patient test the blood for sickle cell before marriage and avoid the marriage between two carriers.'- medical officer.

''However local marriage customs and traditions like dowry etc. are bigger concerns for these patients who mostly belong to lower socio economic groups''- MO

''sometimes the parents of the women sufferer/carrier do not inform her in laws or husband due to fear of family disputes and abandonment.''

Knowledge regarding facilities to be availed by Sickle cell patients was being given by counsellors in District hospital. All the health providers were aware of emolument of Rs. 600 to all sufferers with annual income less than Rs. 21000. And to all the sufferers below 18 years of age. (under Sanjay Gandhi Niradhar Anudan Yojna). They were also being told regarding free transportation facility in state transport buses and concessional transportation facility in railways for SS Pattern patients.

Follow up visits rate:
Rate of follow up visits is satisfactory in case of sufferers but carriers are mostly lost to follow up.
''Only female carrier patients who sometimes visit with the complaints of Dysmenorrhoea, menorrhagia visit here and asymptomatic carriers do not visit and difficult to track for their marriages etc.''

1. Social context
a) Patients' perspective
Socio-economic background of a patient:

- 12 male sufferers and 8 female sufferers were interviewed. The samples were reached through OPD in haematology section of Nashik district hospital.
- 10% of the patients were completely illiterate, 30% had attended only primary education and rest were educated above 5th class.
- 80% of the respondents belonged to below poverty line category.
- 70% respondents belonged to ST category.

Most of them were from distant blocks like Kalwan (25%), Peth (15%), deola (15%) and few were from other districts as well.

Most of them were farmers or farm workers, daily wage workers. Thus all maximum of the patients belong to lower socio economic status.

Health seeking behaviour
85% of the patients reported that they visited the health care facility when they were ill and were tested for sickle cell anaemia only after they showed symptoms.

Only 15% were detected during regular screening and were asymptomatic before testing.

65% patients reported they visited private practitioner on illness as first point of reference and were referred to district hospital at later stages.

35% of the patients reported they did first testing for sickle cell anaemia at private laboratories.

From the patients (sufferers) interviewed 20% were married to carriers. 30 % had not got family screening done. These are closely knit communities and local customs are still prevalent leading to consanguineal marriages and marriages within the community. Marriages between carriers, carriers

and trait persons are still happening. Thus the awareness campaign for the disease has not yet reached the masses and cultural carriers are rendering counselling method ineffective.

Availability, accessibility, affordability and acceptability of treatment

Availability: All (100%) patients reported that they have to come to district hospital for monthly check ups and collecting medicines. The hydroxyurea drug is not available at RH level in district and all the patients reported they have been prescribed this drug which is too expensive to be taken from private providers and they have to visit District hospital for it.

Accessibility: For majority of patients the district hospital is as far as 90 plus kms from their residence and thus travelling every month makes follow up visits cumbersome. From the patients records it was observed that only 50% of the patients are coming from regular monthly visits. Patients give lost work and long distances as main reason behind noncompliance. One five months' pregnant mother from the tribal block (Kalwan) reported due poor roads, she has not been able to visit district hospital since last 4 months and has to take medicines at very high prices from a private provider.

Affordability: All the patients reported the treatment at district hospital was free of cost and drugs were also provided for free. Also, travelling expenses were taken care since free ST transport facility provision is there.Also Rs. 600 is given to all patients under SGNY. They were given cards for free blood transfusion provision in blood banks all over Maharashtra. Thus there was 100% satisfaction with the affordability of treatment.

Acceptability: Only 15% patients reported side effects of the drug like nausea, acidity, constipation and rest were satisfied with the drug treatment. Acceptability of counselling including blood testing before marriage was there by all the sufferers. However they were still not convinced with the fact screening of whole family should be done, few of them told that other members of the family did not go for check-ups as they were showing no symptoms. According to them it was not necessary to get tested till you have some illness. Consanguineal marriages are common in tribal communities. Dowry, stigma are more imp. Concerns than pre-marital testing since all belonged to lower socioeconomic groups.

2. Examining policy options:

For diagnostics:

As observed during interviews significant amount of patients have got done tests for sickle cell anaemia done in the private laboratories. **As on now there was no interlinking of District sickle cell control program with the private providers. This would lead to significant data gaps and it would be difficult to track these patients further.**

In nasik there are 3 types of diagnostic techniques available-

1) Solubility test
2) Electrophoresis
3) HPLC

From the data it is clear that many patients who turn out to be positive during solubility tests are actually neither carriers nor diseased which can be confirmed only during electrophoresis. Thus there is need to alleviate fear in such patients and make sure they go for electrophoretic diagnosis.

Also, solubility testing is not reliable (especially in case of children) and thus there is need for more centres of electrophoresis testing. Currently 3 out of 9 tribal blocks have these centres. **I would suggest there is need for at least 2 more centres in tribal blocks like surgana and Peth which are far from current electrophoresis testing centres.**

Screening

Adolescent screening

There is no specific provision of adolescent screening. However during interviews I have observed that adolescent SCD patients were most worried and frustrated mainly due to concern of stigma and difficulty in finding partner for marriage. Thus **depression screening and specially focussed counselling provision for adolescents can be added to on -going counselling sessions.**

Selective adolescent screening could be effective strategy as this may lead to prevention of marriages between carriers or carriers and trait patients and would check on further inheritance. However area specific screening in tribal blocks should be avoided as there is high level of migration among tribals in Nasik (from anecdotal evidence as well as patients records).

Screening for Renal diseases:

There were significant no. of patients with renal diseases and also death due to renal complication according to records. As of now there is no provision of screening for renal diseases in SCD patients. According to the NHLBI guidelines screening for all individuals with SCD, beginning by age 10, for proteinuria and if result is negative, repeating screening annually is recommended. From the interviews with the staff at DH it was evident that these tests are available at DH and thus can be incorporated.

New born screening

There is no provision for newborn screening as of now. All the health providers reported that newborn screening is not done in Nashik district. All of them explained **that since prenatal screening is done in all ANC mothers new-born screening was not required as such. However given substantially low level of ANC coverage (26% only have got full ANC checkups done- NFHS 4)and no. of non institutional deliveries in Nasik district (15% total and 27% rural- NFHS) there are high chances of missing out of cases in prenatal screening.** According to the NHLBI guidelines, 2014 SCD screening with clinical consideration of confirmatory test within 2 months should be available for all the new borns.

New born screening would allow Identification of SCD for prevention and mortality from the disease- through active secondary prevention measures (Policy brief for Department

of health, Chhattisgarh). Subsequent experience demonstrated that neonatal screening, when linked to timely diagnostic testing, parental education, and comprehensive care, markedly reduces morbidity and mortality from sickle cell disease in infancy and early childhood (Harris MS, 1989). Also, it could lead to a significant decrease in infant and under- 5 mortality which is quite high in tribal blocks of Nasik district. It will also, contribute to primary prevention as screening can be act of alerting the family to the probability of having subsequent children with SCD. Due to the robust nature of dried blood samples means that this need not be confined to hospital deliveries but could be extended to home deliveries which is very useful in inaccessible areas of tribal blocks of Nasik.

Also, there is a favorable health systems context due to RBSK funded by NHM which calls for screening of newborns. The screening for SCDs is not currently emphasized in this scheme but can be added. Also, epidemiological considerations and anecdotal learning point out that the tribal population is Nasik is migrating to non tribal areas in high proportions thus selectively limiting new born screening to tribal parts would not be effective.

Pneumococcal vaccination:
Infection is the major cause of death in infants as well as children younger than age 5 (under 5 mortality due to Pneumonia in the year 2016-17 is 13% of the total U5 mortality ie 44 out of 344.) The pneumococcal vaccine is one of the candidate vaccines for universal immunization programme and is widely offered to infants in the private sector.

Currently there is no provision of giving this vaccine to newborn and or children under 5 years of age. I would recommend, **offering the vaccine as part of the treatment protocol to all the under 5 years age children who are detected with SCD.**

Hydroxyurea drug therapy

Level of provision:
Currently patients with SCD access HU only at the district hoshital level- and this gives only 3/4 weeks of drugs. Because of inability to come again and again- even those who urgently need it- cannot access these drugs on a regular basis. **I would suggest they could be made available at PHC, RH or SDH levels.** During the interviews program co-ordinator told that earlier the drug was available even at Rural hospital level but provision was discarded later due to mismatch in stocks and distribution records. **To overcome this stocks of hydroxyurea can be procured at DH and from their according to the records of patients registered in each RH the drugs can be sent there. Records monitoring can be done by hematology department in DH.**

Currently SCD mothers in Nasik are not advised to discontinue hydroxyurea during the period of breast feeding. **Maternal use of hydroxyurea is a contraindicated during breastfeeding (NHLBI), thus along with the counseling for pregnant women regarding the advantages of breastfeeding they should be advised to discontinue hydroxyurea until there is dire need.**

In nasik, Hydroxyurea is given to all the sufferers of SCD reporting to DH which around 90% of total caseload (above 24 months of age). However there are frequent stockouts in District Hospital which is only centre of availing this drug. It was noted that currently there is considerable financial barriers to access of this drug as the drug is very expensive to be brought from private facility. Thus instead of prescribing the drug to all the patients it can be limited to only those who need it the most. This way the availability of drug during crisis periods can be ensured. According to the NHLBI guidelines it is strongly recommended that in adults with SCD who have **three or more sickle cell-associated moderate to severe pain crises in a 12-month period, treatment with hydroxyurea to be given** and also for adults with SCA **who have sickle cell-associated pain that interferes with daily activities and quality of life. This can be followed here.**

7. Conclusion

Importance of continual program for sickle cell anaemia

The sickle cell anaemia is concentrated in tribal population in Nashik similar to general trend. ST population of 15,64,369 forms 25.6% of total population and is substantially high. Out of patients interviewed carriers belong to ST community and out of sufferers belong to ST community. These are closely knit communities and local customs are still prevalent leading to consanguineal marriages and marriages within the community. Marriages between carriers and even carriers and trait persons are still happening. For them dowry etc. are bigger concerns than blood testing before marriage. From the patients (sufferers) interviewed 20% were married to carriers. 30 % had not got family screening done.

Thus the awareness campaign for the disease has not yet reached the masses and cultural carriers are rendering counselling method ineffective (it also gives rise to question of right of sufferers/carriers of marrying person of their choice). If this situation continues this can emerge as an epidemic in tribal communities.

Awareness component of the program has been ignored since 2015 and there is no data available on it after 2015. There is need for continued awareness campaign and emphasis on community participation**.. The community volunteers can be created and used for creating awareness which may lead to better acceptance.**

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