

**Association for Health Welfare in the Nilgiris
&
Tribal India Health Foundation**



SICKLE CELL DISEASE

Sickle Cell Disease Center

OPERATIONS REPORT VERSION 1.7



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I. The Association for Health Welfare in the Nilgiris (ASHWINI) and the Tribal India Health Foundation (TIHF)

The Association for Health Welfare in the Nilgiris (ASHWINI) is an NGO, founded in 1991, working in the Nilgiris District, Tamil Nadu to improve the health status of 25,000+ tribals in the area. ASWHINI uses a base secondary-care tribal hospital, a cadre of village-based health workers, and a health insurance program to provide clinical and community-based services to the target area. ASHWINI also administers health education and mental health programs to the target community. The Sickle Cell Disease Center is based on the campus of the tribal hospital, the Gudalur Adivasi Hospital, located near the town of Gudalur.

The Tribal India Health Foundation (TIHF) is a US-based nonprofit organization dedicated to improving health systems for tribal populations throughout India. Through a combination of research and development, the Tribal India Health Foundation and its affiliates collect and send medical supplies and provide administrative assistance to partner tribal health initiatives in India. Most importantly, TIHF funds and administers the Sickle Cell Disease Center, based at the Gudalur Adivasi Hospital.

II. Indian Tribal Health and its Correlates

A. An Introduction to the Tribals of India

India, as a whole, has long been characterized with considerable heterogeneity in terms of religion, territory, language, and caste. During the colonial period, the British enumerated and classified India's population into groups and categories, one of which was the category of the tribal or *adivasi* (indigenous people). Prior to outside intervention, many tribal populations had self-regulating economic and political systems. The tribes were greatly dependent on the forest for their daily needs, including food, shelter, instruments, medicine, and even clothing.

With the policy of national development, however, the exploitation of minerals and forest altered the relationship between the tribes with the natural environment. As a result, tribal populations were forced into relinquishing those resources that had been the foundations of their existence, and what followed was the gradual governmental and societal castigation of tribals as a whole. Nowadays, the word "tribal" often brings to mind images of half-naked and women, arrows and spears in their hands, combined with myths of savagery and cannibalism. Thus, a limited knowledge of tribals in India has translated into a widespread propagation of false information and stigmatization of the approximately 100 million persons enumerated as members of Scheduled Tribes, comprising of about 10%-15% of India's population.²

B. The Health Profile of Tribal India and the Burden of Sickle Cell Disease

As a result of geographical and social barriers, relatively few primary and specialty care health facilities exist in tribal areas, and gaping disparities in health status of tribals, as compared to those in metropolitan areas, are evident. Along with a plethora of infectious diseases including malaria, tuberculosis, polio, and cholera, malnutrition and gastrointestinal disorders are pervasive

among tribal populations, and stark deficiencies have been detected in gross amounts of calcium, iron, vitamin A, vitamin C, and animal protein. Certain tribal groups such as the Onges, Jarawas, and Shompens of the Andaman and Nicobar Islands are facing extinction due to endemic diseases, venereal diseases, and an unusually low sex ratio.

With a large population, burgeoning birth rate, and consanguineous marriage practices, there is a dangerously high prevalence of genetic disorders among tribal populations. Along with amino acid irregularities, Glucose-6-Phosphate Enzyme Deficiency, a fatal and genetically carried deficiency in a blood enzyme, is present in about 15 million tribals, who reside in primarily high-incident malaria zones such as Madhya Pradesh, Maharashtra, Tamil Nadu, Orissa, and Assam states. However, it is the presence of sickle cell anemia among tribal populations that has surged to the forefront as a critical public health problem among tribal groups. A congenital hemolytic anemia that results from a defective hemoglobin molecule, the sickle cell disease causes red blood cells to roughen and become sickled. These cells, in turn, result in an impaired circulation, chronic ill health, periodic crises, long-term complications, and premature death. Half of the patients with sickle cell disease die before 20 years of age due to organ failure.

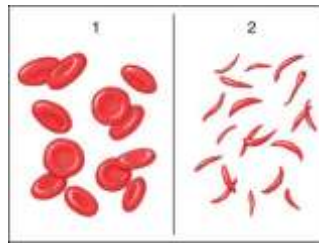
Epidemiological studies confirmed that sickle cell anemia is rampant in the tribal populace, the prevalence of homozygotes for the sickle gene calculated to be over 20% with an estimated five million individuals predicted as carriers. The sickle gene was first described in tribal groups in South India, and studies illustrate its presence in various parts of India including Tamil Nadu, Madhya Pradesh, Orissa, and Kerala. Genetic diseases have traditionally received little attention from urban health services in India, and even less so in tribal areas. As a result, virtually all studies carried out regarding tribal populations and sickle cell disease have strongly recommended that genetic health services be integrated into existing primary health care and medical services to combat the epidemic. Few healthcare delivery systems, however, have implemented these recommendations.

III. Objective: The Goal of the Sickle Cell Disease Center

Post India's independence in 1947, there was a temporary influx of literature on tribals, and tribal research institutes were set up in states that had substantial tribal populations. The purpose of these institutes was to undertake problem-oriented research for the formulation and implementation of development programs in tribal areas. However, though a significant amount of research was carried out in the early stages of these institutes' existence, the output of these institutes has "steadily declined both in quality and quantity." As a result, there have been few sustained interventions employed to combat the spread of congenital disorders in tribal areas.

The purpose of the TIHF/GAH Sickle Cell Disease Center, based at the Gudalur Adivasi Hospital, is to combine research with immediate clinical and community-based interventions to combat the spread of sickle cell anemia in Gudalur and Pandalur taluks in the Nilgiri area. The Gudalur Adivasi Hospital has been treating patients since 1990. The Center aims to devise, implement, and promote a holistic model of sickle cell management, in collaboration with partner organizations, which could be of use to other similarly affected communities in the country. The Sickle Cell Disease Center was started with the objectives of :

1. Identifying SCD in the newborn / or as early as possible by screening and confirmatory tests (hemoglobin electrophoresis).
2. Health education and counseling for SCD patients and relatives to enable the target population to understand the scope of the problem.
3. Prevention of SCD crisis with hydroxyurea and pneumococcal vaccine.
4. Early implantation and treatment of those in crisis.



IV. Sickle Cell Epidemiology and Presentation of the Target Area

The Chetti population of around 5000 in the area is considered non-tribal, though the prevalence of sickle cell anemia is greater than that of resident tribal populations. Our hemoglobin electrophoresis analysis of the cohort of tribals and non-tribals in the area revealed that 719 out of 3730 individuals (19.2%) carried the sickle cell trait, while 91 individuals had the disease (2.43%). The higher prevalence of the sickle cell trait among the Chetti's may be a result of a higher frequency of consanguineous marriages within the relatively small community.

Our analysis by tribe (table 2) in 2005 revealed that the Irula tribe was found to have the highest rate of sickle cell disease (14.2%) while the Bettikorumba tribe demonstrated the lowest disease rate. The Mullukurumba tribe demonstrated the highest carrier prevalence (29%), while the Kattunaickken tribe demonstrated the lowest carrier prevalence of (9.4%).

Table 3 illustrates gender breakdown of sickle cell patients at the Gudalur Adivasi Hospital, while table 4 details the common clinical presentations of the sickle cell patients in question.

Table 1 - Overall Distribution of Sickle Cell Disease and Trait among Cohort (n=3730)

	Trait (HbAS)	Disease (HbSS)
Total (n=3730)	719 (19.2%)	91 (2.43%)

Table 2 - Distribution of Sickle cell Trait (n=299) and Sickle cell Disease (n=46) by Tribe

The Irula tribe was found to have the highest rate of sickle cell disease (14.2%) while the Bettikorumba tribe demonstrated the lowest disease rate. The Mullukurumba tribe demonstrated the highest carrier prevalence (29%), while the Kattunaickken tribe demonstrated the lowest carrier prevalence of (9.4%).

Tribe	No. Sickle cell Trait (% of respective tribe screened)	No. Sickle cell Disease (% of respective tribe screened)
Bettikorumba	47 (11.5%)	6 (1.4%)
Irula/Kota	36 (17.1%)	5 (14.2%)
Kattunaickken	17 (9.4%)	3 (1.6%)
Mullakurumba	43 (29.0%)	6 (4.1%)
Paniya	186 (21.1%)	26 (3.0%)

Table 3 - Age and Gender Wise Percent Distribution of Patients with Sickle Cell Disease (n=46)

Of the sickle cell patients under study, the highest concentration fell in the 5-10 year age group for males (35%), and the 11-20 year age group for females (17.4%).

Age Interval	No. Male (%)	No. Female (%)
1-5	4 (8.7)	2 (4.8)
5-10	16 (35)	1 (2.1)
11-20	5 (11)	8 (17.4)
21-30	1 (2.1)	3 (6.5)
31-40	1 (2.1)	1 (2.1)
41-50	2 (4.8)	2 (4.8)

Table 4 - Percent Distribution of Common Clinical Features among Sickle Cell Tribal Patients in the Inpatient Setting (n=21)

Fever (86%), epigastric tenderness (67%), and splenomegaly (52%) were the predominant clinical features displayed by the sickle cell patients, while systolic murmur and tachycardia were found to be the least frequent clinical feature (19%) displayed.

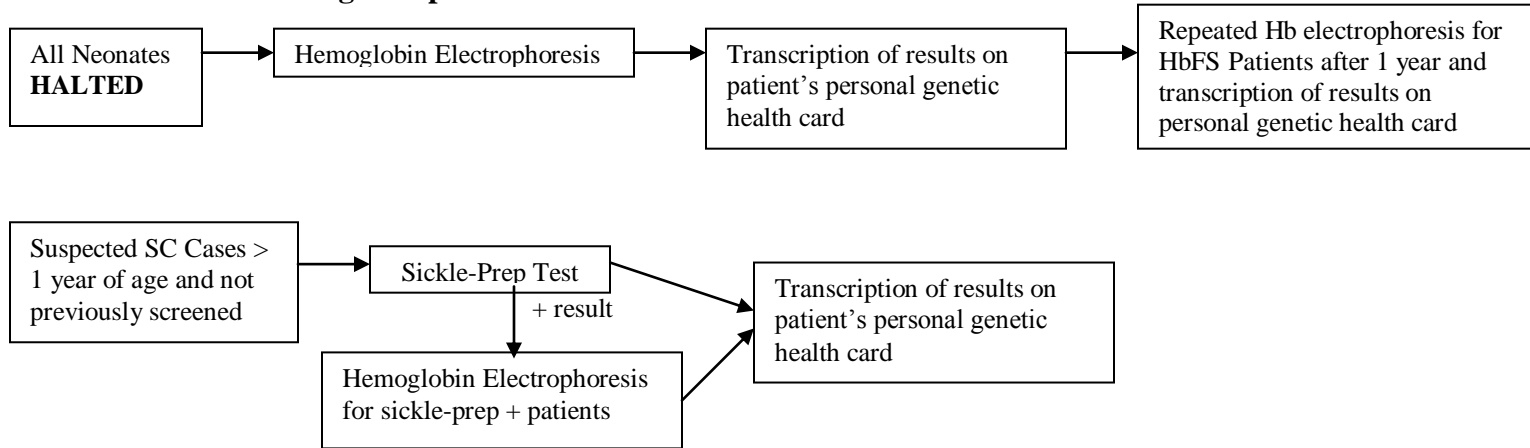
Clinical Feature	Percentage of SCD Patients Displaying Feature
Fever	86%
Epigastric Tenderness	67%
Splenomegaly	52%
Pallor	38%
Edema	29%
Jaundice	29%
Respiratory Infection	24%

Systolic Murmur or Tachycardia	19%
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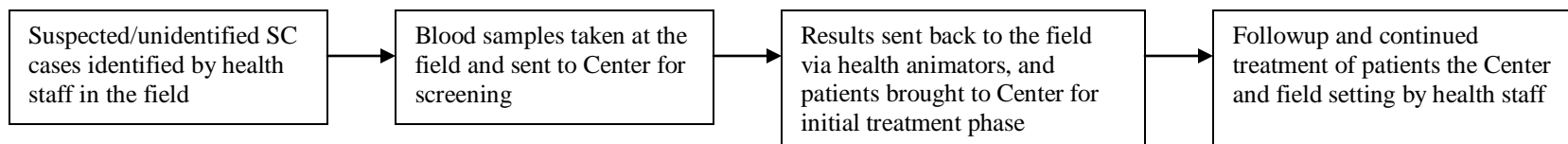
V. Services of the Sickle Cell Disease Center

Sector I: Clinical Services

A. Center-Based Screening Component

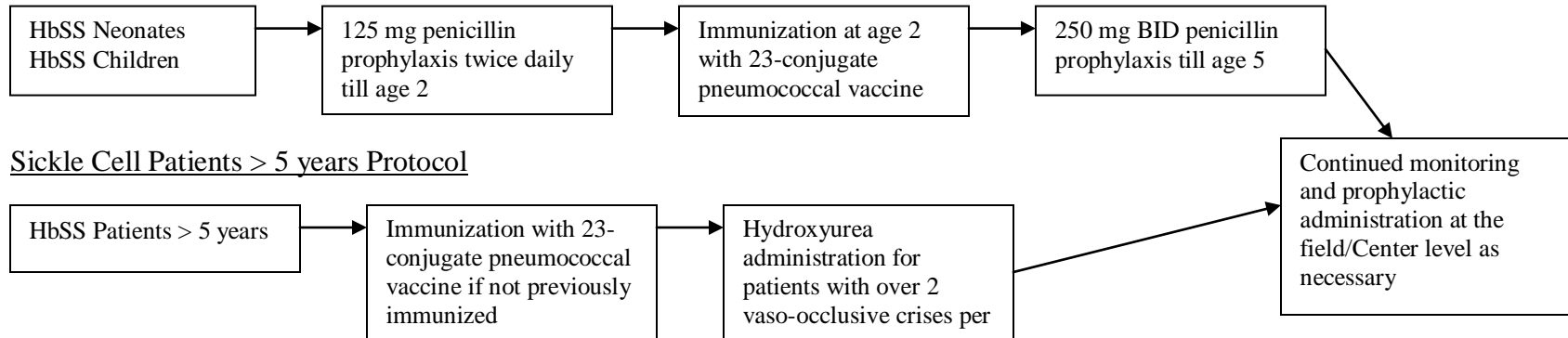


B. Field-Based Identification and Monitoring Component

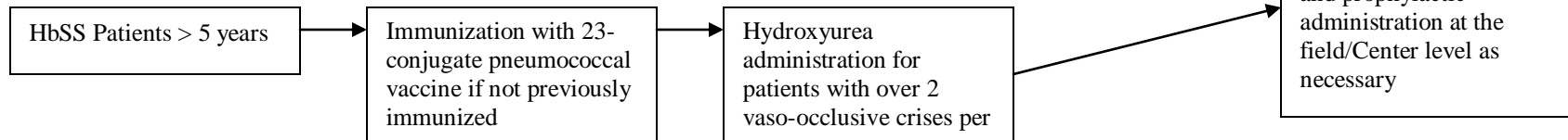


B. Center-Based Treatment Component for Sickle Cell Patients with Field Monitoring

Neonatal and Children < 5 years Protocol

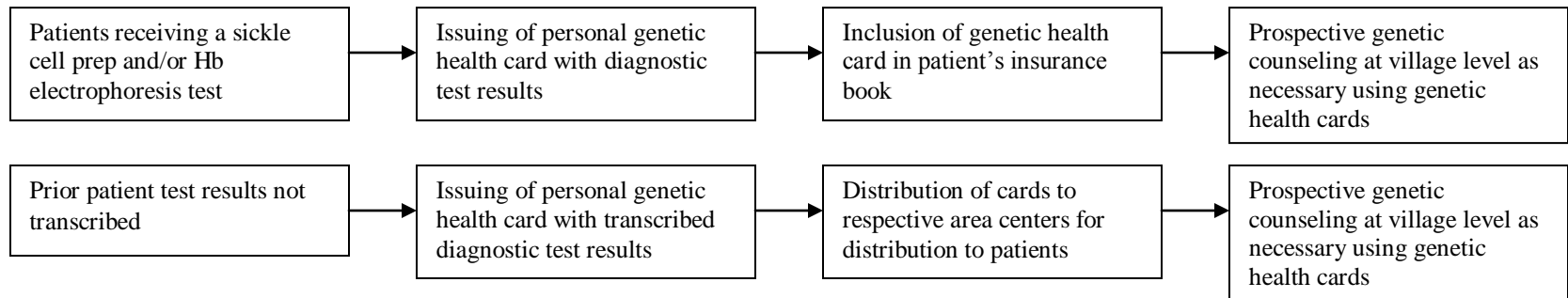


Sickle Cell Patients > 5 years Protocol

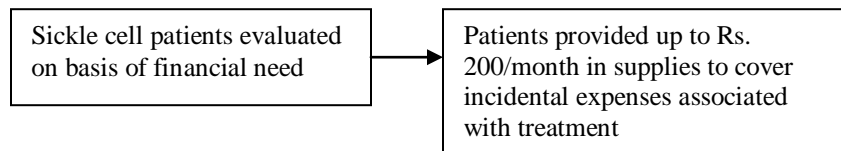


Sector II: Community-Based Services

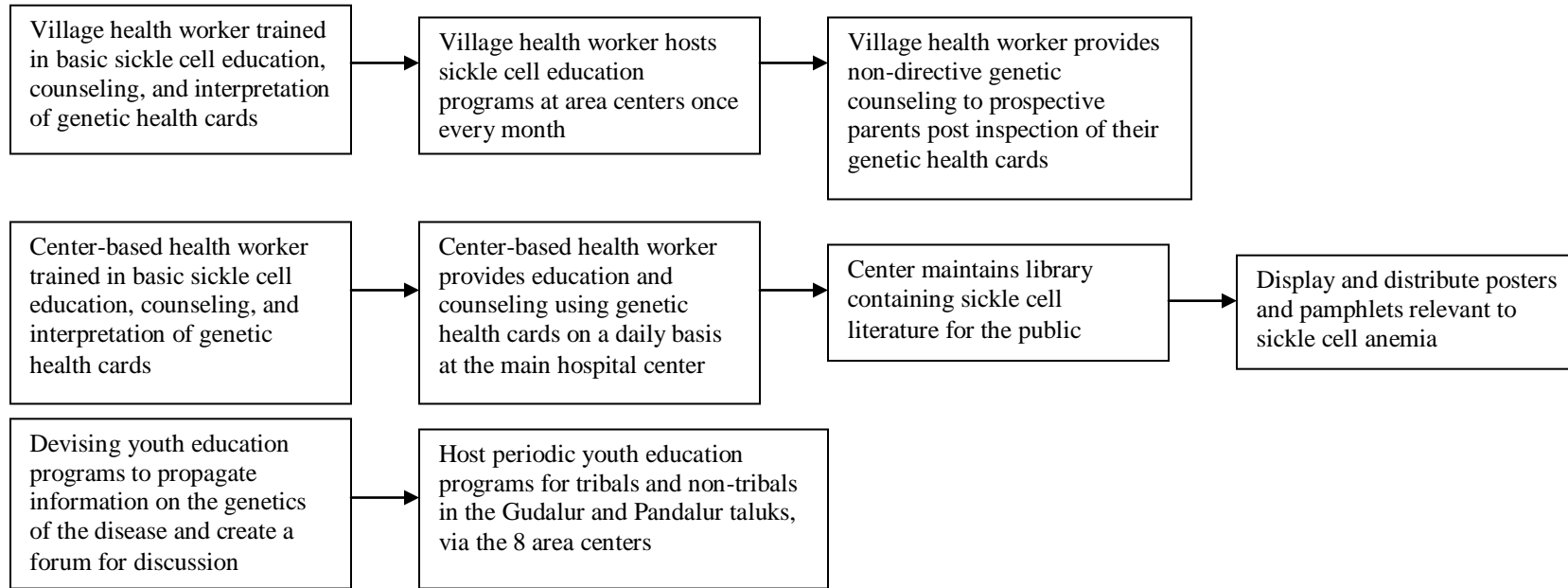
A. Deployment of Genetic Health Cards



B. Sickle Cell Disability Fund

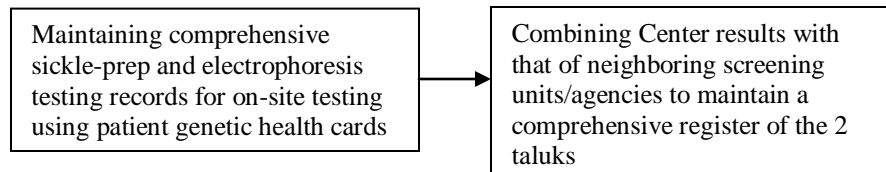


C. Prospective Genetic Counseling and Sickle Cell Education Programs



Sector III: Research & Documentation Services

A. Genetic Documentation



B. Research Topics

- ❖ Testing plant-based antisickling agents for sickle cell disease

- ❖ Evaluating the efficacy and appropriateness of common sickle cell treatment mechanisms among the resident tribal and non-tribal populations
- ❖ Investigating the economics of sickle cell screening and treatment in the area
- ❖ Pursuing active epidemiological studies to determine the rates of sickle cell trait and disease in the target area
- ❖ Investigating community response, attitudes, and knowledge with respect to sickle cell anemia and its associated intervention
- ❖ Health outcomes both mortality and morbidity patterns among resident tribal and non-tribal populations of the Nilgiris

Sector IV: Material and Monetary Resources

A. Patients

- ❖ Tribal and non-tribal patients charged on a sliding scale for screening and treatment of sickle cell disease
- ❖ Counseling and educational services provided by the Center for no cost

B. Government

- ❖ Potential funding for screening, treatment, prophylactics, vaccines, and hydroxyurea
- ❖ Potential funding for education programs and educational materials
- ❖ Potential funding for administrative and infrastructural costs incurred by the Center

C. Research Grants

- ❖ Institutional grants (ICMR etc...) used to further research activities as outlined by the Center
- ❖ Opportunities for medical professionals to conduct on-site research in collaboration with the Center

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Nilgiri District, South India

D. Non-Profit Organizations

- ❖ Funding used to further both clinical and community-based services provided by the Center

E. Medical and Public Health Professionals

- ❖ Expertise used to further the scope and quality of services provided by the Center

VI. Collaborators

The Sickle Cell Disease Center aims to collaborate with the Tamil Nadu and Central Government, and neighboring non-governmental organizations conducting sickle cell-related work in the Nilgiris and other parts in India. Active non-governmental organizations of the area working in the health sector include:

- Swami Vivekananda Medical Mission
- National Adivasi Welfare Association (NAWA)
- Nilgiri Wayanad Tribal Welfare Society
- University of Illinois at Chicago Sickle Cell Disease Center
- American Tamil Medical Association
- NetMargins

In forming active partnerships with these organizations, a sickle cell disease committee may be established to monitor the management of sickle cell disease in the area, allocate government and private funding for sickle cell anemia, and ensure maximum utilization of sickle cell funding for each organization. Additionally, the committee can function as a central body through which all sickle cell funding requests may be routed, in order to minimize duplication of requests and streamline area operations.



Sickle Cell Disease Center
Gudalur Adivasi (Tribal) Hospital
www.tihf.org/scd.htm

Patient Data:

Number of Individuals Screened	6472
Number of Tribal Patients with Sickle Cell Disease	99
Number of Chetty Patients with Sickle Cell Disease	77
Number of Sickle Cell Patients Vaccinated with Pneumococcal Vaccine	51
Active Patients	185

Comprehensive Screening Data (Center + Villages):

Total Individuals Screened	% Trait (HbAS)	% Disease (HbSS)
6472	14.71%	3.01%

Village Screening Data:

Total Individuals Screened	% Trait (HbAS)	% Disease (HbSS)
2693	11.06%	.52%

Year Screening Data (November 2007-November 2008):

Number of Screening Camps	Number of Villages Covered	Total Number of Individuals Screened (Hospital and Field)	% Trait (HbAS)	% Disease (HbSS)
53	76	2026	9.6%	2.4%

Medication/Supply Stock:

<u>Amount of Hydroxyurea (500 mg)</u>	<u>3200 capsules</u>
<u>Number of Pneumococcal Vaccines (Pneumovax)</u>	<u>14 vaccines</u>

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Sickle Cell Inpatients Admissions:

<u>2003</u>	<u>2004</u>	<u>2005</u>	<u>2006</u>	<u>2007</u>	<u>2008</u>
<u>10</u>	<u>13</u>	<u>26</u>	<u>37</u>	<u>36</u>	<u>55</u>