SEWA Rural
Sickle Cell Comprehensive Care Program

BACKGROUND

Sickle cell disease (SCD) is the most prevalent worldwide inherited blood disorder, characterized by sickling of red blood cells leading to complications due to vaso-occlusion and anemia (1). In India, sickle cell disease is associated with significant morbidity and mortality and represents a major public health problem. India is one of the three countries in the world, along with Nigeria and DR Congo, that claims 50% of the world’s sickle cell disease burden (2). The prevalence of sickle cell trait among tribal populations in India varies from 5-34%, with an estimated 18 million Indians with sickle cell trait and 1.4 million with sickle cell disease [3].

Though there is limited nationwide data on outcomes of SCD in India, several studies have documented significant morbidity and mortality from the disease. According to one survey by the Indian Council of Medical Research (ICMR), about 20 per cent of children with sickle disease in India died by the age of two, and 30 per cent of children with SCD in tribal communities die before they reach adulthood [4,5]. Several studies have found severe disease manifestations in 20-50% of SCD populations studied, the majority presenting with severe anemia, acute febrile illnesses, and frequent vaso-occlusive crises [6,7,8,9].

Several simple, cost-effective interventions have been found to greatly improve outcomes in sickle cell disease, including: early diagnosis by neonatal screening, education of patients and families about early recognition of complications, preventive care with prophylactic penicillin and pneumococcal vaccination, regular follow-up and treatment with transfusion and hydroxyurea for severe cases. Despite significant evidence of efficacy, access to these services is limited, especially among tribal communities, traditionally among the most underserved populations in India, that bear the highest burden of sickle cell disease.

There has been recent interest from the Indian government to further sickle cell disease control in endemic areas. The state of Gujarat was the first state in India to implement a Sickle Cell Anemia Control Programme in 2006. In Gujarat, it is estimated that approximately 900,000 tribal people have sickle cell trait and approximately 70,000 people are affected by sickle cell disease [10]. Through the screening program in Gujarat, 5.5 million people have been screened and over 29,000 diagnosed with sickle cell
disease to date. However, coverage of proven interventions among individuals with sickle cell disease seems to be inadequate.

SEWA RURAL

SEWA Rural is a non-profit health and development organization in Jhagadia, South Gujarat, India. It was established in 1980 by a group of physicians and young professionals to alleviate poverty and inequality in the surrounding tribal population with previously limited access to comprehensive healthcare. SEWA Rural operates the local health delivery system, consisting of a 100 bed Kasturba Hospital and OPD clinics, comprehensive eye care program, women empowerment activities, vocational training center, health training center and robust community health worker outreach program to promote a comprehensive health care system. Citizens from the surrounding 1500 villages benefit from these activities. Over the past 35 years, SEWA Rural has had a tremendous impact on improving health outcomes of the community including: a 90% reduction in maternal mortality and 75% reduction in neonatal mortality. So far, more than 100,000 individuals have regained their vision through cataract and other ophthalmologic operations. Almost 2,500 young tribal youth have received employment. (11).

Sickle cell disease is a pervasive problem for the population served by SEWA Rural due to its high prevalence in this mostly tribal area. Many patients are diagnosed when they present with severe complications, and have significant rates of morbidity and mortality. To address the burden of sickle cell disease in the tribal population, we sought to implement a Comprehensive Sickel Cell Program at SEWA Rural.

Figure 1. Distribution of HbS trait in Gujarat - % of allele (12)
SC - scheduled castes, ST - scheduled tribes, GC - general castes
AIMS OF SEWA RURAL SICKLE CELL PROGRAMME

To improve early diagnosis, linkage to care and treatment for patients with sickle cell anemia in order to reduce preventable morbidity and mortality and improve quality of life for sickle cell disease patients through:

1. Development, implementation, and evaluation of a comprehensive sickle cell demonstration program at SEWA Rural that includes screening, community-based, outpatient, and inpatient care interventions.

2. Support development and evaluation of a scalable model of care for sickle cell disease across districts of Gujarat state with significant tribal populations, in partnership with agencies involved in implementation and research at the state level.

SEWA RURAL COMPREHENSIVE SICKLE CELL PROGRAMME COMPONENTS

Patients with sickle cell disease often are diagnosed when they present with severe complications in the hospital. Early diagnosis, linkage to care with close follow-up and monitoring can improve outcomes of sickle cell disease. The SEWA Rural Sickle Cell Program is designed to implement evidence-based interventions like newborn screening for early diagnosis, penicillin prophylaxis, pneumococcal vaccination, and hydroxyurea that have been shown to dramatically reduce morbidity and mortality from sickle cell disease.

1. SCREENING

    Neonatal screening, when linked to active follow-up, parental education, and comprehensive care, has been shown to markedly reduce morbidity and mortality from SCD in infancy and early childhood [1,2]. Universal neonatal screening with close monitoring after diagnosis has become standard of care in the US and Europe. Neonatal screening programs are underway in India, and newborn screening is recommended by the Gujarat Sickle Cell Anemia Control Project, World Health Organization 59th Assembly, and numerous other guidelines in resource-limited settings [2,10,13].

    At SEWA Rural, universal maternal screening for sickle cell disease was implemented in 2011. All women followed in the antenatal clinic or who are admitted and deliver at the hospital are screened for sickle cell disease. Starting in 2014, newborn screening was implemented in partnership with Valsad Raktadan Kendra for any birth from a mother with sickle cell trait or disease. All family members of sickle cell disease cases are also provided testing. Implementation of universal maternal screening and newborn screening for babies of mothers affected by SCT or SCD has led to the diagnosis of over 200 cases of sickle cell disease at SEWA Rural. These patients are now being followed by the comprehensive care program, as described below.

2. OUTPATIENT SICKLE CELL CARE

    Active follow-up, close monitoring, and preventive interventions are essential to achieving the morbidity and mortality benefits of early diagnosis through newborn screening [14]. In 2014, we established a weekly sickle cell disease outpatient (OPD) clinic and disease registry to track all patients who test positive for sickle cell disease. Antenatal, newborn, pediatric and adult medicine patients are followed in the sickle cell OPD. Upon enrollment, patients are entered into the sickle cell registry and followed with clinic visits where they receive evidence-based interventions. The clinic and disease registry are staffed by one physician and one nursing assistant/health counselor.

    Patients have visits at least every 3 months where they receive treatment and monitoring that includes: symptom review and examination for disease complications, hemoglobin and laboratory monitoring, pneumococcal vaccination, daily folic acid, and malaria prophylaxis (FIGURE 2). Patients under age 5 receive daily penicillin prophylaxis. Patients who meet criteria for severe illness are started on fixed low-dose hydroxyurea and followed according to protocol [15, 16]. See appendix 1 for detailed outpatient protocols.

3. INPATIENT SICKLE CELL CARE

    Patients with sickle cell disease are prone to developing severe complications that require inpatient management including vaso-occlusive crises, acute febrile illness, sepsis, severe anemia, acute chest syndrome, stroke, and acute splenic sequestration. To standardize care delivered for acute complications in accordance with national guidelines, we have developed inpatient protocols for management of sickle cell crises for maternal, pediatric, and adult medicine wards to guide management of vaso-occlusive crises, acute febrile illness, acute chest syndrome, and splenic sequestration. See appendix 2 for detailed inpatient protocols.

Figure 2: Components of the Sickle Cell Programme
4. CHRONIC DISEASE REGISTRY AND POPULATION MANAGEMENT

All sickle cell disease patients are entered into the disease registry, which includes information on demographics, disease history, laboratory results, treatments, complications, transfusions, clinic visits, hospital admissions, and mortality. The registry is used to follow quality metrics for performance improvement, and generate lists of patients overdue for routine visits, monitoring, or interventions. The sickle cell counselor conducts outreach to patients who are overdue for care. See appendix 3 for detailed sickle cell disease registry.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Age</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pneumococcal vaccine PSV23</td>
<td>2yr, 7yr</td>
<td>5yrs x 2</td>
</tr>
<tr>
<td>If available, PCV13 vaccine</td>
<td>2mo, 4mo, 6mo, 15 mo</td>
<td></td>
</tr>
<tr>
<td>Penicillin prophylaxis</td>
<td>2mo-5 yrs</td>
<td>Daily</td>
</tr>
<tr>
<td>Folic acid</td>
<td>all ages</td>
<td>Daily</td>
</tr>
<tr>
<td>Malaria prophylaxis</td>
<td>all ages</td>
<td>Daily</td>
</tr>
<tr>
<td>Hydroxyurea</td>
<td>&gt;9mo if indicated</td>
<td>Daily</td>
</tr>
<tr>
<td>CBC</td>
<td>all ages</td>
<td>0.3mo, 0.1mo if on hydroxyurea</td>
</tr>
<tr>
<td>Liver and kidney function tests</td>
<td>all ages</td>
<td>0.12mo, 0.03mo if on hydroxyurea</td>
</tr>
</tbody>
</table>

Figure 3. Outpatient clinic treatment protocol

5. SICKLE CELL COUNSELING AND HEALTH EDUCATION

The sickle cell counselor provides patients and their families with culturally appropriate counseling about the disease process, warning signs of illness, when to seek prompt treatment, and encourages family members to be tested. She staffs a dedicated sickle cell mobile telephone line that patients can reach her with symptoms or concerns. Maintaining the sickle cell registry is one of her primary responsibilities. She also conducts outreach to patients who do not come for regular appointments. She is notified of any new sickle cell diagnoses or when established patients are admitted to enroll patients in the disease registry and provide health education and counseling. See appendix 4 for sickle cell patient education brochure.

COMMUNITY BASED INTERVENTIONS

Future community-based interventions include expansion of our community engagement to include home-based outreach by sickle cell counselors who would provide outreach for screening results, proactive follow-up and tracking of sickle cell disease patients, health education and counseling, early identification of warning signs and referral to outpatient and indoor care as needed. These activities may involve application of SEWA Rural’s innovative mobile technology platform to support sickle cell outreach workers.

MEENA’S STORY

Meena is a 7 year-old girl with sickle cell disease who was hospitalized three times in the past year for severe sickle cell crises, requiring over 10 blood transfusions. She frequently missed school and was unable to do much physical activity without precipitating a pain crisis a year ago. Her family has dealt with pain and death from the illness. Her mother also has sickle cell disease and has been hospitalized many times, and Meena had two older siblings who both passed away as infants from febrile illnesses at home. Presumably, they also had sickle cell anemia putting them at increased risk for serious infections, but had never been tested or received treatment for sickle cell disease. When Meena presented to the SEWA Rural Kasturba Hospital last year, she had severe anemia with a hemoglobin of 4.8 mg/dL and debilitating pain. She slowly improved with treatment and transfusion, and was enrolled in the sickle cell registry on discharge. At her first sickle OPD clinic visit, she was started on hydroxyurea for her frequent crises. Since then, her life has opened up. Her hemoglobin has stabilized and she has had no further pain crises in the past 6 months. She now attends school regularly and is able to play with friends without fear of pain.
PROGRESS TO DATE
1. Initiated universal maternal and newborn screening and testing of all family members of sickle cell patients.
2. In 2014, screened 5812 patients and diagnosed 132 patients with sickle cell disease, including 17 newborns.
3. Enrolled over 230 patients with sickle cell disease for prospective follow-up and comprehensive care and treatment.
4. Implemented a weekly sickle cell OPD clinic staffed by physician and sickle cell counselor.
5. Implemented sickle cell disease registry to prospectively follow clinical outcomes of sickle cell cohort.
6. Over 160 SCD patients have been vaccinated for pneumococcal disease.
7. Approximately 10 SCD patients with severe disease have been started on hydroxyurea.
8. Provided counseling and health education on sickle cell disease to over 200 patients and families.

RESEARCH AND DISSEMINATION
We are evaluating the impact of the SEWA Rural comprehensive care program on health and psychosocial outcomes among the sickle cell disease cohort, in order to elucidate the most effective interventions. We plan to conduct community-based research to identify gaps in current care and implementation barriers to scaling up a comprehensive sickle cell care program in the broader community. Based on this research, we plan to develop interventions to improve coverage and quality of proven interventions for scaleup of a comprehensive sickle cell care model in wider area.

CONTACT INFO
SEWA Rural, Jhagadia, Dist: Bharuch, Gujarat, 393110
Phone number: 091-02645-220021
Fax: 091-02645-220313
Website: http://sewarural.org
Email: sewarural@gmail.com
Dr. Gayatri Desai: shreygayatri@gmail.com
Head, Obstetrics and Gynecology Department, SEWA Rural
Dr. Reena Gupta: reena.gupta@ucsf.edu
Assistant Professor of Medicine, University of California, San Francisco

ACKNOWLEDGEMENTS
Valsad Raktadan Kendra
Department of Health and Family Welfare, Government of Gujarat

APPENDICES
1. Outpatient sickle cell clinic protocol
2. Inpatient sickle cell protocols
3. Disease registry format
4. Sickle cell patient education brochure
REFERENCES


