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# Comprehensive Integrated Care for Patients With Sickle Cell Disease in a Remote Aboriginal Tribal Population in Southern India

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***Background.*** Healthcare delivery for sickle cell disease (SCD) can be challenging, in low resource settings. We studied the feasibility of delivering comprehensive SCD care in a community-based network for remote, economically, and socially disadvantaged tribes in Gudalur, India. ***Procedure.*** We reviewed medical records for all patients followed at the Gudalur Adivasi Hospital. We used published quality of care indicators to benchmark screening and routine healthcare maintenance. ***Results.*** We screened 9,646 individuals (60.4%) under the age of 30 of a population of 25,000 individuals. Of 111 active patients with SCD, 71% have had at least one annual comprehensive clinic visit at a median visit interval of 57 days. We provided pneumococcal immunization and penicillin prophylaxis to 56 (50%) patients and HU to 68 (61%). Median spleen size was 1 cm (range 1–6 cm), mean was Hb 9.3 g/dl and we reported a mean of 0.7 painful episodes/year. Premature deaths occurred in 19 patients at a median age of 23 years due to acute chest syndrome, sepsis, severe anemia, stroke, mesenteric infarction, puerperal sepsis, or sudden unexplained death. Healthcare maintenance met 11 of 17 published SCD quality of care indicators. Average cost was 1,343 Indian Rupees (INR) (approximately US$ 25) per hospitalization and 173 INR (approximately US$ 4) per clinic visit. ***Conclusion.*** High quality comprehensive care for SCD can be delivered for a low income, aboriginal population in India through a community driven network of care. This model can serve as a template for healthcare delivery for SCD in low-income communities. Pediatr Blood Cancer 2014;61:702–705. 

**Key words:** community based; comprehensive care; India; sickle cell disease; tribal

## INTRODUCTION

Sickle cell disease (SCD) is an autosomal recessive disorder caused by a point mutation in the beta globin gene and characterized by vasoocclusion and hemolysis of red blood cells. Disease complications such as anemia, vasocclusive crises, and ischemic injury to multiple organs, impair quality of life and contribute to premature mortality. Advances in SCD care, such as newborn screening, pneumococcal prophylaxis, comprehensive care and hydroxyurea (HU) therapy have improved survival [1]. Substantial barriers remain in the delivery of care, even in economically advanced countries [2]. Recently published indicators of quality of care can serve as benchmarks for care for patients with SCD [3]. SCD is a significant public health problem in India, with over 5200 affected newborns annually [4,5]. India is a large country with numerous ethnic groups wherin SCD is prevalent in numerous ethnic groups, in India. The high prevalence of sickle cell in several distinct groups in India has been the subject of extensive population based studies [6–18]. Among the major ethnic groups with a high burden of SCD are autochthonous tribal peoples known as scheduled tribes or Adivasis. Based on the criteria of the presence of distinctive history, geographical isolation, distinct culture, shyness of contact with the community at large, and economic backwardness, the constitution of India identifies these population groups as scheduled tribes and incorporates several special provisions for the promotion of their educational and economic interests as well as for their protection from exploitation [19]. The scheduled tribes consist of over 645 distinct ethnic groups consisting of over 84 million individuals representing 8% of India's population [20]. Differences in ethnic origins, genetic background, and migration histories of these endogamous tribes, who serve as genetic isolates may explain the variation in the sickle cell trait carrier frequency of 10–33% in these populations [6,8,9,11,12,15–18,21]. In many tribal communities, patients with SCD do not receive comprehensive care for SCD due to the lack of access to health care, socioeconomic deprivation, and geographical and cultural isolation. This study was undertaken to determine the feasibility and sustainability of the delivery of high quality comprehensive care for SCD in extremely marginalized communities in a low-income country.

## METHODS

Gudalur and Pandalur are in the Nilgiris Mountains, in the southern state of Tamil Nadu and have dense forest coverage [20]. Five different Adivasi Tribes (Paniyas, Kattunayakans, Bettakurumbas, Mullukurumbas, and Irulas) with a population of 25,000 scattered in over 300 small hamlets make up approximately 10% of the regional population. ACCORD (Action for Community Organization, Rehabilitation and Development), a non-governmental organization (NGO), was established in 1985 and has been responsible for community organization to secure human rights, health, education, housing, and culture of the tribals. In 1988, the tribals organized Adivasi Munnetra Sangham (Tribal Advancement Council) a representative body of tribals consisting of tribal elders from each tribal hamlet. The Tribal council is the body responsible for making all major decisions related to the welfare of the tribals. It has created and oversees organizations for providing education, healthcare, and agricultural and marketing cooperatives. The Association for Health and Welfare in the Nilgiris (ASHWINI) was established by the Tribal Advancement Council in 1990 to

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provide primary and secondary healthcare for the Adivasis. The executive committee for ASHWINI consists of members of the tribe with a physician serving in an advisory capacity (Fig. 1). We have trained members of the tribal populations as paramedics, nurses, pharmacists, laboratory technicians, and community health workers. Each hamlet has a trained Adivasi Village Health Volunteer who encourages mothers to get their children immunized, pregnant mothers to go for pre-natal checkups, and patients with chronic illnesses to take their medications. Most of the volunteers are women because they were more likely than men to be culturally acceptable in this role. She also identifies and refers patients who are ill, helps Community Health Nurses organize village clinics and maintains an inventory of some basic medications. Eight area centers are each manned by trained Adivasi Community Health Nurses. Community Health Nurses undergo a rigorous 4-year training program including medical, surgical, and community health nursing. They record vital signs, give injections, insert intrauterine devices (IUDs), provide health education, perform antenatal examinations, diagnose and treat common illnesses, provide primary, antenatal and under five care, dispense medicines for chronically ill patients, and conduct village health visits with the Village Health Volunteers. The Adivasi tribal communities own and operate the 40-bed Gudalur Adivasi Hospital (GAH) for secondary care. Most of the staff members are adivasi tribals. GAH has a labor room, an operating room, and a pharmacy stocked with generic medicines. It provides secondary inpatient and outpatient care. We established an electronic health database in 1997. Adivasi families contribute to a group health insurance scheme that covers inpatient, outpatient and prescription benefits and supplements government support for care. We established a comprehensive sickle cell program in 1997. We initiated use of HU and pneumococcal immunization starting in 2005. We have implemented regular community screening starting in 2007. Community Health Nurses assisted by Village Health Volunteers conduct health education sessions followed by mass screening. They screen all individuals 30 years of age and under in the villages with the sodium metabisulfite test [22]. All individuals with positive screening tests undergo cellulose acetate hemoglobin electrophoresis at GAH. The community health nurse gives every family a card with the results of the testing. Those detected to have the sickle cell trait or disease receive genetic counseling through the SCD Centre at GAH. The SCD Centre also facilitates health education and comprehensive care. The Village Health Volunteers and Community Health Nurses provide health education about SCD to patients and family members. Children with SCD under the age of five receive oral penicillin prophylaxis and pneumococcal vaccines. Patients come to the hospital or area centers for monthly check-ups. We treat patients for vasocclusive crises using an established protocol with non-steroidal anti-inflammatory drugs or opioids as needed. Initially, we prescribed HU only to patients who had a history of

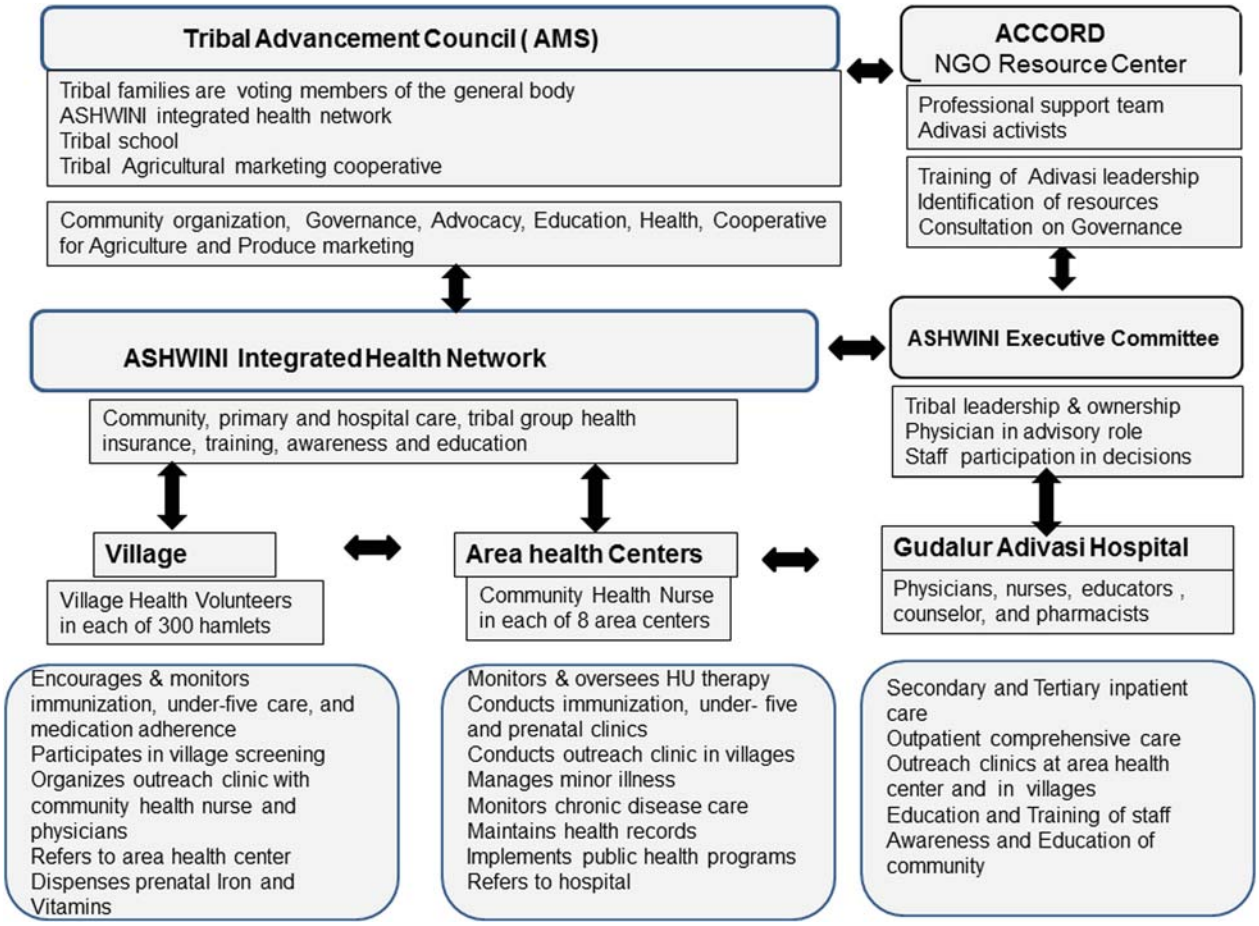


**Fig. 1.** Organizational chart for sickle cell program integrated into network of care.

two or more crises a year. Following the observation of the safety and efficacy of low fixed dose HU [23], we subsequently offered HU to all patients with a lifetime history of a single vasocclusive crisis. We provided each family with a notebook containing records of all health related activities. Community Health Nurses record key health related details, such as live births, deaths, visits to the hospital, blood collection dates, and medications and vaccines provided. We maintain clinical, healthcare utilization and financial data in computerized databases. The affirmative action programs of the Government of the State of Tamil Nadu gives members of this community access to government run educational institutions and to government jobs. The government of Tamil Nadu has identified tribal health as a priority area and recognizes SCD as a special health issue within the scope of tribal health. The government supports outreach services in tribal areas under the aegis of the Tamil Nadu Health Systems project. This project provides financial support for the population screening, free outpatient medication including HU and 23 valent Pneumococcal vaccination, costs of inpatient care at Gudalur Adivasi Hospital as well as supporting village outreach clinics (the operation of a Mobile outreach Van).

## RESULTS

### Health Outcomes

Since the establishment of ASHWINI, there has been a substantial improvement in healthcare indicators for the tribal population. We compared the change in vital health statistics from 1997 to 2006 for Gudalur tribal population to that for the populations of India, Kenya in sub-Saharan Africa, and the United States. The dramatic improvement in health outcomes in Gudalur is evident from the change in infant mortality rate (Gudalur 250/1,000 to 27/1,000; India: 76/1,000 to 50/1,000, Kenya: 61/1,000 to 79/1,000, USA: 8/1,000 to 7/1,000), and maternal mortality rate (Gudalur 630/100,000 to 223/100,000; India: 570/100,000 to 540/100,000, Kenya 650/100,000 to 1,000/100,000, and USA 12/100,000 to 17/100,000) [24–26]. Immunization rates in Gudalur which were an abysmal 5% in 1996 also increased sharply to become comparable with advanced health systems (Gudalur 88%, India 71%, Kenya 83%, and USA 92%) [23,24].

### Sickle Cell Trait Screening Program

Since the establishment of the SCD program, we have screened a total of 9,646 Adivasis, that is, 60.4% of the total eligible population. We identified, 1,089 individuals (11.3% of those screened) as being sickle cell trait carriers. We provided genetic counseling to all of these patients. We detected a total of 137 individuals with SCD indicating disease prevalence of 1.4%.

### Clinical and Laboratory Features and Outcomes

All patients had Hb SS. Median age of patients was 14 years. Patients reported a mean of 0.7 painful episodes/year. Examination revealed median spleen size of 1 cm (range 1–6 cm); mean Hb 9.3 g/dl. There have been 19 deaths from 1997 to 2012, and 9 patients have moved away from the area. Median age of death was 23 years. Cause of death was acute chest syndrome in five patients, sepsis in two, acute severe anemia in two, stroke in one, mesenteric infarction in one, and puerperal sepsis in two and sudden unexplained death in six patients. We provided pneumococcal immunization and penicillin prophylaxis to 56 patients starting at a median age of 2 years prior to the implementation of newborn screening. Of the 111 patients, 68 (61%) are regularly taking HU. Healthcare maintenance for SCD patients at GAH meets 11 of 17 quality of care indicators for routine healthcare maintenance for SCD [27] (Supplemental Table I).

### Healthcare Utilization and Costs of Care

Of 111 active patients, 71% had a visit to the health center at least once in the last 12 months. Patients had a median of seven healthcare visits annually at an average interval of 57 days. Between November 2007 and October 2010, there were 97 hospitalizations for Adivasi patients related to SCD. The average cost of hospitalization is 1,343 Indian Rupees (INR), which is approximately US$ 25. The average cost of an outpatient visit is 173 INR (US$ 4). We provided Pneumococcal vaccine (1,000 INR, US$ 22), free of cost to the patients. All oral medications have a unit cost of 0.30–5 INR (US$ 0.04–0.11). The approximate cost of HU is 150 INR (US$ 3) per month. The median income of a tribal family in 2004 was INR16, 500/year as compared to a median household income in the United States in 2004 of US$ 44,684 [28]. Thus, cost of care when adjusted for income and buying power of the tribal family is US$ 360 a month for HU, US$ 480 for a clinic visit and US $ 3000 for a hospitalization. The median annual cost of clinic visits and HU would represent approximately 18% of the average income of a tribal family. All costs of care including that of transportation and referral to tertiary care centers were and are currently being met through the tribal group health insurance and grants from the state government of Tamil Nadu.

## DISCUSSION

Substantial barriers remain in the delivery of comprehensive care and proven therapies for chronic diseases, even in developed western countries [2]. The availability and utilization of multidisciplinary specialty clinics for different genetic diseases in the United States is uneven. Effective national networks of specialty clinics exist for genetic disorders such as hemophilia and cystic fibrosis. These clinics reach large proportions of the target populations [29]. For other disorders, notably SCD, fewer such centers are available; centers are less likely to be networked, and they are used less widely [29]. Substantial barriers remain in the prescription of, and adherence to disease modifying therapy with HU, the only drug approved by the FDA specifically for SCD [30–33].

We describe the implementation of comprehensive SCD care within the framework of a community-based healthcare network for an extremely socially disadvantaged, economically low-income and geographically remote population in the Nilgiris mountains of southern India. The substantial improvement in access to health care and health outcomes is suggestive of the effectiveness of the network in delivering health care to this underserved population. Comprehensive care for SCD included high uptake of preventive immunization and disease modifying therapy with HU. The healthcare delivery system met the majority of health maintenance quality indicators for SCD [27,34]. The low cost of care and high adherence to clinic visits by the patients is evidence of the effectiveness of the program in providing access to care.

What then are the factors that contribute to the high rate of success in implementing a comprehensive network of care for the Adivasis peoples of the Nilgiris hills, and to what extent are the lessons learned from this population applicable in different populations elsewhere? One of the remarkable features of the Gudalur experience is that efforts to organize the community for the improvement of their social and economic status occurred simultaneously with the development of a network of healthcare. Health and societal issues are multi-dimensional; hence coordinated action in multiple domains may contribute to improvements in multiple areas [35]. Secondly, tribals drawn from the same communities own, administer and, for the most part, staff this modern health network. This is a remarkable achievement considering the extreme socio-economic and educational backwardness of this aboriginal population at the start of the project. These results suggest that even extremely disadvantaged groups can overcome socioeconomic and cultural barriers relevant to health care through community organization and ownership of the health agenda. The remarkable degree of community ownership and involvement in the planning, organization and staffing may have been a crucial ingredient for the success of this health initiative and perhaps the most important lesson that could potentially be translated to other low-income settings. This experience, however, is similar to that observed in other examples of sustained community engagement with a community development ethos, which have resulted in the overcoming of health disparities. Community involvement incorporates the concerns of the community rather than a purely biomedical ethos that may exclude or even marginalize the concerns of the community [36,37]. A third remarkable feature is the complete integration of neighborhood, primary, and secondary care into a network. This is in keeping with current recommendations to provide comprehensive patient-centered care in a Medical Home [38]. This community-based network of care has demonstrated effectiveness not only in the primary care but also in the care of a multisystem genetic disorder such as SCD. This supports the strength of the medical home model of care. This experience is also an example of a rapid improvement in healthcare indicators achieved by a disadvantaged community through the organization of community-based care. Finally, while this study was not designed to determine the natural history of SCD in India, it does provide some intriguing clinical insights. SCD has been considered milder in clinical presentation in India, especially among tribal populations [39,40]. Co-inheritance of alpha Thalassemia and the prevalence of the Arab-India beta globin haplotype may explain the supposed milder phenotype of SCD in India [41,42]. The high rate of pain crises, severe anemia, and premature death due to sepsis, acute chest syndrome, and other complications suggests that the phenotype of SCD in tribal populations in India may not be mild. We speculate that comprehensive care may even have uncovered some of the morbidity by improving early mortality. These data provide the rationale for further study of the phenotype of SCD in these populations.

In conclusion, these data demonstrate that high quality comprehensive care for SCD patients can be delivered in extremely low-income populations in the developing world in a community driven network of care. This experience may serve as a model for delivery of quality care for low-income populations in a variety of different settings.

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