

A Framework for Developing Comprehensive Sickle Cell Disease Programs for Native Americans



Overview, Considerations, and Recommendations



Hari Prabhakar
Hari_Prabhakar@hms.harvard.edu

Version 1.2

Introduction:

According to the US Census estimate in 2003, there are over 2.8 million individuals who identify themselves as American Indian and live in the United States. (Census 2003) There are currently 562 recognized tribes in the United States, the largest being the Navajo, Cherokee, Choctaw, Sioux, Chippewa, Apache, Blackfeet, Iroquois, and Pueblo. The three states with the largest Native American populations are California at 413,382, Arizona at 294,137 and Oklahoma at 279,559. (Census 2000, 2003) 106 of the original Native American languages are still spoken. While around 62% of Native Americans live in urban areas, the residual 38% live on sovereign reservations, trust lands, or areas of tribal jurisdiction or designation. (Hendrix 2008) Due to a combination of poverty, lack of mobility, certain endemic beliefs, and other barriers to healthcare access, a variety of health problems plague Native Americans. A 1990 census indicates that 84% of Native American “Elders” reported an income of less than \$20,000, which is the highest percentage of any ethnic group in the United States. (Hendrix 2008) A variety of issues ranging from infectious diseases among children to chronic disease prevalence, most notably diabetes and cardiac-related issues among adults, are matters of grave concern.

Sickle cell disease is also a matter of concern among those of Native American descent. It is estimated that the prevalence of sickle cell disease among those of Native American descent is around 36.2/100,000 live births, which makes it the third largest minority in the United States affected by the disease, behind African-Americans (289/100,000 live births) and Hispanics living in the Eastern states (89.1/100,000). (Olney 2000) Historically, intermarriage between Native Americans and Blacks in the United States, predominantly in the South, may have contributed to the gene prevalence. (Walton-Raji 2008) Recently, sickle cell disease was classified worldwide as a public health priority by WHO and UNESCO. However, a combination of lack of comprehensive data, infrastructural constraints, autonomy issues surrounding health systems in reservation areas, and cultural beliefs have hindered specific strategies tailored at assuring comprehensive care for sickle cell disease among Native Americans. Given the strong belief systems of Native Americans and the often rural nature of reservations from major centers providing sickle cell disease care, it is important that an effective management system be community-based and culturally sensitive. Based on our experiences in developing a sickle cell management program for indigenous populations in India, we provide the following set of recommendations to help facilitate the process. It is important that key stakeholders including the Indian Health Service, the Bureau of Indian Affairs, major hospitals with sickle cell care facilities, community representatives, and healthcare providers working in Native American areas work jointly to achieve these goals.

Action Plan:

1. Identify areas of high disease prevalence in Native American areas. This may be through a combination of existing epidemiological and statistical data, personal narratives of health professionals working in Native American areas, feedback from organizations working with Native Americans, and community members themselves. Cultural, linguistic, economic, and geographical variations between tribes and areas will have to be considered in planning the program and introducing awareness of the disease. Unfortunately, an accurate assessment of the burden of sickle cell disease amongst Native Americans is difficult due to lack of specificity in the newborn registry programs and in medical reports as to Native American ethnicity status. However, a national sample of hospital discharge information from 1993-2004 provides a preliminary look at areas where sickle cell disease may be most prevalent amongst Native Americans. More on-site data collection is important to fill in the current gaps and identify areas where care is needed most.

State	Frequency of Sickle Cell Hospital Discharges	% of Total Discharges
NJ	66	36.67
NC	42	23.33
FL	31	17.22
NY	11	6.11
MA	8	4.44
MD	6	3.33
PA	6	3.33
WI	4	2.22
SC	3	1.67
VA	2	1.11
CA	1	.56

- Engage with community members of affected areas regarding their perceptions of sickle cell disease. This is called delineating an explanatory model of illness, and requires asking members of the community their thoughts on the disease, its origins, endemic terminology for the disease, its mechanism, and any traditional strategies used for managing the disease. This will allow for planning of a culturally appropriate intervention strategy. Some general cultural attitudes in the Native American community are listed below, as prepared by Levanne Hendrix:

<u>AMERICAN INDIAN</u>	<u>EURO-AMERICAN</u>
1. Cooperation	1. Competition
2. Group Harmony	2. Individual Achievement
3. Modesty and Humility Physical modesty Not putting one's self forward Non-attention seeking behavior (except in Sports)	3. Overt identification of accomplishments Physical exhibition
4. Non-Interference	4. Advice giving, directiveness "Counseling" and "Educating"
5. Silence is valued Ability to listen and wait	5. Points made by aggressive verbal behavior, expression of opinion
6. Emotional Control Contemplation Non-demonstration of anger ☛ other strong emotion	6. Action over inaction Direct confrontation Direct expression of anger
7. Patience Group decision by discussion and consensus	7. Rapid responses Decision making Problem solving
8. Generosity and Sharing Material possessions given away Respect earned by giving rather than saving Upward mobility within non-Indian society not sought	8. Individual Ownership Amassed material property Upward social mobility
9. Indifference toward future planning Saving for one's own benefit not accepted Planning for future generations lost with the land The future, if there is one, "will take care of itself" Time orientation to the "present"	9. Saving for the future (Insurance, retirement, savings account)
10. "Work" done only as needed to feed the family Historically dangerous, risking injury or death	10. "Puritan Work Ethic" Work for work's sake Rigid schedule
11. Indian Time Non-linear, relative to the activity at hand, flexible	11. Eurocentric obsession with time, "time is money"

(Hendrix, 2008)

- Assess potential sources of funding for the program, including the Indian Health Service, Medicare/Medicaid, community-based health insurance, and NGOs working in the area. Costs associated with transportation of patients and coverage of other incidental patient expenses may also have to be factored in to reduce barriers for patients seeking continuous follow-up care for the disease. Cost of medications and therapies has been known to be a major factor in underutilization of medications in Native American communities, especially in areas where Indian Health Service benefits are not available.

4. Identify members of the local Native American community to serve as community representatives of sickle cell management programs in the area. Also identify health professionals, traditional healers, and facilities working in the area and the nearest hospital providing acute management services for the disease to serve as a nodal center. A combination of local health facilities and larger hospitals will allow for propagation of a sickle cell disease program using an institutionalized approach. Plan for the presence of a basic laboratory area in the community to conduct neonatal and adult screening, and regular blood tests associated with sickle cell care.
5. Devise education materials including presentations, posters, banners, and flyers, in collaboration with the Native American community health workers, to propagate awareness of the disease and the availability of local services to manage it. Organize meetings and local fairs, led by the Native American community workers, to promote knowledge and identification of the disease, while continuously gaining feedback from community members on their perception of the disease in lieu of Native American beliefs. Word-of-mouth has shown to be an effective pathway of message propagation in the Native American community.
6. Stock the local health center/sickle cell program with essential medications and laboratory equipment for sickle cell management, as found in the “A Primer for Rural Management of Sickle Cell Disease” publication. Ensure easy accessibility to community members in terms of distance. Explore the possibility of holding screening camps in the community to identify patients and carriers, while also having a neonatal screening program for the disease at a local health center/hospital which provides maternity services.
7. Establish clear linkages with the local facilities and nearby hospitals for transfer of patients in crises or in need of transfusion. Develop a separate records system to keep track of sickle cell patients and carriers in the community. This also includes specialized medical records for sickle cell patients who are on therapy and need frequent follow-up visits.
8. Train local health providers in management of sickle cell disease, and assure that Native American community health workers have proper resources to continue spreading awareness of the disease in their area. Special emphasis will have to be placed on explaining long-term care pathways for sickle cell disease, the importance of complying with any medication regimens prescribed, and the importance of quickly identifying and conveying any disease-related complications. A culture of self-medication, dually taking traditional medicines with allopathic therapies, and at-will consumption of medications will have to be addressed. The following list prepared by Levanne Hendrix provides an

overview of cultural issues that need to be addressed in clinical encounters with Native American patients.

AMERICAN INDIAN AMERICAN	EURO-
Avoidance of direct eye contact as a sign of respect	Direct eye contact considered sign of honesty and sincerity
Handshake lightly, some women touch only the finger tips	Firm handshake denotes power
Information passed by "word of mouth" rather than media, some Internet used	Lectures, Newspapers, TV, Radio, and Internet utilized
Personal information not forthcoming	Self-disclosure valued, "open and honest" communication style
Ideas and feelings conveyed through behavior rather than speech	Verbal expression of ideas and feelings
Words are chosen carefully and deliberately, as the power of words is understood	Verbosity and small talk is appropriate social behavior
Listening is valued over talking	Schools teach speaking over listening, importance of expressing one's opinion
Use of observational skills and non-verbal communication	Verbal and written communication valued
Criticism communicated indirectly through another family member, direct criticism considered disrespectful and rude	Direct criticism used to alter behavior
Withdrawal used as a form of disapproval ("voting with your feet")	Direct expression of disapproval
Request given through indirect suggestion	Directiveness of requests

(Hendrix 2008)

9. Promote home-based identification of complications associated with sickle cell disease, on part of parents and spouses, stressing the importance of early identification and care-seeking. Overt expressions of pain have often been looked down upon the Native American community, and it is important that the necessity of expressing painful episodes and seeking immediate care is conveyed.
10. Explore the cultural appropriateness of genetic counseling in the Native American communities by training members of the Native American community to function as genetic counselors. Should genetic counseling be feasible and acceptable, begin exploring linkages between the local sickle cell programs and prenatal diagnostic facilities in the vicinity. Direct and incidental costs must be explored to ensure that the services are accessible to patients who wish to avail of them.

Conclusion:

The recommendations provided above as part of an action plan are broadly based on experiences taken from working with indigenous communities both in the United States and in India. Given the importance and severity of sickle cell disease and its prevalence among Native Americans, it is important that headway be made in providing essential and comprehensive care using a culturally-sensitive and community-based approach. Engaging the Native American community about the disease and using existing healthcare resources is an important step in moving forward. However, many cultural, political, and geographic barriers will have to be surmounted to ensure that sickle cell management programs are easily accessible to the Native American population.



References

Textual References:

- 1) Hendrix L. Health and Health Care for American Indian and Alaska Native Elders. 2008; Available at: <http://www.stanford.edu/group/ethnoger/americanindian.html>. Accessed 3/25, 2009.
- (2) Olney R. Newborn Screening for Sickle Cell Disease: Public Health Impact and Evaluation. In: Khoury M, Burke W, Thomson E, editors. Genetics and Public Health in the 21st Century. 1st ed. Atlanta, GA: CDC Office of Public Health Genomics; 2000. p. 4.22.
- (3) US Census. Annual Estimates of the Population by Race Alone and Hispanic or Latino Origin for the United States and States: July 1, 2003. 2003; Available at: <http://www.census.gov/popest/states/asrh/tables/SC-EST2003-04.pdf>. Accessed 3/25, 2009.
- (4) US Census Bureau. Census 2000 Summary File 1. 2000; Available at: <http://www.infoplease.com/ipa/A0767349.html>. Accessed 3/25, 2009.
- (5) Walton-Raji A. Researching Black Indian Genealogy of the Five Civilized Tribes. 2008; Available at: <http://www.african-nativeamerican.com/1IntroPage.htm>. Accessed 4/25, 2009.
- (6) HCUP Nationwide Inpatient Sample (NIS). Healthcare Cost and Utilization Project (HCUP). 1993-2004. Agency for Healthcare Research and Quality, Rockville, MD. www.hcup-us.ahrq.gov/nisoverview.jsp

Graphical References:

<http://www.holliston.k12.ma.us/placentino/nativeamericans.jpg>

<http://www.gatewaytosedona.com/image/articles/1463/NativeAmericanDancers.jpg>

<http://library.thinkquest.org/06aug/00440/images/sicklecellincapillary.jpg>

http://images-cdn01.associatedcontent.com/image/A1704/170438/300_170438.jpg

http://sanpedrosula.usvpp.gov/uploads/images/V4milgdgsQwXFc49FeMRPQ/American_Indian_Month.jpg