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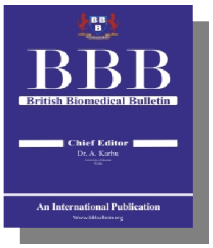

Original

Public Health Challenges of Hemoglobinopathies in Tribal Land in India: A Necessity of Introducing Genetic Services in the Health Care Systems Approach

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A B S T R A C T

There is no doubt that existing health care delivery system is very poor in rural India especially in the localities of backward, marginalized tribal, and under-privileged communities due to several apparent infrastructural lacune, constraints, limitations, and excuses. Added to these adversaries, are the ample prevalence of birth defects/genetic disorders among these poverty stricken vulnerable communities. There were 705 scheduled tribes and subtribes, and 75 Primitive Tribal Groups in India as per 2011 Census, which constituted about half of the indigenous people of the world! A huge mass of dwindling people has virtually no access to genetic services, available only to a few in big cities at exorbitant cost, and utterly lacking in rural India. This epidemiological review related to public health genetics, highlights the genetic burden/magnitude of hemoglobinopathies having glimpses of reproductive and child health, and neonatal/infant mortality in afflicted couples in the three states of India, namely, Chhattisgarh, Madhya Pradesh, and Odisha. With over emphasis laid on bringing awareness and imparting genetic/marriage counseling, carrier detection and establishment of prenatal diagnostic facilities at least in the vicinity of at risk communities. Cost of establishment of such infrastructural facilities is still lower than the potential benefits to the people. Therefore, it is high time to introduce the genetic services in the existing health systems approach.

Introduction

In this ever-changing world, with unique challenges that threaten the health and well-being of the population, it is imperative that the government and the community collectively rise to the occasion and face these challenges inclusively, simultaneously, and sustainable. Social determinants of health and economic issues must be dealt with a consensus on ethical principles – Universalism, justice, dignity, security and human rights. This approach would be of valuable services to humanity in realizing the dream of Right to Health. The ultimate yardstick for success would be, if every person from a remote hamlet to the capital city, experiences the change.

Public health is concerned with disease control and prevention at the population level, through the organized efforts and informed choices of public, private organizations, society, communities and individuals. However, the role of government is crucial for addressing these challenges and achieving health equity. Health challenges are required to be understood and acted upon. Concerted health research, policy development and analysis, program development and evaluation, health systems organization, models of health care financing and operational scientific research are aids to this process. Assessment highlights the need for action, while insights provide the possible solutions.

The focus of this article is to present an overview of the above public health conceptual background and status in relation to hemoglobin disorders in tribal land of India.

Dimensions of Public Health

The World Health Organization[1] definition of health enshrines "Health is a state of complete physical, mental and social well being and not merely the absence of disease or infirmity. A healthy nation is one of the best places to live in. The health of people is the foundation upon which all their happiness and all their powers as a state depend. Original accomplishment of Public Health was "The science and art of preventing disease, prolonging life and promoting physical health and efficiency through organized community efforts for the sanitation of the environment, the control of community infections, the education of the individual in principles of personal hygiene, the organization of medical and nursing services for the early diagnosis and preventive treatment of diseases and the development of social machinery which will ensure to every individual, in the community, a standard of living adequate for maintenance of health" of the population.[2] Thus public health is best identified as a social movement concerned with protecting and promoting the collective health of the community. The province of public health is by no means limited to prevention. In fact, even a small movement by a small village to purify their drinking water source or to stop alcohol drinking is also very much a public health activity.[3]

Epidemiology is the science concerned with the study of determining factors that influence the frequency and distribution of disease, injury, and other health-related events and their causes in a defined human population for the purpose of establishing programs to prevent the spread, control their development including the sum of knowledge gained in such a study.

The importance of public health in the development of India cannot be overemphasized. India is experiencing a rapid health transition. It is confronted both by an unfinished agenda of infectious diseases, genetic disorders, nutritional deficiencies and unsafe pregnancies as well as the challenge of escalating epidemics of non-communicable diseases. This composite threat to the nation's health and

development needs a concerted public health response that can ensure efficient delivery of cost-effective interventions for health promotion, disease prevention and affordable diagnostic and therapeutic health care. The idea is to protect and promote the health of the people of India by facilitating the exchange of information, experience, and research, and advocating policies, programs and practices that improve human public health. Public Health aims to understand and influence the cultural, economic and social determinants of health as well as to study the structure of health systems as efficient channels for health delivery services. Public health is, thus, a discipline built on the academic tradition of inquiry involving research, teaching and professional practice to prevent disease and promote health in populations.

Health system strengthening, human resource development and capacity building and regulation in public health are important areas within the health sector. Contribution to health of a population also derives from social determinants of health like living conditions, nutrition, safe drinking water, sanitation, education, early child development and social security measures. The new agenda for Public Health in India includes the epidemiological transition (rising burden of chronic non-communicable diseases), demographic transition (increasing elderly population) and environmental changes. The unfinished agenda of maternal and child mortality, HIV/AIDS pandemic and other communicable diseases still exerts immense strain on the overstretched health systems. The basic contention is to build human resource capacity for strengthening decentralized health planning, especially at the district level, to improve accountability of health systems, elicit community participation in health, to ensure equitable and accessible health facilities and to bring about convergence in programs and services.[4]

Education and training in public health needs to be interdisciplinary in content so that the pathways of public health action are multi-sectoral in applications. Public health education must include subject areas like epidemiology, population health, genetics, biostatistics, behavioral sciences, health economics, health services management, environmental health, health inequities and human rights, gender and health, health communication, ethics of health care and research. The proposal of interventions needs to be evidence-based, context-specific and resource-sensitive. Potential areas of community participation could be in lifestyle modification in chronic diseases through physical activity and diet modification, and primary prevention of alcohol dependence through active community-based methods like awareness creation and behavioral interventions.[5] Public health has often been defined as a science dealing with the determinants of health defence and prevention of health at the population level, while clinical medicine deals with multiple maladies and their remedies at an individual patient or family level.

Population/Public Health Genetics

Public Health Genetics is rapidly developing medical discipline that provides the knowledge to:

1. Prevent the occurrence of many birth defects;
2. Treat the sequelae of genetic disorders; and;
3. Decrease the burden of chronic disabling diseases.

Genetic services exist to help patients and families understand their specific genetic disease or risk thereof, for improving quality of life within the context of our own psychosocial, ethno-cultural

background. The ultimate goal of genetic services is to reduce mortality and morbidity, and to alleviate suffering associated with genetic/congenital disorders, coming into the domain of public health for individual treatment and management of patients, families, and communities.

Hemoglobinopathies

Hemoglobinopathies are a group of single gene disorders characterized by the production of structurally defective hemoglobin (heme=iron; globin=protein) due to abnormalities in the formation of the globin moiety of the molecule. The most commonly encountered hemoglobins were following alterations:

Hb D^{Punjab} = Glutamic Acid > Glutamine at 121st position

Hb E = Glutamic Acid > Lysine at 26th position

Hb S = Glutamic Acid > Valine at 6th position.

The thalassemias are characterized by an increased/reduced rate of production of normal/abnormal hemoglobin (Hb) due to absence or decreased synthesis of one or more polypeptide globin chains.

Hemoglobin disorders are generally confined to the tropical or subtropical countries of the world because they have probably evolved in the past to subsidize or counter the lethal effects of malaria in the population.[6,7] It has now been convincingly recognized that the heterozygotes of hemoglobin disorders get some immuno-logical and physiological advantage of protection for the non-severity of malaria over the normal individuals against the endemic malaria in tropical and subtropical regions of the world.

World and Indian Scenario

It has been estimated that about 7% of the world population is a carrier of hemoglobinopathies and 3 to 4 lacs babies are born with severe disease worldwide every year. [8] About 60-70 million are estimated to be carriers of sickle cell disease and roughly 1,20,000 homozygotes are added every year in the world. [9] There are 15 million people worldwide who have clinically apparent thalassemia disorders with about 240 million carriers of β-thalassemia alone. [8]

The number of β-thalassemia is 30 million in India with a range of 3-17% prevalence, and the average being 4.2%.[10] Every year 1, 00, 000 babies are born world over with thalassemia major and 15,000 babies are born in India.[11,12] The carrier rate of β-thalassemia gene varies from 1% to 3% in Southern India and from 3% to 17% in North India and 1-3% in rest of the country.[13]

Prevalence/Incidence of Hemoglobinopathies in India

There are regional variations for the three most common structural variants of hemoglobin, i.e. Hb D, Hb E and Hb S; the estimated cumulative allele frequency of these variants has been found to be 5.35% in India.[13, 14] The average allele frequency of the sickle cell gene and hemoglobin D has been observed to be 4.3% and 0.86%, respectively. The hemoglobin E gene constitutes 10.9% in North Eastern region of India.[14,15]

The incidence of births with different hemoglobin disorders per 1000 live-births in a tertiary hospital in Central India (at Jabalpur in Madhya Pradesh) was estimated to know the genetic burden of hemoglobinopathies on the health.[16] It was revealed that only 586 normal children are born against 414 per thousand live-births offspring with hemoglobin disorders. Of the 414 children born with hemoglobin disorders, sickle cell traits constituted 132, followed by homozygous sickle cell disease 101, sickle cell-β-thalassemia 24, and sickle

cell-E disease 1.3 per thousand live-births. On the other hand, β-thalassemia trait contributed 40, followed by thalassemia major 42, hemoglobin E-β-thalassemia 2.7, and hemoglobin E trait 1.3 per thousand live-births. This shows a very heavy burden of birth defects of hemoglobinopathies in Central India.[16]

The contribution of couples with hemoglobinopathies toward neonatal and infant mortality, respectively per thousand live-births was 12.9 and 12.9 (AA/AA), 39.6 and 39.6 (AA/AS), 111.1 and 111.1 (AA/SS), 25.8 and 36.1 (AS/AS), 153.8 and 153.8 (AS/SS); and 160.0 and 160.0 (AA/β-thalassemia trait), 25.6 and 38.5 (β-thalassemia trait/β-thalassemia trait), 20.4 and 40.8 (AS/β-thalassemia trait), 125.0 and 125.0 (AS/Sickle cell-β-thalassemia), and 39.3 and 47.2 [(combined above all categories) except the normal (AA/AA)] for the respective categories of couples in Central India. This revealed a very high mortality in the offspring of carrier couples of hemoglobin disorders in Central India.[17,18] The neonatal mortality rates were 44 and 33 for the state of Madhya Pradesh and for India, respectively for the year 2010 and 2011; and the infant mortality rates were 62 and 47 for Madhya Pradesh and India, respectively for the year 2010 and 2011 as per Registrar General of India (Sample Registration System Bulletin, December 2011).

Chhattisgarh and Madhya Pradesh: Hemoglobinopathies and Allied Disorders

It is fascinating that about half of the world's indigenous communities live in India. They constituted 8.6% of the total population of India as per 2011 census. Tribal communities are highly vulnerable to many hereditary hemolytic disorders that cause a high degree of morbidity and mortality. [19] Pattern of hemoglobin disorders in Central India is still unknown.

The undivided state of Madhya Pradesh was inhabited by 46 tribal communities in Central India that constituted about 23% tribal population of India in 2001. As per 2011 census, the 46 tribal communities in Madhya Pradesh constituted 14.7% of the tribal population (104281034 with 52409823 males and 51871211 females) of India. A randomly conducted study presented the public health challenges of sickle cell disorders, β-thalassemia syndrome and G6PD deficiency in relation to malaria endemicity in scheduled caste and tribal communities of Chhattisgarh and Madhya Pradesh in Central India.[20] High prevalence of the sickle cell disorders was recorded in the tribes of Baiga (22.3%) and Bharia (13.2%) with a range of β-thalassemia trait being 0-3.6% in Madhya Pradesh (Table 1 and 2), followed by Hill Maria (22.5%), Maria (20.2%) and Muria (14.9%) tribes with β-thalassemia trait range of 0-10.4% in Chhattisgarh (Table 3). The G6PD deficiency was varying from 0% to 21.5% in Chhattisgarh and from 1.8% to 12.1% in Madhya Pradesh. The frequency of sickle cell disorders fluctuated from 4.1% to 34.0% among the scheduled tribes of Madhya Pradesh and from 0.9% to 22.5% in scheduled tribes of Chhattisgarh. The range of β-thalassemia trait was variable from 0% to 10.4% in Chhattisgarh and from 0% to 10.0 percent among the scheduled tribes of Madhya Pradesh. The G6PD deficiency range was 1.3% to 9.3% among the scheduled tribes and 0% to 6.9% in scheduled castes of Madhya Pradesh. Among the scheduled castes, the frequency of sickle cell disorders varied from 4.4% to 37.9%, the sickle cell-β-thalassemia being 3.9%. The frequency of β-thalassemia trait was variable between 0 to 10.0 percent among the scheduled castes of Madhya Pradesh.[19,20]

Based on 1251 referral cases studied during March 2010 to March 2012 from a

tertiary hospital, has shown the highest prevalence of sickle cell trait (24.6%) and β-thalassemia trait (12.1%) in the general population of Central India.[10] From the tribal land of Odisha, the highest frequency of β-thalassemia trait (12.7%) was noticed among Paraja Bhuyan tribe of Sundargarh district, followed by Paraja tribe (8.5%) of Koraput district, Dudh Kharia (8.1%) of Sundargarh district, Santhal (8.0%) of Mayurbhanj district, Paik Bhuyan (7.8%) of Sundargarh district.[21,22] High frequency of β-thalassemia trait (10.4%) was recorded among Hill Korwa of Chhattisgarh (Table 2). A very low frequency of β-thalassemia trait (range: 0.4-3.8%) was observed among other major scheduled tribes of Chhattisgarh. The prevalence of β-thalassemia trait (range being 0.2-3.6%) was found very low in Madhya Pradesh. However, the highest frequency of β-thalassemia trait (10.0%) was recorded among Gond tribe of Damoh district in Madhya Pradesh. The frequency of β-thalassemia trait was equally high among scheduled castes such as Jharia caste (10.0%) of Jabalpur district, and followed by Chaudhury (9.0%) of Damoh district, and other backward castes (8.6%) of the state. The frequency of β-thalassemia trait was recorded to be around 3% among Bagata tribe of Andhra Pradesh. The frequency of β-thalassemia trait in primitive tribes of Gujarat (3.1-4.6%), Maharashtra (1.6-3.2%) and Tamil Nadu (0.9-2.3%) was also found very low. In view of the high prevalence of β-thalassemia trait among some tribal communities, emphasis was laid on intervention and prevention through antenatal carrier screening, genetic/ marriage counseling and establishment of prenatal diagnostic facilities in localities of at risk tribes in India.[10]

Odisha State Scenario

It has been estimated that 3-4 million people are suffering from sickle cell disorders in Odisha state (Table 4 and 5). Out of them, two-third million people belong to general and scheduled castes; and about one-fourth of a million constitute the scheduled tribes in Odisha.[23]

The estimate shows that 19.32% of people of Odisha suffer from hemoglobinopathies or every fifth person in Odisha is afflicted with hemoglobinopathies.[23] Out of them, 13.2% have sickle cell disorders (AS= 8%; SS=4%; S-β-Thalassemia=1.2%) or every eighth person in Odisha is afflicted with sickle cell disorder; and 3.8% suffer from β-Thalassemia syndrome (Trait=2.3%; Major=1%; δβ-Thalassemia= 0.5%) or every 25th person in Odisha suffers from β-thalassemia syndrome. It has been estimated that 2% of the population suffers from Hb E disorders (Trait=1.3%; EE=0.4%; E-β-Thalassemia=0.3%) or every 50th person has Hb E disorders. About 0.32% of persons have Hb D disorders (AD=0.2%; DD=0.1%; D-β-Thalassemia=0.02%) or every 313th person or 32 out of 10,000 people have this condition. There is a rare occurrence of Hereditary Persistence of Fetal Hemoglobin (HPFH) in Odisha.[24]

Age and Sex distribution

Since the hemoglobinopathies have in the general autosomal recessive mode of inheritance except the HPFH in which case it has autosomal dominant inheritance, therefore, there should not be any disparity with respective to sex distribution. However, practical observations may show some disparity due to nonrandom mating, ethnicity, caste, eco-geographical location, socio-cultural preferences, biased attitude towards females for treatment, etc. Further, as the hemoglobin disorders are genetically transmitted to the offspring having defective

gene located on autosomal chromosome, therefore, age does not matter for clinical manifestations. Every child born with the defect would either be a male or a female. However, there is also an equally possibility that a child born may be a eunuch or hermaphrodite.

Natural Selection and Prevalence of hemoglobinopathies in India

There is no state in India, which has not been affected either by malaria or by hemoglobinopathies, however, the prevalence does vary from state to state depending upon the breeding structure of the population or prevailing climatic conditions. It has been noted that hemoglobin disorders co-exist either with β-thalassemia or glucose-6-phosphate dehydrogenase (G6PD) enzyme deficiency in malaria endemic areas having an inverse relationship with each other. If the frequency of hemoglobin disorders with most common variants such as Hb S, Hb E or Hb D, increases, then the frequency of either β-thalassemia or G6PD enzyme deficiency decreases in the population and vice versa in malaria endemic areas. [6,7] This is a universal truth in India. For example, sickle cell variant is very common in the populations of central India belt in one hand and the frequency of β-thalassemia or G6PD enzyme deficiency is very low on the other hand in malaria endemic regions. This phenomenon is not only confined to tribal people and tribal areas, but is equally applicable to nontribal people and nontribal regions in India.

The disequilibrium of genetic markers such as various variants of hemoglobin and high occurrence of G6PD deficiency is reflected as the Natural selection mechanism for protection against malaria.[25] Although, no quantitative information (e.g. number of cases, direct evidence of toxicity) available, but the indirect evidence has supported the impact of oxidative stress on G6PD deficient individuals with relatively lower susceptibility to malaria.[26] Roth *et al*.[27] in vitro studies demonstrated that G6PD deficiency in hemizygous males and heterozygous females is equally protected against malaria. Selection can maintain deleterious alleles in the population if there is a heterozygote advantage as in the case of sickle cell trait. The G6PD gene also provides an opportunity to study how selection has affected the genetic variability in the Indian populations.

Malarial parasites break down hemoglobin after invasion.[7] They do so to make room to grow and may also derive nutrition from it. The byproduct of this process, particularly the oxidized iron is potentially toxic to the parasite. Reduced glutathione (GSH) supplies reduce energy to cells under natural mechanisms to overcome the oxidative stress. Any deficiency in the production of GSH in the cell can provide resistance against the malaria parasite.[7]

Mission of Clinical/Population Health Genetics in Public Health

It has two missions:

1. To reduce death, illness and disability from genetic disorders, birth defects and chronic diseases and injuries and to improve the quality of life of individuals, and
2. To protect and promote the health of children with special health care needs by assuring a family-centered, community-based, comprehensive, coordinated and culturally appropriate system of special health care.[28]

Education of patients, parents, professionals, voluntary organizations and the general public about genetics, genetic disorders, birth defects and genetic services is an important activity. Educational services and materials should be available to all.

Hemoglobinopathies Screening Program Requirements[16]

1. Manpower Development & Training.
   - Medical Doctor (Geneticist)
   - Hematologist to handle Laboratory & Blood Transfusion facilities
   - Nurses
   - Laboratory Technicians
2. Genetic Counselor
3. Development of Updated Laboratory Facilities
4. Development of Blood Bank Facilities
5. Multi-specialty consultancy, treatment, and periodic follow up (General Practitioner, Pediatrician, Gynecologist, any other as per requirement)
6. Indoor admission Facilities
7. Funding and Financial Security

Genetic Screening of Burdensome Disorders[28]

Molecular diagnosis is now possible for a large number of genetic disorders. For the strategy of management of genetic diseases, the intake of patients into the program will occur by two routes:

1. Through planned genetic screening (a) Neonatal screening, (b) Pre-pregnancy screening after Marriage;
2. And through individual referrals for medical diagnosis and genetic counseling. Genetic screening has three major objectives: (a) To provide opportunity for medical intervention (treatment), (b) To provide opportunity for counseling about reproductive options; (c) To collect research data pertinent to public health policy and basic knowledge.

At present, there are several institutes/centers throughout India especially in Metro-cities where sufficient facilities are available for detailed investigations and genetic/marriage counseling (Fig. 1).

Pre-requisites for Genetic Counseling[28]

1. Detailed family history
2. Accurate diagnosis
3. Understanding of the medical aspects of the disorder (etiology, natural history, treatment, prognosis, burden)
4. Understanding the inheritance pattern (recurrence risk)
5. Understanding the psycho-social impact of the information
6. Training/experience in counseling techniques
7. Understanding the concepts of health/disease/health care in the appropriate culture.

Genetic/Marriage Counseling

Genetic counseling is meant for

1. Determining the facts: diagnosis, etiology, and inheritance pattern, prognosis, natural history, treatment, and recurrence risks.
2. Transmitting the information to those requesting it in a sensitive, culturally appropriate, understandable way.
3. Supporting the decision making process of the couple.
4. Genetic counseling is non-directive.

Integration of Community Genetic Services into Primary Health Care Systems Approach

Integration of basic public health approaches into the existing primary care and reproductive health clinics is the most appropriate, sustainable and cost-effective approach.[16]

Although some additional training and resources will be required, the potential benefits are considerable.

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Table 1. Spectrum of hemoglobinopathies in the state of Madhya Pradesh in Central India*

Types of hemoglobinopathies	Males	Females	Total	Percentage
Normal	345	318	663	51.8
Sickle cell trait	148	169	317	24.8
Sickle cell disease	49	52	101	7.9
Sickle cell-β-thalassemia	18	9	27	2.1
β-thalassemia trait	55	77	132	10.3
Thalassemia major	10	21	31	2.4
δβ-thalassemia	0	0	0	0.0
Hemoglobin E trait	1	3	4	0.3
Hemoglobin E disease	0	0	0	0.0
Hemoglobin E-β-thalassemia	0	2	2	0.2
Hemoglobin D trait	0	0	0	0.0
Sickle cell-D- disease	0	0	0	0.0
Total	628	651	1279	100.0

*Data from reference No. 10.

Table 2. Genetic burden of community hemoglobinopathies in scheduled tribes and castes of Madhya Pradesh*

Scheduled Tribe	District	N	Sickle Cell Trait %	Sickle Cell Disease %	Sickle Cell Disorders %	β-thalassemia Trait %
Primitive Tribes:						
Baiga	Mandla	1566	15.6	0.5	16.1	0.2
Baiga	Dindori	990	18.4	1.1	19.5	3.6
Bharia	Chhindwara (Inside valley)	183	13.2	0.0	13.2	0.0
Bharia	Chhindwara (Outside)	102	2.9	0.0	2.9	0.0
Scheduled Tribes:						
Barela	Khargone	345	25.5	0.9	26.4	0.0
Barela	Nimar	316	27.2	0.3	27.5	1.3
Bhil	Jhabua	904	20.0	0.9	20.9	0.0
Bhil	Ratlam	433	11.8	0.2	12.0	0.0
Bhil	Nimar	316	27.2	0.3	27.5	1.3
Bhilala	Jhabua	403	30.5	1.7	32.2	0.0
Bhilala	Nimar	370	18.4	0.3	18.7	1.3
Gond	Betul	299	11.4	0.7	12.1	1.0
Gond	Damoh	321	33.0	1.0	34.0	10.0
Gond	Jabalpur	3224	19.0	0.7	19.7	8.7
Gond	Mandla	280	18.6	0.0	18.6	0.0
Gond	Chhindwara	75	4.3	0.0	4.3	0.0
Gond	Chhindwara	158	15.8	2.5	18.3	0.0
Gond	Chhindwara	83	12.0	4.8	16.8	0.0
Gond	Seoni	286	18.9	1.5	20.4	0.0
Gond	Shahdol	252	13.1	1.2	14.3	4.6
Gond	Balaghat	311	15.4	0.6	16.0	2.2
Raj Gond	Damoh	321	10.3	0.3	10.6	3.1
Kol	Satna	290	4.1	0.0	4.1	5.9
Korku	Chhindwara	250	17.2	1.2	18.4	4.8
Korku	Khandwa	301	16.9	0.7	17.6	2.3
Korku	Betul	296	13.8	0.7	14.5	3.9
Patelia	Jhabua	166	20.5	1.8	22.3	0.0
Panika	Shahdol	210	28.6	3.3	31.9	1.4
Pradhan	Dindori	226	28.3	1.8	30.1	0.0
Pradhan	Dindori	990	18.4	1.1	19.5	3.6
Scheduled Castes:						
Balai	Khandwa	276	14.1	0.4	14.5	2.5
Basod	Betul	123	19.5	0.8	20.3	0.0
Basod	Chhindwara	150	22.0	0.0	22.0	4.0
Chaudhry	Damoh	339	18.0	1.0	19.0	9.0
Chaudhry	Shahdol	195	5.1	0.0	5.1	3.6

Jharia	Jabalpur	637	4.4	0.0	4.4	2.3
Jharia	Jabalpur	409	37.9	1.2	10.0	3.9
Katiya	Chhindwara	181	24.9	2.2	27.1	1.1
Mehra	Betul	352	32.4	2.0	34.4	0.3
Mehra	Chhindwara	114	19.8	3.4	23.2	5.2
Mehra	Seoni	216	21.3	0.5	21.8	1.4
Mehra	Balaghat	219	18.3	0.0	18.3	0.0

*Data from reference No. 10.

Table 3. Genetic burden of community hemoglobinopathies in scheduled tribes of Chhattisgarh*

Scheduled Tribe	District	N	Sickle Cell Trait %	Sickle Cell Disease %	Sickle Cell Disorders %	β-thalassemia Trait %
Birhor	Raigarh	270	0.0	0.0	0.0	2.2
Hill Korba	Jashpur	744	1.7	0.0	1.7	8.4
Kamar	Raipur	320	0.9	0.0	0.9	6.6
Hill Maria	Bastar	93	22.5	0.0	22.5	0.0
Maria	Bastar	101	13.9	0.0	13.9	0.0
Bhatra	Bastar	99	13.1	0.0	13.1	0.0
Dhurwa	Bastar	81	6.2	2.5	8.7	0.0
Gond	Ambikapur	127	20.5	0.8	21.3	0.0
Gond	Raipur	157	15.9	0.6	16.5	0.0
Halba	Raipur	122	13.9	0.0	13.9	0.0
Halba	Bastar	95	19.0	1.0	20.0	0.0
Halba	Durg	365	15.6	0.3	15.9	2.4
Kodaku	Sarguja	400	3.0	0.3	3.3	3.8
Oraon	Ambikapur	422	2.1	0.5	2.6	0.0
Oraon	Raigarh & Sarguja	215	0.0	0.0	0.0	1.9

*Data from reference No. 20.

Table 4. Spectrum of hemoglobinopathies in the state of Odisha*

Types of hemoglobinopathies	Males	Females	Total	Percentage
Normal	184	164	348	34.3
Sickle cell trait	171	131	302	29.8
Sickle cell disease	41	36	77	7.6
Sickle cell-β-thalassemia	4	13	17	1.7
β-thalassemia trait	91	94	185	18.2
Thalassemia Major	29	25	54	5.3
δβ-thalassemia	4	5	9	0.9
Hemoglobin E trait	2	7	9	0.9
Hemoglobin E disease	2	1	3	0.3
Hemoglobin E-β-thalassemia	5	2	7	0.7
Hemoglobin D trait	1	1	2	0.2
Sickle cell-D-disease	1	1	2	0.2
Total	533	482	1015	100.0

*Data from reference No. 29.

Table 5. Genetic burden of community hemoglobinopathies in 18 scheduled tribes of Odisha*

Scheduled Tribe	District	N	Sickle Cell Trait %	Sickle Cell Disease %	Sickle Cell Disorders %	β-thalassemia Trait %
Bondo	Malkangiri	165	0.7	0.0	0.7	0.5
Paik Bhuyan	Sundargarh	244	7.4	0.0	7.4	7.8
Paraja Bhuyan	Sundargarh	213	0.9	0.0	0.9	12.7
Paudi Bhuyan	Sundargarh	379	0.0	0.0	0.0	2.1
Didayi	Malkangiri	1014	3.6	0.0	3.6	3.0
Juang	Keonjhar	879	1.8	0.0	1.8	2.3
Dudh Kharia	Sundargarh	422	0.0	0.0	0.0	8.1
Dhelki Kharia	Sundargarh	345	11.9	0.6	12.5	4.1
Kutia Kondh	Kandhamal	65	3.1	0.0	3.1	10.8
Kondh	Kandhamal	254	3.1	0.0	3.1	6.3
Lodha	Mayurbhanj	78	0.0	0.0	0.0	6.7
Saora	Ganjam	177	7.3	0.0	7.3	6.2
Lanjia Saora	Gajapati	74	0.0	0.0	0.0	2.7
Sabar	Ganjam	102	0.0	0.0	0.0	5.9
Bathudi	Mayurbhanj	95	1.0	0.0	1.0	0.0
Bhatra	Nabarangpur	166	15.1	3.0	18.1	6.6
Bhumiz	Sundargarh	116	0.9	0.0	0.9	1.7
Gond	Kalahandi	219	21.0	1.4	22.4	0.5
Kissan	Sundargarh	130	0.0	0.0	0.0	1.5
Kolha	Mayurbhanj	102	0.0	0.0	0.0	2.0
Munda	Sundargarh	96	3.1	0.0	3.1	5.2
Oraon	Sundargarh	104	0.0	0.0	0.0	1.9
Paraja	Koraput	176	13.1	1.7	14.8	8.5
Santhal	Mayurbhanj	100	1.0	0.0	1.0	8.0

*Data from reference No.30.

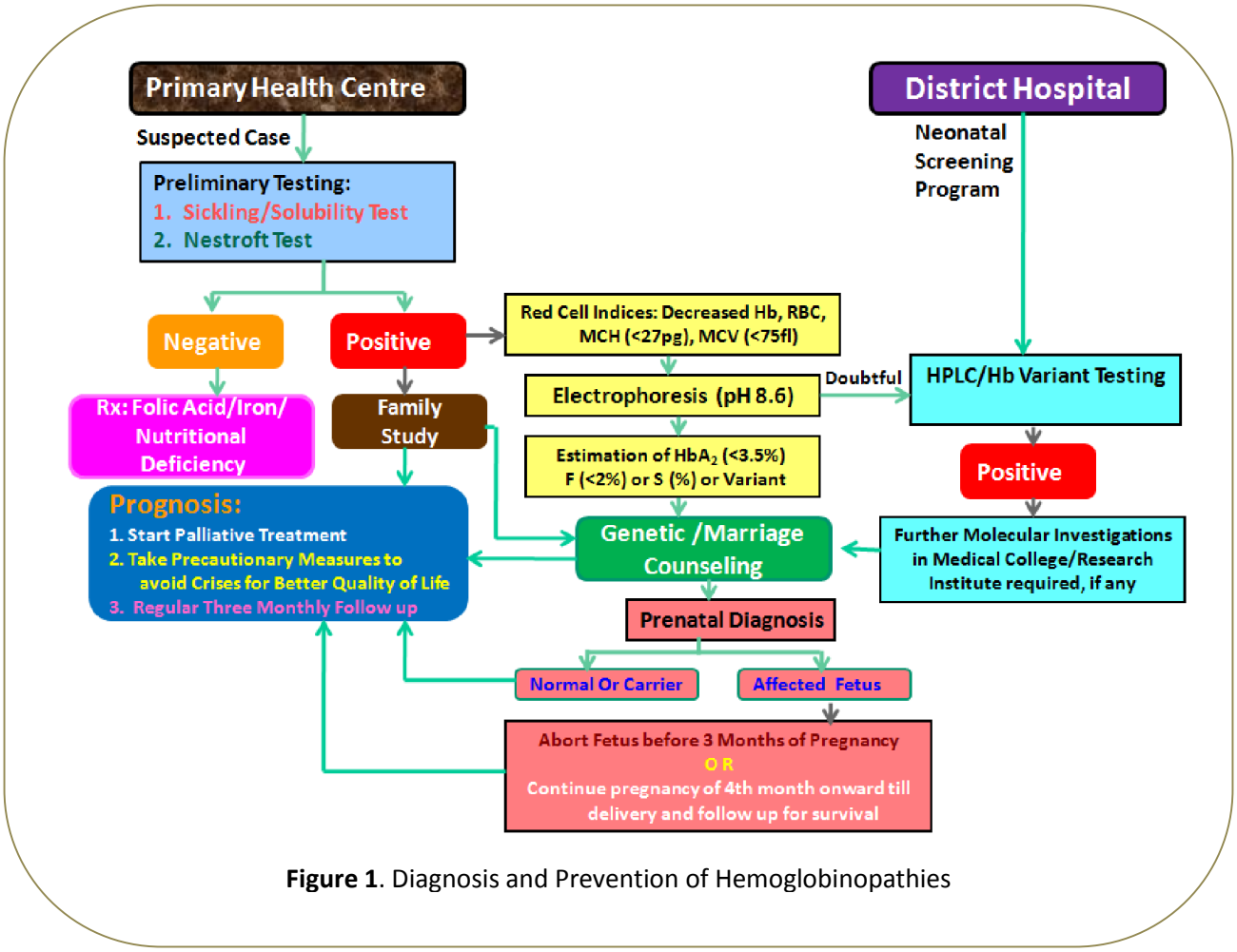

Figure 1. Diagnosis and Prevention of Hemoglobinopathies