"As more and more tests for heterozygosity are developed, predictions can be made with greater and greater reliability about the probability of birth of defective children, and advice can be given to prospective spouses or parents about the desirability of their contributing to the welfare of the human race as a whole by preventing the transmissions of seriously defective genes to the next generation."

Linus Pauling, 1960
1. General Introduction

1.1 Introduction

The tribal population is identified as the aboriginal inhabitants of our country. They are the most vulnerable section of our society living in natural and unpolluted surrounding far away from civilization holding on to their traditional values, customs and beliefs. According to the Indian Constitution 1950 “Any tribe or tribal community or part of or group within any tribe or tribal community as deemed under article 342 are ‘Scheduled Tribes’ for the purpose of the Constitution”.

In general, tribe is a traditional social division consisting of families and communities inter-related by social, economic, cultural, religious, dialect or blood relationships. They are characterized by their primitive traits, geographical isolation, distinct culture and shyness with the community at large and economic backwardness (Article 342, Constitution of India). The Imperial Gazetteer of India (1911) defines a tribe as a “collection of families bearing a common name, speaking a common dialect, occupying or professing to occupy a common territory and is not usually endogamous though originally it might have been so”. Another definition of a tribe by Majumdar (1961) is that “a tribe is a collection of families or group of families bearing a common name, members of which occupy the same territory, speak the same language and observe certain taboos regarding marriage, profession or occupation and have developed a well-assessed system of reciprocity and mutuality of obligations”. Bardhan (1973) defines the tribe as “course of socio-cultural entity at a definite historical stage of development. It is a single, endogamous community with a cultural and psychological makeup”. Chattopadhyaya (1978) defines that “a tribe ordinarily has an ancestor or patron deity. The families or groups composing the larger units are linked through religions and socio-economic functions”.

According to Vidyarthi (1981), the tribe is a social group with definite territory, common name, common district, common culture, behavior of an endogamous group, common taboos and existence of distinctive social and political system, full faith in leaders and self-sufficiency in their distinct economy. Krishnan (1985) defines “tribe is a social group of simple and kind, the members of which speak a common dialect, and
have a common name, a contiguous territory, a relatively single government act together for common purposes, a relatively uniform culture or way of life and a tradition of common descent.” In the Indian context, they are commonly referred to as *Adivasi* (original settlers), *Girijan* (hill dwellers), *Vanya jati* (forest caste men), *Adimjati* (Primitive castes), and *Anusuchit Janjati* (Scheduled tribes). The tribes of India who are unable to defend themselves were gradually forced to recede before the invading hoards of such people, as the Dravidian, Indo-Aryans and Mongolians coming from the West, North West and North East. These people took shelter in the forest and mountain ranges. Those who were left behind in the plains generally disappeared either by absorption or by acculturalization.

1.2 Origin of tribes in India

India is a country with enormous social and cultural diversity since it is positioning on the crossroads of many historic and pre-historic human migrations. Contemporary ethnic populations of India are highly variable, both biologically and culturally (Karve, 1961; Beteille, 1998; Majumder, 1998). The origins and migrational histories of the tribal populations of the Indian subcontinent are not clearly understood. It has been argued that Africa may have made some direct genetic contribution to India, since some tribal populations in Southern India possess phenotypic similarities with Africans, the so-called “Negrito” physical characteristics (Maloney, 1974; Saha *et al.*, 1974; Roychoudhury, 1982; Chandler, 1988; Majumder, 1998). According to Majumder and Mukherjee (1993) a “Negrito element” was widespread throughout India and was eventually forced into a more restricted location in South India.

“Out-of-Africa” hypothesis suggests that the anatomically modern humans originated in Africa about 1,60,000 - 1,50,000 years ago, and then spread outward, completely replacing the local archaic hominin populations outside Africa. India has served as a major corridor for the dispersal of modern humans out of Africa, owing to the positioning of the Indian Peninsula at the crossroads of Africa, the Pacific and the West and East Eurasia. The enormous cultural, linguistic and genetic diversity of the more than one billion people living in the contemporary ethnic India can be attributed to this. The Indian society and culture has been affected by multiple waves of migration and gene flow that occurred in the historic and pre-historic times (Ratnagar, 1995).
According to Guha (1931), the population of India is derived from six main ethnic groups based on their different anthropological stocks. The details are as follows:

(i) Negritos

The Negritos or the Brachycephalic (broad headed) from Africa were the earliest people to inhabit India. They are survived in their original habitat in the Andaman and Nicobar Islands. The Jarewas, Onges, Sentelenese and Great Andamanis tribes are the few groups. Studies have indicated that the Onges tribes have been living in the Andamans for the last 60,000 years (Thangaraj, 2005). The traces of Negritos are seen in patches of some of the forest tribes of the higher hills of the extreme south of India like Irulas, Kotas, Paniyans and Kurumbas and similar traces appear in the inaccessible areas of Assam, Bengal and Burma.

(ii) Pro-Australoids or Austrics

This group was the next to enter India after the Negritos. They represent a race of people, with wavy hair plentifully distributed over their brown bodies, long heads with low foreheads and prominent eye ridges, noses with low and broad roots, thick jaws, large palates and teeth, and small chins. Austrics tribes, which are spread over the whole of India, Myanmar and the Islands of South East Asia, are said to “form the bedrock of the people”. The Austrics were the main builders of the Indus Valley Civilisation. Their language has survived in the Kol or Munda (Mundari) in Eastern and Central India.

(iii) Mongoloids

These people have features that are common to those of the people of Mongolia, China and Tibet. These tribal groups are located in the North-Eastern part of India in states like Assam, Nagaland and Meghalya and also in Ladakh and Sikkim. Generally, they are people of yellow complexion, oblique eyes, high cheekbones, sparse hair and medium height. The tribes like Mundari speaking (Munda Group of Austro-Asiatic Family) Munda, Santal, Ho, Juang, Saora, Gadaba and number of Central Indian Dravidian speaking tribes like the Maria, Muria, Kondh and Oraon come under Mangoloids.

(iv) Mediterranean or Dravidian

These groups came to India from the Southwest Asia and appear to be people of the same stock as the people of Asia Minor, Crete and the Pre-Hellenic Aegeans of Greece.
They are reputed to have built up the city civilization of the Indus Valley, whose remains have been found at Mohenjodaro, Harappa and other Indus cities. The Dravidians must have spread to the rest of India, supplanting Austrics and Negritos alike. Dravidians comprise all the three sub-types, Paleo-Mediterranean, the true Mediterranean and Oriental Mediterranean. This group constitutes the bulk of the scheduled castes in the North India and also has a sub-type called Oriental group.

(v) Western Brachycephals

These include the Alpinoids, Dinaries and Armenois. The Coorgis and Parsis fall into this category.

(vi) Nordics

Nordics or Indo-Aryans are the last immigrants into India. Nordic Aryans were a branch of Indo-Iranians, who had originally left their homes in Central Asia, some 5000 years ago and had settled in Mesopotamia for some centuries. It is believed that the Aryans must have come into India between 2000 and 1500 B.C. Their first home in India was Western and Northern Punjab, from where they spread to the Valley of the Ganga and beyond. These tribes are now mainly found in the Northwest and the Northwest Frontier Province (NWFP). Many of these tribes belong to the "upper castes".

Later, Malhotra (1978) classified the ethnic diversity of Indian subcontinent into four major ethnic groups by their physical features: Caucasoid (European), Proto-Australoid (Aboriginal Australian), Mongoloid (East Asian) and Negritos (African). India has almost all the primary ethnic strains, wherein Caucasoid groups are distributed in most of the regions; Proto-Australoids in West, Central and Southern regions; Mongoloids in the Sub-Himalayan and North-East regions and the Negritos in the Andaman Islands and South India. The dominant ethnic group among the Indian tribal group is Proto-Australoid although groups living in the Sub-Himalayan belt have more of Asian characteristics (Shankarkumar, 2003). It is believed that the ancestor of some of the existing tribes in Southern India entered the country from the Northwest, during pre-historic times.

Ethnic groups inhabiting the Indian sub-continent based on their linguistic affiliation is given in the Table I.
Table I. Different ethnic groups inhabiting the Indian subcontinent based on linguistic affiliation

<table>
<thead>
<tr>
<th>S. No</th>
<th>Ethnic groups</th>
<th>Linguistic affiliation</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Tharu</td>
<td>Indo-European</td>
<td>Kivisild et al., 1999</td>
</tr>
<tr>
<td></td>
<td>Buksa</td>
<td>Indo-European</td>
<td>Kivisild et al., 1999</td>
</tr>
<tr>
<td>2.</td>
<td>Adi</td>
<td>Tibeto-Burman</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Apatani</td>
<td>Tibeto-Burman</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Nishi</td>
<td>Tibeto-Burman</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Naga</td>
<td>Tibeto-Burman</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Tipperah</td>
<td>Tibeto-Burman</td>
<td>Roychoudhury et al., 2001</td>
</tr>
<tr>
<td>3.</td>
<td>Lodha</td>
<td>Austro-Asiatic</td>
<td>Roychoudhury et al., 2001</td>
</tr>
<tr>
<td></td>
<td>Munda</td>
<td>Austro-Asiatic</td>
<td>Roychoudhury et al., 2001</td>
</tr>
<tr>
<td></td>
<td>Santal</td>
<td>Austro-Asiatic</td>
<td>Roychoudhury et al., 2001</td>
</tr>
<tr>
<td>4.</td>
<td>Andh</td>
<td>Indo-European</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Pardhi</td>
<td>Indo-European</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Lambadi</td>
<td>Indo-European</td>
<td>Kivisild et al., 1999</td>
</tr>
<tr>
<td>5.</td>
<td>Jenukurumba</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Kattunaiken</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Soligas</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Koragas</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Kuruchian</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Mullukurunan</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Mullukurumba</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Bettakurumba</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Paniya</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
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<tr>
<td></td>
<td>Yerava</td>
<td>Dravidian</td>
<td>Cordaux et al., 2003</td>
</tr>
<tr>
<td></td>
<td>Irula</td>
<td>Dravidian</td>
<td>Roychoudhury et al., 2001</td>
</tr>
<tr>
<td></td>
<td>Kurumba</td>
<td>Dravidian</td>
<td>Roychoudhury et al., 2001</td>
</tr>
</tbody>
</table>

Source: Cordaux et al., 2003
The overall pattern of the dendrogram tree showed that the Indian populations clustered together, with the exception of the Kurumba, between the Caucasoids and Mongoloids (Figure I).

Source: Thomas et al., 2004

Figure I. Dendrogram tree of global population relationships with five Nilgiri tribes
1.3 Tribal situation in India

India has the largest concentration of tribal population in the world. This land has given shelter to 104.3 million people (Census of India, 2011). There are 533 different tribal communities spread all over India. As per official data, 258 tribal communities speaking about 106 different languages are notified as Scheduled Tribes. The total population of the Scheduled Tribes (ST) in India stood at 8,432,624 as per the 2011 census. The sex - ratio of ST population is 978 females per thousand males, being higher than that of the total population of the country as well as that of Scheduled Castes (SC's). The percentage - proportion of ST's to the total population in States and Union Territories is the highest in Mizoram (94.5%) and Lakshadweep (94.5%) followed by Nagaland (89.1%), Meghalaya (85.9%). Within major states, Chhattisgarh (31.8%) has the highest percentage followed by Jharkhand (26.3%) and Odisha (22.1%). The proportion is lowest in Uttar Pradesh (0.1 %), Bihar (0.9 %), Tamil Nadu (1.0 %) and Kerala (1.1 %).

Madhya Pradesh accounts for the highest percentage - proportion of ST population (14.5%) followed by Maharashtra (10.2%), Odisha (9.7%), Gujarat (8.9%), Rajasthan (8.4 %), Jharkhand (8.4%) and Chhattisgarh (7.8%). In fact, 68 % of the country’s ST population lives in these seven states only (Figure II).

Among the Scheduled Tribes, there are some who belong to more "backward" classes than the others. They have been classified as primitive tribes. They are characterized by low level literacy, declining or stagnant population and are economically backward Primitive Tribal Groups (PTGs) spread over 17 States / UTs and are 75 communities in number.

1.4 Tribal situation in Tamil Nadu

Tamil Nadu is second to Kerala in terms of Human Development Index (HDI). The total population of Tamil Nadu, as per the 2011 Census is 72,147,030. Of this, 7,94,697 are Scheduled Tribes (STs). Thirty six STs have been notified in Tamil Nadu by the Scheduled Castes and Scheduled Tribes Order (Amendment Act, 1976). Of the 5.74 lakhs tribals, 51.03 % of the tribals are males and remaining 48.97 % are females. The main tribes in Tamil Nadu are Malayali, Todas, Kurumbas, Paniyans, Irulas, Kattunayakans, Kanikar, Paliyan, Sholagar, Kadars, Vedars etc. of which Todas, Kotas, Kurumbas, Kattunayakans, Paniyans and Irulas are the Primitive Tribes. Malayali have been notified in Dharmapuri,
Vellore, Tiruvannamalai Pudukkotai, Salem, Namakkal, Villupuram, Cuddalore, Tiruchirappalli, Karur and Perambalur districts, Kurumbaas in the Nilgiri district, Kanikaran in Kanniyakumari district and Shencottah taluk of Tirunelveli district. Kammara, Kotas and Todas have been notified throughout the state except Kanniyakumari district and Shencottah taluk of Tirunelveli district.

Figure II. Tribal dominated states of India
Table II. Status of tribal population in Tamil Nadu

<table>
<thead>
<tr>
<th>S. No.</th>
<th>District name</th>
<th>Total population</th>
<th>ST population</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Ariyalur</td>
<td>7,54,894</td>
<td>10,722</td>
</tr>
<tr>
<td>2</td>
<td>Chennai</td>
<td>46,46,732</td>
<td>10,061</td>
</tr>
<tr>
<td>3</td>
<td>Coimbatore</td>
<td>34,58,045</td>
<td>28,342</td>
</tr>
<tr>
<td>4</td>
<td>Cuddalore</td>
<td>26,05,914</td>
<td>15,702</td>
</tr>
<tr>
<td>5</td>
<td>Dharmapuri</td>
<td>15,06,843</td>
<td>63,044</td>
</tr>
<tr>
<td>6</td>
<td>Dindigul</td>
<td>21,59,775</td>
<td>8,064</td>
</tr>
<tr>
<td>7</td>
<td>Erode</td>
<td>22,51,744</td>
<td>21,880</td>
</tr>
<tr>
<td>8</td>
<td>Kancheepuram</td>
<td>39,98,252</td>
<td>41,210</td>
</tr>
<tr>
<td>9</td>
<td>Kanniyakumari</td>
<td>18,70,374</td>
<td>7,282</td>
</tr>
<tr>
<td>10</td>
<td>Karur</td>
<td>10,64,493</td>
<td>575</td>
</tr>
<tr>
<td>11</td>
<td>Krishnagiri</td>
<td>18,79,809</td>
<td>22,388</td>
</tr>
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<td>12</td>
<td>Madurai</td>
<td>30,38,252</td>
<td>11,096</td>
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<td>13</td>
<td>Nagapattinam</td>
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<td>14</td>
<td>Namakkal</td>
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<td>57,059</td>
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<td>15</td>
<td>Perambalur</td>
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<td>2,584</td>
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<td>16</td>
<td>Pudukkottai</td>
<td>16,18,345</td>
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<tr>
<td>17</td>
<td>Ramanathapuram</td>
<td>13,53,445</td>
<td>1,105</td>
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<td>18</td>
<td>Salem</td>
<td>34,82,056</td>
<td>1,19,369</td>
</tr>
<tr>
<td>19</td>
<td>Sivaganga</td>
<td>13,39,101</td>
<td>790</td>
</tr>
<tr>
<td>20</td>
<td>Thanjavur</td>
<td>24,05,890</td>
<td>3,561</td>
</tr>
<tr>
<td>21</td>
<td>The Nilgiris</td>
<td>7,35,394</td>
<td>32,813</td>
</tr>
<tr>
<td>22</td>
<td>Theni</td>
<td>12,45,899</td>
<td>1,835</td>
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<tr>
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<td>Thiruvallur</td>
<td>37,28,104</td>
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<tr>
<td>24</td>
<td>Thiruvarur</td>
<td>12,64,277</td>
<td>3,034</td>
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<td>25</td>
<td>Thoothukkudi</td>
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<td>4,911</td>
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<td>26</td>
<td>Tiruchirappalli</td>
<td>27,22,290</td>
<td>18,198</td>
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<tr>
<td>27</td>
<td>Tirunelveli</td>
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<td>28</td>
<td>Tiruppur</td>
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<td>5,458</td>
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<tr>
<td>29</td>
<td>Tiruvannamalai</td>
<td>24,64,875</td>
<td>90,954</td>
</tr>
<tr>
<td>30</td>
<td>Vellore</td>
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<td>31</td>
<td>Viluppuram</td>
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<td>32</td>
<td>Virudhunagar</td>
<td>19,42,288</td>
<td>2,294</td>
</tr>
</tbody>
</table>

Source: Census of India, 2011
As per the 2011 Census report of India, among the numerically large STs, Kurumbas have reported the highest growth rate of 43% followed by Malayali (24.2 %), Irulas (12.1 %) and Kattunayakan (5.8 %) as in Table II. They are widely dispersed in 30 districts of Tamil Nadu. The tribal areas of Tamil Nadu can be broadly divided into two major geographical dimensions, viz., the Eastern coastal line, and the mountainous regions of the North and West. The important hill ranges of Tamil Nadu where the tribes are inhabited are the Jawadhi Hills, Yelagiri Hills of North Arcot district, the Kalrayan Hills of South Arcot, the Pachamalai, Kollimalai and Yercadu ranges of Salem, the Anamalai of Coimbatore, the Sitteri Hills of Dharmapuri, the Palani Hills of Madurai, and the Nilgiri Hills. The STs living in the forests of the Nilgiri Hills and in the Eastern and Western Ghats constitute 1% of the total population.

1.5 The tribal situation in the Nilgiri district of Tamil Nadu

The Nilgiris is the least populated district in Tamil Nadu with 7,94,697 (Census of India, 2011). The Badagas, Todas, Kotas, Kurumbas, Irulas, Kattunayakans and Paniyans mainly share the Nilgiri Hills. They constitute the dominant group among different tribal groups in this district, with this district having the maximum tribal population among the districts of Tamil Nadu. According to Census of India (2011) the total tribal population in the Nilgiri district is 32,813, which constitute nearly 3.72% of the Nilgiri district population. Among the main tribes in Tamil Nadu Todas, Kotas, Kurumbas, Kattunayakans, Paniyans and Irulas form the PTs and these groups are not evenly distributed among the six taluks of the Nilgiri district. Around 2,354 of tribes live in Coonoor, around 6,616 of them live in Gudalur taluk, 6,312 live in Kotagiri taluk, 521 live in Kundah taluk and around 10,897 and 6,113 tribal people live in Pandalur and Udhagamandalam taluk, respectively Figure III. The majority of the tribal inhabitance was observed in rural areas in Kotagiri, Kundah, Pandalur and Udhagamandalam while in Coonoor and Gudalur, the major tribes were settled in urban areas (Appendix II).

The tribe wise population is broadly classified as (i) Hill Tribe Foragers (HTF) consisting of Irulas and Paniyans having a total dominant population of 6020 and 7882, respectively; (ii) Hill Tribe Cremating (HTC) consisting of Kattunayankans with a total population of 2480 and Kurumbas having 6552 people; (iii) Hill Tribes Kannada (HTK) consisting of Kotas and Todas with a population of 2024 and 1608, respectively. Among
the six Primitive Tribal Groups (PTGs) it is observed that the Paniyans, Kurumbas and Irulas are the dominant tribal populations in the district (Table III). Moreover, it is observed that the female population is higher compared to the males in all tribal groups except Kattunayankans. A detailed description of each of the PTG is as follows.

(i) Irulas

The Irulas are the largest tribal groups in the Nilgiris. They are forest-based community like the Kurumbas and live mostly on the lower eastern slopes in uni-ethnic settlements or together with Kurumbas, with whom they have economic exchanges and maintain friendly relations. The Irulas grow millets and harvest fruits like lemon, jack, orange and bananas around their settlements. The two ethnic groups help each other in
growing their crops in shifting cultivation. In the Nilgiri district, Irulas are found in the lower regions of the hills. Irulas strictly follow community level endogamy. They also prefer cross-cousin consanguinity. Monogamy is the common form of marriage. Polygamy is commonly observed but polyandry is strictly forbidden in this tribal community.

(ii) Paniyans

The word ‘Paniyan’ means ‘servant’ in both Malayalam and Tamil. They are the traditional farm labors for Chetty landlords. They do not easily mingle with other tribal communities of these areas. The Paniyans usually avoid marrying their cross-cousins. Monogamy is the most common form of marriage among Paniyans whereas, polygyny form of marriage is also found in few settlements in the district. However, polyandry form of marriage is completely absent in Paniyan society. Levirate form of marriage is prevalent in both male and female.

(iii) Kattunayakans

This tribal community is also found only in Pandalur and Gudalur Taluk. They are also known as kadu or shoal nayakans. The Kattunayakans are unique endogamous tribe. In order to regulate their endogamy marriage system they have clan wise exogamy pattern and avoid marrying within clan. Cross-cousin consanguinity marriage is practiced and also they prefer consanguinity of marrying maternal uncle and niece. Sasikumar (1999) reported that there is no ban on the marriage between the members of the same settlement in recent times. Senior sororate system is absent but in few cases they practice junior sororate where the husband engages in marriage or relationship with deceased or infertile wife’s sister.

(iv) Kurumbas

The Kurumbas are the forest based tribes and they provide forest produce to the others, such as honey, bees wax, herbal plants and therapy, baskets, winnows and large grain storage baskets. Kurumbas are the least civilized group in the district and they mostly live in the hill slopes. There are five divisions of Kurumba viz., Allu Kurumbas, Jenu Kurumbas, Betta Kurumbas, Urali Kurumbas and Mullu Kurumbas, each group maintain different ethnic social organisations and social design based on their living habitats. All these five groups follow endogamy to regulate their marriage systems and cross-cousin marriage is practiced except for Mullu Kurumbas who follow monogamy form of...
marriage. Polyandry form of marriage is forbidden, but polygyny system is a status symbol of this community.

(v) Kotas

The Kota people are the blacksmiths and smiths in gold and silver, artisans and musicians. They live scattered in seven regions of the Nilgiri district. Each Kota family provide pots metals-tools such as knives and other metal works to certain number of Toda families and receives ghee, buffaloes, and calves in return. The Kotas are within their cultural frame work and they are strict followers of their culture. Both monogamy and polygamy form of marriage is practiced among them but most of Kotas prefer monogamy. Polyandry is not prevalent among the Kota people.

(vi) Todas

The Todas are the original inhabitants of the Nilgiri Hills and they are one of the most picturesque tribes in India. The Nilgiri Upper Plateau was the habitation zone of the Toda tribe, who were a semi-nomadic pastoral people. They had herds of semi-wild hill buffaloes with different pastures during the dry season from January to June. Their settlements in the midst of shola forests with good surrounding pasture land and running water nearby are called munds (actually mod, which means a herd of cattle). The main source of Toda livelihood was the buffaloes that they breed with great love and care, and their worship is classically described as patriarchal, the Todas actually possess some patrilineal and matrilineal divisions. They were polyandrous (Murray, 1984).

1.6 Economic values of tribes

Tribal economy is intimately connected with the forests. For centuries, the tribal have lived in the fringes of forests and depended entirely on forests for their livelihood. Even today, forest products continue to be the main source of income and sustenance for many tribal communities. These communities live in poverty having very little access to capital assets, health and educational facilities and hardly any protection against vagaries of nature.

However, since the tribal people treated land as a common resource, they rarely had land titles, and thus, lost their lands to outsiders when exploitation of forest resources began on a significant scale. This ensured that a majority ended up as small and marginal landholders.
1.7 Health status of tribe

The health status of tribal population is very poor and worst for primitive tribes because of the isolation, remoteness and being largely unaffected by the developmental process going on in other parts of India. According to Willis *et al.* (2004), they are the most marginalized population groups experiencing extreme levels of health deprivation. The unfavourable health status of tribes in developed countries has been shown across a range of outcomes, including mortality (Bramley *et al.*, 2004), disease (Anand *et al.*, 2001), health behaviours (Gaiser, 1984; Frank *et al.*, 2000) and health care (Finger, 2003; Johnston and Coory, 2005). Few systematic accounts of the health of tribes in developing countries was reported by Escobar *et al.* (2001) and Seale *et al.* (2002).

Research studies on tribes indicate that the primitive tribes have distinct health problems, mainly governed by multidimensional factors like their habitat, different terrain; ecological variable niches, illiteracy, poverty, isolation, superstition and deforestation (Nanjunda, 2010). Most of the tribes are living below the poverty line as defined by Government of India. Different studies and reports have shown that landlessness and poor educational status very often lead to poor utilization of health services (Shukla and Solanki 1985). Tribal communities have become impoverished and marginalized due to their alienation from land and forest. The resources on which their survival depends, loss of livelihood, the question of rights of the adivasis to the natural resources, issues concerning conservation and the role of local communities is a major concern. Prevailing systemic denial of farmland to the landless and land-poor entrenches poverty on the one hand and on the other leaves these marginalized communities to increase the pressure on natural habitats for livelihood (Faizi and Ravichandran, 2008). The resistance of the adivasis to the injustice perpetrated by the state and the responsibility of the state towards the most vulnerable part of our population is often not brought to light. The health status of any community is influenced by the interplay of health consciousness of the people, socio-cultural, demographic, economic, educational and political factors. The common beliefs, traditional customs, myths, practices related to health and disease in turn influence the health seeking behaviour of autochthonous people (Balgir, 2004a).

The health problems need special attention in the context of tribal communities of India. Available research studies point out that the tribal population has distinctive health
problems which are mainly governed by their habitat, difficult terrains and ecologically variable niches (Basu, 2000). The health, nutrition and medico-genetic problems of diverse tribal groups are unique and present a formidable challenge for which appropriate solutions have to be laid out through planning and evolving relevant research approaches. Unhygienic conditions, ignorance and health education are the main factors responsible for their ill health (Basu, 2000). ST populations continue to carry high burdens of ‘diseases of the poor’, namely under nutrition and infectious diseases. High levels of chronic malnourishment have been observed among child and adult populations (Bose et al., 2006).

Almost all the indices of health indicate that the status of tribals was poor (Lakshmi Narayan, 1950). The report of the working group on development and welfare of ST during the Eighth Five Year Plan (GOI, 1989) indicates that diseases like goitre, yaws (infection on skin and bones), malaria and guinea worm were endemic to tribal pockets. Basu (1993a) reported an alarmingly high incidence (40 %) of SCD among the Adiyan tribal group of North Wayanad, Kerala. The author also reported high frequency (19%) of glucose-6 phosphate dehydrogenase red cell deficiency among the tribals of Bastar, Madhya Pradesh.

1.8 Disease among tribes

The common health problems among tribes are malnutrition, which is a big issue inclusive of vitamins A, C, B complex deficiencies, under-nutrition of mothers along with anaemia as a result of food taboos, protein energy under-nutrition and few cases of vitamin deficiencies in children due to general lack of awareness of child care and infant feeding practices. Among most of the tribes, gastrointestinal disorders, particularly dysentery and parasitic infection are very common leading to morbidity and malnutrition, diarrhoea, dysentery, skin diseases and respiratory diseases. It observed that 21% of children suffer at least two bouts of diarrhea every year and 22% suffer from at least two attacks of respiratory infections (IIPS report, 2000). In addition, communicable diseases such as tuberculosis, malaria and STDs are major public health problems. Some tribal groups are also at high risk for sickle cell anaemia. Generally, tribal diets are seen to be deficient in protein, iron, iodine, and vitamins. High prevalence of goitre, among women of child bearing age groups are seen in tribes due to habitation in hilly areas and limited access to sea foods. Most of tribes observed by anthropologists and voluntary organisations appear to have a few common practices regarding maternal and child care; Expectant mothers are expected to restrict their
diet and quantity as there is a common fear that if the baby is too large, delivery would be difficult and might lead to death of the mother (Sahni and Xirasagar, 1990).

Tribes account for 25% of all malaria cases occurring in India and 15% of falciparum cases (Chhotray, 2003). Intestinal helminthiasis is widely prevalent among tribal children (up to 50% in Orissa and 75% in MP) (Chhotray, 2003 and Basu, 1993b). Skin infections such as tinea and scabies are seen among tribals due to poor personal hygiene. Sexually transmitted diseases are relatively more common (7.2% prevalence of syphilis) among tribes of Kolli hills of Tamil Nadu (Kalaivani et al., 2001). Respiratory problems are due to smoke emitted by fireplace in ill-ventilated huts to keep warm during cold nights (Naidu, 2007). Primitive tribal (particularly vulnerable) groups (PVG) of India face special health problems such as genetic abnormalities like sickle cell anaemia-glucose 6-phosphate (G6PD) red cell enzyme deficiency. The health and nutrition related challenges of the vast tribal population of India are as varied as the tribal groups themselves.

1.9 Brief history of SCD

Although the HbS (Haemoglobin S) gene is most common in Africa, SCD went unreported in African medical literature until the 1870s. This is because symptoms were similar to those of other tropical diseases in Africa. The growing knowledge of these conditions of mutation has begun at the turn of 19th century; however African tribal populations were all too familiar with the disease and created their own names for it.

Later as the scientific investigation of the disease was set in motion by Herrick (1910), a Chicago physician gave the first description of SCD, who noted that a student patient from Grenada (West Indies) had an anaemia characterized by unusual “elongated and sickle-shaped” red blood cells. However, Mason (1922) used the term sickle cell anaemia for the first time.

In 1927, Hahn and Gillespie associate the sickling of red blood cells with low oxygen conditions. They found that the change occurs at partial pressures of O₂ prevalent in the body and produces anaemia and other disorders, termed sickle-cell disease.

Later in 1940, Sherman reported that the sickling of Red Blood Cells (RBCs) in the absence of oxygen is caused by a change in the haemoglobin molecule structure.
Pauling *et al.* (1949) using new technique of protein electrophoresis showed that
the haemoglobin from patients with SCD is different than that of normals. This made the
SCD as the first disorder where an abnormality was known to be at fault in a protein.

Allison (1954a) reported a relationship between sickle cell trait and *Plasmodium falciparum* malaria and that the sickle cell gene is maintained as true polymorphism in the
population by having a selective survival advantage to the heterozygotes.

In 1957, Ingram first sequenced sickle haemoglobin and showed that a glutamic
acid at position 6 was replaced by a valine in SCD. Hence using the known information
about amino acids and the codons, he was able to predict the mutation in SCD. This lead to
the identification of the first genetic disorder among human beings.

Flavell *et al.* (1978) prepared maps of the human beta and delta globin genes,
pinpointing the DNA mutation.

Charrache *et al.* (1995) reported that the anticancer drug hydroxyurea is the first to
reduce the frequent, painful complications that characterize SCD.

**1.10 Origin of Sickle cell gene**

Initially the single mutation theory was put forward in which it was conveyed that
a single mutation occurred in Neolithic times in the then fertile Arabian Peninsula
(Lehmann, 1964). It is believed earlier, that the changing climatic conditions caused the
migration of people that could have carried the gene to India, Eastern Saudi Arabia and
down to Equatorial Africa. This hypothesis was supported by citing the distribution of
certain agricultural practices and anthropological evidences, but it is now quite clear that
the sickle cell mutation has occurred as several independent events. By using a series of
different restriction endonucleases, different chromosome structures (haplotypes) are
identified and HbS gene has been found to be linked to certain commonly occurring
haplotypes that are generally different from those bearing the HbA (Haemoglobin A) gene
(Wainscoat *et al.*, 1983; Antonarakis *et al.*, 1984). The study observes that the sickle cell
mutation has occurred on at least three occasions in the African continent where at least
once in either the Arabian Peninsula or the Central India and migration from their home
sites to the other regions has also occurred. This explains that there is wide spread...
chromosomal heterogeneity of B(s) gene cluster haplotypes as compared to the homozygous condition in Africa, Arab or Asia (El-mouzan et al., 1989). However, it is believed that this gene would have developed between 3000 and 6000 generations, approximately 70000-150000 years ago (Kurnit, 1979; Solomon and Bodmer, 1979).

The existence of haplotypes specific to certain regions of the world suggests that the mutant beta globin gene arose separately in Cameroon and Arabo-Indian (or Asian) regions (Oner et al., 1992). All of the areas in question have been are now endemic locations of malarial infection. This observation is consistent with the idea that the high incidence of sickle mutation in these areas is derived from natural selection (Carlson et al., 1994). The mutation that produces sickle haemoglobin occurs spontaneously at a low rate. People with one sickle haemoglobin gene and one normal haemoglobin gene (sickle cell trait) are somewhat more resistant to malaria than people with two normal haemoglobin genes. The widely accepted theory is that HbS offers selective protection against falciparum malaria because of the induction of sickling at physiological oxygen tension by *Plasmodium falciparum* followed by sequestration of parasitized red cells deep within reticulo-endothelial system where microenvironment is hostile for parasite growth (Friedman, 1978; Pasvol and Weatherall, 1979). Thus, people with sickle cell trait would have a better chance of surviving an outbreak of malaria and passing their genes (sickle and normal haemoglobin) to the next generation when they have children. Another theory states that the remarkable stability of sickle gene in Africa remained relatively at a constant level in a population without being eliminated and it is to be thought of as the most widely accepted theory of balanced polymorphism (Allison, 1954a; 1954b; Luzzatto and Reddy, 1970). The global distribution of SCD data points and HbS allele frequency is presented in Figure IVa and IVb.

**1.11 Origin of Sickle cell gene in India**

SCD in India exists mainly in tribal populations, who remain relatively isolated from the mainstream of the society. The likelihood is low that an influx of a sickle cell gene from outside India occurred at a degree to account for rates of heterozygosity that reach up to 35% in some tribes. However, still there has been much debate about the origin of sickle cell gene in India. Inadequate evidence is available to identify whether SCD is indigenous in origin or it had migrated from Africa to India in prehistoric times. With the help of endonuclease restriction enzymes (Kan and Dozi, 1980), the association of sickle
cell gene with 7.6 kb recognition site was found among the Valmiki, Konda Reddy and Koya Dora tribes in southern India (probably Andhra Pradesh) and it is different from its association with 13.0 kb recognition site among the people of Western Africa (Kan and Dozi, 1980). These findings were in favour of a view that the Indian and West African sickle gene mutations arose by separate events. Later on, limited or no attempts were made to further study the origin of sickle cell gene by advanced molecular biology techniques. For many years, the sickle cell gene has been considered confined to people of African ancestry; although the gene was described in southern India, in persons without African origin, as early as 1952 (Lehmann and Cutbush, 1952).

However, over the next 30 years, population screening and the reports of Anthropological Survey of India (Negi, 1972) stated that established sickle cell trait frequencies are up to 35% throughout much of central India with the highest frequencies occurring in Odisha, followed by Assam, Madhya Pradesh, Maharashtra, Uttar Pradesh, Tamil Nadu and Gujarat (Balgir, 1996a; Ambedkar et al., 2001).

1.12 Disease characteristics

The most important protein of Red Blood Cells (RBCs) is haemoglobin (Hb), which is the component that carries oxygen from the lungs to all parts of the body. Haemoglobin consists of four globin chains, two alpha-globin and two beta-globin, each folded around a haem molecule (Schnog et al., 2004). Haemoglobin is the component that carries oxygen from the lungs to all parts of the body. The gene related to sickle cell anaemia is the haemoglobin gene (HBS). The HBB gene provides instructions for making beta-globin. Various versions of beta-globin result from different mutations in the HBB gene. One particular HBB gene mutation produces an abnormal version of beta-globin known as Haemoglobin S (HbS). There are nearly 300 types of Hb, the most being HbA which refers to adult haemoglobin (Figure V).

Other mutations in the haemoglobin gene lead to additional abnormal versions of beta-globin such as Haemoglobin C (HbC) and Haemoglobin E (HbE). HBB gene mutations can also result in an unusually low level of beta-globin; this abnormality is called beta thalassemia. In people inherited with SCD; at least one of the beta-globin subunits in haemoglobin are replaced with haemoglobin S. HbS has a substitution of valine for glutamic acid at the sixth position of the beta globin chain. According to Allen (2005),
sickle cell anaemia is an inherited abnormal Haemoglobin formation due to the presence of Haemoglobin S (HbS).

![Diagram of HbA, HbS, and HbSS molecules]

**Figure V. Protein of Haemoglobin A, Haemoglobin S, Haemoglobin SS**

### 1.13 Mechanism of HbSS

A red blood cell containing only HbS in the absence of normal HbA is capable of carrying oxygen but when the HbS gives up its oxygen to the tissue, the HbS molecules becomes crystalline (Allen, 2005). These crystals are sticky and form long rods inside RBC. The RBC become rigid, inflexible, and sickle-shaped unable to squeeze through small blood vessels and therefore blocks small blood vessels (Figure VI).

The sickle-shaped red blood cells regain their original round shape when deoxygenated, but on repeated oxygenation and deoxygenation, the cells become increasingly hard and brittle and irreversibly sickled (Allen, 2005). The sickle-shaped RBCs die prematurely, which can lead to anaemia. The cells are destroyed by the
reticuloendothelial system once they are sickled. Normal RBCs have a life span of 120 days, but, sickled cells have a life span of only 15 days (Dix, 2001).

1.14 Genetics of Sickle cell

SCD is an inherited blood condition resulting through the inheritance of abnormal genes from both parents (Serjeant, 2006). SCD denotes all genotypes containing at least one sickle gene, in which HbS makes up at least half the haemoglobin present. Usually, individuals receive two copies of the haemoglobin gene, one from each parent. Individuals with SCD do not have any copies of the haemoglobin A gene. Instead, these individuals have two copies of an alternative form of the haemoglobin gene. At least one of those two alternative genes must be haemoglobin S in order to result in SCD. Rarely, a person has one gene for haemoglobin S and another of a rare form of haemoglobin, such as haemoglobin O. The most common type of SCD is haemoglobin SS disease.

An individual receives a copy of haemoglobin A from one parent and a copy of haemoglobin S from the other known as carrier. The carrier state for SCD is often referred to as "sickle cell trait" (Koch et al., 2000). Sickle Cell Trait is not a disease and does not change into SCD later on in life. However, if two individuals with Sickle Cell Trait have a child together, that child is at risk (25%) for having SCD. People who have sickle cell trait
usually have no symptoms and lead normal lives. However, they can pass the sickle cell gene to their children.

(i) Inheritance Patterns

SCD is a recessive gene if two parents have two copies of the HbS gene. Children born to parents with these genes (Figure VII) will have SCD (Bloom, 1995).

(ii) One Parent has Sickle Cell Disease and Sickle Cell Trait

Figure VIII shows that one parent has the SCD HbS genes; therefore, all of that parent’s gametes will carry the HbS gene. The other parent has one HbS gene and one HbA gene. The chances for that parent with both the HbS gene and the HbA gene transmitting either gene are equal, or 50/50 (Bloom, 1995). All children born to these parents will have either SCD or sickle cell trait children with the chances being 50/50 for each.
(iii) **One parent has sickle cell disease and the other parent carries normal genes**

Figure IX illustrates that the HbA gene is carried by the normal parent and the gametes from this parent will carry this particular gene, as well as the SCD parent carrying the HbS gene (Bloom, 1995). All children conceived from these parents will inherit one normal and one sickle cell gene. All children will have the sickle cell trait.

![Figure IX. One parent with sickle cell disease and the other parent carries normal genes](source)

(iv) **Two parents have sickle cell trait**

Figure X illustrates that both parents have an equal chance of transmitting the two genes. If both parents have the sickle cell trait, they will have a 25% chance of having children with SCD, as well as a 50% chance of carrying the sickle cell trait (Bloom, 1995). Also, a slight 25% chance occurs to produce children with normal genes. In this case, 50% of the chances are that one parent will transmit an HbSS gene and the other chances are that the other parent will transmit an HbA gene (Bloom, 1995).

![Figure X. Two Parents with sickle cell trait](source)
(v) One parent is normal and the other has sickle cell trait

All children from this couple (Figure XI) will display the sickle cell trait or have normal genes because half of the gametes of the parent who carries the sickle cell trait will carry the HbS gene, and the other half will carry the HbA (Bloom, 1995). This combination will result in a 50/50 chance of producing a child with normal genes or inheriting sickle cell traits.

Source: Bloom, 1995

Figure XI. One parent with the sickle cell trait and the other as normal

1.15 Types of SCD

There are three main types of SCD: Haemoglobin SS disease where individuals have received two copies of the haemoglobin S gene; Haemoglobin SC disease where individuals have received one copy of the haemoglobin S gene and one copy of the haemoglobin C gene. Haemoglobin SB (beta) Thalassemia disease including SB0 (beta zero) and SB+ Thalassemia disease where the individuals have received one copy of the haemoglobin S gene and one copy of the haemoglobin beta- thalassemia gene. The beta-thalassemia genes result in reduced (B+) or lack of expression (B0) of normal haemoglobin A, resulting in a person either having only haemoglobin S or mostly haemoglobin S with a small percent of normal haemoglobin A.

1.16 Clinical features

According to World Health Organization (WHO) report (2005), Sickle cell anaemia covers a wide spectrum of illness. Most affected people have chronic anaemia with a haemoglobin concentration of around 8 g/dl. In children, sickle-shaped red blood cells often become trapped in the spleen, leading to a serious risk of death before the age of seven from a sudden profound anaemia associated with rapid splenic enlargement or splenic malfunction.
causing an overwhelming infection. Children between 6 and 18 are affected with painful swelling of the hands and/or feet (hand-foot syndrome). Progressive ischaemic atrophy of the spleen from ischaemia results in increasing the risk of sepsis (particularly pneumococcal). Haemolysis results in gall stones formation and related diseases. Sequestration of sickle cells in spleen and liver causes sequestration crisis among these people.

1.17 Epidemiology of SCD

About 5% of the world’s population carries gene responsible for haemoglobinopathies and among them sickle cell disorders account for about 70% of the worldwide haemoglobin disorders (Angastiniotis et al., 1995). It is the second most common haemoglobinopathy in the world. SCD is commonly found among people of tropical countries and transmitted as autosomal recessive character (Gupta et al., 1991). The incidence of this condition is most common in people of Africa, Sub-Saharan, Mediterranean and of Indian origin. As per WHO report of 1983, 60 million carriers of sickle cell and 1,20,000 sickle cell homozygotes are added every year in the world. With a population of 1000 million at the millennium (2000) year and a birth rate of 25 per 1000 liveborns, there would be about 45 million carriers and about 15,000 infants born each year with haemoglobinopathies in India (Balgir, 2000 a). Based on 1981 population census in India, it was estimated that there were 24,34,170 carriers and 1,21,375 sickle cell homozygotes among the tribes of India (Rao, 1988). There were 1,86,096 sickle cell anaemia cases present in the Indian subcontinent according to Bhasin et al. (1994).

SCD is widespread among the ethnic groups or tribal groups of India. This was estimated based on the prevalence rates of sickle cell haemoglobin as there were over 50,00,000 carriers and 2,00,000 homozygous SCD cases among tribals alone in India (Malhotra, 1993). It has been reported that nearly 20 million people suffer from this disease in India (Ghai, 2000). It is well documented that the gene for Sickle cell haemoglobin is located on the short arm of chromosome 11 and has an autosomal inheritance. Hence, it can manifest in two forms viz., Heterozygous (HbAS) carrier and homozygous (HbSS) sufferer.

In India, this gene was first described among tribal group of the Nilgiri hills (Lehmann and Cutbush, 1952), the incidence varied from 5% to 34% and it was mainly restricted to the tribal population. In 1955, Buchi confirmed the presence of the disease in
Veddoids of South India which was followed by Sukumaran et al. (1956) who reported its prevalence in Western India which later spread to central India where the prevalence rate ranged between 9.4 to 22.2 % as per Shukla and Solanki (1985). Gorakshakar (2006) reported that during the last fifty years several groups of investigators conducted hospital based epidemiological surveys among various ethnic groups. According to this survey, it was noted that the prevalence of SCD was found to be 0 to 18 % in North-eastern India, 0 to 33.5 % in western India, 22.2 to 44.4 % in central India and 1 to 40 % in southern India. However, the first review on sickle cell haemoglobin in India was done by Balgir and Sharma (1988) and followed by Balgir (1996a; 1996b; 2001; 2004b) which highlighted the wide spread distribution of the sickle cell disorders in India. The following Table IV depicts the prevalence of sickle cell anaemia among states of India (Rao, 1991).

Table IV. Prevalence of SCD in states of India

<table>
<thead>
<tr>
<th>States</th>
<th>Prevalence of Sickling (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Andhra Pradesh</td>
<td>0-34.6</td>
</tr>
<tr>
<td>Bihar</td>
<td>0-0.6</td>
</tr>
<tr>
<td>Gujarat</td>
<td>0-30</td>
</tr>
<tr>
<td>Karnataka</td>
<td>0-25</td>
</tr>
<tr>
<td>Kerala</td>
<td>0-29.7</td>
</tr>
<tr>
<td>Maharashtra</td>
<td>0-45.4</td>
</tr>
<tr>
<td>Orrisa</td>
<td>0-12.4</td>
</tr>
<tr>
<td><strong>Tamil Nadu</strong></td>
<td><strong>0-35.3</strong></td>
</tr>
<tr>
<td>Utter Pradesh</td>
<td>0-32.6</td>
</tr>
<tr>
<td>West Bengal</td>
<td>0-1.1</td>
</tr>
<tr>
<td>Madhy Pradesh</td>
<td>0-48.5</td>
</tr>
</tbody>
</table>

*Source: Rao (1991)*

The Nilgiri district records the highest tribal population accounting to 5 to 10 % of the total mass of Tamil Nadu. Many studies on the genetics of tribal groups in Tamil Nadu have reported the red cell enzyme and serum protein polymorphisms, such as PGM1, PGH13, ACP, AK, LDH, GLO, ESD, HP, TF, GM and G6PD among the Irulas, Kurumbas and Todas groups (Kirk et al., 1962; 1963; Saha et al., 1976). Sickling trait has been studied among the Irulas, Paniyans, Todas, and Kurumbas (Lehmann and Cutbush, 1952; Lehmann and Sukumaran, 1956; Buchi, 1959; Negi, 1967; Ghosh, 1973; Saha et al., 1976; Ghosh et al., 1977; Das et al., 1977; Ramasamy et al., 1994). Studies have also been
carried out on HLA polymorphism in the Kotas and the Badagas of the Nilgiri Hills (Selvakumar et al., 1987) and the Koyas of Andhra Pradesh. Many of these studies have reported high incidences of SCD (30%) among tribes living in hill areas (Bhatia and Rao, 1987). According to the studies reported by All India Institutes of Medical Sciences (AIIMS, 1990), the prevalence of SCD in three tribes such as Kurumbas, 5.1 % (HbSS) and 20.4 % (HbAS), Paniyans 1.5 % (HbSS) and 13.7 % (HbAS) and Kattunayakans 0.6 % (HbSS) and 11.6 % (HbAS) (AIIMS, 1990) was observed.

1.18 Factors that influence SCD

The wide spectrum of clinical and haematological presentation of SCD has involved considerable attention and several studies have been directed to identify the factors involved in modulating the nature of SCD (Weatherall et al., 1969; Pembrey et al., 1978; Wood et al., 1980; Al-Awamy et al., 1986; El-Hazmi et al., 1990). Carloyn and Harphan (1992) observed that health status of a population is influenced by the environmental conditions, health services, characteristics of the population and socioeconomic conditions. Dietary habits, socio-economic status, hygienic and sanitary conditions, climate and exercise are the factors that influence the health of SCD individuals. It is therefore, estimated that SCD patients living in healthy environments and belonging to a high socio-economic group may suffer from a few complications compared to those SCD patients who are devoid of proper health care and nutrition. In this respect, few environmental and socio economic factors may modulate the SCD presentation also.

(i) Environmental condition

Environmental factors can affect gene expression and genetic variation is a result of the type of environment where an individual lives and how humans reacts to different environmental factors (Juulia et al., 2009). Predominantly high altitude natives are naturally selected for traits that offset the unavoidable environmental stress of severe lifelong high-altitude hypoxia. The hypoxic condition can regulate many physiological and pathological processes in humans through a family of transcription factors such as hypoxia inducible factors (HIFs) (Eltzschig and Carmeliet, 2011; Muaj et al., 2012; Maes et al., 2012; Semenza, 2012). These variables can affect regulations of genes that contribute to phenotype expressions and they may also influence the interaction of disease phenotypes with fitness (Furrow et al., 2011). During the past few years, experimental evidence has
emerged to suggest that environmental factors may influence cellular proliferation attrition in an organ-specific manner (Semenza, 2012). In such cases and in disorders with no genetic background, environmental factors seem to influence and/or cause disease onset, progression and outcome (Rosenberg et al., 2011). The importance of environmental influences and their mechanisms hold great potential for increasing the understanding and further amelioration of SCD. The environmental conditions such as altitude and climatic variables play an important role in modifying the clinical presentation of the SCD. It is also unlikely that the prevalence of haemoglobinopathies is directly influenced by altitude, because the physiological effects of decreasing oxygen concentration are relatively minor within this altitude range (Dirren et al., 1994). When the oxygen level or environmental temperature drops (hypoxia), HbS containing red blood cells can sickle and these sickle cells stick to the walls of blood vessel, clump together and block their blood supply. According to Catherine (2009), sickling can be precipitated by environmental factors such as hypoxia, low pH, cold, and dehydration of the RBC, as well as adhesion molecules and cytokines associated with infections.

(ii) Socio-economic factors

In general, proper care and management of SCD tribal patients is believed to play a significant role in decreasing the clinical severity and associated complications of SCD among tribes. Due to their lower socio-economic status, the tribes are devoid of proper care and management of the disease. The socio-economic status, contributes to all the factors i.e. improved nutrition, proper medical care and prophylactic measure, improved overall health status, clean and healthy environment and good and proper sanitary measures. Knowledge of the demographic and socioeconomic profile of SCD patients is essential to identify their needs, to contribute to improving resource allocation and also to create and implement public health policies that benefit this population (Santos and Neto, 2013). Socioeconomic factors are not necessarily a direct cause of the disease, but also a deterrent to improving the quality of life. People in high distress situations are likely to lack the social support that is needed to improve their health status (Cassel, 1976; Faresjo, 1992).

(iii) Lack of health care delivery

An individual’s health status is influenced not only by the environmental, genetic and social determinants but also disparities of health care system. As Sutton et al. (1999)
states "the failure by healthcare providers to distinguish between addiction, dependence, and tolerance is a major component in the failure of effective management of the sickle cell patient with pain". With its disruptive occurrences of severe pain, frequent hospitalizations and the over abundance of medical complications that it produces, SCD can in itself be an immense challenge for its sufferers. Tribal communities with SCD are especially vulnerable and often face social exclusion because of their habitation in remote hard to reach areas and lack of availability and accessibility of basic health services. Typically, pregnant women or patients with pain crises from remote tribal hamlets will be unable to make it to health facilities in time. The lack of health care disparities among tribes include differences in geography, lack of access to adequate health coverage, communication difficulties, cultural barriers, poor socio-economic background, climate and lack of awareness.

(iv) Lack of awareness of SCD

Due to lack of awareness of SCD, most tribal population tend to fall ill more frequently and have to wait long before seeking medical help. Griffin (1997) reported to the SCD Association of America Dallas Chapter’s newborn screening program that challenges for proper SCD care included “a pervasive lack of awareness of sickle cell risk among the target population; a lack of effective parental notification and referral mechanisms to ensure family access to required testing, education, counseling and support services; inadequate parental health knowledge and health supervision to improve outcomes for children affected by SCD and inadequate medical homes, culturally competent, compassionate, comprehensive and continuous.” This is because in comparison with other chronic diseases, SCD remains one of the least understood and confusing medical conditions for the tribals (Clarke and Clare, 1981).

1.19 Geographical Information System for public health

Great leaps in the field of science are often made at the borders where different fields of research meet. Geographical Information System (GIS) in health research is one kind of field which proves of this. In many public health and epidemiology projects, GIS has emerged as an innovative and important component, sometimes even an essential tool. It is easy to determine spatial relationships between disease occurrence and other information that is geo-referenced differently from the disease data.
According to WHO, “GIS provide ideal platforms for the convergence of disease-specific information and their analysis in relation to population settlements, social, health services and the natural environment, highly suitable for analysis of epidemiological data and finding trends and interrelationships that would be difficult to reveal in tabular format. GIS allows policy makers to easily visualize problems that relate to health and social services and natural environment and accordingly to target resources effectively” (WHO, 2007).

Maps have long been used to describe geographic pattern of disease. Disease mapping is the usual means of presenting descriptive geographic data on disease and creating maps to show the intensity of the disease. The aim of the disease mapping includes:

- Simple description by showing or displaying a visual summary of geographical risk.
- Hypothesis generation by giving clues to cause of disease and/or factors that influence spread by informal examination of maps with exposure maps, components of spatial versus non-spatial residual variability may also provide clues to source of variability.
- Provide estimates of risk by area to inform public health resource allocation.
- Estimation of background variability to the underlying risk in order to place epidemiological studies in context.

GIS is commonly used to combine spatial data from different sources for mapping disease and to identify the causal factors of observed spatial pattern cluster (Robinson, 2000; Graham et al., 2004). Accurate and detailed information on population size and distribution are therefore of significant importance for deriving population-at-risk and infection movement estimates in spatial epidemiological studies (Tatem et al., 2011).

Disease mapping was developed and traced by Koch (2005) from a map of plague outbreaks at Bari, Italy in the seventh century to map AIDS for the entire earth in the present day. Koch reported that John Snow in 1850s mapped the location of cholera cases in Soho, London (Schellenberg et al., 1998; Walter, 2000). Later in 1978, maps were produced to understand the distribution of yellow fever in New York City, USA. Disease outbreak is a result of the combination of social, environmental, and individual disease variables and therefore mapping can be more meaningful for the analysis for spatial expression of the
disease. According to Myers et al., (2000), disease mapping is used to understand the geographical distribution and spread of disease in the past and present and most important it is used to forecast future disease spread or epidemic outbreaks and to identify the factors that may promote or inhibit disease transmission. Disease studies have revealed strong spatial aspects, including disease case location and disease diffusion. The mapping of disease can be used to pinpoint the areas where outbreaks originate and effectively target high-risk areas for early prevention and control thus; mapping spatial aspects of diseases could help people to understand some puzzles of disease outbreak. GIS is currently recognized as a set of strategic and analytic tools for public health, so the design and implementation of an information system for SCD control with GIS capacity should be considered.

Generally population is composed of genetics, gender and age, and is related to humans as biological entities that interact directly with the environment contributing to the development of disease (Meade and Earickson, 2000). Age is one of the most important factors to consider when describing the disease because age-specific disease rates usually show greater variation than rates defined by other attributes. Age is probably the single most important “person” attribute in disease mapping because almost every health-related event or state varies with age. The traditional approach to disease mapping uses age adjustment, which multiplies local disease rates by standard population weights (Pickle and White, 1995; Pickle, 2009). There are number of factors that vary with age such as susceptibility, opportunity for exposure and physiological response of the population.

Gender, typically described in terms of masculinity and femininity, is a social construction that varies across different cultures and over time (Wood, 1997). Gender is one of the complex traits that are particularly useful and important because they include the social dimensions necessary for understanding its impact on health and each has genetic underpinnings, to varying degrees. In general, males have higher rates of illness and death than females do for a wide range of diseases. For some diseases, this sex-related difference is because of genetic, hormonal, anatomic, or other inherent differences between the sexes. On the other hand, the sex-related differences in the occurrence of many diseases reflect differences in opportunity or levels of exposure. The significant differences in the health needs of women and men need to be clearly identified so that they can be reflected in equitable strategies for resource allocation. The risk and spread of diseases are
heterogeneous in space and hence the population distributions and counts should be resolved to higher levels rather than regional estimates (Robinson, 2000).

Therefore, mapping of relevant population attributes of interest, including age, gender, and community is important in order to find the influencing factors of SCD intensity. Age and sex has been used for many years as an important factor to enable researchers to easily investigate spatial trends and the nature of disease distributions. Therefore, there is an urge to map the disease pattern of SCD among tribal groups with respect to sex, age, SCD pattern using GIS.

1.20 Spatial and statistical analysis for SCD

The generation of data on prevalence and distribution of sickle cell disorder is essential for the effective study and management among the tribes. Disease maps (spatial distribution of disease) provides researchers with visual display and suggest thorough pattern of physical facilities and the human environment, useful avenues of research into casual process (Langford et al., 1999). Spatial epidemiology is defined as variation in disease risk or incidences. Therefore, a wider range of studies examine different determinants of health and clustering of health outcomes, either within demographic and socio-economic classification or spatial location (Wennberg, 1999; Bond, 2001; Ricketts and Pope, 2001).

Modelling of spatial analysis includes procedures for testing hypotheses about the causes of disease and the nature and processes of SCD disease transmission. In general, modelling involves the integration of GIS with standard statistical and epidemiologic methods. GIS can support in generating data for input to epidemiologic models, displaying the results of statistical analysis, and modeling processes that occur over space. Exploratory data analysis refers to spatial data that can be mapped in order to investigate the hypothetical relationships between disease and independent variables. The resulting maps from exploratory data analyses are often used to develop potential hypotheses and future research objectives (Clarke et al., 1996; McLafferty, 2003; Carr et al., 2005).

The study conducted by Noor et al., (2006) observed that cost-surface models have been used to estimate journey-time to health facilities in resource-poor settings, but have often focused only on journeys made on foot or else have dealt simplistically with varying modes of transportation (Tanser et al., 2006). Pedestrian journeys made without
mechanised transport are potentially more straightforward to model because average speeds will tend to be relatively similar across different settings and; different categories of paths, tracks and roads offer broadly similar walking speeds, journeys rarely deviate from the most available direct route (Langmuir, 1984). It was observed that the health status of the tribes was comprehensive and area-specific and health-related studies were limited. Most of the available studies were isolated, fragmentary and did not cover the various dimensions of health affecting the tribes like high degree of marital consanguinity, caste/class and geographical endogamy, certain irrational traditions, lack of medical facilities, beliefs aggravating the nutritional status and health, heavy cost of treatment, poor economy, backwardness in all spheres of life, etc.

To monitor and programme an effective control strategy, an assessment of the geographical distribution of the disease is needed (Neeru et al., 2009), through which the cause and spread of the disease can be mapped for better management. Hence, there is a need to map the prevalence of the disease pointing out highly endemic zones and also to identify risk within the tribal groups with respect to sex and age using GIS.

SCD is clinically one of the most important haemoglobinopathies and the outcome is very difficult to predict with very few therapeutic remedies. This disease is recognized as a critical health problem among the indigenous people in the different parts of the country. Though several attempts have been made to delineate the diseased tribes, basic research has stopped at the clinical diagnosis rather than determining the socio-environmental determinants for new incidence.

Technological support has advanced the medical field for high risk diseases diagnosis and treatment however, these genetic disorders lack attention. To aid medical help, a clear demonstration has to be made as to identify the lifestyles, age and gender based distribution, inequities in health access and the severe stress on the burdened. The present research work is a preliminary attempt to investigate the social and environmental influences on the disease burden and aims to approach the disease in a more social manner so as to reduce further burden.

The next section deals with a clear outline on disease status among the tribes and the focus of research problem undertaken in the present study.