A BIOSOCIAL STUDY ON β-THALASSAEMIA

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CERTIFICATE

Certified that the thesis entitled "A Biosocial Study on β-thalassaemia" is the record of original work done by Ms. Mondira Bhowmik, that the contents of this thesis did not form a basis of the award of any previous degree to her, or to the best of my knowledge to anybody else, and that the thesis has not been submitted by her for any research degree in any other University.

In habit and character Ms. Mondira Bhowmik is a fit and proper person for the degree of Doctor of Philosophy.

(A.K. Ghosh)
Supervisor.

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CHAPTER-I

INTRODUCTION

On a world wide basis hereditary forms of anemia are immensely common and affected persons are numbered in millions. Such hereditary disorders may be looked into on two levels: (1) biological level, and (2) familial or societal level. On biological level one should look into various types of manifestations of such diseases and their genetic consequences. On familial/societal level one has to look into the cost of maintaining such patients, psychological impact on the patients themselves and their parents, contributions of the patients to the society in terms of academics, workload, etc. The main purpose of such study should be to find out the types of welfare measures that could be taken for the patients and also to see how the social load could be reduced in the society.

Tyler et al. (1982, 1983) have found out how the families of the patients with Huntington's chorea (a genetic disorder) suffer from tremendous economic hardship to meet the expenses for treatment of those patients and also the patients themselves suffer from various types of disruption in family as well as in conjugal life. They have also

observed that there are other types of problems for such patients, e.g., employment opportunities, social adjustments, etc. Finally, they are of the opinion that the community as a whole is bound to suffer for such patients from social and economic points of view, if no suitable social measures are taken to rehabilitate those patients in proper and reasonable manner.

In the present study we shall consider only thalassaemia (β-thalassaemia). The thalassaemias are a group of inherited disorders of haemoglobin synthesis with varying severity, characterised by reduced rate of production of one or more of the globin chains of haemoglobin. This leads to imbalanced globin chain synthesis and to precipitation of the globin chains which are produced in excess. The major defects in red-cell maturation and survival, which characterise thalassaemia, are the direct result of the deleterious effects of the precipitated globin chains. It is now believed that it is one of the most common single gene disorders in the world. It produces a massive public health problem in many countries. It is true that it occurs in particularly high frequency in a broad belt, stretching from the Mediterranean through the Middle East, Indian subcontinent, Burma, and South-East Asia (Weatherall, 1983).

Ingram and Stretton (1959) have suggested that there are two main types of thalassaemia - (1) α-thalassaemia -

occurring due to reduced rate of α-chain synthesis and (2) β-thalassaemia - resulting from defective production of β chains. This suggestion of Ingram and Stretton (1959) has fully been confirmed by many subsequent studies on the rate of globin chain production.

If the gene is inherited from one of the parents the person will be in a heterozygous state with reference to the thalassaemia gene. In such a case of heterozygous state various terms like thalassaemia trait, thalassaemia minor, thalassaemia minima or Cooley's trait are used to express the underlying genetic heterozygosity. Generally the people in heterozygous state, i.e., with thalassaemia trait, are more or less in perfect health condition, though there can be mild to moderate anemia, slight elevation of HbA_2 (2.5 to 5 per cent), presence of small amount of HbF (if not all) and slight reduction in erythrocyte count. So in such cases, the clinical symptoms are usually nil or minimal (Chatterjea, 1965).

Sometimes the thalassaemia genes may be inherited from both parents, and the person will be genetically homozygote with reference to the thalassaemic gene. Such condition is known as thalassaemia major or Cooley's anemia in which the clinical manifestations are quite severe (Chatterjea, 1965).

Besides these two conditions, some individuals are found to have a thalassaemia gene, along with an abnormal

haemoglobin gene, which they inherit one from one parent and the other from the other parent. Such persons, possessing two dissimilar haemoglobinpatheic genes, are said to be in "double heterozygote" state. In such case of a double heterozygote state (like HbS-thalassaemia, HbE-thalassaemia, etc.), the gene for thalassaemia interacts with the gene of relevant abnormal haemoglobin. such conditions lead to an enhanced production of abnormal haemoglobin, and thereby producing varying degrees of severe clinical disorder. In any case, in both homozygote and double-heterozygote states the thalassaemia patients suffer from severe anemia and many other haemotological problems (Chatterjea, 1965; Weatherall, 1975).

Clinical Picture of α- and β-Thalassaemia

The main cause for α-thalassaemia is due to gene deletion when there is a possibility of losing from 1 to 4 α-genes, four abnormal conditions are found. The loss of a single α-gene does not cause any pathological condition. In a foetus this deficiency of α-chain leads to the production of an excess of τ-chain, which forms τ_4 tetramer or Hb Barts. Around 2 to 3 per cent of haemoglobin is of this form. The condition is known as α-thalassaemia-2. When two α-genes are missing, it leads to a condition known as α-thalassaemia-1; and at birth 5 per cent of haemoglobin is Hb Barts. The

situation is associated with minor blood abnormalities in later life. "When only two α-loci are present they may be on the same chromosome, i.e., the Homologe has lost both α-loci, *or they may be on different chromosomes - the so called* homozygous situation - in which a single gene has been deleted from each chromosome. This 'homozygous state' tends to characterise the situation in African and Mediterranean countries, and the heterozygous one in Asia (Harrison, 1988). When the syntheis of α-chains is defective, the haemoglobin produced is made up of four β-chains, known as HbH (β_4) or of four τ-chains, known as Hb Bart's (τ_4). Individuals with such types of haemoglobin suffer from severe anemia (Poirier et al., 1994). An unusual form of the condition occurs in some *children with mental retardation and an acquired form has* been reported in patients with preleukomia and other myeloproliferative disorders. But the patients with HbH disease have variable degree of anemia and splenomegaly. Haemoglobin values range between 7 and 10 g/dl and the red cells show marked hypochromia and variation in shape and size. A complete absence of α-loci leads to a condition, known as hydrous foetalis. It is fatal in pre-natal development. In such case no α-chains are produced at all but the production of τ-chain continues for longer than normal. *This, however, is insufficient for post natal existence* (Harrison, 1988).

β-thalassaemia is characterised by persistent synthesis of τ-chains beyond the neo-natal period, which results in a variable elevation of foetal haemoglobin (HbF). The β-thalassaemia can broadly be classified into β^{O}-thalassaemia, in which no globin chains are synthesized at all, and β^{+}-thalassaemia, in which there is a reduced rate of β-chain production. These conditions are very heterogeneous, both at molecular and phenotypic levels. The β^{+}-thalassaemia can be further sub-classified at a descriptive level into the severe Mediterranean form and the milder Negro form (Weatherall, 1983).

There is also another well defined sub-group of both β^{O} and β^{+}-thalassaemia, in which the HbA_2 level is normal in heterozygotes. Finally, there are some rarer forms of thalassaemia which have only been found in single families or populations (Weatherall, 1983).

Usually the homozygous state for β^{O}-thalassaemia or the severe forms of β^{+}-thalassaemia or the compound heterozygous states for β^{O} and severe β^{+}-thalassaemia are associated with transfusion-dependent anemia from early in life. However, some patients run a much milder course and the term thalassaemia intermedia is used to describe this condition (Weatherall, 1983).

Weatherall (1975) has very succinctly summarised the thalassaemia syndromes, which are as follows:

Table 1.1

The Thalassaemia Syndromes***

Type of Thalassaemia	Homozygous State	Heterozygous State
β-thalassaemia(with haemoglobin A production)	Severe anaemia; high level of haemoglobin F	Increased level of haemoglobin A_2
β-thalassaemia(with no haemoglobin A production)	Severe anaemic; haemoglobin consists of F and A_2 only	As above
β-thalassaemia(with no haemoglobin A production)	Moderate anaemia; haemoglobin consists of F and A_2 only	Increased level of haemoglobin A_2 and high levels of haemoglobin F (5-15% range)
β-δ-thalassaemia	Severe anaemia; haemoglobin consists of F only	Haemoglobin F in 5-20% range; normal levels of haemoglobin A_2
Haemoglobin Lepore thalassaemia	Severe anaemia; haemoglobin consists of Lepore and F	Normal levels of haemoglobin A_2; haemoglobins F and Lepore present
β-thalassaemia with β-chain	Clinical severity depends on level of haemoglobin A, and proportion of haemoglobin A is produced. Most important are S thalassaemia, C thalassaemia and E thalassaemia.	
α-thalassaemia 1‡‡	Death in utero; haemoglobin consists mainly of Bart's	Difficult to detect in adults; haemoglobin Bart's in 5-10% range in infancy
α-thalassaemia 2‡‡	Not yet recognised	Not detectable in adults; slight elevation of haemoglobin Bart's in infancy
Haemoglobin H disease	-	Probably heterozygous for for thalassaemia 1 and 2 or haemoglobin; Constant Spring
α-thalassaemia 1 and 2 with haemoglobin E	-	Severe anaemia; haemoglobins Bart's, F, E and A present
δ-Thalassaemia	-	Reduced haemoglobin A_2 levels
Thalassaemia-like states	Normal levels of haemoglobin A_2 and F, with clinical picture of thalassaemia	

‡ Probably a distinct form of β-thalassaemia with more haemoglobin F synthesis than in typical β-thalassaemia.

‡‡ May represent different genetic disorders in different populations.

‡‡‡ Adopted from Weatherall (1975).

Characteristics of β-thalassaemia

Basu (1994), while discussing about the characteristics of β-thalassaemia, has made the following observations:

1) In case of severe anemia frequent blood transfusions are needed and often it turns to be fatal in early childhood.

2) Red cells of varying size and distorted forms (poikilocytes) and target cells are present.

3) Foetal haemoglobin (HbF) level rises to 50 to 90 per cent.

4) The percentage of haemoglobin A_2 is variable. The HbA_2 level is elevated in the parents of the persons with thalassaemia major.

5) Erythrocyte count is generally reduced (1,000,000 to 3,000,000 cells for Cmm) and the serum iron concentration is found to be higher than normal.

6) The stained blood shows a marked hypochromia and many of these cells show only a thin rim of haemoglobin.

7) Physical growth is usually retarded.

8) Marked pallor, patchy skin pigmentation, chronic leg ulcers and abdominal distentions are often observed.

9) Bones of cranial vault are thickened and changes in facial bones give some characteristic appearance known as Mongoloid faces.

10) Bone pain and recurring bouts of fever occur from an early age.

11) Progressive splenomgaly is found early and affected individuals fail to gain weight normally and become anemic.

Diagnosis of β-thalassaemia

The diagnosis of β-thalassaemia major is usually possible from the clinical and haematological data as well as from family studies. For all suspected individuals electrophoresis tests are performed to determine the level of haemoglobin F and A_2 in both patients and parents. The high A_2 β-thalassaemia homozygosity can be diagnosed, if both parents are found to have increased haemoglobin A_2 level and the patient also shows an increase in haemoglobin F, usually in excess of 20 per cent of the total haemoglobin. electrophoretic studies on the parents will also indicate whether the patient is truly homozygous for high A_2 β-thalassaemia or heterozygous for high A_2 β-thalassaemia and one of the β-thalassaemic variants (Basu, 1994). The recent evidences indicate that β-thalassaemia heterozygotes may be distinguished from normal infants by age of 3 months, and with certainty by age of 6 months. The HbA_2 level becomes elevated above the normal adult level by about 3 months and is clearly distinguishable from normal by 6 months. An

elevated level of HbF is also a useful indicator for the presence of a β-thalassaemia gene since it seems to be significantly higher than in normal infants, at least upto the age of 1 year (Weatherall, 1983). β-thalassaemia major can be distinguished from severe iron-deficiency anemia by more severe changes in erythrocytes in former disease by presence of large amounts of HbF, raised serum ion level and increased iron content of bone-marrow in thalassaemia major (Basu, 1994).

Growth and Development of β-thalassaemic Patients

Garn (1952) is of the opinion that height and weight are the two body dimensions which are generally taken as good indicators of physical growth. As a matter of fact, human physical growth is influenced by many factors, including genetic constitution (Mills, 1937; Meredith, 1941; Garn, 1952; Greulich, 1957).

The clinical pictures of the thalassaemic children have first been described by Cooley et al. (1927). The patients usually appear to be small for their age. But Smith et al. (1955) have pointed out that physical growth of the thalassaemic persons seems to be normal upto 8 or 10 years of age and then retardation in rate of physical growth starts and final stature becomes shorter than average.

Sarkar et al. (1984) have made a study on stature and weight in relation to some haemoglobin genotypes in a Bengali-speaking population of Calcutta. They have observed that haemoglobin genotypes do not have any influence on stature and weight in adult population. They have further observed that the heterozygotes (comprising HbβE or βT gene) enjoy no advantage in physique over the haemozygotes, which may be due to relaxation of selective pressure. In a significant study on 50 individuals (both sexes), all having diagnosed as suffering from β-thalassaemia major, aged between 2 years 10 months and 28 years 5 months, Johnston and Krogman (1964) have made the following observations:

"1. Children with Cooley's anemia have their growth patterns altered in two ways; a retardation in their normal growth expectation and a retardation in their rates of growth.

2. The retardation in growth rate would indicate a much later expectation for any growth-time-linked event. The expression of the event itself; when it occurs, would be a function of the damage due to the prolonged anemia.

3. With the extension of the growth period, and with the success in prolonging life expectancy, it could be concluded that a curve of growth similar to the normal, but not quite reaching normal values, would emerge for children so affected.

4. Cephalofacial manifestations of orthodontic significance seems to be concentrated mainly in maxillary alveolar bone and in the palate as well."

The pathogenesis of growth retardation in thalassaemic patients is complex and incompletely understood. However, the National Thalassaemic Bulletin (1995) indicates the following factors, which may be responsible for affecting growth:

1) Chronic anemia
2) Endocrine disease
3) Chronic liver disease
4) Iron overload
5) Zinc deficiency
6) Desferal toxicity

Distribution of Thalassaemia

During recent years there have been some reviews depicting the geographical distribution of thalassaemia. No other haemoglobinopathic genetic disorder is known to be as widely distributed as thalassaemia till today.

There are mainly two areas, where prevalence of thalassaemia disease is very high, though it is found in all parts of the world. These two focal areas are as follows:

1) Mediterranean countries including Italy, Sicily, Sardina, Greece, Cyprus, Turkey and parts of North Africa.

2) South-eastern countries including India, Burma, Thailand and Indonesia (Chatterjea, 1965).

Regarding the distribution of this genetic disorder Chatterjea (1965) has mentioned that β-thalassaemia has been reported in China, the Phillippines, Australia, New Guinea and various parts of Africa and the West Indies. He has further mentioned that some isolated instances of β-thalassaemia have been detected from almost every country and in many racial groups.

In India instances of β-thalassaemia have been reported since 1938 from different parts of the country. But β-thalassaemia cases have been studied mainly from the pathological point of view. Basu (1994) has very recently tried to compile all data on β-thalassaemia, so far published on Indian populations. The first instance of β-thalassaemia in India was recorded in a two years old Bengali boy (Mukherjee, 1938). In Bengal the frequency of β-thalassaemia gene has been estimated to be present to the extent of 3.7 per cent among the Bengali Hindus (Chatterjea et al., 1957). Some cases of homozygous β-thalassaemia have also been reported in Bengal (Chatterjea et al., 1957) as well as in Gujarat (Sanghvi et al., 1958; Sukumaran et al., 1959, 1960). Choubisa (1985) has reported that the frequency of β-thalassaemia trait varies from 1.05 per cent in the Mochi to 3.12 per cent in the Ganchha among the scheduled caste

populations of Rajasthan. Gupta and Tiwary (1993) have reported that in Jabalpur (Madhya Pradesh) β-thalassaemia is common in all categories of population, i.e., scheduled castes, backward classes and forward communities and also present in lower frequency among the scheduled tribes. Sharma et al. (1971) have found the presence of β-thalassaemia among the Lohana, Gaud Saraswat and Chitrapur Saraswat in Gujarat. Sukumaran and Haveli (1978), cited by Basu (1994), have found β-thalassaemia among the Warli, Dhodia and Kokana in varying frequencies. Sukumaran (1975), while reviewing abnormal haemoglobins in India, has discussed about the prevalence of β-thalassaemia in various populations of Bombay. In north-eastern states of India a very few studies have so far been carried out. Das and Deka (1980) have compiled the existing data and have shown that the frequency of β-thalassaemia gene is about 2.8 per cent among the Assamese of lower Assam and 0.8 per cent among the Ahom and 1.4 per cent among the Khasis of Meghalaya. The β-thalassaemia trait has been reported in different parts of U.P., Maharashtra, Karnataka, Andhra Pradesh, Kerala, Orissa and Goa (Basu, 1994). However, our purpose is not certainly to show the exhaustive distribution of thalassaemia in India, but to focus the point that β-thalassaemia is found all over the country, though its concentration is more in eastern sector, particularly in West Bengal.

Treatment of β-thalassaemia

The treatment for homozygous β-thalassaemia consists of monthly transfusions and iron chelation therapy and bone marrow transplantation.

Iron overload is a major clinical problem in this group of disorders, particularly in β-thalassaemia. In severe homozygous form of β-thalassaemia. (β-thalassaemia major) the affected children require regular blood transfusions.

Since iron losses in man are not significantly increased in response to iron overload, a large excess of iron storage steadily accumulates with progressive damage to the liver, endocrine glands, etc. Death from cardiac iron loading usually occurs at the end of the second decade of life (Pippard, 1983).

An excessive dietary iron absorption contributes to iron overload in the patients with β-thalassaemia major particularly if their blood transfusion regiments do not maintain haemoglobin level sufficient to suppress their own erythopoisis. Increased absorption from the gastro-intestinal tract provides the main route for iron loading in patients with β-thalassaemia intermedia. These patients have clinical features dominated by chronic anemia and extreme erythroid hyperplasia, but inspite of their not requiring regular blood transfusion they may eventually develop iron overload

associated with liver cirrhosis and multiple endocrine deficiencies. However, there are great individual variations in the rate of iron loading, and at present there is no simple way to predict which patient with β-thalassaemia intermedia will develop potentially fatal iron overload by the middle of life. It is clear that iron loading occurs at predictable rate in the patients with transfusion-dependent thalassaemia. So a careful individual assessment of the degree and rate of iron loading is therefore essential to remove iron overload (Pippard, 1983).

Treatment of Iron Overload in Thalassaemia

Iron chelation therapy, along with regular blood transfusion, is essential for normal growth and development of the β-thalassaemia patients. So iron chelation therapy should be introduced as soon as possible to prevent organ damage due to iron overload. Iron chelation is being achieved by desferrioxaimine (DF, Desferal). DF needs to be injected subcutaneously over 8 hours daily with the help of a portable electronic pump which costs around Rs.20,000 to Rs.25,000, besides Rs.4500 for desferal (National Thalassaemic Bulletin, 1995). The drug was not widely used until the demonstration that prolonged infusions produced much greater urinary iron excretion than equal dose given by intramuscular bolus injection. With this form of therapy it is now possible to

obtain negative iron balance even in very young transfusion-dependent thalassaemic children (Pippard, 1983).

However, this form of therapy is available to less than 10 per cent of the patients, primarily due to its high cost. In India only less than 3 per cent of the thalassaemia patients can receive some DF. Several side effects have been observed on regular DF therapy. In view of its high cost, decreasing compliance and cumulative toxicity, efforts were made over the last 30 years to develop an effective and safe oral iron chelator (National Thalassaemia Bulletin, 1995).

Recently a new capsule Deferiprone/kelfer has been evaluated in 17 countries at 32 centres. In India after an extensive trial on about 200 affected children in Bombay, Calcutta, Delhi, and Chandigarh got the best response. Iron excretion by Deferiprone depends on the dose, frequency of administration and ferritin level. Deferiprone has been used in 2 to 4 divided dose of 50 to 100 mg/day. In this dose it excretes 10 to 20 mg of iron in iron overload individuals and only 1 to 2 mg of iron in normal persons. Overall deferiprone has been found to be effective in excreting iron and in this dose it is able to deplete iron store. It mobilises iron from saturated transferring liver and other compartments containing excess of iron. The patients with HbF/β-thalassaemia achieve negative iron balance faster on the same dosage schedule. Long term administration helps bringing the

serum feritin level within the normal range. India is the only country in the world, where multicentric study at four centres, on over 225 cases, have been conducted to determine the effectiveness and safety of deferiprone in the multitransfused thalassaemia children (National Thalassaemia Bulletin, 1995).

Bone Marrow Transplantation

Bone marrow transplantation offers complete cure as defective erythroid stem cells are replaced by normal erythroid precursor cells. However, transplantation can be administered in those cases where HLA compatible donors can be found. The transplantation has to be performed early before the child becomes overly iron immunized by transfusions. But even under ideal circumstances bone marrow transplantation is currently associated with about 25 per cent mortality (Kan, 1986). The cost of bone marrow transplantation varies from Rs.3.5 lakhs to Rs.6.0 lakhs, depending on complications of post transplantation. It is reasonable to expect the following results from bone marrow transplantation in thalassaemia:

i) 60 per cent complete cure with good quality of life.

ii) 20 per cent relapse of thalassaemia.

iii) 20 per cent death due to transplant related complications (National Thalassaemia Bulletin, 1993).

But repeated packed cell transfusions continue to remain the main pillar of treatment. Haemoglobin level has to be maintained near normal level and all efforts are to be made to ensure that haemoglobin level does not fall below 10 gm/dl. Washed red cells minimise the blood transfusion reactions. Leucocyte filters should be used, if thalassaemic children develop blood transfusion reaction repeatedly (National Thalassaemia Bulletin, 1993).

Polymorphism

It may be pointed out that of all haemoglobinopathies thalassaemia accounts for the highest infant mortality rate in this country. Chatterjea (1968), while discussing about haemoglobinopathy in Bengal, has pointed out that β-thalassaemia, either in homozygous form or in combination with HbE-appears to be the commonest form of haemoglobinopathy and in fact the incidence of HBE β-thalassaemia has been much higher than that of homozygous β-thalassaemia since the latter is more severe than the former, and many homozygotes die too early.

Now the question arises how gene frequency is being maintained in a population in spite of continued loss of genes through early deaths of homozygotes. This cannot be

explained on the basis of any highly exaggerated mutation rate. Such loss of genes are supposed to be counter-balanced by some biological advantages enjoyed by the heterozygotes against baneful influences of malarial parasites (and perhaps other infections, iron-deficiency and some other environmental stresses). This illustrates the concept of balanced polymorphism (Chatterjea, 1968). In one study (Bhattacharya et al., 1982) it is postulated by examining overall reproductive performance (taking fertility and infant mortality together) that the heterozygotes will eventually suffer from a selective disadvantage in a place like Calcutta, where malaria was claimed to have been eradicated about three decades ago. But with changing situation at present and with reemergence of malignant malaria (often reported in newspapers) it is urgently felt that we shall have to review and reassess the whole situation (Ghosh, 1996).

As it is well known that the persons with thalassaemia disease, i.e., in homozygous states, generally do not survive upto the reproductive age. But now with advancement of medical technology, many of the thalassaemia patients, particularly in urban areas, survive beyond 30/35 years of age (Ghosh, 1996). So with the help of modern medical treatments the expectancy of life of such patients has become significantly greater than what it was even in the middle of

the present century. Consequently, such improved chance of survival of the patients with thalassaemia disease has created a series of social and biological problems, which are critical and relevant for future biological and social evolutions. However, such problems do not occur in the populations of simple cultures, living in remote rural areas. The reason is that they hardly get any opportunity to take the advantage of improved medical facilities since they just cannot afford such huge expenses. Consequently, such deleterious genes are steadily removed from populations through the process of natural selection (Ghosh, 1996).

But the situation in urban area is quite different from that in rural areas. All modern medical facilities are available in urban areas, and the people try their best to avail themselves of such opportunities even if these are very expensive. So in modern societies the individuals with such genetic disorder not only survive and enter into reproductive age-group but also have started contributing such deleterious genes to next generation. Consequently, genetic load in the population is steadily increasing and concomitantly social load increases too in such societies. In this context, Roberts (1975) says: "In modern human society deleterious genes assume an importance beyond that which they confer on the life of the individual himself. They present a far wider range of problems than survival and reproduction imply,

problems not only of treatment often expensive and long continued but also of day to day care and maintenance".

In a study in Calcutta (Das et al., 1983), it is observed that the reproductive fitness of the homozygous β-thalassaemia persons is around 0.03, which means that it has increased from 0 per cent to 3 per cent. It means that the rate of segregation of thalassaemia gene has increased to a great extent in view of the fact that previously the segregation of such genes used to take place only through heterozygotes and now homozygotes as well as heterozygotes are passing such thalassaemia genes to the next generation. Consequently, the frequency of thalassaemia gene has steadily been increasing in every generation. It is not only coming through some fresh mutations but also accumulating through higher rate of segregation. Chatterjea et al. in 1957 reported that the frequency of thalassaemia gene among the Bengalees was around 3 per cent, but is now reported by S. Majumder that about 10 per cent of the Bengalees carry thalassaemia gene (*The Statesman*, Calcutta, dated 14.2.94). It means that the frequency of thalassaemia gene has steadily been increasing over last 40 years. In some reported item published in *The Sunday Observer* (New Delhi, Bombay, dated February 6, 1994), that about 4 per cent of Indian population carries thalassaemia trait, 5000 thalassaemia children are born every year in India and 30 million carriers will

continue to pass on this dreadful disorder to their children. According to Dr. Dilip Bhattacharya, the founder of Thalassaemia Society of India in Calcutta there has been an increase of 50 per cent in the number of thalassaemia cases in the last 10 years and every year there is an addition of 250 patients to the thalassaemia society of India (*The Statesman*, Calcutta, dated 14.2.94).

Such limited information proves to what extent the genetic quality of the population, especially in the eastern part of the country, where this genetic disease is very much prevalent in higher frequency, is deteriorating, and thereby to what extent biological load is increasing in Indian population and it is well known that if genetic load (i.e., biological) increases, social load is concomitantly bound to increase in the population. It in turn certainly tells upon both physical and social health of the population.

So in order to understand the physical and social health of a population a combination of demographic, biological and socio-cultural approach is needed. However, so far no serious in depth study has been done, excepting a few isolated surveys from biological point of view. But the use of only one of these approaches is bound to result in partial understanding of the situation. The disease may be a biological one but it does have some social implications on the population. Similarly some social factors may be directly

responsible for passing of a particular disease from one generation to another. With this view in mind we intend to take up a study on bio-social aspects of β-thalassaemia from in and around Calcutta with the following objectives:

1) To determine the physiological symptoms of β-thalassaemic individuals;

2) To find out the rate of physical growth of the thalassaemic patients (upto 20 years);

3) To assess socio-economic and psychological pressures on the parents and the patients themselves;

4) To understand ability of the patients to learn (e.g. education) to work in terms of physical and/or intellectual output, and to attend to social activities;

5) To find out mode of treatment and average cost;

6) Finally, to suggest what type of comprehensive social measures should be taken for such genetically handicapped people and what type of economic reliefs may be given to the parents of such patients.

With this small introduction we shall present our findings in subsequent chapters.

CHAPTER-II

MATERIALS AND METHODS

In this chapter, we shall deal with the materials collected for the present study and the method that have been applied.

Data for the present study on "A Bio-Social Study on β-Thalassaemia" were collected in and around Calcutta and the fieldwork was done during the period between May, 1994, and September, 1995. Data were collected from 380 thalassaemic patients from the following hospitals:

1. The Thalassaemia Society of India
2. Kothari Medical College
3. Bhoruka Research Centre
4. Balananda Brahma Chari Hospital
5. Assembly of God Church.

And for comparison data on 364 normal individuals were collected from the following schools:

1. Jodhpur Boys' School
2. Sishuniketan
3. Udbastu Girls' School.

Both the samples were divided on the basis of sex and age groups, which have been shown in the following table (Table-2.1):

Table-2.1

Classification of Sample by Age and Sex (upto 20 years)

Age group (Years)	Thalassaemic		Controlled	
	M	F	M	F
Below 1 year	5	2	-	-
1 year	-	1	-	-
2 years	9	5	-	-
3 years	13	8	7	10
4 years	23	11	20	20
5 years	22	15	45	15
6 years	27	9	11	7
7 years	22	11	17	6
8 years	21	12	32	11
9 years	11	11	22	9
10 years	20	10	20	13
11 years	16	11	16	8
12 years	17	9	7	2
13 years	7	5	8	4
14 years	8	3	13	4
15 years	6	3	9	12
16 years	4	2	2	3
17 years	2	-	2	2
18 years	7	3	3	2
19 years	-	-	2	-
20 years	-	1	-	-

Data were collected on haematological, demographic and social parameters and also anthropometric measurements.

Demography:

The nature of demographic data, collected for the present study, was based on those parameters, suggested by WHO (1964, 1968). These are as follows:

Individual records:

Name, data of birth, age, religion, caste, occupation, educational status, age at marriage, etc. were collected.

Fertility records:

These include pregnancy history of each mother, present age of mother, total number of live births, birth order, age and sex of each offspring.

Mortality records:

These include total number of conceptions, number of dead children, sex, age, date of birth, age at death, cause of death, number of reproductive wastages (abortion, still birth), etc.

Socio-Cultural Parameters:

These include monthly income, monthly expenditure, attendance of children in school, their performances, participation in extra-curricular activities, expenditure on

diseased child, average cost for treatment, etc. were recorded.

All the above mentioned data were collected from the parents. From the patients, the following information were collected:

Information such as name, age, sex, religion, education, father's name, father's occupation, father's income were collected. In order to understand the health status of the patients, information such as health problem at present, when detected, the symptoms, any difference after treatment, total number of transfusions taken till date, frequency of hospital visits, feelings after prolonged treatment, etc. were collected. Data on age at menarche, were collected from the adult girl patients.

Anthropometry:

The anthropometric measurements were taken on 133 thalassaemic females and 240 thalassaemic males. An equal number of measurements were also collected from the normal individuals who were not known to be suffering from thalassaemia or any other disease at the time of fieldwork. In the Controlled sample there were 128 females and 236 males, aged 3 years to 20 years. As per recommendation by the International Biological Programme (Weiner and Lourie, 1981), the following measurements were taken: height, weight, chest girth (inhale), chest girth (exhale), humerus diameter,

bicondylar femur diameter, biceps girth, calf girth, and for skinfold measurements Biceps, triceps, subscapular, supra-illiac, and calf were taken.

Anthropometric Measurements:

To take all these measurements we essentially followed the methods suggested by Sen (1994) and Das and Deka (1992-93) and adopted all the landmarks from Sen as well as from Das and Deka.

Height-Vertex:

It measures the vertical distance from floor to vertex.

Vertex:

It is the highest point on the head, when the head is on eye-ear plane. This measurement was taken with the help of an anthropometer keeping the subject on a erect posture. The measurement was taken standing on his/her back. The anthropometer was placed at the back and between the heels of the subject, as close to the subject as possible.

Weight:

The body weight was taken on a spring weighing machine, asking the subject to stand on it with an erect posture and light apparel. The weighing machine was checked from time to time with a known weight.

Chest Girth (Inhale and Exhale):

The measurement was taken with a steel tape from the front of the subject on the level of the fourth pair of ribs (in the case of adult females, it was taken from the back). Reading was recorded separately at the time of inhalation and exhalation.

Bicondylar Humerus Diameter:

The subject's elbow was bent to a right angle and the width across the innermost parts of the lower end of the humerus was taken. The measurement is usually oblique since the medial epicandyle of the humerus is lower than the lateral pressure when exerted to compress the tissues. Rod compass was used to take this measurement.

Bicondylar Femur Diameter:

The subject was asked to sit on a table with knees bent to a right angle and the width across the outermost parts of the lower end of the femur was measured. Pressure was exerted to compress the tissues.

Bicep Girth:

The subject's arms hang relaxed just away from his/her side, and the circumference was taken horizontally at the marked level. This measurement was taken with the help of a steel tape.

Calf Girth

The subject sits on a table with the leg hanging freely. Maximum circumstance, obtained by moving the tape vertically up and down the leg, is taken horizontally.

Skinfold Thickness Measurement (left):

Ponderx skinfold caliper graduated in cm and mm was used for the measurement of skinfold thickness at biceps, triceps, subscapular, supra-illiac and calf.

Triceps:

The fold of the subject was picked up at the dorsal side of the left upper arm cm above the level of the upper arm girth taking care that the fold should be in a line along the length of the arm.

Biceps:

The fold was taken in front of the upper arm on the same level as in the case of biceps region, taking care that the fold should be parallel to the length of the arm.

Subscapular:

Fold was picked up just below the inferior angle of the left scapula. It had to be vertical but slightly inclined downward and laterally parallel to the natural cleavage line of the skin, i.e., at about 30° angle to the vertical line.

Supra-Illiac:

The skinfold is picked up approximately 1 cm above and 2 cm medial to the anterior-posterior illiac spine.

Calf:

This measurement was taken on the level of the maximum circumference of calf on medial border, parallel to the length of leg.

These anthropometric measurements were taken on both the thalassaemic patients and the normal individuals with a view to finding out the growth pattern of the thalassaemic patients.

Haemtological Data

Haemtological data were collected from the hospital records as it would be much more authentic. The following information about the patients were collected from the hospital records: Date of birth, date of diagnosis, previous transfusion, total haemoglobin before and after transfusion.

It may be noted that the hospitals in Calcutta do not keep all relevant records on physiological parameters about the thalassaemic patients. Consequently, we had to be satisfied with those data mentioned above.

Statistical Methods

The statistical methods (Cambers, 1958) which have been adopted in the present study, are as follows:

Mean:

The best known and most useful form of average is the arithmetic mean, usually referred to as the 'mean' or the 'average'. It is defined as a value which can be obtained by dividing the total values of various items in a series by the total number of items. It is worked out as under:

$$\text{Mean } (\bar{x}) = \frac{\Sigma x_i}{N} = \frac{X_1 + X_2 + \ldots + X_n}{N}$$

where

Σ = Symbol for Summation;

X_1 = Value of the ith Item, X_i; = 1 = 1,2 ...., n

N = Total number of Item.

Standard Deviation

By far the best and the most useful measurement of scatter is the standard deviation. Standard deviation is defined as the square root of the mean of the square of the deviations of observations from their arithmetic mean. It is computed as follows:

$$SD = \sqrt{\frac{\Sigma (x-\bar{x})^2}{N}}$$

Here $(x-\bar{x})$ represents the deviation of an individual reading from the mean

$\Sigma(x-\bar{x})^2$ the sum of the square of all such deviation. The divisor is taken as N.

Standard Error of the Mean (S.E.)

When we estimate the standard deviation of the sampling distribution from the number and standard deviation of a single sample, the estimated value is called standard error of the mean and it is calculated as follows:

$$S.E. = \frac{SD}{\sqrt{N}}$$

where

SD is the standard deviation of the sample

N is the number of observations.

The Chi-Square (X^2) Test

The chi-square is used in the investigation of a number of different problems. It can be used whenever we wish to determine weather or not the observed frequencies in a particular category differ significantly from those which would be expected to fall in that category under a certain set of theoretical assumption, or hypothesis. In the present study, the chi-square is used for testing the difference

among different sample proportions (Blalock, 1972; Parker, 1973). These form of statistics is as follows:

$$\chi^2 = \frac{\Sigma(O-E)^2}{E} = \frac{(O_1-E_1)^2}{E_1} = \frac{(O_2-E_2)^2}{E_2} + \ldots + \frac{(O_n-E_n)^2}{E_n}$$

where O is the observed frequency and E the corresponding expected one.

The value obtained is then compared with that given in the table of chi-square with (N-1) degree of freedom (DP). In case of 2x2 table, the number of (DP) is (R-1) (C-1), where R and C are the number of rows and column respectively. The expected frequency is calculated as (Row total) (Column total)/(Grand total) and the formula for determining χ^2 value for 2x2 table is as follows:

$$\chi^2 = \frac{(ad - bc)^2 N}{(a+b)\ (b+d)(c+d)(a+b)}$$

where N = a + b + c + d

The other measurements include the following:

1) Weight for Age (%)

2) Height for Age (%)

3) Weight for Height (%).

These percentages were calculated following ICMR (1972) standard.

Difference between Two Means

To find out the significant difference between the two means, we have applied 't'-test as suggested by Chambers (1958).

Classification of Data

In the present study, we have followed the following classifications:

Educational Level:

The individuals, who are unable to read or write, have been grouped as illiterate. Those, who could read and write and had education upto Class-IV, have been placed under the category of primary level. From Class-V to Class-X have been placed under secondary category, XI and XII in the high secondary. However, this classification is arbitrary.

Economic Classification:

In the present study, we have broadly classified the population into three economic groups with the help of the following interval, estimation based on standard deviation which is as follows:

Above ($\bar{a}$ + 4.SD) = High Income Group.

($\bar{a}$ + 4.SD) to ($\bar{a}$ - 4.SD) = Middle Income Group.

Below ($\bar{a}$ - 4.SD) = Low Income Group.

In the present study, the average per capita monthly income of 337 households has been calculated following the method given by Khongsdier (1997):

$$(\bar{a} - 4.SD) = Rs.913.59 - \frac{4 \times 1218.84}{337} = Rs.\ 648.01$$

$$(\bar{a} + 4.SD) = Rs.913.59 + \frac{4 \times 1218.84}{337} = Rs.1179.17$$

Participation in Games:

Participation in games of the thalassaemic patients has been classified into 3 categories, namely, 'Indoor', 'Outdoor' and 'Indoor and Outdoor'. The Indoor games here refer to Carom, Chess, Ludo, Chinese Checker, etc. and Outdoor games refer to football, cricket, volleyball, basket ball, etc.

The physical activities of the thalassaemic patients have been classified into: 'Active', 'average' and 'inactive'.

i) Active - includes those, who regularly take part in games.

ii) Average - includes those, who at times take part in games.

iii) Inactive - includes those, who do not take part in any game at all.

Regarding the classification of performance in school we have divided them into satisfactory and not satisfactory. By satisfactory we mean that those children who come under certain standard (i.e. regular attendance, good in study, etc.) and in the not satisfactory group, we have included those children, who do not perform according to the expected standards of the school.

In the subsequent chapters, we shall present our findings and we shall compare the present findings with relevant data available till now.

CHAPTER-III

BIOLOGICAL ASPECTS

In the present study data on 380 β-thalassaemic patients have been collected. Out of these 380 patients, 14 patients (4 females and 10 males) came to the hospitals without having any specific symptom but they came for routine check up and β-thalassaemia was detected in them.

Section-I - Observations based on Primary Data

Table 3.1

Symptoms of the Patients before Detection of Thalassaemia*

	Symptoms	Female		Male	
		No.	%	No.	%
1)	Fever	18	13.74	42	17.87
2)	Fever and loss of appetite	21	16.03	30	12.77
3)	Anemic and loss of appetite	6	4.58	10	4.26
4)	Fever with vomitting tendency and loss of appetite	25	19.08	35	14.89
5)	Fever and swelling of body	5	3.82	17	7.23
6)	Fever with cold and cough and and loss of appetite	16	12.21	47	20.00
7)	Fever with vomitting tendency and enlarged spleen	8	6.11	12	5.11
8)	Fever with vomitting tendency, loss of appetite and anemic	25	19.08	34	14.47
9)	Jaundice, physically under-developed, vomitting tendency, and loss of appetite	7	5.34	8	3.40
	Total	131	99.99	235	100.00

* 10 male and 4 female patients were detected for thalassaemia during routine check-up.

Table 3.1 shows whatever symptoms the patients had before the detection of β-thalassaemia. All symptoms have been classified into 9 categories. It is seen that in case of male patients, 42 (i.e., 17.87 per cent) had fever only, whereas 30 (i.e., 12.77 per cent) had both fever and loss of appetite and 35 (i.e., 14.89 per cent) had fever with vomitting tendency and loss of appetite. About 47 (i.e., 20.00 per cent) individuals had fever with cold and cough and loss of appetite. 34 (i.e., 14.47 per cent) individuals had fever, with vomitting tendency, loss of appetite, besides anemic condition. 12 (i.e., 5.11 per cent) individuals had enlarged spleen with fever and vomitting tendency. There are also 8 (i.e., 3.40 per cent) individuals, who were suffering from Jaundice, vomitting tendency, loss of appetite, and physical under-development. It is seen that there were 10 (i.e., 4.26 per cent) individuals had anemic condition and loss of appetite, whereas 17 (i.e., 7.23 per cent) individuals had fever and swelling of body.

In the present sample there are altogether 131 female patients with β-thalassaemia, out of which 25 (i.e., 19.80 per cent) had fever with vomitting tendency and loss of appetite and another 25 (i.e., 19.08 per cent) individuals had all these symptoms in addition to anemia. 8 (i.e., 6.11 per cent) patients were suffering from fever, with vomitting tendency and enlarged spleen, and 21 patients (i.e., 16.03

per cent) had only fever and loss of appetite. There were 18 patients (i.e., 13.74 per cent) had only fever, while 6 patients (i.e., 4.58 per cent) had anemic condition, along with loss of appetite. There were also 5 patients (i.e., 3.82 per cent) had fever, along with swelling of body. There are 7 (i.e., 5.34 per cent) individuals who were suffering from jaundice with vomitting tendency, loss of appetite and physical under-development. There were 16 (i.e., 12.21 per cent) individuals, who reported that before the detection of thalassaemia they were suffering from fever, with vomitting tendency and enlarged spleen. However, of all these symptoms, reported by both male and female patients the following are comparatively more common:

1) fever;

2) fever with vomitting tendency and loss of appetite;

3) fever with vomitting tendency, loss of appetite and anemic condition;

4) fever with cold and cough and loss of appetite.

Table 3.2

Age of Detection of Thalassaemia

Age in years	Male		Female		Total	
	No	%	No	%	No	%
< 1	110	44.90	64	47.41	174	45.79
1 - 5	114	46.53	57	42.22	171	45.00
6 - 10	17	6.94	13	9.63	30	7.90
11 - 15	4	1.63	0	0.00	4	1.05
16 - '20	0	0.00	1	0.74	1	0.26
Above 20	0	0.00	0	0.00	0	0.00
Total	245	100.00	135	100.00	380	100.00

Table 3.2 shows the age of detection of thalassaemia in case of both male and female patients. It is found that thalassaemia was detected in case of 44.90 per cent of males and 47.41 per cent of females before 1 year of age. Combining both male and female together, it is seen that β-thalassaemia was detected for 45.79 per cent of all patients before completion of 1 year of age. In case of individuals, aged between 1 and 5 years, β-thalassaemia was detected for 46.53 per cent of males and 42.22 per cent of females. Combining both male and female patients together, it is seen that 45 per cent of all patients β-thalassaemia was detected within 5 years of age. Only in case of 7.90 per cent of all cases (male-6.94 per cent and female-9.63 per cent) β-thalassaemia

was detected between 6 and 10 years of age. In case of 4 (i.e., 1.63 per cent) male individuals β-thalassaemia was detected between 11 and 15 years of age. Only in 1 (i.e., 0.74 per cent) female β-thalassaemia was detected as late as 20 years of age. So it may be said that β-thalassaemia is generally detected in most of the cases within 5 years of age and very rarely beyond that age. When the detection of β-thalassaemia is as late as 20 years it means that for one reason or the other the patient never consulted any specialist for treatment. However, such late detection is a very rare phenomenon.

Table 3.3 shows the frequency of hospital visits by the β-thalassaemic patients. In the present sample it is seen that nearly 30.79 per cent of all patients visit hospitals at least once in a month, whereas 40.26 per cent of them pay visit twice in a month. It is also found that only 2.11 per cent of all β-thalassaemic patients visit hospital once in 2 months and 0.26 per cent of them visit hospital once in a year. This table further shows that the β-thalassaemic patients, both males (i.e., 41.22 per cent) and females (i.e., 39.26 per cent), aged between 6 and 10 years, visit hospital much more regularly. It is further found that the β-thalassaemic patients, both female (i.e., 29.63 per cent) and male (i.e., 27.35 per cent), are regular visitors to hospitals. In the very early age, i.e., less than 1 year of

Table 3.3

Frequency of Hospital Visits by Age and Sex

Frequency of Visits	Age Group in Years													
	< 1		1-5		6-10		11-15		16-20		20+		Total	
	M	F	M	F	M	F	M	F	M	F	M	F	No	%
Once in a month	4	1	24	17	27	13	11	10	6	2	1	1	117	30.79
Twice in a month	1	-	22	14	43	27	23	12	5	2	2	2	153	40.26
2 to 3 times in a month	-	1	-	-	2	1	2	-	-	-	1	-	7	1.84
Thrice in two months	-	-	17	7	20	10	13	6	-	1	-	-	74	19.47
Thrice in a month and more	-	-	4	-	8	1	4	2	-	-	1	-	20	5.26
Rare visits:														
a) Once in a year	-	-	-	-	-	1	-	-	-	-	-	-	1	0.26
b) Once in two months	-	-	-	2	1	-	1	1	2	1	-	-	8	2.11
Total	5	2	67	40	101	53	54	31	13	6	5	3	380	99.99
%	(2.04)	(1.48)	(17.35)	(29.63)	(41.22)	(39.26)	(22.04)	(22.96)	(5.30)	(4.44)	(2.04)	(2.22)		

age, only 2.04 per cent of males and 1.48 per cent of females visit hospitals. However, it is seen that as age advances, the β-thalassaemic patients' regular visit to hospitals decrease. It may be due to the fact that many of the β-thalassaemic patients die early in age.

Table 3.4

Total Number of Transfusions Taken by the Patient till the Date of the Present Investigation

Age in Years	Male					Female					Total		
	No	Range	Mean	S.D.	±S.E	No	Range	Mean	S.D.	±S.E	Mean	S.D.	±S.E
< 1	5	2-60	4.00	1.67	0.75	2	8-12	10.00	2.00	1.41	5.71	3.24	1.22
1 - 5	67	1-110	43.31	25.73	3.14	40	5-90	41.55	23.84	3.77	42.65	25.05	2.42
6 - 10	101	4-342	111.59	72.17	7.18	53	5-200	97.62	52.33	7.19	106.79	66.34	5.35
11 - 15	54	5-400	142.33	92.10	12.53	31	35-300	147.94	73.82	13.26	144.38	85.93	9.32
16 - 20	13	36-300	172.00	82.42	22.86	6	72-400	237.00	122.93	50.19	192.53	101.65	23.32
Above 20	5	106-350	225.20	90.18	40.33	3	216-600	348.67	177.81	102.66	271.50	143.22	50.64

Table 3.4 shows the number of transfusions, taken by the patients till the date of the present investigation. It is found that for the β-thalassaemic patients, aged below 1 year of age, the number of transfusions taken is not as much as when the patients are older. For the β-thalassaemic male patients, aged below 1 year, the mean number of transfusions taken is found to be 4.00±0.75 and that for female patients

10.00±1.41. The male β-thalassaemic patients, aged between 1 and 5 years, the mean number of transfusions taken is 43.31±3.14. The same for the female patients is 41.55±3.74. When the β-thalassaemic patients are aged between 6 and 10 years, the mean number of transfusions taken by males and females is found to be 111.59±7.18 and 97.62±7.19, respectively. The mean number of transfusions taken by male and female β-thalassaemic patients, aged 11-15 years, is 142.33±12.53 and 147.94±13.26 respectively. In case of the β-thalassaemic patients, aged 16-20 years the mean number of transfusions taken is 172±22.86 by males and 237±50.19 by females. In case of the β-thalassaemic patients, aged above 20 years, the mean number of transfusions taken by males is 225.20±40.33 and the same is 348.67±102.66 for females. Combining both sexes together the mean number of transfusion taken by the β-thalassaemic patients are 5.71±1.22, 42.65±2.42, 106.79±5.35, 144.38±9.32, 192.53±23.32 and 271.50±50.64 for the age groups less than 1, 1-5, 6-10, 11-15, 16-20 and 20+ years, respectively. It shows that as age advances the β-thalassaemic patients are bound to take more and more transfusions.

Table 3.5

Health Condition Before Transfusion

Health Condition	Age in years	Male		Female		Total	
		No	%	No	%	No	%
Serious Problem	< 1	2	2.30	0	0.00	2	1.47
	1 - 5	25	28.74	11	22.45	36	26.47
	6 - 10	33	37.93	20	40.82	53	38.97
	11 - 15	20	22.99	15	30.61	35	25.74
	16 - 20	5	5.75	2	4.08	7	5.15
	20 +	2	2.30	1	2.04	3	2.21
	Total	87	100.00	49	100.00	136	100.01
No Serious Problem	< 1	3	1.90	2	2.33	5	2.05
	1 - 5	42	26.58	29	33.72	71	29.10
	6 - 10	68	43.04	33	38.37	101	41.39
	11 - 15	34	21.52	16	18.60	50	20.49
	16 - 20	8	5.06	4	4.65	12	4.92
	20 +	3	1.90	2	2.33	5	2.05
	Total	158	100.00	86	100.00	244	100.00

Table 3.5 and 3.6 show health condition before and after transfusion respectively. It is seen from Table 3.5 that out of 380 β-thalassaemic patients, 136 individuals (87 males and 49 females) had serious health problems, and 244 (158 males and 86 females) had no such serious health problem before taking transfusion. In this table, age-wise and sex-wise break-up have been given, which is self-explanatory.

Table 3.6

Difference After Treatmemt (Transfusion)

Condition with Transfusion	Age in years	Male		Female		Total	
		No	%	No	%	No	%
Improve-ment	< 1	3	1.91	2	2.47	5	2.10
	1 - 5	41	26.12	29	35.80	70	29.41
	6 - 10	71	45.22	31	38.27	102	42.86
	11 - 15	30	19.11	13	16.05	43	18.07
	16 - 20	9	5.73	4	4.94	13	5.46
	20 +	3	1.91	2	2.47	5	2.10
	Total	157	100.00	81	100.00	238	100.00
						(62.63)	
No Improve-ment	< 1	2	2.27	0	0.00	2	1.41
	1 - 5	26	29.55	11	20.37	37	26.06
	6 - 10	30	34.09	22	40.74	52	36.62
	11 - 15	24	27.27	18	33.33	42	29.58
	16 - 20	4	4.55	2	3.70	6	4.22
	20 +	2	2.27	1	1.85	3	2.11
	Total	88	100.00	54	99.99	142	100.00
						(37.37)	

After detection of β-thalassaemia, when the patients start taking transfusions regularly, the situation abruptly changes. Table 3.6 shows that how conditions of the patients have changed after having transfusions from time to time. It is found that nearly 62.63 per cent of all β-thalassaemic patients have felt an improvement in their health condition, whereas 37.37 per cent have not found any difference in their health status before and after transfusions. However, since this table is self-explanatory, it needs no further discussion.

Table 3.7

Menarcheal Age in β-thalassaemia Girls

Population	N	Mean	S.D.	± S.E.	Range in years	
Female thalassaemia patient	11	13.82	1.64	0.49	11 to 20+	Present study
Bengali speaking women with thalassaemia trait	48	13.63	1.21	0.12	10 to 16	Bhattacherya et.al 1982
Bengali speaking normal women	853	13.21	1.26	0.04	9 to 18	Bhattacherya et.al 1977

In the present study data on menarche have been collected on 40 β-thalassaemic girls aged 11+ years. Out of 40 girls with β-thalassaemia major, it is seen that only 11 (i.e., 27.50 per cent) of them have had menarche. Table 3.7 shows that the mean menarcheal age among the girls with β-thalassaemia major is 13.82±0.49 years. It is seen from the table that in the Bengali speaking women with β-thalassaemia trait the mean menarcheal age is found to be 13.63±0.12 years (Bhattacharjee *et al.*, 1977) and that for the normal girls is 13.21±0.04 years. So the mean menarcheal age among the girls with thalassaemia major is higher than that among the women with β-thalassaemia trait and the normal females. Since the

present sample is too meagre, we have not ventured to perform any statistical test on these data.

Section-II - Observations based on Secondary Data

In this section we have used the secondary data, taken from the hospital records.

Table 3.8

Total Haemoglobin Before Transfusion

Age Group in years	Male					Female				
	N	Range g/dl	Mean	S.D.	±S.E.	N	Range g/dl	Mean	S.D.	±S.E.
0 - 5	48	3.4-11.3	7.54	1.84	0.27	30	5.0-10.0	8.31	1.28	0.23
6 - 10	60	3.3-10.1	8.34	7.53	0.99	38	5.1-9.8	7.58	1.20	0.20
11 - 15	28	4.2-8.8	7.08	1.14	0.22	14	5.0-9.2	6.94	1.40	0.37
16 - 20	6	6.0-11.0	7.51	1.65	0.68	6	4.9-7.8	6.61	0.90	0.37

In the male β-thalassaemic patients, aged up to 5 years, before taking any transfusion, the mean total haemoglobin content was 7.54±0.27 g/dl. and the same in female β-thalassaemic patients aged upto 5 years, 8.31±0.23 g/dl (Table 3.8). In case of male β-thalassaemic patients, aged between 6 and 10 years, the mean total haemoglobin content was 8.34±0.99 g/dl. and the same in the β-thalassaemic girls of the same age-group was 7.58±0.20 g/dl. The mean total haemoglobin content was 7.08±0.22 g/dl. and 6.94±0.37 g/dl. in male and female β-thalassaemic patients, aged 11-15 years respectively. In the age-group 16-20 years,

the mean total haemoglobin content was 7.51±0.68 g/dl. in male β-thalassaemic patients and the same was 6.61±0.37 g/dl in the female β-thalassaemic patients.

Table 3.9

Total Haemoglobin After Transfusion

Age Group in years	Male					Female				
	N	Range g/dl	Mean	S.D.	±S.E.	N	Range g/dl	Mean	S.D.	±S.E.
0 - 5	25	6.8-12.8	10.44	1.69	0.38	18	7.2-14.2	10.87	1.70	0.40
6 - 10	44	5.2-13.2	9.45	1.72	0.26	30	7.8-15.6	10.06	1.45	0.27
11 - 15	22	6.5-12.0	9.26	1.39	0.30	12	6.5-12.0	9.29	1.56	0.45
16 - 20	4	8.0-10.0	8.65	0.82	0.41	4	7.1-10.7	8.80	1.29	0.64

Table-3.9 shows the total haemoglobin content in the β-thalassaemic patients after receiving transfusions. It is found that in the age-group upto 5 years, the mean total haemoglobin content is 10.44±0.38 g/dl in males and the same is 10.87±0.40 g/dl in females. In the age group 6-10 years the mean total haemoglobin content is found to be 9.45±0.26 g/dl and 10.06±0.27 g/dl in males and females respectively. The mean total haemoglobin content of 8.65±0.41 g/dl is found in the male β-thalassaemic patients, aged 16-20 years, and the same in the female β-thalassaemic patients of the same age group is 8.8±0.64 g/dl. The point to be noted here is that as age advances the total haemoglobin content, even after repeated transfusions, start falling. It may also be

noted that even after receiving repeated transfusions, the β-thalassaemic patients, both male and female, never reach the normal total haemoglobin level, as recommended by the W.H.O. (The W.H.O. Scientific Group, 1968a has recommended the haemoglobin level of 13.00 g/dl for adult males and 12.00 g/dl for the non-pregnant adult females). It shows that the β-thalassaemic patients cannot sustain the total haemoglobin level in their blood in spite of getting transfusions time and again.

Table 3.10

Performance of Splenectomy

Age Group in years	Male			Female		
	No	Yes	%	No	Yes	%
Upto 5	48	3	6.25	30	2	6.67
6-10	61	12	19.67	38	10	26.32
11-15	31	16	51.61	14	4	28.57
16-20	+6	2	33.33	6	3	50.00

Table 3.10 shows performance of splenectomy on the β-thalassaemic patients by sex and age-group. It is seen that in the age-group up to 5 years only 6.25 per cent of males and 6.67 per cent of females β-thalassaemic patients have had to undergo splenectomy. In the age group 6-10 years, 19.67 per cent of males and 26.32 per cent of females with β-thalassaemia major have undergone splenectomy. In the age group 11-15 years the percentage of the β-thalassaemic

patients, who have undergone splenectomy, has increased to 51.61 in males and 28.57 in females. In age group 16-20 years, nearly 33.33 per cent of all male β-thalassaemic patients and 50 per cent of all female β-thalassaemic patients have undergone splenectomy. However, the total data in the age group 16-20 years, are very few, and consequently the results cannot be relied upon. The point to be noted here is that as age advances, more and more patients with β-thalassaemia major undergo splenectomy. It holds good for both sexes.

Unfortunately, none of the hospitals in Calcutta, which are dealing with the β-thalassaemic patients keep in their records the differential blood count before and after transfusions. Consequently, in spite of our best attempt we have not been able to examine the changes in differential count in the β-thalassaemic patients before and after transfusions.

Mode of Treatment and Average Cost

Thalassaemia, one of the most common genetic disorders, can be detected by various haemotological tests very early in life. Recent evidences have shown that the β-thalassaemia heterozygotes can be distinguished from the normal infants by age of 3 months and with certainty by the age of 6 months. The HbA_2 level becomes elevated above the normal adult level by about 3 months and is clearly distinguishable from normal

by about 6 months (Weatherall, 1983). Once a child is diagnosed as having β-thalassaemia, various tests are performed to determine the level of differential count, total count of W.B.C., total count of R.B.C. abnormal cells, R.B.C. morphology, total haemoglobin level, etc.

The principle modes of treatment of thalassaemia are as follows:

1) Repeated blood transfusion to maintain near normal haemoglobin (over 10 g/dl.) level;

2) Removal of iron with Iron-chelating agent;

3) Bone marrow transplantations.

Blood transfusions continue to remain the main pillar of the treatment. Haemoglobin level has to be maintained near normal level and for this a patient has to visit hospital at least once in a month to even 3 to 4 times in a month, depending upon the condition of the patient. After taking about 40 transfusion, patients are advised to take Iron chelating agent to remove excess iron from the body.

Iron overload occurs in patients with β-thalassaemia due to repeated blood transfusion and iron absorption from the gut. Excessive iron deposition in various organs is responsible for their dysfunction and complication of therapy. In absence of adequate chelation therapy most of the β-thalassaemic patients succumb to the disease by second

decade of life. So the patients are advised to undergo chelation therapy to maintain a negative iron balance.

Iron chelation is done by desferrioxamine (DF, Desferal) which has to be administered subcutaneously over 8-10 hours for 5 to 6 times per week with the help of a infusion pump. Subcutaneous administration of D.F.O. often results in local pain and swelling, while the prolonged used produces decreased visual activity and hearing loss. Because of high cost of desferal and decreasing compliance with advancing age, a new oral iron chelator named Deffriprone/Kelfer has been evaluated and it is used in 2 to 4 divided doses of 50 to 100 mg/day. It is found to be much cheaper than Desferal.

Average Cost

Infusion Pump: It costs Rs.20,000 to Rs.25,000/-. Each dose of Desferal costs Rs.200/-. If injected 5 to 6 times per week, the total cost comes to be Rs.4800/- per month. Blood Bag (each transfusion) costs Rs.495/- to Rs.645/-.

So on the whole, the approximate expenditure, incurred for a patient per month for proper growth and development and to lead a normal life, comes about Rs.5295/- to Rs.5445/- plus some other expenditures.

The treatment that gives more relief for longer period to the β-thalassaemic patient is Bone-marrow transplantation.

However, it is done only in few selected places like CMCH, Vellore and Bombay. Here erythroid stem cells are replaced by normal erythroid precursor cells. However, transplantations can be administered in those cases where HLA compatible donors can be found. The cost of Bone-Marrow transplantation varies between 2.5 lakhs and 6 lakhs, depending on complication of past transplantation. So it is generally not within the reach of ordinary individuals. Few patients from Calcutta who have undergone bone marrow transplantation, have been helped by various N.G.O's. In the present sample, not a single case has been found to have undergone bone marrow transplantation.

Section-III - Growth and Development

In order to understand the growth and development processes in the patients with β-thalassaemia major, 8 anthropometric and 5 skinfold measurements have been taken into consideration. They are height, weight, chest girth (inhale and exhale), humerous diameter, bicondylar femur diameter, biceps girth, calf girth, and 5 skinfold measurements such as biceps, triceps, subscapular, supra-illiac and calf.

Table 3.11

Means and Standard Deviations of Anthropometric Measurements (for Females)

Anthropometric Traits		Age Group in Years															
		5				6-10				11-15				16-20			
		N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.
Height	C	45	106.04	0.71	4.79	46	124.76	1.65	11.20	30	146.86	1.61	8.82	7	154.66	1.58	4.17
	T	24	90.98	2.41	11.82	32	115.91	1.40	7.93	23	128.98	1.65	7.90	4	134.00	4.14	24.48
Weight	C	45	15.50	0.29	1.93	46	24.00	1.16	7.87	30	36.70	1.52	8.35	7	46.43	2.69	7.13
	T	28	12.23	0.38	2.03	36	18.72	0.56	3.36	25	27.26	1.26	6.30	6	29.66	3.12	7.65
Chest Girth (inhale)	C	44	52.42	0.47	3.10	46	59.51	1.01	6.89	30	71.38	1.29	7.04	7	78.21	2.22	5.88
	T	32	51.13	0.71	3.99	43	57.69	0.66	4.39	30	65.92	1.10	6.00	7	66.93	2.28	6.03
Chest Girth (exhale)	C	44	51.26	0.49	3.23	46	57.82	0.10	6.76	30	69.48	1.34	7.35	7	76.36	2.28	6.05
	T	32	49.91	0.71	4.02	43	56.28	0.68	4.47	30	64.00	1.06	5.83	7	65.00	2.23	5.91
Humerous Diameter	C	45	4.24	0.04	0.24	46	4.78	0.06	0.38	30	5.35	0.05	0.27	7	5.39	0.06	0.16
	T	33	3.85	0.07	0.38	43	4.28	0.06	0.38	30	4.87	0.07	0.36	7	5.13	0.21	0.54
Bicondylar Femur Diameter	C	45	3.64	0.04	0.27	46	4.10	2.04	0.30	30	4.67	0.04	0.22	7	4.40	0.13	0.34
	T	31	4.26	0.10	0.57	43	4.78	0.09	0.56	30	5.16	0.85	0.46	7	5.44	0.15	0.40
Bicep Girth	C	45	15.42	0.19	1.27	46	17.13	0.39	2.63	30	19.97	0.49	2.67	7	22.21	0.84	2.22
	T	34	14.21	0.38	2.21	43	14.97	0.27	1.75	30	16.62	0.22	1.22	7	15.71	1.44	3.81
Calf Girth	C	45	20.00	0.21	1.40	46	23.52	0.44	3.00	30	27.74	0.54	2.94	7	30.97	1.09	2.88
	T	35	18.72	0.45	2.64	43	20.75	0.35	2.30	30	23.16	0.29	1.60	7	29.24	1.24	3.27

Continued Table 3.11

Anthropometric Traits		Age Group in Years															
		5				6-10				11-15				16-20			
		N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.
Skinfold Thickness																	
Biceps	C	45	3.78	0.21	1.40	46	3.63	0.30	2.05	30	5.37	0.42	2.30	7	4.57	0.63	1.67
	T	33	3.09	0.24	1.38	43	3.19	0.26	1.67	30	3.33	0.29	1.60	7	3.00	0.64	1.69
Triceps	C	45	7.06	0.32	2.17	46	7.78	0.62	4.23	30	9.87	0.75	4.10	7	13.00	1.59	4.21
	T	33	6.18	0.29	1.64	43	6.35	0.31	2.02	30	7.37	0.34	1.85	7	6.86	1.27	3.36
Subscapular	C	45	5.09	0.24	1.60	46	6.33	0.45	3.07	30	9.33	0.67	3.66	7	10.86	1.52	4.02
	T	32	4.44	0.27	1.52	43	4.30	0.29	1.91	30	5.20	0.19	1.05	7	5.29	1.19	3.15
Supra-illiac	C	45	4.33	0.34	2.28	46	6.46	0.71	4.80	30	7.80	0.65	3.53	7	9.29	1.41	3.73
	T	31	3.42	0.20	1.15	43	3.58	0.30	1.98	30	4.97	0.32	1.74	7	4.86	1.06	2.80
Calf	C	45	8.49	0.29	1.89	46	9.24	0.42	2.84	30	11.46	0.52	2.85	7	12.86	1.15	3.04
	T	33	7.24	0.37	2.13	43	7.33	0.31	2.05	30	8.20	0.47	2.57	7	8.43	1.70	4.50

Table 3.11 shows various anthropometeric measurements and skinfold thickness for both β-thalassaemic and normal girls. It is seen that in the age group upto 5 years the mean height is 106.04±0.71 cm. in normal girls and that in the β-thalassaemic girls 90.98±2.41 cm. The mean weight in the Controlled sample is found to be 15.5±0.29 kg. and in the β-thalassaemic girls the same is 12.23±0.38 kg. The mean chest girth, inhale and exhale, in the Controlled sample is

52.42±0.47 cm. and 51.26±0.49 cm. respectively and those in the β-thalassaemic girls are 51.23±0.71 cm. and 49.91±0.71 cm. respectively. The mean humerous diameter is found to be 4.24±0.04 and 3.85±0.07 cm. in the Controlled and β-thalassaemic samples respectively. The mean bicondylar femur diameter is found to be 3.64±0.04 cm. and 4.26±0.10 cm. respectively in the normal individuals and β-thalassaemic patients. The mean biceps girth among the normal individuals is 15.42±0.19 cm and that in the β-thalassaemic girls is 14.21±0.38 cm. The mean calf girth in the Controlled sample and β-thalassaemic patients is found to be 20.00±0.21 cm. and 18.72±0.45 cm., respectively.

The mean biceps thickness in the Controlled sample is found to be 3.78±0.21 mm. and in the β-thalassaemic patients it is 3.09±0.24 mm. So far as triceps is concerned, in the Controlled and the β-thalassaemic samples the mean measurement is found to be 7.06±0.32 mm. and 6.18±0.29 mm. respectively. The mean subscapular thickness in the Controlled and β-thalassaemic samples is found to be 5.09±0.24 mm and 4.44±0.27 mm. respectively. The mean supra-illiac thickness in the Controlled and β-thalassaemic individuals is found to be 4.33±0.34 mm. and 3.42±0.20 mm. respectively. The mean calf thickness is found to be 8.49±0.29 mm in the Controlled sample and 7.24±0.37 mm. in the β-thalassaemic individuals.

In the age-group 6-10 years the mean height of the Controlled females is found to be 124.76±1.65 cm. whereas that in the β-thalassaemic girls is 115.91±1.40 cm. The mean body weight of the Controlled and β-thalassaemic girls is 24.00±1.16 kg. and 18.72±0.56 kg. respectively. The mean chest girth (inhale) in the Controlled and β-thalassaemic samples is found to be 59.51±1.01 cm. and 57.69±0.66 cm. respectively. The mean chest girth (exhale) in the Controlled sample and β-thalassaemic girls is found to be 57.82±0.10 cm. and 56.28±0.68 cm. respectively. The mean humerous diameter is 4.78±0.06 cm and 4.28±0.06 cm in the Controlled and β-thalassaemic samples respectively. The bicondylar femur diameter is found to be 4.10±0.04 cm. in the Controlled sample and 4.78±0.09 in the β-thalassaemic girls. It is found that the mean biceps girth in the Controlled and β-thalassaemic samples is 17.13±0.39 cm. and 14.97±0.27 cm. respectively. In the Controlled sample and β-thalassaemic individuals the mean calf girth is found to be 23.52±0.44 cm. and 20.75±0.35 cm. respectively.

The mean biceps thickness in the Controlled sample is 3.63±0.30 mm. and that in the β-thalassaemic sample 3.19±0.26 mm. In the Controlled and β-thalassaemic samples the mean triceps thickness is found to be 7.78±0.62 mm. and 6.35±0.31 mm. respectively. The mean subscapular thickness in the Controlled and β-thalassaemic samples is 6.33±0.45 mm. and

4.30±0.29 mm. respectively. In the Controlled and β-thalassaemic samples the mean supra-illiac thickness is found to be 6.46±0.71 mm. and 3.58±0.30 mm. respectively. In calf thickness the Controlled sample shows the mean value of 9.24±0.42 mm. and that in the β-thalassaemic sample is found to be 7.33±0.31 mm.

In the age-group 10-15 years the mean height in the Controlled sample is found to be 146.86±1.61 cm. whereas the same in the β-thalassaemic patients is 128.98±1.65 cm. The mean weight in the Controlled sample and β-thalassaemic patients is found to be 36.70±1.52 kg. and 27.26±1.26 kg. respectively. The mean chest girth (inhale) in the Controlled sample and β-thalassaemic patients is 71.38±1.29 cm. and 65.92±1.10, respectively. Similarly, the mean chest girth (exhale) in the Controlled sample and β-thalassaemic patients is 69.48±1.34 cm. and 64.06±1.06 cm. respectively. In the Controlled sample the mean humerous diameter is found to be 5.35±0.04 cm. and in the β-thalassaemic patients is 4.87±0.07 cm. The mean bicondylar femur diameter in the Controlled and β-thalassaemic samples is 4.67±0.04 cm. and 5.16±0.85 cm. respectively. In the Controlled sample the mean biceps girth is found to be 19.97±0.49 cm. and that in the β-thalassaemic patient is 16.62±0.22 cm. In Controlled and β-thalassaemic samples the mean calf girth is found to be 27.74±0.54 cm. and 23.16±0.29 cm. respectively.

The mean biceps thickness in the Controlled and β-thalassaemic samples is 5.37±0.42 mm. and 3.33±0.29 mm. respectively. Similarly, the mean triceps thickness in the Controlled sample is 9.87±0.75 mm. and in the β-thalassaemic sample it is found to be 7.37±0.34 mm. So far as subscapular thickness is concerned, the mean value is found to be 9.33±0.67 mm. in the Controlled sample and 5.20±0.19 in the β-thalassaemic sample. It is found that the mean supra-illiac in the Controlled sample is 7.80±0.65 mm. and in the β-thalassaemic sample it is found to be 4.97±0.32 mm. The mean calf thickness in the Controlled and β-thalassaemic samples is 11.46±0.52 mm. and 8.20±0.47 mm. respectively.

In the age group 15-20 years it is seen that the mean height is 154.66±1.58 cm. in the Controlled sample and 134.00±4.14 in the β-thalassaemic sample. The mean weight is found to be 46.43±2.69 kg. in the Controlled sample and 29.66±3.12 kg. in the β-thalassaemic sample. The mean chest girth (inhale) in the Controlled sample is 78.21±2.22 cm. and that in the β-thalassaemic sample 66.93±2.28 cm. The mean chest girth (exhale) in the Controlled and β-thalassaemic samples is found to be 76.36±2.29 and 65.00±2.23 cm. respectively. The mean humerous diameter in the Controlled and β-thalassaemic samples is found to be 5.39±0.06 cm. and 5.13±0.21 cm. respectively. In the Controlled and β-thalassaemic samples the mean bicondylar femur diameter is

4.40±0.13 and 5.44±0.15 respectively. The mean biceps girth in the Controlled sample is found to be 22.21±0.84 cm. and that in the β-thalassaemic sample 15.71±1.44 cm. In the Controlled and β-thalassaemic samples the mean calf girth is found to be 30.97±1.09 and 24.04±1.24, respectively.

In the Controlled and β-thalassaemic samples the mean biceps thickness is found to be 4.57±0.63 mm. and 3.00±0.64 mm. respectively. So far as triceps thickness is concerned, the Controlled sample shows the mean value of 13.00±1.59 mm. and in the β-thalassaemic sample the same is 6.86±1.27 mm. The mean subscapular thickness in the Controlled and β-thalassaemic samples is found to be 10.86±1.52 mm. and 5.29±1.19 mm. respectively. The mean supra-illiac thickness in the Controlled sample is 9.29±1.41 and in the β-thalassaemic sample it is found to be 4.86±1.06 mm. The mean calf thickness in the Controlled and β-thalassaemic samples is found to be 12.86±1.15 mm. 8.43±1.70 mm. respectively.

The entire data for both the Controlled and β-thalassaemic groups have been classified into four age-groups. It is all through observed that in all measurements, excepting bicondylar femur diameter, the mean values in the Controlled sample are higher than those in the β-thalassaemic sample. The above discussion on various anthropometeric measurements is on the female samples.

Table 3.12

t-value for the Difference in Respect of Various Anthropometric Traits Between Controlled and Thalassaemic Females

Age Group	Height	Weight	Chest Girth (inhale)	Chest Girth (exhale)	Humerous Diameter	Bicondylar Femur Dia.	Bicep Girth	Calf Girth	Biceps	Triceps	Subcapular	Supra-illiac	Calf
0 - 5	5.99*	6.84*	1.98*	1.56	4.84*	5.76*	2.85*	2.58*	2.16*	2.06*	1.80	2.28*	2.69*
6 -10	4.09*	4.11*	1.50	1.28	5.77*	6.90*	4.55*	4.93*	1.11	2.06*	3.79*	3.74*	3.66*
11-15	7.76*	4.78*	3.22*	3.17*	5.58*	5.48*	6.24*	7.47*	4.00*	3.07*	5.93*	3.96*	4.65*
16-20	12.11*	9.32*	5.88*	9.39*	3.25*	13.89*	10.31*	10.76*	4.59*	7.98*	10.42*	8.27*	8.29*

* Significant at 5% level of probability.

In Table 3.12 shows the results of t-test, carried out in respect of all anthropometric measurements, considered in the present study between the Controlled and β-thalassaemic female samples. It is seen that the mean height in the Controlled sample of all age groups is significantly higher than that in the β-thalassaemic sample of all those age-groups at 5 per cent level of probability. Similarly, this is true for body weight, humerous diameter, bicondylar femur diameter, bicep girth, calf girth, triceps, supra-illiac and calf. In case of chest girth (inhale) the mean value in the Controlled sample is significantly higher at 5 per cent level of probability in all age groups, except in the age group 6-10 years. In case of chest girth (exhale) it is also seen that there is significant difference between the Controlled and β-thalassaemic samples in the age groups 11-15 years and

Female Sample

Growth Curve for Height of Controlled & Thalassaemic Girls

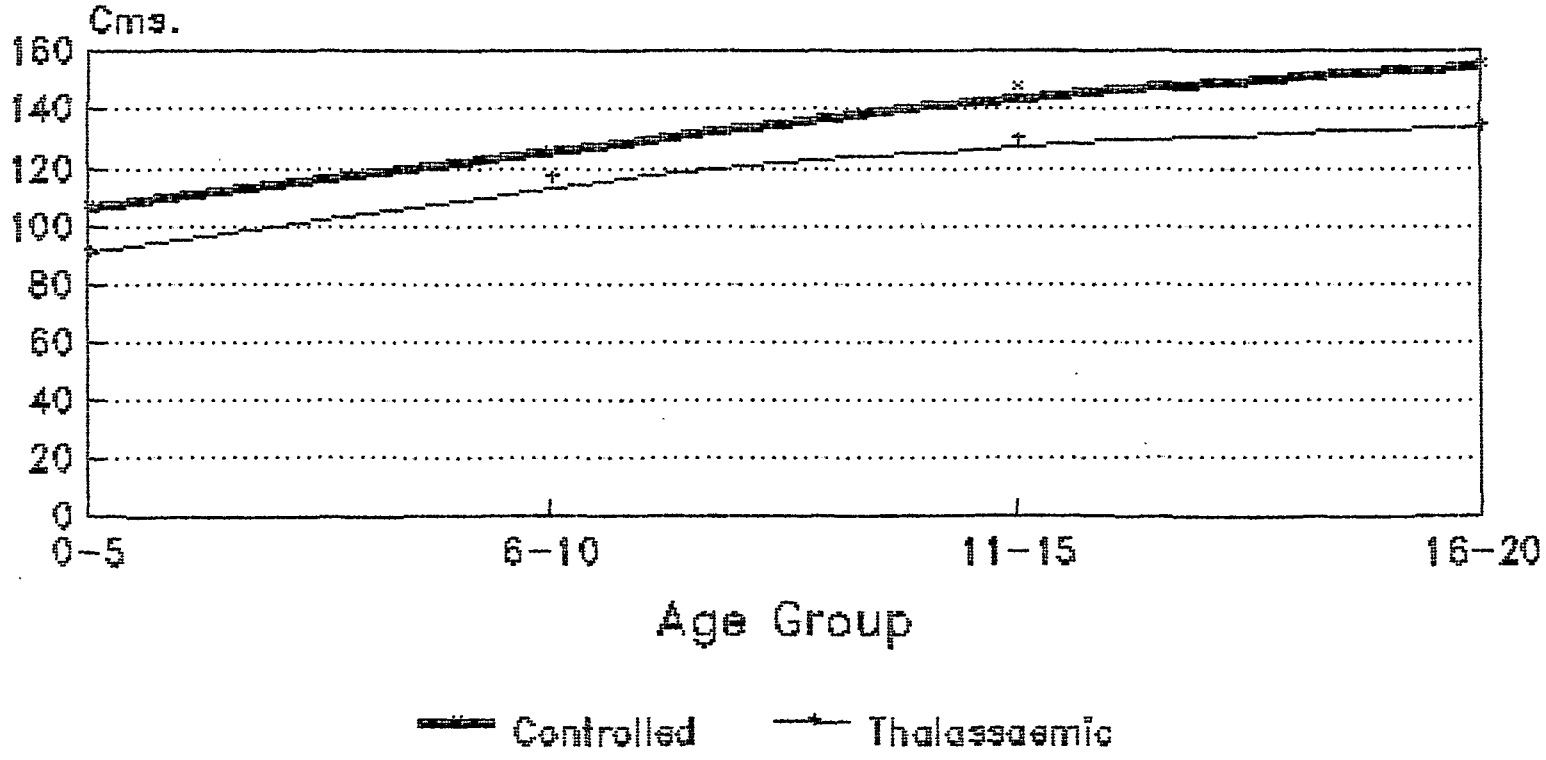

Fig. 1

Female Sample

Growth Curve for Weight of Controlled & Thalassaemic Girls

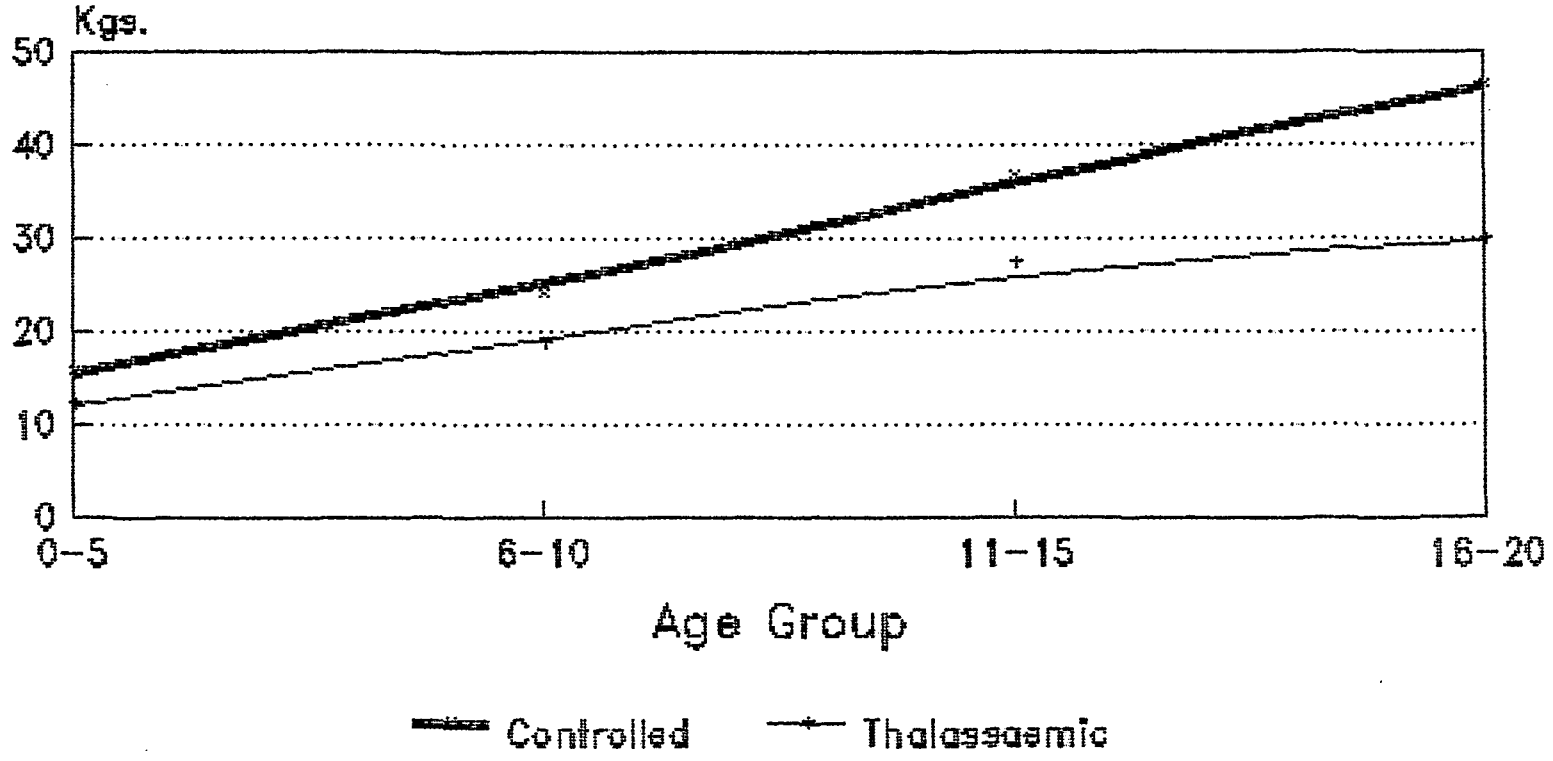

Fig. 2

16-20 years. However, it is also observed that though the mean value of chest girth (exhale) is higher in the Controlled sample in the age groups less than 5 years and 6-10 years than that in the β-thalassaemic patients, the differences between the Controlled and β-thalassaemic samples are not statistically significant. So far as the mean subscapular thickness is concerned it is found that the β-thalassaemic patients are having significantly lower value than the Controlled sample in all age groups, except in the age-group 0-5 years in which the mean value of the Controlled sample is insignificantly higher than that in their β-thalassaemic counterparts. It may be noted that So far as the bicondylar femur diameter is concerned, the β-thalassaemic patients are having significantly higher mean value in all age-groups than the Controlled sample. In fine, it may be noted that the β-thalassaemic patients, in respect of all measurements, excepting bicondylar femur diameter, are having significantly lower values than their normal counterparts.

Figure-1 shows growth curve for height between the Controlled sample and β-thalassaemic girls. It shows that the growth curve is lower in β-thalassaemic girls in comparison to the Controlled sample.

Figure-2 shows growth curve in respect of weight between the Controlled and the β-thalassaemic girls. It is

Female Sample

Growth Curve for Chest Girth of Controlled & Thalassaemic Girls

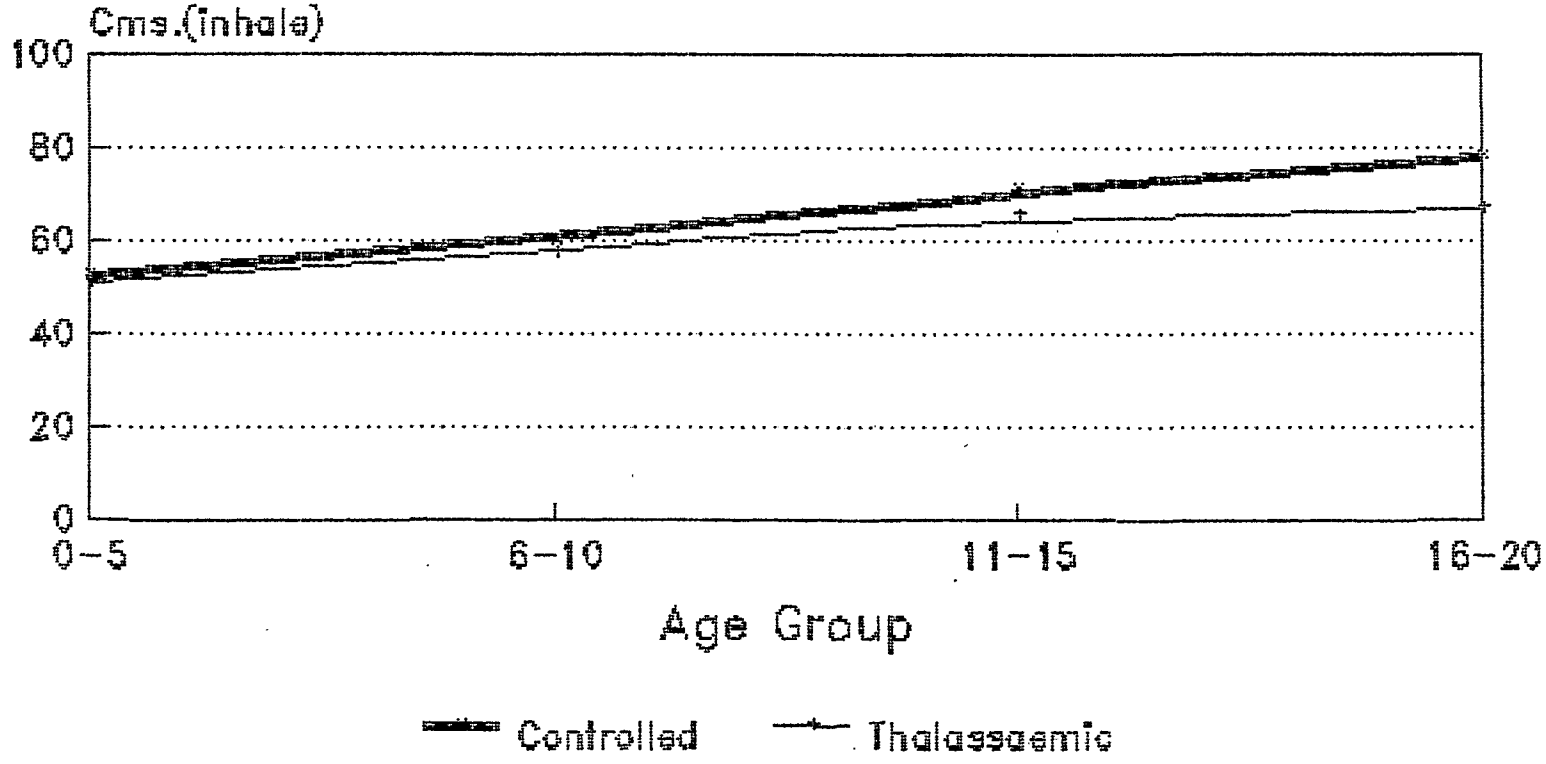

Fig. 3

Female Sample

Growth Curve for Chest Girth of Controlled & Thalassaemic Girls

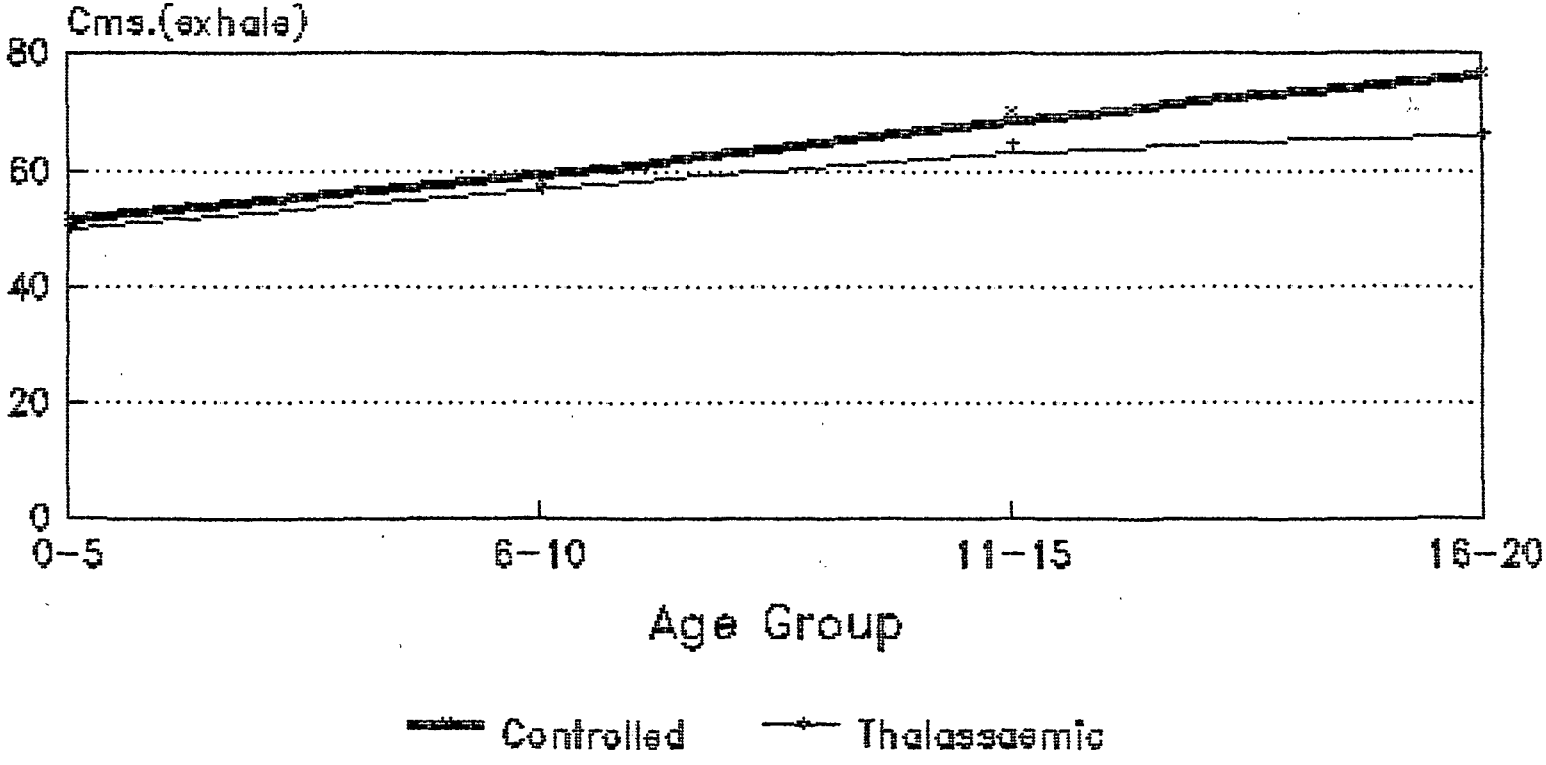

Fig. 4

Female Sample

Growth Curve for Humerous Diameter of Controlled & Thalassaemic Girls

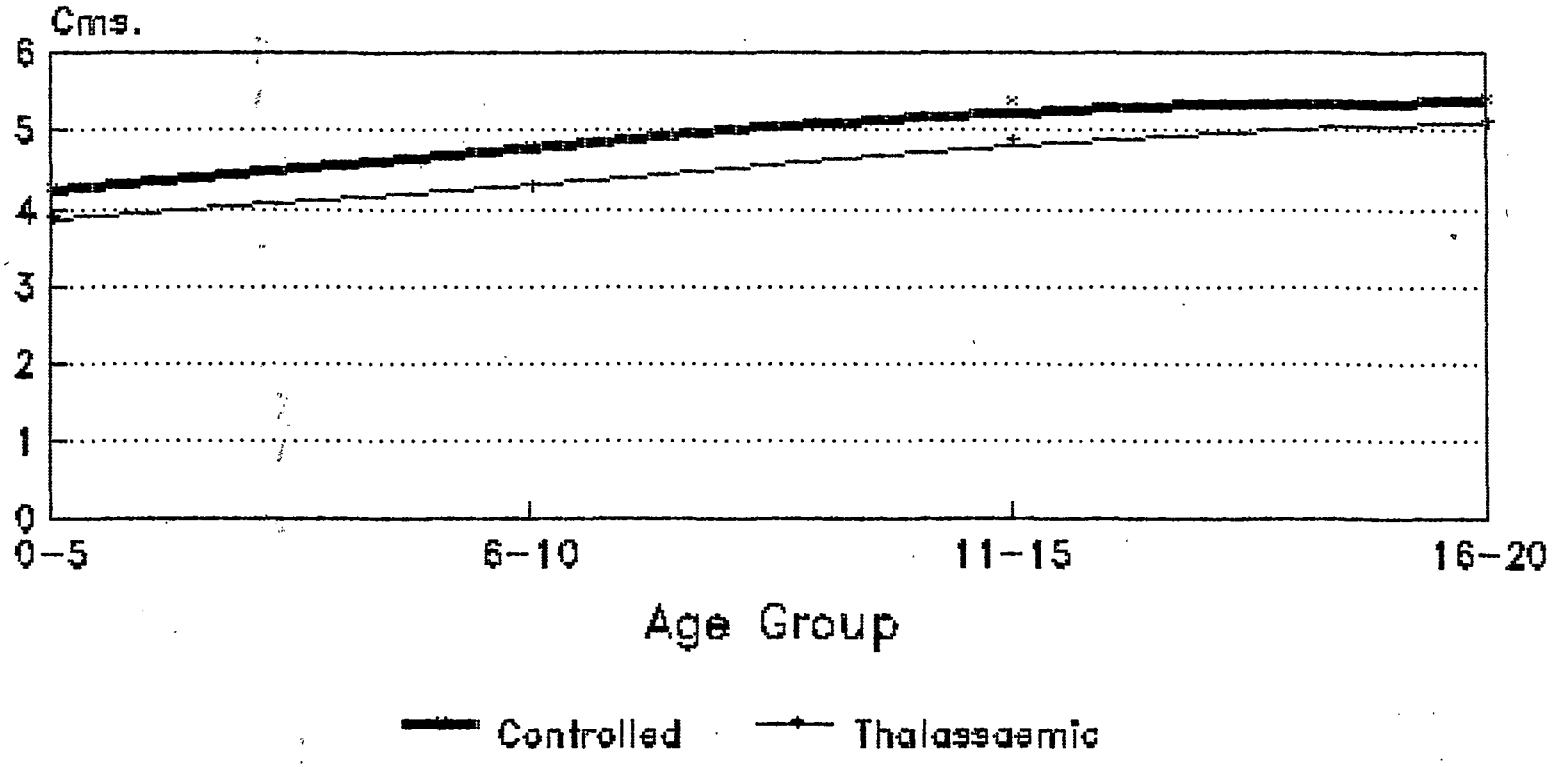

Fig. 5

Female Sample

Growth Curve for Bicondylar Femur

Dia. of Controlled & Thalassaemic Girls

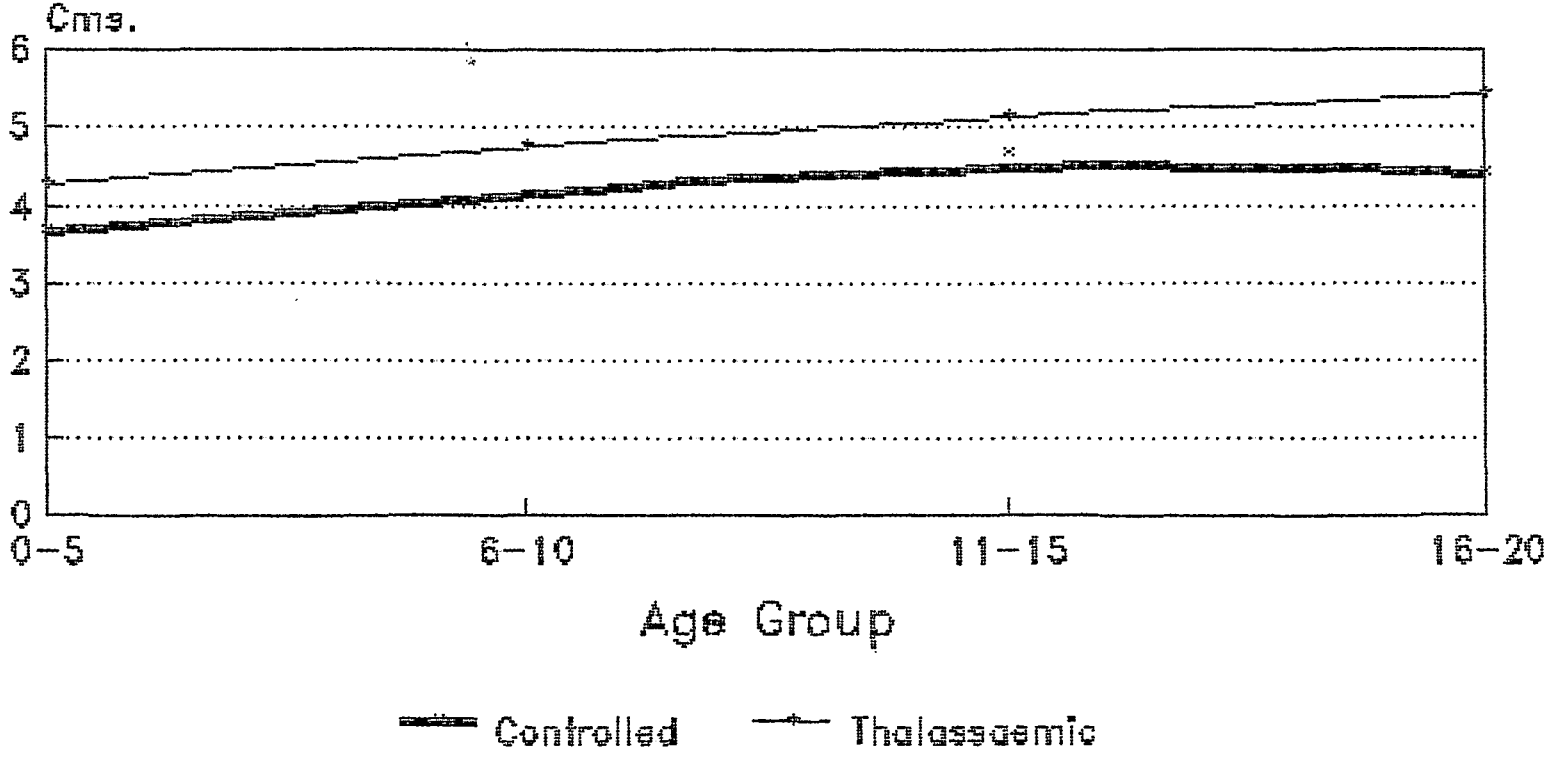

Fig. 6

Female Sample

Growth Curve for Bicep Girth of Controlled & Thalassaemic Girls

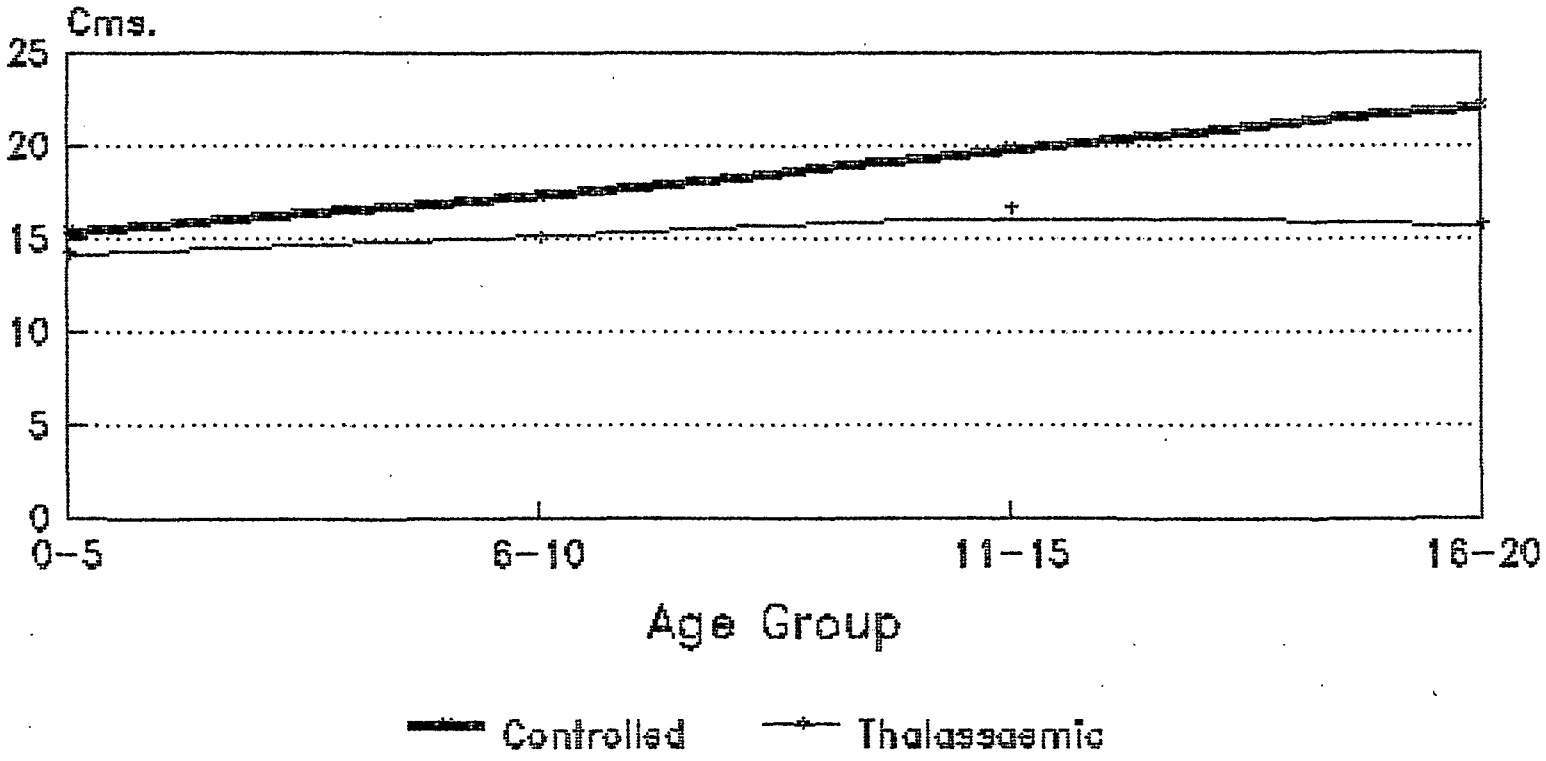

Fig. 7

Female Sample

Growth Curve for Calf Girth of Controlled & Thalassaemic Girls

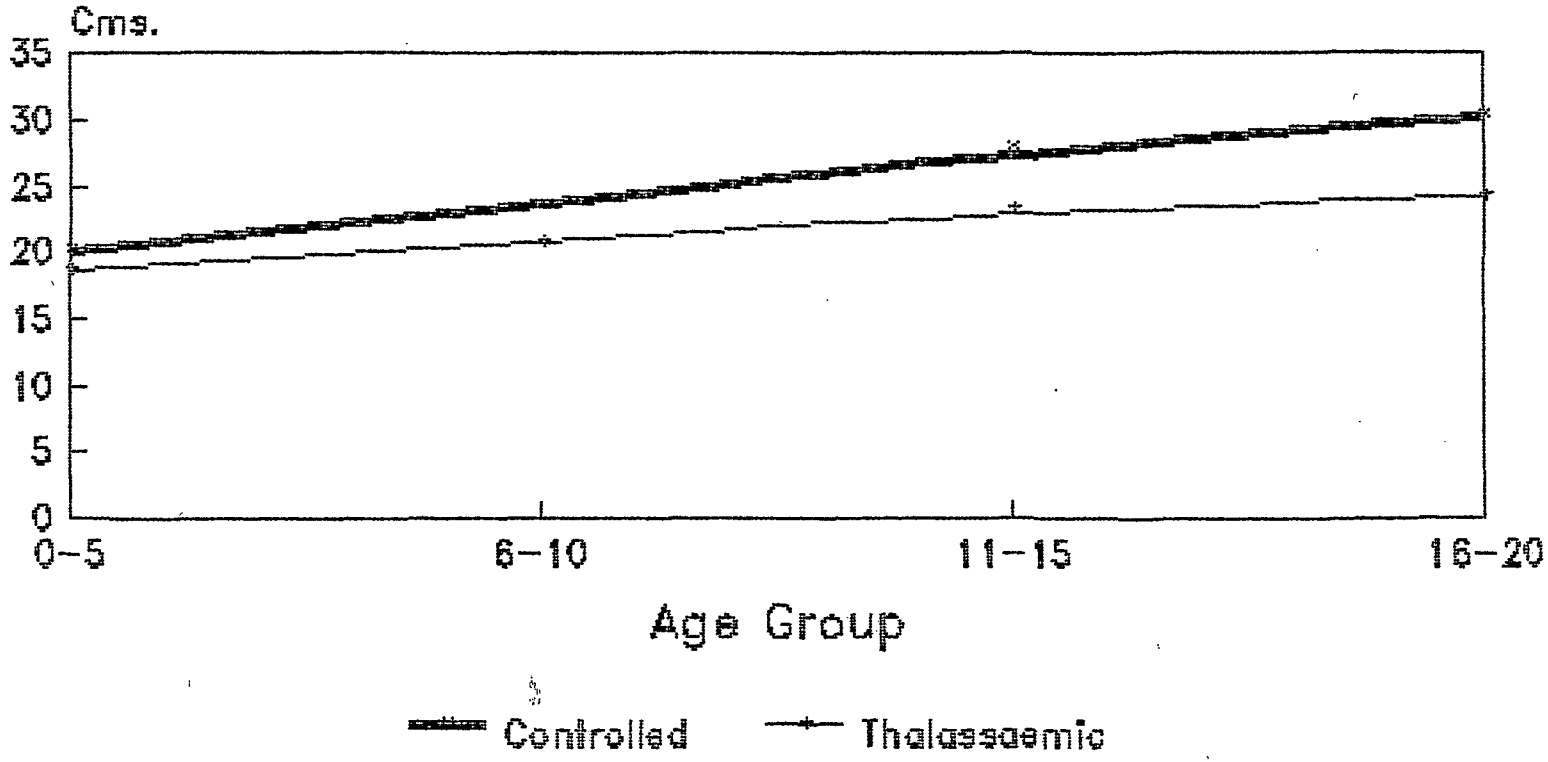

Fig. 8

found that in all age groups the weight is lower in the β-thalassaemic girls than in the Controlled sample.

Figure-3 shows growth curve for chest girth (inhale). It indicates that as age advances the difference between the Controlled and the β-thalassaemic girls becomes broader.

Figure-4 shows that the growth curve for chest girth (exhale) between the Controlled and the β-thalassaemic girls is the same as in Figure-3.

Figure-5 shows that growth curve for humerous diameter between the Controlled and β-thalassaemic girls. The difference between them remains by and large same from lowest to the highest age group and the β-thalassaemic girls shows lower value in comparison to the normal girls.

Figure-6 shows the growth curve for bicondylar femur diameter between the Controlled and the β-thalassaemic girls. It is seen that the normal girls show lesser value than the β-thalassaemic girls and as age advances the difference between them increases.

Figure-7 shows the growth curve in respect of biceps girth. The curve indicates that girls have greater mean values in all age groups in comparison to their β-thalassaemic counterparts and as age advances the difference between them also increases.

Figure-8 depicts a comparison in respect of calf girth between the Controlled and β-thalassaemic girls. It shows

Female Sample

Growth Curve for Skinfold Thickness at Biceps of Con. & Tha. Girls

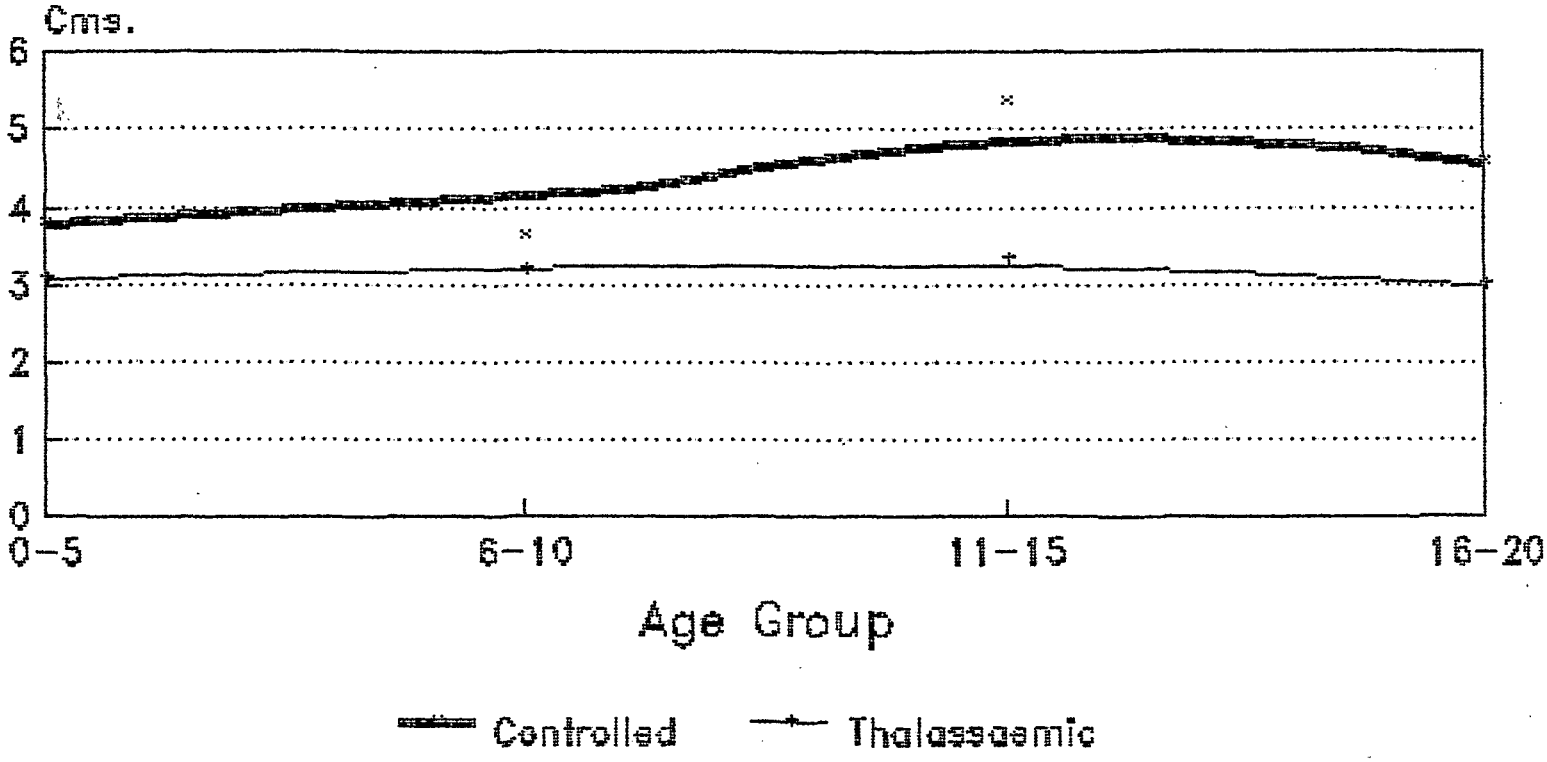

Fig. 9

Female Sample

Growth Curve for Skinfold Thickness at Triceps of Con. & Tha. Girls

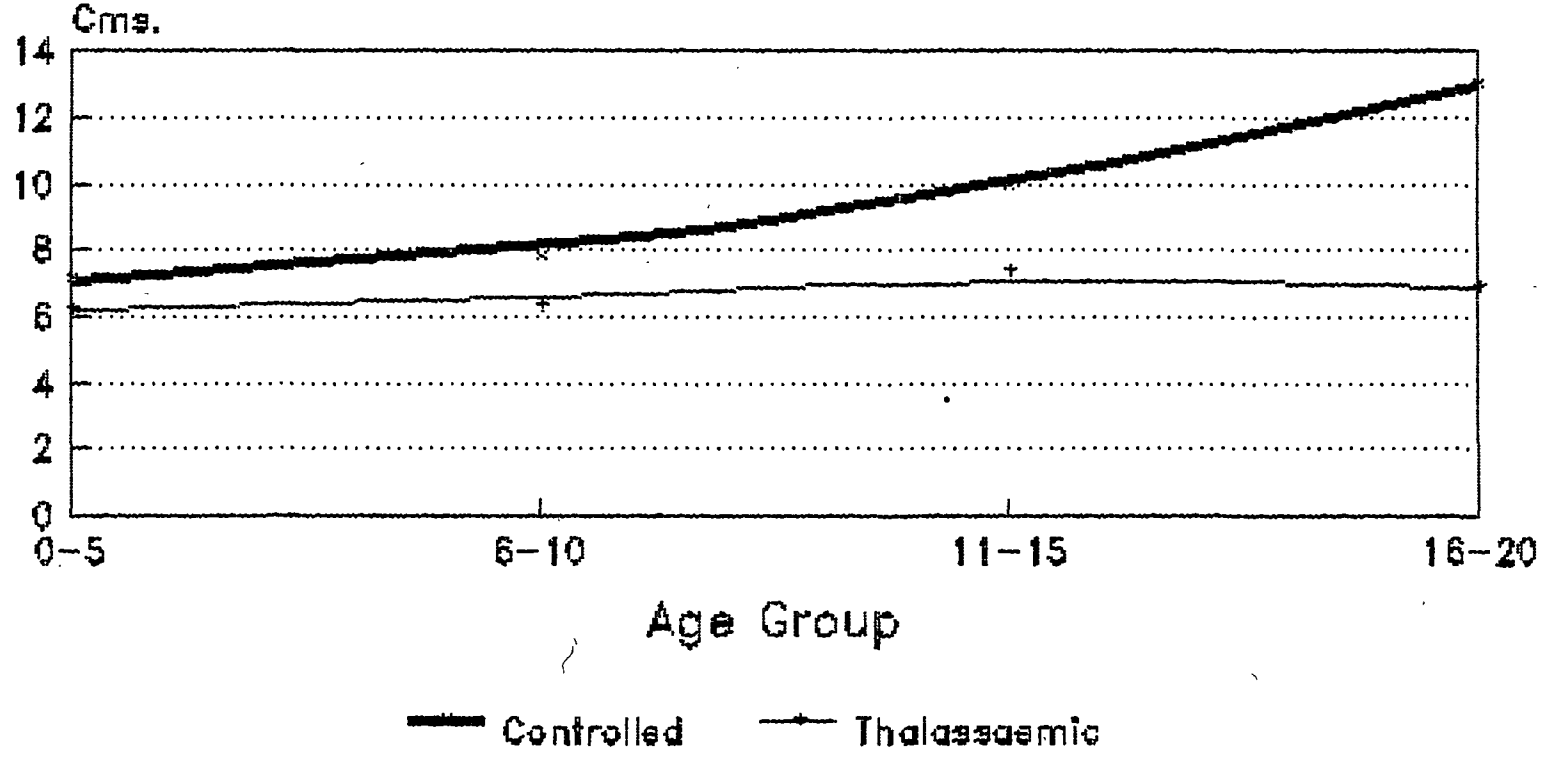

Fig. 10

Female Sample

Growth Curve for Skinfold Thickness at Subscapular of Con. & Tha. Girls

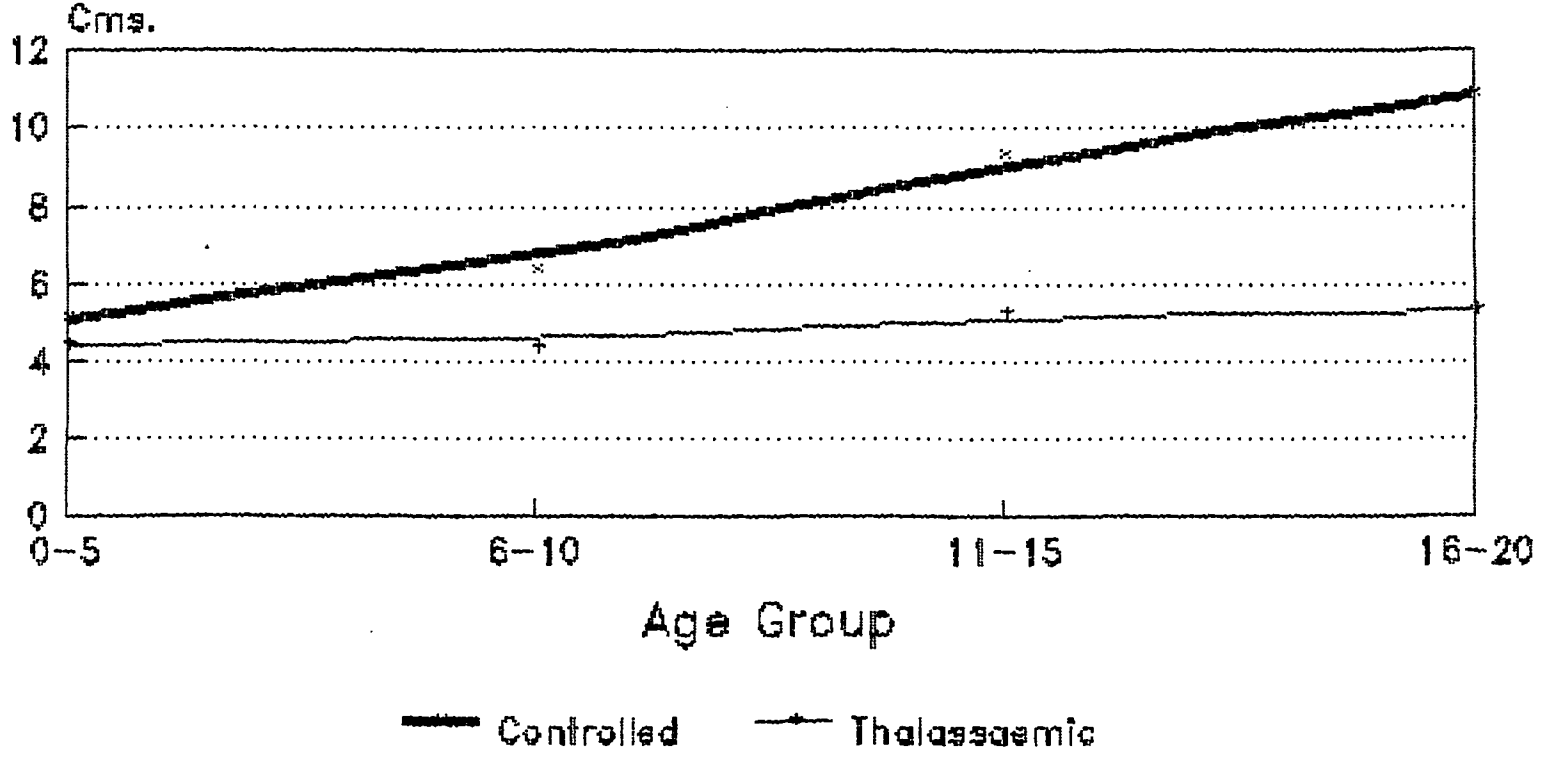

Fig. 11

Female Sample

Growth Curve for Skinfold Thickness at Supra-illiac of Con. & Tha. Girls

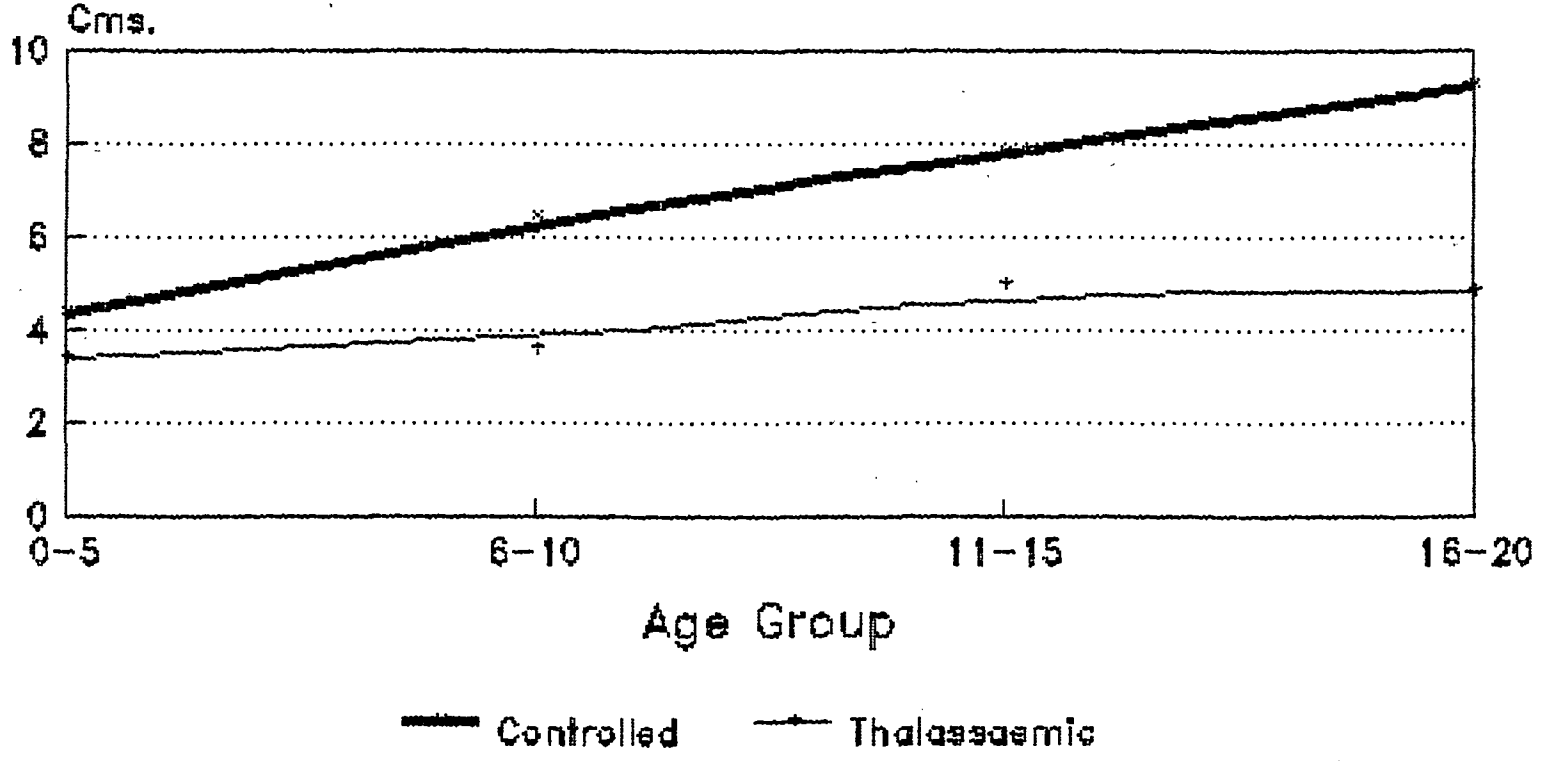

Fig. 12

that the normal girls always have mean higher value in all age groups in comparison to the β-thalassaemic girls and the difference between them in respect of this character broadens as age advances.

Figure-9 shows a comparative picture between the Controlled and β-thalassaemic girls in respect of biceps. It is seen that the normal girls always have a higher value than the β-thalassaemic girls and the difference between them starts broadening from the age-group 6-10 years.

Figure-10, shows growth curve in respect of triceps between the Controlled and the β-thalassaemia girls. It is clear that the normal girls in all age-groups have higher values than their β-thalassaemic Counterparts and the difference between them gets broadening from the age group of 6-10 years.

Figure-11 depicts a comparative picture in respect of subscapular thickness between the Controlled and the β-thalassaemic girls. It shows that the normal girls from the very beginning have higher values in comparison to their β-thalassaemia counterparts and the difference between them gets broadening from the age group 6-10 years.

Figure-12 shows growth curve in respect of supra-illiac thickness between the Controlled and the β-thalassaemic girls. It is seen that normal girls always have higher values in all

Female Sample

Growth Curve for Skinfold Thickness at Calf of Controlled & Thalassaemic Girls

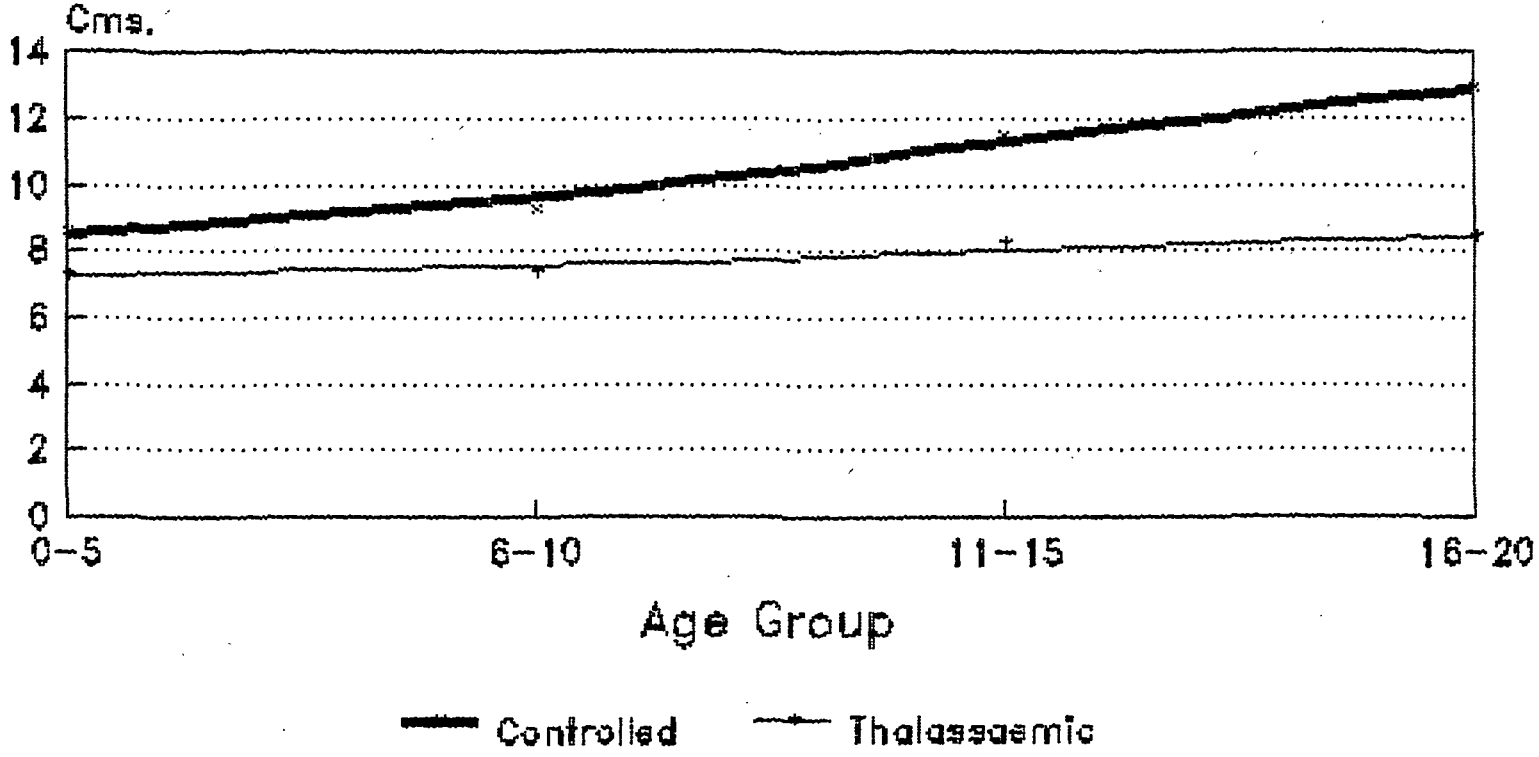

Fig. 13

age groups and as age advances the difference between them broadens.

Figure-13 shows growth curve in respect of calf thickness between normal and β-thalassaemic girls. It is seen that normal girls always have higher values in all age groups and as age advances the difference between them broadens.

Table 3.13

Means and Standard Deviations of Anthropometric Measurements (for Males)

Anthropometric Traits		Age Group in Years															
		5				6-10				11-15				16-20			
		N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.
Height	C	72	106.72	0.73	6.23	101	127.96	0.83	8.29	53	153.68	1.75	12.76	9	169.54	1.61	4.83
	T	37	95.84	1.21	7.38	70	116.55	1.19	9.84	27	134.54	1.67	8.66	5	148.94	3.68	8.23
Weight	C	72	16.62	0.34	2.91	101	24.56	0.55	5.68	53	42.34	1.54	11.22	9	55.39	3.54	10.63
	T	46	12.72	0.43	2.93	78	19.62	0.39	3.48	31	26.97	0.86	4.81	7	41.71	2.72	7.19
Chest Girth (inhale)	C	72	53.69	0.44	3.74	101	61.82	0.56	5.67	53	74.54	1.03	7.47	9	82.61	2.44	7.33
	T	41	51.95	0.57	3.65	105	58.48	0.44	4.56	52	64.17	0.79	5.68	9	75.57	2.07	6.20
Chest Girth (exhale)	C	72	52.43	0.44	3.77	101	59.96	0.55	5.57	53	72.08	0.97	7.08	9	79.61	2.55	7.66
	T	41	51.63	0.56	3.39	105	57.02	0.44	4.52	52	62.36	0.77	5.53	9	73.12	1.85	5.55
Humerous Diameter	C	71	4.46	0.03	0.23	101	5.00	0.032	0.33	53	6.03	0.08	0.57	9	6.06	0.14	0.43
	T	63	3.90	0.04	0.35	105	4.38	0.05	0.38	52	4.98	0.08	0.57	9	5.89	0.13	0.38
Bicondylar Femur Diameter	C	71	3.73	0.03	0.31	101	4.66	0.03	0.31	53	5.05	0.05	0.33	9	5.47	0.17	0.51
	T	64	4.48	0.07	0.54	105	4.98	0.05	0.52	52	5.38	0.09	0.66	9	6.16	0.27	0.81

Continued Table 3.11

Anthropometric Traits		Age Group in Years															
		5				6-10				11-15				16-20			
		N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.
Bicep Girth	C	72	15.66	0.17	1.44	101	17.35	0.24	2.44	53	21.22	0.42	3.08	9	23.52	0.94	2.82
	T	66	13.96	0.16	1.27	105	14.80	0.15	1.56	52	15.72	0.25	1.82	9	19.02	0.58	1.74
Calf Girth	C	72	20.72	0.23	1.99	101	24.08	0.24	2.40	53	29.87	0.46	3.31	9	33.72	1.08	3.23
	T	66	18.06	0.22	1.82	105	20.49	0.23	2.39	52	21.94	0.35	2.55	9	26.60	1.05	13.24
Skinfold Thickness																	
Biceps	C	71	3.51	0.20	1.66	101	3.92	0.25	2.48	53	4.58	0.36	2.66	9	3.78	0.73	2.20
	T	65	2.85	0.12	0.98	104	2.79	0.13	1.28	52	3.06	0.21	1.55	9	2.78	0.31	0.92
Triceps	C	71	6.52	0.29	2.47	101	7.12	0.40	4.03	53	8.92	0.58	4.24	9	7.33	1.17	3.50
	T	65	5.48	0.19	1.58	104	5.33	0.17	1.68	52	5.83	0.34	2.45	9	6.89	0.60	1.07
Subscapular	C	71	4.73	0.26	2.20	101	5.68	0.34	3.40	53	7.19	0.43	3.14	9	8.67	0.90	2.71
	T	64	3.64	0.12	0.94	104	3.48	0.13	1.32	52	3.73	0.17	1.21	9	5.44	0.69	2.06
Supra-illiac	C	71	3.30	0.29	2.41	101	4.29	0.36	3.59	53	6.26	0.56	4.11	9	7.11	0.99	2.96
	T	64	2.78	0.16	1.27	104	2.65	0.14	1.39	52	2.81	0.21	1.51	9	5.44	0.90	2.71
Calf	C	71	8.11	0.31	2.62	101	8.47	0.41	4.16	53	10.62	0.68	4.95	9	9.67	1.52	4.57
	T	65	7.09	0.28	2.26	104	6.14	0.21	2.19	52	7.40	0.47	3.36	9	10.11	1.35	4.04

Table 3.13 shows the mean value of anthropometric traits, taken on both Controlled and β-thalassaemic male samples for age group 0-5 years. It has been found that the

mean height in the Controlled sample is 106.72±0.73 cm. and in the β-thalassaemic sample 95.84±1.21 cm. The mean weight in the Controlled sample is found to be 16.62±0.34 kg. and that in the β-thalassaemic sample 12.72±0.43 kg. The mean chest girth (inhale) is 53.69±0.44 cm. in the Controlled sample and 51.95±0.57 cm. in the β-thalassaemic sample. The Controlled sample shows the mean value of chest girth (exhale) is 52.43±0.44 cm., whereas the mean value in the β-thalassaemic patients is 51.63±0.56 cm. In the Controlled and β-thalassaemic samples the mean humerous diameter is found to be 4.46±0.03 cm. and 3.90±0.04 cm. respectively. It is found that the mean bicondylar femur diameter is 3.73±0.03 cm. in the Controlled sample and the same in the β-thalassaemic sample is 4.48±0.07 cm. The mean biceps girth in the Controlled and β-thalassaemic sample is 15.66±0.17 cm. and 13.96±0.16 cm. respectively. So far as calf girth is concerned, it is found that the mean value in the Controlled sample is 20.72±0.23 cm. and the same in the β-thalassaemic sample 18.06±0.22 cm. In the Controlled and β-thalassaemic samples the biceps thickness is 3.51±0.20 mm. in the Controlled sample and 2.85±0.12 mm. in the β-thalassaemic sample. In case of triceps thickness the Controlled sample shows the mean value of 6.52±0.29 mm. whereas the β-thalassaemic sample shows the mean value of 5.48±0.19 cm. In subscapular thickness the mean value of 4.73±0.26 mm. is

found in case of the Controlled sample and 3.64±0.12 mm. in case of the β-thalassaemic sample. In the Controlled and the β-thalassaemic samples the mean value of supra-illiac thickness is 3.30±0.29 mm. and 2.78±0.16 mm. The mean calf thickness in the Controlled sample is 8.11±0.31 and that in the β-thalassaemic sample 7.09±0.28 mm.

In the age group 6-10 years the mean height of the Controlled sample is found to be 127.96±0.83 cm. and in the β-thalassaemic sample 116.55±1.19 cm. The mean body weight of the Controlled and the β-thalassaemic samples is found to be 24.56±0.55 kg. and 19.62±0.39 kg. respectively. So far as the chest girth (inhale) is concerned, the Controlled sample shows the mean value of 61.82±0.56 cm. and that in the β-thalassaemic patients is 58.48±0.44 cm. It is found that the mean chest girth (exhale) in the Controlled and β-Thalassaemic samples is 59.96±0.53 cm. and 57.02±0.44 cm. respectively. So far as humerous diameter is concerned, the mean value in the Controlled and β-thalassaemic samples is 5.00±0.03 cm. and 4.38±0.05 cm. respectively. The mean value of bicondylar femur diameter is 4.66±0.03 cm. in the Controlled sample and 4.98±0.05 cm. in the β-thalassaemic sample. So far as biceps girth is concerned, the Controlled sample shows the mean value of 17.35±0.24 cm. and in the β-thalassaemic sample 14.80±0.15 cm. The Controlled sample shows the mean value of calf girth is 24.08±0.24 cm., whereas

the β-thalassaemic sample shows that the mean value of the same measurement is 20.49±0.23 cm.

So far as biceps thickness is concerned, the mean value is found to be 3.92±0.25 mm in the Controlled sample and 2.79±0.13 mm. in the β-thalassaemic sample. The mean value of triceps thickness in the Controlled sample is 7.12±0.40 mm. and that in the β-thalassaemic sample is 5.33±0.17 mm. So far as subscapular thickness is concerned, the mean value in the Controlled sample is found to be 5.68±0.34 mm. and the same in the β-thalassaemic sample is 3.48±0.13 mm. The Controlled sample shows that the mean supra-illiac thickness is 4.29±0.36 mm. and the same is 2.65±0.14 mm. in the β-thalassaemic sample. The calf thickness in the Controlled sample shows the mean value of 8.47±0.41 mm. and the β-thalassaemic sample shows the mean value of 6.14±0.21 mm.

In the age group 11-15 years, it is found that the mean height of the Controlled sample is 153.68±1.75 cm. and that in the β-thalassaemic sample is 134.54±1.67 cm. The mean body weight is found to be 42.34±1.54 kg. in the Controlled group and 26.97±0.86 kg. in the β-thalassaemic sample. It is found that the mean chest girth (inhale) is 74.54±1.03 cm. in the Controlled sample and 64.17±0.79 cm. in the β-thalassaemic sample. The Controlled sample shows that the mean value of chest girth (inhale) is 72.08±0.97 cm., whereas the same value in the β-thalassaemic sample is 62.36±0.77 cm. The mean

humerous diameter in the Controlled sample is 6.03±0.08 cm. and that in the β-thalassaemic sample is 4.98±0.08 cm. The Controlled and the β-thalassaemic samples show the mean value of bicondylar femur diameter is 5.05±0.05 cm. and 5.38±0.09 cm. respectively. The mean biceps girth is 21.22±0.42 cm. in the Controlled sample and 15.72±0.25 cm. in the β-thalassaemic sample. The Controlled sample shows that the mean value of calf girth is 29.87±0.46 cm., whereas the mean value of calf girth in the β-thalassaemic patients is 21.94±0.35 cm.

The mean biceps thickness in the Controlled sample is 4.58±0.36 mm. and 3.06±0.21 mm. in the β-thalassaemic sample. The mean triceps thickness is found to be 8.92±0.58 mm. in the Controlled sample and 5.83±0.34 mm. in the β-thalassaemic sample. The mean subscapular thickness is 7.19±043 mm. and 3.73±0.17 mm. in the Controlled and β-thalassaemic samples respectively. The mean supra-illiac thickness is 6.26±0.56 mm. in the Controlled group and 2.81±0.21 mm. in the β-thalassaemic sample. The mean calf thickness is found to be 10.62±0.68 mm. in the Controlled group and 7.40±0.47 mm. in the β-thalassaemic sample.

In the age group 16-20 years, it is found that the mean height in the Controlled group is 169.54±1.61 cm. and that in the β-thalassaemic sample is 148.94±3.68 cm. The mean body weight in the Controlled and β-thalassaemic group is found to

be 55.39±3.54 kg. and 41.71±2.72 kg. respectively. The mean chest girth (inhale) in the Controlled group is 82.61±2.44 cm. and that in the β-thalassaemic sample is 75.57±2.07 cm. The Controlled sample shows that the mean chest girth (exhale) is 79.61±2.55 cm. and in the β-thalassaemic sample the mean value is found to be 73.12±1.85 cm. In the Controlled and β-thalassaemic samples the mean humerous diameter is found to be 6.06±0.14 cm. and 5.89±0.13 cm. respectively. The mean bicondylar femur diameter in the Controlled and β-thalassaemic sample is found to be 5.47±0.17 cm. and 6.16±0.27 cm. respectively. The mean biceps girth is 23.52±0.94 cm. and 19.02±0.58 cm. in the Controlled and β-thalassaemic samples respectively. The mean calf girth in the Controlled group is 33.72±1.08 cm. and the same in the β-thalassaemic group is 26.60±1.05 cm.

The Controlled and β-thalassaemic samples show the mean biceps thickness as 3.78±0.73 mm. and 2.78±0.31 mm. respectively. The Controlled group is having the mean triceps thickness as 7.33±1.17 mm., whereas the same in the β-thalassaemic group is 6.89±0.60 mm. The subscapular thickness in the Controlled group is 8.67±0.90 mm. and 5.44±0.69 mm. in the β-thalassaemic sample. The mean supra-illiac thickness in the Controlled group is 7.11±0.99 mm. and that in the β-thalassaemic group is 5.44±0.90 mm. The mean calf thickness

in the Controlled and β-thalassaemic samples is 9.67±1.52 mm. and 10.11±1.35 mm. respectively.

From the above description of anthropometric measurements in the β-thalassaemic patients and the Controlled sample, it is seen that in all the anthropometric measurements, considered in the present study, the mean value is lower in the β-thalassaemic sample than in the Controlled sample, and it holds good for all the 4 age-groups considered for the present purpose. The only exception is noted in case of bicondylar femur diameter, which shows higher mean value in the β-thalassaemic group in comparison to the Controlled group and it is true for all 4 age group in the present study.

Table 3.14

t-value for the Difference in Respect of Various Anthropometric Traits Between Controlled and Thalassaemic Males

Age Group	Height	Weight	Chest Girth (inhale)	Chest Girth (exhale)	Humerous Diameter	Bicondylar Femur Dia.	Bicep Girth	Calf Girth	Biceps	Triceps	Subcapular	Supra-illiac	Calf
0 - 5	7.70*	7.11*	2.42*	1.12	5.27*	9.30*	7.28*	8.36*	2.83*	2.95*	4.20*	1.57	2.44*
6 -10	7.82*	7.26*	4.65*	4.17*	10.63*	5.49*	9.01*	10.80*	4.01*	8.38*	6.04*	4.35*	5.06*
11-15	7.91*	8.71*	7.98*	7.85*	9.88*	3.35*	11.25*	13.72*	3.65*	4.60*	7.48*	5.77*	3.90*
16-20	14.37*	8.46*	6.61*	5.78*	2.50*	6.10*	11.50*	13.24*	3.59*	1.07	7.97*	3.51*	0.61

* Significant at 5% level of probability.

Table 3.14 shows the result of the t-tests carried out in respect of various anthropometric traits between the

Male Sample

Growth Curve for Height of Controlled & Thalassaemic Boys

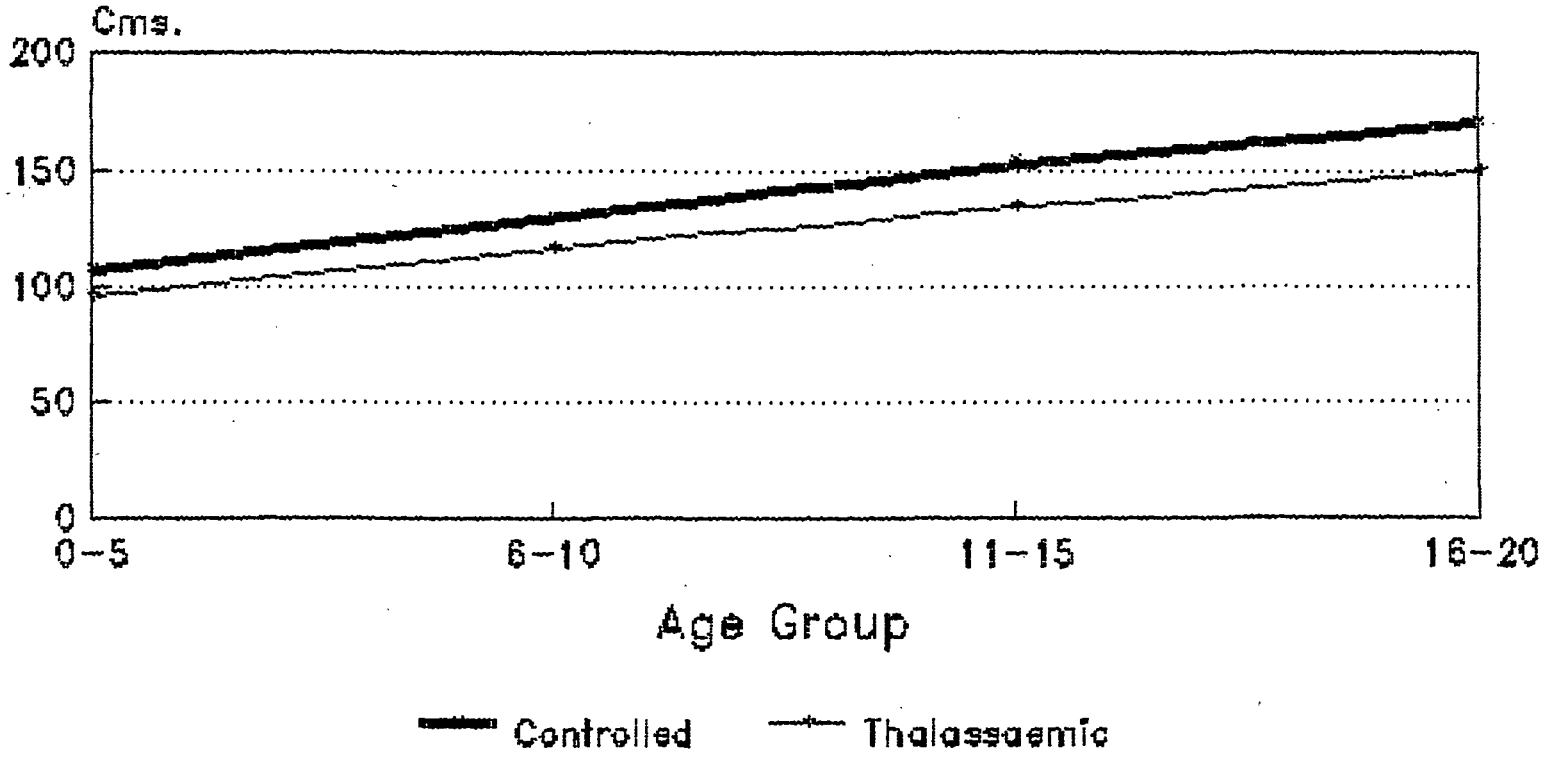

Fig. 14

Controlled and β-thalassaemic samples. It is seen that the height is significantly greater in the Controlled group in comparison to the β-thalassaemic sample, and it is true for all age groups. Similar findings have been noticed for weight, chest girth (inhale), humerous diameter, biceps girth, calf girth, biceps, subscapular and supra-illiac. This is the general trend. However, a few exceptions have been noticed such as chest girth (exhale) in the age group 0-5 years, no significant difference is found between the β-thalassaemic group and Controlled sample. Similarly, in the age-group 16-20 years in case of triceps thickness and calf thickness no statistically significant differences in mean values between the Controlled and β-thalassaemic samples have been found. Further a point to be noted here is that so far as bicondylar femur diameter is concerned, the β-thalassaemic patients have shown the greater mean value in all four age groups in comparison to those in the Controlled group.

Taking into consideration all anthropometric traits in both male and female, it can be suggested that body dimensions are by and large significantly greater in the Controlled sample, i.e., among normal people, than in the β-thalassaemic patients.

Figure-14 shows the growth curve for height between the Controlled and β-thalassaemic boys. It is found that in all

Male Sample

Growth Curve for Weight of Controlled & Thalassaemic Boys

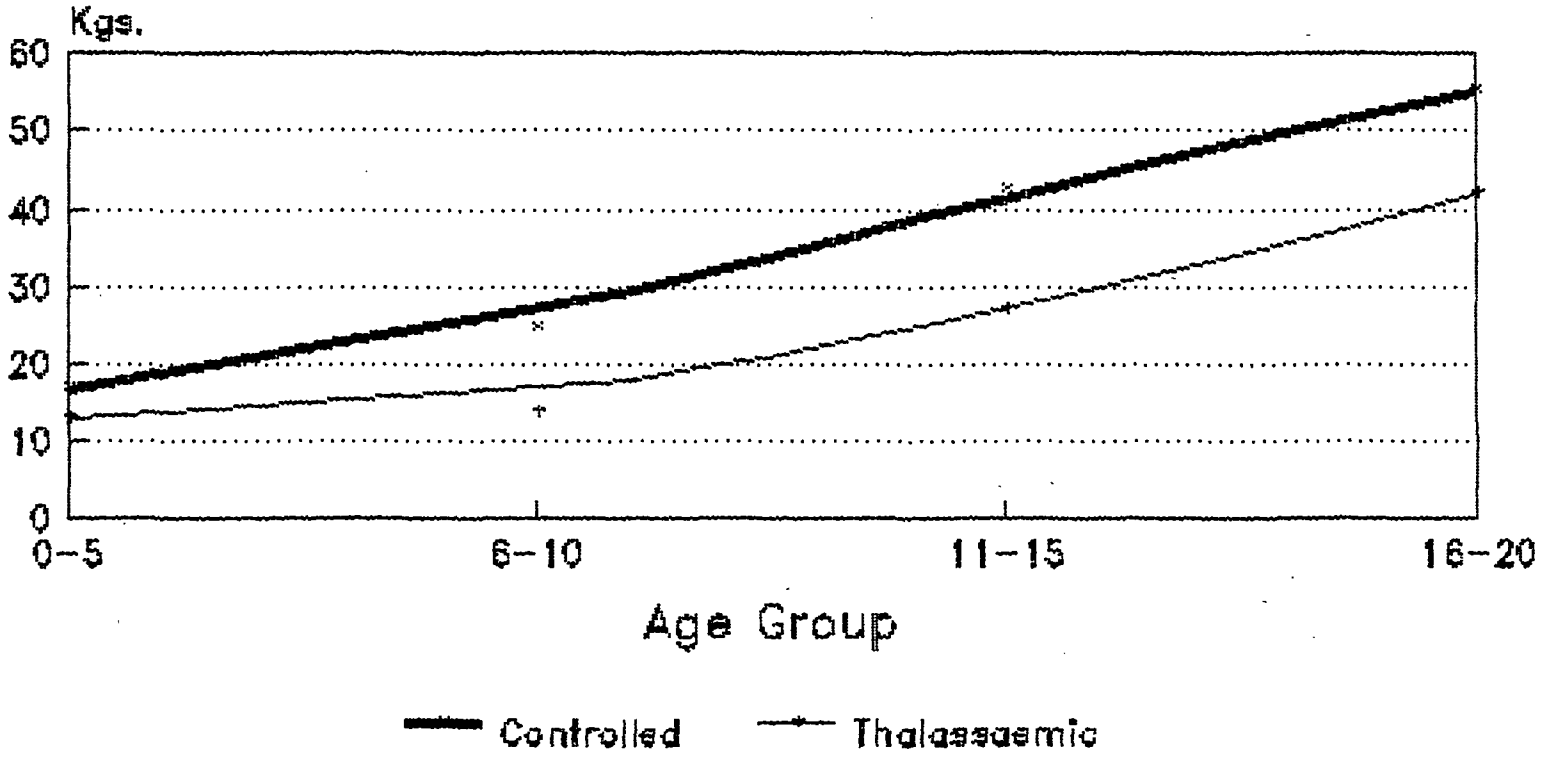

Fig. 15

Male Sample

Growth Curve for Chest Girth of Controlled & Thalassaemic Boys

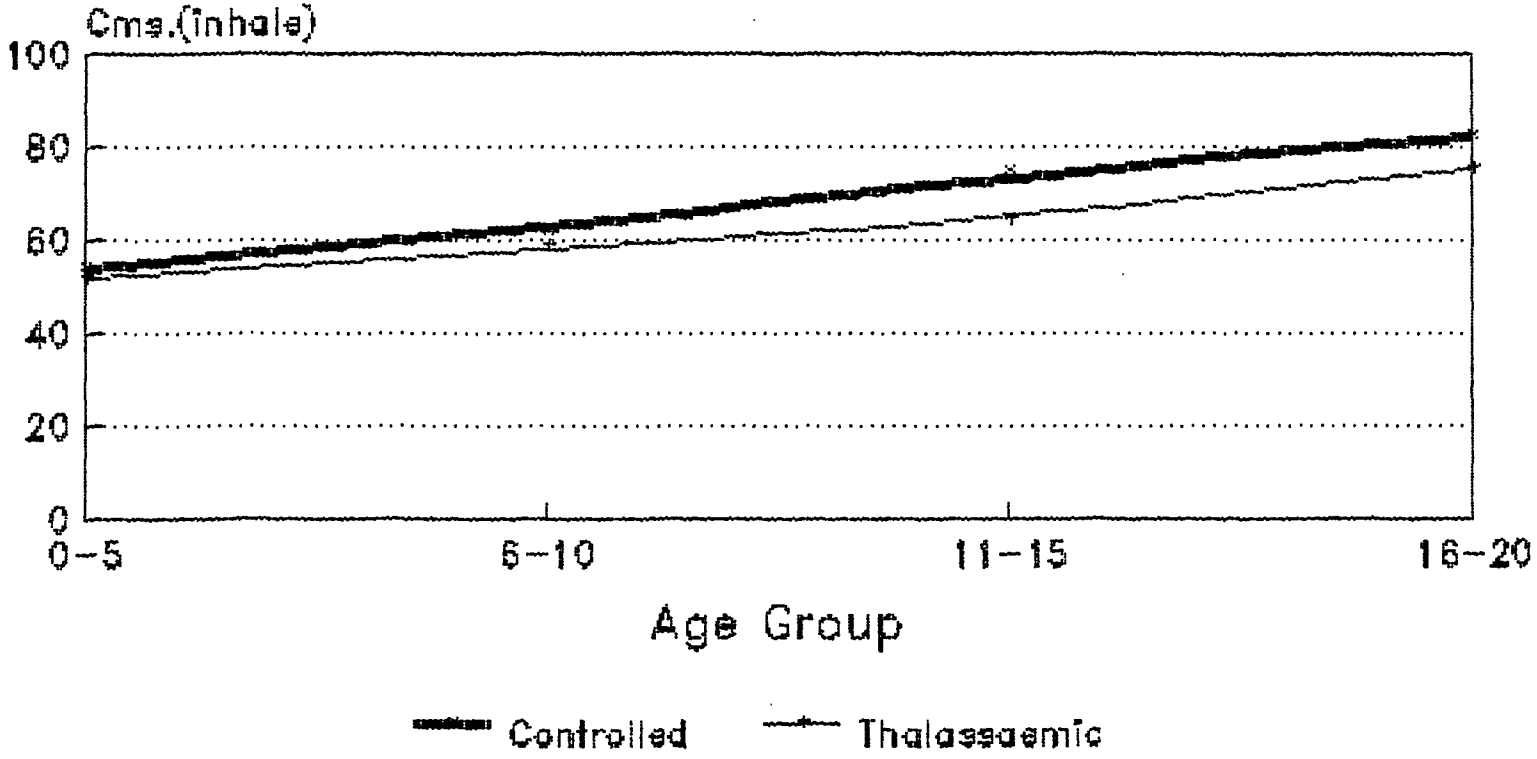

Fig. 16

Male Sample

Growth Curve for Chest Girth of Controlled & Thalassaemic Boys

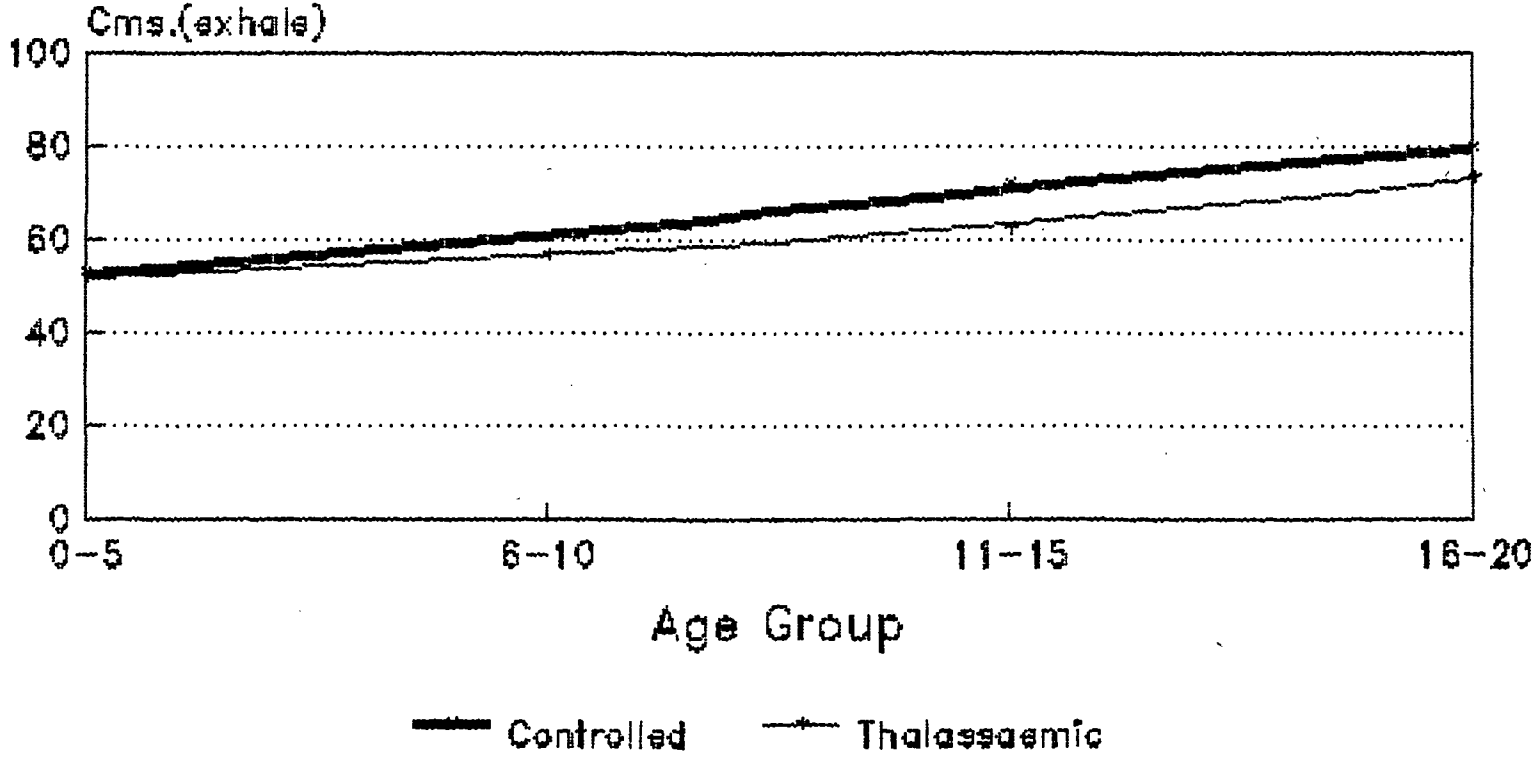

Fig. 17

Male Sample

Growth Curve for Humerous Diameter of Controlled & Thalassaemic Boys

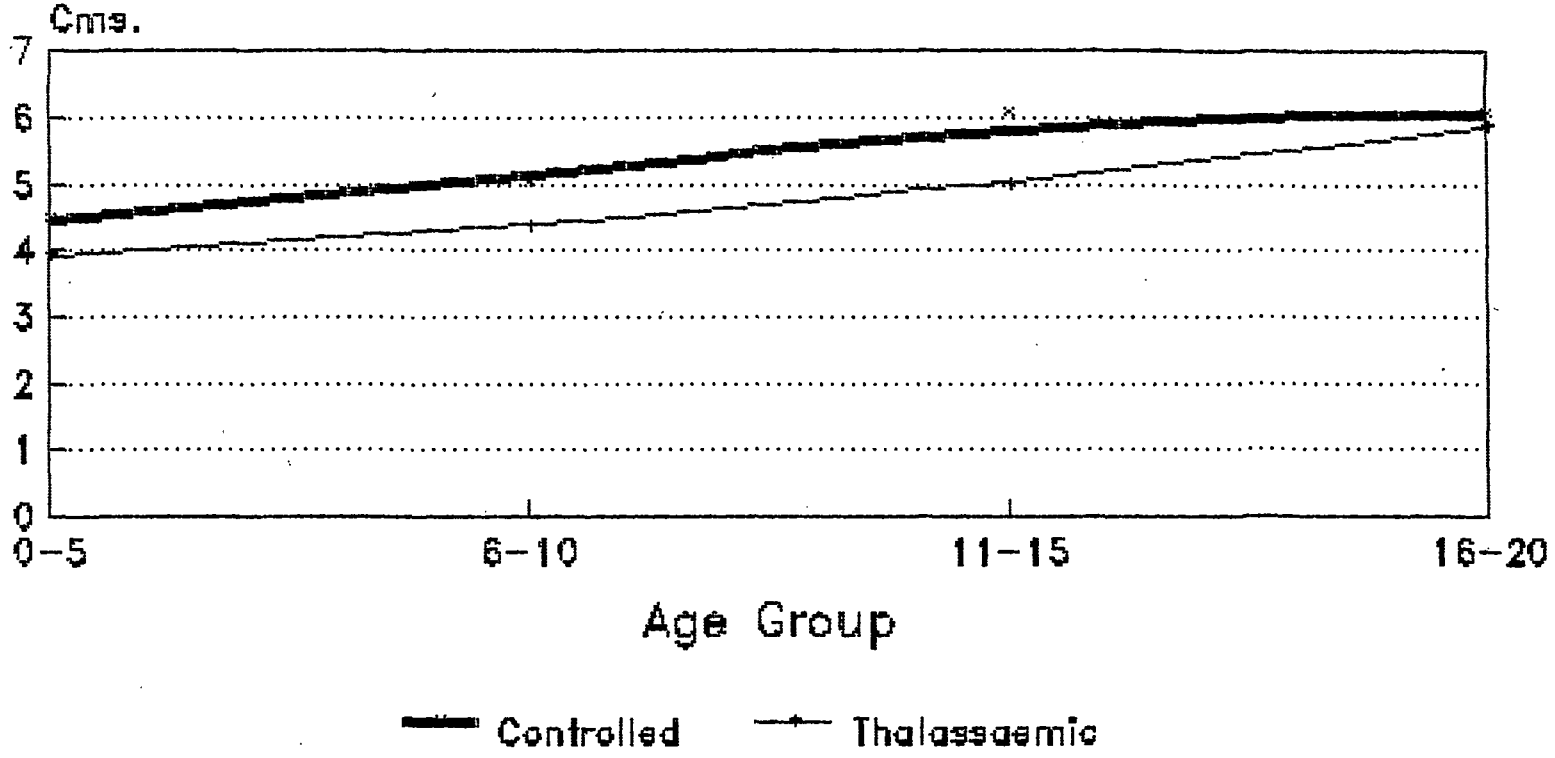

Fig. 18

Male Sample

Growth Curve for Bicondylar Femur Dia. of Controlled & Thalassaemic Boys

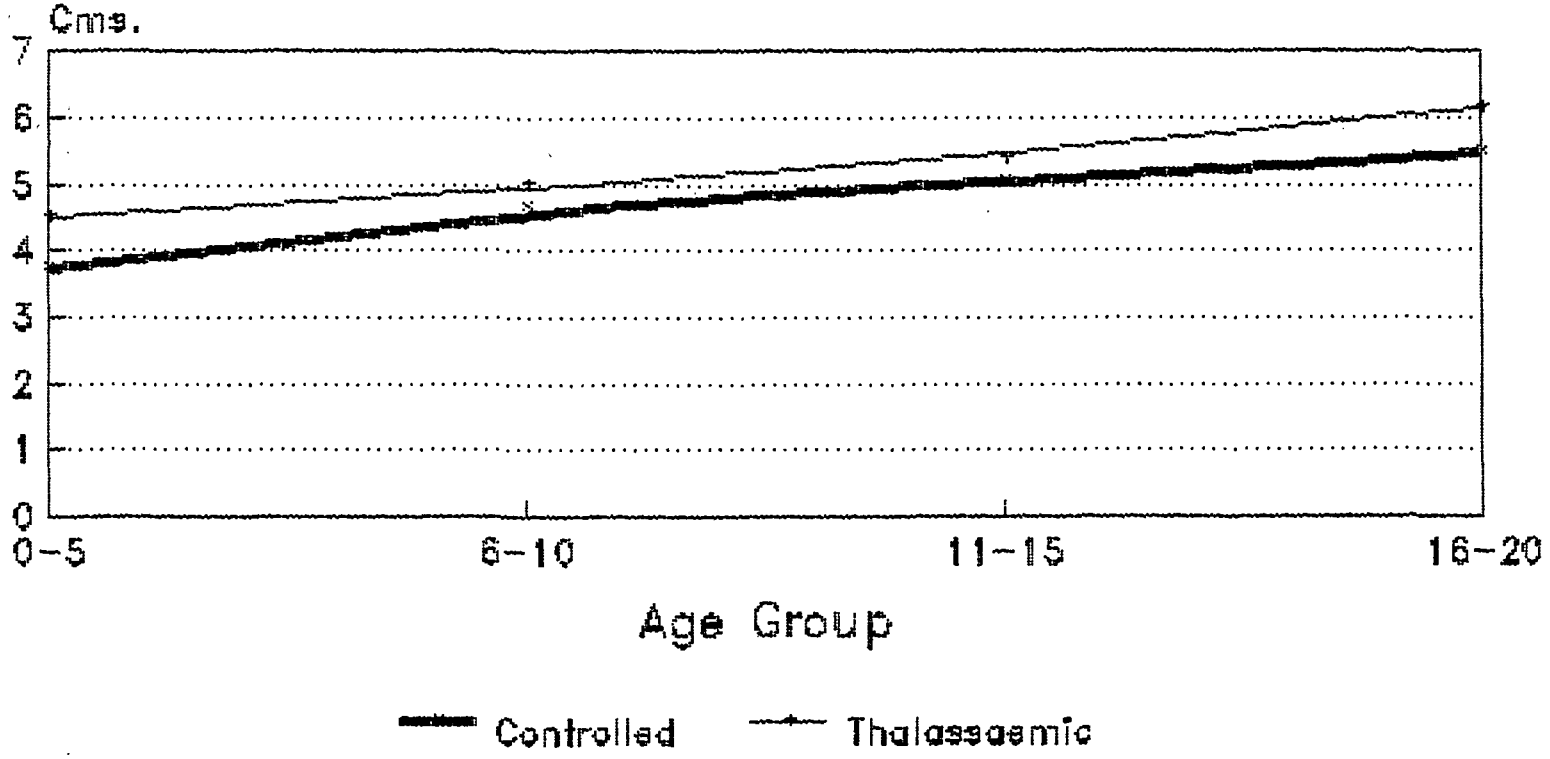

Fig. 19

age-groups the height is lower in the β-thalassaemic boys than in the Controlled sample.

Figure-15 shows the growth curve for weight. It shows that the growth curve is lower in the β-thalassaemic boys in comparison to the Controlled sample. It further shows that the rate of growth in the Controlled sample further accelerates in the age-group 6-10 and continues to do so till age group 16-20 years.

Figure-16 shows the growth curve in respect of chest girth (inhale). It is seen that the normal boys always have higher value in all age-groups in comparison to the β-thalassaemic boys.

Figure-17 shows the growth curve for chest girth (exhale) between the Controlled and β-thalassaemic boys and is by and large same as in figure-16.

Figure-18 shows the growth curve for humerus diameter between the Controlled and β-thalassaemic boys. It is found that the growth curve is lower in the thalassaemic boys in comparison to the Controlled sample. The curve further shows that the difference between them reduces with the advancement of age.

Figure-19 shows the growth curve for bicondylar femur diameter. It shows that the Controlled sample have lower values in comparison to the β-thalassaemic boys for all age-groups.

Male Sample

Growth Curve for Bicep Girth of Controlled & Thalassaemic Boys

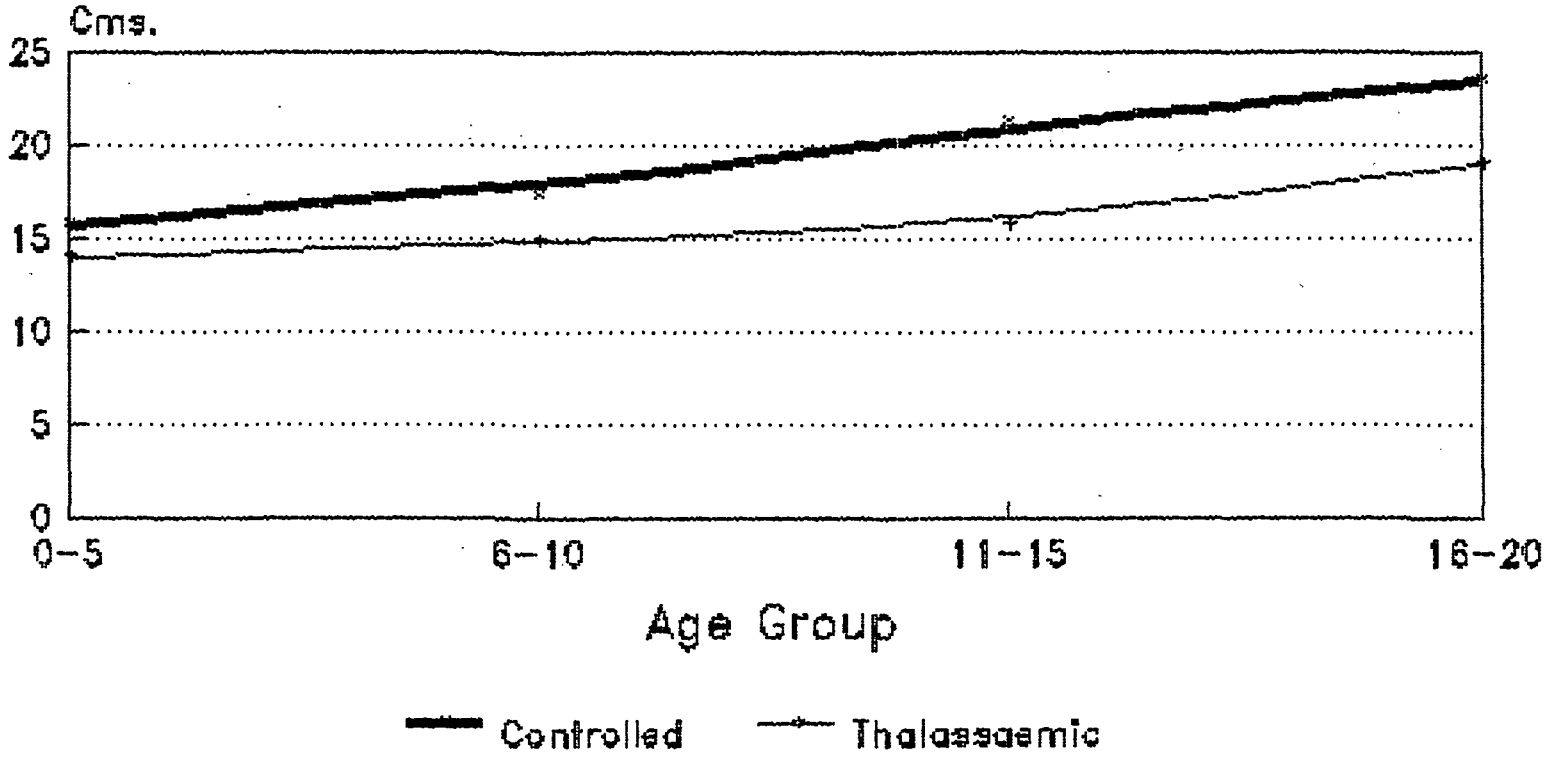

Fig. 20

Male Sample

Growth Curve for Calf Girth of Controlled & Thalassaemic Boys

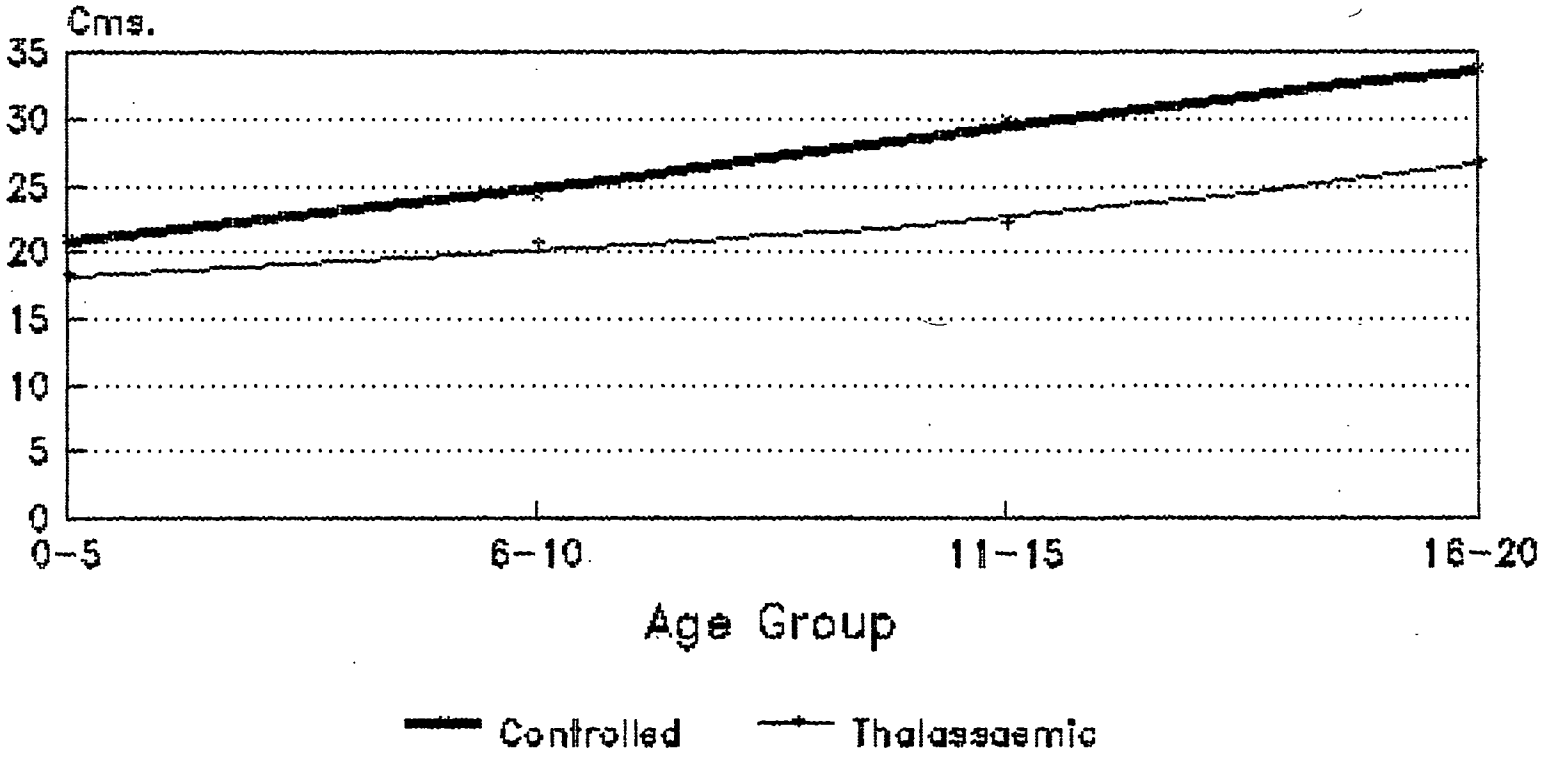

Fig. 21

Male Sample

Growth Curve for Skinfold Thickness at Biceps of Con. & Tha. Boys

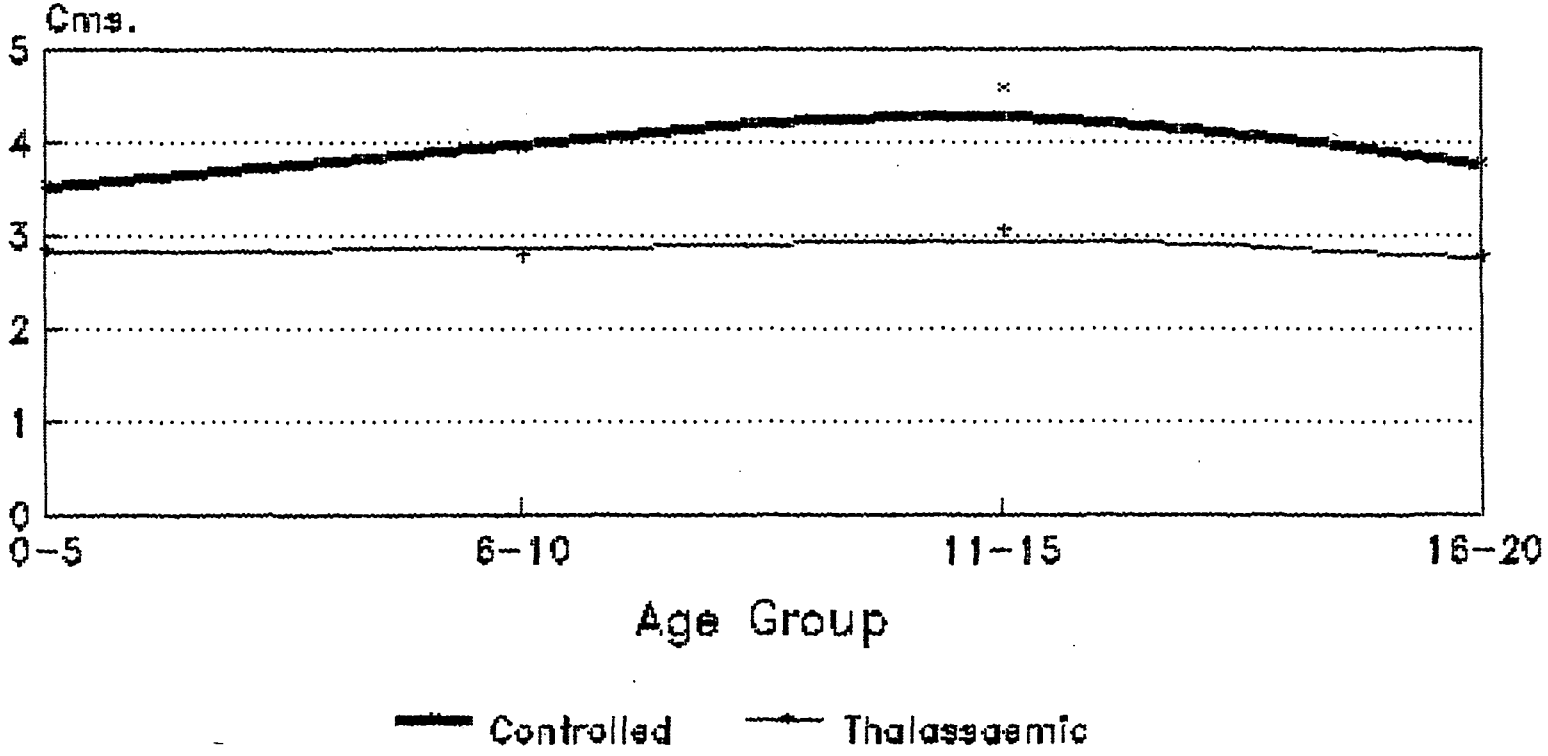

Fig. 22

Male Sample

Growth Curve for Skinfold Thickness at Triceps of Con. & Thalassaemic Boy

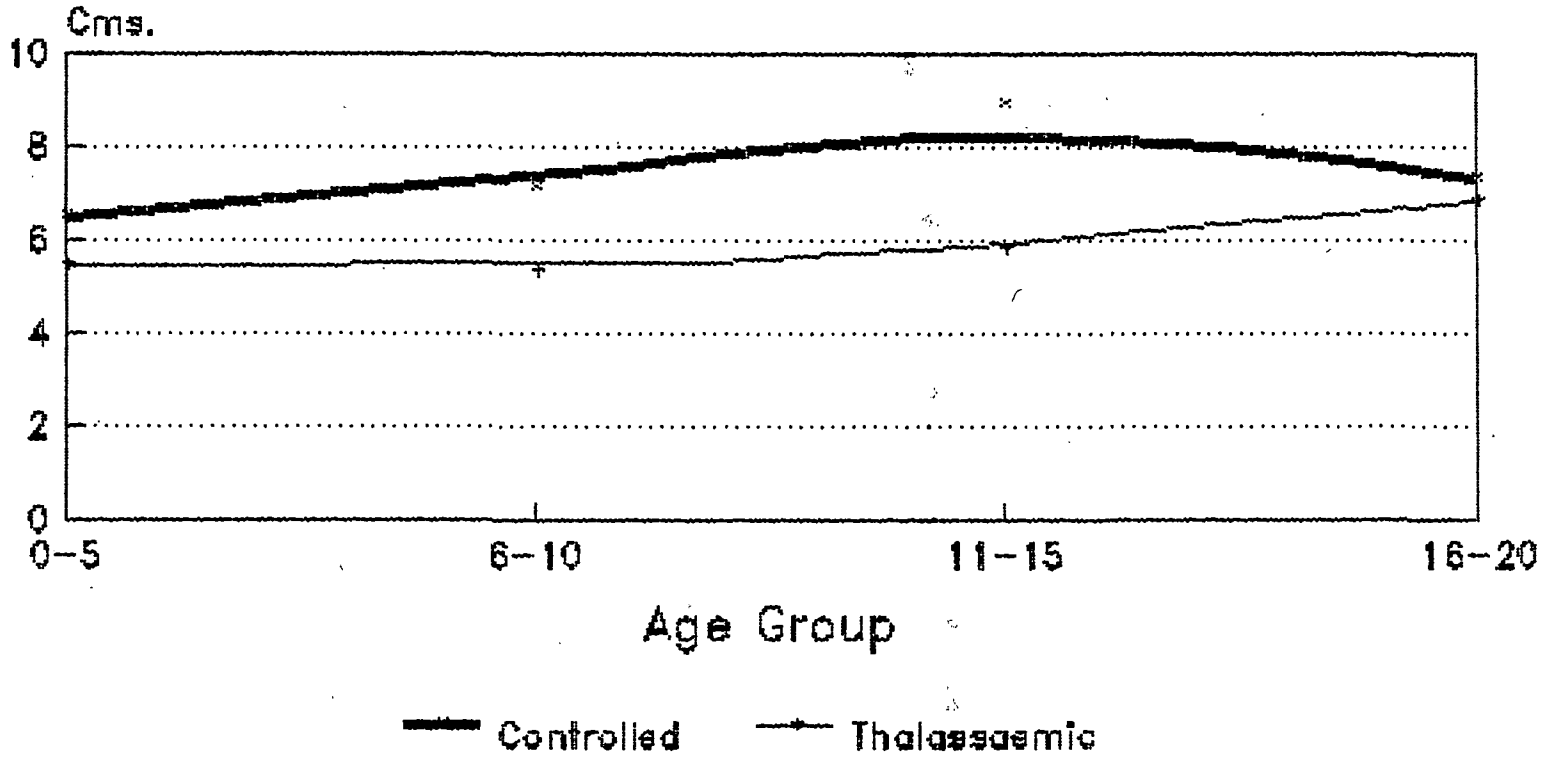

Fig. 23

Male Sample

Growth Curve for Skinfold Thickness at Subscapular of Con. & Tha. Boys.

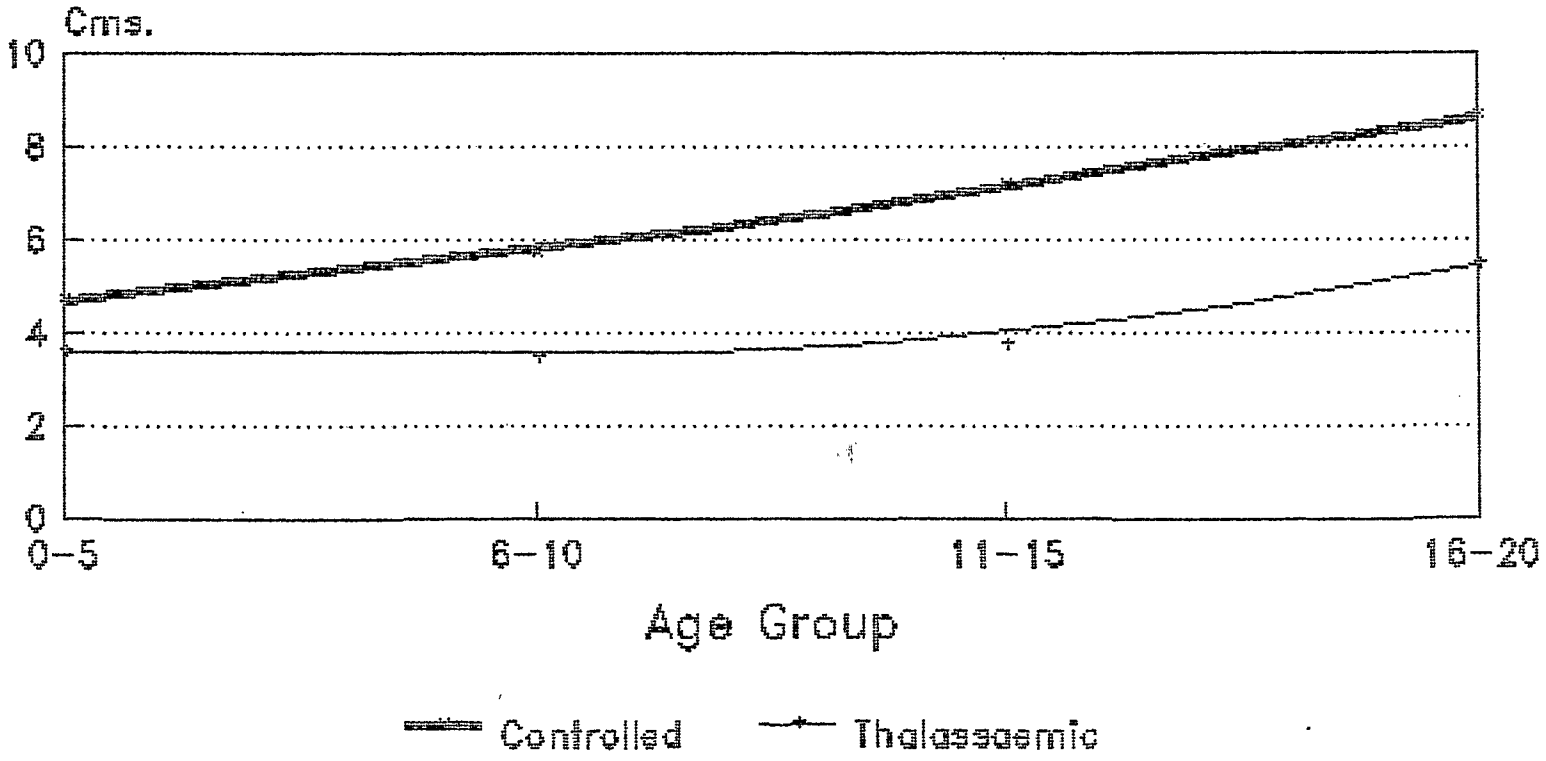

Fig. 24

Figure-20 shows the growth curve in respect of biceps girth between the Controlled and β-thalassaemic boys. It is seen that the normal boys always have the higher growth rate than the β-thalassaemic boys and the growth rate further accelerates from 6-10 years of age.

Figure-21 shows the growth curve in respect of calf girth. It is seen that β-thalassaemic boys have a lesser value in comparison to the Controlled sample and the difference between them further widens with the advancement of age.

Figure-22 shows the growth curve for biceps between the Controlled and β-thalassaemic boys. The curve indicates that the β-thalassaemic boys have almost the same value from the lowest to the highest age groups and is certainly lower than that in the Controlled sample. However, the difference between them starts decreasing from the age group 10-15 years and onwards.

Figure-23 shows the growth curve in respect of triceps which shows that the difference between the Controlled sample and β-thalassaemic boys. The difference between them broadens from 6-10 years of age and then it slowly drops from 11-15 years and becomes almost minimal in 16-20 years.

Figure-24 shows the growth curve in respect of subscapular thickness between the Controlled and β-thalassaemic boys. From the very beginning the β-thalassaemic

Male Sample

Growth Curve for Skinfold Thickness at Supra-Illiac of Con. & Tha. Boys

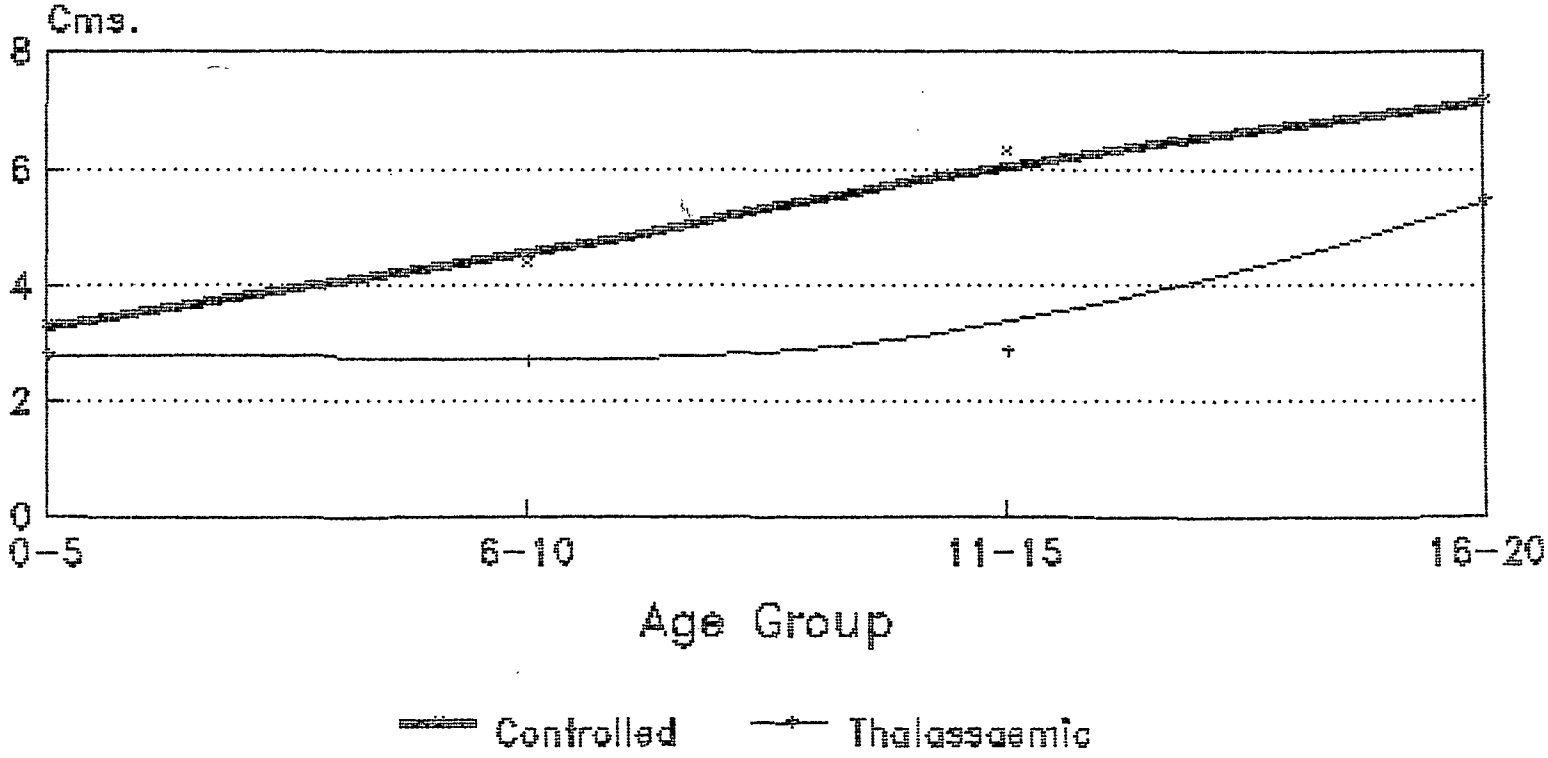

Fig. 25

Male Sample

Growth Curve for Skinfold Thickness at Calf of Controlled & Thalassaemic Boys

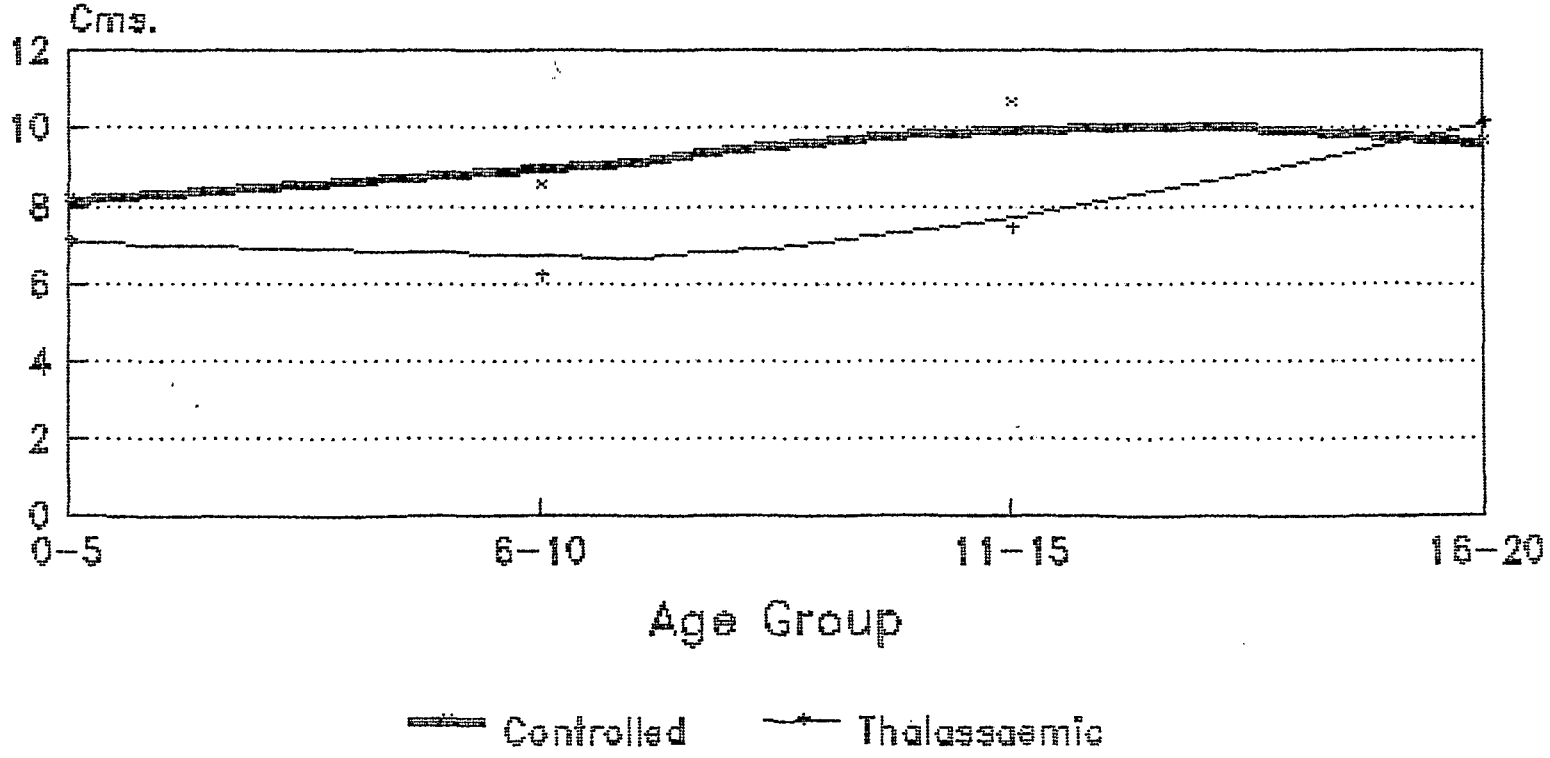

Fig. 26

boys have a lesser value in comparison to the Controlled sample and the difference broadens from the age-group of 6-10 years.

Figure-25 shows the growth curve in respect of supra-illiac thickness between the Controlled and β-thalassaemic boys. The curve shows that the Controlled sample in all ages have a higher value in comparison to the thalassaemic boys. The difference between them broadens from 6-10 years and the difference reduces in the age-group 16-20 years.

Figure-26 shows the growth curve in respect of calf thickness between the Controlled and the β-thalassaemic boys. It is seen that initially the normal boys have higher value than the β-thalassaemic boys and the differences between them keeps on increasing till the age group 11-15 years, and thereafter, the growth curve of the β-thalassaemic boys approaches that of the normal boys and then it is slightly more in the thalassaemic boys in comparison to the normal boys in the age group of 16-20 years.

Table 3.15 shows the height for age, weight for age, and weight for height in both the Controlled and β-thalassaemic girls for all 4 age-groups, i.e., upto 5 years, 6-10 years, 11-15 years, and 16-20 years. So far as the height for age in the age group upto 5 years is concerned, it is seen that the mean value in the Controlled group is

111.76±0.72 and that in the β-thalassaemic group 100.17±0.77 cm. The same in the age-group 6-10 years is 103.60±0.92 cm. in the Controlled group and 98.13±1.08 cm. in the β-thalassaemic sample. For the age-group 11-15 years, it is seen that height for age is 102.67±0.90 in the Controlled group and 95.01±0.73 in the β-thalassaemic group. In the age-group 16-20 years the same is 102.18±1.16 cm. in the Controlled group and 99.20±2.29 cm. in the β-thalassaemic group. The point to be noted here is that height for age is always greater in the Controlled group in comparison to the β-thalassaemic group.

Table 3.15

Anthropometric Indices for Females

Traits		0-5				6-10				11-15				16-20			
		N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.
Height for Age	C	27	111.76	0.72	3.74	40	103.60	0.92	5.85	24	102.67	0.90	4.40	6	102.18	1.16	2.85
	T	24	100.17	0.77	3.79	33	98.13	1.08	6.13	27	95.01	0.73	3.50	6	99.20	2.29	5.62
Weight for Age	C	27	119.05	2.43	12.62	40	112.31	0.90	4.04	24	108.00	4.03	19.72	6	111.56	7.08	17.34
	T	24	100.36	3.57	17.51	33	95.87	3.07	17.10	27	90.46	2.36	11.30	6	77.36	7.93	19.42
Weight for Height	C	27	102.50	2.35	12.23	40	104.97	2.86	18.09	24	102.92	3.08	15.10	6	107.97	5.58	13.66
	T	24	101.60	3.50	17.14	33	100.04	2.16	12.24	27	104.90	1.92	9.23	6	79.82	6.89	16.88

When weight for age is calculated, it is seen that in age-group upto 5 years, the mean is 119.05±2.43 kg. in the Controlled group and 100.36±3.57 in the β-thalassaemic group.

In case of age-group 6-10 years, the same is 112.31±0.90 in the Controlled group and 95.87±3.07 in the β-thalassaemic group. In the age group 11-15 years the mean weight for age is 108.00±4.03 and 90.46±2.36 in the Controlled and β-thalassaemic group respectively. In the age group 16-20 years, the mean weight for age is 111.56±7.08 and 77.36±7.93 in the Controlled and β-thalassaemic groups. It is found that in all age-groups weight for age is always greater in the Controlled group in comparison to the β-thalassaemic group.

When weight for height is examined, it is found that in age group upto 5 years, 6-10 years, 11-15 years, and 16-20 years the mean values in the Controlled group is 102.50±2.35, 104.97±2.86, 102.92±3.08, and 107.97±5.58 respectively. Similarly in the β-thalassaemic group the same is 101.60±3.50, 100.04±2.16, 104.90±1.92 and 79.82±6.89 for the age groups upto 5 years, 6-10 years, 11-15 years, and 16-20 years respectively. It may be seen that weight for height in each of this four age groups is greater in the Controlled sample than in the β-thalassaemic sample. The only exception is in the age 11-15 years where the weight for height is slightly more in the β-thalassaemic group.

Table 3.16

Anthropometric Indices for Males

Traits		0-5				6-10				11-15				16-20			
		N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.	N	Mean	± S.E.	S.D.
Height for Age	C	36	108.71	0.78	4.66	65	107.02	0.55	4.47	41	105.79	0.81	5.21	8	103.94	0.83	2.35
	T	32	100.00	1.17	6.60	59	96.82	0.79	6.08	42	93.84	0.73	4.71	8	90.89	1.38	3.90
Weight for Age	C	36	118.07	2.78	16.69	65	121.36	2.48	20.01	41	133.29	3.85	24.67	8	123.93	7.67	21.70
	T	32	98.85	3.19	18.02	59	97.77	2.00	15.39	42	90.66	2.29	14.81	8	90.52	4.79	13.55
Weight for Height	C	36	108.53	2.05	12.30	65	107.63	1.95	15.69	41	116.97	2.98	19.09	8	115.29	5.98	16.91
	T	32	103.22	2.08	11.79	59	105.70	1.57	12.08	42	106.36	2.03	13.18	8	116.33	7.65	21.65

Table 3.16 shows the height for age, weight for age, and weight for height in males. It shows height for age in the age groups upto 5 years, 6-10 years, 11-15 years, and 16-20 years. The mean height for age in the β-thalassaemic group is 100.00±1.17, 96.82±0.79, 93.84±0.73, and 90.89±1.38, respectively. The same for age groups upto 5 years, 6-10 years, 11-15 years, and 16-20 years in the Controlled group is 108.71±0.78, 107.02±0.55, 105.79±0.81 and 103.94±0.83 respectively. The weight for age in the age groups upto 5 years, 6-10 years, 11-15 years, 16-20 years in the β-thalassaemic patients is 98.85±3.19, 97.77±2.00, 90.66±2.29 and 90.52±4.79 respectively and the same in the Controlled sample is 118.07±2.78, 121.36±2.48, 133.29±3.85 and 123.93±7.67 respectively.

So far as the weight for height is concerned, in the age groups upto 5 years, 6-10 years, 11-15 years and 16-20 years in the β-thalassaemic group the mean is found to be 103.22±2.08, 105.70±1.57, 106.36±2.03 and 116.33±7.65 respectively, and in the Controlled group the same is 108.53±2.05, 107.63±1.95, 116.97±2.98 and 115.29±5.98 respectively. So far as the height for age, weight for age, and weight for height in males are concerned, it is found that in all age-groups these values are greater in the Controlled group than in the β-thalassaemic group, excepting the weight for height in the age group 16-20 years in which the mean value in the β-thalassaemic group is marginally greater than that in the Controlled group.

When the estimations are made for the height for age, weight for age, and weight for height it is seen that in case of both males and females the Controlled group shows greater value in comparison to the β-thalassaemic group. It shows that the growth rate in the β-thalassaemic patients is certainly slower and/or retarded in comparison to their normal counterparts and it holds good for each age. However, we shall discuss this point further at an appropriate place.

Table 3.17

t-test Between Thalassaemic and Controlled Samples

Traits		Age Groups (in years)			
		0-5	6-10	11-15	16-20
Height for Age	M	6.19*	10.60*	10.96*	8.10*
	F	10.99*	3.85*	6.61*	1.16
Weight for Age	M	4.54*	7.40*	9.51*	3.69*
	F	4.33*	3.24*	3.76*	3.22*
Weight for Height	M	1.82	0.77	2.94*	0.11
	F	0.21	1.38	0.55	3.17*

* significant at 5% probability

Table 3.17 shows the results of 't'-test between the β-thalassaemic patients and normal individuals in respect of height for age, weight for age, and weight for height. In case of height for age it is found that for all age groups there are significant differences between the β-thalassaemic and normal males. This is true for the females of all age-groups, excepting for the age group 16-20 years which shows no significant difference between the β-thalassaemic and normal females. In respect of weight for age significant differences exist between the β-thalassaemic and normal individuals, and it holds good for both sexes. In respect of weight for height no significant difference is noticed between the β-thalassaemic and normal individuals, excepting

Female Sample

Growth Curve for Height for Age of Controlled & Thalassaemic Girls

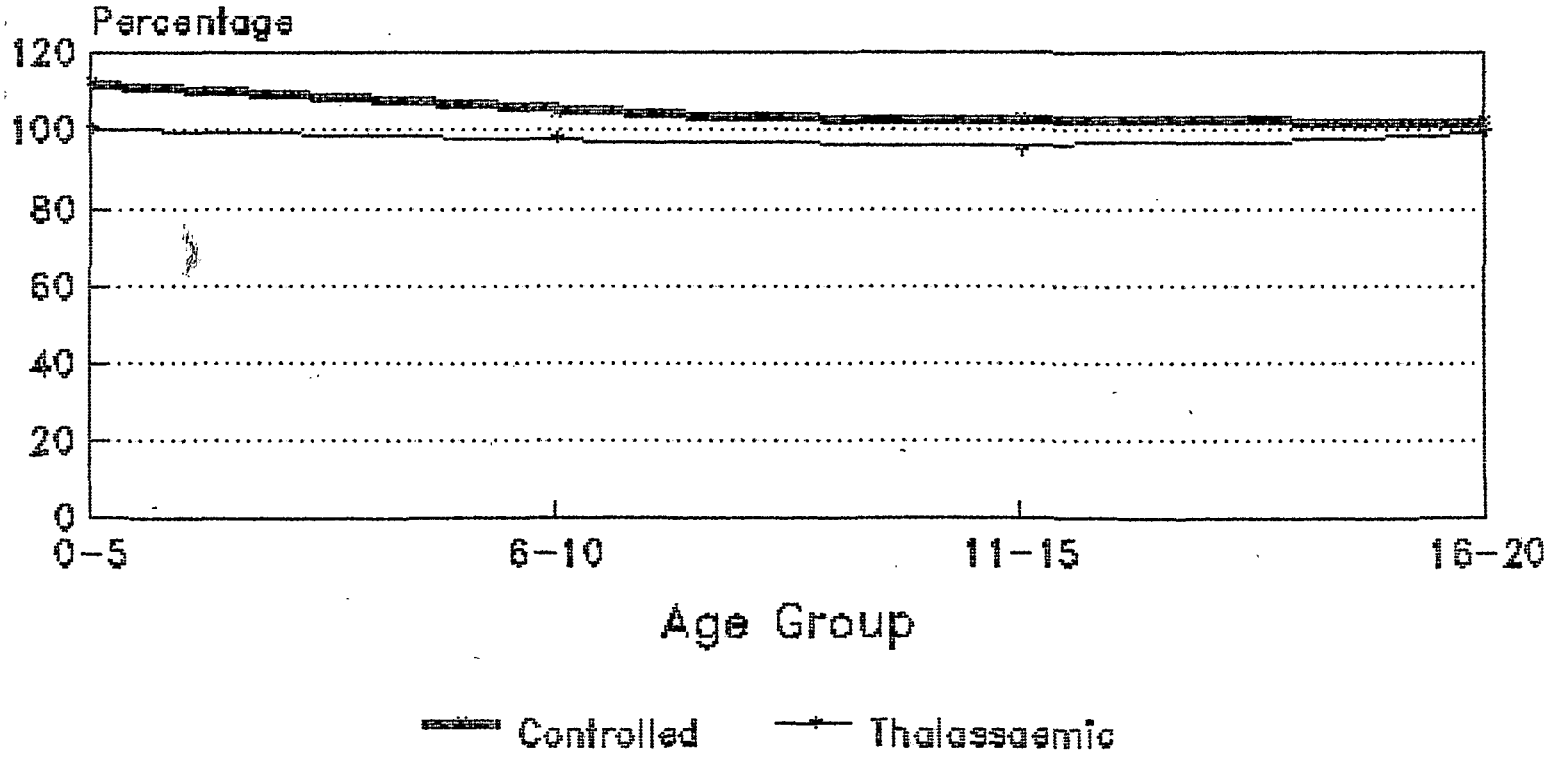

Fig. 27

Female Sample

Growth Curve for Weight for Age of Controlled & Thalassaemic Girls

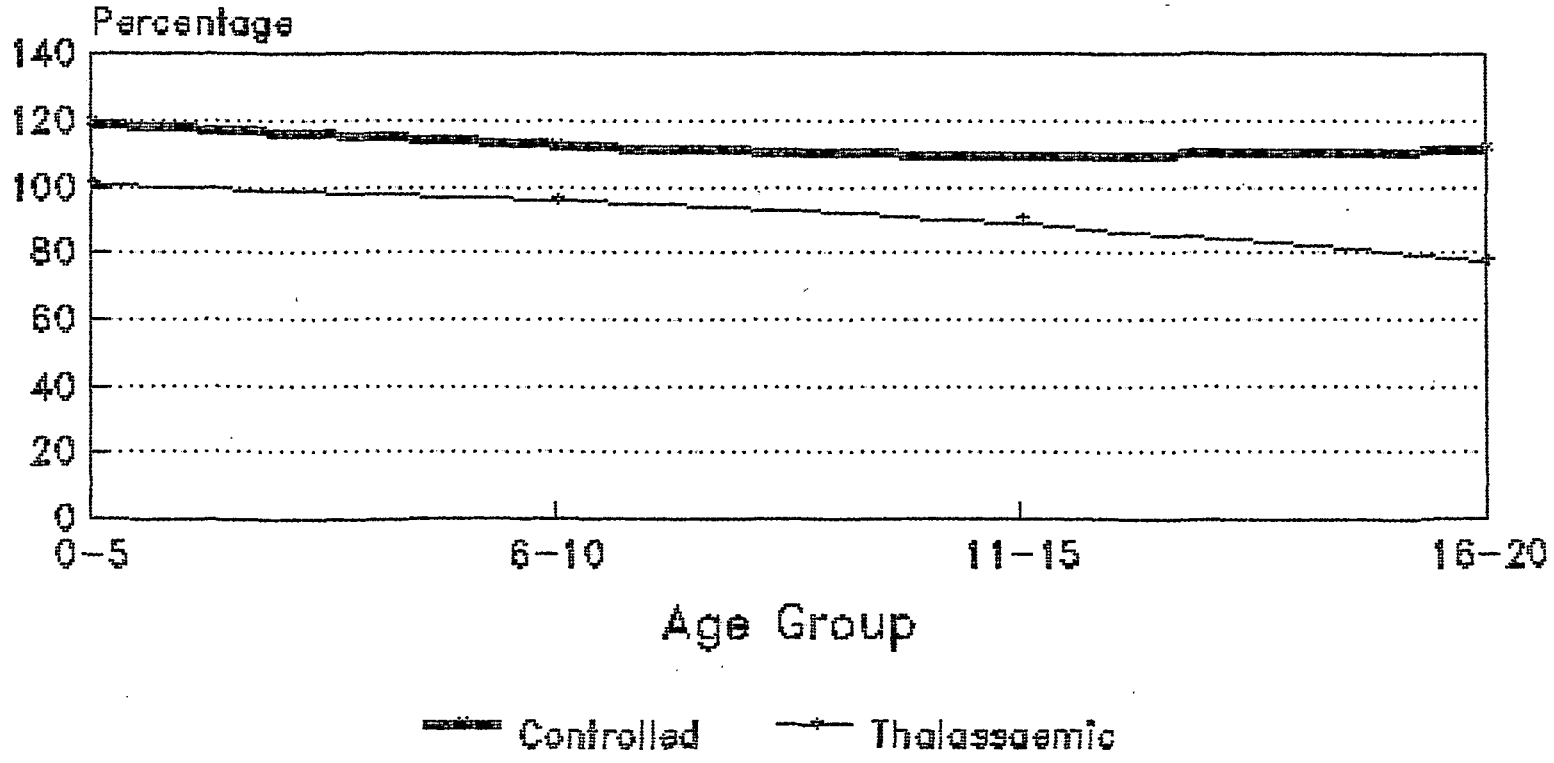

Fig. 28

Female Sample

Growth Curve for Weight for Height of Controlled & Thalassaemic Girls

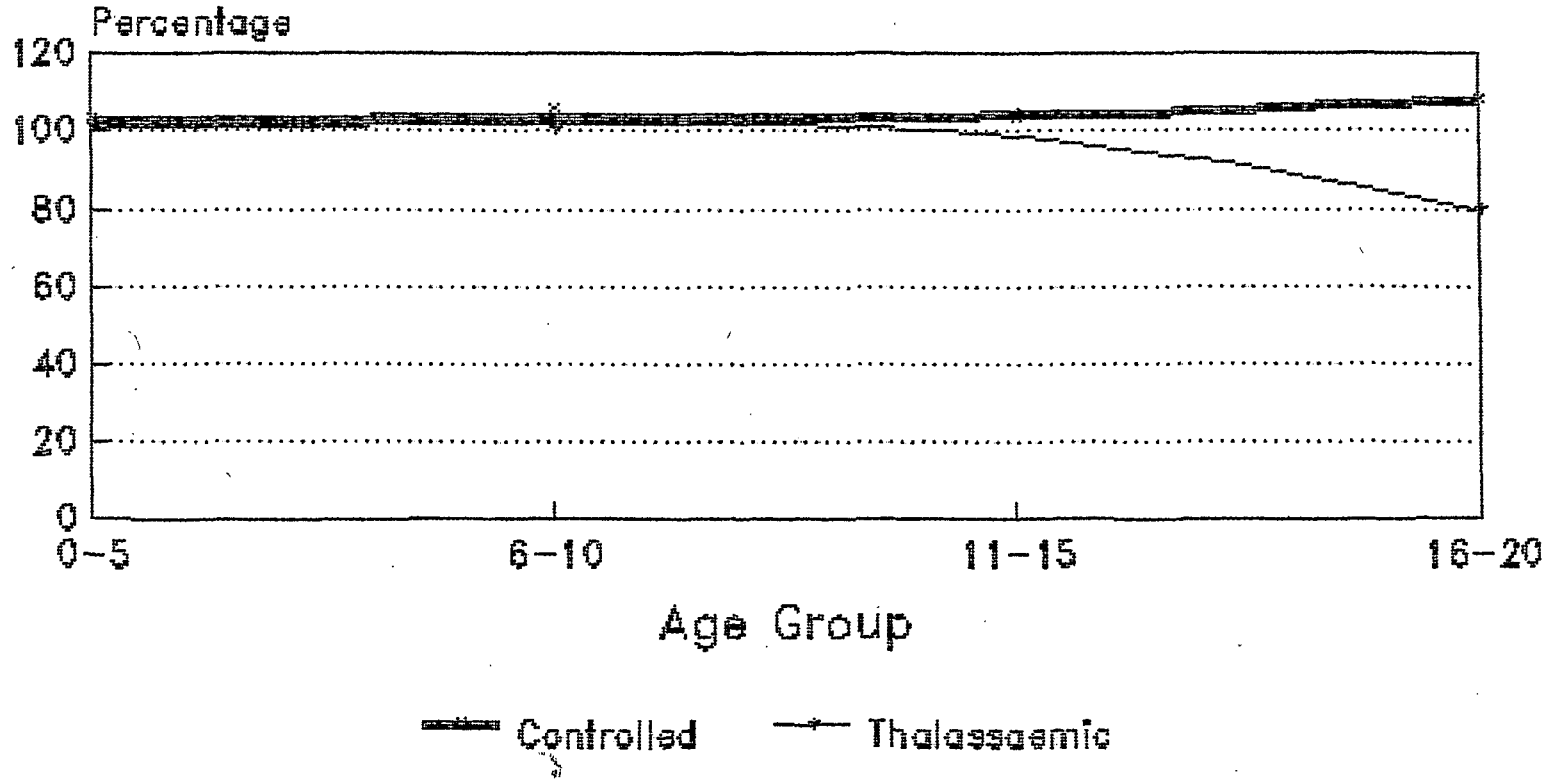

Fig. 29

in the age group 11-15 years in which the β-thalassaemic males significantly differ from the normal males and in the age group 16-20 years the β-thalassaemic females significantly differ from the normal females. However, it may be observed that height for age, and weight for age are greater in normal individuals in comparison to the β-thalassaemic counterparts. But in respect of weight for height no such difference between the normal and β-thalassaemic individuals is generally found.

Female

Figure-27 shows the growth curve in respect of height for age between the Controlled and β-thalassaemic girls. The curve shows that in all four age-groups in β-thalassaemic females are having lesser value than the Controlled sample. However, the differences between them gradually decreases with advancement of age.

Figure-28 shows the growth curve in respect of weight for age between the Controlled and β-thalassaemic girls. It shows that the Controlled sample throughout have higher mean values than the β-thalassaemic girls and with advancement of age, i.e. from 10-15 years the difference between them becomes broader.

Figure-29 depicts a comparative picture in respect of weight for height between the Controlled and β-thalassaemic

Male Sample

Growth Curve for Height for Age of Controlled & Thalassaemic Boys

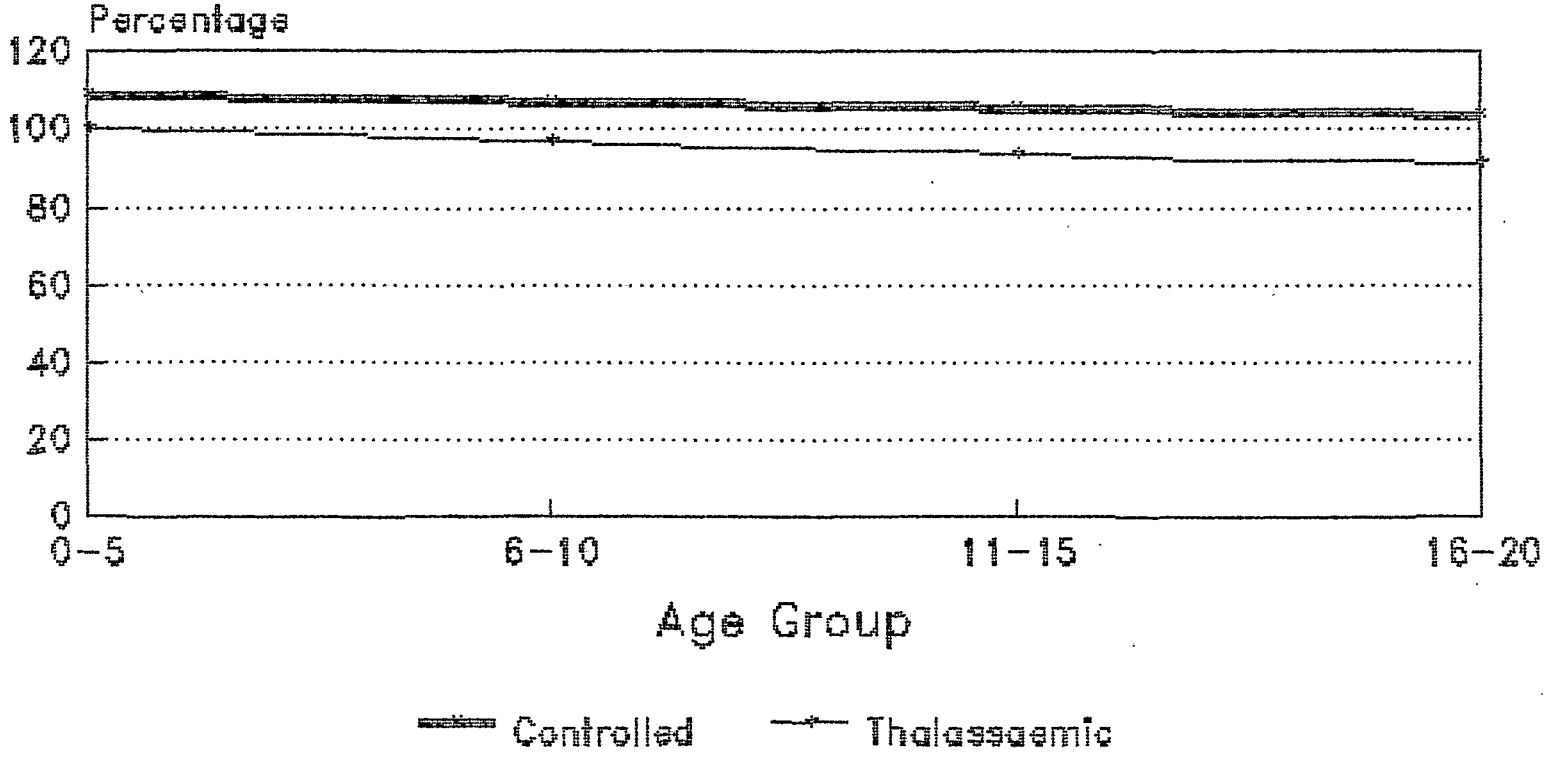

Fig. 30

Male Sample

Growth Curve for Weight for Age of Controlled & Thalassaemic Boys

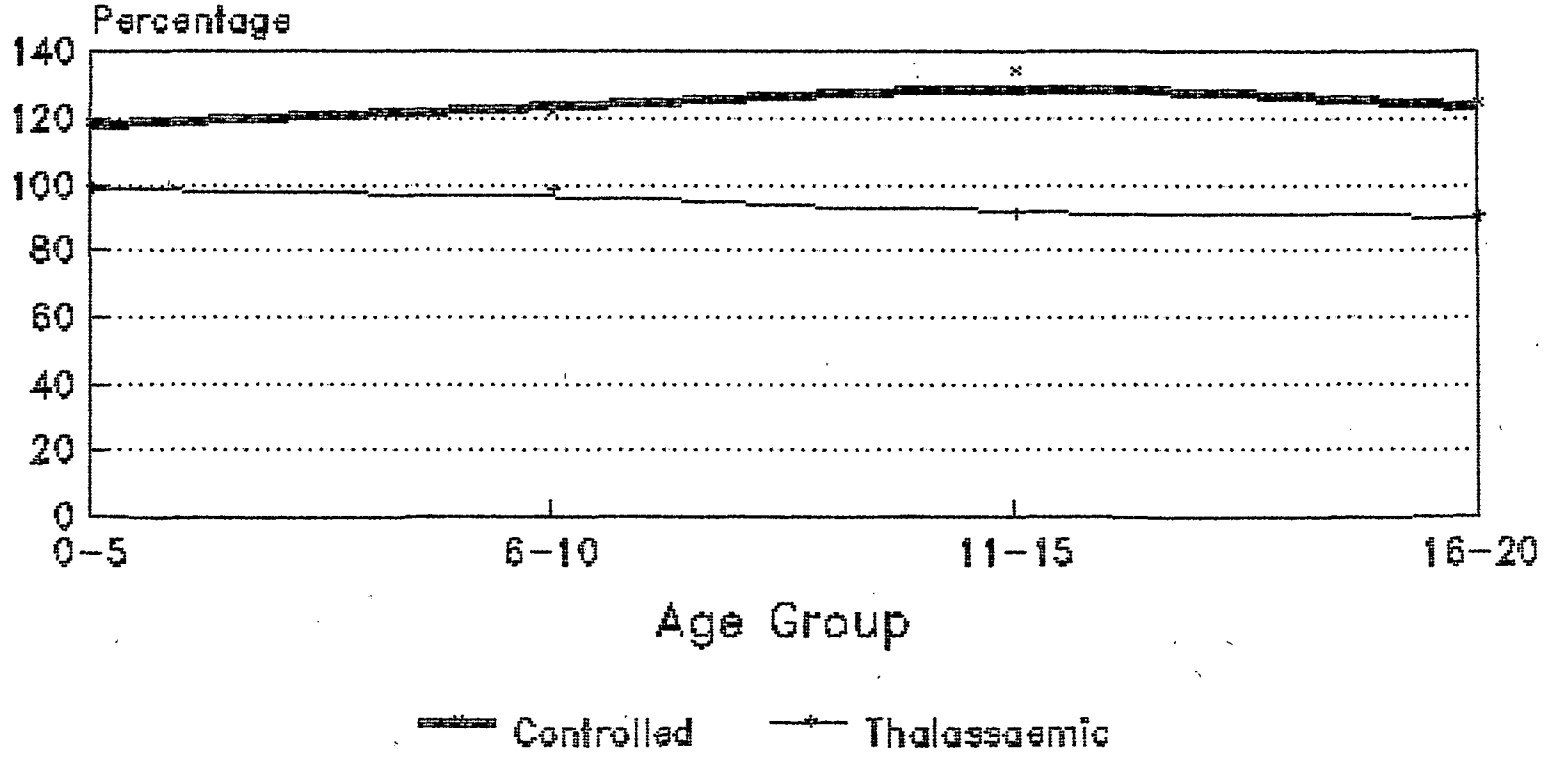

Fig. 31

Male Sample

Growth Curve for Weight for Height of Controlled & Thalassaemic Boys

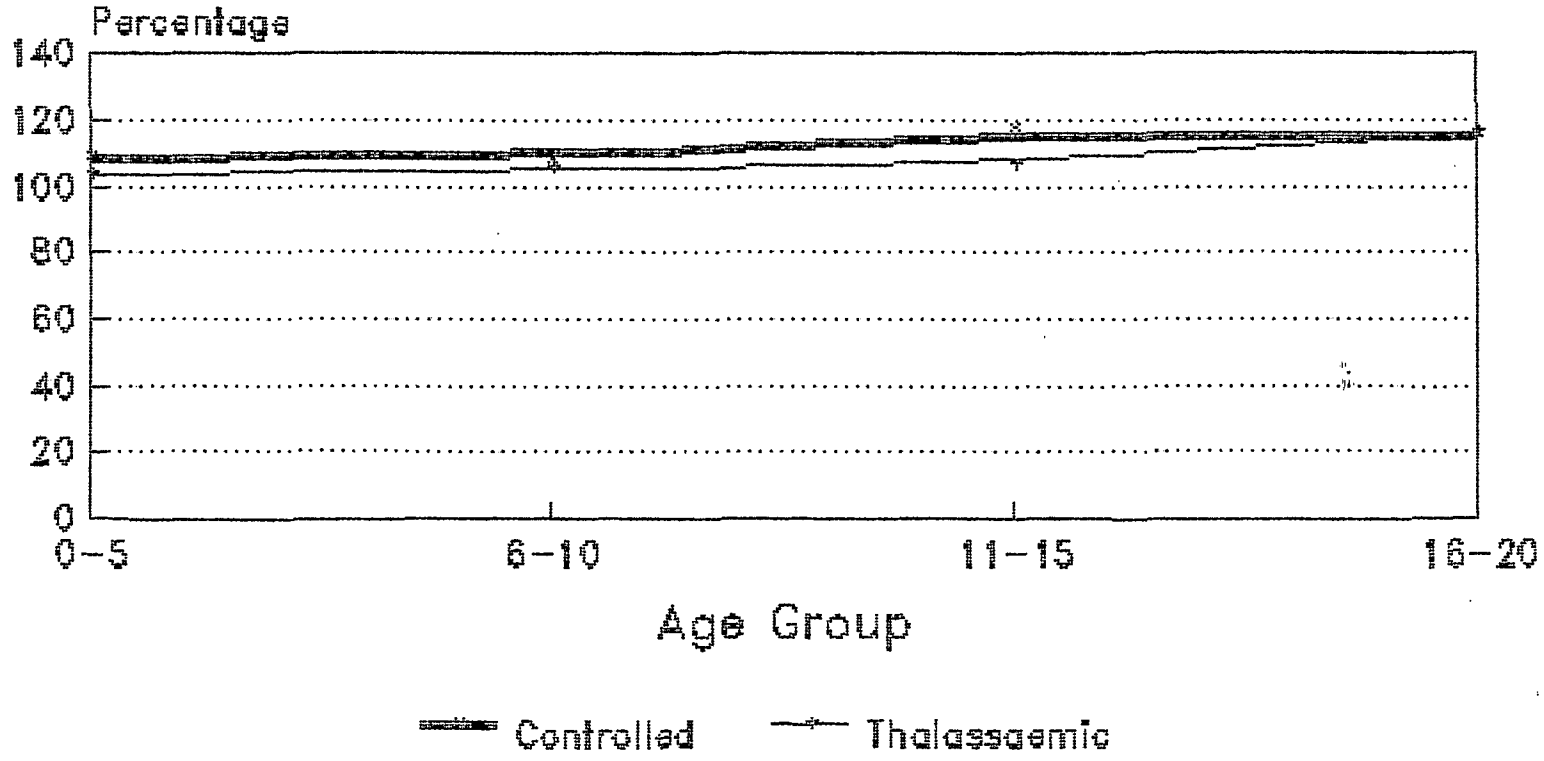

Fig. 32

girls. The curve shows that the mean value between the Controlled and β-thalassaemic sample is more or less same upto the age-group of 15 years and thereafter the value drops in case of the thalassaemic girls and the difference between them widens.

Male

Figure-30 shows the growth curve in respect of height for age between the Controlled and β-thalassaemic boys. It is seen that β-thalassaemic boys have lesser value than the Controlled sample in all four-age groups, though the difference between them has slightly broadened in the age group 16-20 years.

Figure-31 shows the growth curve in respect of weight for age between the Controlled and β-thalassaemic boys. It shows that the normal boys have higher values than the β-thalassaemic boys and the difference between them broadens from 6-10 years and continues to do so till the age group 16-20 years.

Figure-32 shows the growth curve in respect of weight for height between the Controlled and β-thalassaemic boys. It is seen that since the earliest age-group the value of weight for height in the normal boys is greater than the β-thalassaemic boys till the age group 11-15 years and

thereafter, the difference between them gets lesser and lesser till it becomes same in the age group 16-20 years.

Table 3.18 shows the rate of increment in respect of 13 anthropometric and skinfold traits. The rate of increment for the age-group 6-10 years have been calculated on the basis of the measurements found in the age group upto 5 years. Similarly, the rate of increment for the age group 11-15 years has been calculated on the basis of the measurements found in the age group 6-10 years and that for the age-group 16-20 years, on the basis of the age group 11-15 years.

In all anthropometric measurements and skinfold thickness it is found in males aged 6-10 years, the rates of increment in physical growth are greater in normal children than that in β-thalassaemic children excepting in case of height, body weight, humerous diameter and calf thickness. In the age group 11-15 years, the rates of increment of the normal children are greater in all anthropometric measurements and skinfold thickness excepting triceps thickness, for which the β-thalassaemic children show greater rate of increment. In the age-group 16-20 years all male β-thalassaemic children show greater rates of increment than the normal children excepting in case of skinfold thickness of biceps and triceps for which greater rates of increment are found in case of the normal children.

Table 3.18

Rate of Increment (%) in Physical Growth

Anthropometric Traits (in cm.)		Normal and Thalassaemic Children							
		Male (age group in yrs)				Female (age group in yrs)			
		0-5	6-10	11-15	16-20	0-5	6-10	11-15	16-20
Height	C	-	16.60	16.73	9.35	-	15.00	15.05	5.04
	T	-	17.77	13.37	9.67	-	21.15	10.13	3.75
Body Weight	C	-	31.48	42.02	23.56	-	35.42	52.91	20.96
	T	-	35.14	24.52	35.34	-	36.67	31.32	8.12
Chest Girth (inhale)	C	-	13.15	17.07	9.77	-	11.91	23.90	9.57
	T	-	11.17	11.14	12.24	-	11.39	12.48	1.51
Chest Girth (exhale)	C	-	12.56	16.81	9.46	-	11.33	16.79	9.00
	T	-	9.45	8.56	14.72	-	11.32	12.20	1.46
Humerous Diameter	C	-	10.80	17.08	0.50	-	11.29	10.65	0.56
	T	-	34.70	12.05	15.45	-	10.26	11.91	5.07
Bicondylar Femur Dia.	C	-	19.95	7.72	7.68	-	11.22	12.21	1.59
	T	-	10.04	7.43	12.66	-	10.87	7.36	5.15
Bicep Girth	C	-	9.80	18.24	9.78	-	9.98	14.22	10.13
	T	-	5.74	5.85	17.35	-	5.08	9.99	5.79
Calf Girth	C	-	13.95	19.38	11.42	-	14.89	15.25	10.43
	T	-	11.91	6.61	17.52	-	9.83	10.41	4.46
*Biceps	C	-	10.00	14.85	21.16	-	4.13	32.40	17.50
	T	-	2.15	8.82	10.07	-	7.21	4.40	11.00
*Triceps	C	-	8.43	3.00	17.83	-	9.25	21.10	24.15
	T	-	2.81	22.64	15.38	-	2.52	13.84	7.43
*Subscapular	C	-	16.73	21.00	17.07	-	19.62	32.15	28.23
	T	-	4.59	6.70	31.43	-	3.26	17.31	31.43
*Supra-illac	C	-	23.25	31.31	11.95	-	32.97	17.18	16.04
	T	-	4.90	5.69	48.35	-	4.74	27.96	2.06
*Calf	C	-	4.14	20.34	9.82	-	8.12	19.44	11.04
	T	-	4.19	17.02	26.81	-	1.23	10.61	2.73

* Skinfold thickness; C denotes control; T denotes thalassaemia.

In female, aged 6-10 years, the normal girls show greater rates of increment in physical growth in all measurements, excepting height, body weight and biceps skinfold thickness than their β-thalassaemic counterparts of the same age group. In the next age group, i.e., 11-15 years, all normal girls show greater rates of increment in physical growth than their β-thalassaemic counterparts excepting in case of humerous diameter and supra-illiac skinfold thickness. In the age group 16-20 years the β-thalassaemic girls, however, show greater rate of increment in physical growth for humerous diameter bicondylar, femur diameter and subscapular skinfold thickness than their normal counterparts of the same age groups.

It is seen from the above discussion that the rate of increment in physical growth of the β-thalassaemic children, boys and girls, is slower than that of the normal children, though there are some exceptions in one or the other measurement. So one can by and large infer that the physical growth of the β-thalassaemic children is retarded at least upto 15 years of age, and thereafter though the rate of increment in physical measurements improves, the β-thalassaemic patients generally never catch-up the normal children in physical growth.

CHAPTER-IV

SOCIAL ASPECTS

In this chapter various social aspects will be dealt with. It covers demographic composition of the families with β-thalassaemic patients, economic conditions of the parents of the β-thalassaemic children and their psychological stresses and economic hardships. Besides, academic and extra-curricular activities of the β-thalassaemic patients will also be discussed.

Table 4.1

Thalassaemic Patients by Age and Sex

Age group (in years)	Male		Female		Total	
	No.	%	No.	%	No.	%
< 1	5	2.04	2	1.48	7	1.84
1-5	67	27.35	40	29.63	107	28.16
6-10	101	41.22	53	39.26	154	40.53
11-15	54	22.04	31	22.96	85	22.37
16-20	13	5.31	6	4.44	19	5.00
20+	5	2.04	3	2.22	8	2.11
Total	245	100.00	135	99.99	380	100.01

Table 4.1 shows age and sex of the β-thalassaemic patients. In the present study we have been able to collect information on 380 β-thalassaemic patients of which 245 (i.e. 64.47 per cent) are males and 135 (35.53 per cent) are females. It further shows that 1.84 per cent, 28.16 per cent, 40.53 per cent, 22.37 per cent, 5.00 per cent and 2.11 per cent of all β-thalassaemic patients belong to the age groups <1, 1-5, 6-10, 11-15, 16-20 and 20+ years respectively. It is found in the present sample that of all male β-thalassaemic patients 2.04 per cent, 27.35 per cent, 41.22 per cent, 22.04 per cent, 5.31 per cent and 2.04 per cent belong to the age-groups <1, 1-5, 6-10, 11-15, 16-20 and 20+ years respectively. Similarly, of all β-thalassaemic female patients 1.48 per cent, 29.63 per cent, 39.26 per cent, 22.96 per cent, 4.44 per cent and 2.22 per cent belong to the age groups <1, 1-5, 6-10, 11-15, 16-20 and 20+ years, respectively. It is found that most of the β-thalassaemic patients, male or female, belong to the age group 6-10 years. However, it is seen that as age advances, particularly after 15 years of age, the frequency of β-thalassaemic patients gradually decreases.

Table 4.2 show the frequency of the β-thalassaemic patients by religion and caste. In the present sample there are 336(88.42 per cent) β-thalassaemic patients, belonging to Hindu religion and all of them are Bengali speaking.

Table 4.2

Thalassaemic Patients by Religion and Caste

Religion/Caste	Male	Female	Number	Percentage
A. Hindu				
i) Brahmin	32	20	52	13.69
ii) Kayastha	61	37	98	25.79
iii) Mahisya	41	14	55	14.48
iv) Sodgop	15	10	25	6.58
v) Yadav	6	4	10	2.63
vi) Misc. castes	64	32	96	25.27
vii) Other castes (non-Bengalee)	2	3	5	1.32
B. Muslim	20	14	34	8.95
C. Christian	4	1	5	1.32
Total	245	135	380	100.00

Besides, there are 5 β-thalassaemic patients who are non-Bengalee, though belonging to Hindu religion. In the present sample there are 34(8.95 per cent) β-thalassaemic patients, who are Islam by faith and 5(1.32 per cent) who are Christian by faith. However, both the Muslim and Christian β-thalassaemic patients are all Bengalee speaking individuals. Among the Hindu β-thalassaemic patients 13.69 per cent, 25.79 per cent, 14.48 per cent, 6.58 per cent, 2.63 per cent and 25.27 per cent belong to the Brahmin, Kayastha, Mahisya,

Sodgop, Yadav and miscellaneous castes respectively. So the present sample shows that most of the β-thalassaemic patients belong to Kayastha Caste, followed by Mahisya and Brahmin castes. In the present sample only a very few β-thalassaemic patients are Christian by faith.

Table 4.3 shows the overall reproductive performance of the mothers of the β-thalassaemic patients. This table indicates total number of pregnancies, live births, surviving children, dead children and reproductive wastage. While calculating the mean number of live births, all individual live births, not total pregnancy, have been considered, since some twin births are there in the present sample. The mean number of live births are found to be 1.67±0.13, 1.84±0.13, 2.67±0.23, 1.80±0.19, 1.40±0.21 and 2.23±0.31 respectively of the Brahmin, Kayastha, Mahisya, Sodgop, Yadav and miscellaneous castes of the Bengalee speaking population. The mean of number of live births among the Muslim mothers is found to be 3.32±0.37 and that of the Christian mothers 2.00±0.49. Similarly among the non-Bengali Hindu Caste mothers, the mean number of live births is found to be 1.80±0.33. It shows that among the Muslim the mean number of live births is highest (3.32 per cent), which is followed by Mahisya (2.67 per cent). In case of all other groups the mean number of live births is around 2.00.

Table 4.3

Reproductive History of the Mothers with ß-Thalassaemic Children

Religion/ Caste	No. of Mothers	No. of Pregnancies	No. of L.B	Live Births			No. of S.C.	Surviving Children			No. of Dead Child	Dead Children			Abortion		Still Birth		RW*	
				Mean	S.D.	±S.E.		Mean	S.D.	±S.E.		Mean	S.D.	±S.E.	No	%	No	%	No	%
Hindu																				
a) Brahmin	48	104	80	1.67	0.94	±0.13	72	1.50	0.68	0.10	8	0.17	0.47	±0.07	23	22.33	1	0.97	24	23.30
b) Kayastha	96	223	177	1.84	1.24	0.13	151	1.57	0.91	0.09	26	0.27	0.62	0.06	46	20.62	0	0.00	46	20.62
c) Kalhisya	49	153	131	2.67	1.63	±0.23	112	2.29	1.44	0.20	19	0.38	0.66	0.09	22	14.37	0	0.00	22	14.37
d) Sodgop	25	56	45	1.80	0.94	0.19	41	1.64	0.69	0.14	4	0.16	0.61	0.12	11	19.64	0	0.00	11	19.64
e) Yadav	10	19	14	1.40	0.66	0.21	14	1.40	0.66	0.21	0	0.00	0.00	0.00	5	26.32	0	0.00	5	26.32
f) Other Caste	96	247	214	2.23	1.24	0.31	183	1.91	0.94	0.10	31	0.32	0.78	0.08	33	13.36	0	0.00	33	13.36
g) Other Caste (Non-Bengalee)	5	13	9	1.80	0.75	0.33	8	1.60	0.49	0.22	1	0.20	0.40	0.18	4	30.76	0	0.00	4	30.76
Muslim	31	113	103	3.32	2.04	0.37	87	2.81	1.86	0.33	16	0.52	0.67	0.12	8	7.08	2	1.77	10	8.85
Christian	5	12	10	2.00	1.10	0.49	8	1.60	0.48	0.22	2	0.40	0.80	0.36	2	16.67	0	0.00	2	16.76

Note: *Reproductive Wastage (Abortion & Still Birth together)

The mean number of surviving children is found to be 1.50±0.10, 1.57±0.09, 2.29±0.20, 1.64±0.14, 1.40±0.21 and 1.91±0.10 respectively among the Brahmin, Kayastha, Mahisya, Sodgop, Yadav and miscellaneous castes of the mothers of the β-thalassaemic patients. Among the Muslim mothers the mean number of surviving children is found to be 2.81±0.33 whereas the same among the Christian mother is 1.60±0.22. Among the non-Bengalee Hindu Caste the mean number of surviving children is 1.60±0.22.

So far as the frequency of dead children is concerned, it is found that the mean number of dead children in the various Hindu Caste groups, Muslim and Christian varies between 0.16±0.12 (in the Sodgop) and 0.52±0.12 (in the Muslim). However, it shows that the frequency of dead children in these populations is very low. It may be due to the fact that they, being urban dwellers, have been enjoying better medical facilities.

Regarding reproductive wastage it may be noted that the frequency has been calculated on the basis of total number of pregnancies. It is found that the frequency of reproductive wastage among the Brahmin is 23.30 per cent, whereas among the Kayastha the same is 20.62 per cent, among the Muslim mothers the frequency of reproductive wastage is 8.85 per cent. However, it is seen that the frequency of abortions in this caste and religious groups varies from 7.08 per cent to

22.33 per cent. It shows that the frequency of abortions among the mothers of the β-thalassaemic patients is considerably high.

Table 4.4

Thalassaemic Patients - By Birth Order

Birth Order	Thalassaemic Patients	
	Number	Percentage
1	199	52.37
2	89	23.42
3	45	11.84
4	20	5.27
5	9	2.37
6	10	2.63
7	8	2.10
Total	380	100.00

Table 4.4 shows the occurrence of the β-thalassaemic patients by birth order. It is seen that nearly 52.37 per cent of all β-thalassaemic patients belong to the first birth order and 23.42 per cent and 11.84 per cent are of second and third birth orders respectively. It is further observed that there is an inverse relationship between occurrence of β-thalassaemia and number of birth order, which means that as the number of birth order increases, birth of a baby with β-

thalassaemia decreases. Births of most of the β-thalassaemic patients are either in first, second or third birth orders.

Table 4.5

Educational Achievements of the Parents

Education	Number	Percentage
1. Illiterate	28	7.67
2. Primary	22	6.03
3. Secondary	167	45.76
4. Higher Secondary	41	11.23
5. Graduate	107	29.32
Total	365	100.00

Table 4.5 shows educational achievement of the parents of the β-thalassaemic patients. It is found that nearly 7.67 per cent of such parents are illiterate, while 6.03 per cent of them have had education upto the primary level. The majority of the parents (45.76 per cent) have had education upto the secondary level. Only 11.23 per cent of all such parents have read upto the higher secondary level. 29.32 per cent of all such parents are found to be graduate.

Table 4.6

Economic Condition of the Families*

Income Group	Number	Percentage
L.I.G. (Below Rs.648.01)	171	50.74
M.I.G. (Rs.648.01 to Rs.1179.17)	88	26.11
H.I.G. (Above Rs.1179.17)	78	23.15

* 28 of the parents did not say about their income. They have not been included.

Table 4.6 shows the economic condition of the families in which there are β-thalassaemic patients. The economic condition of the families has been classified into 3 categories namely, lower income group (L.I.Gr.) - monthly income less than Rs.648.01, middle income group (M.I.Gr.) - monthly income between Rs.648.01 and Rs.1179.17 and high income group (H.I.Gr.) - monthly income above Rs.1179.17. The method of classification of economic condition has already been explained in the Chapter-II. In the present study we have altogether 365 families. However, since the information about monthly income of 28 families is not available, the present classification is based on 337 families. From Table-4.6 it is seen that nearly 50.74 per cent of the families belong to the lower income group whereas 26.11 per cent and 23.15 per cent of all families belong to the middle income group and high income group respectively.

Table 4.7

Economic Condition of the Families and the Frequency of β-thalassaemic children

Income group	No.of Normal children	%	Total No. of β-thalassaemic children	%	Total No.of surviving children
L.I.G.	199	52.65	179***	47.35	378
M.I.G.	36	29.57	90**	71.43	126
H.I.G.	36	30.77	81*	69.23	117

* includes 3 twins

** includes 1 twin sister and 1 brother

*** includes 3 twins.

28 of the parents who did not say about their income have not been included and their 30 children including 2 twins have also been excluded.

X^2 (between 3 income groups) 31.981, df=2, P<0.05.

X^2 (between Low & Middle Income Group) 22.01, df=1, P<0.05

X^2 (between Low & High Income Group) 17.15, df=1, P<0.05

X^2 (between Middle & High Income Group) 0.1405, df=1, P>0.05.

Table 4.7 shows the economic condition of the families and the frequency of β-thalassaemic children. It is found that in the families, belonging to the lower income group, there are altogether 378 children out of which 199, i.e. 52.65 per cent are normal and 179, i.e. 47.35 per cent are β-thalassaemic. In the families, belonging to the middle income

group, out of total number of 126 children 36, i.e. 29.54 per cent are normal and 90, i.e. 71.43 per cent are β-thalassaemic. In the families with high income there are altogether 117 children out of which, 36, i.e. 30.77 per cent are normal and 81, i.e. 69.23 per cent are β-thalassaemic.

The X^2 value shows that there are significant differences in the frequency of β-thalassaemic children among these three income groups (X^2-31.981 df=2, P <0.05). But the X^2 value shows that there is no significant difference between middle and high income groups in respect of the frequency of β-thalassaemic patients (X^2=0.1405, df=1, P>0.05). But the X^2 value between middle and low income groups in respect of the frequency of β-thalassaemic patients shows that there is significant difference (X^2=22.01, df=1, P<0.05). Similarly, the X^2 value between high and low income groups, in respect of frequency of β-thalassaemic patients, shows that there is statistically significant difference (X^2=17.15, df=1, P<0.05). Since the present sample of the β-thalassaemia patients has been collected from 5 hospitals in Calcutta, it may well be assumed that the people, belonging to the lower income group, bring their diseased children more in hospitals than their counterparts, belonging to either the middle income group or the high income group. The reason for doing so is that the average expenditure on a β-thalassaemic patient in any hospital is much lower than that in any

private clinic. Consequently, the people, belonging to the lower income group, have no choice but to rely on hospitals for the treatment of their diseased children, whereas the people, belonging to the high income group can afford to spend a huge amount on their diseased children for treatment in any private clinic and those belonging to the middle income group, though cannot afford as much as the people in the high income group, even prefer going to private clinics to government hospitals.

Table 4.8

Expenditure for Treatment of β-thalassaemic Children

Expenditure per month	Number	Percentage
Upto Rs.500.00	98	27.84
Rs.501-1000	132	37.50
Rs.1001-1500	46	13.06
Rs.1501-2000	36	10.23
Rs.2001-2500	14	3.98
Rs.2501-3000	11	3.13
Rs.3001-3500	2	0.57
Rs.3501-4000	7	1.98
Rs.4000+	6	1.71
Total	352	100.00

Table 4.8 shows how much expenditure per month on a β-thalassaemic patient is incurred by the parents. It is seen that the minimum amount of Rs.500/- p.m. is spent on a β-thalassaemic child whereas it is seen that some parents spend as much as Rs.4000/- and more p.m. on a β-thalassaemic child. However, it is found that for 27.84 per cent of all β-thalassaemic patients the minimum expense is Rs.500/- or less per month, whereas for 37.50 per cent of the β-thalassaemic patients the same varies between Rs.501/- and Rs.1000/-. For nearly 1.71 per cent of β-thalassaemic patients the parents spend more than Rs.4000 per month. It is seen that richer the parent, more money is spent for treatment of their β-thalassaemic children in comparison to the parents, belonging to the lower and middle income groups. However, it is well understood that irrespective of economic condition, the parents of the β-thalassaemic children have to spend a substantial amount of their monthly income on their diseased children.

In the present study there are 380 β-thalassaemic patients, belonging to different economic groups. However, 28 parents of 30 thalassaemic patients could not tell about their monthly income. Consequently, to calculate the average expenditure on treatment of the β-thalassaemic patients, those 30 thalassaemic patients have not been included. Out of

those 30 patients 16 are getting free treatment at various hospitals in Calcutta.

Table 4.8A

Mean Expenditure per month on β-thalassaemic Patients

Economic Group	No.of β-thalassaemic Children	Expenditure*		
		Mean(Rs.)	S.D.	± S.E.
L.I.G.	179	714.39	474.90	35.44
M.I.G.	90	1082.33	602.89	63.55
H.I.G.	81	2158.64	1626.28	180.74

* Overall Mean expenditure: 1143.24±57.43 per month.

Table 4.8A shows the average expenditure on the β-thalassaemic patients per month. There are 179 β-thalassaemic patients, belonging to the lower income group. The mean expenditure per month per patient is Rs.714.39±35.44. There are 90 β-thalassaemic patients in the middle income group and the mean expenditure per month per patient is found to be Rs.1082.33±63.55. In the high income group there are 81 β-thalassaemic patients, and it is found that the mean expenditure per β-thalassaemic patient per month is Rs.2158.64±180.74. Combining all the patients of these three economic groups together, it is found that the average expenditure on treatment of the β-thalassaemic patients per month is found to be Rs.1143.24±57.43. From the above table,

it is seen that the well-to-do families can afford to spend much more money on their β-thalassaemic children for their treatment in comparison to those belonging to either the lower income group or the middle income group. It is further noticed that the parents of the middle income group spend much more money for treatment of their β-thalassaemic children than what their counterparts in the low income group can afford.

Table 4.9

Economic Hardhsip of the Parents

Income Group	Economic Hardship		No. Economic Hardship		Total
	No.	%	No.	%	
L.I.G. (Below Rs.648.01)	160	93.57	11	6.43	171
M.I.G. (Rs.648.01 to 1179.17)	59	67.05	29	32.95	88
H.I.G. (Above Rs.1179.17	27	34.62	51	65.38	78

In the present study, we have collected information regarding economic hardship faced by the parents of the β-thalassaemic children. Altogether we have been able to collect their information from 337 parents, belonging to the low, middle and high income groups. In the low income group there are 171 parents of which 160, i.e. 93.57 per cent have

admitted that they have been facing tremendous economic hardship for the treatment of their diseased children. In the middle income group there are altogether 88 parents out which 59, i.e. 67.05 per cent are having tremendous economic the hardship for meeting the expenses for their diseased children. In the high income group, out of 78 parents only 27, i.e., only 34.62 per cent have expressed such feeling of hardship. From this table it is quite clear that in the low economic condition, economic hardship to meet expenses on medical treatment of a β-thalassaemic child is much more than that either in the middle income group or in the high income group.

Educational and other Activities of the β-thalassaemic Patients

Table 4.10 shows schooling of the β-thalassaemic patients. All β-thalassaemic patients have been classified into 5-age groups, namely upto 5, 6-10, 11-15, 16-20 and above 20 years. It may further be noted that in the age groups upto 5 years, all those β-thalassaemic children aged 2 years and below have not been included since they have not reached the school going age. In the present sample there are altogether 380 β-thalassaemic patients, out of which 7 are below 2 years of age. So, the present analysis has been made on 373 β-thalassaemic patients.

Table 4.10

Schooling of the β-thalassaemic Patients

	Age(in years)	Male		Female		Total	
		No.	%	No.	%	No.	%
Attended School	Upto 5	23	13.37	14	15.05	37	13.96
	6-10	87	50.58	42	45.16	129	48.68
	11-15	50	29.07	29	31.18	79	29.81
	16-20	11	6.40	5	5.38	16	6.04
	Above 20	1	0.58	3	3.23	4	1.51
	Total %	172 (64.91)	100.00	93 (35.09)	100.00	265	100.00
Did not Attend	Upto 5	44	63.77	26	65.00	70	64.81
	6-10	14	20.29	11	27.50	25	23.15
	11-15	4	7.25	2	5.00	6	5.56
	16-20	2	2.90	1	2.50	3	2.78
	Above 20	4	5.80	0	0.00	4	3.70
	Total %	68 (62.96)	100.01	40 (37.04)	100.00	108	100.00

* Figures in parenthesis indicate percentage of the total
1) Total males = 240; % of school going = 71.67
% of not going to school = 28.33
2) Total female = 133; % of school going = 69.92
% of not going to school = 30.08

Of 373 β-thalassaemic patients, it is found that 265, i.e. 71.04 per cent, have been attending school, whereas 108, i.e. 28.95 per cent β-thalassaemic patients have not gone to any school. It is seen that out of 265 β-thalassaemic

patients, who have been attending school, 172 (64.91 per cent) are boys and 93 (35.09 per cent) are girls. Similarly, it is found that out of 108 β-thalassaemic patients, who are not attending any school, 68 i.e. 62.96 per cent are boys and 40 i.e. 37.04 per cent girls. It may further be noted that out of 240 male β-thalassaemic patients, 71.67 per cent are attending school, whereas 28.33 per cent are not going to any school. Similarly out of 133 β-thalassaemic girl patients, 69.92 per cent are attending school and 30.08 per cent are not going to any school. It is further observed that most of the β-thalassaemic patients, boys or girls, aged between 6 and 10 years, are going to school. It is true for both sexes that as age advances the frequency of attending school gradually decreases.

Table 4.11 shows the present educational achievement of the β-thalassaemic patients. It shows that 20.35 per cent of males and 18.28 per cent of females have been going to pre=primary school, whereas 51.74 per cent of males and 50.54 per cent of females are in the primary school. Similarly, 23.84 per cent of males and 29.03 per cent of females are in the secondary level. It is further seen that only 2.91 per cent of males and 2.15 per cent of females are in the higher secondary level. Only 2 β-thalassaemic males have been found to be graduate. It is true that the β-thalassaemic patients

Table 4.11

Educational Achievement by Age and Standard of Education

	Age in Years											
	1-5		6-10		11-15		16-20		20+		Total	
	M	F	M	F	M	F	M	F	M	F	M	F
Pre-Primary	22	10	12	7	1	0	0	0	0	0	35 (20.35)	17 (18.28)
Primary	1	4	73	34	14	7	1	1	0	1	89 (51.74)	47 (50.54)
Secondary	0	0	2	1	35	22	4	3	0	1	41 (23.84)	27 (29.03)
Higher Secondary	0	0	0	0	0	0	5	1	0	1	5 (2.91)	2 (2.15)
Graduate+	0	0	0	0	0	0	1	0	1	0	2 (1.16)	0 (0.00)
Total											172 (100.00)	93 (100.00)

generally do not go beyond the secondary level, though a very few try to achieve higher education. It may perhaps happen due to the fact that the β-thalassaemic patients suffer from various health problems since very early in childhood, and with advancement of age, health problems are getting multiplied.

Table 4.12

Performance in School

Age (in years)	Performance in School					
	Satisfactory		Not Satisfactory		Total	
	M	F	M	F	M	F
1-5	12 (52.17)	6 (42.86)	11 (47.83)	8 (57.14)	23	14
6-10	52 (59.77)	22 (52.38)	35 (40.23)	20 (47.62)	87	42
11-15	26 (52.00)	11 (37.93)	24 (48.00)	18 (62.07)	50	29
16-20	6 (54.55)	2 (40.00)	5 (45.45)	3 (60.00)	11	5
20+	1 (100.00)	3 (100.00)	0	0	1	3
Total	97 (56.40)	40 (47.31)	75 (43.60)	49 (52.64)		

In Table 4.12, performance of the β-thalassaemic patients in school has been classified into two groups: satisfactory and non-satisfactory. The details of this classification has already been described in the Chapter-II. It is seen that irrespective of age-groups the male β-thalassaemic patients have been performing more satisfactorily than their female counterparts. It is found that of 56.40 per cent of male β-thalassaemic patients perform satisfactorily, whereas 47.31 per cent of female β-thalassaemic patients do the same. It is also observed that

among the β-thalassaemic patients 43.60 per cent and 52.64 per cent of male and female respectively have not been performing satisfactorily. It means that nearly 50 per cent of all β-thalassaemic patients just cannot do well in their academic pursuits. It seems that their serious health problems prevent them from doing well in educational matter.

Table 4.13

Participation in Extra Curricular Activities

Partici-pation in	Age(in years)	Male		Female		Total	
		No.	%	No.	%	No.	%
Play	1-5	64	28.44	36	31.86	100	29.58
	6-10	98	43.56	48	42.48	146	43.20
	11-15	49	21.78	24	21.24	73	21.60
	16-20	11	4.89	4	3.54	15	4.44
	20+	3	1.33	1	0.88	4	1.18
	Total	225	100.00	113	100.00	338	100.00
No Play	1-5	3	20.00	4	20.00	7	20.00
	6-10	3	20.00	5	25.00	8	22.85
	11-15	5	33.33	7	35.00	12	34.29
	16-20	2	13.33	2	10.00	4	11.43
	Total	15	99.99	20	100.00	35	100.00

Grand total : Male 240: Play - 93.75 per cent
No Play - 6.25 per cent
Female 133: Play - 84.96 per cent
No Play - 15.04 per cent.

Table 4.13 shows how many of the β-thalassaemic patients participate in extra-curricular activities. It may be noted that in this analysis only those β-thalassaemic patients, who have been attending educational institutions have been included. It is seen that out of 240 β-thalassaemic boys, nearly 93.75 per cent (225) participate in games, whereas 6.25 per cent of them (15) never take any part in any sort of games. It is further observed that out of 133 β-thalassaemic girls 84.96 per cent (113) do participate in games and the rest (20), i.e., 15.04 per cent, do not take part in any game. So it shows that somehow, the boys take part more in games than the girls. It is further observed that both boys and girls aged up to 15 years take more part in games in comparison to those who are beyond 15 years of age. It happens perhaps due to the fact that as age advances, physical disabilities of the β-thalassaemic patients increase, and that prevents them from taking part in any extra-curricular activity.

Table 4.14 shows the nature of participation in games of the β-thalassaemic patients. It is found that out of 225 β-thalassaemic male patients, who have been participating in games, 34.22 per cent participate only in indoor games, whereas 22.22 per cent participate in outdoor games. It is also seen that nearly 43.56 per cent of them participate in both indoor and outdoor games. Out of 113 β-thalassaemic

Table 4.14

Nature of Participation in Games by the Thalassaemic Patients

Age in Years	Nature of Participation										
	Indoor		Outdoor		Indoor & Outdoor		Total		Grand Total		
	M	F	M	F	M	F	M	F	No	M %	F %
1 - 5	27 (42.19)	20 (56.76)	8 (12.50)	5 (13.51)	29 (45.31)	11 (29.73)	64 (100.00)	36 (100.00)	100	64.00	36.00
6 -10	23 (23.47)	20 (42.55)	29 (29.59)	6 (12.77)	46 (46.94)	22 (44.68)	98 (100.00)	48 (100.00)	146	67.12	32.88
11-15	21 (42.86)	12 (52.00)	9 (18.37)	5 (20.00)	19 (38.78)	7 (28.00)	49 (100.01)	24 (100.00)	73	67.12	32.88
16-20	4 (36.36)	3 (75.00)	4 (36.36)	0 (0.00)	3 (27.27)	1 (25.00)	11 (99.99)	4 (100.00)	15	73.33	26.67
20+	2 (67.67)	1 (100.00)	0 (0.00)	0 (0.00)	1 (33.33)	0 (0.00)	3	1 (100.00)	4	75.00	25.00
Total	77 (34.22)	56 (49.56)	50 (22.22)	16 (14.16)	98 (43.56)	41 (36.28)					

girls 49.56 per cent and 14.16 per cent participate in indoor and outdoor games respectively. But 36.28 per cent of them participate in both indoor and outdoor games. However, it is seen that by and large the male β-thalassaemic patients participate more in games than the female β-thalassaemic patients.

Table 4.15

Degree of Physical Activities of the Thalassaemic Patients

Age (in Years	Active		Average		Inactive		Total	
	M	F	M	F	M	F	M	F
1 - 5	41	18	22	13	4	9	67	40
6 -10	40	18	40	25	21	10	101	53
11-15	13	7	26	17	15	7	54	31
16-20	5	4	6	1	2	1	13	6
20+	1	1	3	1	1	1	5	3
Total	100 (41.67)	48 (36.09)	97 (40.42)	57 (42.85)	43 (17.92)	28 (21.05)	240	133

Table 4.15 shows the degree of physical activities of the β-thalassaemic patients by age and sex. The classification of degree of physical activities has already been described in the Chapter-II. It is seen that out of 240 male β-thalassaemic patients, 41.67 per cent are physically active, whereas 17.92 per cent are physically inactive. For the rest (i.e. 40.42 per cent), physical activities are average. In case of 133 β-thalassaemic female patients 36.09 per cent are physically active, 21.05 per cent are physically inactive, and 42.85 per cent are average in their physical activities. It shows that the male β-thalassaemic patients are on an average better active physically than their female counterparts. The point to be noted here is that as age advances, physical activities of the β-thalassaemic patients, both boys and girls, decline steadily.

Table 4.16

Concept of Parents About Marriage of Their Diseased Child

Undecided	Yes	No	Depends on the Children
117 (32.05)	88 (24.11)	140 (38.36)	20 (5.48)

Table 4.16 shows the opinions of the parents regarding marriage of their β-thalassaemia children. It shows that 38.36 per cent of the parents, who are having β-thalassaemic children, are quite sure that the diseased children should not marry. The reason that they have put forward, is that such mental agony of the parents and physical discomforts of the diseased children should not be encouraged and consequently, the parents are of the opinion that they will not allow their diseased children to go for marriage. On the other hand, 88 (i.e., 24.11 per cent of the parents with β-thalassaemic children are, however, in favour of allowing their diseased children to go for marriage. There are 117 such parents (32.05 per cent) who have been found to be undecided about marriage of their β-thalassaemic children. Also it is found that there are 20 (i.e., 5.48 per cent) parents, who have no personal opinion regarding marriage of their β-thalassaemic children but they are willing to leave it to their diseased children.

Table 4.17

Reasons in Favour & Against Marriage of Thalassaemic Children

Positive	No	%
a) Only if the doctor permits	4	4.55
b) Only if he/she gets permanently cured	42	47.73
c) For continuation of lineage	18	20.45
d) To lead a normal life	13	14.77
e) For security	5	5.68
f) Being a girl child have to get married	6	6.82
Negative	**No**	**%**
a) Because of his/her sickness	118	84.29
b) Don't want to pass it to the next generation	10	7.14
c) They won't survive that long	10	7.14
d) Might be tortur for sick children	2	1.43

Table 4.17 further shows the reasons, given by the parents in favour of marriage of their β-thalassaemic children as well as the reasons against marriage of their β-thalassaemic children. It is found that nearly 47.73 per cent of the parents are in favour of getting their β-thalassaemic children married, if the diseased children are permanently cured. It shows that these parents are virtually ignorant of the fact that this disease is genetic in nature and incurable. It is also seen that 20.45 per cent of all parents are in favour of getting their β-thalassaemic children married with the idea of continuation of their lineages. It again indicates that these parents are ignorant about the serious genetic nature of the disease and are conservative in

their outlook, and somehow they want to see that their lineage should continue without thinking of its impact on families as well as on society at large. It is found that 14.77 per cent of the parents are in favour of seeing that the diseased children are married since they feel that marriage can bring security to their diseased children; 6.82 per cent of the parents, who are having β-thalassaemic girl children, feel that the girls should get married under any circumstances. These also show the conservative outlook of some parents. However, 4.55 per cent of the parents have not yet made up their minds, but will depend on doctor's advice, though they are inclined in favour of getting their diseased children married. It shows that all these parents are either ignorant about the serious nature of the disease or they are ready to ignore all consequences for one or the other personal reason.

In the present study it is seen that there are 140 parents, who are quite aware of the serious nature of this disease and are firmly against marriage of their diseased children. 84.29 per cent of such parents are against marriage of their diseased children for the reason that the disease is incurable. 7.14 per cent of such parents do not want that such disease to pass on to the next generation. Another 7.14 per cent of the parents are sure that their diseased children will not survive for long. 1.43 per cent of the parents are

of the opinion that marriage of such diseased children means torture to these sick people.

Table 4.18

Parental Anxiety

	Always	At times	Not at all
Parental Anxiety about the diseased child	38 (92.60%)	27 (7.40%)	0

Table 4.18 shows to what extent the parents of β-thalassaemic children remain anxious about their diseased children. It is found that 100 per cent of such parents remain anxious about their diseased children. Nearly 92.60 per cent of all parents are always anxious about their β-thalassaemic children, whereas 7.40 per cent of such parents at times become very anxious.

Table 4.19

Freedom Given to the Diseased Children

	Allow At Times		Never Allowed	
	No	%	No	%
Freedom given to diseased children to move alone	203	55.62	162	44.38

Table 4.19 shows to what extent the parents of the β-thalassaemic children allow their diseased children freedom to move alone. Nearly 44.38 per cent of such parents never

allow their β-thalassaemic children to move alone freely, whereas 55.62 per cent of such parents at times allow their diseased children some freedom to move alone. However, it clearly shows that 100 per cent of such parents always live with some amount of anxiety about their diseased children.

Table 4.20

Socialization of the Diseased Children

Taking Diseased	Yes		No	
	No	%	No	%
Taking diseased child for pleasure trips	325	89.04	40	10.96
Visiting relatives/friends along with the diseased child	334	91.51	31	8.49
Attendance to social gathering along with diseased child	358	98.08	7	1.92

Table 4.20 shows the socialization process of the β-thalassaemic children. It is seen that nearly 10.96 per cent of the parents never go out for any pleasure trip with the diseased children and 8.49 per cent of them never visit their friends or relatives, along with their diseased children. It is also seen that nearly 1.92 per cent of such parents never attend any social gathering with their diseased children. It shows that a considerable number of parents is hesitant to take their diseased children for any social gatherings and

for their diseased children they are ready to sacrifice their own personal pleasures.

In this chapter, demographic compositions of the families in which there are some β-thalassaemic patients, have been described and also educational achievement of the β-thalassaemic children, economic condition of the parents, expenditure on treatment of β-thalassaemic patients, performance of the β-thalassaemic children in school and their involvement in extra-curricular activities and their socialization processes have been discussed. In this chapter psychological as well as economic problems of the parents of β-thalassaemic children have also been dealt with. In a subsequent chapter, the implications of all these findings will be further discussed.

CHAPTER-V

DISCUSSION

It is well known that with rapid advancement of medical technology various types of diseases of non-genetic origins (that is, environmental) have to a great extent been controlled and are successfully being cured. But the disorders which are of genetic origin can neither be controlled nor cured till today with advancement of medical sciences. It is true that modern medical technology has considerably brought about improvement in modes of treatment for such genetic disorders and consequently the average longevity of persons, suffering from various genetic disorders, has considerably increased. As a result the chance of survival of those individuals with various types of genetic disorders must have some serious social and *biological implications which are certainly relevant and* critical to man's future biological and social evolutions. For example, the inherited retinoblastoma the frequency of which at present is 1 in 40,000 is likely to increase to 1 in 15,000 in future after successful medical therapy (Reddy 1992). However, till today such problems have not occurred with the populations of simple culture who hardly get opportunity to take advantage of modern medical facilities.

In those societies deleterious genes are speedily removed through the process of natural selection. But the situation is not like that in modern societies which enjoy modern medical facilities. In modern society individuals with genetic disorder not only survive and enter into reproductive age group but also are likely to contribute those deleterious genes to the next generations and thereby there is every likelihood that both genetic and social load in such societies will further increase in future.

With this idea in view in the present study we have dealt with the case of homozygous β-thalassaemia patients in order to find out various physiological symptoms of disease, assessing the growth pattern of the thalassaemic children and various social and economic implications of such disease on the parents of the patients and the patients themselves. In this connection one can recall what Roberts (1975) has said, while working on genetic disorders, particularly hemophilia. He is of the opinion that in modern human societies the deleterious genes have assumed a greater importance beyond that which they confer on the life of the individuals. He has further suggested that it is not the problem of treatment, which are often very expensive and long continued, but of day to day care and maintenance, and finally he has suggested that with advancement of medical technology there are adverse effects on the society as such. As suggested earlier, in the

present study we have taken into consideration the homozygous β-thalassaemia patients and have tried to examine the various social problems cropping up not only with thee expensive mode of treatment but also various social and economic hardships on the parents of such patients.

The present study has been divided into two parts, namely biological aspects and social aspects. In biological aspects we have dealt with the symptoms of thalassaemia, to what extent the thalassaemic patients have to, visit hospitals, health condition of the thalassaemic patients before and after transfusion, requirement of operation, i.e., splenectomy, general mode of treatment of the thalassaemic patients and finally the physical growth of the thalassaemic patients.

The social aspects cover demographic composition of the families with thalassaemic patients, economic condition, psychological stresses and economic hardships of their parents. It also includes academic and extra-curricular activities of the thalassaemic patients.

In the present study we have collected data on 380 β-thalassaemic patients out of which, 14 per cent never had any visible symptoms, though on doctors' advice their parents brought them to hospitals and thalassaemia in them was detected. However, of all symptoms the most prominent ones are fever with vomitting tendency, loss of appetite, and

anaemic condition. To the best of our knowledge, nobody has ever suggested such symptoms by which a layman can suspect the presence of thalassaemia. It is well known that in homozygous β-thalassaemia there are severe anaemia and elevation of HbF and HbA_2, higher serum-iron concentration and some other haematological characteristics (Basu 1994). The point to be noted is that thalassaemia is generally detected before 1 year of age, though in many cases, perhaps due to negligence and ignorance, it is detected as late as within 5 years. This is by and large universally true.

In the present study, we have examined the frequency of hospital visits by the thalassaemic patients. It is seen very clearly that as age advances, the frequency of hospital visits by the thalassaemic patients also increases. The reason for such increased frequency of visits is that with advancement of age the thalassaemic patients start suffering more and more and they are compelled to visit hospitals in greater frequency for medical treatments and temporary comfort. However, to the best of our knowledge, no one has ever tried to examine this aspect.

In the present study we have examined the feelings of the patients before and after taking transfusions. It is well understood that transfusion from time to time is absolutely needed for any thalassaemic patient. In this connection, it may be recalled what Hardisty and Weatherall (1982) have

suggested "It has now been established quite unequivocally that homozygous thalassaemic children grow better and develop far fewer complications if their haemoglobin level is maintained as close to normal as possible... Immediately after diagnosis a full blood group genotype should be obtained and once steady-state, haemoglobin level has been reached which requires transfusion, the child should be started on a regular transfusion regina". The importance and method of transfusion for management of this genetic disorder have already been reviewed by Weatherall and Clegg (1981), Medell and Berdoukas (1981). So, we have not made any attempt to discuss the importance of blood transfusion in case of the thalassaemic patients. In the present study we have seen that nearly 64.21 per cent of all thalassaemic patients had never faced any serious problem before taking transfusion, whereas 35.79 per cent of them certainly had serious problems before taking transfusions. However, it is found that nearly 62.63 per cent of all thalassaemic patients have certainly felt an improvement in their health condition after having transfusions, whereas the rest have never felt so. Perhaps it may be due to the fact that those, who are older people and have had transfusions several times, can hardly make any difference in their health condition before and after transfusion. In this connection it may be mentioned that so far no body has made any attempt to find out the feelings of

the thalassaemic patients, about their health condition before and after transfusions. What we suggest now is that one has to look into the feelings of the thalassaemic patients regarding transfusions, though it is an established fact that transfusion is a 'must' for any homozygous thalassaemic patient.

In the present study we have made an attempt to see how menarche is delayed in the girls, suffering from β-thalassaemia major. It is seen that the mean age at menarche in case of homozygous β-thalassaemic girls is 13.82±0.49. Bhattacharjee et al. (1982) have reported that the mean age of the Bengali speaking women with β-thalassaemia trait is 13.63±0.12 years. But Bhattacharjee et al. (1977) have reported that the mean age at menarche among the normal Bengali girls is 13.21±0.04 years. It shows that in comparison to the normal girls and the girls with thalassaemia trait the mean menarcheal age in the girls with β-thalassaemia major is further delayed. Chatterjea (1965) has also made a similar observation. He has said, "the secondary sex characters appear late and menstruation is delayed and scanty" in case of girls with homozygous thalassaemia. The present study supports the findings of Chatterjea (1965).

Swarup Mitra et al. (1969), Ajmani et al. (1977) and many others have observed that usually the total haemoglobin

level is lower in those, who have abnormal haemoglobin genes and/or thalassaemia genes than in those with normal haemoglobin genotype (HbA/A). In the present study we have examined the total haemoglobin level among the thalassaemic patients by age and sex. It has been found that the total haemoglobin level in case of the male thalassaemic patients varies between 3.4 to 11.3 g/dl. and that in the female thalassaemic patients between 5.0 to 10.0 g/dl, depending on age of the patients. The WHO (1968a) has recommended that the total haemoglobin level in case of adult normal males should not be lower than 13.0 g/dl. and that for non-pregnant normal adult females 12.0 g/dl. So, in the present study, it is seen that no thalassaemic patient, male or female, can maintain that level of total haemoglobin level as recommended by WHO (1968a) Sukumaran (1975) has suggested that the mean total haemoglobin level in case of males and thalassaemia major is 4.7±0.30 g/dl. and that in females with thalassaemia major is 4.4±0.10 g/dl. In the present study it is found that the thalassaemic patients, either male or female, have slightly better mean value of the total Hb level than what Sukumaran (1975) has suggested. Das et al (1982) have suggested that among the minor male thalassaemic patients (0-14 years) the total haemoglobin level is 5.73±0.30 and that among the adult (15 years+) male thalassaemic patients is 9.98±0.30. They have further observed that the total haemoglobin level among

the minor girls with thalassaemia major is 5.98±0.62 g/dl. From that point of view in the present data the initial total haemoglobin content in the thalassaemic patients, male and female, is slightly higher. However, the present findings in no way contradict the findings of Sukumaran (1975) and Das et al. (1982) since the point to be noted is that the total haemoglobin content in the thalassaemic patients is always lower than that in the normal persons, minor or adult.

When the total Hb content in the person with β-thalassaemia major is examined after they have had some rounds of transfusion, it is found that in all cases the total haemoglobin content in thalassaemic patients, both male and female, has risen to certain level, though not to the level recommended by the WHO (1968). In this connection, it may be mentioned that Choudhury et al. (1997) have observed that there is no consistent pattern in rise of fall in the total haemoglobin content after transfusions and they have further observed that if there is rise in the Hb level after transfusion, the rise in the Hb level is not statistically significant. It may be mentioned that the UKTS News (September, 1993), quoted by the National Thalassaemia Bulletin (1993), has suggested that "Ideally the post transfusion level should be no more than 15 g/dl and pre-transfusion should be no less than 10 g/dl." However in the present study neither of the two has been observed.

The result of splenectomy have already been reviewed by Weatherall and Glegg (1981). Medell and Berdoukas (1981) and Hardisty and Weatherall (1982) have suggested that there is good evidence that young children are more prone to serious infections after splenectomy and the operation should be avoided in the first five years of life. In the present study it is seen that more than 6 per cent of the infants below 5 years of age have undergone splenectomy. It is further observed that as age advances with the thalassaemic patients splenectomy is performed more and more on them. The indications for splenectomy are on increase in transfusion requirements and it is mostly done, when physical discomfort for a large spleen increases. In the present study it is found that (combining both sexes together) nearly 22.22 per cent of the thalassaemic patients have undergone splenectomy. in the present sample majority of the patients belong to the lower income group and most of them just cannot afford the cost of operation. Consequently, in the present sample around 22 per cent of them, mostly belonging to the middle and high income groups, have gone for splenectomy. Since no such data are available we have not been able to compare the present findings with what is happening in other parts of the country.

In the present study we have not been able to collect information on differential blood count, osmotic fragility,

packed cell volume, alkali resistant haemoglobin, etc. since none of the five hospitals in Calcutta, where the present study has been done, keep those records properly. It is unfortunate that in spite of our best attempt we have not been able to study all haemotological aspects of the thalassaemic patients.

In the present study we have examined the growth patterns of the thalassaemic individuals and have made a comparison with the normal individuals. Garn (1952) has suggested that height and weight are the two body dimensions which should be taken as good indicators of physical growth. Mills (1937), Wolff (1940), Grenlich (1957) and others have suggested that human physical growth is influenced by many factors including nutrition, genetic constitution, etc. Sarkar et al. (1984) have observed that haemoglobin genotypes and also β-thalassaemic genotypes do not have much influence on height and weight in adult population. Sanctis and Pinamonti (1995) have observed that pathogenesis of growth retardation in thalassaemic patients is complex and incompletely understood. They have suggested some factors like chronic anemia, endocrine disease, defeseral toxicity, etc. as the main factors affecting growth. However, in the present study we have taken into consideration 8 anthropometric and 5 skinfold measurements to understand the growth pattern of the β-thalassaemic individuals. When we

compared each of the measurements, taken on the thalassaemic individuals, male and female separately, with the normal male and female individuals by age-groups it is seen that the growth of the thalassaemic individuals in almost each of these measurements, is significantly lower in comparison to those in the normal individuals. It is further observed that the rate of increment in physical growth of the β-thalassaemic individuals is much slower than that of the normal individuals. It is true that there are a few exceptions in one or the other measurements. But it can be said that the physical growth of the β-thalassaemic persons is certainly retarded at least upto 15 years of age. It is further observed that after 15 years of age the rate of increment in physical measurements of the thalassaemic persons certainly improves, though it never catches up the rate of growth of the normal persons. The present findings do not corroborate with the observations, made by Sarkar et al. (1984). In this connection it may be noted that Johnston and Crogman (1964) have studied the pattern of growth in children with thalassaemic major in the USA. They have inferred that there is a retardation in normal growth expectation and a retardation in the rate of growth. They have further observed that a growth curve drawn on the measurements of the thalassaemic children looks similar to that drawn on the normal but not quite reaching normal values. The present

study fully corroborates the findings of Johnston and Crogman (1964).

As mentioned earlier in the present study we have tried to examine some social implications of thalassaemia disease.

In the present study we have seen that generally the fertility level of the mothers with thalassaemic children is very low. The reason is perhaps that most of the thalassaemic children were born between first and third birth orders. Consequently, the parents generally do not look for larger family size. But the reproductive wastage among the mothers with thalassaemic children is fairly high. However, it is seen that there is an inverse relationship between occurrence of thalassaemia and number of birth order, i.e. as the number of birth order increases, the probability of birth of a baby with thalassaemia decreases.

Since thalassaemia is a genetic disorder there is no correlation between occurrence of thalassaemia and educational standard of the parents. However, when parents are better educated they try to restrict their families after birth of a thalassaemic baby.

When economic condition of the families in which there are thalassaemia babies, is classified into 3 groups - low income group, middle income group and high income group, an interesting point emerges. It is found that the frequency of thalassaemic children is more in middle income and high

income families than that in the lower income group. The reason for this is probably that in the low income group many of the thalassaemic babies die early since the parents belonging to the low income group just cannot afford to give their diseased children proper medical treatment, which is very expensive and long continued. On the other hand the parents in the high income group are certainly in a better position to provide better medical treatment to their thalassaemic children. The parents, belonging to the middle income group, may not be that financially sound like the people of the high income group, but even then they do certainly try their best to provide proper medical treatment as far as possible to their thalassaemic children, in spite of their severe economic hardship.

In the present study it has been seen that the average expenditure on the thalassaemic children incurred by the parents of high income group is much higher than that incurred by the parents, belonging to the middle income group. The average expenditure per month on a thalassaemic child is found to be lowest in the low income group families. It is certainly understandable that the parents, belonging to the low income group, just cannot afford so much expensive treatment for their thalassaemic children, and consequently many of their children must have died in early age. In Table-4.9, it is presented the data on economic hardship, faced by

the parents of the thalassaemic children. It is found that nearly 95 per cent of all the parents, belonging to the low income group, are facing tremendous economic hardship to maintain their thalassaemic children. Even among the parents, belonging to the middle income group nearly 67 per cent of them have admitted such economic hardship, whereas nearly 35 per cent of the parents, belonging to high income group, have had such economic burden for the maintenance of their thalassaemic children. It clearly shows that lower the economic condition of the parents, higher is the economic burden on them for the maintenance and treatment of their thalassaemic children. In this connection one may recall what Tyler et al. (1982) have observed in England while studying the socio-economic burden of Huntington Chorea in South Wales. They have also observed that as the age of the person, suffering from Huntington Chorea, advances, cost of treatment also increases. It creates a serious economic burden on the family. However, in a place like England also where there are a lot of social security for the ailing persons, the families are facing tremendous economic burden for the treatment of their family members, suffering from Huntington's Chorea.

In a place like Calcutta, where there is hardly any social security for the diseased people, the entire burden of their treatment is borne by the family members. Consequently, one can well assume that the parents, belonging to the low

income group, have to deprive their normal children with a view to meeting all expenses for the treatment of their thalassaemic children. The same is by and large true for the parents, belonging to the middle income group.

So far the thalassaemia patients are concerned, it is found that nearly 29 per cent of them do not go for schooling, perhaps for their bad health condition. It is further observed that as age of the thalassaemia patients advances, the frequency of attending schools drops sharply. Generally, they do not go beyond the secondary level. It is found in the present study that nearly 50 per cent of the thalassaemic patients do not perform satisfactorily in school. Perhaps due to their poor health condition they have to take time off often from their studies. It may be assumed that for their poor educational performances it may, in future, create problems for them to find out suitable jobs, and that in turn may create further economic burden on them.

It is further observed that so far physical activities of the thalassaemic children are concerned, nearly 20 per cent of them have become inactive by 15 years of age and nearly 40 per cent of them by that age cannot perform heavy physical activity. It shows that altogether nearly 60 per cent of the thalassaemic patients become dependent for their day to day work on others. It indirectly creates problems not only on the thalassaemic patients themselves but also on

other normal members of their family. It may be noted that in England most of the children with haemophila are engaged in little or no physical exercise (Roberts, 1975). The same is by and large true for the thalassaemic children here.

In the present survey data on parents' idea of getting their thalassaemia children married have been collected. It is found that nearly 25 per cent of the parents having thalassaemic children firmly believe that their thalassaemic children should marry, whereas 38.36 per cent of them have expressed their opinion against marriage of such thalassaemic children. Nearly one-third of the total parents till the time of the present survey are found to be undecided. It shows that a good number of parents are ignorant about the serious genetic nature of this disease and many of them are found to be conservative in their outlook since they believe in all traditional system.

In the present survey it is found that nearly 93 per cent of all parents remain always extremely anxious about health condition of their thalassaemic children, whereas 7 per cent of such parents at times become anxious of their diseased children. However, it shows that nearly 100 per cent of all parents spend their time in anxiety for their thalassaemic children. Consequently, nearly 100 per cent of the parents never allow their thalassaemic children to move

freely, though 56 per cent of the parents rarely allow their thalassaemia children some freedom for moving alone.

In the present study, it is found that nearly 11 per cent of the parents with thalassaemic children never go out for any pleasure trip, nearly 9 per cent of them never take their thalassaemic children to visit their relatives or friends and 2 per cent of them never attend any social gathering along with their thalassaemic children. It shows to what extent the parents are depriving themselves and their other normal children from pleasures of life and many of them avoid visiting friends/relatives/or any other social gathering with thalassaemic children since they feel embarrassed to explain to others every now and then of health condition of their diseased children. Roberts (1975) has reported that nearly 50 per cent of the families with hemophiliac children never go out for holidays, considering the health condition of their diseased children and a fairly good number of mothers are unable to take part in any social activities outside their house. The situation is by and large same for the families with thalassaemic children.

In the present study, we have dealt with some biological aspects of β-thalassaemia and tried to find out some possible social implications of such serious genetic disorder. However, there are a lot of limitations in the present study, for example:

1) We have not tried to find out the present frequency of thalassaemia gene among the Bengalees. It has been reported by Dr. S. Majumder (*Statesman*, Calcutta dated 14.2.94) that the frequency of the thalassaemic gene is about 10 per cent among the Bengalees at present. It shows that the frequency of thalassaemia gene among the Bengalees has steadily been increasing over the last 40 years, since the time, Chatterjea et al. (1954) reported the frequency of thalassaemic gene was around 3 per cent. It needs an urgent study to determine the exact frequency.

2) The present study is confined only among the thalassaemic patients aged upto 20 years. As a matter of fact the average longevity of the thalassaemic patients have at present increased to 30/35 years (Ghosh, 1996). It shows that many of the thalassaemic patients, with the help of modern medical technology, now enter not only into the reproductive age group but also marry and reproduce. In this connection, it may be noted that Das et al. (1983) have reported that the reproductive fitness of the β-thalassaemic patients is now around 0.03, which means that it has increased from 0 to 3 per cent. It shows that the rate of segregation of the thalassaemic gene has increased to a great extent in view of the fact that previously the segregation of such genes used to take place only through heterozygotes and now both homozygotes and heterozygotes are passing such thalassaemic

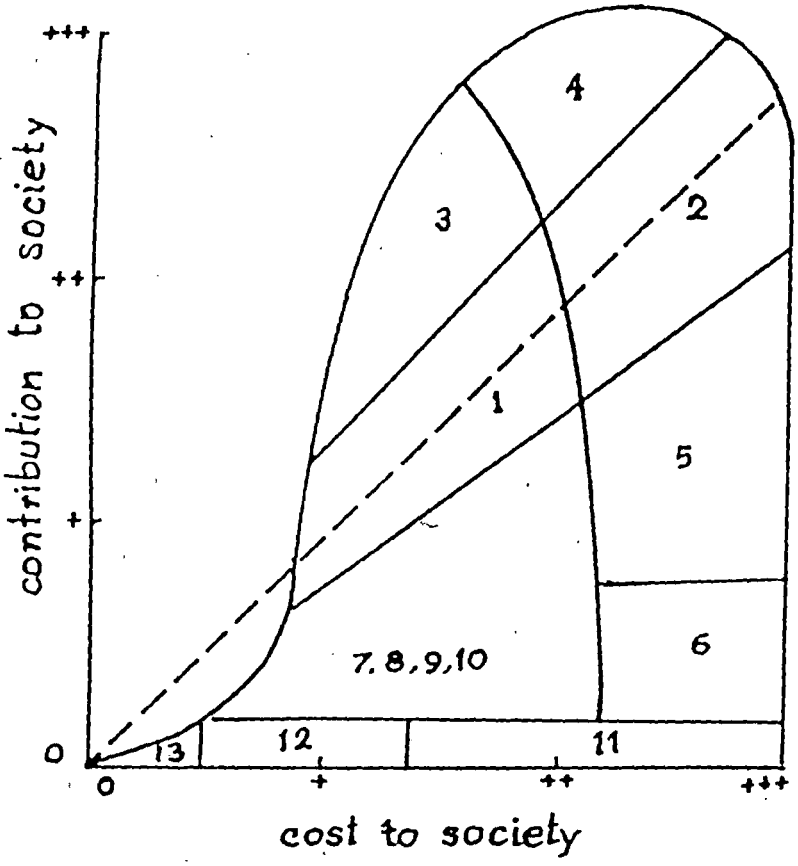

Fig. 33: Social fitness of human genotypes.

genes to the next generations. It warrants an immediate study to find out the exact rate of segregation of the thalassaemic gene.

3) In the present study we have not included the married thalassaemic persons. Consequently, it has not been possible for us to find out the social implications of this genetic disorder on conjugal life.

4) In the present study, we have not been able to collect data on all haematological parameters before any treatment given to a thalassaemic patient. It is also needed that such data on haematological parameters must be looked into after giving treatment to the thalassaemic patients. It will help us to understand to what extent modern treatment can remove discomfiture of the thalassaemic patients.

As mentioned above there are some serious lacunae in the present study, which we shall try to make up in our future study.

In this connection, one can mention, the observations, made by Wright (1960), on social fitness of human genotypes. It is admitted that evaluation of social fitness of human genotype is very difficult. However, Wright has treated the problem in terms of the balance between the contributions to society by individuals of different genotypes and what they cost to the society. Figure-33, adopted from wright (1960) shows the following categories of individuals:

1) Those - who contribute to the society approximately what they cost at relatively modest levels. It includes most of the people - ordinary persons, law abiding citizens, etc.

2) Those - in whom a balance between contribution and cost at relatively high levels. It includes professional persons, technicians, experts, specialists on whose education, society invests heavily, and who receive standard of living above the average by dint of their labour.

3) & 4) Those - make extra-ordinary contribution at either low or high costs to the society. For example, it includes high class artists or experts, persons of genius and others.

5) Those persons - whose capacities are like those in the first and second category. But they repay to the society much less than the cost of their maintenance.

6) It includes criminal and anti-social persons who are otherwise of normal mental capacity.

7) Those - who have sub-normal physical constitution and health. It seems that most of the thalassaemic patients should be included in this category since society pays a lot for them and their return to the society is minimal.

8) Those persons - who are of low intelligence but sufficient to take care of themselves under existing social conditions.

9) & 10) Those persons - who are normal in childhood, but experience early mental or physical breakdown. In this category many of the more severe hereditary diseases like Huntington's chorea come.

11) Those - who are incapacitated physically or mentally throughout life.

12) Those - who die before maturity. It means that they have no opportunity to make any appreciable contribution to society.

13) Those - who die at or before birth.

It is seen that each of these categories includes a great number of genotypes. The categories 1 to 5 are the normal persons, who keep society going and the rest i.e. 6 to 13 in which social cost is much more than returns and they become burden on society. Most of the genetic disorders have become heavy burden on society.

In the present study we have seen that with the development of medical technology the mode of treatment for thalassaemic patients has tremendously improved, and consequently, it may be assumed that many of them are surviving for quite some time and entering into reproductive age group. But there is very little opportunity for them to make any positive contribution to the society. On the other hand, the society is paying heavily on welfare and physical well-being of those thalassaemic patients. It is not

unreasonable to assume that with advancement of medical technology, the rate of segregation of the thalassaemic gene, as suggested by Das et al. (1983), is increasing in the population which means genetic load is accumulating at faster rate in the society. It is well known that if genetic load increases in society, social load is bound to increase concomitantly in view of the fact that society will have to maintain many more thalassaemic patients than it used to do a few decades earlier.

With all these ideas we may suggest some measures by which we can prevent the spread of this genetic disorder and by the process we can reduce to a great extent the social load. The suggestions are as follows:

1) The government must come forward to take up the responsibility of all medical treatment for all thalassaemic patients in this country as it is normally expected that any welfare state would do.

2) Alternatively, if the government itself cannot take up this responsibility the government should reimburse the parents the whole amount spent by them for the treatment of their thalassaemic children.

3) The government must introduce immediately a national register for all thalassaemic patients in this country.

4) The government should make all efforts to encourage and motivate people to go for marriage counselling.

5) The government should make a legislation to prevent marriage between two heterozygotes, i.e., those who are the carriers of this deleterious gene and also the legislation should prevent the thalassaemic patients from marrying.

With this idea in future we hope to make a further study on the β-thalassaemic patients and that may help us understand the biological and social implications of this dreaded disease.

CHAPTER-VI

SUMMARY

MAJOR FINDINGS:

Biological Aspects

1) Symptoms: The symptoms before detection of thalassaemia show that 17.87 per cent of all males and 19.08 per cent of all females had only fever and 16.03 per cent of females and 12.77 per cent of males suffered from fever and loss of appetite. 19.08 per cent of females and 14.89 per cent of males had fever with vomitting tendency and loss of appetite. About 20 per cent of males and 12.21 per cent of females suffered from fever with cold and cough, loss of appetite. 19.08 per cent of females and 14.47 per cent of males suffered from fever with vomitting tendency, loss of appetite, besides anaemic condition.

2) Out of 380 thalassaemic patients, 174 (45.79 per cent) were detected before one year of age. In case of 171 (45 per cent) individuals it was detected by five years of age. It means 90.79 per cent of all the patients it was diagnosed before five years of age.

3) It is found that the majority (i.e. 40.26 per cent) of the patients visit hospital twice in a month. It is

seen that the patients aged 6 to 10 years, visit hospitals much more regularly.

4) Regarding health condition before and after transfusions it is found that out of 380 thalassaemic patients 136 individuals had serious health problems before transfusion and 244 individuals had no such serious problem. So far as the health status after transfusion is concerned, 62.63 per cent of all thalassaemic patients felt an improvement in their health condition and 37.37 per cent had not found any difference in their health status.

5) It has been found that the mean age at onset of menarche among the girls with β-thalassaemia major is 13.82±0.49 years, and that in the girls with β-thalassaemia trait is 12.63±0.12 years and for the normal girl it is found to be 13.21±0.04 years. It shows that the onset of menstruation is delayed in case of the homozygous β-thalassaemia girls.

6) Mean total haemoglobin level before transfusion shows:

Age group	Male	Female
Upto 5 years	7.54±0.27 g/dl	8.31±0.23 g/dl
6-10 years	8.34±0.99 g/dl	7.58±0.20 g/dl
11-15 years	7.08±0.22 g/dl	6.94±0.37 g/dl
16-20 years	7.51±0.68 g/dl	6.61±0.37 g/dl

It shows that the mean total haemoglobin is much lower in the β-thalassaemia patients than that in normal persons recommended by the WHO (1968).

7) The mean total haemoglobin level after transfusion shows:

Age group	Male	Female
Upto 5 years	10.44±0.38 g/dl	10.87±0.40 g/dl
6-10 years	9.45±0.26 g/dl	10.06±0.27 g/dl
11-15 years	9.26±0.30 g/dl	9.29±0.45 g/dl
16-20 years	8.65±0.41 g/dl	8.80±0.46 g/dl

It shows that even after having repeated transfusions the total haemoglobin level in the β-thalassaemic patients male or female, never reached the recommended level.

8) Performance of splenectomy shows:

Age group	Male			Female		
	No	Yes	%	No	Yes	%
Upto 5 years	48	3	6.25	30	2	6.67
6-10 years	61	12	19.67	38	10	26.32
11-15 years	31	16	51.61	14	4	28.57
16-20 years	6	2	33.33	6	3	50.00

It shows that as age advances, more and more thalassaemic patients had to undergo splenectomy.

Altogether 13 anthropometric measurements including 5 skinfold thickness have been taken on 380 (245 males and 135

females) β-thalassaemic patients and 364 (236 males and 128 females) normal individuals aged between 3 and 20 years.

9) In all measurements it is found that the rate of growth in the β-thalassaemic patients is significantly lower than that in normal individuals, excepting in bicondylar femur diameter, where the thalassaemic patients are having higher value than the control sample.

10) It is found that the rate of increment in physical growth of the thalassaemic patients (boys and girls) is lower than that in the normal individuals, though there are a few exceptions.

11) It is found that physical growth of the thalassaemic patients is retarded upto 15 years of age and thereafter though the rate of increment in physical measurements improves, the thalassaemic patients never catch up the normal individuals in physical growth.

12) In the present study we have described the mode of treatments of β-thalassaemia major. It is found that the average cost of treatment of thalassaemia patients comes around Rs.5445/-. It is seen that majority of the thalassaemic patients in the present sample just cannot afford so much money for treatment. Consequently most of them are not having full course of treatment.

Social Aspects

13) It is seen that out of 380 thalassaemic patients, 245 (64.47 per cent) are males and 135 (35.53 per cent) are females. It is also seen that most of the thalassaemic patients, either male or female belong to the age group 6-10 years. It has also been noticed that with advancement of age, particularly after 15 years, there is a decrease in the frequency of thalassaemia patients.

14) The distribution of the thalassaemic patients by religion/caste shows that it is more or less equally prevalent among all castes and religious groups.

15) The reproductive history of the mothers of the thalassaemic patients shows that the mean number of live births is highest (3.32 per cent) among the Muslims, followed by the Hindu Mahisya (2.71 per cent). It is also found that the mean number of surviving children is the highest among the Muslim (2.84±0.33) and the lowest (1.60±0.22) among the Christian. The present study shows that the frequency of dead children varies between 0.16±0.12 in the Sadgope and 0.52±0.12 in the Muslim. A very high frequency of abortions has been noticed in the study, which varies from 7.96 per cent to 22.33 per cent among the mothers of the thalassaemic children.

16) It is seen that nearly 52.31 per cent of all thalassaemic patients belong to the first birth order. It has

also been observed that there is an inverse relationship between the occurrence of the thalassaemia and number of birth order. It shows that as the number of birth order increases, the probability of birth of a baby with thalassaemia decreases.

17) Educational achievement of the parents shows that 7.67 per cent are illiterate, whereas 29.32 per cent are graduates. Majority of the parents (45.76 per cent) have had education upto the secondary level.

18) Economic condition of the families with thalassaemia patients shows that 50.74 per cent of the families belong to the lower income group, 26.11 per cent to the middle income group, and 23.15 per cent to the high income group.

19) The frequency of thalassaemia children in relation to economic condition shows that out of 350 thalassaemic children 179 belong to the low income group, 90 belong to the middle income group, and 81 of them to the high income group.

20) It is found that the mean expenditure per month per thalassaemic patient in the low income group is Rs.714.39±35.44. In the middle income groups Rs.1082.33±63.55 and in the high income group Rs.2158.64±80.74. Combining all the patients of three economic groups together, it is found that the average expenditure on treatment of the thalassaemic patients per month is Rs.1143.24±57.43.

21) It is found that nearly 95 per cent of all parents, belonging to the low income group, are facing tremendous economic hardship to maintain their thalassaemic children. Even among the parents of the middle income group nearly 67 per cent of them have admitted such economic hardship, whereas nearly 35 per cent of the parents belonging to the high income group have had such economic burden on them for maintenance and treatment of their thalassaemic children. It clearly shows that lower the economic condition of the parent, higher is the economic burden on them.

22) Educational achievement of the thalassaemic children shows that out of 373 patients 2365 (71.67 per cent) have been attending school whereas 108 (28.95 per cent) have not attended any school. It further shows that with advancement of age the frequency of thalassaemic children's going to school decreases.

23) Educational achievement of the thalassaemic patients shows that 20.35 per cent of males and 18.82 per cent of females have been going to the pre-primary schools whereas 51.74 per cent of males and 50.54 per cent of females are in the primary schools. Similarly, 23.84 per cent of males and 29.03 per cent of females are in the secondary level. However, it shows that the thalassaemic patients generally do not go beyond the secondary level, which may be

due to various health problems, faced by them since childhood.

24) Educational performances of the thalassaemic patients show that irrespective of age groups the male thalassaemic patients have been performing more satisfactorily than their female counterparts. It further shows that nearly 50 per cent of all thalassaemic patients just cannot do well in their academic pursuits which may be again due to various health problems.

25) It has been found that nearly 93.75 per cent of the males and 84.96 per cent of the females participate in games. It shows that boys take more part in games than the girls. It has also been found that both boys and girls aged upto 15 years, take more part in games in comparison to those who are above 15 years of age.

26) Physical activities of the thalassaemic patients show that nearly 41.67 per cent of male and 36.09 per cent of female thalassaemic patients are physically active. It further shows that with advancement of age activities of the thalassaemic patients gradually declines.

27) Opinion of the parents regarding marriage of the thalassaemic children shows:

In favour	Against	Undecided	Depends on Children
88(24.11)	140(38.36)	117(32.05)	20(5.48)

28) In the present study it is found that nearly 93 per cent of all parents remain always extremely anxious about the health condition of their diseased children. Only 7 per cent of the parents become anxious at times. However, it shows that nearly 100 per cent of such parents spend their time in anxiety for their thalassaemic children. Consequently, nearly 100 per cent of the parents never allow their thalassaemic children to move freely, though 56 per cent of the parents rarely allow their thalassaemic children some freedom to move alone.

29) It is found that nearly 11 per cent of the parents with thalassaemia children never go out for any pleasure trip. Nearly 9 per cent of them never take their thalassaemic children to visit their relatives or friends and 2 per cent of them never attend any social gatherings along with their thalassaemic children.

We have seen in the present study how recent medical advancement has brought about an improvement in the mode of treatment for the β-thalassaemic patients. Consequently, the average longevity of the β-thalassaemia patients has increased considerably, as a result of which a good number of β-thalassaemic patients not only enter into the reproductive age groups but also they have started contributing the deteriorious genes to the next generation. As a result the genetic load in the population is steadily increasing on the

other hand it is seen that the thalassaemic patients have very little opportunity to contribute to the society. it is well known that society spends a lot for maintenance of such thalassaemic patients. Consequently, it shows that as genetic load increases in the society, social load concomitantly increases. We have made certain suggestions to decrease both genetic load and social load. The suggestions are as follows:

1) The government must come forward to take up the responsibility of all medical treatment for all thalassaemic patients in this country as it is normally expected that any welfare state would do.

2) Alternatively, if the government itself cannot take up this responsibility the government should reimburse the parents the whole amount spent by them for the treatment of their thalassaemic children.

3) The government must introduce immediately a national register for all thalassaemic patients in this country.

4) The government should make all efforts to encourage and motivate people to go for marriage counselling.

5) The government should make a legislation to prevent marriage between two heterozygotes, i.e., those who are the carriers of this deleterious gene and also the legislation should prevent the thalassaemic patients from marrying.

With this idea in future we hope to make a further study on the β-thalassaemic patients and that may help us understand the biological and social implications of this dreaded disease.

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