



FEDERATION of Indian Thalasseemics

National Thalassemia Bulletin

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Thalassemia

is

Preventable

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Support

National Update on Thalassemia October 7-8, 1995, Chandigarh



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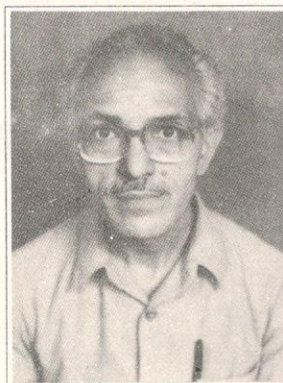
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EDITORIAL

Large number of parents of Thalassemic children were excited with the news item which appeared in the Hindustan Times on 23rd December, 1995.

Blood stem cell transplant is akin to bone marrow transplant and is not any new development.

Thalassemia is genetic inherited disorder resulting in a defect in the stem cells. Bone marrow transplant offers cure as the defective stem cells of thalassemic patient are replaced by the normal stem cells from HLA matched sibling. Possibility of having HLA matched sibling is nearly 35 percent, while the possibility of getting HLA matched donor in a general population is nearly one in million. Therefore, all over the world bone marrow transplantation is being done with HLA matched sibling. As large number of patients needing bone marrow transplantation cannot undergo BMT, therefore bone marrow registries have been started in many countries in an effort to find the HLA matched donor from the general public. Such match if available is called unrelated donor.

With the advent of GROWTH FACTORS (GF) the stem cells from the bone marrow can be easily mobilised in adequate numbers which can be collected with the use of cell separators. These stem cells can be transplanted in the recipient. Procedure of BMT or stem cell transplant remain the same. The only advantage of stem cell transplant is that donor need not undergo repeated bone marrow aspiration for collection of stem cells from the bone marrow. Any centre undertaking BMT can easily perform the stem cell transplantation. Thus the Singapore event is not a new development to offer a cure for Thalassemia.

We would like to extend our heartiest congratulations to the team of dedicated doctors of Singapore General Hospital on their historic feat.

Dr. V.P. Choudhary

GROWTH AND DEVELOPMENT IN THALASSEMIA MAJOR

— V. De Sanctis, A. Pinamonti



Growth begins when a baby is conceived, and continues throughout life. It is complex process, influenced by a variety of factors that are only beginning to be understood.

Cause of growth failure include

1. genetic factors
 2. intrauterine insult
 3. environmental factor
 4. chronic illness
 5. endocrine deficiency and hormonal abnormalities
- inherited disease of skeletal growth.

The endocrine system influences growth through the activity of growth hormone (GH) and other hormones. Growth hormone-releasing factor is produced by the hypothalamus and crosses the hypothalamic-pituitary portal system to stimulate the anterior pituitary gland to release growth hormone (GH). Growth hormone in turn is believed to exert part of its promoting effect through the mediation of somatomedins, also known as insulin-like growth factors (IGF).

Somatostatin produced in the hypothalamus inhibits while GRF stimulate GH secretion, respectively.

Because children grow at characteristic rates at different ages, an understanding of pattern of normal growth is a fundamental prerequisite for the evaluation of abnormal growth. In order to judge whether a child has normal growth and maturational development, standard normal curves are required. Since there are great differences in growth rates, adult heights, and age of puberty onset among different ethnic groups, it would be of great advantage to use standards based on ethnic background for each child assessed. Unfortunately, such curves are not available.

The Tanner-Whitehouse charts, which are based on English children, are widely used internationally.

Accurate measurement of stature requires strategies appropriate to the patient's age.

Supine length is generally taken from birth until two years.

In older children, the preferred method of measurement utilizes a precision-built stadiometer.

In absence of a commercially available stadiometer, a suitable alternative can be made at a modest cost.

Segmental measurements, such as measurement of the trunk may yield the first indication of skeletal dysplasia.

The bone age may be delayed for a variety of reasons so it is not very helpful in finding the cause of short stature.

ETHIOPATHOGENESIS OF GROWTH RETARDATION

The pathogenesis of growth retardation in thalassemic patients is complex and incompletely understood. The main factors affecting growth are listed below:

- Chronic anaemia
- Endocrine disease
- Chronic liver disease
- Iron overload
- Zinc deficiency
- Desferal toxicity

Since the management of thalassemia has changed progressively over years some factors responsible for growth retardation, such as anaemia and folic acid deficiency, are no longer important because they are corrected by transfusions. (In India in almost all the towns & even in metropolitans growth retardation can still be seen by chronic anaemia and folic acid deficiency.)

Iron overload resulting from repeated blood transfusions is always present in thalassemia in spite of iron chelation therapy, and has been thought to be the cause of the endocrine abnormalities. This is supported by histological studies of different endocrine glands. Iron toxicity in the tissue cells is a complex process involving free radical formation and lipid peroxidation resulting in mitochondrial, lysosomal and sarcolemmal membrane damage.

Other possible factors contributing to the growth retardation are:

a) individual susceptibility to the toxic effects of iron overload, b) chronic liver disease with alteration in the hepatic generation of IGF-I, c) zinc deficiency and d) a direct effect of desferrioxamine (Desferal, DFX; Ciba-Geigy) on the bones. In addition, a recent experimental observation suggested that increased amounts of circulating free haemoglobin may inhibit cartilage growth.

ENDOCRINE DISORDERS

GH-IGF-axis

Although growth hormone secretion appears to be normal in many thalassemic patients with growth retardation, there is evidence indicating an impaired growth hormone secretion in some of them.

A list of the classification of the GH-IGF disorders is given below:

1. Growth Hormone Insufficiency
2. Hypothyroidism
3. Delayed Puberty & Hypogonadism

We do not know with certainty how common this endocrine pathology is.

Growth Hormone Insufficiency

In 1990 Dr. Torresani measured urinary excretion of GH in 38 of our thalassemic patients from Ferrara, aged 7 to 24 years. A GH excretion similar to that in non thalassemic patients with GH deficiency was found in 12 patients (in 20% of the patients between 7 and 12 years, in 40% between 12 and 15 years and in 25% over 15 years):

Treatment with recombinant hGH from mammalian cells was given for a year to 15 thalassemic patients (8 males and 7 females) with short stature, reduced peak response of GH (below 10ng/ml) after stimulation with arginine and clonidine respectively,

The results of growth velocity for the year before, during and after treatment are:

Responders — Growth velocity 4 cm per year greater than the previous year;

Partial Responders — Growth velocity equal to 2-4 cm per year above the previous year;

Non-Responders — Growth velocity less than 2 cm per year above the previous year.

Hypothyroidism

This endocrine complication is common in patients who are anaemic and/or in poorly chelated but it is rare in patients who are regularly transfused and chelated with DFX. In fact in the last 10 years there have been no new cases of primary hypothyroidism among our patients.

Delayed puberty and hypogonadism

In a collaborative study of the endocrine complications in 2535 thalassemic patients followed in 29 Pediatric and Haematology Departments throughout Italy, lack of pubertal changes was the most common finding in the patients. 48% of males and 39% of females over the age of 15 years were prepubertal. The Hypothalamic-pituitary-gonadal axis in our patients assessed by functional studies has shown that thalassemics who develop hypogonadism have anterior pituitary damage with intact gonads. However, today the majority of well chelated young thalassemic patients can expect to achieve full sexual maturity and enjoy the physical, emotional and psychological benefits of adolescence and adulthood.

ZINC DEFICIENCY

Thalassemia major may have a mild zinc deficiency, particularly if they have an abnormal glucose metabolism.

Zinc is required for many metabolic activities which

are essential for DNA synthesis, cell division and protein synthesis.

DESFERIOXAMINE TOXICITY

DFX therapy is essential to prevent the deleterious effects of iron overload in heart liver and endocrine glands. In the last 7 years a reduction in growth velocity and rickets-like skeletal radiological changes associated with joint stiffness have been observed in 12 thalassemic patients, aged from 3 to 16 years, who were regularly chelated (50-70 mg/Kg/d, 5-7 times/week) during childhood and pubertal maturation.

The pathogenesis of toxic lesions of DFX is not yet clear. Different mechanisms may be postulated: a) chelation of the trace elements, b) direct effect of the drug and its metabolites, c) inhibition of cellular proliferation.

CONCLUSIONS

In chronic disease, such as thalassemia, there is more than one growth factor which can cause growth retardation. Not all of the factors are operative in every child at any one time, but many of the factors are interrelated.

Iron overload in different endocrine glands and liver damage, caused by viral infection, in the presence of iron overload, may play a role in the development of endocrine disorders. If this is correct, prevention of endocrine complications also depends on prevention of liver infections.

Our study indicates that an ideal therapeutic regime, which both prevents the toxic effect of iron overload and does not lead to a toxic effect of continuous subcutaneous DFX chelation therapy, has not yet been found, and therefore additional studies and long term follow up are needed.

थैलासीमिया मेजर में वृद्धि व विकास

प्रो. वी. डी. सैंक्टिस

भूणावस्था से आरंभ हो कर जीवन भर वृद्धि का क्रम चलता रहता है। यह एक बहुत ही विषम तरीका है जोकि कई कारणों से प्रभावित होता है।

वृद्धि में रुकावट के निम्न मुख्य कारण हैं:-

1. अनुवांशिक
2. गर्भावस्था का तिरस्कार
3. पारिवारिक परिस्थितियां।
4. जीर्ण रोग
5. अन्तःस्रावी ग्रन्थियों की विकृति
6. कंकाल तंत्र संबंधी अनुवांशिक कारण।

“ग्रोथ हारमोन” (GH) व अन्य “हारमोनस” द्वारा अंतःस्रावी ग्रन्थियां शरीर की वृद्धि को प्रभावित करती हैं। ग्रोथ हारमोन रिलीजिंग हारमोन (GRF) हाइपोथैलमस में बनता है तथा पिट्युटरी में जाकर एंटीरीयर पिट्युटरी ग्लैंड को उत्तेजित करके ग्रोथ हारमोन को निकालता है। GH का कुछ प्रभाव सोमाटोमेडिन

के द्वारा पड़ता है जोकि इंसुलिन प्रकार के वृद्धि जनक कारक (IGF) है।

हाइपोथैलमस से ही उत्पन्न सोमाटोस्टेटिन GH को रोकता है जबकि GRF उसको उत्तेजित करता है।

थैलासीमिया के रोगियों में वृद्धि की रुकावट एक बहुत ही पेचीदा प्रश्न है जिसकी पूर्ण जानकारी प्राप्त नहीं है। इनमें वृद्धि की रुकावट के निम्न मुख्य कारण हैं:-

1. देर तक रहने वाली खून की कमी
2. अन्तःस्रावी रोग
3. जिगर के पुराने रोग
4. लोहे की अधिकता
5. जस्ते की कमी
6. डैस्फराल की विषाक्तता

अब पहले से बेहतर रक्त संचारण की सुविधा प्राप्त है अतः खून की कमी व Folic Acid की कमी के कारण वृद्धि में रुकावट कम

देखने को मिलती है। (यद्यपि भारत में लगभग सभी छोटे शहरों व कुछ हद तक बड़े शहरों में भी इन कारणों से वृद्धि में रुकावट बहुतायत में पाई जाती है।)

लोह निष्कासक दवा देने के बावजूद थैलासीमिकस में लोहे की मात्रा अधिक ही पाई जाती है। अन्तःस्रावी ग्रन्थियों में विकृति का यह मुख्य कारण है इसकी पुष्टि इन ग्रन्थियों की जांच से हो जाती है।

अन्य मुख्य कारण:-

- ❖ व्यक्ति विशेष की लोह की अधिकता प्रति ग्रहणशीलता
- ❖ हाल ही में एक कारण और जो देखा गया है वह है कि रक्त में मुक्त हीमोग्लोबिन की अधिकता, यह अस्थियों के ऊपर की उपास्थियों की वृद्धि में रुकावट डालता है।

अन्तःस्रावी विकृतियां

यद्यपि बहुत से थैलासीमिकस में ग्रोथ हारमोन उचित मात्रा में निकलता है तथापि कुछ रोगियों में यह कम स्रवित होता है।

GH-IGF से संबंधित विकृतियां निम्न प्रकार हैं :-

1. ग्रोथ हारमोन की कमी
2. थायराइड की कमी
3. किशोरावस्था का देरी से आरम्भ व जननांगों के विकास में कमी।

ग्रोथ हारमोन की कमी

1990 में डा. टोरेसनी ने 38, 7-24 वर्ष के थैलासीमिकस के मूत्र में GH की मात्रा नापी। इनमें से 12 रोगियों (जिसमें 20% 7-12 वर्ष, 40% 12-15 वर्ष तथा 25% 15 वर्ष से बड़े थे) में GH की उतनी ही कमी पाई गई जितनी कि थैलासीमिया रहित GH की कमी से प्रभावित व्यक्तियों में पाई जाती है।

15 थैलासीमिया रोगियों (8 पुरुष व 7 स्त्री) जिनका कद छोटा था व आर्जीनीन व क्लानीडीन से उत्तेजित करने उपरान्त GH का अधिकतम असर 10ng/ml से कम था उनमें Recombinant hGH का प्रयोग किया गया। इसका निम्न प्रभाव देखने में आया :-

Responders — पिछले वर्ष की अपेक्षा Growth Velocity 4 CM अधिक हुई।

Partial Responders — पिछले वर्ष से 2-4 CM अधिक वृद्धि

हुई।

Non-Responders — पिछले वर्ष की अपेक्षा 2 CM से भी कम वृद्धि हुई।

थायराइड की कमी

यह विकृति उन रोगियों में अधिकतर देखने में मिलती है जिनमें खून की कमी रहती है या लोहे की मात्रा अधिक होती है। यदि रक्त संचारण समय पर किया गया हो व अतिरिक्त लोहे को पूरा निकाला गया हो तो यह दोष नहीं पाया जाता।

प्रजननांगों के विकास में कमी व किशोरावस्था के आगमन में देरी

इटली में 2535 थैलासीमिकस पर पाया गया कि 48% लड़के व 39% लड़कियां 15 वर्ष तक भी किशोरावस्था को प्राप्त नहीं कर पाये थे। प्रजननांगों में विकास न होने का कारण anterior pituitary gland में क्षति का पाया जाना था। जबकि अब लोह निष्कासक दवायें उपलब्ध हैं। अतिरिक्त लोह रहित थैलासीमिकस में शारीरिक व मानसिक रूप से पूर्ण स्वस्थ किशोरावस्था व व्यस्कता का पाया जाना संभव है।

जिंक की कमी

थैलासीमिकस में थोड़ी जस्ते की कमी पाई जाती है विशेष रूप से यदि वे प्रमेह से पीड़ित हों।

डैस्फराल की विषाक्तता

शरीर से लोहा निकालने के लिये विश्व भर में डैस्फराल का प्रयोग किया जाता है। पिछले सात वर्षों में 3-16 वर्ष के 12 बच्चों (जिनको 50-70 ng/kg/day 5-7 times/week के हिसाब से डैस्फराल दिया जाता था) में रिक्कटस के समान लक्षण पाये गये व वृद्धि में कमी पाई गई।

(भारतीय परिस्थितियों में इसकी कल्पना भी नहीं की जा सकती।)

निष्कर्ष

थैलासीमिया में शारीरिक वृद्धि में रुकावट के एक से अधिक कारण हैं। सभी कारण किसी भी बच्चे में एक साथ नहीं होते परन्तु ये सब एक दूसरे से जुड़े हुये हैं। अतिरिक्त लोहे से अन्तःस्रावी ग्रन्थियों में विकार व वाईरल इन्फेक्शन की वजह से जिगर में विकृति शारीरिक विकास को बहुत बुरी तरह प्रभावित करती है।

Report from Ahmedabad

Thalassemia & Sickle Cell Society of Ahmedabad organised a lecture on Thalassemia & Bone Marrow Transplantation on Saturday, January 6, 1996 at Hotel Mest, Conference Hall, Navrang Pura, Ahmedabad. The Chief Speaker was Dr. Alok Shrivastva, Haematologist, C.M.C. & Hospital, Vellore. Lecture was followed by Question-Answer session and ended with a dinner.

He gave his results of B.M.T. at Vellore which were comparable with that of Italy.

Report from Thalassemia Society of Burdwan

Thalassemia Society of Burdwan plans to organise a walk on 12-02-96 for creating awareness on

Thalassemia. It will start from Burdwan Town Hall & will conclude at Rajwari University.

The society is looking after 120 patients. It has got licence for Blood Bank. The Blood Bank will start functioning within 2-3 months.

Report from Chandigarh

Mr. Rakesh Gupta an active member of Thalassemia Children Welfare Association, Chandigarh organised a Blood Donation Camp on Saturday 25th November 1995 at 1221 Sector 19-B, Chandigarh. Mr. Pawan Kumar Bansal, Member of Parliament, Chandigarh inaugurated the camp. It was sponsored by Agarwal & Company, Chandigarh.

NATIONAL UPDATE ON THALASSEMIA, OCTOBER 7-8, 1995 CHANDIGARH

A national update on thalassemia was jointly organised by the Thalassaemic Children Welfare Association, Chandigarh and Departments of Hematology, Pediatrics and Transfusion Medicines, Postgraduate Institute of Medical Education & Research, Chandigarh on October 7-8, 1995 in P.G.I., Chandigarh. The conference was a grand success which was the result of collective efforts by Dr. Gurjiwan Grewal, Dr. S.K. Agnihotri, Dr. R.K. Marwaha and Dr. Sudarshan from P.G.I. and Executive members of Association. Prof. Amrit Tiwari, Sub-Dean of the P.G.I. inaugurated the conference and Prof. B.N.S. Walia, former Director, P.G.I. and the guest of honour delivered a touching address and appreciated the efforts and activities of the T.C.W.A. Mr. M.S. Rekhi, Sr. Vice-President, T.C.W.A. while appreciating the relationship and smooth co-ordination between the Association and the P.G.I. doctors appealed the community for their involvement in the interest of Thalassaemics and to adopt preventive measures to check further increase in the number of patients.

Scientific sessions were addressed by eminent experts including Dr. David Dennison, Associate Professor of Medicine, C.M.C. College & Hospital, Vellore, Dr. K.C. Das, Kuwait University & Hematology Unit, Kuwait, Dr. V.P. Chaudhary, All India Institute of Medical Sciences, New Delhi, Dr. M.B. Aggarwal from Bombay Hospital &

Medical Research Centre, Dr. M.R. Lokeshwar, Bombay, Dr. Nishi Madan, Dr. V.K. Khanna, Dr. Siddharth Sen all from Delhi, Dr. Gurjeewan Grewal, Dr. S.K. Agnihotri and Dr. R.K. Marwaha from the P.G.I. highlighted the Advances in the Diagnosis & Management of Thalassemia.

An insight into the molecular basis of these disorders and the identification of common DNA mutations prevalent in the Indian Population have made possible the setting up of parental diagnostic facilities. Various cost-effective methods of screening for carrier status were discussed with due emphasis being laid down differentiating features between a beta-thalassemia carrier and iron-deficiency anemia. The recent introduction of an oral iron chelator, at roughly 1/5th of the cost of its parenteral counterpart, has hold out hope to those who could not afford chelation therapy in the past. Bone-marrow transplantation, from a fully matched, HLA identical sibling donor has been carried out in over 40 cases in the C.M.C., Vellore with 70-75% success rates. Drugs to regulate the HbF switch have been used with varying degrees of initial success. Butyrates, which held out considerable promise a few years ago, have not stood the test of time. A presentation on gene therapy focussed on the problems which need to be overcome before this form of treatment could be made available to

patients.

A panel discussion, on each of the two days, stimulated a great deal of interest. The activity enabled parents and patients to obtain clarifications regarding various aspects of treatment from the experts.

The necessity and feasibility of setting up regional referral centres, providing comprehensive facilities, were discussed in the valedictory function. It was emphasised that the various thalassemic societies in India must amalgamate into a national body so as to have a voice that matters. Beginning was made in 1994 when more than eight such associations formed a common platform under the banner of Federation of Indian Thalassemics.



A group of Thalassaemic Children in front of Stall of Federation of Indian Thalassemics

Eminent persons who are helping the Association for welfare of the Thalassaemic children were honoured on behalf of the Association during the concluding session of National Update.

On October 7, Vir Deva Foundation, Chandigarh organised highly entertaining and educative variety programme in Tagore Theatre which was dedicated to Thalasseemics, as a part of the update programme. Mr. Rakesh Arora, President Vir Deva Foundation and Mr. M.S. Rekhi of T.C.W.A. while addressing the audience (900) touched upon Thalasseemia and its causes, treatment and management of Thalasseemic patients. A Souvenir was also released by General Manager of the Indian Express.

The proceedings of the update were also covered by the press and electronic media extensively.

It was also decided to adopt the year 1996 as Awareness year for Thalasseemia. The request is being made to all the societies and FIT for the needful to be done in this regard.

Congrates

Thalasseemics of Ajmer have joined hands and formed a Society under the banner of Ajmer Region Thalasseemic Welfare Society. Thalasseemics of Ajmer and adjoining area may contact its Secretary Mr. Iswar Parvani at 18/2 Mundri Mohalla for help and co-operation.

Welcome

Haryana Thalasseemia Welfare Society has joined "FIT".

Its head office is at 'D' Park, Model Town, Delhi Road, Rohtak, Haryana.

Thalasseemics from Rohtak and adjoining area may contact its President Mr. Ashok for help and co-operation.

National Thalassemia Welfare Society

A Perfect Health Mela was organised at Jawaharlal Nehru Stadium Grounds by Govt. of Delhi & H.C.F.I., New Delhi in collaboration with various National & International Health organisations including UNICEF & W.H.O. from 27th October, 1995 to 5th November, 1995. It was inaugurated by Chief Minister Mr. Madan Lal Khurana. Daily a number of senior ministers, bureaucrats, politicians, film stars and sports personalities visited the Mela. National Thalassemia Welfare Society took active participation by putting up a stall. Electronic view boxes were made showing various steps of Management & Inheritance of Thalassemia. Over 20 banners were put with following main slogans.

- ❖ स्वस्थ दिखने वाले पति-पत्नी थैलासीमिया बच्चे के जन्मदाता हो सकते हैं।
- ❖ यदि आप पंजाबी, सिंधी, गुजराती अथवा बंगाली हैं तो आपके लिये थैलासीमिया की जानकारी अति आवश्यक है।
- ❖ विवाह अथवा गर्भाधारण से पूर्व थैलासीमिया की जांच,

कभी नहीं आने देगी आपके बच्चे को थैलासीमिया की आंच।

- ❖ थैलासीमिया बच्चों का जीवन आपके रक्तदान पर निर्भर है।
- ❖ आपका रक्तदान = थैलासीमिया बच्चों की आयु सीमा।

As our stall was only next to organiser's one it was highly attended. Awareness literature was distributed freely.

National Thalassemia Welfare Society also organised a picnic for a second successive year. It was held at Qutab Minar lawns on Sunday the 24th December, 1995. Over 200 members of thalassemic families participated. Racing, Tambola, Songs, Dances, Jokes and many other activities marked the event. Thalassemics were rewarded for their performances. Dr. V.P. Choudhary gave the history of Qutab and adjoining monuments. Contributory lunch was served. All the children were given a carry back gift.

Special gift were given to Mr. Ashok Sachdeva, Mrs. Neelam Kohli and Mrs. Kamlesh for their outstanding work during the year.

STEM CELL TRANSPLANT FROM UNRELATED DONORS FOR THALASSEMIA

— Dr. David Dennison

Stem cell transplants from fully matched unrelated donors have been done in small numbers in the United States and Europe for patients with blood cancer and the initial results are encouraging. In the coming years many more will be done and we will get a better idea of how successful they are. The Singapore General Hospital has apparently successfully done one such transplant for a patient with thalassemia major. The technology is not difficult and any major transplant centre like ours can do it.

The procedure is identical to a bone marrow transplant except that instead of harvesting the marrow from the donor's hip bones, stem cells are collected from the donors blood using a machine called a cell separator. The advantage for the donor is that he or she does not have to undergo general anaesthesia. The advantage for the patient is that rejection which is a major complication in unrelated donor bone marrow transplants

may be less of a problem here. The disadvantage for the patient is that graft versus host disease can be a very serious and life threatening complication in unrelated donor bone marrow transplants. The incidence of graft versus host disease is not lower in stem cell transplants. We are waiting for more centres abroad to start doing them in order to gauge the results before we start doing them on our good risk patients. It is very necessary for many more patients to undergo this kind of transplantation before we can draw any firm conclusion. The correct, scientific and ethical way to do this kind of a study is to do it on patients who have very little chance of survival with conventional treatment with blood transfusions and iron chelation. If successful in more than 50-60% of patients then it may be offered to patients who are good risk. This may take a few years. However I believe that this may become an effective way of transplantation in the future.

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JUNEJA

PROPERTIES

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A-10, Gujranwala Apartments
Vikas Puri, New Delhi-110 018

Tel: 550 5043, 561 7667

NATIONAL THALASSEMIA WELFARE SOCIETY (Regd.)

KG-1/97, Vikas Puri, New Delhi-110 018 Tel: 550 7483

SPECIAL THALASSEMIA CLINIC

National Thalassemia Welfare Society organises Thalassemia Check up Clinic on **2nd Sunday** of every month at **Charitable Medical Clinic, Lajpat Bhawan**, Near Vikram Hotel, Near Mool Chand flyover, Lajpat Nagar, New Delhi.

Facilities

- ❖ Growth Monitoring
- ❖ Chelation Therapy
- ❖ Serum Ferritin Assay for Rs. 150/- only
- ❖ Inj. Engerix (Hepatitis B vaccine)
Rs. 175/- for Children below 10 years
Rs. 350/- for Children above 10 years
- ❖ Thalassemia Screening

For appointment Tel 550 7483

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Any person can become a member of the society.

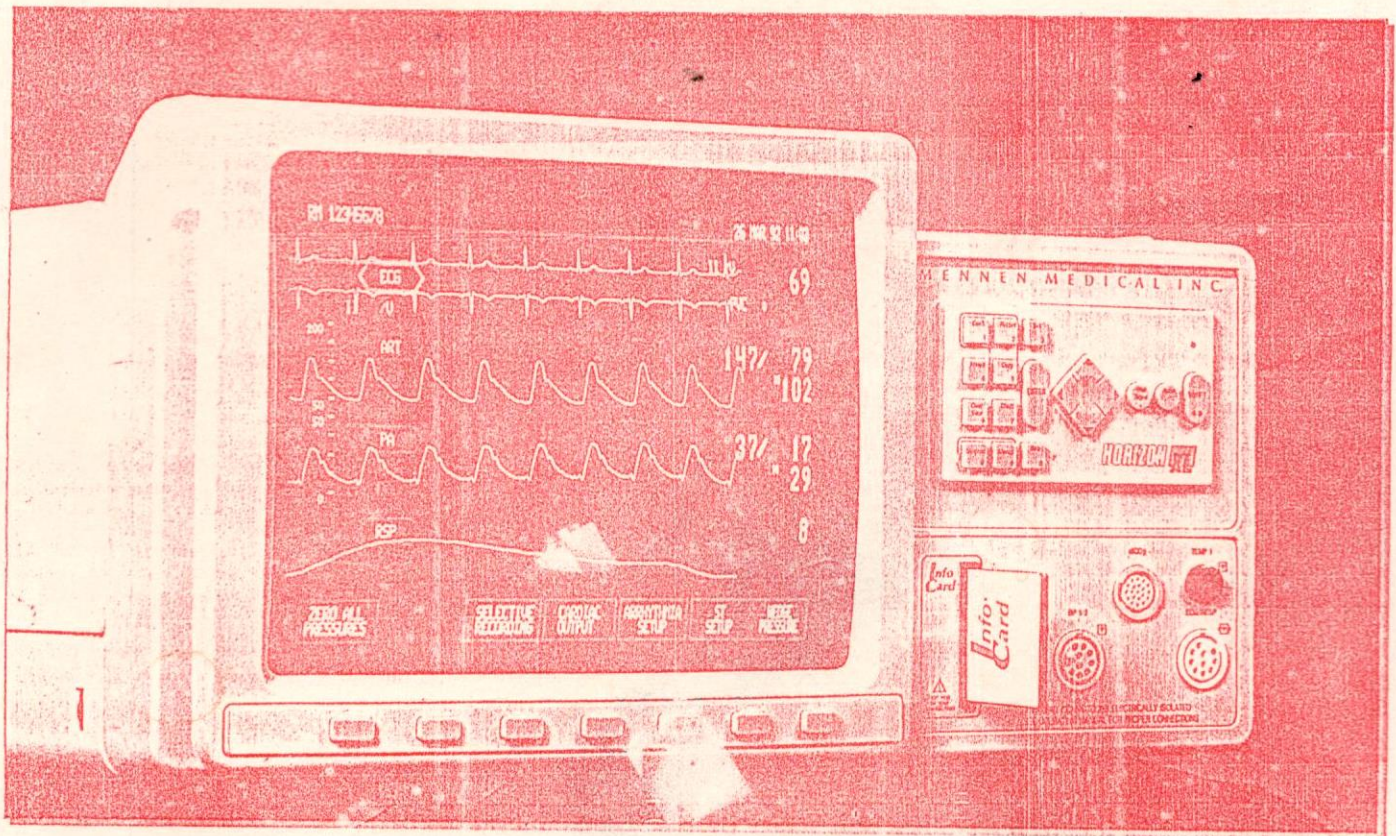
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Deferiprone

Abridged Prescribing Information **Composition:** Kelfer-250/500 Each capsule contains Deferiprone 250 mg/500 mg. **Indications:** Transfusion haemosiderosis, especially in cases of thalassaemia, other haemolytic anaemias, aplastic anaemia and myelodysplastic syndromes, acute iron poisoning, siderosis associated with liver cirrhosis and for the diagnosis of iron-storage diseases. **Dosage and Administration:** 50-75 mg/kg body weight daily in 2-4 divided doses. **Contraindications:** Hypersensitivity to deferiprone. **Warnings and Precautions:** Kelfer should be administered with caution in patients whose serum ferritin levels are below 1000 ng/ml and in patients with impaired hepatic and renal function. Kelfer is not recommended in children below 2 years of age. Reversible impairment of cardiac function may occur in patients with severe iron overload undergoing combined treatments with Kelfer and vitamin C. **Pregnancy:** Deferiprone is not recommended for use in pregnant women. **Side Effects:** GI disturbances, joint pains and swelling are reported. Agranulocytosis, neutropenia and zinc depletion may occur. **Patient Monitoring:** The minimum monitoring essential for deferiprone therapy: (1) Haemoglobin, total and differential white cell counts and platelet counts at 3-4 weekly intervals or whenever clinically indicated (2) Serum ferritin at 3-4 monthly intervals. **Note:** If the total white cell count drops to less than 3000/cmm or Absolute Neutrophil Count (ANC) falls to less than 1000/cmm or platelet count falls to less than 1,00,000/cmm, the drug should be discontinued. In case the patient develops severe joint pain, swelling or difficulty in squatting/walking and no relief is obtained by administering ibuprofen/diclofenac or any other suitable NSAID, the therapy should be discontinued. The drug should not be restarted if joint pains recur. **Presentation:** Container of 50 capsules.

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