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FEDERATION of Indian Thalassemics

National Thalassemia Bulletin

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KELFER LAUNCHED

The long awaited oral iron chelator

L-1/DEFERIPRONE/KELFER

now available for Therapeutic use in Thalassemia in India

Dr. George J. Kontoghiorghes, Inventor of drug, receiving a momento from Dr. S.K. Kacker, Director, All India Institute of Medical Sciences, at the Symposium on Thalassemia & Deferiprone during the launching of Kelfer at Jawahar Lal Auditorium, AIIMS, New Delhi 4th & 5th March, 1995 Dr. Manorma Bhargava celebrating the occasion.

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FIT Executive Meeting

— Secretary's Report

You will be glad to learn that FIT has been registered and nine Thalassemic societies in India have already enrolled with it. List of member societies is given on left side. We hope that others will be joining soon.

FIT executive meeting was held on 4th March 1995, New Delhi. Six Societies were represented and the meeting lasted for over 3 hours. A few of the following important points discussed and decisions taken there upon.

International Thalassemia Day

Various forms of its celebrations were discussed and it was left to the individual societies for final decision. However it was agreed that all societies should celebrate it along with banner of FIT.

Awareness Programme

It was felt that it was the urgent need of the hour that FIT should produce literature for "Awareness" in such a way that it can be used by all member societies in our country.

General Facilities

It was decided that FIT should approach the authorities to get the

1. Handicap benefits
2. Concession for Air and Bus travel
3. Income Tax benefit on medical expenses for care of Thalassemic children.

Letter Heads

All members desired that all societies who are member of FIT should mention on their letter heads "Member of Federation of India Thalassemics" along with its logo.

Future Activities

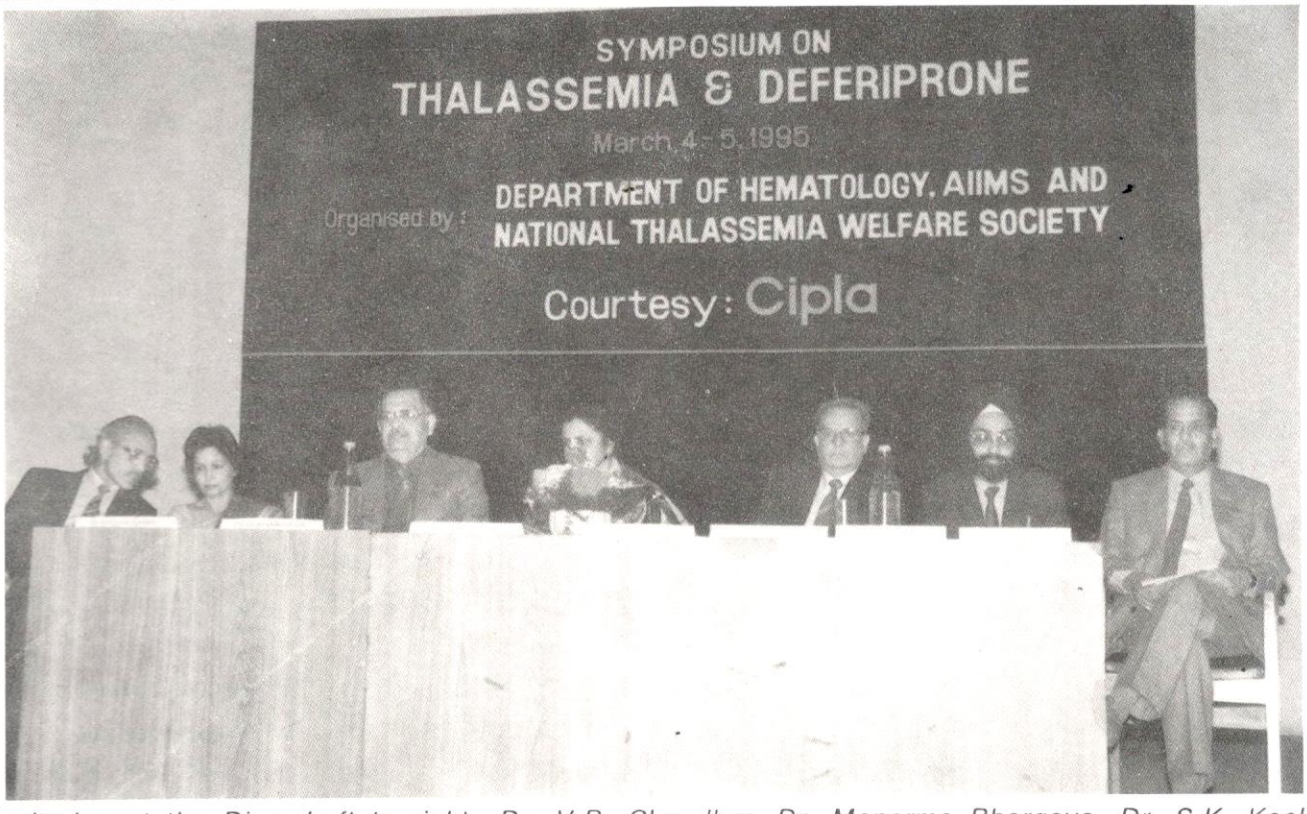
Thalassemic Children Welfare Association, Chandigarh informed that they will be organising the next National Thalassemia Conference in Sep./Oct. 1995 at Chandigarh. Members desired it should be held under the banner of FIT to which Mr. S.P. Ajmani President of T.C.W.A. agreed.

Custom & Excise Relief on Kelfer

Members felt that steps should be taken to get custom relief on Maltol (raw material used in manufacturing of Deferiprone) and excise relief on bulk drug Deferiprone.

Dr. J.S. Arora

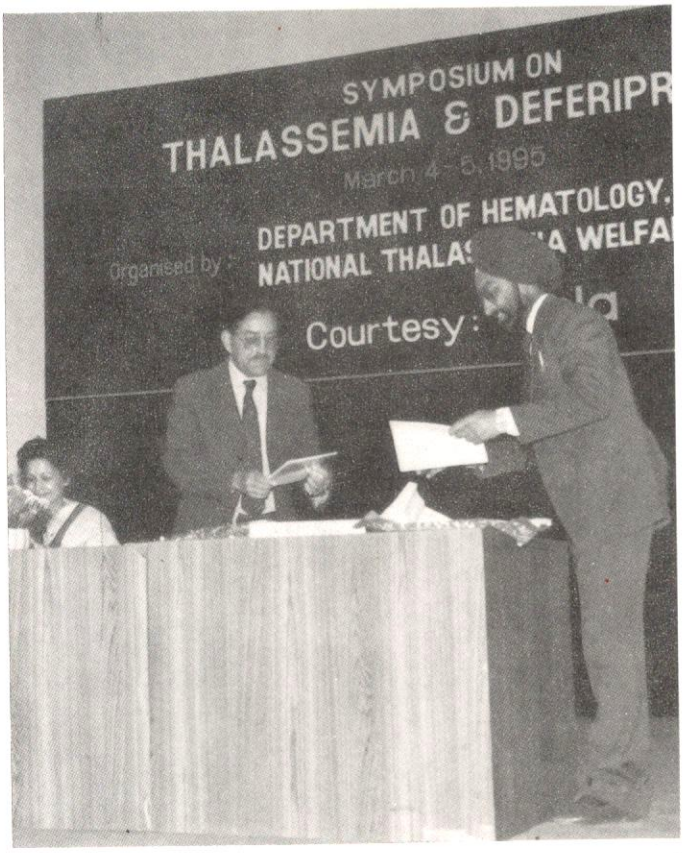
KELFER LAUNCHED



Dignitaries at the Dias: Left to right – Dr. V.P. Choudhry, Dr. Manorma Bhargava, Dr. S.K. Kacker, Miss Surrender Saini, Dr. P.K. Dave, Dr. J.S. Arora, Mr. S.P. Ajmani

As the results of successful trials of L-1/Deferiprone/Kelfer from abroad and Bombay poured in, it became burning topic among the thalassemics. As soon as the trials went multicentric in India the people were eagerly watching their results. Deliberations of Cyprus International Conference and the results of Indian multicentric trial presented at Bombay Conference and then at National Thalassemia Conference Delhi made the thalassemics restless. Patients/ Parents individually, through societies and then Federation approached Govt of India and Cipla Ltd. for its early launching. Even after the trials were over Drug Controller of India, Director

Miss Surrender Saini, Padma Bhushan awardee, noted social worker, Chairman-Delhi Social Welfare Advisory Board & President of N.T.W.S. lightening the lamp during the inauguration ceremony.



Prof. L.M. Nath, Dean - AIIMS, receiving a copy of Souvenir from Dr. J.S. Arora

blessings in the form that its users are benefited. A *pooja* ceremony was held at Chhatarpur Temple on Maha Shivratri the 28th Feb. 1995 and *Shri Akhandpath Sahib* followed by *Kirtan & Ardas* was held at Gurudwara Rakab Ganj Sahib on the day of launch i.e. 5th March, 1995.

Thalassemic parents were overjoyed. As with any other drug Kelfer too has some side effects and like in all new drugs scientists advised its use under close monitoring. To appraise the Doctors and Parents National Thalassemia Welfare Society in collaboration with Deptt. of Haematology, AIIMS organised a Symposium on Deferiprone on 4th & 5th March, 1995. 1st day was purely scientific and over 200 medical professionals attended this session. On the 4th March it was inaugurated by Prof. L.M. Nath, Dean, AIIMS. Dr. J.S. Arora released a souvenir on this occasion. It contains detailed articles on Oral iron chelation one by Dr. George J. Kontoghiorghes & another by Dr. M.B. Agarwal, General of Health Services, Govt. of India, Indian Council of Medical Research & others experts from various Institutions took more than a year before the seal of approval was put by Govt. of India. At last the Day came and the long awaited oral iron chelator L-1/Deferiprone/ Kelfer was launched in Indian market for therapeutic use in thalassemics on 5th March, 1995 at AIIMS.

As the drug was to be launched first time in the world it was decided to pray the almighty to shower his

Prof. Dr. S.K. Kacker, Director AIIMS discussing the prospects of Kelfer with Dr. M.B. Agarwal & Mr. Harish Chawla

Distinguish speakers & Chairpersons during the Symposium
Left to right – Dr. M.B. Agarwal, Haematologist, Bombay Hospital & M.R.C., Bombay and Secretary T.S.C.S. Bombay, Dr. V.P. Choudhry, Addl. Prof., Deptt. of Haematology, AIIMS, Dr. R.K. Marwaha, Addl. Prof., Deptt. of Paediatric, P.G.I., Chandigarh, Dr. George J. Kontoghiorghes, Prof., Deptt. of Haematology, Royal Free Hospital, London, Dr. A.K. Saraya, Former Head, Deptt. of Haematology, AIIMS, Dr. I.C. Verma, Head, Genetic Unit, AIIMS, Dr. H Pati, Assoc. Prof., Deptt. of Haematology, AIIMS

Prevention of Thalassemia — a necessity by Dr. I.C. Verma and Future development in management of Thalassemia by Dr. V.P. Choudhry. Dr. V.P. Choudhry while welcoming the faculty thanked Dr. George for coming all the way to India to bless his baby (Deferiprone). He stressed the need of chelation therapy. He also gave the overview of Thalassemia management and the poor facilities prevailing in the country. Dr. George J. Kontoghiorghes the invertor of drug gave clear picture of Deferiprone trial world wide and its present status.

Prof. Nishi Madan while speaking on epidemiology showed a short clinical study in which 5.5% Delhiites were found to be thalassemia traits.

Dr. Gogtay a representative of Cipla Ltd. mentioned the monitoring and distribution of drug in detail on both days. He said the present cost of a 500 mg Cap. is Rs. 19/- and that of 250 mg is Rs. 11/-.

Dr. P.K. Dave, Medical Superitendent, AIIMS receiving the momento from Miss Surrender Saini

On 5th of March Prof. Dr. S.K. Kacker, Director, AIIMS while inaugurating the Symposium congratulated Dr. George J. Kontoghiorghes who invented the drug and Dr. M.B. Agarwal, Dr. V.P. Choudhry, Dr. R.K. Marwaha and Dr. S. Chandra who worked along with

A view of the audience
Front row – Mr. Goverdhan Kalra (Thalassemia Society of Jaipur), Dr. Y.K. Hamied (M.D. Cipla), Mr. Harish Chawla (Director Cipla), Dr. G.J. Kontoghiorghes (Inventor of Drug), Mr. S.V. Iyer (President T.S.C.S., Bombay)
Second row – Dr. (Mrs.) Raj Chawla, W/o Mr. Harish Chawla, Dr. (Mrs.) Madhu Choudhry, W/o Dr. V.P. Choudhry, Dr. (Mrs.) Usha Gupta (Blood Bank Officer, AIIMS)

other colleagues on this drug in India. He said, "this is a true example of co-ordinated research work between pharmaceutical industry and medical profession". He appreciated Dept. of Haematology and National Thalassemia Welfare Society for working hand in glove to alleviate the sufferings of thalassemic families. He said that with future advances thalassemics will have bright future and science will be able to eradicate this disorder.

Miss Surrender Saini, a noted social worker and President of the Society said that I am very happy to learn that Kelfer is being launched. "Today is a red letter day in the history of Indian thalassemics. With the introduction of Kelfer now treatment will be easy and within the reach of most of the thalassemics. They will grow normal having all the faculties and will live a normal life." "We had personally met the Finance Minister to get the relief in custom and excise so that the drug may be made more economical." She said, "I pray to God the patients should benefit from Kelfer. I am always ready to devote my services for welfare of thalassemics".

While speaking on the occasion Dr. Manorma Bhargava, Head of Haematology, AIIMS inform

that their Department has contributed for the research work on Kelfer and assured the thalassemics for continued support for improvement in medical care. She thanked Dr. George for developing the Deferiprone and Managing Director, Cipla Dr. Y.K. Hamied for bringing the drug to India and hope that Cipla will continue to work for welfare of thalassemia.

Dr. Y.K. Hamied, M.D. Cipla and Mr. Harish Chawla, Director, Cipla thanking Dr. (Mrs.) Manorma Bhargava, Head of Haematology Department, AIIMS for co-operatation during the trial of drug and in organising this Symposium.

5th March's deliberations were specifically tailored to the needs of thalassemic patients and their parents. All the Indian speakers used Hindi as their communication language. Dr. M.B. Agarwal very clearly spoke on all the ifs & buts, use & abuse, pros & cons, plus & minus points of Kelfer in a lay man language so that every body can understand the drug very well. The remaining doubts were cleared during long Question-Answer session. Over 500 thalassemic parents (150 odd from outstation) left their seats with satisfaction and clear minds to choose between KELFER & DESFERAL. At the close of session Dr. Y.K. Hamied congratulated Dr. George J. Kontoghiorghes, Dr. M.B. Agarwal and all others who worked on the drug. He said it took a dedicated work of over 15 years to bring this drug into the market and Cipla was associated with it since 1988. He appreciated Dr. V.P. Choudhry the Organising Secretary of the Symposium for successful organisation of the two day session and assured the parents that Cipla will continue to work for welfare of thalassemia. The day long session ended with lunch. Both the sessions were open to all, no registration fees was charged and sponsored by Cipla.

Dr. Manorma Bhargava, Mr. Harish Chawla & Dr. I.C. Verma (Head – Genetic Unit, AIIMS) at the end of session

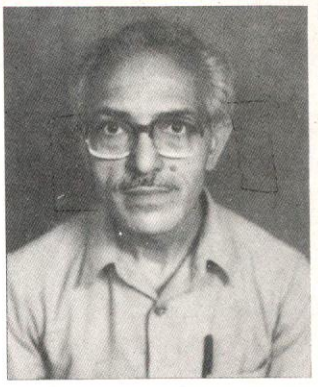
The event was widely covered in all the leading newspapers and telecasted in National News.

During last 4 months many thalassemics who were not on any iron chelation have started taking Kelfer, even the few who were earlier on Desferal have preferred to switch over to Kelfer.

We wish the Kelfer should stand the test of time.

CURRENT STATUS OF ORAL IRON CHELATOR: DEFERIPRONE

—Dr. V.P. Choudhry
Addl. Prof., Deptt. of Haematology, AIIMS, New Delhi



Over 117 million people are heterogygotes for haemoglobinopathies in Asia and 105,978 babies with hemogygous state are born every year. Among these, nearly thirty millions are carriers for thalassemia alone in India and 8-10 thousands babies with thalassemia major are born every year in India alone. Safe blood transfusions at regular intervals is the only way of hope for these children. Most thalassemic children develop iron overload as a result of transfusional iron overload or increased iron absorption from the gut. Iron chelation therapy along with regular blood transfusions is essential for normal growth and development with this form of therapy few lucky ones have reached third decade in India and forth decade in the world.

Chelation therapy should be introduced as soon as possible to prevent organ damage as a result of iron overload. Iron chelation is being achieved by desferrioxamine (DF, Desferal) which is a fungal product isolated from streptomyces pilosus. DF needs to be injected subcutaneously over 8 hours daily with the help of a portable electronic pump. This form of therapy is available to less than 10% of patients worldwide primarily because of its high cost and decrease in compliance with the advancing age. In India only less than 3% of thalassemic patients could receive some DF., while in Cyprus 45% of the health budget is spent on DF. Several side effects have been observed on regular DF therapy In view of its high cost, decreasing compliance and cumulative toxicity, efforts have been made over the last 30 years to develop an effective, and safe oral iron chelator. Over four hundreds of chelators have been evaluated in vitro and in animals. Among these only 15 have reached the state of clinical evaluation in humans and of these only 1,2-Dimethyl -3 hydroxypirid-4-one (L-1/Deferiprone/Kelfer) has been evaluated in 17 countries at 32 centres. So far over eight hundred patients have undergone treatment with Deferiprone over several years. Iron excretion by Deferiprone depends on the dose, frequency of administration and ferritin level. India is the only country in the world, where multicentric study at four centres involving over 225 cases have been conducted to determine the effectiveness and safety of Deferiprone in the multitransfused thalassemic children.

Physical & Chemical Properties:

It is crystalline substance which is stable at room temperature and in the acidic pH. It forms red colour complexes with iron and has high affinity for iron. Three molecules of L-1 are needed to bind with one molecule or iron. Soluble forms of iron are rapidly mobilised by L-1 whereas iron bound to proteins is mobilised slowly. L-1 mobilises iron from iron storage such as ferritin and hemosiderin. Mobilisation of iron from ferritin and hemosiderin is very slow, whereas from transferrin and lactoferrin is completed within hours.

International Study on Deferiprone:

Worldwide clinical studies were initiated over several centres from 1987. Over eight hundred patients and volunteers have received L-1 in seventeen countries for variable periods and even as long as for 4 years. These studies have revealed that iron excretion by L-1 depends upon the dose, frequency of administration and the iron overload. It has been used in 2-4 divided doses of 50-100 mg/kg/day. In this dose it has been shown to excrete 10-120 mg of iron in iron overloaded individuals and only 1-2 mg of iron in normal persons. Overall L-1 has been observed to be effective in excreting more iron than transfused by blood cell transfusions and in this dose it is able to deplete the iron stores. It appears that iron is mobilised from saturated transferrin, liver and other compartments containing excess of iron. Patients with HbF/B-thalassemia achieve negative iron balance faster on the same dosage schedule. Long term administration have resulted in bringing the serum ferritin levels within the normal range.

L-1 is bitter in taste, soluble and rapidly absorbed from the stomach and intestine. It is metabolised to a glucuronide conjugate and is cleared from the blood within 1-2 hours. It is excreted in the urine as a glucuronide conjugate, unchanged L-1 or L-I bound to iron. Drug is not excreted through stools.

Indian Study:

Maximum number of thalassemic children have under-

gone treatment with Deferiprone in India. Initial studies in India were started in August 1989 at LTMG hospital and JJ Group of hospitals in Bombay. First study involved 52 cases while in the second study 70 patients were evaluated. Subsequently a multicentric study was initiated at Bombay, Delhi, Chandigarh and Calcutta to evaluate the efficacy and safety of the drug in patients with thalassemia. In the initial studies at Bombay drug was administered in doses of 25, 50, 75 and 100 mg/kg/day. It was clearly demonstrated that iron excretion was directly dependent on the daily dose. Serum ferritin levels fell by 50% after one year in patients receiving either 75 or 100 mg/kg/day. Overall urinary iron excretion on equivalent dose of L-1 and desferioxamine (DFO) was identical as the L-1 was being given daily. In addition to ordinary iron excretion 20-30% of iron is also excreted through faeces. DFO is used for 5-6 days a week in most protocols to improve the compliance. Since the Deferiprone is administered daily with interruption. Therefore, the overall efficacy of Deferiprone is almost equal to that of DFO. Results at AIIMS has revealed that the serum ferritin levels fell to nearly 50% of the initial levels in patients receiving 75 mg/kg/day of Deferiprone. Fall in serum ferritin associated with lightening of skin colour. Many patients have achieved the serum ferritin levels below 2000 ng/ml on long term therapy despite continuous transfusions. Iron excretion was higher in patients with E-beta thalassemia. Similarly iron loaded patients of thalassemia intermedia who are not transfusion dependent also show a rapid decline in serum ferritin levels along with the decrease in level of liver iron content.

Thus Deferiprone appears to be an effective agent in bringing the negative iron balance when administered in the doses of 50 or 75 mg/kg/day. However, it can be used in higher doses in the presence of very high serum ferritin levels.

Adverse Effects:

Long term administration of Deferiprone have established that it has only transient adverse effects. Over 400 patients have been able to continue the drug without any complaints or complications. Its side effects are dose dependent. Mild nausea, abdominal pain, vomiting have been observed in nearly 5% of cases. All these symptoms are mild and subside on continuation of therapy except in an occasional patient when it became essential to withdraw the drug.

Myelotoxicity presenting as neutropenia has been observed in 1% of cases. Development of neutropenia is not associated with the dose or duration of therapy. Patients may succumb to associated infection or septicemia. One patient in our study developed neutropenia and died of diptheria infection. Unfortunately one cannot predict the development of myelotoxicity. It is advisable to monitor the counts at 3-4 weekly interval. However, total and neutrophil count are essential whenever patient on L-1 develops fever. Such cases should be initiated with broad spectrum antibiotics and the Deferiprone should be stopped if blood testing is not possible immediately.

Pain over joints and muscles is seen in 10-30% of cases. In majority of them pain is either mild without affecting day to day activity of the individual or it subsides with analgesics. However, in small number of patients drug has to be withdrawn because of arthritis. Some centres have observed that the development of arthropathy is dependent on the dose and on ferritin levels. However, we and others feel that arthropathy is neither dependent on dose of L-1 nor on initial serum ferritin levels. Myelotoxicity and arthropathy are reversible. The pathogenesis of both these complications is not well understood. However, idiosyncratic mechanism may be responsible for myelotoxicity. It has been postulated that arthropathy could be secondary to re-distribution of iron into the synovial membrane or articular cartilage. Most studies including ours have evaluated ANF, RF positivity with the development of arthropathy. All these studies have clearly shown that these immunological parameters lack specificity with musculoskeletal complications. ANF & RF positivity in these cases may be secondary to non specific stimulation of the immune system following multiple transfusion. Other uncommon side effects include dermatopathy following zinc deficiency.

GOOD NEWS

Drug controller of India has at last given the permission in January 1995 to CIPLA to make the drug available for chelation of iron for patients with iron overload. Availability of this drug will benefit the large number of thalassemic patients, as now the chelation therapy will be within the reach. This orphan drug at last has been released. However, a regular monitoring will be essential. It will be unfortunate that patients with mylotoxicity and skeletomuscular involvement will not be able to use this drug. They will have to resort to expensive, painful and inconvenient DFO therapy. Without iron chelation therapy most thalassemic patients used to succumb to iron toxicity by 15-20 years of age. With the advent of Deferiprone, majority of patients will be able to initiate chelation therapy at an early age to lead a normal life as the drug is cheap and within the reach of the majority. Thus a bright future awaits for the thalassemic children.

मौखिक लोह निष्कासक दवा डैफरीप्रोन की ताजा स्थिति

— डा॰ वी॰पी॰ चौधरी
हिन्दी रूपान्तर डा॰ जे॰एस॰ अरोड़ा

एशिया में 11.7 करोड़ लोग हीमोग्लोबिन संबधित रोगों से प्रभावित हैं तथा 105978 बच्चे हर वर्ष पैदा होते हैं। इसमें से भारत में 3 करोड़ लोग थैलासीमिया के कैरीयर हैं तथा 8-10 हजार बच्चे प्रतिवर्ष थैलासीमिया मेजर के पैदा होते हैं। सुरक्षित नियमित रक्त संचारण पर ही इनका जीवन निर्भर है। थैलासीमिक बच्चों में रक्त संचारण व आंतो से लोह अवशोषण द्वारा शरीर में लोहे की मात्रा बढ़ जाती है। थैलासीमिक्स में सामन्य वृद्धि के लिये नियमित रक्त संचारण के साथ-साथ अतिरिक्त लोहा निकालना भी अति आवश्यक है। लोह निष्कासन जल्द से जल्द आरंभ कर देना चाहिए ताकि विभिन्न अंगों को नुकसान न हो। यह प्रक्रिया त्वचा के नीचे धीरे-धीरे विशेष पंप द्वारा 8-10 घंटे में डैस्फराल injection देकर की जा सकती है। दुनिया में 10% व भारत में केवल 3% लोग ही इसका खर्चा बर्दाश्त कर सकते हैं। इसके अतिरिक्त आयु बढ़ने के साथ-साथ बच्चे इसका विरोध करने लगते हैं। साईप्रस का 45% स्वास्थय बजट केवल डैस्फराल पर ही खर्च होता है। डैस्फराल की मंहगाई, नुकसानदायक प्रभाव व बड़े बच्चों में विरोध को देखते हुए पिछले 30 वर्षों में करीब 400 से अधिक रसायनों पर परीक्षण किये गये जिनमें से 15 पर मानवीय प्रयोग किये गये। केवल एल-वन, डैफरीप्रोन या कैलफर ही एक मात्र ऐसा है जिसे 17 देशों के 32 केन्द्रों में 800 से अधिक थैलासीमिक्स पर आजमाया गया। केवल भारत में ही सबसे अधिक 4 केन्द्रों पर 225 बच्चों पर इसके प्रयोग किये गये। कुछ बच्चों को 4 वर्ष तक लगातार यह दवा दी गई।

उपरोक्त प्रयोगों से पता लगा कि एल-वन द्वारा लोहे का निष्कासन दवा की मात्रा, देने की अवधि व शरीर में लोहे की मात्रा पर निर्भर करता है। इसको 50-100 मि॰ग्रा॰/प्रति किलो/प्रति दिन के हिसाब से 2-4 भागों में विभाजित करके दिया गया। इससे थैलासीमिक्स में 10-120 मि॰ग्रा॰ लोहा निकला जबकि आम आदमी में केवल 1-2 मि॰ग्रा॰ लोहा ही निकला। कुल मिलाकर ऐसा देखने में आया कि जितना लोहा रक्त संचारण से शरीर में जाता है उससे अधिक लोहा शरीर से निकलता है तथा लम्बे समय तक इसका प्रयोग करने से शरीर में लोहे की मात्रा सामान्य हो जाती है। यह 1-2 घंटे में आंतो से अवशोषित होकर शरीर से बाहर निकल जाता है।

भारत में अगस्त 1989 में LTMG अस्पताल व JJ Group of Hospital बम्बई में इसका प्रयोग आरंभ हुआ। इसमें पहली बार 52 तथा बाद में 70 बच्चों को लिया गया। इसके बाद बम्बई, दिल्ली, चंडीगड़, कलकत्ता में एक साथ परीक्षण किये गये। एक वर्ष में फैरीटीन की मात्रा लगभग आधी रह गई। एल-वन की लोह निष्कासक क्षमता डैस्फराल के बराबर आंकी गई। हालांकि कैलफर द्वारा लोहा केवल मूत्र से निष्कासित होता है परन्तु क्योंकि डैस्फराल सप्ताह में केवल 5 बार देना ही संभव है और कैलफर मुख द्वारा लेने के कारण रोज़ लिया जा सकता है अत: कुल मिला कर दोनो द्वारा समान लोहा निकलता है। बहुत से बच्चों में फैरीटीन 2000 से कम करने में भी सफलता मिली। E-बीटा थैलासीमिया व थैलासीमिया Intermedia में इसके प्रभाव और अच्छे पाये गये।

नुकसानदायक प्रभाव

लगभग 400 थैलासीमिक्स में बिना किसी दुष्प्रभाव के डैफरीप्रोन को काफी लंबे समय तक दिया जा सका। इसके दुष्प्रभावों में हल्का सा ज़ी मिचलाना, पेट दर्द, उल्टी आना आदि 5% बच्चों में देखा गया। ये सभी लक्षण दवा के लगातार लेने से समाप्त हो जाते हैं। केवल एक बच्चे में इन कारणो से दवा को बंद करना पड़ा।

रक्त में श्वेत कणों की कमी या Neutropenia 1% बच्चों में देखने को मिला। यह दुष्प्रभाव दवा की मात्रा अथवा देने की अवधि से कोई संबंध नही रखता। AIIMS में एक बच्चे में यह दुष्प्रभाव देखने को मिला तथा डिपथीरिया से इसकी मृत्यु हो गई। दुर्भाग्यवश इसकी कोई पूर्व घोषणा नही की जा सकती अत: प्रति 3-4 सप्ताह पर इसकी जांच करवा लेनी चाहिए। जब भी कैलफर दवा लेने वाले थैलासीमिक को बुखार हो वह

तुरंत दवा बंद कर'ले और Hb, TLC, DLC व प्लेटलैट आदि की जांच करवा ले तथा उसको उच्च स्तर के जीव रोधक दवा (broad spectrum antibiotic) देने चाहिए। (*नोट:* पुन: कैलफर दवा देने से पूर्व अपने चिकित्सक से परामर्श अवश्य कर ले।) 10-30% थैलासीमिक्स में जोड़ो का दर्द भी देखने को मिला। इनमें से अधिकतर में दर्द बहुत हल्का होता है तथा दर्द निवारक दवा देने से खत्म हो जाता है। परन्तु कुछ में इस दर्द के कारण डैफरीप्रोन को बंद करना पड़ता है। उपरोक्त दोनों दुष्प्रभाव दवा बंद करने से स्वत: ही समाप्त हो जाते हैं। इन दुष्प्रभावों का कोई स्पष्ट कारण अभी तक ज्ञात नहीं हो पाया है। डैफरीप्रोन के प्रयोग से 1% बच्चों में जिंक धातु की कमी के कारण त्वचा पर प्रभाव देखा गया।

शुभ सूचना

जनवरी 1995 में औषधि नियंत्रक ने सिपला कंपनी को इस दवा को थैलासीमिक्स के इलाज के लिये बनाने की आज्ञा दे दी है। अब बहुत अधिक बच्चे इसके प्रयोग से लाभान्वित हो पायेगें यद्यिप इसका प्रयोग केवल चिकित्सीय देख रेख में ही करना होगा। जिन बच्चों में इस दवा के दुष्प्रभाव देखने को मिलेगें उनको दुर्भाग्यवश मंहगी व दर्दकारक डैस्फराल पर ही निर्भर रहना पड़ेगा। बिना लोह निष्कासन के एक थैलासीमिक का जीवन 15-20 वर्ष से अधिक नहीं होता। डैफरीप्रोन/कैलफर के सस्ता होने के कारण आरंभ से ही थैलासीमिक्स इसका प्रयोग कर पायेगें। अत: आने वाले समय में थैलासीमिक बच्चों का भविष्य उज्ज्वल होगा।

Kelfer Launch in Ahmedabad

Left to right – Dr. Dilip Shah, Dr. R.B. Shah, Dr. George J. Kontoghiorghes, Dr. M.B. Agarwal
Extreme right – Mr. M.D. Golani

Thalassemia Sickle Cell Society of Ahmedabad in collaboration with Indian Academy of Paediatrics, Gujarat Branch, organised a symposium on Advances in oral iron chelators. Dr. George J. Kontoghiorghes, Dr. M.B. Agarwal and Dr. R.B. Shah addressed over 120 paediatricians. T.S.C.S., Ahmedabad organised another meeting on Kelfer. Over 100 Thalassemic parents benefited from it.

Comparative Study of Kelfer & Desferal

Adopted from Information/Literature supplied by the Companies

	KELFER	DESFERAL
Route of Administration	Oral	Slow S.C./I.V. with Infusion Pump over 8-12 hrs.
Excretion	Through Urine. Excretion is proportional; to dose, frequancy of administration and iron over load.	Through Urine & Faeces. Excretion related to the dose, route, duration of administration & iron over load
Dose	50-100 mg/kg/day. 7 days a week.	20-60 mg/kg/day 5-7 times a week.
Effect of Vit. C	May enchance iron excretion	150-250 mg. Enhances iron excretion
Cost	500 mg Cap. Rs. 19/- & 250 mg. Cap. Rs. 11/-	Rs. 127/- per vial of 500 mg. + cost of accessories (approx. 30/- per day)
Cost per child (20 kg. & Ferritin > 2500 ng)	Approx. Rs. 60/- per day	Approx. Rs. 300/- per day

Adverse reactions:

Kelfer:—Transient agranulocytosis & neutropenia (1-2%). Transient musculo-skeletal and joint pain (10-20%).

Gastric intolerance (2-6%) – anorexia, nausea, vomiting, gastric discomfort & altered taste, disappear on continuation of therapy. Zinc deficiency (1%) leading to dermatopathy.

Desferal:—Pain, swelling, erythema, burning, pruritus, wheals and rash at the site of injections, occasionally accompanied by fever, chills and malaise, anaphylactic/anaphylactoid reactions with or without shock, angioedema.

Generalised rash pruritus, urticaria. Nausea, vomiting, diarrhoea, abdominal cramps.

Visual defects, retinopathy, optic neuritis, cataracts, corneal opacities. Tinnitus, hearing loss.

Neurological disturbances, dizziness, convulsions. Peripheral sensory neuropathy, paraesthesia.

Growth retardation, bone abnormalities. Leg cramps, malaise, bone pain.

Hypotension, Adult respiratory distress syndrome, Impaired renal/hepatic function.

I.V. administration may cause neutropenia, thrombocytopenia and pancytopenia.

Warnings/Precautions:

Kelfer:—Hb, TLC, DLC & platelet counts should be done 3-4 weekly intervals or whenever clinicaly indicated.

If the patient develop fever or sore throat immediate white cell count should be done. If the TLC drops to 3000/cmm or Absolute Neutrophil Count falls to less than 1000/cmm or platelet count falls to less than 1,00,000/cmm, the drug should be discontinued immediately and should not be restarted. Appropriate antibiotic therapy should be instituted. The patient may have to be hospitalised till the counts come back to normal.

In case of severe joint pain, swelling or difficulty in squatting/walking and no relief is obtained by administering suitable drug the therapy should be discontinued. The drug should not be restarted if joint pain recur.

Iron Chelation is must

Vit. C should not be started until the Kelfer therapy is in progress for 2 weeks. In patient with severe iron over load undergoing combined treatment with Kelfer & Vit. C reversible impairment of cardiac function may occur. Such patients should be monitored.

Use with caution in Renal/Hepatic impairment.

Desferal:—Rapid I.V. infusion may lead to collapse

If patient under treatment with Desferal develop fever, treatment should be temporarly discontinued, bacteriological test performed and suitable antibiotic therapy started at once. After the infection has resolved treatment with Desferal can be resumed.

Desferal with high dose Vit. C (> 500 mg) may cause cardiac function impairement. Reversible on withdrawl.

Message

Let us forget our anxieties and worries for the time being and cheer up the release of the much awaited drug L1-Kelfer!

In India we are lucky to have the choice between the two drugs Desferrioxamine and Deferiprone. The first, being very expensive, is beyond the reach of most of the families. The discomfort faced by our children while taking the prick almost every night is also disturbing.

CIBA-GEIGY continues to ignore the requests made by Indian Thalassemic Associations to reduce the price of Desferal. In such conditions the availability of Kelfer in our country is indeed a matter of relief for all thalassemics.

Since chelation is very important for the growth and the good quality of life it is imperative for the parent to decide between the two drugs - Desferal or Deferiprone. The final choice lies with them.

CIPLA needs to be congratulated. It has made us proud to release the drug for the first time in our country. We HOPE that the welfare of thalassemics is their primary concern.

We wish all the best to our thalassemia children and hope that no child is left without chelation in the near future.

SHOBHA TULI

Report from Burdwan

On 8th May the International Thalassemia Day, Thalassemia Welfare Society of Burdwan organised a cycle rally, riders carrying the posters on Prevention of Thalassemia. A documentary film for social awareness, a blood donation camp and a cultural evening was also organised on this day.

National Thalassemia Welfare Society

Initiates

Dr. B. N. Dara Award

N.T.W.S. initiates Dr. B.N. Dara award to an Indian Medical Professional for dedicated work in the field of Thalassemia. Dr. B.N. Dara is a retired leading paediatrician of Jaipur.

Award carries a cash prize of Rs. 1500/-, citation along with a momento.

The Blood Transfusion Day

On the Blood Transfusion Day,
I go with joy and gay,
Where I meet my friends,
And learn how to adopt happy trends.
When the needle pricks in my hand,
All tell me not to cry,
So that the temprament of doctor
Does not go high.
When blood enters in my hand,
I feel that someone irrigates dry land.
When my transfusion is over,
I feel myself as strong as bulldozer.
Again, I feel joy and gay,
As there is no other way.

(Jagdeep Singh)
851, Phase-II
S.A.S Nagar

Report from Chandigarh

Celebration of International Thalassemia Day on May 8, 1995

The Thalassemic Children Welfare Association circulated leaflets, booklets & brochures and the articles were contributed for publication in major news papers.

A seminar addressed by the eminent Doctors of the P.G.I., Social Workers & Spokesman of the T.C.W.A. was organised on 8th May, 1995 in Govt. Senior Higher Secondary School, Mohali, which was attended by the grown up students, educationist & representatives of social and religious organisations.

Dr. R.K. Marwaha, Addl. Professor, Department of Paediatrics, P.G.I., Chandigarh advised that before marriage, the individuals should undergo Thalassemic screening tests, facility for which is now available at P.G.I. Dr. S.K. Agnihotri, Incharge Blood Bank, P.G.I., Chandigarh elaborately explained the shortage of quality blood to meet the requirement of Thalassemic and other patients. She appealed the community to volunteer for blood donations. Smt. Usha Talwar, Social Worker, Blood Bank Society, P.G.I., Chandigarh also delivered a motivational talk for blood donation & cited some examples, when the individuals donated the blood more than 40 times and are still hale and healthy. The Principal, Govt. Sr. Higher Secondary School, Mohali, advised the students and teachers to carry out the message to their dears and near ones and to adopt preventive measures. Mr. M.S. Rekhi, Vice President, T.C.W.A. informed that the association has been giving financial assistance to the patients belonging to weaker section of the society. He appealed for financial assistance to the association in cash or kind and also adopt some needy patients, for which the society has to come forward.

A blood donation camp was held in P.G.I., which was inaugurated by the Director Dr. B.N.S. Walia. More than 244 persons (parents, relatives & friends of the patients) donated the blood. Same day the Thalassemic Children who participated in painting, poems, story writing competition etc. were encouraged in the prize distribution function. The students who got more than 80% marks in their final examinations were also honoured. The association organised another camps on May 12, in High Ground, Air Force Station, Chandigarh in which more than 90 donors participated. Third consecutive camp was conducted on June 8, 1995 in the P.G.I., Chandigarh in which 415 volunteers donated blood. Fourth blood donation camp in this series will be held on 29th July, 1995 at P.G.I.

NATIONAL UPDATE ON THALASSEMIA

October 7-8, 1995 P.G.I., Chandigarh

A 'National Update on Thalassemia' is being organised by the Department of Haematology, Paediatrics and Transfusion Medicine, P.G.I. Chandigarh and the Thalassemia Children Welfare Association on October 7-8, 1995. Various aspects of the epidemiology, molecular defects, diagnosis and management will be discussed by eminent experts working in this field. Special sessions will be arranged for open interaction between parents, patients and the guest faculty.

For further details contact:

Dr. R.K. Marwaha, Organising Secretary, Department of Paediatrics, PGIMER, Chandigarh-160012, Phone (0172) 541031-39 Extn. 506, 236; Fax (0172) 540401.

FIT News

International Thalassemia Day Celebrations by FIT

Distinguished speakers during the meet
Left to right – Dr. V.K. Khanna, Head Thalassemia Unit, S.G.R. Hospital, Prof. I.C. Verma, Prof. V.P. Choudhry, Prof. S.K. Sarin, Deptt. of Gastroenterology, G.B. Pant Hospital

National Thalassemia Welfare Society and Thalassemics India celebrated International Thalassemia Day jointly on Sunday, the 7th May, 1995 under the banner of FIT.

Early in the morning a blood donation camp was organised at Ashok Vihar, New Delhi in collaboration with IMA, Delhi North Zone and Ashok Vihar Yuva Munch. It was inaugurated by D.C.P. Mr. Karnail Singh who also donated blood on the occasion. All the donors were given a gift of 2 Pearl Pet jars in a carry bag printed with "Donate Blood" & "Prevent Thalassemia" on either side. Around 200 volunteers donated blood for the noble cause. Active participation of Dr. H.C. Gupta, Mr. Raj Kumar Garg and Dr. Gautam Bose was highly appreciable.

A Child-Doctor-Parent meet was organised in the evening at Constitution Club, New Delhi. Dr. I.C. Verma, Dr. V.P. Choudhry, Dr. Vijay Sarin and Dr. V.K. Khanna held a panel discussion and satisfied the quest of audience. A need was felt that all the voluntary blood donation be screened for Hepatitis C. Moderator of the discussion Dr. V.P. Choudhry informed the house that he has already approached the authorities to make the Hepatitis C screening mandatory in all the blood banks.

Prizes and gifts were given to over 20 Thalassemic children who attended any distinction in any activity at their school and to those who performed any cultural activity on that day followed by Lucky draw prizes.

All the Thalassemics were given a carry home gift of 'audio cassettes'.

The meeting ended with a contributed dinner.

This issue is sponsored by CIPLA LTD.

National Thalassemia Welfare Society (Regd.)

(R. No. S/26823, Registered under Societies Registration Act XXI of 1860)
KG-1/97, Vikas Puri, New Delhi-110 018. Phone: 550 7483 Fax: 91-11-559 1202, 559 8879

NATIONAL THALASSEMIA WELFARE SOCIETY

COMMITTED FOR

A AWARENESS IN THE COMMUNITY

B BETTER FACILITIES

C COST REDUCTION IN THERAPY

D DRUGS AND ANCILLARY FACILITIES

E EDUCATION

F FAMILY SUPPORT

G GREATER COORDINATION AMONG SOCIETIES IN INDIA

MEMBERSHIP

Any person can become a member of the society.

Charges	Inland	Foreign
Patron :	Rs. 5000	$ 500
Life :	Rs. 500	$ 50
Annual :	Rs. 100	– —

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	Inland	Foreign
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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

Note: Cipla has kindly agreed to open from 1-4 P.M. on this day for Kelfer supply.

विशेष थैलासीमिया क्लीनिक

नैशनल थैलासीमिक वैलफेयर सोसाईटी की ओर से ***प्रतिमाह दूसरे रविवार*** को एक विशेष थैलासीमिया क्लीनिक का आयोजन ***लाजपत भवन, लाजपत नगर,*** निकट विक्रम होटल/मूलचन्द फ्लाई ओवर में किया जाता है।

सुविधाये:-

- नियमित वृद्धि परीक्षण
- लोह निष्कासक दवा
- फैराटीन परीक्षण मात्र 150/- रूपये में
- Hepatitis inj.
  10 वर्ष से कम आयु के लिये 175/- रूपये
  10 वर्ष से अधिक आयु के लिये 350/- रूपये
- थैलासीमिया माइनर/मेजर की जांच

पूर्व समय निर्धारण के लिये 5507483 पर सम्पर्क करें।

नोट:- इस दिन कैलफर दवा के वितरण के लिये सिपला अपना कार्यालय 1–4 बजे तक खोलने के लिये सहमत हो गई है।

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