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FEDERATION of Indian Thalassemics

National Thalassemia Bulletin

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2nd National Thalassemia Conference '97

CARE And CONTROL

27th – 28th December, AIIMS, New Delhi

Eminent speakers on Dias during panel discussion at the end of day *(Report inside)*

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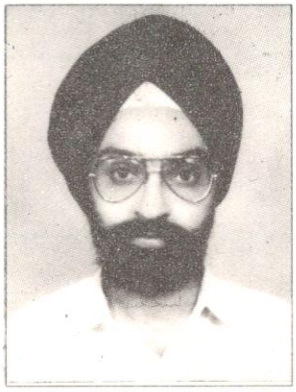
EDITORIAL



Once again summer is approaching fast and you can see long queues of desperate, anxious, tired, dejected parents of Thalassemic children at Blood Banks. A few lucky enough to get a pouch of blood while others have to take a chance for the next day, day after and day after.....

Blood Bank authorities are not to be blamed for this. Climatic conditions & Exams followed by Vacations in colleges inhibit the procurement of blood. Conditions will be worse this summer since Supreme Court has ordered a ban on professional donors; though a laudable step, it will further increase the scarcity of blood.

Thalassemic parents has to play an important role in motivating their friends, relatives and general public for voluntary blood donation. They should also help their local Society in organizing blood donation camps.

Thalassemic Children Welfare Association, Chandigarh collected over 3000 units of blood in 1997 which is almost 50% of their requirement for over 300 patients taking transfusion at PGI, Chandigarh. If Thalassemic parents of other cities also co-operate with their Societies, blood situation can be improved there as well.

It is estimated that if only 5% of healthy population donates blood even once a year nobody will die just because of shortage of blood. Overall India 60 lakh units of blood are required annually, however collection is just 30 lakh units and the shortage is likely to increase to 40 lakh units by the end of the century. In India just 0.3% people donate blood once a year while voluntary blood donation is 5% in U.S.A & 10% in Switzerland.

Above figures show the ignorance, lack of awareness & fear psychosis among the Indian community. **May God! good sense prevail among Indians so that they also volunteer themselves for regular blood donation.**

Dr. J.S. Arora

Bone Marrow Transplantation In Thalassemia

— *Guido Lucarelli, Claudio Giardini and Emanuele Angelucci (Pesaro, Italy)*

Thalassemia refers to various types of hereditary anemias identified by a reduced production of one of the globin chains that form the haemoglobin molecule. In ß Thalassemia there is a deficient or absent synthesis of ß globin chains that constitute the adult hemoglobin molecule which causes several deleterious effects on erythrocyte production and survival. Haemolysis and ineffective erythropoiesis lead to a chronic anaemia with erythroid marrow hyperplasia: this determines an increase in the plasma iron turnover and increased iron absorption. A progressive iron overload is associated with the above mechanism and is the manifestation of packed red cell transfusional regimen adopted to correct the anaemia. Transfusion and regular iron chelation with deferoxamine constitute the conventional treatment for severe ß Thalassemia. Homozygous thalassemia which once resulted in early death has become a chronic disease compatible with prolonged survival although it remains a progressive disease.

Bone marrow transplantation

The first successful transplant in ß Thalassemia was performed in Seattle on December 5, 1981 in an untransfused 14 month old child. At the same time a 14 year old thalassemic patient who had received 150 red cell transfusions was transplanted at Pesaro in December 17, 1981, but he had reoccurrence of thalassemia after rejection of the graft. The experiences that followed, which were promptly reported in the literature by the group of Pesaro, were disappointing, with the first series of patients transplanted using high doses of cyclophosphamide and total body irradiation (TBI) showing a high percentage of failures related to marrow rejection and early toxicity. From 1983, a modification of a conditioning regimen originally proposed by Santos for Leukemia was adopted. This included a combination of busulfan (BU) and cyclophosphamide (CY) without use of radiation. In 1990, a retrospective evaluation made in 222 consecutively transplanted patients led us to categorize patients under 16 years of age into three prognostically different classes of patients. The risk factors considered included the presence of hepatomegaly (enlargement of more than 2 cm below the costal margin), the presence of liver fibrosis in the pretransplant liver biopsy, and the quality of iron chelation received before transplantation. The quality of chelation was considered adequate when deferoxamine therapy was initiated within 18 months after the first transfusion and administered subcutaneously for 8-10 hours for at least five days each week. Chelation was defined as inadequate if there was any deviation from this requirement. As of September 1995, 761 transplants in thalassemic patients have been performed in Pesaro, 714 from HLA identical siblings, 22 from phenotypically identical parents, 24 from partially matched family members, and one from an HLA-identical unrelated donor. In this review we update our experience in bone marrow transplantation in Thalassemia from HLA-identical donors in 697 patients, with the last patient transplanted on December 29, 1994, and the analyses performed on September 30, 1995.

Bone Marrow Transplantation in Class I patients

Class I patients are identified by absence of hepatomegaly, regular iron chelation therapy performed before transplant, and absence of fibrosis at the pretransplant liver biopsy.

Between June 1983 and December 31, 1994, 111, Class I patients under the age of 16 years were transplanted using a conditioning protocol consisting of BU 14 mg/kg and CY 200 mg/kg. The graft versus host disease (GVHD) prophylaxis consisted of weekly methotrexate (MTX) before December 1985 and cyclosporine A alone from this date onwards. The median age of this group of patients was 4 years (range 1-16 years);the median number of transfusions received pretransplant was 40 (range 4-304; the median serum ferritin level was 1171 ng/ml (83-5200). Thirty percent of patients had serological markers of Hepatitis B (HBV) infection and 31% (10 out of 32 patients tested) had evidence of Hepatitis C(HVC).

In patients receiving marrow transplants for thalassemia, liver fibrosis has never been observed before the age of three years. In view of the known hazards of the liver biopsy procedure in very young children, patients under the age of 3 years did not undergo liver biopsy unless hepatomegaly was present; such infants were considered not to have liver fibrosis. In the remaining 79 patients , a pretransplant liver biopsy was performed. Liver iron overload was graded with semiquantitative estimation according to previously published criteria. Using this procedure, 42 patients (53%) had a mild, 35 (44%) a moderate and two patients (3%) a severe liver iron overload. A quantitative estimation of the liver iron content has been obtained for 28 patients; the median liver iron concentration was 7.0 mg/gm of dry tissue (range 1-24).

In a Kaplan-Meier analysis, the probabilities of survival, event free survival, rejection and non rejection mortality are respectively 95%, 90%, 5% and 5% with a maximum follow up of about 12 years. Five patients died from causes related to the transplant. Six patients experienced rejection of the graft and all reconstituted autologous marrow. The last death occurred on day 1244, and the last episode of rejection on day 365. Two additional patients died from causes unrelated to the transplant

procedure and the disease itself (car accidents) at day +933 and +3190; these patients have been censored from the Kaplan-Meier analysis at the day of the event.

Thirty patients (27%) presented with acute graft-versus-host disease AGVHD, grade II to IV, and 12 patients (12%) developed a clinical form of chronic GVHD. From Januay 1986, all class I patients had received AGVHD prophylaxis consisting of cyclosporine A, 5mg per kilogram intravenously daily from day 2 through day 5, followed by 3 mg per kilogram daily until the patient was unable to tolerate oral administration at a daily dose of 12.5 mg per kilogram. A dose of cyclosporine was then tapered from day 60 until the drug was discontinued after one year (protocol 6).

Bone marrow transplantation in class II patients

Between June 1983 and December 1994, bone marrow transplants were performed in 293 class II patients. These patients are identified by the presence of either hepatomegaly,a history of irregular chelation performed before transplant, or histological evidence of liver fibrosis, or various combinations of two of the above risk factors. The mean age was nine years (range: 1-16), the approximate total number of transfusions was 116 (range: 2-430), and the mean serum ferritin level was 1913 ng/ml (33-8547). One hundred and sixteen patients (39%)had serological markers of hepatitis B infection, and 40% (24 of 60 patients tested) of hepatitis C. The pretransplant liver biopsy (total number of biopsies performed and evaluable:273 showed in 64 patients (23%) a mild, in 162 (59%) a moderate, and in 47 (17%) a severe iron overload. Liver fibrotic damage was absent in 69 patients (26%), mild in 98 (37%), moderate in 75 (29%), and severe in 23 (9%). Quantitative estimation of the liver iron content has been obtained for 51 patients; liver iron concentration ranged from 0.8-43 mg/gm, with a median of 10.0. This group includes 3 patients who, although affected by a genetically different disease (double heterozygotes, Sickle cell/β thalassemia) , were all regularly transfused and chelated from a young age; these patients have been evaluated and conditioned for transplant following the same strict schedule already in use for thalassemia.

The probabilities of survival, event-free survival, rejection, and non rejection mortality are respectively 86%, 82%, 6% and 15% with a maximum follow-up of 12 years. Forty Three patients died from transplant-related causes ; 22 patients died within the first 100 days (most of them during the aplastic phase and, during the early engraftment, from septic-haemorrhagic causes an acute GVHD); and 21 patients died after the first 100 days, 17 of them during the first year while on immunosuppressive therapy with cyclosporine A. Four patients died after the first year post transplant: 2 of them from complications of a severe form of chronic GVHD, and 2 from septic shock that suddenly caused death in a 24 hour period after the first symptoms began, most likely related to the splenectomised state. An additional patient died from accidental causes at day +1700, and he has also been censored from the analysis at the time of death. Seventeen patients rejected the graft: 12 of them have survived with autologous reconstitution, under transfusional support. Seventy seven patients (28%) presented with acute GVHD, grade II to IV; 44 patients (17%) developed chronic GVHD.

Bone marrow transplantation in class III patients

Between June 1983 and March 1989, 55 patients were included in class III, since they presented all the aforementioned risk factors and had been transplanted after a total dose of BU 14 mg/kg. The results of this clinical experience, when analysed in 1990, were considered unsatisfactory because of high incidence of early mortality due to toxicity and infections. It was thought that this toxicity was a consequence of high dose CY in patients with preexisting liver damage, and admission of class III patients to protocol 6was interrupted.

Beginning in March 1989, new conditioning protocols with a reduced dose of cyclophosphamide were introduced in search of less toxic adverse effects and according to the experiences in malignancies. Between March 1989 and December 31, 1994, 110 patients were included in class III. The median age of the overall group, historical and post-March 1989 (165 patients), is 11years (range 3-16),the median number of pretransplant transfusions 150 (range 4-435), the median serum ferritin level 3,187ng/ml (range 604-17,450). Ninety four patients (57%) had serological markers of hepatitis B infection and 46% (38 out of 82 patients tested) of hepatitis C. The pretransplant liver biopsy showed a mild iron overload in 16 patients(10%),a moderate form in 60 patients (35%), and a severe iron overload in 89 patients (55%); 41 patients (24%) had mild fibrosis, 54 (32%) moderate, and 70 (44%) a severe fibrotic liver damage. Quantitative estimation of the liver iron content has been obtained for 35 patients; liver iron concentration ranged from 3.0 to 47.0, with a median of 15.1 mg/gram.

The conditioning regimens studied since March 1989 included BU 14-16 mg/kg, a dose of CY variable from 120- 160 mg/kg and, in some protocols, the use of antilymphocytic globulin (ALG). In a Kaplan-Meier curve, the probabilities of survival, event free survival, rejection and non rejection mortality for this group of 110 patients are 77%, 54%, 33% and 20% respectively, with a maximum follow-up of six years (Figure 4). Twenty three patients died from causes related to the transplant. Thirty three patients rejected the graft; 25 of them are alive with return of the thalassemia, and one patient has been successfully retransplanted. Fifteen patients (16%) developed Grade II to IV acute GVHD, and seven 10% developed chronic GVHD. The most recent analysis of a Kaplan-Meier curve including the subgroup of the 48 most recent class III patients, transplanted since June

1992 after busulfan 14mg/kg and a total dose of 160mg/kg of cyclophosphamide and with a maximum follow-up of 37 months, shows 85% survival, 68% event free survival, 25% rejection,and 15% non rejection mortality. From the data presented here, the introduction of new protocols with a reduced dose of CY represents a promising step in the attempt to reduce the transplant related mortality in this patient population. In fact, mortality is reduced from 37% to 20% (15% with the last modification of the conditioning regimen), although the rejection rate at the same time varied from 13% to 33%.

Bone Marrow Transplant in adult Thalassemic Patients

The early experience with transplantation for patients over 16 years was disappointing, with 4 of 6 patients dying of AGVHD and 2 patients dying 9 months and 6 years , respectively, after recurrence of the thalassemia [16]. In October 1988, bone marrow transplantation in patients older than 16 years was restarted after categorization of the patients into class II or class III and the adoption of the protocols used for these classes in younger patients. Through December 31, 1994, 86 adult thalassemic patients have been transplanted. The age ranged from 17 to 32 years (median 19). The median serum ferritin level was 1888 ng/ml (range: 322-9071). Sixty-two patients (73%)had serogical markers of hepatitis B infection and 87% (59 out of 68 patients tested) of hepatitis C. Liver biopsy was performed before transplant in all 86 patients. Eighteen patients (21%) had mild,41 (47%) moderate, and 27 (31%) severe iron overload; with regard to fibrosis, 26 patients (30%) had mild, 20 (23%) moderate, and 40 patients (46%) severe liver fibrosis. In some of these patients, the histological documentation was liver cirrhosis. all patients had a history of irregular iron chelation at the time of the pretransplant evaluation. This group also includes two patients affected by double heterogeneous sickle-cell/ß-thalassemia. By a Kaplan-Meier analysis, the probabilities of survival,event-free survival, rejection and non rejection mortality are 66%, 63%, 3% and 35%, respectively. Twenty-seven patients (32%) died from causes directly related to the transplant, mainly of sepsis or hemorrhage within the first 100 days. One patient, who was splenectomised before transplant, died from septic shock on day +1363. Two patients experienced rejection of the allogeneic marrow and are alive with complete autologous reconstitution. Twenty-three patients (27%) presented with acute GVHD. grade II to IV, and 10 patients (16%) developed chronic GVHD.

Mortality and causes of death

A total of 145 patients (21%) died. The incidence of infections was 37% (54 patients), with a prevalence of fungal (23 patients) and viral (18 patients) infections. The large majority of these lethal infections occurred during the period of bone marrow aplasia. Thirteen patients did not recover autologous hemopoeitic reconstitution after rejection of the allograft and died of causes directly related to the prolonged marrow aplasia. Acute and chronic GVHD have been direct or indirect causes of death in 34 patients (23%).An unusual cause of death observed only after bone marrow transplantation in thalassemia, has been sudden cardiac tamponade, which occurred in six cases (4%). The mortality for veno-occlusive disease in this patient population has been lower than expected in transplants done in patients with a high rate of liver damage and with BU as part of the preparative regimen. Three patients had late fulminant infections with fatal sudden shock. This complication has been attributed to the fact that these patients had been splenectomized years before the transplant and therefore could have had a higher susceptibility to encapsulated bacterial infections. For this reason immunisation against encapsulated bacteria such as pneumococcus, haemoplilus and meningococcus and/or long term antibiotic prophylaxis is highly recommended in these patients. Four patients who experienced rejection of the allograft and had complete autologous reconstitution died from causes related to progression of the disease itself (cardiac and infectious causes) between day +2213 and day +3180 post transplant. Four patients died from causes unrelated to the transplant and the original disease (car accidents); these patients as already mentioned have censored from the Kaplan-Meier analysis at the time of the event.

Conclusions

Today allogeneic bone marrow transplantation represents the only therapeutic modality for the eradication of ß-Thalassemia major. Several bone marrow transplant centres have ongoing clinical experience in this setting. The first thalassemic patient was transplanted more than 13 years ago. Since then the results of marrow transplantation have improved steadily, with major progress in the management of transplant related complications. This is due to the use of cyclosporine, more effective treatment of cytomegalovirus infection, improvement of aseptic techniques, and evolution of systemic antibiotic therapy. At the moment the patient in class I has a 5% probability of mortality, 4% probability of rejection and 91% probability of disease free survival postransplant. In patients in classes II and III,the organ damage related to iron overload and to acquired blood borne viral infections, is more advanced and therefore transplant related mortality is higher. However these are patients who have developed progressive and significant organ damage while receiving conventional treatment. The survival expectations of such patients are poor in the absence of intervention by marrow transplantation. Hopefully in the future the improved management of graft-versus-host disease and the development of technologies for bone marrow transplantation from unrelated donors may expand the pool of potential candidates.

2nd National Thalassemia Conference '97

National Thalassemia Welfare Society in collaboration with Dept. of Haematology, AIIMS organised 2nd National Thalassemia Conference at Jawahar Lal Nehru Auditorium, All India Institute of Medical Sciences, New Delhi.

Federation of Indian Thalassemics, Indian Society of Haematology & Blood Transfusion, Paediatric Haemato-Oncology chapter of I.A.P., Indian Academy of Paediatrics, Delhi Branch & Delhi Society of Haematology were also associated with it. Wide publicity for its organisation was given in official publications of these organisations & through liberal distribution of brochures.

The Conference was inaugurated by Dr. P.K. Dave, Director, AIIMS. While inaugurating the Conference he said "Thalassemia is a commonest inherited disorder in India & yet the general population is not aware of disease & its inheritance". He said Medical Institute is fully aware of the magnitude of this disorder & many Faculty members have dedicated themselves to provide facilities for Control of Thalassemia.

In her Presidential Address, President of the Society, Ms. Surrendar Saini expressed deep concern about the pathetic condition of Thalassemics. She stressed the need for voluntary blood donation & reduction in cost of treatment. She thanked Mr. Harish Chawla, Director, CIPLA Ltd. for taking keen interest in welfare of Thalassemics and reducing the cost of Kelfer by almost 40%.

Dr. V.P. Choudhry, Prof. of Haematology, AIIMS said "The present Conference will provide a forum to update the knowledge of doctors and parents in various facets of thalassemia. It will facilitate in improving the care of thalassemics in this country as the doctors and parents will be well equipped with the current state of the art".

Mr. M.S. Rekhi, President of FIT in his address said " Such conferences addressed by International and National experts of repute , who are working tirelessly in Thalassemic field help not only Paediatricians and Haematologists but also parents of thalassemic children who struggle hard to maintain their children and are always anxious and interested to know advances being made in this field which give them a ray of hope for the future".

Prof. Mammen Chandy, Prof. & Head of Haematology, Dept. of Medicine, CMC Hospital, Vellore receiving 1st Dr. B.N. Dara Award from Prof. Manorama Bhargava, Prof. & Head, Dept. of Haematology, AIIMS, New Delhi. Mr. Kamal Manchanda enjoying the celebration.

Dr. J.S. Arora, General Secretary of the Organising Committee and National Thalassemia Welfare Society gave an overall view of the Society & its activities. He announced the first Dr. B.N. Dara Award conferred to an eminent Haematologist Dr. Mammen Chandy for his pioneer work in BMT in Thalassemia in India. He also announced the best Social Worker Award to Mr. Ashok Sachdeva for his selfless services.

Prof. M.C. Maheshwari, Dean of AIIMS released the Souvenir which contained abstracts and articles by eminent participating faculty.

Dr. V.N. Sardana, Joint Director, NACO (National AIDS Control Organisation), Dr. George J. Kontoghiorghes, inventor of 1st oral iron chelator, Deferiprone, Dr. Harish Chawla, Director, Cipla Ltd. and Dr. J.S. Arora, General Secretary, NTWS discussion the current status of Thalassemia Management in India

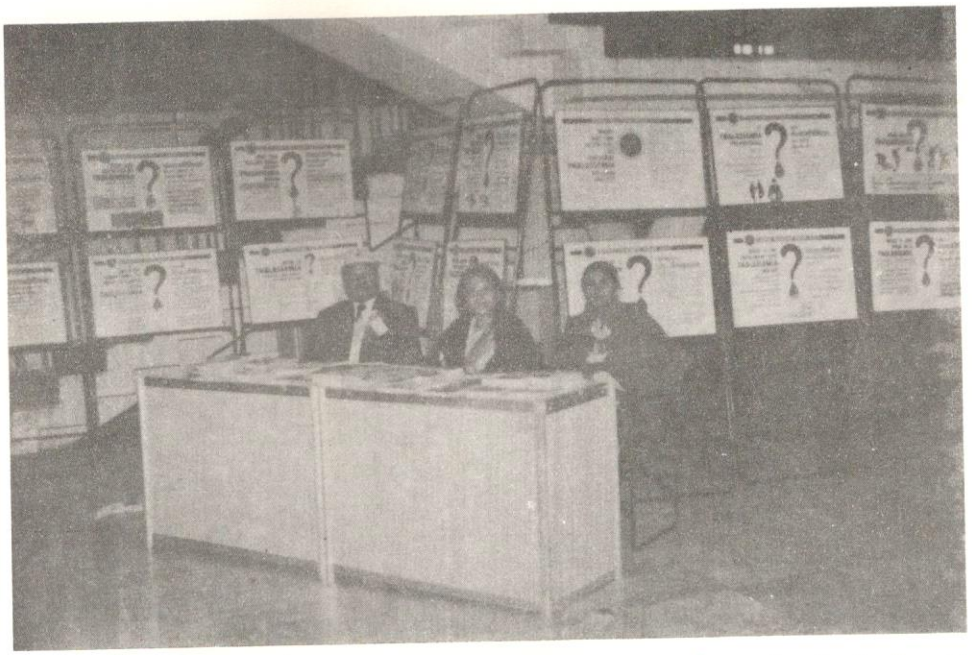
The message from TIF Chairman, Panos Englezos was addressed during the inaugural session. While congratulating the Chairman & members of Organising committee for their initiative and tireless efforts in organising this great Scientific event he stated " I would like to stress that such efforts are highly appreciated and form the basis for ameliorating the devastating problems associated with this hereditary disorder".

Symposium on following topics were held: Epidemiology & Diagnosis of Thalassemia, Transfusion Therapy, Complications of Thalassemia, Transfusion Transmitted infections, Stem Cell Transplantation, New Developments, Splenectomy, Doubts in Thalassemia, Chelation Therapy, Role of Thalassemic Societies, Thalassemia Control Program & Psychological Problems.

Talk on BMT in Thalassemia by Dr. Mammen Chandy & International data on Deferiprone by Dr. George J. Kontoghiorghes was well attended & highly appreciated. Thalassemic families were excited to hear Dr. N.K. Mehra when he stated that "We have recently established Asian Indian Donor Marrow Registry of unrelated voluntary donors for those patients who fail to find an HLA identical sibling in the family. Data on 1500 HLA typed healthy unrelated individuals is already in the computer". So far one patient could find an HLA identical donor". He said we have a goal of registering 1 million volunteers from all ethnic & racial background. AIDMR has also established links with most International Registries through BMDW Computer Network. The primary researches are made free of cost and further investigation if required are chargeable by the Donor Centre.

While presenting the data on Thalassemia in North India, Dr. Mamta Sharma, Head of Paediatrics, DDU Hospital presented a beautiful study on 373 thalassemic children attending Thalassemia Clinic run by National Thalassemia Welfare Society at Lajpat Bhawan, Lajpat Nagar, New Delhi.

In her study she found 61.39% belonged to families migrated from West Pakistan. Hepatomegaly was more marked than Splenomegaly. At the time of registration only 30.43% were monitoring Hb more than 9%. It was concluded that proper follow-up of these patients had a



Mr. Ashok Sikka, Ms. Monisha & Ms. Monika at the Exhibition Stall put up by the Society. Displayed panels showing Problems, Magnitude & Prevention of Thalassemia both in English & Hindi

positive impact on improving their Hb level & proper iron chelation.

Dr. V.N. Sardana, Joint Director, NACO spoke on the National Policy on Blood Transfusion Services & Strategic plan for Prevention & Control of AIDS in India. He said during past 3 years 154 Zonal Blood Testing Centres have been established providing linkages to other blood banks for testing HIV. He said "NACO is providing financial support to all the public sector blood banks by giving contingency grant for purchase of test Kits, reagents, glassware & blood bags". On the basis of Hospital beds, with a criterion laid down by WHO of 6-16 (average 11) units of blood per hospital bed annual requirement is estimated as 60 lac units/annum & there is shortage of blood to the extent of 50% at the existing level, shortfall is likely to increase by about 40 lac units by the end of Ninth Plan" he said.

Dr. M.B. Agarwal a renowned name among Thalassemic families in India & most experienced person in oral Iron Chelator in India presented a study on Combination Chelation Therapy of Deferiprone & Desferrioxamine on 16 patients. Combination therapy was offered to patients who could not afford full dose of Desferral or where Kelfer alone was either ineffective or it caused significant dose related side effects. All except 2 patients on Combination therapy could continue to treatment without adverse effects and with good efficacy.

Prof. S.K. Sareen, Prof. & Head, Dept of Gastroenterology, G.B. Pant Hospital while speaking of Hepatitis B status in India said " Blood Transfusion expose these patients to a high risk of HBV infection, specially in India, because of the i) High prevalence of HBV carrier state in India ii) Use of blood from professional donors in India who have high HBV prevalence rates (10-20%) iii) fact that blood is not screened for HBV in all blood banks iv) the test kits are insensitive v) Presence of mutant forms of HBV.

Over 400 patients, parents & medical professionals participated. An open dialogue session in Hindi was also held at the end of session on both days for active participation of parents and patients with experts. The events were widely covered in leading newspapers.

International Thalassemia Day (8th May)

FIT will be celebrating International Thalassemia Day on Saturday, the 9th May, 5 p.m. onwards at Constitution Club, Rafi Marg, New Delhi–110001

- Panel Discussion
- Awards to Outstanding Thalassemia Children
- Cultural Activity
- Dinner

Contact: Dr. J.S. Arora
Ms. Monisha
Ms. Sunita (AIIMS)

Ms. Shobha Tuli
Ms. Vandana (SGRH)

INAUGURATION OF NEW BLOOD TRANSFUSION ROOM FOR THALASSEMICS IN P.G.I. CHANDIGARH

Dignitaries at the Dias during the inauguration ceremony. Left to right Mrs. Shobha Tuli, Board Member, TIF, Mr. M.S. Rekhi, President, FIT, Dr. Lata Kumar, Prof & Head, Dept. of Paed., PGI, Prof. B.K. Sharma, Director, PGI, Prof. B.N.S. Walia, Former Director, PGI, Dr. J.S. Arora, General Secretary, FIT, Dr. S.K. Bhattacharya, Principal of TTT Institute

TCWA Chandigarh functioning for the last 10 years was facing acute shortage of space for providing even modest services to Thalassemics due to increase in their number to 340 from about 70 in 1987. On the request of TCWA, the management of PGIMER, Chandigarh allocated 765 sq.ft. space in new Advanced Paediatric Centre (A.P.C.) (an imposing architectural landmark inaugurated by the President of India in 1997).

TCWA decided to furnish this room suitably by raising funds from the parents of Thalassemics and other benovolents at a cost of Rs. 6.35 lacs. The blood transfusion room has been air conditioned and is furnished with ultra modern furniture, fridge and water cooler for the comfort of Thalassemic children. Two jumbo-sized televisions with VCR & music system have also been installed in the room for the entertainment of children during their stay. The walls are decorated with paintings & sceneries to give aesthetic touch to the room. The provision of a computer has also been made for keeping up-to-date record of each child for proper follow-up.

While speaking at this occasion, Director of PGI, Prof. B.K. Sharma said that they at the P.G.I. are all concerned for Thalassemics and try to go out of the way in providing medical services to Thalassemic children. He further indicated that in the Ninth Plan, they have proposed establishment of BONE MARROW TRANSPLANTATION UNIT. In his address Mr. M.S. Rekhi, Vice President, TCWA appealed to the doctors of P.G.I. to come forward in raising funds for the TCWA so that this association could also provide the life saving drug (Kelfer) to all the Thalassemics registered with it.

Dr. Mrs. Lata Kumar, Prof. & Head of Paediatrics stated that the department of Paediatrics at P.G.I. is always ready to help the association in this noble cause by providing timely medical advice to these children and their parents. They have a team of dedicated doctors under the leadership of Dr. R.K. Marwah, Additional Professor, Paediatrics who are fully devoted to the cause of Thalassemics.

Dr. B.N.S. Walia, Former Director of PGI in his address mentioned that he is happiest man to see this association grow with every passing year. In Democracy it is the collective voice which counts.

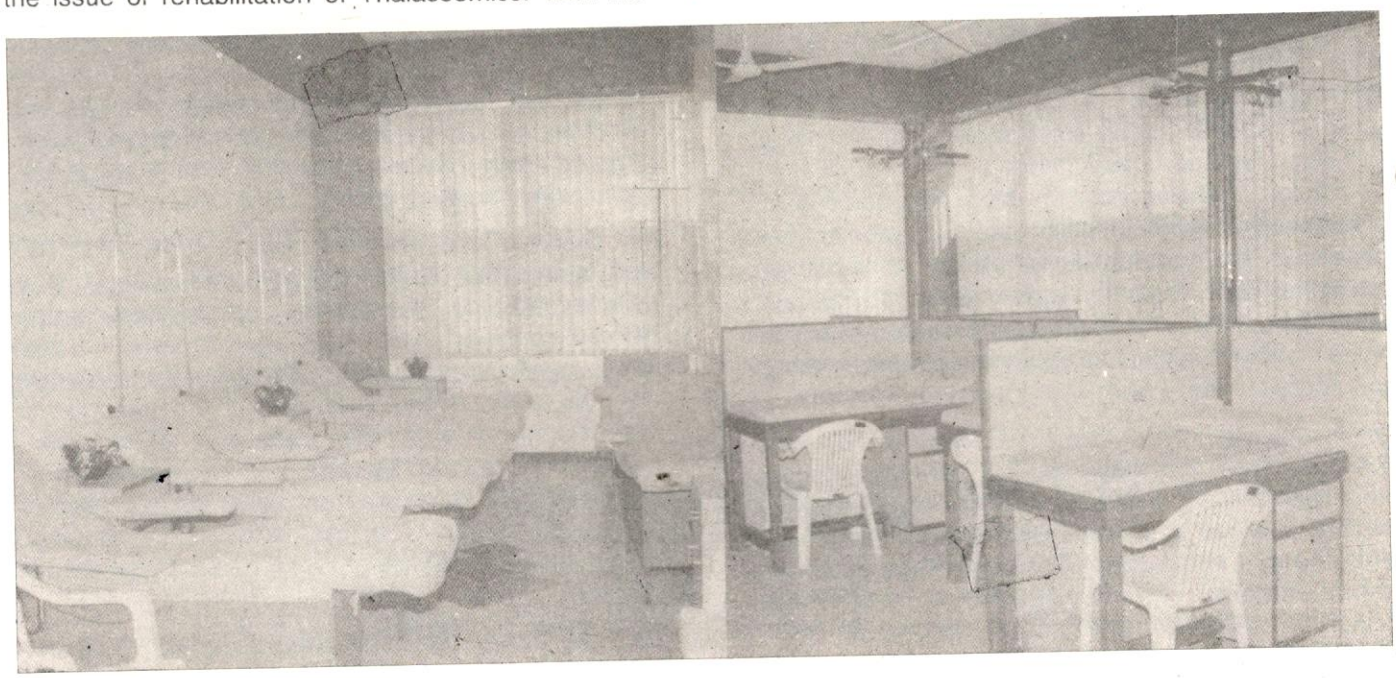
Mrs. Shobha Tuli a member on Board of Director of TIF also lauded the efforts of TCWA and PGI for this commendable job. She said that she happened to visit different countries and she can say with proud that this Thalassemic unit is not only the best in the country, but also the world over. While highlighting the role of Thalassemia International Federation (T.I.F), she mentioned that the organisation is trying to draw the attention of Govts. in different countries for effective awareness about the Thalassemia and its management through the good offices of World Health Organisation (W.H.O.).

Dr. J.S. Arora, General Secretary of FIT while congratulating TCWA & PGI for creating a separate well equiped transfusion unit for Thalassemics, also

Prof. B.K. Sharma, Director PGI, Chandigarh inaugurating the newly developed Thalassemia Unit. Others seen in the picture are: Prof. B.N.S. Walia, Dr. J.S. Arora, Dr. Lata Kumar, and Dr. R.K. Marwah, Addl. Prof. of Paediatrics, PGI, Chandigarh

highlighted the role of Federation of Indian Thalassemics (FIT) an All India body. He stated that the FIT was instrumental in getting the release of World's first oral iron chelator KELFER and reduction in its price recently. The FIT is trying further to reduce the cost through waiving of custom and other taxes. The FIT is co-ordinating with TIF for disseminating the latest knowledge about the management and prevention of this disease. Dr. Arora and a number of other speakers also raised the issue of rehabilitation of Thalassemics. With the better management facilities available at present, more and more Thalassemics are reaching adulthood. Thus there is need to think about their rehabilitation. The FIT is approaching the Govt. for providing them facilities at par with disabled persons, if possible without being labelled as disabled.

At the outset, Sh. S.S. Khattar, General Secretary of the TCWA read his report and dwelt on organisational arrangements of the association, facilities being provided



Newly developed, beautifully decorated 25 beded Thalaseemia Unit at PGI, Chandigarh

to the Thalassemics, educational and awareness programmes etc undertaken by the association. He further mentioned that TCWA is planning to raise a capital fund of about Rs. 2.00 crores for providing iron chelation therapy (Kelfer) to all the children registered with it. The Association is persuing the U.T. Adminstration for exempting sales tax on life saving drug i.e. Kelfer as done by Delhi Adminstration. He also stated that there is a need to start screening test facilities on a large scale at PGI, Chandigarh and other Regional Medical Centres such as Patiala, Amritsar, Rohtak and Shimla etc. and also establishment of thalassemia units at these centres,so that the children from far off places of these states need not travel long distances to Chandigarh for blood transfusion and medical advice.

Sh.S.P.Ajmani, the president of TCWA proposed vote of thanks to all the dignitaries for their presence at the occasion .He also expressed his thanks on behalf of the association to all those who donated funds liberally for furnishing this new Blood Transfusion Unit at the PGI and helped earlier in raising funds and holding blood donation camps. In the end a film on Thalassemia, "Fighting for the Red in the Blood" prepared by the Technical Training Teachers Institute, Chandigarh was released by Dr. S.K. Bhatacharya, Principal of the Technical Training Teachers Institute.

Report from Burdwan, Gosaipara

For treatment of Thalassemic children we have built a hospital in front of Agriculture Farm, at Burdwan, 100 Km. away from Calcutta. This hospital is the only heaven for the thalassemic children of whole North Bengal & Bhagalpur district of Bihar.

To build this hospital we have raised donation also from passengers of local train. We started the hospital with 10 patients and at present about 200 patients come regularly for treatment. Unfortunately, neither guardians of the child nor we have the resources to give them Desferral injection or oral Kelfer capsules. We are holding seminars with students of higher classes also requesting the prospective bride & bridegrooms to have premarital gene examination. On the other hand we are working house to house visit in the villages around Burdwan to survey the situation and asking them to have their gene to be examined.

In 1993 we held a camp for thalassemia screening and found 10% carriers at that time. We have taken continuous propaganda to see that the patients of thalassemia do not increase.

NATIONAL THALASSEMIA WELFARE SOCIETY (Regd.)

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

SPECIAL THALASSEMIA CLINIC

N.T.W.S. 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. 150/- for Children below 10 years
  Rs. 300/- for Children above 10 years
- Thalassemia Screening

For appointment contact:
Dr. J.S. Arora, Tel 550 7483

Note: Cipla has kindly agreed to open from 12.30 P.M.—4.30 P.M. on this day for Kelfer supply.

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Any person can become a member of the society.

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A GLOBAL BREAKTHROUGH IN THALASSAEMIA

The world's first oral iron chelator

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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