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FEDERATION of Indian Thalassemics

National Thalassemia Bulletin

Vol. 2 No. 1 March, 96

International Conference on Thalassemia New Delhi, 25th to 27th October, 1996

Distinguished International speakers:

Dr. Mike Augastiniotis	Cyprus
Dr. B. Wonke	London
Dr. M. Walker	London
Dr. N. Olivieri	Canada
Dr. D. Sanctis	Italy
Mr. Panos Englezos	Cyprus
Mr. G. Constantinou	Cyprus
Dr. T.N. Patric	Singapore

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- Hepatitis & Osteoporosis
- Cardiac Complications
- Oral Chelator & other novel treatments
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- Stem Cell
- Parents Associations

The Conference is open to medicos, para-medicos, patients, parents, social workers & others.

For any other informaiton contact:

Thalassemics India
C-1/59, Safdarjung Enclave, New Delhi–16.

Thalassemia Awareness Walk, Burdwan 27th February, 1996

Report Inside

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EDITORIAL

CORD BLOOD BANK

The London and South East Zone's new cord blood bank opened officially on 8 December 1995, based at the North London Blood Transfusion Centre. It will be the first of its kind operating, in full, within the National Blood service. (Other such facilities do exist, most notably in Newcastle and Bristol, but none of them has yet reached the position where they are actively collecting cord blood units and storing them ready for use by patients other than on a very small scale or for research purposes).

The new cord blood bank represents an additional treatment to patients suffering from disease such as leukaemia.

With the discovery that umbilical cord blood (or more commonly termed 'cord blood') contains stem cells, a promising alternative to bone marrow transplants has been opened up. For example, cord blood stem cells do not require such stringent matching to patients, so it is easier to find a suitable match; cord blood can be stored for a long period of time and is ready for immediate use (thus eliminating the need to trace a donor). Furthermore, the new cord blood bank intends to collect donations from the broadest possible ethnic mix, ensuring a better chance of a match being found compared to the current low rate of donors from ethnic minorities on the bone marrow register.

So far, the use of stem cells retrieved from cord blood has been limited to transplants to children suffering from diseases like leukaemia. (The amount of cells available in cord blood is quite small and would not be enough to treat adults. Work is already well underway to discover how these stem cells could be encouraged to grow in the laboratory so that sufficient quantities could be produced for adults.) Evidence so far has proved that the new treatment is effective. The Lancet recently reported the outcome of 44 sibling cord blood transplants. World-wide fifty transplants have taken place between patients and unrelated donors.

The cord blood bank is being housed in a specially converted of North London's Deansbrook Road site in Edgware, North London. It is situated alongside the Centre's established and innovative tissue bank facility.

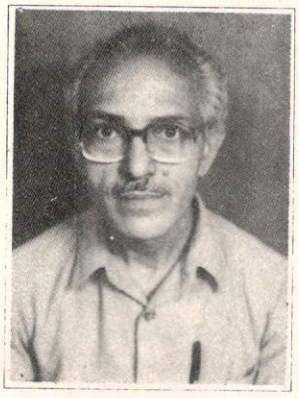
At Deansbrook Road, the cord blood bank aims to recover and store 5,000 donations from the placentas of babies born at the nearby Wellhouse NHS Trust. The hospital's ethics committee has approved the project. In each instance, mothers will have to provide formal consent for their umbilical blood to be used by the bank and their participation will be purely voluntary. Work is underway to ensure that all involved with the Trust's maternity services, as well as local GPs, midwives, hospital and community paediatricians and regional screening services, are aware of the new scheme.

Editor's Note: We understand that the first Cord Blood Transplant in a thalassemic in the U.K. will be carried out this year at the Hammersmith Hospital.

(From U.K.T.S. News Review)

PREVALENCE OF HEPATITIS C IN THALASSEMIA

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



Children with diseases such as aplastic anemia, PRCA, refractory anemia, leukemia and other malignancies, thalassemia and hemoglobinopathies require multiple blood transfusions. Among these disorders, thalassemia & hemoglobinopathies are the most common. All these children are at higher risk of developing blood transmitted infections. Among various infections hepatitis B, C, D & HIV have long term implications with a fatal outcome. Present communication deals with hepatitis C regarding its prevalence, natural course of the disease & its prevention.

Hepatitis C virus (HCV)is a RNA virus identified in 1989 and is responsible for majority of parentally transmitted non A and non B hepatitis. Presently only anti HCV is commercially available for its diagnosis and management. Initially the antigen (recombinant yeast polypeptide C 100-3) was against a small HCV genome and had a high false positivity. Now, several recombinant proteins from various regions of HCV genome have been developed for testing anti HCV with little false positivity (II/III generation Anti HCV tests).

PREVALENCE:

Prevalence of hepatitis C has been evaluated by several workers in multitransfused thalassemic children (Table I). Prevalence of hepatitis C has varied between 3-62.2% of cases. However, the data from various Institutions in Delhi over the last two years have revealed that prevalence of hepatitis C has increased significantly. In a pilot study, five hundred & fifty donors were screened for prevalence of hepatitis C at MAMC and associated hospitals, 11 of them (2%) were positive for anti HCV. Thus it clearly shows that prevalence of HCV has increased in donors which serve as the major source of HCV infection in those requiring multiple blood transfusion. Incidentally prevalence of hepatitis C in this pilot study is higher than prevalence of hepatitis B in some studies. However screening of blood for HBsAg is mandatory for all blood banks.

NATURAL COURSE OF HEPATITIS C

Wonke and her colleagues have observed that 10 of 21 children positive for anti HCV have progressed to chronic active hepatitis or cirrhosis. Similarly Resti & his colleagues over 13 years period in 78 thalassemics observed that over 55% of children progressed to chronic hepatitis. In the absence of long term studies on HCV infection in thalassemics, may I refer to a large multicentric study to elucidate the natural course of HCV infection in which it was observed that nearly 50% of patients developed histological evidence of chronic hepatitis. Elevated levels of ALT along with persistence of Anti HCV were observed in patients who progressed to chronic hepatitis. Ten to fifteen percent of patients progressed to cirrhosis & finally to hepatocarcinoma 7-18 years after the onset of NANB (HCV). While in a Japanese study the persistence of Anti HCV was observed in 75% of patients.

Our understanding of the natural course of HCV infection is evolving. Factors such as (a) age at exposure (b) duration of infection (c) degree of liver damage at first exposure, (d) risk of recurrent exposure etc. have greater influence on the progression of the HCV infection to the development of chronic active hepatitis, cirrhosis or hepatocellular carcinoma. The risk of development of cirrhosis is higher as compared with hepatitis B infection. Thus the hepatitis C infection is more serious infection as compared with hepatitis B and till date there is no vaccine with which the individuals at higher risk can be prevented.

MANAGEMENT:

Interferon is the accepted form of therapy for HCV infection. Initial studies referred as non A non B but which were later confirmed as cases of HCV

infection. Interferon was found to be an effective form of therapy. In the larger studies on 166 patients of non A non B hepatitis when treated with 3 million units of Interferon alfa-2b, three times a week for 6 months, resulted in normalization of ALT in 38% cases along with significant reduction in lobular and periportial inflammation. Interferon therapy has been used in small number of thalassemic children with hepatitis C. In all these studies the normalization of ALT levels and loss of HCV RNA in the serum were considered for the successful management. Forty to seventy percent cases responded to treatment but, unfortunately 40-60 percent of children relapsed on follow up. Alpha-2b Interferon has been approved by the FDA of USA as a drug for the management of hepatitis C. Unfortunately, it is expensive and cost of therapy for six months will be nearly one lakh per child. Such a high cost of therapy is beyond the reach of majority of families.

PREVENTION:

Hepatitis C can be easily prevented by screening all the donors and the blood found to be positive for Anti HCV by II or III generation tests should be discarded. It is of utmost important that the spread of hepatitis C should be controlled as (a) nearly 50% of patients with hepatitis C progress to cirrhosis (b) its cost of treatment is expensive & is beyond the reach of the majority and (c) nearly 50% of patients relapse soon after the therapy and (d) as yet there is no vaccine against hepatitis C. Thus there is an urgent need, that IAP should take up the matter with NACO or other Government Authorities so that routine screening of blood for hepatitis C is initiated and made mandatory for all blood banks with immediate effect.

Report from Ahmedabad

Annual Report of Thalassemia & Sickle Cell Society of Ahmedabad:

Two blood donation camps were organised by the members with the help of Shri Mahavir Jain Vidyalaya & other local group.

40 patients were screen for Hepatitis B & Hepatitis C also free of charge. Surprisingly two members were detected having Hepatitis C carrier. Four patients were positive for Hepatitis B.

Engerix B vaccine was given to all the members of society free of charge. This activity is continued for new member also.

Every month 40 thalassemics are given Blood transfusion free of charge by the society.

Dr. R.B. Shah, President, Dr. Alok Shrivastav, Hematologist, C.M.C.H., Vellore, Dr. Dilip Shah, Vice President, Mr. M.D. Golani, Secretary, on occasion of Lecture on "Bone Marrow Transplant" organised by TSCSA.

One meeting was held on 6th January, 96 to give detailed information on Bone Marrow Transplant. The meeting was addressed by Dr. Alok Shrivastav who was kind enough to explain in Hindi.

One of our member, Master Darshan Patel, has undergone B.M.T. & he is doing well.

थैलासीमिया में हैपेटाईटिस 'सी' का वितरण

— डा॰ वी॰ पी॰ चौधरी

एपलास्टिक एनीमिया, ल्यूकीमिया, थैलासीमिया व अन्य हीमोग्लाबिन संबंधी रोगों में विशेषकर थैलासीमिया में बार-बार रक्त संचारण करना पड़ता है। इन बच्चों में रक्त संचारण संबंधी रोगों से प्रभावित हहोने की अधिक संभावना रहती है। अनेक संक्रामित रोगों में से हैपेटाईटिस बी, सी, डी व HIV का प्रभाव दीर्घकालिक व घातक होता है।

हैपेटाईटिस विषाणु की खोज 1989 में हुई। इसे ए व बी अतिरिक्त हैपेटाईटिस फैलाने में मुख्य कारक माना जाता है। हैपेटाईटिस सी (HCV) का वितरण विनिमय कई विशेषज्ञों द्वारा थैलासीमिक बच्चों में आंका गया। इसका वितरण 3-62.2% आंका गया। दिल्ली के विभिन्न चिकित्सालयों में पिछले 2 वर्ष में देखने को मिला कि HCV से प्रभावित बच्चों की संख्या दिन प्रतिदिन बड़ रही है। मोलाना आज़ाद मैडीकल कालेज व संलग्न अस्पतालों में 550 रक्तदाताओं के परीक्षण में पाया गया कि उनमें से 11 (2%) में HCV पाया गया अत: यह निष्कर्ष निकलता है कि रक्तदाताओं में HCV का वितरण बड़ रहा है।

HCV की प्राकृतिक जीवन वृत्ति

डा॰ वॉन्की व उनके साथियों ने पाया कि 21 में से 10 HCV प्रभावित बच्चों में रोग का दीर्घकालिक प्रभाव व सिरहोसिस पाया गया। इसी प्रकार Dr. Resti व उनके साथियों ने भी 78 थैलासीमिक बच्चों में 13 वर्ष तक अध्ययन करने के बाद निष्कर्ष निकाला कि 55% में दीर्घकालिक ज़िगर रोग हो जाता है। विभिन्न संस्थानों के अध्ययन से परिणाम निकलता है कि HCV प्रभावित 50% बच्चों में ज़िगर के पुराने रोग (chronic hepatitis) व 10-15 प्रतिशत में सिरहोसिस व अंतत: HCV रोगक्रमण के 7-18 वर्ष पश्चात् ज़िगर का कैंसर हो जाता है। एक जापानी अध्ययन में 75% में HCV पाया गया। HCV संक्रमण की जीवन वृत्ति का पूरा ब्योरा अभी प्राप्त नहीं है।

रोग के दीर्घकालिक प्रभाव, सिरहोसिस अथवा ज़िगर के कैंसर होने में निम्न कारण मुख्य रूप से प्रभाव डालते हैं :

अ) संक्रमण के समय रोगी की आयु
ब) संक्रमण का समय अर्थात् संक्रमण कितनी देर तक रहता है
स) प्रथम संक्रमण के समय ज़िगर का नुकसान
द) बार-बार संक्रमण का खतरा।

हैपेटाईटिस बी की अपेक्षा हैपेटाईटिस सी में सिरहोसिस का खतरा कहीं अधिक है और दुर्भाग्य की बात यह है कि हैपेटाईटिस सी वैक्सीन अभी तक नहीं बनी है।

चिकित्सा

HCV संक्रमण में "Interferon" नामक दवा बहुत ही उपयुक्त साबित हुई है। 166 HCV रोगियों में Interferon alfa-2b के 30 लाख युनिट सप्ताह में तीन बार 6 माह तक देने पर 38% रोगियों में ज़िगर के बड़े हुये एंजाईम में कमी आंकी गई। HCV से संक्रामित थैलासीमिया बच्चों में Interferon देने पर 40-70 प्रतिशत में लाभ देखा गया परन्तु दुर्भाग्यवश 40-60 प्रतिशत में रोग का पुनरार्वतन भी देखने को मिला। यह दवा अमेरिका की खाद्य व दवा नियंत्रण प्राधिकरण द्वारा मान्य है परन्तु 6 माह का खर्चा लगभग 1 लाख रुपये आने के कारण अधिकतर परिवारों के बजट के बाहर है।

बचाव

HCV संक्रमण से बचाव का एक ही रास्ता है - रक्तदाताओं के रक्त में HCV का परीक्षण व संक्रमित रक्त का त्याग। HCV प्रसार का नियंत्रण अति आवश्यक है क्योंकि :

अ) 50% HCV संक्रमित रोगियों को सिरहोसिस हो जाता है,
ब) इलाज बहुत मंहगा है,
स) चिकित्सा के उपरांत भी 50% रोगियों में पुनरावर्तन पाया जाता है,
द) अभी तक HCV के लिये कोई वैक्सीन नहीं बनी।

पश्चिमी देशों में रक्त दाताओं में HCV का परीक्षण सरकार की ओर से आदेशात्मक है।

भारत सरकार पर भी इस बात का दबाव डाला जाना चाहिए कि भारत में भी सभी Blood Banks में HCV का परीक्षण अनिवार्य कर देना चाहिए।

Report from Thalassemia Welfare Society of Burdwan

Thalassemia Welfare Society, Burdwan organised a walk on 27th February, 96 in order to make people aware of the disease Thalassemia. Different schools along with Burdwan St. Xavier's School joined the walk. The Vice Chancellor of Burdwan University delivered a speech on Thalassemia.

At present we are getting ready for setting up our own Blood Bank for the benefit of the patients. We are in need of the machine Eliza Reader. A request is being made to philanthropist for this noble cause.

Kota Joins

The Thalassemia Society of Kota joins the 'Federation'. Thalassemics from Kota and adjoining areas may contact its:

President: Dr. P.P. Gupta
Prof. & Head
Department of Paediatrics
Jay Kay Lon Hospital
(Medical College),
Kota–324001
Phone: (O) 25358
(R) 27812

Secretary: Dr. A.L. Bairwa
24-B, Banker's Colony
Guman Pura, Kota–324007
Phone: (R) 26774

NATIONAL THALASSEMIA WELFARE SOCIETY (Regd.)
KG-1/97, Vikas Puri, New Delhi–110 018 Tel: 550 7483

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

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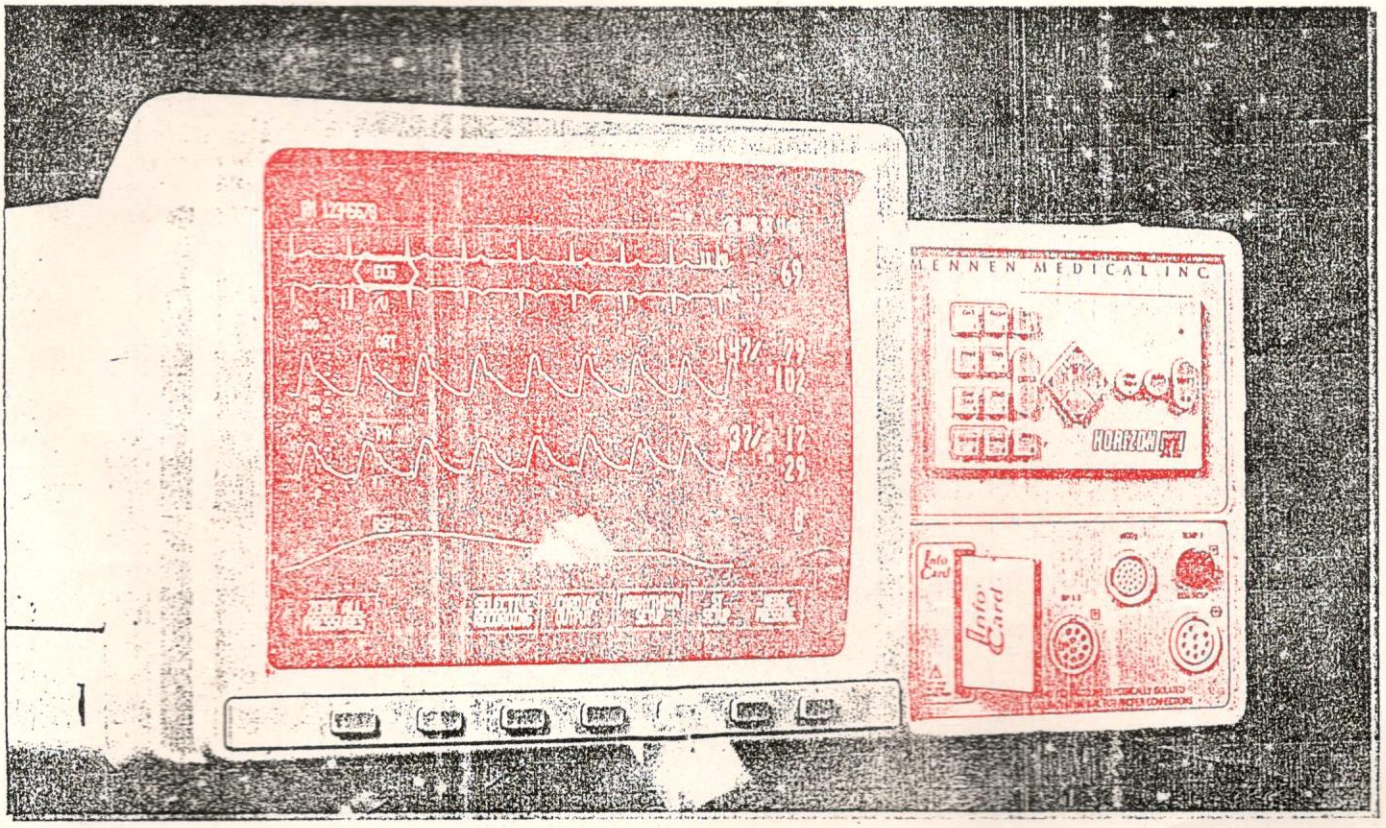

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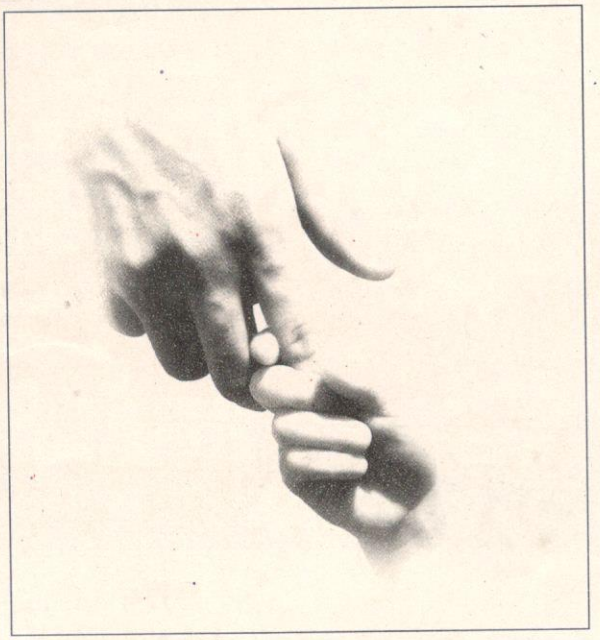

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