FEBRUARY 2010 ISSUE # 14

PTPP
Pertubuhan Thalassaemia Pulau Pinang

THALWATCH

BEYOND THALASSAEMIA

THE OFFICIAL NEWSLETTER OF THE THALASSAEMIA SOCIETY OF PENANG

INSIDE THIS ISSUE:

KEM THALASSAEMIA KE-13

Tarikh :17-18 October 2009

Tempat: Hotel Royal Penang

Tema: " The Adolescent Thalassaemic — Living Life Fully "

KEM THALASSAEMIA KE-13

13th Thalassaemia Camp

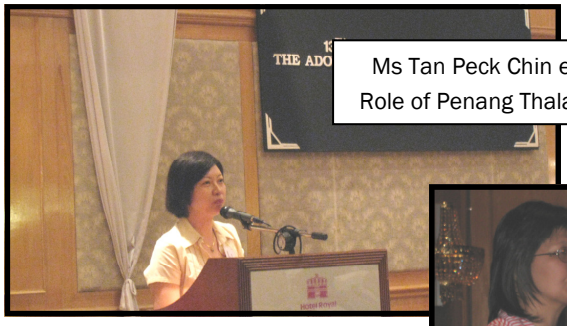
"The Adolescent Thalassaemic – Living Life Full"

Organized by :

Pulau Pinang Thalassaemia Society

Hari Pertama 17 October 2009 (Sabtu)

Morning Registration



Ms Tan Peck Chin explained on the Role of Penang Thalassaemia Society

Dr Goh Ai Sim updated on Thalassaemia Registry in Penang

OVERVIEW

- Burden of thalassaemia in Malaysia
- Distribution of Thalassemia patients in Penang according to ethnic group, age and gender
- Types of Thalassemia patients
- Blood transfusion
- Serum Ferritin
- Chelation therapy
- MRI T2*
- Other related services

Prof. Dr. Vip Viprakasit talked on Optimising Chelation Therapy

The 13th Camp had a total of 105 participants.

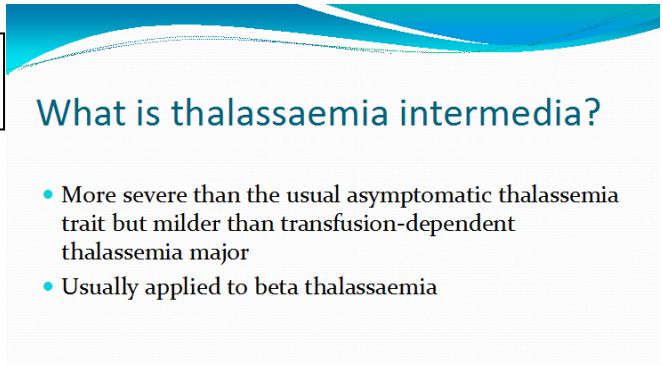
Outline:

- How to optimise iron chelation therapy
  - Update on iron monitoring
  - Choices of iron chelation therapy
    - DFO
    - DFP and Combination therapy
    - DFX
  - Key success factors for your chelation

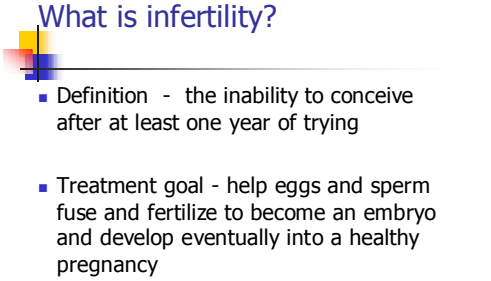
KEM THALASSAEMIA KE-13

Various Topics, Speakers and Presenters in the 2 days Camp

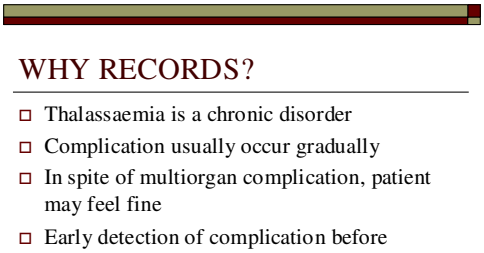
Dr. Angeline Yeoh shared on Thalassaemia Intermedia



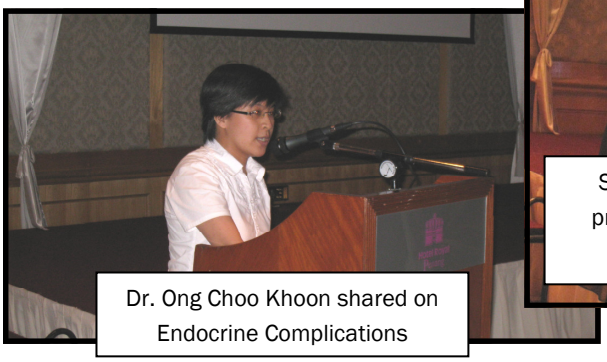
Dr. Narinder Singh Shadan discussed on Fertility Issues



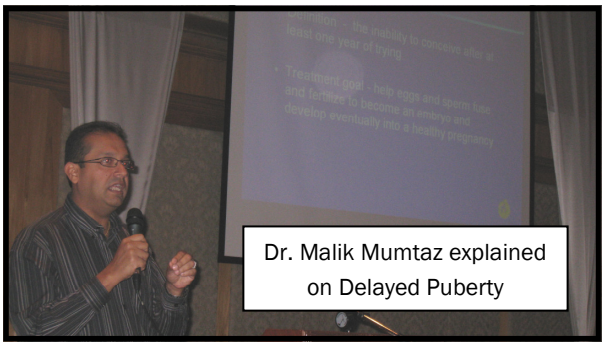
Dr. Dan Giap Liang shared on Growing Up with Thalassaemics



S/N Zailawati Mamat presented on 'Keeping Records'



Dr. Ong Choo Khoon shared on Endocrine Complications

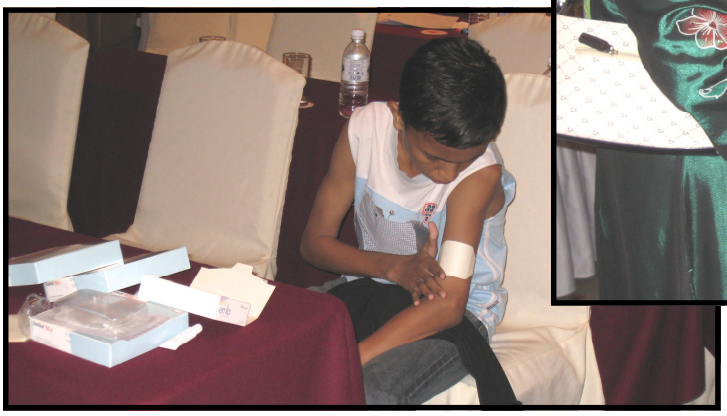


Dr. Malik Mumtaz explained on Delayed Puberty

KEM THALASSAEMIA KE-13

Desferal Session

My Pump is ***my Friend***



Total of 12 patients participating in the desferal session

KEM THALASSAEMIA KE-13

Aktiviti-Aktiviti Hari Kedua 18 October

Dr. Foong Wai Cheng's role-playing session with participants under 16 age group

Uncle Kasi hosted fun and games with children

4TH THALASSAEMIA WORKSHOP FOR DOCTORS, NURSES AND HEALTH PERSONNEL

TOPICS DISCUSSED

19th October 2009, ACC Auditorium, Hospital Pulau Pinang

> Challenges of thalassaemia management in developing countries

> Short statue and delayed puberty

> Management of infertility in thalassaemia

> The alpha thalassaemia syndromes

> Overview of laboratory diagnosis for hemoglobinapathy

> Molecular diagnosis of thalassaemia

RAYA OPEN HOUSE

Sambutan Hari Raya

12 October 2009, ACC Day Care Hematology Penang General Hospital

OUTDOOR ACTIVITES

The Race & Exercise For Fun

REPORT ON STARWALK 2009

By Charendee Soon

On 13th of December 2009, Sunday, my family and I entered Starwalk 2009 organised by Star Publications (Malaysia)Bhd and Penang Amateur Athletic Association. We checked in the reporting centre in SMK Raja Tun Uda around 7.10 in the morning and were given a yellow coloured wristband.

We started our walk at 7.15 a.m. from SMK Raja Tun Uda. There were thousands of participants entering this Starwalk 2009. Some of the participants took part in the Competition Category while others including me and my family participated in the Non-Competition Category. We were given 2 hours to finished the walk.

My brother and I leading the walk when we raced among us. I was inspired by Dr. Fong's father's spirit. His determination was beyond many other man whom were 30 years younger than him. He continued the race without any signs of giving up despite of his age even he is exhausted in the end of the walk. I reached the finishing point at 8.45 a.m and finished the walk in 1 hour and 15 minutes time. I was satisfied with my result as i am faster than my brother who was physically prepared.

Snacks and drinks are provided in penang International Sports Arena(PISA) for all the walkers to recover the energy lost during the walk. My family and I entered the main hall of PISA to watch the Variety Show. Prizes were given out during the show. There were performence performed by school students from Kuala Lumpur. There were even fancy dress competition for kids. There were also a dance group called the B-Boys to skow some of their dance moves which were krumping and street jazz. I enjoyed the slick moves of the dancers as I like street dance.

Second runner up, first runner up and championfor each group in the Competition Category from Men to Women were given a pewter medallion and cash up to RM250. After prizes were given to the winner, the Variety Show continued with its activities. Before the show ended the host asked all yhe audience of all ages to do some chicken dance. Little coorperation were given though. The programme is continued with Lucky Quiz Contest which was the lucky draw. From all the prizes provided for the Lucky Quiz Contest such as brand new handphones from Nokia N97 to iPhone 3GS, LG LCD TV to LG Home Theatre System and air conditioners only the 5 grand prizes stole the audience's attention which were brand new Modenas GT110 motorcycle.

All the audience wished to get their hands on those Modenas motorbike including myself but unfortunately when the host announced the winner of those motorbike, I am not one of them. After the winner claimed their prizes the Starwalk 2009 was officially ended. Some went back with joys in their eyes and with prizes in their hands but many went back with disapointment including me because I did not win anything from the lucky draw. Disapointed I am but proud was what I felt too because I proved to the world that *Thalasaemics Boleh*(Thalasaemics Can). It means that I can do anything I want to even with Thalasaemia if I really put my heart and effort into it.

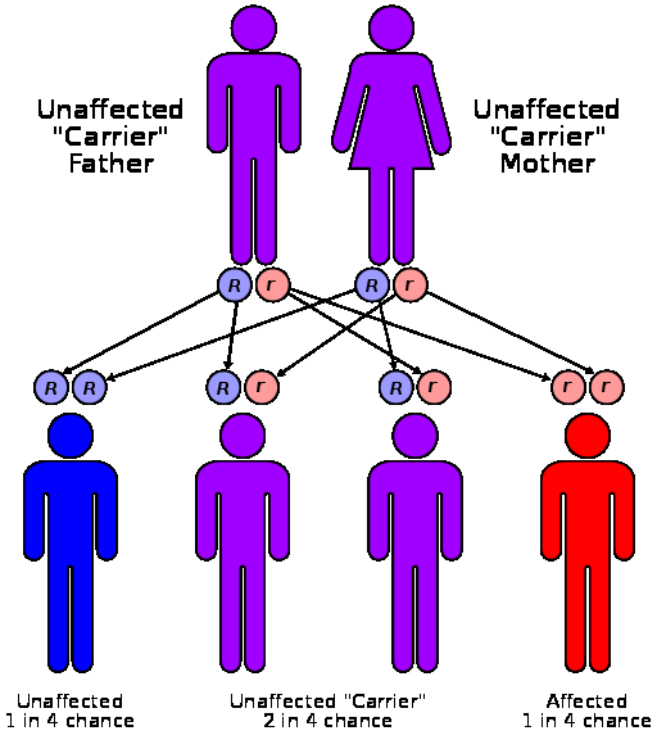
PROJECTS FOR YEAR 2010

14th Thalassaemia Camp

May 2010 – in conjunction with World Thalassaemia Day.

On going twice a year camp for thalassaemics and families, and their carers.

Aimed at providing psychological support in thalassaemia management for both affected families and carers. Topics depend on feedback on what needs to be addressed and appropriate speakers are brought in accordingly. This camp is to give an opportunity for socializing amongst participants to form the important camaderie much needed to support their life-long management.



15th Thalassaemia Camp

Sept/Oct 2010

More of an educational camp addressing difficult topics. Invited speakers participate both at this camp as well as Thalassaemia Workshop for Medical Personnels which follows the camp.

5th Thalassaemia Workshop for Doctors, Nurses and Health Personnels.

Sept/Oct 2010

This workshop is held at the General Hospital, with Paediatric and Hemotology Depts alternating in co-orgainising with the Thalassaemia Society. Targeted at doctors, nurses as well as laboratory personnels, and any other that is relevant in the management of thalassaemia.

1st Thalassaemia Workshop for Laboratory Personnels.

Aimed at creating awareness and training for laboratory personnels in the diagnosis of thalassaemias and most importantly, the traits.

1st Workshop in Basic Nursing Care for Thalassaemia

Specially structured for monitoring and nursing skills unique to the management of thalassaemia.

1st Counselling Workshop in Thalassaemia.

With screening now available to the public, properly trained personnels are needed to handle affected individuals. Counselling is also much needed for young adults with thalassaemia as they cope with social pressures.

MEDICAL ARTICLE

Using MRI to Measure Liver Iron Levels

Associate Professor Tim St. Pierre, School of Physics, The University of Western Australia

Introduction

Access to MRI methods for measuring heart and liver iron levels is becoming more widespread. MRI-based measurement techniques are yielding data that are proving very useful in clinical trials to access the efficacy of new iron chelation therapies in groups of patients. They are also increasingly being used to monitor individual patients in routine clinical practice.

Liver Iron Concentrations (LIC)

There are 4 key reasons why the precise and accurate measurement of iron levels in the liver is important in both the early detection and ongoing management of patients with thalassaemia:

1. LIC provide an accurate estimate of total body iron loading
2. Under normal circumstances, the liver is primary site of iron storage.
3. Patients with elevated LIC have been shown to be at greatest risk of future cardiac complications and premature death.
4. There is a longitudinal (but not crosssectional) relationship between liver and heart iron levels suggesting that a high LIC prospectively predicts cardiac iron loading and cardiac disease, and as such acts as an early warning signal of possible future cardiac complications.

Measuring LIC using MRI

Several MRI-based methods have been developed to measure LIC. The most widely accepted methods fall into 2 main categories: (i) relaxometry methods based on measuring R2 and (ii) relaxometry methods based on measuring T2*.

Conclusions

High liver iron concentrations are high risk factors for future cardiac disease in thalassaemia major patients, even in the absence of cardiac loading as measured by cardiac T2*. Monitoring of liver iron concentrations to aid in chelation dosing decisions requires techniques with sufficient accuracy and precision to measure clinically relevant changes in LIC.

Disclosure

Tim St. Pierre is a Board Member and Shareholder of Resonance Health, provider of the FerriScan® service.

The above excerpts are from UK Thalassaemia Society Magazine September 2009 Issue #115

(www.ukts.org)

Definitions & Terminology

MRI = magnetic resonance imaging: the use of nuclear magnetic resonance of protons to produce proton density images

Chelation = the process of removing a heavy metal from the bloodstream

Latest News from the Medical Literature

Complited by Dr Michael Angastiniotis

Magnetic Resonance Imaging of the Heart

Cardiac MRI (CMR), and in particular the T2* measurement, has now become part of routine monitoring of thalassaemia patients in most dedicated centres. The early detection of iron deposition in the heart is a major advantage for patients, since the reversal of even advanced heart complications has been convincingly demonstrated by intensifying iron chelation, either by continuous IV Deferoxamine or combining Deferoxamine with Deferiprone. In order to decide on the timing of intensification of treatment, it is important to know the risks associated with the various T2* values and their association with complications such as heart failure and arrhythmia.

Endocrinology

An important observation from Milan was published recently (Scacchi M et al, Enr J Endocrinol, 2009, Oct). They have found subtle impairment of adrenocortical function in some adult thalassaemia patients. This, they suggest, may become clinically important in case of major stressful event, such as a severe infection. We are accustomed to performing endocrinology monitoring during adolescence, having in mind mainly growth and sexual maturation, while in adults reproductive ability, diabetes and rare deficiencies are sought. Adrenocortical function is important, and in some stressful situations a deficiency may be dangerous. The authors recommend assessment of adrenocortical function in all adult patients.

Thromboembolic Complications

Well known in sickle cell diease, thromboembolic phenomena have only recently been emphasised in thalassaemia intermedia. One manifestation is stroke, and a common finding in sickle cell patients is asymptomatic brain infarction. Back in 1999, a group from Palermo (Manfre L et al, AJR, 1999, 173:1477-80) described changes on MRI imaging of the brain—ranging from mild atrophy to large infarcts—in 6 out of 10 thalassaemia intermedia patients who had presented no neurological symptoms or signs. Recently, Beirut has confirmed these observations (Taher A et al, J Thromb. Haemost, 2009, Oct). Of 30 patients with thalassaemia intermedia who had been splenectomised in the past, 18 (60%) had evidence of one or more white matter lesions on brain MRI. The age-range was 18-54 years. Brain MRI is recommended in this group of patients, but from what age and how frequently has yet to be defined.

The above excerpts are from Thalassaemia International Federation (TIF) Magazine November 2009 Issue #56

(www.thalassemia.org.cy)

Definitions & Terminology

Arrhythmia = an abnormal rate of muscle contractions in the heart

Endocrine = a gland which secretes hormones directly into the blood stream