APRIL 2016 ISSUE # 22

THALWATCH

BEYOND THALASSAEMIA

THE OFFICIAL NEWSLETTER OF THE THALASSAEMIA SOCIETY OF PENANG

Salam sejahtera kepada semua.

Suatu tinjauan tentang kehidupan seseorang talasemia yang memerlukan kemasukkan darah setiap satu atau 2 bulan di alam dewasa mereka telah dilakukan pada bulan Julai hingga Ogos 2015 (**Impact of Thalassaemia on Quality of Life and Employment among Adult Thalassaemia Patients in Penang (ITQLEAP).**

Secara ringkas, kebanyakkan mendapat pekerjaan yang memberi kepuasan kerja dan ingin di termia sebagai seseorang yang normal. Kadar pekerjaan (employment rate) mereka secara keseluruhan adalah lebih rendah dan ada kaitan sedikit sebanyak dengan tahap pendidikan. Namun begitu, mereka yang mencebur dalam tinjauan ini adalah amat kecil. Dengan itu, keputusan tinjauan ini tidak dapat memberi gambaran kehidupan seseorang talasemia dewasa yang 100% hakiki. Diharap tinjauan yang lebih mendalam and luas dapat dilakukan untuk memberi gambaran yang lebih utuh.

Tinjauan ini telah dibentangkan di Kem Thalassaemia 2015 dan akan dibentangkan pada PMC Research Day 2016.

Dilampirkan dibawah merupakan abstrak tinjauan ini dalam Bahasa Inggeris.

Sekiansahaja.

Dari
Dr WC Foong
(bagi pihak semua yang menjayakan tinjauan ini)

Impact of Thalassaemia on Quality of Life and Employment among Adult Thalassaemia Patients in Penang (ITQLEAP)

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BACKGROUND
Advances in the management of Thalassaemia, a chronic blood disorder, have resulted in longer life expectancy and a near normal life[(1,4)]. However, patients with thalassaemia were reported to face more adulthood challenges especially in the workforce[(2)]. Their unemployment risk was higher and many face discrimination[(2,3)]. The overall unemployment rate in Malaysia is 3.1%. This research aimed to explore their quality of life, adulthood challenges and employment status.

METHOD
All transfusion-dependent thalassaemia patients aged 18 years and above from Penang Hospital and Seberang Jaya Hospital were approached either in person or through phone calls between July and August 2015. A self-administered questionnaire selected from WHOQOL-BREF[(5)] and SF-36[(6)] questionnaires was given to those who consented. Results collected were analysed using SPSS version 46.

RESULTS
Of a total of 80 eligible patients, 62 consented. Half of the participants suffered from additional clinical complications and 29% were required to seek hospital care at least twice a month. The unemployment

rate among this group was 14.5% and three of them were still studying. There were no participants in working in the Protective and Healthcare sectors. 30% unemployed participants did not complete their secondary education level compared to 18% with an occupation (p = 0.69). Those unemployed were more likely to have more medical problems compared to their working counterparts (58% against 49%;p= 0.41). Mean score for quality of life, physical health, psychological health, social relationship and employment satisfaction were 63.79±2.05, 60.40±2.08, 61.94±1.95, 69.39±2.87 and 59.80±2.49 respectively. There was no significant difference between the scores for each domain with the employment status. Most of the comments made by the participants were about their ability to be a normal person with similar capabilities.

CONCLUSION

The quality of life in the adult thalassaemics is not related to their employment status.The employment rate is low and proportionate to the level of education. Presence of medical complications of thalassaemia contributes to risk of unemployment. The limitation of this study is its small sample size which may not reflect an exact picture of all thalassaemia patients. Therefore, this study calls for future studies which recruitof all adult working thalassaemia in the whole of Malaysia.

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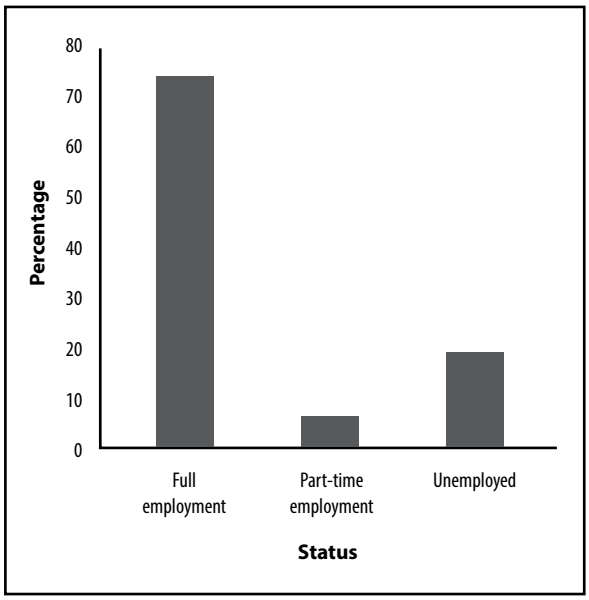
3. Pakbaz Z, et al. Education and Employment Status of Children and Adults with Thalasemia in North America. Pediatric Blood Cancer 2010; 55(4): 678-683
4. George E Beta-thalassemia major in Malaysia, an on-going public health problem. Med J Malaysia 2001; 60(1): 397-400.
5. The World Health Organization Quality of Life (WHOQOL) – BREF
6. SF-36c2 Healthy survey

APPENDIX:

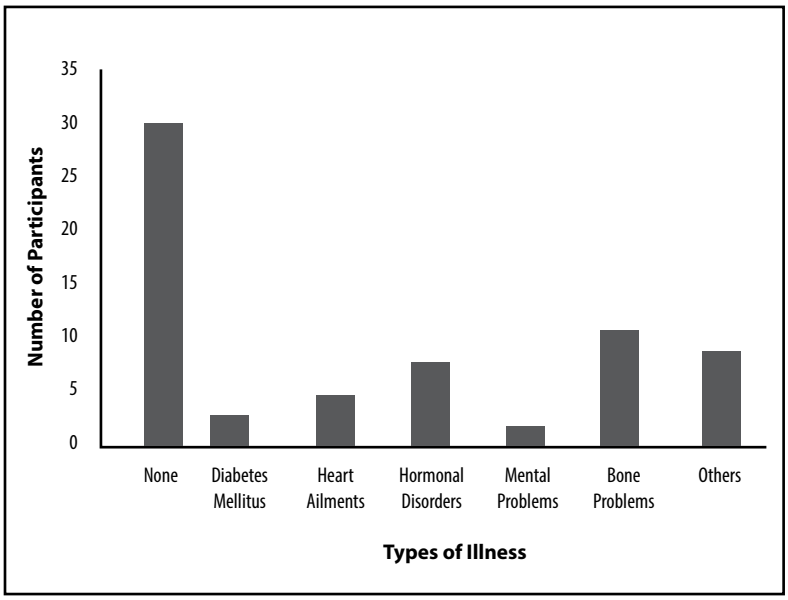
Participants comments:

- Terimalah pesakit thalassaemia seperti rakan-rakan anda yang lain
- Ambil tahu tentang talasemia, jangan hanya menilai berdasarkan apa yang dilihat, fahami keadaan kesihtan pesakit, *hanya dedah status kepada orang yang tertentu, *kadang kala letih bila dekat dengan tarikh temujanji
- Pesakit talasemia sama saja dengan orang lain, Cuma istimewanya, kami kena tambah darah
- Dengan harapan orang sekeliling menerima kami sebagai orang normal
- Saya ingin mereka tahu bahawa saya juga manusia biasa. Saya tiada masalah dengan keluarga, saudara dan kawan rapat. Cuma rakan yang baru ingin berkenalan tidak boleh terima keadaan ini. Saya juga ada perasaan untuk disayangi dan dihargai. Lantak la orang tak suka ka pap, janji kita bahagia.
- Jangan kata kami pesakit talasemia tidak boleh melakukan kerja atau aktiviti, sebenarnya kami mampu
- I am thalassemic, so what?
- Jangan bezakan pengidap talasemia dgn org yg normal kern kadang-kadang penghidap talasemia boleh memberikan prestasi yg sama malah lebih baik dr org normal
- Faham dengan keadaan diri saya dan sekadar mana kemampuan yang saya mampu lakukan. Kerana kesihatan kadang kala terjejas. Tidak mahu sentiasa dipaksa melakukan walaupun mereka tahu tahap kesihatan saya
- Memahami keadaan pesakit talasemia dan dpt bergaul dgn mereka dan tidak memandang rendah akan pesakit talassemia
- Saya ingin sihat spt org normal dan juga ingin mendapat sokongan drprakan-rakandan juga keluarga

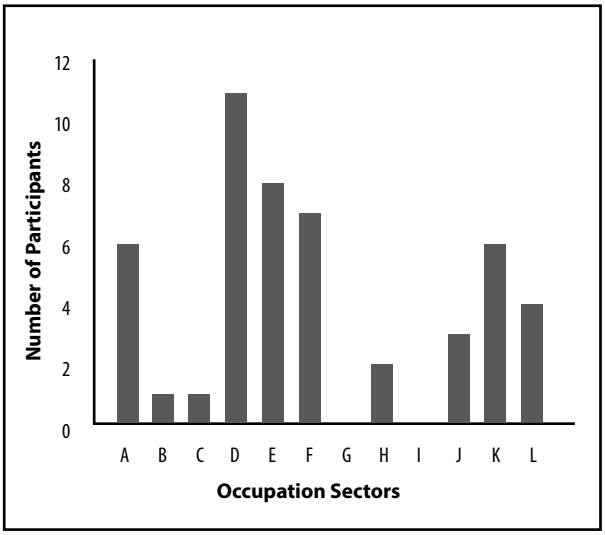
Employment Status



Presence of Other Medical Illness



Occupation



A: Business

B: Engineering / Architecture / Design

C: Farming / Fishing

D: Sales and marketing

E: Skilled craft and related trade
eg. Plumber, machine operator, seamstress

F: Food preparation and serving related

G: Protective service eg police, army, security guard

H: Education and training

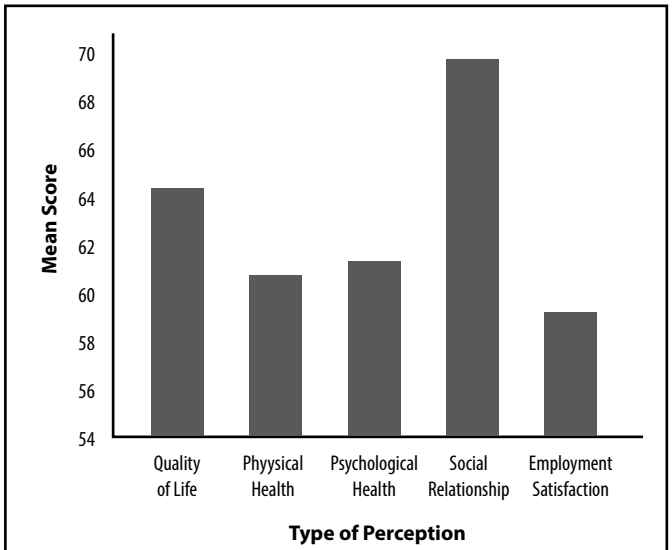
I: Healthcare practitioners and technical support

J: Elementary occupations eg cleaner, labourer, street vendors (excluding food)

K: Office and administrative support

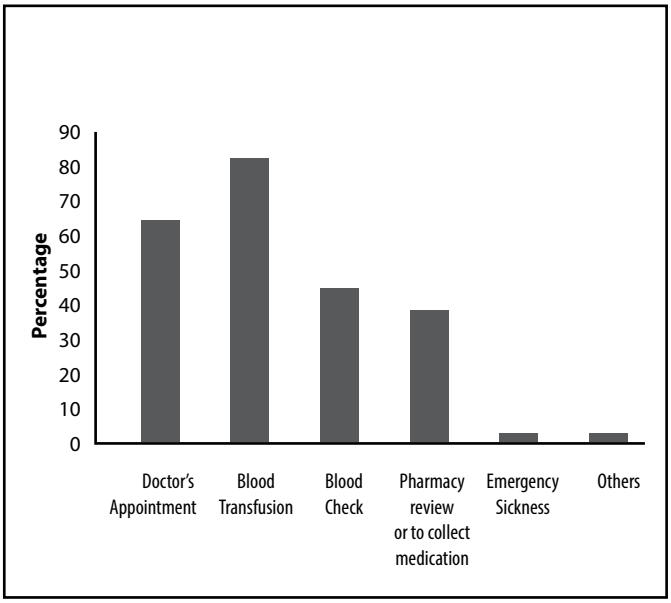
L: Others

Life Perception and Acceptance



Note (for this study): A mean score of 50 and below is taken as poor perception and acceptance in life. A mean score of 75 or more is considered excellent.

Reasons for Hospital Visits



BERITA TERKINI UNTUK RAWATAN TALASSEMIA

Ubat Luspatercept merupakan ubat kajian baru untuk merawat anemia yang berkaitan dengan penyaki ttalassemia dan akan menjalani kajian fasa ketiga di seluruh dunia.

Ubat ini meregulasikan pertumbuhan dan pematangan sel darah merah pada peringkat lewat. Ia diberi secara suntikan setiap 3 minggu.

Data awal menunjukkan bahawa ubat ini boleh meningkatkan takat haemoglobin dan mengurangkan keperluan darah di kalangan pesakit thalassemia. Dengan lebih spesifik, 65% pesakit NTDT mencapai peningkatan haemoglobin sekurang-kurangnya sebanyak 1g/dL manakala 47% bertambah sebanyak 1.5g/dL.

Untuk pesakit TDT yang menerima transfusi darah yang kerap, 79% dapat mengurangkan keperluan darah sebanyak 20% dan 57% pesakit diperhatikan mengurangkan keperluan darah sebanyak 50%.

Ini telah memberi kesan yang positif terhadap kualiti hidup dan juga mengurangkan takungan besi di dalam hati.

Kesansampingan ubat ini adalah minima dan kebanyakannya tidak serius seperti kesakitan pada tempat suntikan, sakit otot, sakit kepala, sakit tulang, rasa letih dan ruam.

Kajian secara rawak fasa ketiga ia itu “the BELIEVE trial” akan bermula di kalangan pesakit beta talassaemia major dan TDT bermulatahun 2016 dan Hospital Pulau Pinang adalah antara institusi yang dipilih untuk menjalankan kajian tersebut. Rawatan pengelatan besiakan diteruskan sewaktu menjalani rawatan luspatercept.

Daripada :

Dr. Goh Ai Sim
Consultant Haematologist
Hospital Pulau Pinang.

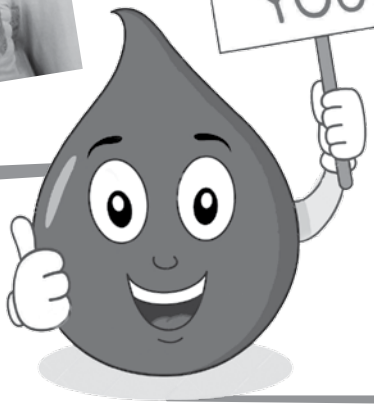
26TH AGM

10 ACC WORKSHOP

KL Public Awareness

Federal Meeting

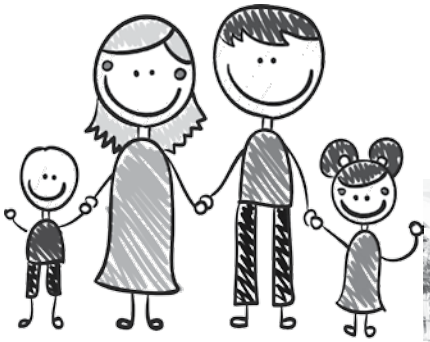
Activities



Blood Donation

Buka Puasa

Sabah Public Awareness



Hari Keluarga

Activities


Thalassaemia Pulau Pinang


LOLLIPOPS
STALL NO. 30
TICKET
MINERAL WATER
STALL NO. 30
TICKETING

Pertubuhan Thalassaemia Pulau Pinang

PESTA MAKANAN AMAL

HOME-COOKED, FRESHLY-BAKED, HALAL, VEGETARIAN, STREET FOOD, FRESHLY-BREWED

槟地中海贫血症协会
义卖会筹获 13 万元

（槟城 7 日讯）槟州地中海贫血症协会于周日举行义卖会，获得 68 个摊位参与，主要是筹募活动基金。该协会主席柯诗萍说，目前已筹获 13 万令吉活动经费，希望通过义卖会可筹获 15 万令吉的目标。

这场义卖会也邀请槟州福利、爱心社会及环境委员会主席彭文宝前来主持开幕礼。他致词时指出，槟州地中海贫血症协会与联邦地中海贫血症协会于 11 月份，首次在槟州举行地中海贫血症教育营，获得良好反应。他说，此类型活动应该多举行，以便提高人们对地中海贫血症的认识，也能让各单位互相分享经验及资源。#

➡ 诗萍和协会委员们主持剪彩仪式。彭文宝（左3起）、柯

Thalasseamia: Quick Facts

Parvinder Singh s/o Amar Singh (Ph.D)
Science Department
Institute of Teacher Education
Penang Campus
Malaysia

Thalassemia is a common public health problem in Malaysia and 1 out of every 20 Malaysians are carriers of this genetic disorder. The disorder results in excessive destruction of red blood cells, which leads to anemia. People with Thalassemia disease are not able to make enough hemoglobin, which causes severe anemia. Hemoglobin is found in red blood cells and carries oxygen to all parts of the body. When there is not enough hemoglobin in the red blood cells, oxygen cannot get to all parts of the body. Organs then become lack for oxygen and are unable to function properly.

There are two primary types of Thalassemia disease; Alpha Thalassemia disease and Beta Thalassemia disease as summarized in Figure 1

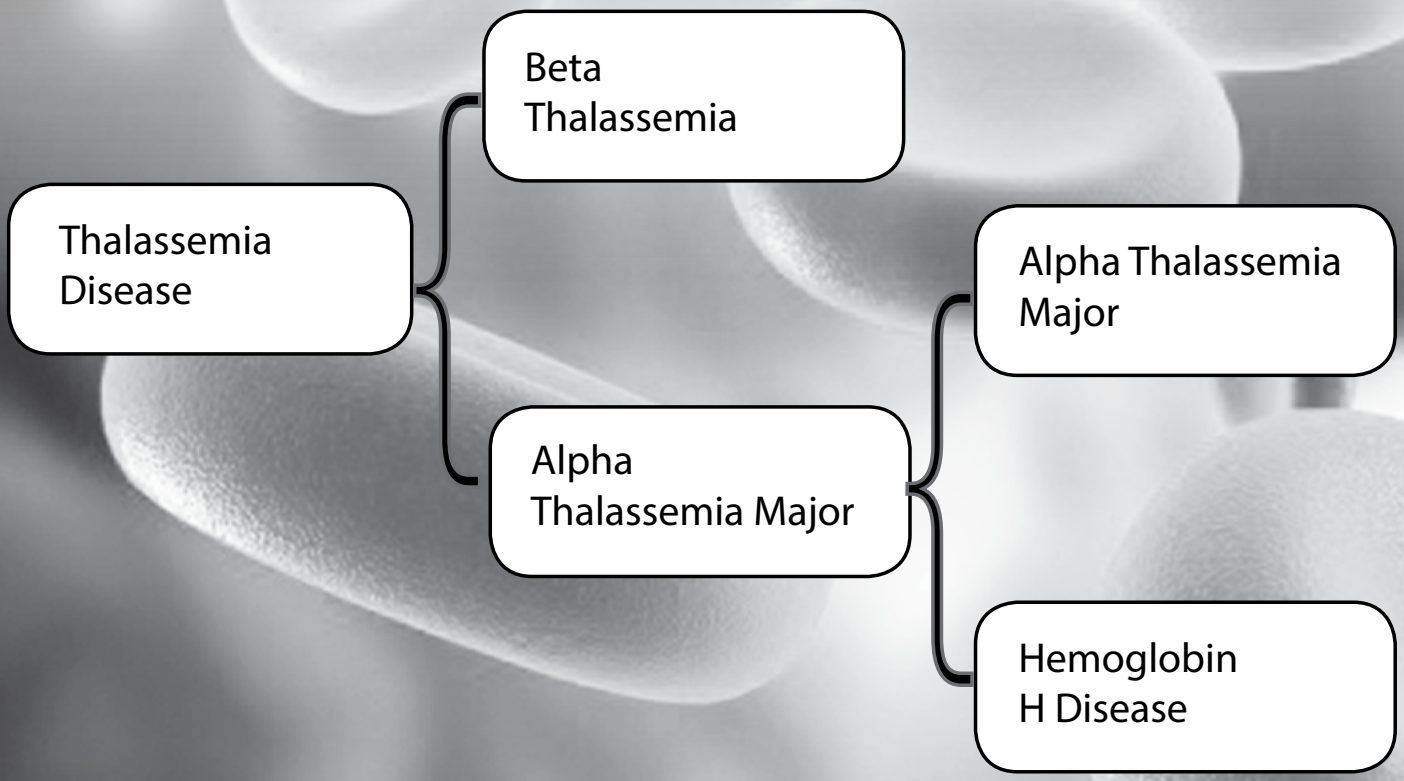


Figure 1 Thalassemia Disease

Beta Thalassemia Major is a serious illness. Symptoms appear in the first two years of life and include paleness of the skin, poor appetite, irritability, and failure to grow. Proper treatment includes routine blood transfusions and other therapies.

There are two main types of Alpha Thalassemia disease. Alpha Thalassemia Major is a very serious disease in which severe anemia begins even before birth. Pregnant women carrying affected fetuses are themselves at risk for serious pregnancy and delivery complications. Another type of Alpha Thalassemia is Hemoglobin H disease. Hemoglobin H disease can cause bone issues. The cheeks, forehead, and jaw may all overgrow. Additionally, hemoglobin H disease can cause: jaundice, which is a yellowing of the skin or the whites of the eyes, an extremely enlarged spleen and malnourishment

A wide variety of associated symptoms of Thalassemia commonly appear in the first two years of life. Symptoms may include; Skin with pale, jaundiced or anemic appearance Slow growth Poor appetite Fatigues easily. At risk for infections Thalassemia is a serious medical condition. If left untreated, it can result in medical complications that may lead to death. Complications may include: Enlarged liver, spleen, and heart,Thin and brittle bones

Thalassemia trait is not an illness and will never turn into one because it is not contagious. A trait carrier of thalassemia will always be a trait carrier. The genetic trait is passed down from parents to children. It is very much important for the public to to increase knowledge and cultivate positive attitudes toward prevention of Thalassaemia.

Activities

3rd National Thalassaemia Camp

3rd National Thalassaemia Camp

In conjunction with

21st Pulau Pinang Thalassaemia Camp

Theme:"Empowerment Through Information"
Date: 14 & 15 November 2015
Venue: Hotel Royal, Penang

Participants

Patients:	Adults - 63 children - 22
Parents:	38
Speakers:	Foreign - 4 , local - 6, society members - 9
Nurses:	20
Society members :	8
Volunteers:	13
Others:	53 (Kedah - 5, Perlis - 7, Johore - 2, Kelantan - 5, Selangor - 5, K.L. - 4, Perak - 13, Medan - 1, Sabah - 9 & Sarawak - 2)

Day 1 – 14 November 2015

Registration of participants started at 8.00 am. Being the 3rd National Thalassaemia Camp as well, there was a total of 236 participants from West and East Malaysia.

1.Welcome Address by Penang President – Ms. Janice Kua, in all aspects of thalassaemia and be participative & ask questions as all the experts are here at this camp for them.

She also announced that a fund raising food fair will be held on 6 12.15 at Komplex Penyayang, Penang and coupons are still available.

2. Hanoi Experience – Mr Doraisingam Rethinam – a parent

He was able to learn and refresh his knowledge, through the many questions posed by the many participants, some of whom were from rural areas but who were willing to learn.
Mr Dorai reminded participants to take advantage of opportunities to learn by attending camps and encouraged participants to be interactive and ask questions as a lot of money is spent to organize such activity.

3. Annual Camp – Does It Help? Cik Nur Ezzati Bt. Rosli

Izzati said all Thalassaemics are grateful and thankful to Mdm. Swee Hong who founded the Penang Thalassaemia Society in 1988.

She is a Thalassaemic diagnosed at age 1 year and is now 19 years of age.
Annual camps like this is certainly a help to all Thalassaemics as:

i) they will gain an abundance of knowledge and updates through invited expert speakers and doctors who share and impart their wide knowledge & experiences.
ii) Thalassaemics are encouraged & reminded to be consistent in their treatment regime to benefit from it.
iii) it helps to be constantly reminded they are as normal as any individual.
iv) when meeting new participants/parents they are able to give encouragement, help and advice
v) attendance at yearly camps allows them to meet up with each other and encourage each other to face challenges in their daily lives.

In conclusion she thanked the society for organizing yearly camps like this, the doctors, nurses and her parents for the encouragement & motivation given her each time she felt like giving up

4. Self-management and self-motivation

Sharing by:

i) ***Che Amal Mufidah*** *– age 15 years Paediatric patient & confirmed Beta-Thalassamia at 3 years old.*
She has been carrying out desferal chelation herself at home since aged 9 years. She was taught this since aged 6 years by her mother who then rewarded her with small gifts each time. Desferal chelation has not interrupted her studies as the pump is easy to carry with her. She takes her oral medication every morning. Presently her S. ferratin level is 1100. She thinks that if both her parents, the doctors & nurses have worked so hard to care for her health, she should now be responsible to look after her own health. Both her elder brother & sister, also Thalassaemics & successfully caring for their own health have been good examples for her to emulate. She is confident that any challenges can be overcome if the body is healthy and that her ambitions for her future will be realised.

ii) Cheah Hong Yi – *18 year old college student & a Beta-Thalassaemia Major*
He asked thalassaemics what is it that is stopping them from enjoying life. He says it is important to have a positive and "beautiful mindset" and upon "waking up to take a second to think about what a privilege it is to be alive and healthy." He further advised that whatever they want to be, they should not be afraid but to motivate themselves to find their own happiness as whatever they want is out there waiting for them. He says "Thalassaemia can be cured" and advised all thalassaemics to: "Wake up early. Drink coffee. Work hard. Be ambitious. Keep your priorities straight, your mind right, and your head up. Do well, live well, dress really well. Do what you love and love what you do. It is time to start living. However, motivation without perseverance is only temporary. So make it permanent."

iii) Puan Noorhafiza Binti Noorhamdan – *a working adult who was born in 1977.*
She received her education in Alor Star, Kedah and describes herself as a "devoted daughter,a loving sister, a loyal wife and a dedicated volunteer." Her journey as a thalassaemic began at 1 year and 8 months old when she was diagnosed with E-B. Thalassaemia. Her parents were shocked as no one in the family was ever diagnosed with such an illness.
Treatment available then was blood transfusion. Despite her illness, from 1955-2002 she worked at A&W, Guardian Pharmacy, Nandos and since December 2002 as a Government employee.

She gets her strength from:

- her believe in God
- the strong support she receives from her husband, family, friends and the Kedah Thal. Society.

She tells herself to always stay strong and confident, strive for the best, not to give up easily and believe that despite her illness she can contribute to society. 10 years ago she had no knowledge regarding E-BThalassaemia. She came from HKL with S.Feritine level of 10,800. Dr. Goh introduced her to the Penang Thalassaemia Society where she gained awareness & knowledge about thalassaemia and she used the knowledge gained to help her friends. This illness is not an obstacle, instead it drives her to do her best in whatever role she is assigned to. She is able to fulfil her duty as an employee in the civil service, take care of her own treatment and contribute to her society. She says as a patient, one should raise awareness regarding thalassaemia to the public and prove to them that thalassaemics are able to live normally. Society should also change its views about thalassaemia and give them the opportunity to show their capability in work, education and living normal lives. A patient must also fully cooperate with the doctors and nurses and contribute & exchange knowledge and updates among members. She says thalassaemia is not a burden as with proper treatment and medication thalassaemics are able to live life to the fullest.

iv) Puan Adura Helmi – *a parent who has 2 daughters, the eldest a thalassaemic and a normal younger child.*
In the beginning, she found it difficult to accept that her child has thalassaemia, until after having participated at camps like this where she met other thalassaemics who gave her the motivation and moral support she needed. Her responsibilities now are to:

a) see that her child carries out treatment and also keeps all appointments as prescribed by the doctor.
b) ensure that her daughter does not miss any classes except when she has to go to the hospital for her treatment.

She thanked all the doctors and nurses who are committed and dedicated in treating her daughter to enable her to fulfil her aspirations.

5. New Combination in Chelation Therapy – Dr. Farrukh Shah
Consultant Haematologist, Whittington Hospital, England

Dr. Farrukh reminded that iron chelation is important as iron overload has:
a) Fatal complications e.g.
   i) cardiac – dysrhythmia & heart failure
   ii) Infections
   iii) Liver – cirrhosis, viral hepatitis, failure
b) Non fatal complications e.g.
   i) growth failure
   ii) abnormalities in sexual development & fertility
   iii) diabetes
   iv) hypothyroidism
   v) hypoparathyroidism
   vi) osteoporosis

Chelators in clinical use are Desferrioxamine (DFO), Deferiprone (DFP) & Exjade(DFX)
Presently, multiple regimes is possible:
- Monotherapy using DFO, DFP & DFX
- Combination therapy DFO / DFP
  DFP / DFX
  DFX / DFP

Evidence for combination being:
Combined DFO + DFP chelation therapy is effective in reducing myocardial iron,
DFP + DFO reduces severe myocardial IOL and improvement in IOL with DFX + DFO, & improved cardiac T2* with DFX + DFO.
She went on to discuss complications and management of the side effects of the chelators in use.

She concluded with this message, that:
- iron overload is a serious cause for both morbidity & mortality
- chelation therapy is needed to safely manage transfusional iron overload
- side effects from all these chelation agents are manageable
- well managed patients have a very good prognosis

6. Understanding Complications of Transfusions – Dr Anizah Bt .Arshad,
Transfusion Medical Specialist,
Hospital Pulau Pinang

Dr. Anizah explained the aim of blood transfusion therapy in thalassaemia is to suppress extramedullary haemopoiesis while minimising complications of under transfusion and maintaining normal well being. A blood transfusion can sometimes cause reactions which are rarely life threatening. Common symptoms are fever, chill and rashes which can be treated with medications. However, patients are carefully monitored while they are being transfused. She went on to talk about prevention & management of Acute and Delayed Transfusion Reactions-Immunogenic and non immunogenic, also complications due to bacterial contamination.

Pn Noorasyikin Md Saad- patient
Pn Noorasyikin spoke on the causes, symptoms, prevention & management of the side-effects and adverse reactions during and after transfusions.

7. Optimising Bone Health in Thalassaemia – Dr Farrukh Shah

Dr. Farrukh explained causes for bone problem is multifactorial, difficult to manage and debilitating.
As It is unclear what can provide long lasting benefits, prevention is the best option.
Common bone problems are :
a) osteoporosis due to problems with endocrine, anaemia and desferrioxamine
b) osteomalacia which occurs because of vitamin D deficiency, lactose intolerance.
c) degenerative changes as one gets older

She went on to talk about their prevention and advised thalassaemics to do their chelation well to stop iron from damaging the bones & hormone making glands, keep the Hb level high, exercise regularly, eat a healthy diet rich in calcium and avoid smoking & alcohol.

A healthy diet, vitamin D supplements & sunlight will prevent osteomalacia

8. TDT and NTDT – A Revision – Dr Wong Sing Ai, Physician, Hospital Kepala Batas, Penang

Transfusion Dependent Thalassaemia (TDT) refers to patients who require regular lifelong transfusion for survival. Non-transfusion Dependent Thalassaemia (NTDT) are a group of patients who do not require regular RBC transfusion for survival. However some may require occasional /frequent transfusions in certain circumstances. It is important to differentiate between NTDT and TDT to prevent placing patients on lifelong transfusion therapy unnecessarily. NTDT patients should be monitored closely to avoid development of complications. Blood transfusions should be individually tailored to meet NTDT patients' needs. Many NTDT patients may develop iron overload and require treatment with an iron chelator even in the absence of transfusion therapy. Early recognition, diagnosis and appropriate management are essential to prevent development of complications and improve outcomes.

9. When & Why I Need Splenectomy – Prof. Suthat Fucharoen, Consultant Paediatric Haemotologist, Siriraj Hospital, Bangkok, Thailand

Prof. Suthat started by revising the anatomy of the spleen and its functions.
Hypersplenism refers to ill effects resulting from increased splenic functions which may be improved by splenectomy.
Some indications for splenectomy:

TDT patients
- annual blood requirement exceed 1.5 times of those splenectomised
- splenic enlargement accompanied by symptoms like pain & feeling of satiety
- massive splenomegaly causing concern of possible splenic rupture

NTDT patients
- worsening anaemia leading to poor growth & development
- transfusion therapy is not possible
- chelation therapy is unavailable
- hypersplenism with worsening anaemia, leucopenia or thrombocytopenia resulting in recurrent bacterial infections
- splenomegaly with symptoms of pain and early satiety
- massive splenomegaly causing concern of possible splenic rupture

Removal of the spleen causes lower and incomplete adaptive immune response against bacteria and the patient becomes susceptible to infections.
Prof's. recommendations are:
- advice patients to avoid food /water that may be contaminated
- give prophylactic immunization against common organisms
- patients (especially splenectomised ones) with fever/other signs of bacterial infections be considered as emergency cases
- stop DFO chelation
- broad spectrum antibiotics be started to cover both gram negative and gram positive organisms
- provide adequate supportive care e.g. bring down body temperature, adequate hydration & blood transfusion to keep safe level of haemoglobin.

10. What to do when having fever – Dr. Dan Giap Liang, Consultant Paediatrician and Paediatric Cardiologist, Island Hospital

Fever < 37.5 C considered low fever, but still needs to be alert. A proper thermometer should be used to check temperature.
Fever is when:
- temperature > 38.5 C in a child
- temperature > 39 C in an adult
- rigor

may be life threatening, seek treatment immediately

In Thalassaemics:
- when s.ferritine high
- diabetics
- splenectomised patients
- anaemia

What causes fever ? Infections of the:
- throat
- skin
- stomach
- brains
- ears
- chest/lungs
- urinary tract
- blood

Vital signs to check
- Blood pressure & pulse
- Blood circulation (capillary refill)
- Respiration

Investigations
- Blood/urine
- X'ray
- Spinal fluid

Treatment
- i/v infusion
- high doses of antibiotics
- further investigations and close monitoring

Dr. Goh added that adult patients seeking treatment late when they have infections e.g.fever is still a problem. She also stressed that treatment must be sought from Doctors who are well versed in Thalassaemia as against those who are not.

11. Impact of Thalassaemia on Quality of Life & employment among adult thalassaemic patients In Penang – presented by Dr Foong Wai Cheng
Lecturer & Consultant Paediatrician, Penang Medcal College

A survey was carried out for the above purpose in June 2015 – September 2015.
Objectives of the survey was to:
i) measure quality of life (QOL) among adult transfusion dependent thalassaemia (TDT) patients.
ii) explore the impact of thalassaemia in general on employment among TDT adult patients.

Number who consented to participate was **67.** (Penang & Seberang Jaya Hospitals)
Their mean age: was 29.03 years.

Gender		Race		Marital status	
Males	32	Malays	51	Married	21
Females	35	Chinese	16	Single	44
				Divorced	2

Education level

No formal education	Primary	Lower secodary	Higher secondary	Tertiary
3	2	7	37	18

Employment status

Unemployed	Employed part time	Employed full time	Self employed
15	4	39	9

Monthly Household Income

No income	< RM 2000	RM2000-RM5000	RM5000
13	40	13	0

Occupation: Participants are involved in sales/ marketing, skills craft, food industry, administration work, business, studying/training, architect, engineering and others e.g. protective service, healthcare, farming, fishing, agriculture.

The questionnaire required answers for routine hospital visits, other health problems, self- perceived health status, unwell days, QOL, social, psychological & physical health, job satisfaction & conducive work environment.

Results were:
1. Quality of life: fair with most days feeling well mentally and physically.
2. Majority has worked before and are still working. They are employed full time and are fairly satisfied with their jobs. They find their working environment fairly conducive.
3. Their frequent visits to the hospital did not affect the conduciveness of the working environment.
4. Physical, psychological & social health is generally fair, in range of 50-74 regardless if they are self employed, employed full time or part time or unemployed.

Therefore the disease does not cause any problems jn employment.
Sample size is small (Penang only). For the future can consider:
- Whole of Malaysia
- Questions prepared to obtain qualitative data
- Comparison with controls e.g. caregivers, siblings, cousins

At the end of the presentation, Prof. Suthat suggested:
- a larger sample size
- to include more parameters e.g. QOL related to good income of parents
- insurance coverage – whether thalassaemics will be able to get insurance coverage.

12. Chelation sessions

Participants grouped themselves and held discussions and were advised on solutions to their problems e.g. side effects. Proper & correct chelation was stressed for their well being.

1. S/N Zailawati i/c Desferal chelation with: 6 adult patients & 3 paediatric patients present.
2. Dr. Goh Ai Sim who took charge of the 21 patients on L1.
3. Dr. Foong Wai Cheng with 12 patients on Exjade

Day 2 – 15 November 2015

After breakfast children < 12 years of age reported to the volunteers to be taken around sight-seeing on the "Hop on Hop off" bus, while the rest continued with the planned programme

1. Overview of Thalassaemia in Malaysia – Dr Hishamshah Mohd. Ibrahim
Consultant Paediatrician, Senior Consultant Paediatrician
Haemotologist & Oncologist, Hospital Kuala Lumpur

Before the start of the National Thalassaemia Program

- Management of patients was sub optimal resulting in:
  - High morbidity & mortality
  - Premature deaths due to complications of iron overlad
  - Multiple end-organ complications
- Medical personnel were inadequately trained
- There was poor public awareness of the problem
- There were no population screening & preventive measures
- Epidemiological & outcomes data was in adequate, so
- Unable to plan & monitor performances

The National Thalassaemia Prevention & Control Program was introduced in 2005, where emphasis of the strategy & policy was:

- Public awareness & health education — Preventive
- Population screening & laboratory diagnosis — component
- Aim: for all to know their status & be well informed about it
- Management and treatment of patients — Curative
- Thalassaemia Registry — component

To give best possible care + retrospective genetic counselling + pre-marital counselling + voluntary population based screening +family screening + occasional prenatal diagnosis (PND) + abortion

Dr Hishamshah also revealed that Thalassaemia in Malaysia (June 2014)

Diagnosis	%
Beta thalassaemia major	40.2
HbE-_Beta thalassaemia	32.7
Beta thalassaemia intermedia	9.8
HbH Disease	11.3
Others	6

Prevention program is still not very successful due to religious and cultural Issues so thalassaemic babies are still being born. The challenge to reduce the birth rate of Thalassaemic babies remains.

<u>Perception of parents towards PND & termination of pregnancy</u>
Refuse PND - 29%
Agreed for PND and abortion - 28%
Agreed for PND but refuse abortion - 43% (70%-80% Muslims)

<u>Perception of Health Care Workers to counsel about PND</u>
42% are not agreeable to discuss about termination of pregnancy fpr Thalassaemia.
Some of the reasons cited are condition is not serious enough, abortion is not legal, abortion is not allowed by religion & there is still lack of knowledge in this area

<u>Practice of Parents – no Family Planning in Thalassaemia families</u>
2 siblings withTDT - 71.4%
3 siblings with TDT - 14.3%
4 siblings with TDT - 14.3%

In September 2005 – September 2006, there was an attempt to screen for thalassaemia carriers among 16 year old adolescents in N.E. District, Penang. – an area of 119 sq.km. & population of about 483,000 served by 1 Government & 9 private hospitals, 37 secondary schools with a Form 4 (16 yrs.old) population of 7281 students.
There was difficulty in getting consent for screening from PARENTS.
Only 59% of parents consented to screening and 86.7 % of those screened had normal result.

Present concerns

- Ineffectiveness of prevention program
  - inadequate screening of target groups
  - inadequate PDN facilities
  - awareness is superficial – not the desired practice
- Attitude of Malaysians to preventive strategies
  - No f/planning in families with thalassaemia
  - PND & Termination of Pregnancy (TOP) - ? unacceptable options
  - Screening - no consent & fear of stigmatization
- Attitude & practice of health care professionals
  - Non-directed counselling
  - Unwilling to discuss preventive strategies & TOP
  - Unaware of preventive strategies

Future strategy direction : **PREVENTION –** applying principle of effective genetic screening :

i) which will be made accessible to all target population
ii) to offer wide range of preventive options
iii) for long term feasibility & sustainability
iv) in line with the nation's policy & agenda for:
   - compulsory screening of 16 year old adolescents
   - reduced carrier couple
   - reduction of new thalassaemia

There was a very lively discussion at the end of the talk – whether above preventive strategies will be possible as Muslim law & its enforcement is different in each state etc.
The school will be a good institution to promote awareness for early prevention, but thalassaemics themselves should not hide their problem. Instead there should be representation by the whole Thalassaemia fraternity to speak in "One Voice" - that thalassaemics can lead normal lives

2. Progress of GeneTherapy – Prof. Suthat who introduced the topic by talking about:

Treatment of Thalassaemia

1. Conventional treatment
   - Blood transfusion
   - Iron chelation
2. Haemoglobin F Stimulation
3. Treatment of complication
   - Infections
   - Heart failure etc.
4. Cure
   - Bone marrow and stem cells transplantation
   - Gene Therapy (correcting abnormal genes)

He informed that:

- a study undertaken has demonstrated continued promise of gene therapy in B thalassaemia major regardless of genotype
- LentoGlobin BB305 Drug product results in sufficient early haemoglobin production to require minimal or no transfusion support for thalassaemia patients
- These data demonstrates the feasibility of a multi-centre international collaboration with centralized stem cell transduction
- The safety profile is consistent

3.T2* Results – Danger Points – A/Prof. Tan Ru San,
Consultant Cardiologist, National Heart Centre, Singapore

Prof. Tan revealed that according to the survival cohort of birth, those born after 1980's, mortality due to iron overload was much improved. It was reported in UK that iron overload deaths took a sudden dip with the introduction of T2* to measure heart iron.in 2000 – 2003.

Why the need to measure Heart iron

Heart failure from heart iron overload is the commonest cause of death in transfusion dependent thalassaemics. Echocardiogram, serum ferritin & liver iron measurements cannot reliably predict heart iron overload. MRT2* scan can detect heart iron overload early, so that intensive treatment can be started and also be used to monitor improvement during treatment.

He concluded by saying that low heart T2* is associated with an increased frequency of cardiac complications. Intensive chelation in iron overloaded patients with heart failure results in an improvement in heart T2* as well as heart pump function. Heart iron overload is reversible. Conventional methods e.g. liver iron, ferritin & echocardiographic abnormalities are inadequate. Heart iron can be assessed accurately and non-invasively the MR T2* technique e.g.in monitoring guiding treatment & comparing treatment strategies. Early detection and treatment should save lives.

4. Fertility & Reproduction in TDT – Dr Amma Kyei-Mensah,
Consultant Obstetrician & Gynaecologist, Whittington Hospital, England

Dr. Amma says that with advances in Thalassaemia , successful pregnancies are occurring more often. If the body is in good shape, pregnancy.will be successful. Iron chelation ideally from infancy is the key, therefore full compliance to treatment & advice is needed to reduce iron damage to important organs & glands that produces hormones e.g. pituitary glands, therefore no periods, pancreas – diabetes, underactive thyroid etc.

If treatment is vigorously maintained, thalassaemics may not need fertility treatment, they may even get pregnant spontaneously, and also there will not be any complications. Therefore it is in the hands of Thalassaemic patients themselves to have normal deliveries and healthy babies. Fertility treatment if required is in the form of hormonal injections to stimulate egg release in women and to stimulate sperm production in males but which may take a longer time to respond.

5. Challenges with Paediatric Thalassaemics – Dr Shoba Anne Thomas,
Paediatric Haemotologist & Oncologist,
Hospital Pulau Pinang

Paediatric challenges are diverse as in the different age groups physical, physiological & psychological demands differ.

Infants & Toddlers are dependent on parents who may be in shock and grief.
They feel the pain & discomfort of treatment therefore it is crucial that parents accept the situation as early as possible to build a strong relationship with the child to help him through the treatment & to build a trusting relationship with health care workers.

The school-going child who will be aware that he is different – in looks, having to miss classes due to treatment etc. Parents & caregivers play an important role to explain the disease & encourage the child to feel confident. However they should be cautious and not to overprotect the child.

Adolescents are the most difficult group to deal with. They may be rebellious and refuse to comply with treatment especially in chelation therapy. So they need to know that they have the power to control the quality of their lives.

6. Challenges with adult thalassaemia- Dr Goh Ai Sim, Consultant
Haemotologist, Hospital Pulau Pinang

Chelation Therapy has ben funded by the Government since 2006, but challenges with adult thalassaemia still exist e.g.

a) treatment related
- side effects of drugs, working hours
  transfusion problems etc.

b) patient related
- compliance problem which can also be deliberate
- communication with health staff
- lack of knowledge
- frequent absentism from work
- transfer to other states/places for higher education
- getting employment
- getting insurance coverage

c) hospital related
- budget, resources
- space (daycare, ward transfusion)
- manpower (trained doctors & nurses)

d) policy (night/weekend transfusion)

Dr Goh made known the cost for treatment: -

Blood filter each	RM 70
RBC 1 unit	RM 200
Blood investigations	RM 500 every 6 months
Pump each	RM 2,500
Thalaset each	RM 10
MRI T2*	RM 1200

Cost of Drugs for chelation :	Deferiprone 9 tablets/day	RM 29
	Deferiprone syrup 45 ml/day	RM 54
	Desferrioxamine 2g/day	RM 43
	Deferiserox (exjade) 1.5 g/day	RM 274

Dr Goh concluded by informing that the estimated cost to treat a thalassaemia major till age 30 years is RM 3 million.

In spite of all these challenges, she quoted Mark Twain “Challenges are what make life interesting; overcoming them is what makes life meaningful.”

7. Accepting who I am – sharing by Nur Edzianni Rosli - a B thalassaemia Major patient

Nur Edzianni, born in 1991 had been diagnosed since age one. She showed pictures of herself from infancy till adulthood and apart from those showing her undergoing treatment, the rest were that of a young girl enjoying life to the fullest attending camps sponsored by PTPP. With support & encouragement from her parents and healthcare workers, and the knowledge obtained from such activities, she has kept herself healthy and went through college and is now gainfully employed at Kolej Pergigian Pulau Pinang. She has not hidden her problem from anyone and she is thankful her boss is understanding of her periods of absentism for treatment.

8. Importance of record keeping – S/N Elliyana Bt. Tajidin,
Paediatric Daycare, Penang Hospital
Munir Bin Md Noor
a HbH adanal Constant Spring patient

S/N Elliyana informed that good & proper record keeping is essential for
- comprehensive management
- optimisation of treatment
- early detection of complications before irreversible damage to organs

Records are also filed according to types for easy accessibility.

Records kept are
a) Thalassaemia protocol
b) Patient summary, family tree
c) DNA Hb analysis
d) Immunization, growth chart
e) Investigation & Blood transfusion flow chart

All records are important for monitoring for:-
- iron overload
- growth & development
- drug toxicity
- infection

Munir Bin Md. Noor,, born in 1976, from a family of 3 siblings has been diagnosed since age 9 years & has been transfusion dependent since then. He fully understands the aims as described by S/N Elliyana and importance of these records as reference for his caregivers to monitor his condition, also for him to keep his appointments on time. He has been credited as one who has been keeping records well.

9. Round table discussion on compliance to chelation

Participants were divided into 6 groups. From their feedback, they all understood the aims of chelation & its importance.

Common side effects expressed by all groups were - Mouth ulcers, edema, rashes, fever, nausea, gastric pain, vomiting, diarrhoea, sleepiness. Suggestions & tips to overcome/manage above effects were discussed and shared.

Having heard promising information in the treatment of thalassaemia, the take home message to all thalassaemics throughout was to take good care of themselves, maintain complete compliance to treatment & investigations as there is definitely hope for a better tomorrow

The camp was brought to a closure with the children of the society showing off their talent in the various activities and performances.

Reported By Ms Wong Ah Soo
10.12.2015

Taniah, Nur Ezdianni & Redzuan

Assalamualaikum dan Salam Sejahtera. Saya dilahirkan pada 06.06.1991 di Aman Specialist. Pada usia dalam lingkungan empat belas bulan, mama mendapati keadaan saya pucat, kerap demam dan suka menangis pada waktu malam. Mama membawa saya ke klinik untuk mendapatkan rawatan. Ahkhirnya, saya dibawa ke GH dan mendapati HB saya sangat rendah. Selepas itu, saya telah disahkan menghidap penyakit Thalasemia Major. Bukan mudah untuk keluarga saya menerima berita ini. Tetapi keluarga saya sangat kuat dan sabar dengan dugaan yang menimpa.

Saya masih lagi teringat, setiap malam mama dan abah membawa saya ke rumah Madam Khoo untuk membantu saya menyuntik desferal. Setiap kali mahu suntik desferal, saya akan lari satu rumah dan menjerit kerana takut dengan jarum. Mama sangat tegas dalam pemakanan saya. Budak seusia saya tidak diberi makan coklat, tidak dapat minum air milo dan makanan yang mempunyai iron. Mama akan sentiasa memantau apa yang saya makan. Setiap bulan, tempat yang istimewa bagi saya ialah Wad 004 di mana saya akan menjalani tranfusi darah. Masih teringat kami semua akan beratur depan Aunty Saw untuk tunggu giliran cucuk. Selepas pukul satu, kami semua kena bawa darah ke wad.

Saya suka menghadiri kem, kerana di situ saya akan mendapat ilmu-ilmu yang penting dan dapat berjumpa dengan rakan-rakan. Alhamdulilah, pada 20.02.2016 saya telah berkahwin dan akan berpindah ke Johor.

Saya bersyukur Allah masih memberikan saya kesihatan yang baik dan saya ingin mengucapkan jutaan terima kasih kepada keluarga saya yang menjaga saya dari kecil, member saya kata-kata semangat dan selalu bersama saya. Terima kasih yang tidak terhingga kepada Pertubuhan yang saya sayangi kerana selalu ada untuk pesakit Thalasemia. Saya akan merindui kamu semua. Doakan saya ya di tempat baru.

Ezdianni

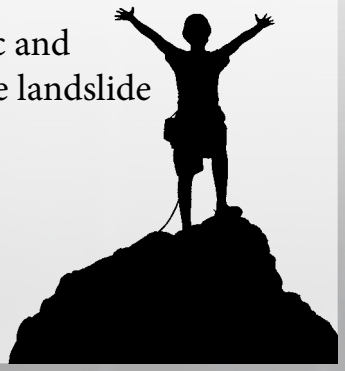
ANNOUNCEMENT 2016

A HISTORIC CLIMB - Penang Thalassaemia Kinabalu Challenge

30 August - 2 September 2015

Dear all,
We, the Penang Thalassaemia Society is indeed sad to inform the public and members that the climb was cancelled by the Sabah authority due to the landslide on June 06 2015.

Thank you.
Yours truly,
PTPP Managing Committee



AGM Ke27
Perhimpunan Tahunan

Tarikh : AHAD 22/5/2016
Masa : 8.30 am - 12.30 pm
Tempat : Orkid Room,
Kompleks Masyarakat Penyayang
Jalan Utama, 10450 Penang.

Makanan ringgan akan disediakan • Sebuah bas akan disediakan bagi ahli Seberang Jaya - maximum 40 orang saja • Kehadiran anda sangat dihargai.

22nd Camp

Date :24-25/9/2016
Masa :8.30 am - 5.30 pm
Tempat: Hotel di Pulau Pinang

Sila hubungi pejabat untuk keterangan lanjut.

T2* Workshop for health personnel @ Seberang Jaya Hospital

Date : Tuesday, 27/9/2016
Time : 8.30 am - 2.00 pm

11th ACC Workshop for Health personnel @ Penang General Hospital

Date : Monday , 26/9/2016
Time : 8am - 5 pm
Venue : ACC building, 4th.Flr,
Hospital Pulau Pinang.

Announcement 2016

World Thalassaemia Day 2016

Public Awareness @ Penang General Hospital Foyer

Date : Friday 6/5/2016
Time : 8am - 3 pm

Heritage Walkathon - Sunday 29/5/2016
@ Beach Street, Penang.

Members , please call office for further details.

Please call office for further enquiries : 04 2272133

This newsletter is published by:

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