

Thalassaemia

A U S T R A L I A

Unifying support and genetics

Thalassaemia Australia Winter 2010 Volume 2 Issue 6

Quarterly



Contents

2010 Committee of Management Meetings	2
Internet resources	3
Community education and advocacy – Support for the Victorian Spleen Registry	4
Grief, loss and chronic illness workshop	5
Research: Compliance, adherence, concordance	6
Research: Magnetic Resonance Imaging and Iron Chelation Therapy	8
Fundraising and events	10
Special feature – Thalassaemia Society of NSW	12
Kid's page	16

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Assisting Victoria's diverse cultural communities in understanding thalassaemia

Thalassaemia Fact Sheets Launch – May 6 2010

Thalassaemia is traditionally referred to as a Mediterranean condition and often associated with people who have a Mediterranean heritage. In Australia, the post war migration from Southern Europe and the Middle East has elevated the prominence of Thalassaemia. Subsequent migrant communities from Southern Asia, and Africa are changing the demographic profile of Thalassaemia affected persons.

It is therefore important to ensure that information on Thalassaemia is available and accessible to all affected persons, in their primary language and is culturally appropriate.

On May 6 2010, in Richmond, Thalassaemia Australia in conjunction with the Murdoch Childrens Research Institute and the Victorian Multicultural Commission, launched its new education and information campaign on thalassaemia, relevant to all culturally and linguistically diverse emerging communities.

The launch was chaired by Mr George Lekakis – Chairperson of the Victorian

Multicultural Commission and formally opened by Hon. Ms Janice Munt – Parliamentary Secretary for Health, Member Legislative Assembly Mordialloc.

Special guests at the launch included:

- Mr Christos Salamanis – Greek Consulate General;
- Ms Maria Vamvakinou – Federal Member for Calwell;
- Dr Paul Fennessy, Manager, Genetics & Health Technology, Department of Health;
- Dr Chris Barnes – Director of Henry Ekert Haemophilia Treatment Centre at the Royal Children's Hospital; Assoc.
- Prof. Sylvia Metcalfe – Group Leader, Genetics Education and Health Research, Murdoch Childrens Research Institute;
- Dr. MaryAnne Aitken – Group Leader, Genetics Education, Murdoch Childrens Research Institute;
- Dr Jim Vadolas – Group Leader, Cell and gene therapy, Murdoch Childrens Research Institute.

(cont. page 2)

Thalassaemia Australia Inc.



Committee of Management 2010

Mr. Sotirios Katakouzinis – President
Mrs. Maria Triantafyllou – Vice President
Mr. George Ambatzidis – Treasurer
Ms. Mary Konstantopoulos – Secretary
Ms. Lien Sam
Mr. John Wilson
Dr. Jim Vadolas
Mrs. Bessy Mougos

Committee meeting dates for 2010

All meetings held at: Thalassaemia Centre,
333 Waverley Road, Mt Waverley 3149

All welcome!

Tuesday 18 May 2010
Tuesday 22 June 2010
Tuesday 20 July 2010
Tuesday 17 August 2010
Tuesday 14 September 2010
Tuesday 19 October 2010
Tuesday 16 November 2010 – also AGM
Tuesday 14 December 2010

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Assisting Victoria's diverse cultural communities in understanding thalassaemia (cont. from page 1)

Under the banner: *Assisting Victoria's diverse cultural communities in understanding thalassaemia*, Thalassaemia Fact Sheets were translated into nine new languages: Arabic, Cantonese, Italian, Greek, Mandarin, Sinhalese, Nuer, Tamil and Vietnamese; to address the needs of our culturally and linguistically diverse community. These translations were sponsored by the Victorian Multicultural Commission and Novartis Oncology.



This event highlighted the importance of the raising awareness of thalassaemia and the need to support current research, in order to find a cure for this condition. It also coincides with International Thalassaemia Day, celebrated by everyone touched by thalassaemia.

These fact sheets are now available on CD and can be downloaded from the Thalassaemia Australia website at www.thalassaemia.org.au.

We would in particular like to thank all who attended and made this a very successful launch of a much needed resource. Support from the Australian Red Cross Blood Service, Orphan Australia and Novartis is also important and much valued.



If you have an event or story you would like publicised please send the details to the newsletter editor at 333 Waverley Road, Mount Waverley VIC 3149, Ph: 03 9888 2211, Fax: 03 9888 2150 or email info@thalassaemia.org.au. Please include the date and time of the event; a description in 20-30 words; venue address; any costs involved and a contact name and phone number and/or email address for public enquiries.

This newsletter is supported by an unrestricted educational grant from Novartis Oncology. All content presented in this newsletter has been independently prepared by Thalassaemia Australia.



Connecting people with services

Infoxchange Service Seeker is Australia's most extensive directory of community services, currently containing more than 240,000 records.

Service Seeker is freely available online 24 hours a day, seven days a week. The directory is readily searchable by agency name, keyword and free text search and can be refined by geographic area.

The information can also be provided in hardcopy and CD-ROM directories and is the basis for the S2S electronic referral system.

The records provide not only contact and address information, but also detailed and precise descriptions of the services offered by the organisations providing those services.

For more information about **Infoxchange Service Seeker**, visit www.serviceseeker.com.au.



How can the internet help you manage your health?

Come along and find out:

- how to source quality and trusted health information
- the best health sites to use on the internet
- how to better understand and manage your health care



Information specialists from The Royal Melbourne Hospital and the State Library of Victoria are offering FREE computer-based workshops for the public. Bookings are essential.

When: Choose one of these six workshops:
Wed 10 March, 4-6pm Fri 19 March, 10am-12 noon
Wed 7 April, 4-6pm Fri 23 April, 10am-12 noon
Wed 5 May, 4-6pm Fri 21 May, 10am-12 noon

Where: State Library of Victoria, 328 Swanston Street Melbourne (Melway ref: 1B N1)

Who: Everyone is welcome but bookings are essential

How: email bookings@slv.vic.gov.au or phone 8664 7099



New website: www.betterhealth.vic.gov.au

Visit the Better Health Channel's new website to find up to date health information, recipes and fact sheets.

Community education and advocacy:

Support for the Victorian Spleen Registry

You may be aware of the great work that is done by the Victorian Spleen Registry to support our members and patients that have had their spleens removed.

In recent times, the Victorian Spleen Registry sort the support of Thalassaemia Australia to assist them in securing funding for their future.

Thalassaemia Australia staff, Victorian Spleen Registry staff and other consumer groups met with Department of Human Service, Dr Rosemary Lester, Deputy Chief Medical Officer and Mr Michael Batchelor (Immunisation Program) to discuss the issues that affect people with reduced splenic function and the need for the Victorian Spleen Registry to continue raising awareness and educating patients, their families and medical practitioners. Thalassaemia Australia emphasised the needs of its patient group, and how to date they have been assisted by the Spleen Registry.

As a result of these discussions, the Victorian Spleen Registry will receive a further allocation of funds for the near future. This is a great result for all concerned and we wish them continued success. We again encourage you all to become members of the Victorian Spleen Registry, and visit their website at www.spleen.org.au.

What is the spleen?

The spleen is the size of a small fist and is situated on the left hand side of the body tucked under the ribs. Its main role is to help the body combat some specific and occasionally severe infections. When the spleen is absent or poorly functioning, this person is at increased risk of bacterial infections.



2010 Community based placement

Greetings! We are two Medical students from Monash University on our Community based placement here at Thalassaemia Australia this year.



As you may already know, Thalassaemia Australia has been participating in the Community Based Placement program for the past three years, and each year, two medical students would be placed here to work on a community health promotion project. For example, last year Marc and Ruchith helped to improve the education resources for schools.

This year, our project will involve everybody! We'd love to find out more about what you feel towards Thalassaemia Australia, so as to help TA to help you better!

To do this, we need all your enthusiastic participation. It is really simple – all you have to do is fill out a short multiple-choice survey. This survey will ask you briefly about your opinions as to what you would like TA to do for you, and which areas you would like TA to focus even more on. Don't worry, you don't have to put your name and contact details, everything is going to be anonymous and your responses will be kept absolutely confidential.

Where can you get this survey then? We will be emailing you the online survey link – so keep a look out! It is very convenient, just a click of the finger! Copies of the survey will also be available at Monash Medical Centre, just where you normally would go for your blood transfusions.

All family and friends and even non-members of TA are invited to participate, and as long as you are above 18, and understand English, you are eligible to help us! This is because we also want to see from their perspective, as a non-member, what they would expect of TA to do for its members and for thalassaemia patients in Australia. This way, we would be able to help TA become better-rounded in its care for you!

If you aren't going to MTU any time soon, and would rather not do the survey online, you could also ring us to post you the survey. All you have to do is fill it in and return it to us either by dropping in the form at the confidential folder at the hospital or by post. If you completed the survey online, that's already automatically returned and you don't have to do anything else. A final report will be released at the AGM.

We really appreciate your help, and look forward to seeing you around this year!

Jane Doan and Xuan Qi Koh



Grief, loss and chronic illness workshop

Thalassaemia Australia attended a workshop on Grief, loss and chronic illness, held by the National Association of Grief and Loss.

The aims of this workshop were:

- To increase understanding of the links between loss, grief and chronic illness.
- To enable participants to increase their confidence to assist people experiencing loss and grief who also have a chronic illness.
- To provide information on and practice in loss and grief counselling in the context of chronic illness.

Areas that were of particular importance to the Thalassaemia Australia community included:

- The 'Normal Grief Storm' by Dr Graham Fulton – which highlights a list of emotions, thought processes that can be associated with grief and loss;
- Loss – can be many things to many people, and is unique to the individual;

- The level of grief and loss experienced by a person is dependant on the coping strategies available to them;
- Anticipatory loss;
- The importance of giving a person 'permission' to grieve and 'naming' their loss;
- Grieving people need information and social support;
- The Flinders Program of Chronic Illness.

Thalassaemia Australia endeavours to continue to increase its knowledge of grief and loss issues, to report these to our members and to offer support its community wherever possible.

For further information regarding Grief and Loss, please call the Australian Centre for Grief and Bereavement on 03 9265 2100.



Daryl & Ros Nankervis

Bereavement Daryl Nankervis

It is with great sadness that we pass on to you some sad news from Ros Nankervis, of the passing of her husband Daryl Nankervis. Daryl died suddenly on 30 April 2010 at the age of 64.

Some of you may remember that Ros Nankervis (nee Smith) was a nurse at the Royal Children's Hospital some time ago.

Over the years, Ros and Daryl continued to take a great deal of interest in the welfare and lives of many of the patients and member of TSV/Thalassaemia Australia.

Thalassaemia Australia extends its condolences and best wishes to Ros and her family during this sad time.

May his soul in peace.

Medication | Update

Ferriprox oral solution (250mL) is now available in Australia. Ferriprox is indicated for iron overload in patients with thalassaemia major who are unable to take desferrioxamine therapy; or in whom desferrioxamine therapy has proven ineffective. Speak to your physician today and discuss the if Ferriprox oral solution is right for you.

Research:

Compliance, adherence, concordance – what's in a word, and does it matter?

Compliance, or adherence, by patients to prescribed therapies is an important aspect of patient care that directly affects health outcomes. The topic is of particular relevance in thalassaemia, where adherence to treatment, especially iron chelation, has a direct impact on survival. For this reason, TIF decided to include presentations from both sides of the patient-doctor relationship in the programme of the 2nd Pan-European Conference on Haemoglobinopathies. We thank both speakers, Dr Chris Sotirellis and Dr Antonio Piga, for their interesting contributions to the debate.

The patient's perspective:

Dr Chris Sotirellis, Vice-President, UK Thalassaemia Society

Dr Chris Sotirellis is an aeronautical engineer by profession, and the Vice-President of the UK Thalassaemia Society. He pointed out that adherence to medication has been a big problem since the time of Hippocrates, who wrote that patients often lied about taking their medication. In modern days, the terminology describing the phenomenon of “taking medication as prescribed” has a significant effect on patients’ attitudes.

From compliance to adherence

Compliance, the term traditionally used, refers to “the extent to which the patient’s behaviour matches the recommendations from the prescriber”. It implies submission or involuntary conformity: “Comply or else...”. It implies passivity on the part of the patient, who, if not compliant, is seen as a rebel, incompetent, or a nuisance. In this approach, the patient’s beliefs are not interacting with the doctor’s, or they are seen as an obstacle to treatment. The higher status of the doctor in relation to the patient is implicit.

The term compliance has now largely been replaced by adherence. Adherence refers to “the extent to which the patient’s behaviour matches agreed recommendations from the prescriber”. It evokes a sense of being closely attached to a set regimen, following the rules, guidelines or standards. However, adherence implies a more active role than compliance: a collaboration with the physician and a self-motivated decision to adhere to advice. In this model, patient acceptance is based on trust and agreement is part of the process.

The next level: concordance

According to the Royal Pharmaceutical Society of Great Britain (1997) – there are two sets of equally cogent health beliefs – that of the patient and that of the doctor. The patient should convey

his/her health beliefs to the doctor, and the doctor should enable this to happen. “The intention is to assist the patient to make as informed a choice as possible about the diagnosis and treatment, about benefit and risk and to take full part in a therapeutic alliance. Although reciprocal, this is an alliance in which the most important determinations are agreed to be those made by the patient”.

This leads to the concept of concordance. Concordance is not synonymous with either compliance or adherence. It is based on the notion that consultations between clinicians and patients are a negotiation between equals, and acknowledges that individual patients may differ in their evaluation of the risks and benefits of a particular medicine. Health professionals are concerned on the quality of their prescribing, aiming at the best possible clinical outcomes. At the same time they are urged to be patient-centered and evidence-based. Yet these two can conflict.

Reasons for non-adherence

Thalassaemia patients are asked to comply with many treatments, including blood transfusions, chelation, treatments for hepatitis, bone disease, diabetes, heart, hormones, etc. It can feel as if life is reduced to “getting treatment”. There is a need to respect the patient’s “normal life” priorities and the patient’s time to fulfil them. Do even so-called centres of excellence respect these needs, for example by providing after-hours transfusion with proper staffing and other support to minimise hospital visits? Is there a willingness to work with the patient? The patient can easily be marginalised and seen as a burden to society, yet with the right care he/she can contribute enormously to society.

Non-adherence can be intentional or involuntary. It may relate to the quality of information (clarity, evidence, source), the impact of the regimen on daily life (costly, painful), the physical and mental capacity of the patient, or his/her social isolation, his/her ability to absorb more of the burden of uncertainty and treatment, or his/her self-image. The treatment offered cannot be predicated solely on the doctor’s views, but also by an understanding of the

real burden on the patient’s life. This understanding comes by “listening” to the patient, and not keeping everything on a clinical level.

What patients expect from their doctors

Patients with chronic conditions use reasoning and judgement to make decisions. In doing so, they must grapple with irreducible uncertainty concerning their life, including its duration and its content (career, having a family, paying off a mortgage, etc.). Medical practitioners often ignore these long-term goals and the uncertainties that affect the daily lives of patients. Yet the practitioner is the only “buffer” the patient has between science, the health services and the pharmaceutical industry. This is the real privilege doctors have, and their power and status derives from it, so they should value it and use it wisely. The doctor’s motivation should always be clear in seeking the optimum for their patients and in their role of buffer between the patient, services and industry.

Environmental and social factors which influence the patient include the interpersonal relationship between the doctor and patient, as well as the quality of support from family members and friends. Most patients intuitively know their doctor’s attitude and willingness to understand these things. The patient requires respect for his/her time, respect for the priorities of his/her life, and honesty on the part of the doctor. The patient hopes that the doctor will get out of her/his routine in order to help, accepting the patient’s “good” and “bad” days, their fears and uncertainties. The patient expects the doctor to share in these battles and be an ally, helping to reduce the burden of the disease and the treatment. Chris says: “The doctor must accept that I am more than just a thalassaemia patient: I am just as complex a being (if not more complex) than she/he is!”

The doctor's perspective:

Dr Antonio Piga, University of Torino Thalassaemia Centre

Dr Antonio Piga heads one of the best known reference centres in the world for treatment of thalassaemia. In his view, the terms “compliance”, “adherence” and “concordance” all refer basically to the same thing: the voluntary cooperation of the patient in following a prescribed regimen, including timing, dosage and frequency.

In general, the adherence rate for long-term treatments is around 50% and much lower for lifestyle prescriptions. In thalassaemia, adherence – particularly to iron chelation – is crucial to survival and better health. Dr Piga reviewed methods for assessing compliance, and emphasised the obstacles to optimal compliance, both emotional and physical.

Obstacles to compliance

One of the obstacles is the so-called “patch” phenomenon: Transfusion does not cure; it compensates the anaemia, giving life and well-being, but also carrying damaging factors as viruses and iron, which has to be removed continuously by chelation. Even when successful in maintaining optimal chelation, the patient is always between the devil (iron toxicity) and the deep blue sea (chelation toxicity). Further, the patient cannot directly feel or experience the effects of both iron damage and iron chelation, so she/he can only realise how harmful it can be after many years

of poor chelation, when damage is already done. Compliance is therefore a function of trust, that is to say of the quality of the doctor-patient relationship. Other factors for non-compliance include body image issues, the daily reminder of the condition and feeling “different”, and the need to be constantly committed to the treatment.

On the emotional side, chronic disease may expose both patients and health professionals to feelings of impotence, frustration, negation, refusal or disinterest. Patients and doctors alike may be susceptible to search for a “magic solution”. Patients, perhaps, do not always realise that their doctors also experience such feelings, since the doctors may activate defensive mechanisms that range from emotional detachment to over-involvement. Both prevent the possibility of a well-balanced and close doctor-patient relationship.

The challenge of the patient's choice

Health professional attitude and health services models have evolved over the years towards greater cooperation, although there are still doctors whose attitudes could do with improving. The traditional paternalistic model (“the good patient obeys”) is unfortunately still popular. The information transfer model (“the capable patient chooses adequately”) leaves the incapable patient behind. The ideal model, the alliance model (“let’s face it together”) is difficult to be achieved. There are still remarkable differences between countries and cultures, however.

According to modern thinking, the patient’s view takes precedence. This raises challenging questions about choice and responsibility: if the patient’s choice falls substantially short of “safe” levels of treatment according to evidence based guidelines, then the doctor may be left with a burden of responsibility that is hard to manage – emotionally, ethically and legally. Patient-centred care, and what the patient desires or can bear, may come into conflict with good medical practice and seriously compromise desirable health outcomes.

How to achieve concordance with good medical outcomes?

In thalassaemia, the problem of compliance has been alleviated to some extent by the advent of oral chelators, but it is noteworthy that they have not done away with the problem.

Key elements to improve compliance include full and sincere information to the patient; accurate assessment of compliance with a listening and caring approach; and removal of practical obstacles to optimal compliance. More generally, a change in the culture of the doctor-patient encounter is needed. This does not mean just improving communication skills, although these are important, but an evolution of the whole relationship to be based on mutual respect – both for the doctor’s professional opinion, and for the patient’s personal decisions.

Compliance is therefore a function of trust, of the quality of the health professional-patient relationship, which considers the emotional as well as the physical obstacles to chelation, and should be a well-balanced and close one. Compliance, therefore, can well be considered as a marker of the quality of care.

This article appeared in the Thalassaemia International Federation, TIF Magazine, Issue 57, April 2010, reprinted with thanks.

Research:

Thalassaemia today with respect to Magnetic Resonance Imaging and Iron Chelation Therapy

by Dr Vasili Berdoukas, Haematologist, Children's Hospital of Los Angeles. Department of Hematology/Oncology

Introduction

Up until the start of this century, the main methods for assessing the degree of iron load in patients on regular blood transfusions, particularly those with thalassaemia, were by ferritin levels and liver iron concentration with biopsy by measuring the amount of iron excreted in the urine after a given dose of either desferrioxamine (DFO – Desferal™) or deferiprone (DFP – Ferriprox™). As cardiac disease due to iron overload, was the most common cause of death in thalassaemia despite chelation therapy, there was more or less no accurate and non-invasive method of measuring cardiac iron.

Ferritin is an acute phase reactant (a substance that reacts with another in a chemical reaction). This means that the ferritin could increase because the patient had an infection or some other problem and could also be low in the presence of vitamin C deficiency.

Liver iron concentration (LIC) did give information with respect to the total body iron load. In most centres this was performed by liver biopsy. The latter was regarded as the gold standard. The problems related to it were that it was invasive, the measurement of iron was not standardized and that the distribution of iron throughout the liver was not homogenous.

The iron excretion in the urine in response to chelation, though relatively easy to perform, was not very accurate and depended on many factors, such as the varying percentage difference of excretion in the urine compared to the faeces, the vitamin C status of the patient and the accuracy of the urine collection.

Magnetic resonance imaging has revolutionized the approach to the assessment of iron load. It can assess the degree of iron load in any organ of interest. Not only can we now assess the total body iron load by evaluating the LIC, but the iron can also be assessed in specific organs. The liver can be assessed by the test called T2* (or R2* which is 1000/T2*) and the T2 (R2). In thalassaemia the additional organs of interest are the **heart, pancreas and pituitary gland**. The specific test for these organs is the T2* (R2*).

Magnetic resonance imaging of the heart also allows the most accurate assessment of cardiac function, even though it does not completely replace the echocardiographic studies.

In a number of centres in groups of patients who have been treated only with DFO, it has been found that significant percentages of patients have had excessive cardiac iron loading and a worrying percentage who have very heavy iron loading. It has also been found that practically all patients who had reduction in their cardiac function had cardiac iron.

Therefore, today MRI is an essential study for patients with thalassaemia. In patients on chelation, it is usually recommended once they are old enough to have the study without needing to be sedated. If it is considered necessary in younger children, then it may be justified to perform it with appropriate anaesthesia or sedation.

MRI Values for Heart and Liver

The following tables demonstrate the significance of the values that are acquired by MRI.

Heart			
Value	Normal	Mild–Moderate	Heavy
T2* (ms)	> 20	10-20	< 10
R2* (sec -1)	< 50	50-100	> 100
LVEF	> 61%		

If the LVEF is between 56-61% there is concern that the patient has abnormal cardiac function. If it is less than 56% this is regarded as definitely needing intervention.

A recent study has shown that patients with T2* in the heavy loaded zone i.e. cardiac T2* < 10ms have a 52% risk of

developing cardiac failure within 12 months of the MRI that shows such iron load. In these patients, it is considered a matter of urgency to reduce the amount of iron in the heart.

Frequency of MRI Testing

In general the frequency of having MRI depends on the values found at the time of the first MRI. If the patient does not have excessive hepatic or cardiac iron and there are no changes made to his chelation therapy and compliance to the chelation remains acceptable, then it is reasonable to repeat the study every two years. If the patient has excessive cardiac or hepatic iron, then it is reasonable to review annually. If the cardiac T2* is in the danger zone (i.e. < 10 ms) then repeat could be performed at six monthly intervals if possible.

Tailoring of Chelation Therapy

The following discussion is very general. In all cases the choice of **chelation therapy should be made after discussions take place between the patient and physician** taking into account the patient's past history with respect to transfusions and chelation therapy, previous compliance with the chelation the patients have been prescribed and adverse reactions that the patients have experienced.

Younger Patients and those who cannot have an MRI for any reason

In this situation any of the three available chelators may be reasonable. In this case it is important to assess the amount of iron received and continuing to be received through transfusions, the mean annual ferritin levels and the expected compliance of the patient to the chelation that will be prescribed.

In Europe the first line of treatment is considered to be DFO therapy. Ferriprox or Exjade may be reasonable for patients in whom DFO has been perceived to be unacceptable or does not reduce the mean ferritin to satisfactory levels. If there are any concerns about heart function, then as Ferriprox has been shown to be cardioprotective. In the future, perhaps the combined use of Ferriprox and Exjade may be an option for therapy in such cases.

Patients with T2* higher or equal to 20 ms with normal or light hepatic iron (LIC <7 mg/g dry weight):

Maintain current chelation therapy. There is however some recent data that indicates that we should be aiming in all patients to bring the liver iron levels to completely normal. This would mean LIC of <0.8mg/g/dry weight. It seems that such levels can only be achieved with the use of intensive combination therapy with DFO and Ferriprox and subsequently still need some form of combination therapy for those levels to be maintained.

Patients with normal cardiac T2* (>20ms) and mild to moderate hepatic iron (LIC <7-15 mg/g dry weight)

For this situation, it is important to determine whether the patient has been complying with their treatment. This would require in depth consultation with the patient and doctor and adjustments made to treatment. If the patient is receiving only DFO it may be reasonable to check the patients Vitamin C levels and if low

provide supplementation. If compliance with DFO is considered inadequate and highly unlikely to improve, it may be reasonable to change to either Ferriprox or Exjade with adjustments made according to the changes found in subsequent MRI studies. If the patient has been receiving Exjade or Ferriprox for a long period of time, their dosage should be maximized and consideration given to adding DFO to their regime.

Patients with normal cardiac T2* (>20 ms) and heavy hepatic iron (LIC >15 mg/g dry weight)

In this situation, it is important to attempt to reduce the LIC to normal levels. If the chelation being received has been given for a long period of time, then dose of the chelators should be maximized. If the dose is already maximal, then combination of DFO and Exjade or DFO and Ferriprox should be considered until levels have reduced and adjustments to the regime can be made.

Patients with light or moderate cardiac iron overload (10 < T2* < 20ms)

In this case it depends on the level of the T2* and also the degree of iron load. If the T2* is < 15ms and/or hepatic iron is elevated, then it seems combination of Ferriprox and DFO is the option most clearly likely to effect the most rapid reduction in both cardiac and hepatic iron. If only the T2* is < 15ms with low liver iron, then Ferriprox may be a reasonable option. If the Cardiac T2* is between 15-20ms, then either Ferriprox or Exjade may be prescribed and combination of Ferriprox and DFO if there is excessive liver iron. As the data on Exjade has not decisively been shown to be effective in reducing cardiac iron, it would be essential to repeat the MRI at least annually if not more often.

Patients with severe cardiac iron overload (Cardiac T2* <10 ms)

In this situation it is essential to reduce cardiac iron as a matter of relative urgency as there is the abovementioned high risk of development of cardiac failure. In this case, optimal treatment is the combination of DFO and Ferriprox. If Ferriprox is contraindicated, then the best option is 24 hour infusions of DFO usually via an implantable intravenous infusion system. This treatment will also improve the liver iron concentration irrespective of its level.

If overt heart failure is present, then initially patients should be treated with continuous intravenous DFO and oral Ferriprox until the cardiac function is stabilized and then start on subcutaneous DFO and oral Ferriprox.

Conclusion

There have been great advances in the management of thalassaemia over the last 50 years. It is essential for patients to have an excellent working relationship with their physicians but the most important part of their care is to accept and use the treatment prescribed. Nothing in the above article can replace the relationship with the medical staff, the guidance and compliance with treatment. Today with the ability to monitor and control iron, it is hoped that we can improve survival and reduce morbidity so that patients live a normal life.

Dr. Vasili Berdoukas is a consultant to ApoPharma Inc. the International distributor of Ferriprox and has a confidentiality agreement with Novartis Inc. with respect to the development of Exjade.

Fundraising and events

Athenian Tavern Night Fundraiser

Friends and families of Thalassaemia Australia gathered at one of Melbourne's favourite Greek restaurants ATHENIAN Tavern in Box Hill for a night filled with excellent Greek food, music, loads of dancing and above all great company.

Whilst the band rested, our President Sotirios Katakouzinis welcomed everyone for coming and thanked everybody for showing their support. The raffle was drawn and we congratulated our lucky winners and we hope they all enjoyed their prizes. We would like to thank our sponsors Galanis Wines, Brady Road Cellars and of course Athenian Tavern for their support once again.

Thalassaemia Australia raised \$1,740 to go towards Thalassaemia Research on the night and our special thanks goes to Mr. C Lolis that has been donating to Thalassaemia Australia over many years and the Society of Kondias Lemnos St. Demitrios and Kondias Lemnos Senior Citizens Club.



Galanis
WINES



An evening with the members of the Gregorios Lodge No.865

The beauty of working for Thalassaemia Australia is that sometimes we have the privileged of attending some interesting and different events.

On 11 March 2010, we were invited to the Gregorios Lodge No. 865 of Freemasons Victoria to receive a cheque for \$861.30 raised by members of the Gregorios Lodge to support the work of Thalassaemia Australia and patients.

TA at the request of Worshipful Master Andrew Karaconstantis gave a short presentation on Thalassaemia and addressed the ways in which community groups such as the Gregorios Lodge can support and promote the awareness of this condition throughout the community and within their own society.

Our evening with the Freemasons was highlighted by the camaraderie shown amongst its members and the warm welcome and hospitality received by Thalassaemia Australia staff.

If you would like to know more about Freemason's Victoria, contact Andrew Karaconstantis on 0433 303 155 or email andkara@bigpond.net.au.

Thalassaemia International Federation Message

May 8 2010 – International Thalassaemia Day

The informed patient: knowledge is power

"The improvement of understanding is for two ends: first, our own increase of knowledge; secondly, to enable us to deliver that knowledge to others."

John Locke (1632–1704)

This year's theme for 8 May has two aspects. The first is the patient's knowledge and information. At TIF we believe that an informed patient is an empowered patient: the more you know, the better you can manage your condition and your life. The more you know, the better you can interact with your doctors and other carers. And when a lot of knowledgeable, strong individuals come together and join efforts, their collective voice will be even stronger.

Knowledge is power

How often have doctors, nurses and other health professionals said about their patients, particularly those with chronic conditions: "My patients understand their disease better than I do!" The experience and knowledge held by the patient can greatly benefit patient care and quality of life, something which gradually is becoming more recognised by the medical world.

Today's knowledgeable patients can make decisions regarding their treatment and work in partnership with their doctors, rather than simply being passive recipients of instructions. But in order to do this, the patient needs the power that comes through education and information. This has long been, and still is, one of TIF's main educational aims.

Sharing the knowledge

The second aspect, as so wisely expressed by the great Enlightenment philosopher, is sharing the knowledge. Patients working together with other patients, parents, volunteers and health professionals – creating associations, getting involved in health policy formulation at a local, then national and even international level. We can really make a difference, not just for ourselves, but for others, too!

The idea of the 2010 slogan is to encourage every patient to become educated, but also to recognise her/his already-existing knowledge, to use it and share it.



Fundraising: Thalassaemia Association of WA

The Thalassaemia Association of WA is currently fundraising. We are selling Entertainment Books for \$65 each. If you would like to purchase a book please contact Teresa Alvaro on 0400 533 036 or email ta42@hotmail.com. The \$12 proceeds from each book sold will go towards the Thalassaemia Association of WA. The fundraising will help our society to continue to fulfil the aims of our society and educate our wider community on thalassaemia.

Good luck to all the Thalassaemia Society's around Australia in their fundraising events!

Letter from the Centre Coordinator



Hello everyone,

I would like to welcome our NSW Patients and Members to the new format of our newsletter.

This is our first newsletter merged with Thalassaemia Australia and we are happy to be able to do this and give our Patients and Members news on a national level to keep you all well informed on what is going on around us.

I would also like to thank Thalassaemia Australia for giving us this opportunity as well as Novartis for funding this project. I am sure you will all enjoy the extra information and reading!

I would also like to keep you all updated on one of my major objectives for 2010 is community education. A programme was developed for schools and community groups within NSW.

I am happy to let you know that we carried out a presentation to the Bangladesh Community of 50 adults, it was very well received and I was notified that everyone left well informed.

We have also been able to reach out to one of the High Schools in our community who invited us to talk about our philanthropic organisation, as well as our community awareness stands which we held at our four main treating hospitals around Sydney. It has been a great experience to get out there and give the public an opportunity to listen to our real life experiences and also explain the importance of donating blood.

Moving forward we have a fantastic Dinner Dance organised by the Thalassaemia Society of NSW (details opposite) which I am looking forward to and hope to see a lot of you there!



Raising awareness for Thalassaemia around International Thalassaemia Day!

This year the Thalassaemia Society of NSW held community awareness stands at our four main treating Hospitals for the week leading up to and the week after International Thalassaemia Day.

Not only did we go out and educate the community about Thalassaemia but we also promoted the importance of blood donation and how it keeps our patients alive.

We found this to be of great interest to everyone who we approached or came up to talk to us and had an overwhelming response by all, we look forward to doing it all over again.

We were able to not only educate adults but also put smiles on children's faces as we offered our special white and red printed balloons.

I wish to thank not only the members of the society and staff who attended and helped out on the day but also to Australia Red Cross Blood Services and Sydney IVF for brochures, stickers and keyrings provided to had out on the day.

If you would like the Society to come and hold a community awareness stand at or near your work place please email your request to: coordinator@thalnsw.org.au.

Thank you!

Nancy Lucich
Centre Coordinator



THE THALASSAEMIA SOCIETY OF NSW



DINNER DANCE

WHEN: SATURDAY 26TH JUNE 2010

WHERE: MYTILENIAN HOUSE,
225 CANTERBURY RD, CANTERBURY

TIME: 6.30PM-12.00AM

PRICE: ADULTS \$60, CHILDREN \$30.

*INCLUDES AN ASSORTMENT OF GREEK MEZETHES, SEAFOOD & MEAT PLATTERS AND FRUIT PLATTER. DRINKS INCLUDE BEER, WINE AND SOFT DRINKS.

*SPIRITS CAN BE PURCHASED ON THE NIGHT WITH A PROPORTION OF PROCEEDS DONATED TO THE THALASSAEMIA SOCIETY OF NSW.

*CHILDREN'S PRICE ASSIGNED FOR AGES 3-12 YEARS.

**FOR MORE INFORMATION AND TICKET SALES
PLEASE CALL MARIANNE 0418970527 OR
MARIA 98962749 / 0406754437**

THE THALASSAEMIA SOCIETY OF NSW THANK YOU FOR YOUR SUPPORT, PROCEEDS OF THIS EVENT
WILL GO TOWARDS ONGOING RESEARCH AND SERVICES FOR OUR PATIENTS.



Diary dates

- | | |
|--|--|
| 26 June 2010
Dinner Dance –
see opposite page | 26 September 2010
Thalassaemia Society
of NSW AGM |
| 26 August 2010
POW Patient Focus
Group | 12-14 November 2010
Patient Family &
Friends Camp |

Introduction to the new Counsellor – Amy Elzahaby

Thalassaemia Society of New South Wales

Hello my name is Amy Elzahaby, I am the Counsellor for the Thalassaemia Society of NSW. I provide counselling sessions and support to our patients and their families.

In my role I work closely with the patients and the Clinical Nurses in the main Thalassaemia treating hospitals in NSW which are Sydney Children's Hospital, Randwick, Prince of Wales Hospital, Randwick, Children's Hospital, Westmead and Royal Prince Alfred Hospital at Camperdown.

I welcome all patients and their families, and encourage anyone who may require any counselling advice, support, information or just a chat to contact me.

My working days are Mondays, Thursdays and Fridays from 9am to 4pm. Email me at counsellor@thalnsw.org.au or call 0437 044 830.



Do something special. Give blood.
Call 13 14 95 or visit donateblood.com.au



POW PATIENT FOCUS GROUP

for all thalassaemia & sickle cell patients

Thursday 26th August 2010
from 6pm to 7:30pm
10 West Tea Room

Enquiries to Emily Allen
CNC Thalassaemia/Transfusion
on 02 9382 4982 or
Emily.Allen@sesiahs.health.nsw.gov.au



Thalassemia and reproductive genetics in the 21st century

Planning a family has always been especially important for carriers of thalassemia. Being aware of the diagnosis from an early age is pivotal for young people who are carriers of thalassemia. Having prior knowledge of the information and options can help carriers and their partners make more informed choices in their reproductive years.

Recent advances in reproductive genetics have significantly enhanced the array of reproductive options available to carriers of thalassemia. One in particular – pre-implantation genetic diagnosis or “PGD” – has opened up greater choice for carriers of this disorder. Indeed PGD is becoming the benchmark for couples planning a family where there are concerns about a genetic condition that has significant implications for a child's health and well-being.

Carriers of thalassemia have several options:

- Use preimplantation genetic diagnosis (PGD) at Sydney IVF, which allows IVF embryos that are not affected by thalassemia to be specifically selected for transfer.
- Have a natural pregnancy and have the developing baby tested at around 11 weeks in the pregnancy. If the baby is found to have thalassemia, termination of the pregnancy is an option.
- Have an unassisted pregnancy, but be prepared for the 1 in 4 chance that the child will be affected by thalassemia.

PGD is simply the combination of IVF and genetics. The improvements in IVF success rates and genetic technology have turned PGD from frontier bioscience in the early 1990s into everyday medicine in the 21st century. In Australia we are approaching almost 1,000 babies born following PGD with the added surety that comes from thirty years of experience with IVF. Between 1-2% of all Australian babies are now conceived using IVF.

In thalassemia, PGD enables a couple to start a pregnancy knowing that the embryo has been tested and shown not to carry the faulty thalassemia gene before it is implanted in the uterus.

PGD at Sydney IVF

Sydney IVF is one of the very few centres in Australia with the vital combination of IVF and genetics facilities to perform sophisticated PGD tests successfully.

The biopsy

Couples wishing to use PGD to select embryos that are not affected by thalassemia will need to have an IVF cycle to create embryos, just like couples with infertility undergoing an IVF

treatment. PGD requires the biopsy or removal of cells from each embryo for analysis.

At Sydney IVF, our advanced embryo culture techniques allow us to wait until the embryos have reached the optimum fifth day of development when they can have a hundred or more cells, and then remove three to four at a time. Other clinics conduct the biopsy at Day 3 of the embryos' development when they consist of just six to eight cells, and only a single cell is removed, greatly reducing the number of opportunities for success.

By waiting until embryos have reached the blastocyst stage, Sydney IVF scientists can select cells from the trophectoderm, the part of the embryo that will go on to form the placenta. The inner cell mass, the part that will become the baby, is not touched.

The biopsy illustrated

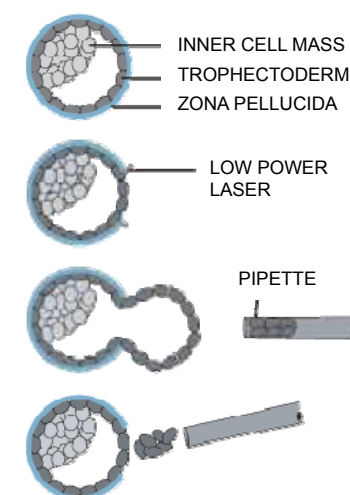
At Day 3 of the embryo's development, a small hole is made in the outer layer of the embryo (the “zona pellucida”) using a delicate laser beam.

The embryo continues development until Day 5 or 6 when it becomes a blastocyst, characterised by the separation of cells into trophoblasts (which go on to become the placenta) and the inner cell mass (which go on to become the fetus).

Trophoblasts are drawn out through the hole using a hollow suction tube called a biopsy pipette. The required cells are separated from the others using the laser and collected separately.

The remaining cells quickly realign and the embryo goes on developing.

If you have or are a carrier of thalassemia, you can discuss whether PGD is applicable with your geneticist or physician, or by calling Sydney IVF on 9229 6420.





Chocolate Chip Cookies

Makes: 20 cookies (Get mum or dad to help!)

Ingredients

- 240g unsalted butter
- 300g (1 1/2 cups) firmly packed soft brown sugar
- 1 large egg
- 1 tsp vanilla extract
- 300g (2 cups) plain flour
- 1/2 tsp bicarbonate of soda
- 350g milk chocolate chips

Method

- Preheat the oven to 190°C.
- Line two large baking sheets with non-stick baking paper.
- Place the butter and sugar in the bowl of an electric mixer and beat on high for five minutes until pale.
- Add the egg and vanilla and beat for a further minute.
- Sift the flour and bicarbonate of soda into the bowl.
- Add a pinch of salt, then use a wooden spoon to carefully fold in the flour until well combined.
- Fold in the chocolate chips.
- Roll two tablespoons of the mixture into a ball, then flatten slightly onto a baking tray lined with non-stick baking paper. Repeat, leaving a fair amount of space between each biscuit to allow for spreading. (You may need to bake the cookies in batches.)
- Bake for 15-20 minutes (the biscuits should have just started to darken at the edges but will still be a little soft).
- Remove from the oven and use a palette knife to lift biscuits from trays, then transfer to a wire rack to cool.

Enjoy!



Q: Why don't acrobats perform in the winter?

A: Because they only do summer-saults!



Hey all,

Well winter is finally here and so is the worlds greatest sporting event FIFA World Cup 2010. Our hosting country South Africa will be all a buzz as too Australia for our Aussie Socceroos. Go aussie goooooooooooooo bring us the world cup. Good luck to all teams and hope your dream team does well!

For all those late night munchers during the World Cup fest, we have included a nice recipe for Giant Chocolate Chip Cookies served of course with a warm or cold glass of milk.

Colour me in!



Q: What do snowmen eat for breakfast?

A: Snow Flakes.