HAEMOPHILIA NEWS

A Publication of the Haemophilia Society of Singapore | MICA (P) 250/06/2006 | Vol 11 August 2009

WHAT'S INSIDE

PRESIDENT'S MESSAGE

A Missed Opportunity

In late February this year, a request was passed to me for a talk on haemophilia at the Republic Polytechnic at Woodlands Avenue 9. The organizer was the student's welfare department, and the audience were the year 1 and 2 students. The date was set for mid May 2009, and finally 22nd May 2009 was decided upon.

At first, I was wondering , with publicity given about haemophilia over the years, what there was to further inform these students. The message came back that these don't read hard print, and they are not yet fully aware of the issues of inclusiveness in our society.

Well then, I shall start with the basics of understanding what haemophilia is and what the social cost is for the person with haemophilia. So I turned for help to Dr Koh Pei Ling, who immediately e-mailed a set of her slides, and with her permission, 50 sets of printed notes were made ready for the lecture.

On the appointed day, I rearranged my appointments, left at 11.45am for the talk at 12.30pm. The campus of Republic Polytechnic was very impressive. I went to the reception desk as instructed, and waited to be met. After 15 minutes, I was apologetically informed that the talk had been cancelled. Due to miscommunication, the email was sent only that morning.

Well, a great disappointment, not with the organizers, as these things do happen, but at the lost chance to inform these future movers of our society about what it's like to live with haemophilia. We think that it is like playing an old record to speak about what we know. But there are always those who have not heard before and every opportunity is an opportunity not to be missed. Haemophilia is not a disease.

I left, not knowing if there will be a repeat request. But I left some copies of the printed notes with the organizer.

Dr Tan Hooi Hwa

Interview with Dr Tien Sim Leng,

Senior consultant, Department of Haematology, SGH

1. **How would a person know if he is a haemophiliac? What are the signs/symptoms?**
   Dr Tien: The signs and symptoms of haemophilia include:
   (1) Bleeding - bruises, joint bleeds.
   (2) Predisposition to the condition due to family history.
   (3) Blood tests for Clotting Factor Assays.

2. **Total number of haemophiliacs in Singapore?**
   Dr Tien: There are a total of 307 in registry (inclusive of Haemophilia A, Haemophilia B and Von Willebrand's Disease (VWD).

3. **Of these, how many have classic haemophilia?**
   Dr Tien: There are 212 cases of classic haemophilia in the registry.

4. **Demographics of the Singapore haemophiliac population (ethnicity, age groups, etc). Of these, how many are children?**
   Dr Tien:
   Breakdown based on ethnicity:

	Chinese	**Malay**	**Indian**	**Others**	**Total**
Haemophilia A	168	27	13	4	212
Haemophilia B	27	2	0	3	32
Von Willebrand's Disease (VWD)	54	9	0	0	63

   Breakdown based on age group:

	0-13yrs	**14-18yrs**	**>19yrs**	**Total**
Haemophilia A	33	20	159	212
Haemophilia B	4	5	23	32
Von Willebrand's Disease (VWD)	2	2	59	63

   Breakdown based on severity:

Haemophilia A				
Severity	**0-13yrs**	**14-18yrs**	**>19yrs**	**Total**
0%	15	3	53	71
1-5%	13	10	47	70
>5%	5	7	59	71
Grand Total:	**33**	**20**	**159**	**212**

   Breakdown based on severity:

Haemophilia B				
Severity	**0-13yrs**	**14-18yrs**	**>19yrs**	**Total**
0%	3	1	2	6
1-5%	0	2	7	9
>5%	1	2	14	17
Grand Total:	**4**	**5**	**23**	**32**

5. **What % of patients are treated in SGH? Where do the rest get treated?**
   Dr Tien: About 80% are treated in SGH. The rest are seen at NUH or KKWCH or private hospitals.

6. **Any other useful information on haemophilia or haemophiliacs?**
   Dr Tien:
   - To learn self infusion so that they can do home treatment.
   - To infuse the factor as soon as possible and not to delay treatment.
   - To take care of their veins.
   - To have some form of exercises, such as swimming, which will help strengthen the muscles. Strong muscles help to protect and support joints. May reduce joint bleeding.
   - Those who had financial difficulties are referred to the medical social worker for help.
   - Patients are also advised to join a support group such as the haemophilia society of Singapore.
   - Genetic counselling is available when needed.
   - Caregivers are encouraged to learn more about haemophilia and learn how to infuse for their sons or husbands.

MOH VWO Excellence Seminar 2009

Ministry of Health organized the half day seminar for Charities and IPCs on Friday, 6 February 2009. The seminar topic was "Beyond Compliance: Embracing the Spirit of Good Governance".

The keynote lecture was delivered by Ms Caroline Oliver, Chief Executive of Good to Govern Ltd. She spoke on "Doing The Right Things The Right Way - Balancing Purpose with Compliance". Mr Gerard Ee, Chairman of National Kidney Foundation, another speaker spoke on "Turning a Dream Board into Reality". This was followed by a panel discussion cum Q&A on "Governing without Managing".

In Ms Caroline Oliver's delivery, VWO's were encouraged to practise correct governance and to comply with regulations not just for complying sake. The importance of the Board was emphasized, that it "had to do the right thing to achieve its purpose", to ensure that the "purpose" is achieved, to be focused on the "purpose" by monitoring, reviewing and being accountable for it. The Board must lead and strive for transparency. For the larger VWO, there should be a distinct separation of the Board ole - to govern - and that of the Executive team - to manage. Overlapping of roles, unclear accountability, unchecked conflict of interests and the danger of "a slave with two masters is a free man" should be avoided.

Mr Gerard Ee spoke on the importance of getting the correct person to serve on the Board. The prospect must be carefully sourced and need to be acquainted with the organisation's mission, vision and to know what is expected of him and the duration of his term. Board roles must be clearly spelt out.

During the panel discussion, it was highlighted that Board members need to understand what was happening in the organization and not only be aware of the agenda for that day's meeting. Those organizations with paid executive staff shared that Board members can be supportive of their work and not to "interfere" unnecessarily with their executive role. The importance of communication between the Board and the executives was stressed.

Summarised by:
Fie Fie Sheyo / Wee Ai Choo

Wellness Talk on 21 March 2009

By Dayrid Foo

On the 21st March 2009, the Society organized a Wellness Talk at The Minding Centre, whereby members were given a treat to learn some simple basic meditation. The instructor was Mr Piya Tan, who patiently guided and taught Members the various ways of meditating. We were given first-hand experience to try out techniques such as the "Breath" and "Loving-Kindness" meditation. At the end of the sitting, a Q&A session was held and from there, various positive feedback were given by our Members with regards to the practice.

By having learnt these basic techniques and practising them daily, I realized that it has indeed improved my general well-being tremendously. Stress levels are reduced and life has been fitter, happier. The mind is more peaceful and clear too. When I have bleeds, I do not chalk up negative emotions too. Meditation is definitely my number one recommendation to anyone seeking a happy life.

June 2009
Mushroom Farm Outing

By Dayrid Foo

On the 20th June, the Society organized a trip where we all had a chance to take a behind-the-scenes look at how mushrooms were being grown and packed at one of Singapore's most renowned farms - Mycofarm.

At around 1400hrs, we all assembled at Newton Hawker carpark to prepare our departure for the farm by bus. The crowd turnout was very good and members, along with their families, came with much anticipation as the outing coincided with father's Day. It was also a god time for members to interact and have fun too!

When we reached the farm's location in Seletar, we were greeted by the in-house guide and taken to the first "greenhouse" where Shiitake mushrooms were being grown. The guide explained to us how the mushrooms were cultured and packed, their origins as well as their health benefits! It is said that by eating Shiitake mushrooms daily, we can reduce our cholesterol levels by as much as 25%! Thereafter, we were taken to another "greenhouse" to be introduced to another kind of mushroom known as the Japanese Oyster Hiratake Mushroom. This mushroom's appearance is very beautiful because it is grown in a bunch, just like white flowers! And like most mushrooms, they contain high amount of nutrients and are easy to cook!

A little tip here for those who buy mushrooms: the white spots that we see on the mushroom caps are NOT fungal growth. Rather, they are just signs that the mushroom were still in their growing stages when they were picked!

After touring the "greenhouses", we were taken for a live cooking demonstration. We were taught on how to cook the various kinds of mushrooms using simple and healthy ways. Everyone got to try samples of the cooking and I will have to agree that it was very delicious!

When we had pleased our taste buds, we proceeded to the final stop in our tour, which was the retail shop. In there, members got the chance to buy various kinds of mushrooms that were grown in Mycofarm. Most of us got back on the bus with big red plastic bags full of mushrooms and I must say, that sure is a great start for healthy eating!

We left Mycofarm for Newton hawker carpark at around four in the afternoon. Though the trip was short, it was highly educational and satisfying. I hope all members who went for the outing felt likewise and had a joyfully enhanced Father's Day!

Comprehensive Care and Hemophilia: Facing the challenges and exploring the benefits

Alison Street, MD
WFH Vice-President Medical

This World Hemophilia Day we celebrate the benefits of provision of comprehensive care. And there are many, including better survival rates, less disabilities, and an increased ability to manage the medical and social needs of people with hemophilia and other inherited rare bleeding disorders. However, in many countries there are also significant challenges, including the lack of resources and funding.

What are these challenges? Hemophilia is an uncommon disorder and can be expensive to treat when using factor concentrates. The endorsement of factor concentrates by the World Health Organization, by including them on its Essential Medicines List, is critical support for negotiations with funding authories. Our challenge is to persuade governments that supporting hemophilia treatment helps ensure a safe and adequate blood and blood product supply to the whole community.

In countries where access to a sufficient supply of safe concentrates exists, there is the challenge of complacency. It is quite easy for both persons with hemophilia and their medical caregivers to take this access for granted. We must all remember that government financial support of these programs and treatment products is constantly under review. Governments need frequent reminders that treating hemophilia ends up being less expensive than covering the medical and social costs of disabilities due to ineffective hemophilia care. We must continue to lobby together for government support and funding.

Another challenge is the lack of professional encouragement for clinicians to practice in this field. Comprehensive hemophilia care centres are developed and defined by the commitment of a multidisciplinary group of healthcare professionals who want to work together as a team. In an era of international shortage of physicians and nurses, clinicians who choose to develop their careers in hemophilia care need to be valued and supported within their hospitals and need to promote their work to their professional colleagues and through the hemophilia community. This requires training and continuous development to further skills in hemophilia care.

The goals of comprehensive care and chronic disease management are to develop systems for care planning and delivery which use evidence-based medicine where available, promote clinical teamwork, and support clinical information systems with a patient-focused approach. These tools, when available to committed clinicians, permit flexibility in dealing with the many complex aspects of hemophilia care. The World Federation of Hemophilia has long encouraged the alliance of multidisciplinary clinicians working together to provide services that address the needs of the hemophilia community.

Studies show that mortality and hospitalization rates of people with hemophilia decrease when they are treated at a hemophilia treatment centre

The benefits of comprehensive care are far reaching and central to treating the physical, emotional, psychological, social, and educaitonal needs of people with bleeding disorders and their families. Studies show that mortality and hospitalization rates of people with hemophilia decrease when they are treated at a hemophilia treatment centre and that resources are used more appropriately with prompt treatment, leading to more effective use of replacement therapy. Patients benefits from education available through such centres and learn from each team member how to manage different aspects of their condition.

This World Hemophilia Day gives us the opportunity to explore not only how we can address the challenges to comprehensive care, but also how we can highlight the benefits. Each step towards engaging members of the healthcare team leads to improved treatment for all those with hemophilia and other rare bleeding disorders.

New Developments In Research

Longer acting concentrates and platelet-delivered factor VIII for patients with inhibitors

Alok Srivastava, MD
WFH Vice-President Communications & Public Policy

Two studies were published recently that highlight exciting new developments for treating hemophilia.

LONGER ACTING CONCENTRATES

Dr. Philip Fay and his group from Rochester, New York, USA, have been working on developing a more stable factor VIII protein. Factor proteins are present in the blood, but once activitated, they gradually disappear. The time taken for the activity of a particular factor to halve is called its "half-life". The half-life of factor VIII is 8-12 hours.

In 2008, Fay and his colleagues first reported their work (*Blood,* 2008; 112: 2761-9), in which they showed that carefully designed molecular changes in factor VIII could increase the stability of both factor VIII and its activated form (cofactor VIIIa). The benefit of these new molecules would mean that factor VIII would survive longer in the blood, meaning less frequent infusions for patients.

The initial results of *in-vitro* (in the test-tube) assessment of the function of these molecules are promising, showing a severalfold increase in their stability. However, more work needs to be done, including *in-vivo* (within a living being) studies in animal models to assess whether the use of modified molecules leads to the development of inhibitors and to determine their efficacy in controlling bleeding. This would be done before human studies can be undertaken. Although there is a long way to go, the prospect of having to use replacement therapy much less frequently is an extremely attractive option.

PLATELET-DELIVERED FACTOR VIII

Another new development is the delivery of factor VIII specifically to the site of bleeding by platelets. Dr. Robert Montgomery and his group from Milwaukee, Wisconsin, USA, have discovered a way to do this and described it in a recent paper (*Blood* 2008; 112: 2713-21).

The Milwaukee group first created mice that expressed factor VIII in their platelets by inserting the actor VIII gene into hematopoietic stem cells (the source of all blood cells) but with a promoter (a sequence of DNA required to switch on a gene) that is expressed only in platelets. These genetically modified stem cells were then transplanted into hemophilic mice with induced inhibitors. Recipient mice had to receive whole body irradiation as "conditioning", a process required for these stem cells to engraft. These stem cells produced platelets that carried factor VIII in them. Hemophilic mice with these transplanted stem cells survived clipping of their tails, which is usually fatal in such animals.

This is a new approach to managing patients with inhibitors. The factor VIII in platelets is protected from being destroyed by circulating inhibitors. In addition, it is delivered to the exact site of bleeding by the platelets, which are part of the first response to any bleeding. It also offers a form of gene therapy for all patients with hemophilia. Much more work will need to be done, including studies in larger animals, to assess the safety and efficacy of this approach. However, this certainly is a significant development in the field.

New Inhibitors Web Section Provides Resource for Patients

Mark Brooker
WFH Senior Public Policy Officer

The World Federation of Hemophilia (WFH) has launched a new website section focused on inhibitors in hemophilia. Inhibitors are a serious medical problem that can occur when a person with hemophilia has an immune reaction to normal treatment with clotting factor concentrates. The new web section explains inhibitors, who is at risk of developing them, what are the signs and symptoms, and what are the treatment options, including immune tolerance induction. It also includes a resource centre with links to publications on inhibitors as well as links to other websites.

For people with hemophilia who have access to sufficient supplies of safe clotting factor concentrates, the development of inhibitors is the number one safety concern. Bleeding is very hard to control in someone with hemophilia who develops inhibitors and treating inhibitors is expensive and time consuming. Studies of children with severe hemophilia A show that about 25 to 30 per cent of them develop inhibitors. Fewer individuals with hemophilia B develop inhibitors, approximately 1 to 6 per cent. There is ongoing research into the prevention and treatment of inhibitors and the WFH will update the section whenever new information is available.

The content of the inhibitors web section was developed and reviewed by the WFH inhibitors working group, chaired by Dr. Keith Hoots. The working group includes healthcare professionals and patient representatives.

Forth Coming Events

1. ***Annual General Meeting***

   23rd August 2009 (Sunday)
   1.30 - 3.30pm
   Medical Alumni Association Auditorium
   Outram Road
   (Annual Merit Awards for students will be presented)

If you would like to give your support towards the welfare of people with haemophilia in whatever way, please drop us a line or send your donations to:

The Haemophilia Society of Singapore
Farrer Road P.O. Box 0273 Singapore 912810

If you require a tax exempt receipt, please supply your NRIC No. / Business Reg. No.