

bloodmatters

A Newsletter for the Haematology Association of Ireland Nurses' Group

April 2007

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Letter from the Editor

Welcome to the 1st 2007 edition of Blood Matters – and welcome to several new members of the HAI nurses committee – more details inside.

We also have reluctantly said farewell to our previous Chairperson, Caitlin McCoy who has offered an invaluable contribution to the HAI Nurses Group for many years. Thank you Caitlin!

We are delighted to once again offer a prize winning crossword – and the solution to the Myeloma crossword from our previous edition. Here's another chance to boost your finances by €50!

A case study is included overleaf regarding the unusual condition of Paroxysmal Nocturnal Haemoglobinuria (PNH). We learn how the role of a haemovigilance practitioner can ensure the delivery of a high quality service and optimise patient safety and education throughout the transfusion process.

This edition also concentrates on the patients' perspective, through both the presentation of a poem written by a haematology patient as he awaited his consultation in out patients and a candid account of living with leukaemia by a 16 year old young woman.

We look forward to seeing you all on Friday 25th May at the annual HAI nurses' spring study day, in the Hilton airport hotel, Dublin. The theme for the day is chronic haematological conditions and promises a wide and interesting selection of presentations. Remember attendance is FREE to all HAI members!!

Yours, on behalf of the HAI nurses committee

Caroline McCaughey
Newsletter Editor

Membership Information

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A Patient's perspective - Alison's story

My name is Alison Balfe and I'm 16. I was diagnosed with leukaemia when I was 14.

I went to my local doctor because I had noticed I had no energy and I had no colour in my face, especially my lips. I just had a blood test at the Doctor's and went home again not thinking there was anything wrong with me. The next day the Doctor rang and told my mam to bring me back to him straight away. From there we were told to straight to Our Lady's Hospital for Sick Children, Crumlin where I stayed overnight waiting to find out what the problem was. The next day a team of doctors from haematology came to see me, it was then I was told I had leukaemia.

My treatment which would take two and a half years started that day. The first ten months would be intense and the other months involved a treatment once every four weeks. The treatment at first was hard going due to the effects of 'chemo' and also the shock I suffered from not being well. I stayed in Crumlin for ten days and then went home. I stayed at home for the start of the treatment but after a couple of weeks I decided it was time to go back to school. I was in third year at the time and I wanted to sit my Junior Cert that year. I went to school when I could, but a lot of time I was too tired. Even though I didn't go that much, going to school cheered me up, seeing my friends and feeling like me again.

After ten months I started on the slower part of the treatment. This was a big difference. I had more freedom and I had more energy. My life felt like it

was getting back on track. I could do more things I hadn't been able to do. I'm in sixth year now and my treatment finished in February 2007.

I have a brother of 12 and a sister and best friend who's 19. I have lots of other friends too; the ones from school, my friends from dancing and my best friend Orla who lives beside me. They were there for me through it all. Orla was there for me the most. If I was feeling really bad we would just sit at home and do nothing.

My two loves in life are going out dancing and watching football (me being a huge Arsenal fan!) or watching any sport. I took a year off from dancing when I was sick and when I started back last year I realized how much I missed it. I recently started playing camogie too.

During the summer I went to the 'Hole in the Wall' Gang Camp in Connecticut through Barretstown. I met many new friends there. Everyone there was either undergoing treatment, some for many years, or finished. It was really enjoyable and I had a great holiday. We spent nine days on the camp and then went on to New York for two days. There were people from all different countries there.

In school engineering, home economics and German are my favorite subjects. I'm part of the stage crew in my school too and we are putting together the talent show and graduation in the school. When I leave school I hope to do media or journalism and become a sports reporter/writer.

The HAI committee wish Alison all the very best and are proud to facilitate the beginning of her career in media by publishing her thoughts in Blood Matters!!!

Case History - PNH

Patient with Paroxysmal Nocturnal Haemoglobinuria (PNH); Transfusion Requirements and the role of Haemovigilance.

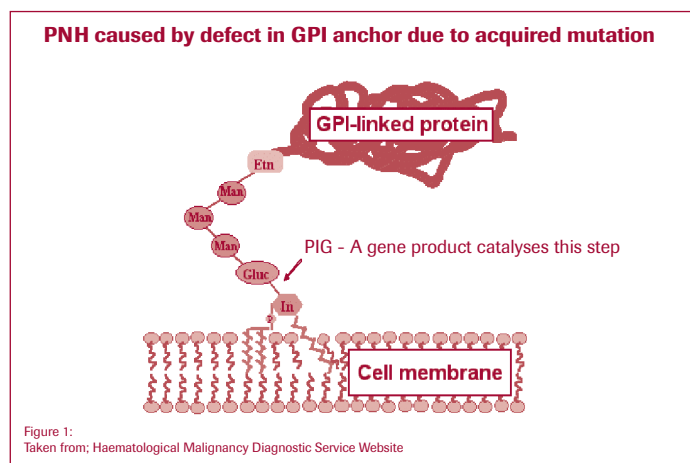
Author: Patricia Gardiner, Transfusion Surveillance Officer, Midland Regional Hospital Mullingar, Co Westmeath

Definition

“PNH is an acquired haemopoietic stem cell disorder characterized by intravascular haemolysis, thrombotic events and bone marrow failure”. (Granata et al., 2004)

Pathogenesis

PNH results from a clonal expansion of a haematopoietic stem cell which has acquired a somatic mutation in PIG-A gene. (Parker et al., 2005) The cells derived from the abnormal clone are deficient in surface proteins normally attached to the cell membrane by a glycosylphosphatidylinositol



(GPI) anchor.

Structure of GPI Anchor

The phenotype is thought to result from lack of CD59 (MIRL: membrane inhibitor of reactive lysis) and CD55 (DAF: decay accelerating factor) which protect against complement.

For poorly understood reasons, possibly due to escape of the PNH clone from immune-mediated damage, there is suppression of normal haemopoiesis which allows the PNH clone to flourish. In 10% of cases, this amounts to actual aplastic anaemia.

The effects of the mutation (X-linked) and the depression of normal haemopoiesis make up PNH. Haemolysis is probably due to the action of complement on abnormal red cells. (Rosse, 2005). The mechanism of thrombosis is thought to be due

to the lack of CD59 from the platelet membranes. (Jarva and Meri, 1999). In some cases the abnormal clone may eventually disappear but the disease can re-activate especially in times of severe infections. (Besa and Woermann, 2005).

Case History

- This 84 year old male was admitted with haemoglobinuria in 2000.
- Laboratory investigations indicated intravascular haemolysis
- Flow cytometry confirmed PNH clone presence in 93% of white blood cells and 13% of red cells. The measured PNH clone is often much lower in red blood cells due to haemolysis of abnormal red cells or due to prior transfusion.
- Since 2000, 35 hospital admissions with 120 units of RCC transfused.
- Never required a Platelet transfusion
- No haemolytic transfusion reactions to date.
- Medications to manage his PNH: Warfarin, Folic acid and Feospan. Generally presents with haemoglobinuria. During acute phases serum urea, reticulocyte count, total bilirubin and LDH are elevated. Serum ferritin, serum haptoglobin have been reduced and his urine has been strongly positive for urinary haemosiderin.

Component Requirements and Haemovigilance Intervention

Treatment of Anaemia

- Iron therapy when deficient only as it may provoke a reticulocytosis and increase haemolysis. (Besa and Woermann 2005)
- Folic acid supplements because of the increased cell turnover by the bone marrow.
- Blood transfusion therapy is initiated at haemoglobin levels of 7 g/dl. monitoring for signs of iron overload. The transfused cells have a better survival than the patient's cells as the defect is intrinsic to the patients own red cells.
- Declined to participate in trial of monoclonal antibody treatment (Eculizumab).

Pre-Transfusion Screening

Patient Blood Group and Rh status	
Antibody Status:	Anti-Chido Antibodies: <ul style="list-style-type: none"> • These antibodies are only found as immune antibodies. • Do not cause red cell destruction. (Mollison 1993). Anti-Jka Antibodies: <ul style="list-style-type: none"> • Usually IgG antibodies • Can cause delayed haemolytic transfusion reactions and • Can disappear rapidly after stimulation (Hoffbrand et al., 2002).
This patient has been phenotyped	<ul style="list-style-type: none"> • IBTS screen for specific Antigen Negative Red cells • Issued least incompatible
RCC Selection	<ul style="list-style-type: none"> • RCC are leucodepleted • Suspended in saline-adenine-glucose-mannitol (SAGM) additive (minimal residual plasma) therefore washed RCC are no longer required. (Hoffbrand, 2002). • Group specific RCC to avoid any complement activation

Haemovigilance

Haemovigilance is a quality process which aims to increase safety in transfusion from the donor to recipient (Faber, 2004). The EU Directive 2002/98/EC has provided a legislative mandate for Haemovigilance. (NHO, 2004). The Directive 2002/98/EC requires:

- full traceability of blood components (Article 14)
- mandatory reporting of adverse reactions and events (Article 15)
- the provision of a quality Haemovigilance System

A quality Haemovigilance System is achieved by:

- Developing and implementation of evidence based transfusion guidelines
- Haemovigilance education and inductions for all staff involved in transfusion
- Quality improvements through clinical audit.
- Risk Management/Adverse Event/Reactions/Near Miss Reporting and root cause analysis provide systematic methods of monitoring processes.

Specific Haemovigilance issues for this patient

1. Good communication between the clinicians and the laboratory medical scientists is

essential to ensure this patient receives specific antigen negative blood.

Safety features include:

- Transfusion Request Forms must provide patient diagnosis and transfusion history
 - Laboratory electronic records of patient diagnosis, antibody status and transfusion requirements.
2. Availability of “Blood transfusion Patient Information Leaflets”, outlining risks, benefits and signs and symptoms of transfusion reactions.
 3. **A Transfusion Alert notice** is affixed to the patient’s medical notes as per NHO (2003) Annual Report. This notice:
 - alerts clinical staff to patient’s need for specialised blood components
 - prompts medical staff to contact the laboratory in advance of an impending transfusion
 - assists in the early notification of the IBTS to provide adequate time to screen for specific antigen negative units.
 4. The National Blood Users Group (2004) advocates the practice of **conducting routine transfusions** where possible during the day. This practice permits close patient observation with increased staffing levels in the event of a transfusion reaction. This patient is observed for signs of transfusion reactions. Guidelines and training for managing severe acute reactions are provided as per NHO (2004) Annual Report.
 5. The NBUG (2001) have highlighted risks of transfusion and advocated avoiding unnecessary transfusion, administration on a unit by unit basis and the assessment of the patient between units. Following a recent trial of Eculizumab, Hillmen et al. (2006) demonstrated stabilization of haemoglobin levels without transfusions was achieved in 49% of the patients assigned to Eculizumab.

Summary

This patient with PNH is supported with RCC transfusion in times of acute intravascular haemolysis. His transfusion management is complicated due to the presence of allo-antibodies so extra vigilance is required during transfusion. Haemovigilance is a quality tool for assessing and promoting safety in blood transfusion for patients and staff.

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The HAI 2006 annual conference

On the 3rd and 4th of November 06 we enjoyed a highly successful Nurses' Group programme at the HAI conference in the Clarion Hotel, Cork City, with a record number of nurses and AHP's registering for the event.

The programme incorporated an eclectic combination of presentations from both nursing and medical clinicians. Friday's programme commenced with an update on emerging therapies in the management of Myeloma and was followed by a fascinating exploration of the present and future options for haematology patients facing infertility as a result of their disease and/or treatment. Prior to lunch, a nurse lecturer, who also has a degree in Law, gave us insight into legal dilemma's and challenges which can and will face the haematology nurse. Despite these somewhat frightening implications we still managed to enjoy a lovely lunch on completion of the morning session!

The afternoon got off to a good start discussing care of the adolescent patient; then the focus changed to a debate on how to optimise the morale of nurses through reflection and group discussion of critical incidents. The morning session concluded with dialogue in respect to interpretation of tissue typing results prior to the nurses joining our medical and scientific colleagues at the annual Liam O'Connell lecture. Regardless of being interrupted by a fire alarm and evacuation of the building, Prof. Greaves eventually delivered an intriguing lecture concerning predisposition to acute leukaemia!

Following poster viewing on Friday evening the committee voted for best nursing poster, the prize for which was awarded to Helen Hume from the Ulster Hospital, for her poster outlining the implementation and impact of the care of the dying pathway.

Consequent to a night of festivities, there were a few bleary eyes on Saturday morning; however a lively and informative talk on "immunology made easy" kick started the day. Subsequently the nursing management and prevention of febrile neutropaenia was explored – no mean feat in itself as we learned how the haematology unit in Cork additionally comprises the local burns unit!

We were delighted to have such a good turn out at nurses AGM where a presentation was made by our outgoing Chairperson, Caitlin who explained how the HAI were taking on charitable status at the

advice of our auditors, with effect from April 2007. We also received an update from the treasurer and the new committee members were nominated and seconded.

The final two presentations were definitely worth waiting for and those who resisted the lure of shopping in Cork were not disappointed! Several case studies were presented regarding unusual and interesting haematology patients, and last but not least a most informative and challenging exploration of cultural issues was led by Marvelle Brown. Positive evaluations were received regarding not only the content but also venue of our conference, and we plan to incorporate any suggestions which you made into the plans for HAI, Sligo 2007. The HAI nurses conference seems to get better year on year – see you in Sligo!

Poetry Corner

I Have Stood

I have stood
On this edge
For so long

Watching and waiting
Never letting go
To the hope
Or the fear
Of something coming.

What is driving
This tension
Always there
At some level?

What else could
Living be?
Lose the edge
At your peril.

This was written by a male patient who was attending a haematology outpatient department when he experienced a sudden feeling of impending doom... He wrote this poem in response; realising that fear was not a negative force in his life, rather the very driving force that motivated every significant achievement. Poem is reprinted with the kind permission of the author.

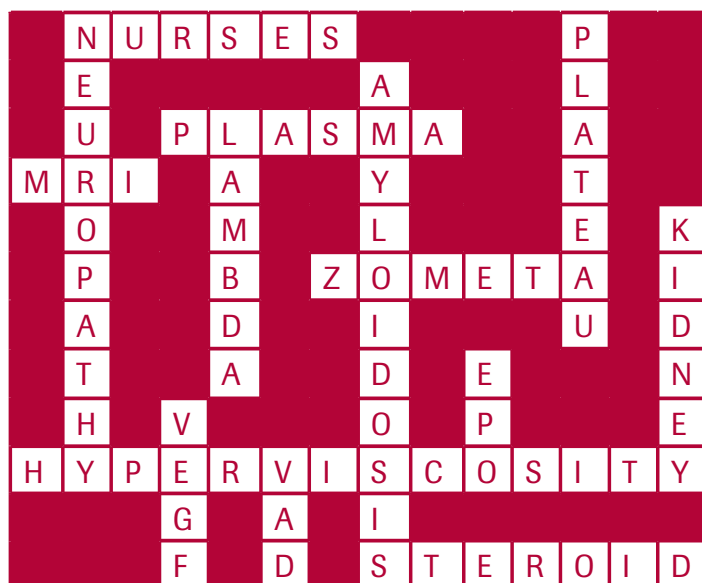
Calling all poets!!

If any of you would like to contribute a poem of your own, or that of a patient, (with permission), please feel free to send it to susan.piggott@belfasttrust.hscni.net

Myeloma

Crossword Solution

(to put you out of your misery!!!).
Sure it wasn't so difficult after all??!!



Cryptic clues

Clues ACROSS

- 1 – They're magic!
- 4 – A fancy screen
- 6 – A metal detector!
- 8 – It's good stuff...but watch your jaw!
- 11 – A sticky situation
- 13 – The only perk

Clues DOWN

- 1 – Bad with your nerves
- 2 – The kind of flat you want
- 3 – A deposit which may affect your balance
- 5 – Light links
- 7 – If I drink I'm not so stressed!
- 9 – This might hurt... your budget
- 10 – A four letter F word!!
- 12 – Orange juice

Winner of the Myeloma crossword....and €50 richer

Ruth Thompson, Deputy Regional Haemovigilance Practitioner, N Ireland. Congratulations!!

Forthcoming Events

25th May 2007

Haematology Association of Ireland Nurses Group.
Spring study day - Chronic Haematological Disorders.
Airport Hilton Hotel
Contact: Susan Piggott @ Belfast City Hospital
Tel: 0044 2890 329241
Bleep: 0675
Email: susan.piggott@belfasttrust.hscni.net

30 April–2 May 2007

British Society for Haematology
47th Annual Scientific Meeting
Bournemouth International Centre, UK
Contact: <http://www.b-s-h.org.uk/confer00.htm>

1st June 2007

European Blood and Marrow Transplantation UK
Nurses and Allied Profession Group
Study day Lancashire County Cricket Club,
Old Trafford Manchester
Contact: <http://www.ebmt.co.uk/>

7th - 10th June 2007

European Haematology Association
12th Congress Neue Messe Vienna, Austria
Contact: www.ehaweb.org

21-23rd June 2007

Advanced Haematology Nursing
Faculty of Medicine, Imperial College
16th Annual International Conference.
Contact: marvelle.brown@tvu.ac.uk

19th–21st October 2007

Haematology Association of Ireland Annual
Conference
Clarion Hotel, Sligo
Contact: susan.piggott@belfasttrust.hscni.net

9th November 2007

Myeloma UK Myeloma UK Education Day for Nurses
Great Southern Hotel, Cork Airport, Kinsale Rd, Cork
Contact: Kirsty on 0044315573332 or email:
kirsty@myeloma.org.uk

9th November 2007

European Blood and Marrow Transplantation UK
Nurses and Allied Profession Group
Study day Birmingham Motor Museum
Contact: <http://www.ebmt.co.uk/>

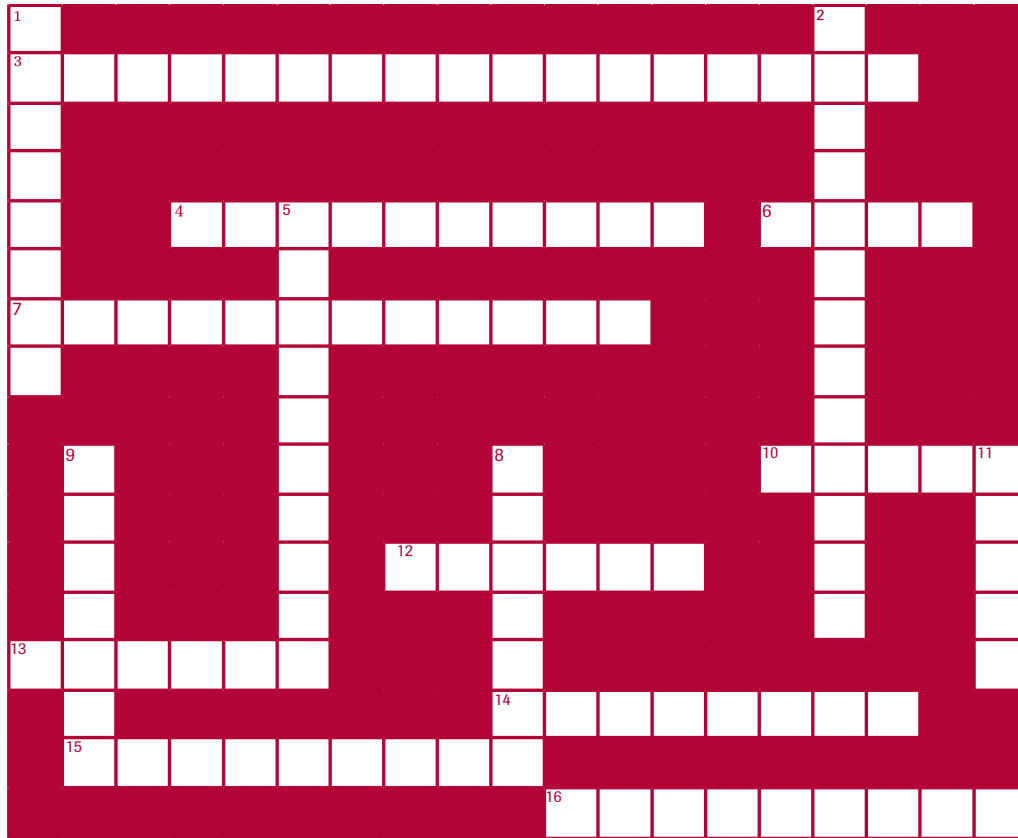
December 8-11, 2007

The American Society of Hematology
49th Annual Meeting and Exposition
Georgia World Congress Center
Atlanta, Georgia
Contact: <http://www.hematology.org/meetings/2007>

Another Prize Winning Crossword.

WIN €50!!

In this crossword all the cryptic clues are related specifically to the topic of **transplantation**. The winner will be randomly selected from all correct entries. Closing date for the competition is **Wednesday 1st August 2007**. Entries should be posted to Caroline McCaughey, Practice Educator, QUB School of Nursing and Midwifery, 50 Elmwood Avenue, Belfast, BT9 7ST



Cryptic Clues

Across

- 3 – Defences disabled
- 4 – Across the foliage
- 6 – Foolhardy
- 7 – After the shampoo
- 10 – Liquid life
- 12 – Prize winning vegetable
- 13 – Transplant by Kellogg’s
- 14 – It started with a locked room (2 words)
- 15 – A hard working car
- 16 – Before an expletive

Down

- 1 – A spinster playing tennis
- 2 – Just my type
- 5 – Family mushroom
- 8 – A farmer’s role
- 9 – A mythological 3 in 1
- 11 – Not a kebab!

NAME _____

JOB TITLE _____

WORK ADDRESS _____

CONTACT NUMBER _____