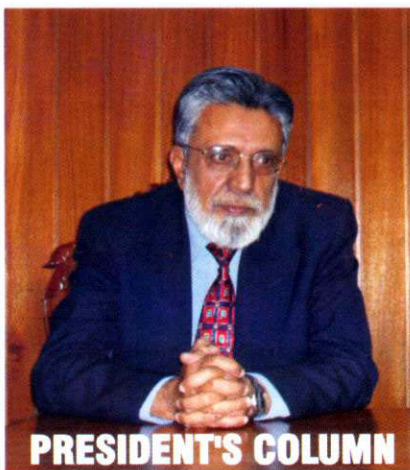


# Newsletter



Volume 3 No 2, April-June 2008

## Pakistan Society of Haematology



**PRESIDENT'S COLUMN**

Let me first apologize for the delay in this issue of the newsletter. Mostly it is because of me. Because of my heavy commitments in my new position I could not write this column in time. Any how the issue is now in your hands.

As you know this is the election year and you are going to elect new office bearers of the Society. Only advice that I can offer is that while nominating and electing the office bearers, please look for people who can devote time for the matters of the Society and for the advancement of Haematology as an independent specialty. Most important issue at hand is to determine whether Haematology, at undergraduate level, is part of Pathology (Basic and Special) or more than that. This can not be determined unless curriculum of Haematology for MBBS and BDS is well defined. The Executive Committee has constituted a Committee to do that. Once the curriculum is defined, it can easily be determined that at what levels a qualified Haematologist be involved in teaching at undergraduate level.

This will also determine the course which postgraduate training in Haematology should take (Clinical, Laboratory or combined).

Blood Transfusion Services, in near future, are going to take new shape for the sake of safety of both recipients and donors. Although all Provinces and Federal Government have promulgated legislations to ensure safe blood transfusion through Blood Transfusion Authorities, they lack uniformity. A uniform legislation is going to be introduced shortly. There is also a plan for separating blood collection, screening, processing and supply of blood and products from blood transfusion in hospitals. The "organized" blood transfusion services will require a large number of appropriately qualified and well trained doctors and technicians. It is high time that the Society should seriously look into this and prepare for provision of required number of trained professionals.

It was decided long ago that all haematology events organized in the country, particularly by members of the Society, will also carry the logo of the Society. I have pointed out earlier that some of our colleagues are not doing that. Members have every right to promote their respective Institutions but at the same time they have an obligation to promote the Society as well. I expect that members will take care in future. After all, almost all the participants in the events belong to PSH.

Another important area is coordination of events. This was decided that all members before finalizing the dates of their event will inform the central office to ensure that their activity is not interfering in activity organized by another colleague. I urge once again to take care in future.

Holy month of Ramadan has started. I extend my heartiest felicitations to all of you at this occasion. I pray that this month leaves lasting effect on our lives.

With regards,  
**Masood Anwar**



## Upcoming Events

### **11<sup>th</sup> Annual Conference Pakistan Society of Haematology**

11<sup>th</sup> Annual Conference of Pakistan Society of Haematology will be held from 13-15 February 2009. Further details and schedule of the conference will be announced by organizers in the near future and will also be available on PSH website. For further details please contact Dr Mussarat Niazi Secretary Conference at PSH Conference Secretariat Pathology Department Khyber Medical College Peshawar.

### **Elections of PSH office bearers**

Elections for PSH office bearers will be held by end of this year. Those members (excluding life members) who have not cleared their dues are requested to clear it as soon as possible so that they are eligible to vote. A list of those who are eligible for voting will be sent to all members in the near future.

### **Establishment of Haematology Ward and OPD at PGMI Hayat Abad Medical Complex Peshawar**

A 20 bedded haematology ward and OPD has started functioning from 1<sup>st</sup> of July 2008 at PGMI Hayat Abad Medical Complex Peshawar. It will greatly facilitate timely treatment to the local patients suffering from malignant and non-malignant haematological disorders. Besides this it will also boost the development of clinical haematology.

## **Long-term safety of hydroxyurea in sickle cell disease and thalassaemia**

**Dr Parvez Ahmed**

Hydroxyurea (HU) is known to have beneficial effect in the sickle cell disease by promoting increased production of fetal haemoglobin (Hb F)<sup>1</sup>. It has considerably improved the prognosis of patients with this disease and with modern therapy mean life expectancy of sickle cell disease is about 50 years. Hydroxyurea has also improved quality of life in these patients and many patients now reach reproductive age. A significant benefit of hydroxyurea has also been reported in patients suffering from beta thalassaemia intermedia<sup>2</sup> and even thalassaemia major<sup>3, 4, 5</sup>. This effect is due to partial amelioration of globin chain imbalance since increased availability of ( $\Gamma$ ) Gamma chain can bind excess ( $\alpha$ ) Alpha chains. Clinical trials have shown that this partially restored balance between  $\alpha$  and non  $\alpha$ -globin chains is associated with beneficial effects in thalassaemia patients. Sustained discontinuation of red cell transfusions have been reported in small number of cases suffering from beta thalassaemia major. Hydroxyurea can be used in patients suffering from thalassaemia intermedia to minimize or even obviate the need for regular transfusions and concomitant iron overload<sup>2</sup>. G-gamma Xmn1 polymorphism has been reported to be particularly associated with better responses to hydroxyurea<sup>4, 5</sup>.



# Newsletter



In view of easy availability and low cost more and more patients of sickle cell and beta thalassaemia are being put on prolonged hydroxyurea treatment. Concerns about long-term effects of hydroxyurea have been raised regarding mutagenic effects inducing chromosomal and teratogenic aberrations. The effects however have not been noted in two independent clinical studies even after long-term exposure to hydroxyurea<sup>3</sup>.

HU has been associated with impaired spermatogenesis in mammals, resulting in testicular atrophy, a reversible decrease in sperm count, abnormal sperm morphology and motility. Chromatin structure of germ cell is also affected with increased apoptosis. First case of possible adverse effect in human was development of hypogonadism with testicular atrophy and gynecomastia in a 68-year old male with essential thrombocythaemia who received HU for 8 years<sup>6</sup>. The effects reversed with testosterone treatment. Recently a large case series involving 108 semen samples in 44 patients of sickle cell disease has been published<sup>7</sup>. In this study there was decrease in ejaculate volume, sperm count, motility, normal morphology and vitality between pre and post-hydroxyurea treatment samples. Overall fertility in this population was however reported to be conserved.

Due to improvements in treatment, majority of the patients with sickle cell disease and a large number of patients with thalassaemia intermedia and thalassaemia major are likely to attain reproductive age. The alterations in the semen analysis highlight the need for follow up semen analysis in patients who are on long-term treatment with HU till large-scale prospective studies settle this issue.

The potential effect of HU on male fertility requires evaluation and would be a major concern necessitating advice on sperm cryopreservation as a preventive measure to preserve male fertility. This may also represent a possible obstacle for HU treatment in young patients with sickle cell disease and thalassaemia.

## References:

- 1- Charache S, Terrin ML, Moore RD, Dover GJ, Barton FB, Eckert SV, et al. Effect of hydroxyurea on the frequency of painful crises in sickle cell anemia. Investigators of the Multicenter Study of Hydroxyurea in Sickle Cell Anemia. *N Eng J Med* 1995; 332:1317-22.
- 2- Karimi M, Darzi H, Yavarian M. Hematologic and clinical responses of thalassemia intermedia patients to hydroxyurea during 6 years of therapy in Iran. *J Pediatr Hematol Oncol.* 2005; 27: 380-5.
- 3- Yavarian M, karimi M, Bakker E, Harteveld CL, Giordano PC. Response to hydroxyurea treatment in Iranian transfusion-dependent -thalassemia patients. *Haematologica* 2004; 89: 1172-8.
- 4- Bradai M, Pissard S, Abad MT, Dechartres A, Ribeil JA, Landais P, et al. Decreased transfusion needs associated with hydroxyurea therapy in Algerian patients with thalassemia major or intermedia. *Transfusion* 2007; 47: 1830-6.
- 5- Koren A, Levin C, Dgany O, Kransnov T, Elhasid R, Zalman L. Response to hydroxyurea therapy in beta-thalassemia. *Am J Hematol.* 2008; 83: 366-70.
- 6- Beeson M, Antwi EK, Vesely DL. Hydroxyurea-induced hypogonadism. *Endocrinologist* 1999; 9: 389-90. Berthaut I, Guignédoux G, Kirsch-Noir F, de Larouziere V, Ravel C, Bachir D, et al. Influence of sickle cell disease and treatment with hydroxyurea on sperm parameters and fertility of human males. *Haematologica* 2008; 93: 988-93.



# Newsletter



## Your Views and News

**Dear Colleagues:** Your contributions to PSH newsletter are backbone to its success. The response so far has been lukewarm. Please send short communications, case reports, scientific activities and developments in your departments and issues of common interest. Photographs of scientific events/meetings are also welcome.

## Update Address

Please update your addresses in case there is any change in it. All members are requested to email us their mobile/phone contact and email address.



### Correspondence

#### **Dr Parvez Ahmed**

Consultant Haematologist

Armed Forces Bone Marrow Transplant Centre, Rawalpindi

Tel: 051-56130771, 051-56134011 (Office)

051-56134043, 0515580460 (Residence)

Mobile: 03008561288, Email: [parvez101@yahoo.com](mailto:parvez101@yahoo.com)

**Sending you special wishes across the miles for a joyous and a wonderful Eid**



 **LEUKOKINE Inj.**  
Filgrastim / r-metHuG-CSF

**Medac Disodium Pamidronate**  
(Disodium Pamidronate 30mg)