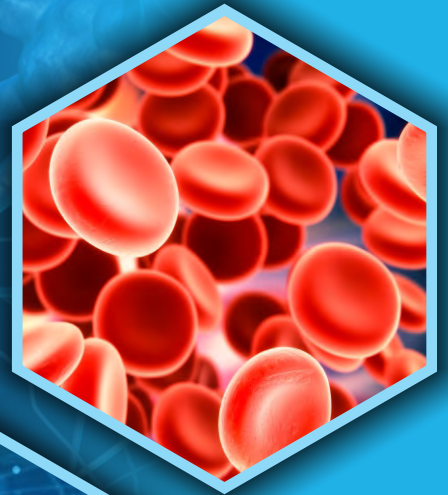




# HAEMATOLOGY UPDATES

Vol. 13 No. 4, October - December 2018



## Events:

- 13th FCPS Haematology Intensive Course, 2018
- PSH Monthly Meeting at KEMU, Lahore
- PSH Monthly Meeting at SZH, Lahore
- PSH Monthly Meeting at Peshawar
- 10th PSH National Haematology Symposium, Mirpur-AJK

## Inside This Issues

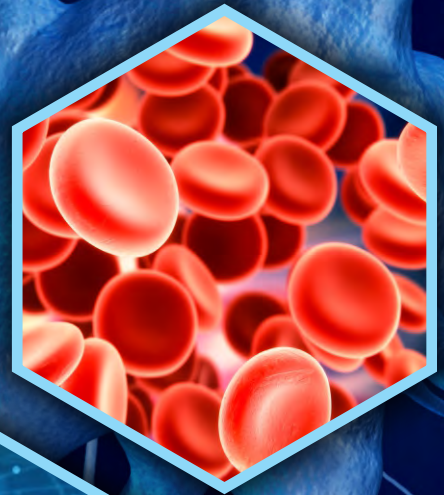
- National Registry Performs
  - Fanconi's Anaemia
  - Glauzman Thrombos Asthenia
- 11 Case Reports
- Choosing Wisely Canada
- Quality Manual for Laboratory
- Haemcon2019, Karachi-Pakistan



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# Views & News

The Pakistan Society of Haematology updates is published on a quarterly basis and is a quick guide to all the happenings in the haematology community. To improve the updates, your comments and suggestions are welcome. We further encourage you to send us write ups and photographs of any PSH event in your city/province and they would be featured in our upcoming updates. For contact, please refer to our corresponding address.

This updates was designed and edited by:

**Dr. Munir Ahmed**

## CORRESPONDENCE

**Dr. Saima Farhan, Secretary PSH**

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## President's Column

Our Dear Colleagues

Assalam-o-Alaikum,

My bundle of thanks to PSH members for support and trust in Pakistan Society of Haematology. I humbly request all of you to pray Almighty Allah to give us good health, courage and strength to fulfill this responsibility.

By the grace of Allah, our different Working Groups have finalized their recommendations; their proposals and final comments will be presented in upcoming Haemcon2019, Karachi. I request all PSH members to participate with their colleagues in this Mega Event of Haematology, in which a large number of prominent National and International Haematologist will present latest developments in the field of Haematology, BMT, Coagulation Medicine, Transfusion Medicine, Cellular Therapy and Gene Therapy. So never miss this wonderful academic and social get-together event to meet your friends.

Allhamdulillah we have started a long awaited dream of "Haematology Registry" for different diseases Diamond Blackfan Anaemia, Congenital Dyserythropoietic Anaemia, Fanconi's Anaemia, Glanzman Thrombasthenia, Bernard Soulier Syndrome, Factor-VII Deficiency, Factor-X Deficiency and Factor-XIII Deficiency. Kindly register your patients in these registries to gather Pakistan database for different Haematological condition. A Bone Marrow Transplant Registry is already functioning smoothly with the untruing efforts of our PSH Rawalpindi/ Islamabad Chapter.

Pakistan Society of Haematology have signed "MOU" for collaboration with International Society of Haematology (ISH), International Society of Laboratory Haematology (ISLH), International Society of Thrombosis and Haemostasis (ISTH), Korean Society of Haematology (KSH), Japan Society of Haematology (JSH), Turkish Society of Haematology (TSH), International Haemophilia Academy, Asia Pacific Bone Marrow Transplant (AP-BMT) and European Blood & Marrow Transplant (EBMT).

We are also in process of understandings with National & International Societies for more integrated approach for multidisciplinary management.

PSH National Symposiums in different corners of Pakistan to facilitate Haematology services have been arranged in Quetta, Hyderabad, Bahawalpur, Multan, Sahiwal, Faisalabad, Abbottabad, Mirpur (AJK) and Peshawar. In next few months PSH National Symposiums will be held in Sargodha, Larkana, Muzaffarabad (AJK), Gilgat and Swat.

Last but not the least, my dear friends the strength our PSH lies in unity and to increase PSH Family, kindly increase the new members.

With Thanks & Allah Hafiz

**Prof. Dr. Nisar Ahmed,**  
President,  
Pakistan Society of Haematology







*Join us!*



Pakistan Society of Haematology has been serving as a platform to raise awareness about the role of Haematology in the medical field. Also working to bring the hematologists all over from Pakistan and abroad, so that they can share their experiences and knowledge.

The current office bearer of PSH has adopted the strategy to expand the learning opportunities in the form of symposium, seminars and workshops to far flung areas of Pakistan. I appreciate all those who have been a part of this struggle.

Hope such activities will continue in the future. However it cannot be possible without the efforts of all our prestigious members. So kindly volunteer yourself, present innovative ideas and even organize your own event or activities to promote Haematology as a specialty.

All interested kindly email their details with areas of interest on [psh.org.pk@gmail.com](mailto:psh.org.pk@gmail.com),

**Dr. Saima Farhan,**  
Secretary/ Treasures  
Pakistan Society of Haematology



## About PSH

Pakistan Society of Haematology was formed in 1996 with the aim of promoting advancement of haematology, BMT and transfusion medicine in the country. Presently it has more than **500** members and we all should make efforts to enroll every haematologist in the country. We request all our members to take special interest in extending the membership to all those haematologists around you who have not yet registered with PSH. Website was launched and has been very active in the recent past. We are trying to rejuvenate the website "<http://www.psh.org.pk>. The website would be interactive and provide on line forum for sharing views with other haematologists, and case discussion with the experts. Other features will be facility to download online membership form, newsletter, list and addresses of the members. Hopefully the website will be more operational within this month InshaAllah.

### PSH History

#### Gen Masood Anwar

PSH was raised as "Pakistan Society of Haematology/Transfusion Medicine (PASHT)" in 1991. A meeting was held at 5 pm on Friday Nov 22, 1991. Professor Dr Mohammad Khurshid, Brig (later Lt Gen) Muhammad Saleem, Dr Khalid Zafar Hashmi, Dr Nasim Siddiqui, and Dr Abdul Hayee attended the meeting as members in presence of Prof A. V Hoffbrand. In this meeting Dr Khurshid presented a brief outlay of the necessity to create such a society. He also pointed out that Dr. Abdul Hayee, Dr. Khurshid, Dr KZ Hashmi and Brig Saleem had met at Bahawalpur and agreed on the general principles that the first meeting would be held along with the International conference of Pathology.

Though initial work was comprehensive, governing body and meetings of PASHT were not held regularly. In Sept 1994 it was proposed by Gen Muhammad Saleem to meet all PASHT members during Pakistan Association of Pathology (PAP) conference at Quetta. Dr. Muhammad Khurshid in consultation with Gen Saleem, Prof. Abdul Hayee, Dr. Khalid Zafar Hashmi proposed a provisional constitution of PASHT for the discussion in meeting

Haematologists from all over the country met on Saturday 9 March 1996 at Hotel Pearl Continental Rawalpindi in order to form a society. It was unanimously agreed that official name of society will be "Pakistan Society of Haematology" with official abbreviation of "PSH". It was also decided that until elections for office bearers the society matters will be looked after by a committee as under

- a. Dr. Muhammad Khurshid
- b. Dr. Ehsan-ul-Allah
- c. Dr. Abdul Hayee
- d. Dr. Khalid Zafar Hashmi
- e. Dr. Khalid Hassan
- f. Dr. Masood Anwar will act as Co-ordinator





## About PSH

4. A general body meeting of PSH was held at Peshawar on 2 and 3 Nov 1996. Election for office bearers were carried out as follow

- a. Lt. Gen. Muhammad Saleem President
- b. Prof. Muhammad Khurshid as Vice President
- c. Dr. Khalid Hassan as Secretary/treasurer

Later in Oct 1997 appointment of vice president was renamed as president elect.

List of past presidents includes

1. Lt. Gen. M. Saleem
2. Prof. Dr. Abdul Hayee
3. Prof. Dr. Abdul Khaliq
4. Prof. Dr. Muhammad Khurshid
5. Prof. Dr. Khalid Zafar Hashmi
6. Maj. Gen. Masood Anwar
7. Prof. Dr. Khalid Hassan
8. Maj. Gen. Suhaib Ahmed
9. Prof. Dr. Samina Naeem
10. Gen. Muhammad Ayyub

List of past secretaries includes

1. Dr. Khalid Hassan
2. Maj. Gen. Masood Anwar
3. Prof. Fazle-e-Raziq
4. Dr. Salman Naseem Adil
5. Dr. Shaheena Kauser
6. Brig. Nadir Ali
7. Maj. Gen. Pervez Ahmed
8. Dr. Nadeem Ikram
9. Dr. Humera Rafiq
10. Brig. Tariq Mehmood Satti

5. PSH was registered with Govt of Pakistan on 8 August 1998(RS/ICT/298 dated 8 Aug 1998 as non political and non sectarian body to promote advancement of haematology including transfusion medicine through encouragement of research, teaching and technical methods. The body will also organize scientific meetings, publication of scientific material, and affiliation with other National and international organizations. Members of Governing body included

Lt. Gen. Muhammad Saleem as President  
Dr. Khalid Hassan as General secretary  
Dr. Birgees Mazhar Qazi as member  
Dr. Waseem Iqbal as member  
Dr. Hassan Abbas Zaheer as member  
Dr. Mobina Ahsan Dhodhy as member  
Dr. Farah Yasin as member  
Col. Masood Anwar as member

It was also decided that First National conference will be held on 4 Oct 1998. Since then Annual conference is held regularly in all capital cities of Pakistan. The society is publishing a quarterly newsletter and providing a forum to the haematologists all over the country contributing as advisors in haematology, consultants, researchers and mentorship. Currently the Governing body includes

- Prof. Dr. Nisar Ahmed as president
- Gen. Parvez Ahmed as President elect
- Dr. Saima Farhan as Secretary



## Executive Committee

New Executive committee was elected during 19<sup>th</sup> Annual Conference of Pakistan Society of Haematology held at Lahore from 16th-18th February 2017. Following are the office bearers of executive committee.

### PRESIDENT

Prof. Dr. Nisar Ahmed  
0300-4330196  
dr\_nisarahmed@hotmail.com

### PRESIDENT ELECT

Maj. Gen. Pervez Ahmed  
0300-8561288  
parvez101@yahoo.com

### SECRETARY/TREASURER

Dr. Saima Farhan  
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dr\_saima99@yahoo.com

## MEMBERS

### ARMED FORCES

Brig. Ch. Altaf Hussain  
Brig. Maqbool Alam  
Brig. Saqib Qayyum

### ISLAMABAD

Prof. Dr. Ayesha Junaid

### PUNJAB

Dr. Muneeza Junaid  
Dr. Manzoor Hussain  
Prof. Dr. Arif Hussain

### SINDH

Prof. Dr. Muhammad Irfan  
Prof. Dr. Salman Naseem Adil  
Dr. Muhammad Nadeem

### BALUCHISTAN

Prof. Dr. Chandi Kapoor

### KPK

Dr. Shah Taj Khan

### AZAD KASHMIR

Dr. Zahida Qasim (Mirpur)

### OFFICE ASSISTANT

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itsme.immy@gmail.com

### OFFICE ASSISTANT

Mr. Abdul Aleem  
0334-4391558  
aleemtospeak@gmail.com





## National PSH Coordinators

### RAWALPINDI/ISLAMABAD

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### KARACHI

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0300-2160765

### LAHORE

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### QUETTA

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0300-8380847

### PESHAWAR

**Prof. Dr. Shah Taj Masood**  
shahtajmasood@yahoo.com  
0333-9118335

## PSH National Advisory and Steering Committee

- Gen. Muhammad Saleem
- Prof. Abdul Hayee
- Prof. Muhammad Khurshid
- Prof. Abdul Khaliq
- Prof. Khalid Zafar Hashmi
- Gen. Masood Anwar
- Prof. Khalid Hassan
- Prof. Yasmin Lodhi
- Prof. Tahir Jameel Ghazi
- Maj. Qaiser Husnain
- Col. Ghulam Rasool
- Prof. Farzana Amjad
- Prof. Nouman Malik
- Prof. Fozia Butt
- Gen. Suhaib Ahmad
- Prof. Samina Naeem
- Gen. Muhammad Ayub
- Prof. Fazle Raziq
- Prof. Javed Asif
- Brig. Muhammad Amin
- Col. Farooq Khatak
- Dr. Barjees Mazhar Qazi
- Prof. Saeed Ahmed Malik
- Prof. Nighat Yasmin Ashraf
- Brig. Jalil Anwar
- Prof. Waseem Iqbal
- Dr. Syed Iftikhar Abdi
- Brig. Ehsan Alvi
- Brig. Zahoor ur Rehman
- Prof. Luqman Butt
- Brig. Farhat Abbas Bhatti
- Brig. Nadir Ali
- Brig. Muhammad Ashraf
- Prof. Tahira Zafar
- Prof. Zeba Aziz
- Dr. Madoodul Manan
- Prof. Muhammad Hirani
- Prof. Zahoorul Latif
- Dr. Mian Muhammad Sharif
- Prof. Mussarat Niazi
- Prof. Muhammad Saeed Talpur
- Prof. Atifa Shoab



## PSH Working Group

### PSH Notice

The current office of Pakistan Society of Haematology (PSH) has done much hard work over the course of two years in form of organizing different academic activities, seminars and workshops to promote Haematology in Pakistan. As you all know in the 1st Executive Council Meeting, working groups were formed and assigned tasks. It is requested to all lead persons to present their progress report in General Body Meeting in 21st annual PSH meeting "Haemcon2019" 14th – 16th March, 2019 Aga Khan University Hospital, Karachi. Let us know if any help from PSH office is required.

Thanks,

**Dr. Saima Farhan**

Secretary / Treasures

Pakistan Society of Haematology

#### I. Working Group JPSH (Journal of Pakistan Society of Haematology)

**Lead Person Prof. Dr. Khalid Hassan**

Cell: 0333-5178210

Email: kh\_pims@yahoo.com

- **Members:**
  - Prof. Dr. Shahida Mohsin
  - Dr. Bushra Moiz
  - Dr. Nadeem Ikram
  - Brig. Nadir Ali
  - Dr. Anum Wasim

#### II. Working Group Blood & Marrow Transplant

**Lead Person Maj. Gen. Parvez Ahmed**

Cell: 0300-8561288

Email: parvez101@yahoo.com

- **Members:**
  - Maj. Gen. Tariq Mehmood Satti
  - Prof. Dr. Salman Naseem Adil
  - Brig. Qamar-un-Nisa Chaudhry
  - Dr. Saima Farhan
  - Col. Mehreen Ali

#### III. Working Group Academic/ Curriculum Development:-

**Lead Person: Gen. Saleem Ahmed Khan**

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Email: saleem003@hotmail.com

- **Members:**
  - Maj. Gen. Parvez Ahmed
  - Prof. Dr. Salman Naseem Adil
  - Prof. Dr. Nisar Ahmed
  - Dr. Bushra Moiz
  - Brig. Nuzhat Salamat
  - Dr. Shah Taj
  - Dr. Saima Farhan

#### IV. Working Group Scholarship and Financial Aid:-

**Lead Person: Brig. Nuzhat Salamat**

Cell: 0321-8514354

Email: nuzhatsalamat@yahoo.com

- **Members:**
  - Dr. Lubna Zafar
  - Dr. Muneeza Junaid
  - Dr. Saima Farhan
  - Dr. Tariq Ismail





## PSH Working Group

### V. Working Group Transfusion Medicine:-

**Lead Person** Brig. Maqbool Alam  
Cell: 0321-5196521  
Email: maqboolalam448@hotmail.com

- **Members:**
  - Prof. Dr. Hassan Zaheer Abbas
  - Dr. Saba Jamal
  - Dr. Ayesha Junaid
  - Dr. Zahida Qasim
  - Dr. Manzoor Hussain
  - Dr. Saima Farhan
  - Dr. Nazish Saqlain

### VI. Working Group Paediatric Haematology:-

**Lead Person** Prof. Dr. Nisar Ahmed  
Cell: 0300-4330196  
Email: dr\_nisarahmed@hotmail.com

- **Members:**
  - Lt. Col. Tariq Ghafoor
  - Dr. Aslam Shaikh
  - Dr. Tooba Fateen
  - Dr. Faiza Rafiq

### VII. Working Group of Coagulation Medicine:-

**Lead Person** Brig. Ch. Altaf Hussain  
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Email: altaf444@gmail.com

- **Members:**
  - Prof. Dr. Mona Aziz
  - Prof. Dr. Samina Amanat
  - Prof. Dr. Arif Hassan
  - Brig. Saqib Qayyum
  - Dr. Naghmana Mazhar

### VIII. Working Group Benign Haematology:-

**Lead Person** Prof. Dr. Muhammad Irfan  
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- **Members:**
  - Prof. Dr. Nadeem Samad Shaikh
  - Prof. Dr. Mubeena Dhodi
  - Prof. Dr. Ayesha Ehsan
  - Dr. Usman Shaikh
  - Col. Nighat Shahbaz

### IX. Working Group Malignant Haematology:-

**Lead Person:** Maj. Gen. Tariq Mehmood Satti  
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Email: tariqsatti@yahoo.com

- Members:**
- Prof. Atifa Shoaib
  - Dr. Nadia Sajid
  - Col. Nighat Shahbaz
  - Dr. Muhammad Idrees
  - Dr. Irum Iqbal
  - Dr. Hayat-ul-Allah

### X. Working Group Molecular Haematology:-

**Lead Person:** Maj. Gen (R). Suhaib Ahmed  
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- **Members:**
  - Dr. Jaweria Ejaz
  - Dr. Bibi Kulsoom
  - Col. Muhammad Naeem
  - Dr. Saima Mansoor
  - Dr. Ayesha Khalid



## PSH Working Group

### XI. Working Group Immunophenotyping:-

**Lead Person: Dr. Asad Hayat**  
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• **Members:**

- Dr. Imran Nazeer
- Dr. Sajjad Ahmed
- Dr. Natasha Alvi
- Dr. Ayesha Imran

### XII. Working Group Appropriate and Sensible use of resources and Investigations:-

**Lead Person: Prof. Dr. Mona Aziz**  
Cell: 0333-4271736  
Email: monaakhlaq@hotmail.com

**Members:**

- Dr. Anum Wasim
- Dr. Ambreen Hamid
- Dr. Asma Sadia
- Dr. Sadia (LNH)
- Dr. Sarwar Khan

### XIII. Working Group Lab Accreditation standards:-

**Lead Person** Dr. Tariq Mehmood  
Cell: 0321-4642978  
Email: tariqm@skm.org.pk

• **Members:**

- Prof. Dr. Ayesha Juanid
- Col. Ghulam Rasul
- Dr. Rabia Ahmed
- Dr. Fouzia Tabassum

## SCHEDULE OF PSH MONTHLY MEETING

City	Coordinator Name	Date	Time
Lahore	Dr. Muneeza Junaid	2 <sup>nd</sup> Tuesday of the Month	09:00am to 10:00am
Karachi	Prof. Dr. Bushra Moiz	Last Friday of the Month	08:00am to 09:00am
Quetta	Prof. Nadeem Samad Shaikh	Last Friday of the Month	09:00am to 10:00am
Rawalpindi/ Islamabad	Brig. Ch. Altaf Hussain	Last Thursday of the month	03:00pm to 05:00pm
Peshawar	Prof. Dr. Shah Taj Khan	3 <sup>rd</sup> Thursday of the month	12:00pm to 01:00pm





## Highlights

### 13 FCPS Haematology Intensive Course, 2018

Department of Hematology and Transfusion Medicine  
The Children' Hospital & Institute of Child Health, Lahore.

FCPS is regarded as the highest degree achieved by the doctors in any specialty in Pakistan. Exams are always stressful for students. One way to manage exam stress is to do crash courses that help students in revising their course syllabus and give them a quick revision. The Department of Hematology and Transfusion Medicine at The Children's Hospital & Institute of Child Health, Lahore has taken the pride of arranging such intensive courses for the last 10 years with full devotion. Pakistan Society of Haematology and CPSP have been the key collaborating bodies. Above all the support of Haematology faculty and enthusiasm of students is evident throughout all such courses. The 13th FCPS Haematology Intensive course was arranged by the Department of Haematology and transfusion medicine, The CH & ICH, in collaboration with Department of Pathology, Shaukat Khanum Memorial Cancer Hospital & Research Center Lahore from 10th -13th October 2018. Residents-on-training from different institutes of the country participated in the course. Dr. Nazish Saqlain, Assistant Professor at The Children's Hospital & Institute of Child Health, Lahore coordinated and organized the course.



## Highlights

### 13 FCPS Haematology Intensive Course, 2018

It was a four-day event which included different sub-sections of Haematology. Day 1 was dedicated to Transfusion Medicine. The course was inaugurated on 10th Oct 2018 by Prof. Masood Sadiq, Dean, The Children's Hospital & Institute of Child Health, Lahore. Welcome address was delivered by Prof. Dr. Nisar Ahmed, Professor of Paediatric Haematology, BMT & Transfusion Medicine, The Children's Hospital and Institute of Child Health, Lahore in the presence of Guest of Honor, Prof. (retd) Dr. Abdul Hayee enlightening lectures were delivered by Brig. Maqbool Alam, Prof. Saba Jamal and Dr. Farheen Karim. It was followed by hands-on training for the residents.



Coagulation Medicine is considered to be an essential part of Haematology; day 2 consisted of interactive lectures and hands-on training regarding Coagulation. The speakers included Prof. Bushra Moiz and Prof. Samina Amanat who discussed interactive cases enabling participants to understand the diagnosis and management of bleeding and thrombotic disorders. Quality assurance (QA) and quality control (QC) is an integral part of diagnostic haematology. Quality assurance sessions included application of West guard rules, assessment of external quality control data and assessment of internal quality control data. After formal presentation on QA and QC by Dr Muhammad Shariq, Assistant Professor at Aga Khan University, participants were given practice questions. The session was well attended and appreciated by all the participants.





## Highlights

### 13 FCPS Haematology Intensive Course, 2018

On day 3 and 4, Morphology sessions took place. On 12th October 2018 lectures were delivered by Gen. Tariq Mehmood Satti, Gen. Suhaib Ahmed, Dr. Tariq Mehmood and Dr. Muneeza Junaid covering important topics of Hemolytic anemia, Haemoglobinopathies and Flowcytometry. Morphology practical session was carried out on 13th Oct 2018, at The Shaukat Khanum Memorial Cancer Hospital, Lahore which consisted of more than 25 important cases. The cases were later discussed by Prof. Tahir Sultan Shamsi, Prof. Shahtaj Masood, Dr. Tariq Mehmood, Dr. Asad Hayat, Dr. Ambreen Hamid, Dr. Ayesha Imran and Dr. Saima Farhan in an interactive session. At the end, Prof. Dr. Nisar Ahmed thanked the distinguished guests and distributed the certificates of participation to all attendees.







## Monthly Meeting

### PSH Monthly Meeting at KEMU, Lahore

The PSH monthly meeting was held in King Edward Medical University, Lahore, in Department of Pathology on Wednesday, 31st October, 2018.

1st case selected was presented by Dr. Sarah Farrukh (PGR-III), a 6-year old female presented in the hematology clinic with complaints of pallor since birth. CBC shows Hb-8g/dl, TLC-6 x 109/ L, Plt-225 x 109/ L. Patient is transfusion dependent since birth, usually requiring transfusion every month. Family history was significant two elder siblings having same complaints. Serum B12 and serum folate level are normal. HB-electrophoresis was also normal. Bone marrow aspirate shows markedly reduced erythroid precursors. Final diagnosis was ***Diamond Black Fan Anemia***.

2nd case selected was presented by Dr. Qurat-ul-Ain Ayaz (PGR-III), 22 years old male presented with complaints of pallor and yellow discoloration of sclera for 4-years. On general physical examination, tenderness on right hypochondrium was found. CBC shows Hb-6g/dl, TLC-9.6 x 109/ L, Platelet-422x 109/ L, MCV-65. 6fl and MCH-20.8pg. Serum ferritin shows 265ng/ml. ALT-182U/l, AST-108U/l, STB 4.6mg/dl, and Serum LDH-575. Provisional diagnosis of haemolytic anaemia was made which can be secondary to enzymopathy, membranopathies or hemoglobinopathies.

Coombs test was negative, G6pd levels were normal, no spherocytosis on examination of peripheral smear was found. Hb-Electrophoresis shows; HbA-48.5%, HbF-47.2% and HbA2-4.3%. Final diagnosis was Thalassemia Intermedia.





## Monthly Meeting

### PSH Monthly Meeting at SZH, Lahore

The PSH monthly meeting was held in Shaikh Zayed Hospital, Lahore, in Department of Pathology on Tuesday, 11th December, 2018.

Topic selected for discussion was a 16-year old female, who presented in the Hematology clinic with complaints of loose motions and fever on-and-off for last 3-months with nocturnal spikes. Fever was relieved temporarily by antibiotics and antipyretics. CBC shows Hb-10.1, TLC-1.9, Plt-144, MCV-96.6, and MCH-26.1. On peripheral smear, normocytic and normochromic blood picture, no MP was found (Malaria ICT negative). Typhidot and Dengue serology were negative. No lymphadenopathy or any organomegaly are found on CT Chest, abdomen and pelvic. Bone marrow aspirate shows frank megaloblastosis. On infectious screen, Brucella antibodies were found positive.

Final diagnosis was made malabsorption syndrome caused by bacterial infection (genus brucella).







## Monthly Meeting

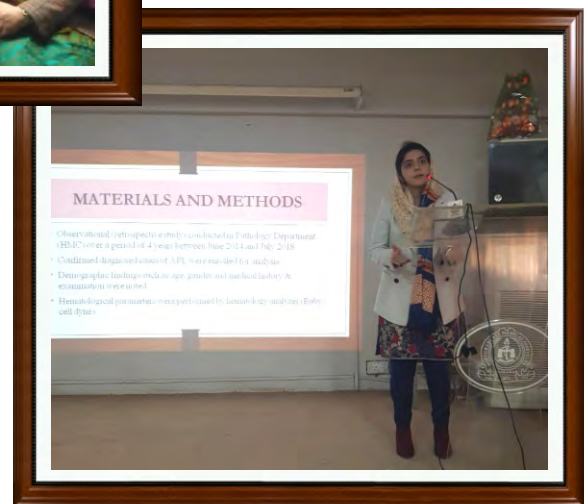
### PSH Monthly Meeting at Peshawar

Monthly PSH meeting Peshawar, was held on 24th of January'2019 at Pathology department, Hayatabad medical complex, Peshawar. It was attended by the senior consultants, fellows and post graduate trainees. Prominent among them were consultants from Khyber Teaching Hospital, Khyber Medical College, Lady Reading Hospital and North West General Hospital.

A case report by Dr Mehr Jabeen, was unique as a 60 years old known hemophiliac A, refractory to every hemostatic treatment was referred for further evaluation; investigations carried out showed time dependent inhibitors against factor VIII.

It was followed by a detailed presentation on Thrombophilias by Dr Khizar Abdullah. Questions asked by trainees were elaborately answered by the consultants and their experiences were shared. Different academic cases were also discussed in detail.

The session was concluded by Prof. Dr. Shahtaj Khan, with a vote of thanks to all the participants and guests followed by formal tea.







## SYMPOSIUM

### 10th PSH National Haematology Symposium, Mirpur

10<sup>th</sup> PSH National Haematology Symposium was conducted in Mirpur, the beautiful city of Azad Jammu & Kashmir on 2<sup>nd</sup> February, 2019 at Divisional Head Quarter Teaching Hospital, Mirpur AJK.

The first session began with recitation of verses from Holy Quran and Naat e Rasool Maqbool ﷺ. Dr. Zahida Qasim, Head of Pathology Department, Divisional HQ Hospital, Mirpur welcomed the participants. Dr. Farooq Ahmed Noor, MS of Divisional HQ Teaching Hospital, Mirpur was Chief Guest. The session was hosted by Dr. Ayesha Shoukat. The Chief Guest expressed his views and congratulated the Pathology Department of the Hospital, Head of Department Dr. Zahida Qasim and president of PSH, Prof. Dr. Nisar Ahmed for the Symposium that provided a platform for learning new updates and knowledge regarding Haematology.

The first presentation was on Genetic counseling & Antenatal Diagnosis of Thalassemia by Gen.(R) Suhaib Ahmed, CEO, Genetic Resource Center, Rawalpindi. After this informative lecture, the second presentation was on Pancytopenia: Evaluation & Management by Gen.(R) Parvez Ahmed from Quaid-e-Azam International Hospital, Islamabad. After this brain storming lecture Prof. Lubna Naseem Consultant Haematologist PIMS, Islamabad expressed her views on Rational use of Blood and Blood Products. Brig. Qamar-un-Nisa, Consultant Haematologist AFBTC/NIBMT, Rawalpindi gave very useful information on Bone marrow transplantation indications and procedure.

The next distinguished speaker was Dr. Nadeem Ikram Associate Professor RMC, Peshawar. He shared his knowledge on thalassemia management and its challenges. Dr. Saima Farhan Assistant Professor of Haematology, The Children's Hospital & ICH, Lahore gave a talk on Rare Bleeding Disorders. The last presentation of the session was on An Approach to Bleeding Disorders by Dr. Shazia Yaseen Assistant Professor of Paediatric Haematology from the Children's Hospital & Institute of Child Health, Lahore

The souvenirs & Shields were presented to Speakers and organizing committee and the symposium was concluded by vote of thanks by Dr. Zahida Qasim, Head of Pathology Department, Divisional Head Quarter Teaching Hospital, Mirpur and specialized Health Secretary Mirpur.

After lunch the organizing committee took all the Speaker and Participants for visit to Mangla Fort, Mangla Museum and Army Water Sports Club. All participants enjoyed this best trip. It was anticipated that symposium like this one should be organized in future as well to upgrade Haematology in best ways.





**SYMPOSIUM**

**10th PSH National Haematology Symposium, Mirpur**







## Case Report

### CO-INHERITANCE OF HAEMOGLOBIN D WITH $\beta$ THALASSAEMIA MUTATION

Dr. Saima Mansoor Bugvi, Prof. Nisar Ahmed

Department of Haematology, Transfusion Medicine and Bone Marrow Transplant,  
The Children's Hospital & Institute of Child Health, Lahore

#### ABSTRACT

Genetic disorders of haemoglobin are quite prevalent in Pakistan due to traditional practices of consanguineous marriages. Thalassaemia is the commonest genetic haemoglobin disorder in Pakistan. Beta thalassaemia has a carrier rate of 5-7%. It is estimated that each year around 5000 new births of beta thalassaemia major takes place. It is also estimated that there will be over 50,000 children suffering from thalassaemia major in the coming years in Pakistan.<sup>1</sup> Hb-D is mostly confined to Punjab. Haemoglobin D can be inherited as a homozygous or a heterozygous trait with other haemoglobinopathies. Though haemoglobin D-Punjab is commonly seen, a heterozygous trait with beta thalassaemia is a very rare presentation. Here, we present a rare case of co-inheritance of haemoglobin D-Punjab and beta thalassaemia in a 06-year-old female of Pakistani origin. She came to us with pallor and weakness noted by her parents for the past three months. No history of similar complaints in the past. On examination, she was pale and with mild splenic enlargement. Her height and weight were below her normal age. On investigation, there was moderate anaemia, peripheral blood film showed marked anisopoikilocytosis. Haemoglobin electrophoresis showed co-inheritance of haemoglobin D-Punjab and beta thalassaemia. She was given hydroxyurea and folic acid supplements. Parents were counseled about her disease and advised regular follow-up. Haemoglobin electrophoresis of her parents and siblings were done which confirmed carrier state of beta thalassaemia in the mother and Presence of haemoglobin D in the heterozygous state in the father. Molecular testing of the family is also done.

**Keywords:** Anaemia, Genetic disorders of haemoglobin, haemoglobin D, consanguinity

#### CASE REPORT

A 6 years old girl resident of Lahore presented at children hospital and institute of child health with complaints of Pallor and Failure to thrive since three months noted by her parents. No other active complaints. She is a product of consanguineous marriage. There is a history of thalassaemia in one of her maternal uncle. On general physical examination Pallor was positive. There was no jaundice and Koilonychia. Lymph nodes were not palpable. On systemic examination, spleen was palpable just below left costal margin. All other systemic examinations were unremarkable. Laboratory investigations revealed Haemoglobin 7.2 g/dl, Red Blood Cell (RBC) count  $4.33 \times 10^{12}/l$ , White Blood Cell (WBC) counts  $7.2 \times 10^9/l$ , platelet count  $629 \times 10^9/l$ , Mean Corpuscular Volume (MCV) 54.5fl Mean Corpuscular Haemoglobin (MCH) 16.6pg, Mean Corpuscular Haemoglobin Concentration (MCHC) 30g/dl, Reticulocyte count was 5.5% (0.5-2.5%). Absolute reticulocyte count was  $252 \times 10^9/l$  ( $50-100 \times 10^9/l$ ). Peripheral blood film showed marked anisopoikilocytosis with prominent target cells, schistocytes and microspherocytes. There is macrocytosis and polychromasia. LDH and serum ferritin levels were normal. Direct and indirect Coomb's tests were negative ruling out any autoimmune causes.





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Sickling test was negative, thereby ruling out sickle cell disease which is quite prevalent. After clinical examination and routine investigations, some sort of haemoglobinopathy was considered as one of the differential diagnosis as it is very common but underdiagnosed in this area. Hb electrophoresis was performed to rule out haemoglobinopathies.

#### Haemoglobin electrophoresis of index case

Hb-A2	6.70%
Hb-F	5.60%
Hb-D	80.70%

Findings of Haemoglobin electrophoresis are suggestive of compound heterozygosity of HbD / beta thalassaemia. The child is advised tablet hydroxyurea 500mg once a day, tab Folic acid once daily. Monthly Follow up is advised and child monitored for anaemia and drug complication.

Extended family screening is proceeded. Complete blood counts along with haemoglobin electrophoresis of the parents were carried out which confirmed thalassaemia trait in the mother and haemoglobin D trait in the father. The proband has two siblings, both are sisters. One sister was found heterozygous for HbD the other sister has normal HPLC. Genetic Counseling and mutational analysis was also done and future prospects for further children discussed.

#### Haemoglobin electrophoresis of father

HbA	50.2%
HbA2:	1.1%
HbD:	48.7%

#### Haemoglobin electrophoresis of mother

Hb-A2:	5.1%
Hb-A:	94.9%

#### DISCUSSION

Haemoglobin protein is formed from two pairs of globin chains each with a haem group attached. Four chains are associated in the form of tetramer. Hemoglobin A, the major haemoglobin consists of 2Alpha and 2beta chains, found in adults and children. Haemoglobin A2 (2-3.3%) consists of 2Alpha and 2delta chains. Haemoglobin F (0.2-1) % consists of 2Alpha and 2 gamma chains found in small quantities in adult life.<sup>2</sup> Mutations in genes encoding haemoglobin chains are present in about 7% of worldwide population. Mutations can cause qualitative, defects and quantitative defects in haemoglobin protein. Qualitative defects alter the amino acid sequence of the



## Case Report

### CO-INHERITANCE OF HAEMOGLOBIN D WITH $\beta$ THALASSAEMIA MUTATION

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protein resulting in structural variants of haemoglobin, such as Hb D, S, and G. In quantitative defects there is reduction in the output from that gene, such as thalassaemias<sup>3</sup>. An individual can also have a combination of two or more of these abnormalities. Haemoglobin D results from point mutation in the beta-globin gene (HOB) in the base of the 121 codon(GAA→CAA) with the substitution of glutamine for glutamic acid (Glu>Gln)<sup>4</sup> Out of 16 sub-types of Haemoglobin D, Hb-D Punjab or Los-Angeles is most common with 0.86% in Indo-Pakistan continent and 3.6% alone in Punjab<sup>4,5</sup>. Haemoglobin D can occur in heterozygous state, homozygous state and in compound heterozygous state. In heterozygous condition it presents no clinical or hematological alterations, and Hemoglobin electrophoresis will show bands of Hb-A and Hb-D while HbA2 will be normal. The Homozygous form Hb DD is also clinically silent<sup>6</sup>.

Haemoglobin electrophoresis of homozygous state will show single band in the region of HbS/D, there will be no Hb A and again HbA2 will be normal. In Compound heterozygotes Hb D is co inherited with either another abnormal haemoglobin and/or along with alpha or beta thalassaemia. There are only a few case reports available on co-inheritance of haemoglobin D with alpha or beta mutations. Individuals with compound heterozygosity for Hb-D beta thalassaemia can have mild to moderate disease. Patients may manifest moderately severe anaemia with splenomegaly and chronic haemolysis while others may show very mild disease. In a review of case series, three compound heterozygous individuals were followed only one case has clinical symptoms. The other two were found in routine prenatal screening, this demonstrates the variability in the clinical picture of coinheritance of haemoglobin D with beta thalassaemia<sup>7</sup>. In our case the child presented earlier at the 06 years of age with obvious pallor. She was growth retarded and her height and weight were below her normal age. We separated all haemoglobin molecules and variants; we found very high percentages of haemoglobin D-Punjab and co-inheritance of beta thalassaemia trait with levels of HbA2 of 6.7%. Lastly, molecular studies as they are recommended as a final investigation were also done which revealed frameshift 8-9 mutation and haemoglobin D the affected child. These mutations were traced back and were found in her parents in the heterozygous state.

### CONCLUSION

The coinheritance of HbD-Punjab with  $\beta$ -thalassaemia can be found in Pakistani population. Therefore, a better understanding of HPLC chromatogram and electrophoretogram patterns and clinical features of this combination is useful for genetic counseling, prevention, and control programs for thalassaemia and hemoglobinopathy.

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## Case Report

### OUTCOME OF TOTAL SPLENECTOMY IN A 6 YEAR OLD BOY WITH HEREDITARY SPHEROCYTOSIS

**Dr. Javaria Fatima, Dr. Shazia Yaseen, Prof. Dr. Nisar Ahmed,**

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#### ABSTRACT:

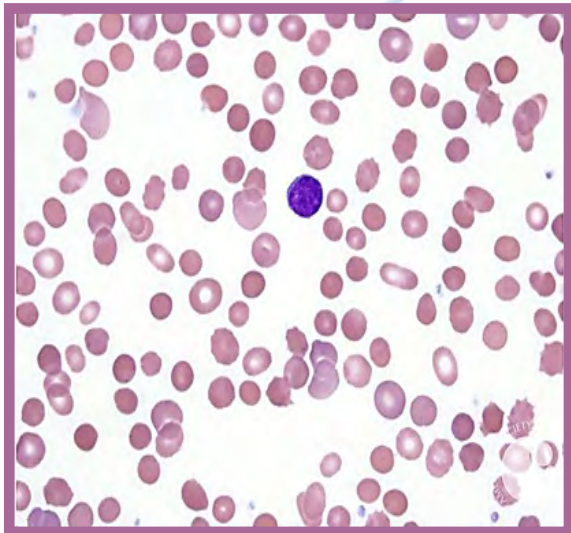
Hereditary spherocytosis (HS) is the most common of hereditary hemolytic anemias due to membrane dysfunction. The disease severity is highly variable with mild, moderate and severe forms. Palpable splenomegaly is present in about 50-60% of patients in paediatric population. Splenectomy significantly reduces the severity of anemia by increasing lifespan of spherocytic red cells.

#### INTRODUCTION:

HS is a common, inherited hemolytic anemia that occurs in all racial groups but is particularly common in Caucasians. The underlying primary molecular lesion is heterogeneous; it may involve several erythrocyte membrane proteins with ankyrin being the most commonly affected followed by spectrin, protein band 3 and more rarely protein 4.2. Red cells of patients with HS have a decreased surface to volume ratio leading to their trapping and destruction during their passage through the splenic cords. Therefore, surgical total splenectomy by removing the main site of red cell destruction lengthens the red cell lifespan.

#### CASE REPORT:

Patient named ABC, 6 years old boy, resident of Jhang, referred to Haematology department of Children Hospital Lahore on 15th March 2017. He presented with complaints of pallor, jaundice and abdominal distension for the last 3 years. The family history was positive for jaundice in father 10 years back. On examination, he was pale and jaundiced with massive splenomegaly and no lymphadenopathy. CBC counts revealed Hb of 7.1g/dl, WBC count of 3.5, and platelets 90,000. RBC indices showed MCV of 78fl, MCH 29pg and MCHC of 36g/dl. Differential leucocyte count was neutrophils 50%, lymphocytes 45%, monocytes 03% and eosinophils 02%.



No blasts or atypical cells were seen. Peripheral smear examination revealed anisopoikilocytosis, hypochromia, microcytosis, spherocytes and polychromasia. Corrected Reticulocyte count was 07%. Coombs test was performed and it was negative. Osmotic fragility was done which turned out to be positive on two separate occasions. Patient history, family history and lab findings supported the diagnosis of HS. Patient was started on folic acid 5mg OD. On follow-up, patient presented with abdominal fullness, weakness and repeated chest infections. He was on regular transfusions. On examination, spleen was 6cm below costal margins and also confirmed on abdominal ultrasound. Cytopenias were evident on CBC. A diagnosis of hypersplenism was made and splenectomy was planned and done on 30th April 2018, after immunization. Patient was put on prophylactic antibiotics after splenectomy and is on regular follow-up. His CBC counts have improved significantly, he, no longer, requires regular transfusions and is doing well.





## Case Report

### **OUTCOME OF TOTAL SPLENECTOMY IN A 6 YEAR OLD BOY WITH HEREDITARY SPHEROCYTOSIS**

**Dr. Javaria Fatima, Dr. Shazia Yaseen, Prof. Dr. Nisar Ahmed,**

#### DISCUSSION:

In paediatric patients, hemolytic anemia is commonly associated with hemoglobinopathies, erythroenzymopathies or red cell membrane disorders like hereditary spherocytosis. HS has been reported in all ethnic groups. The diagnosis of HS is generally straightforward, based on a combination of clinical examination, family history and on laboratory investigations. The median age at diagnosis is about 5 years. About 75% of cases have a positive family history. Regarding the clinical features, patients have varying degrees of anemia and jaundice and most children have mild to moderate enlargement of spleen.

Splenectomy is indicated in HS to relieve symptoms due to anemia or splenomegaly, reverse growth failure or skeletal changes due to over robust erythropoiesis and prevent recurrent gallstones. A lifelong risk of bacterial infection has been recognized for many years as a concomitant cost of splenectomy. Recently, it has been demonstrated that splenectomy also confers a significant risk of delayed vascular events in patients with HS. Still, in patients with usual dominantly inherited spherocytosis, removal of spleen abates the hemolytic process and corrects the anemia. When considering splenectomy for HS in a pre pubertal boy, it is to be remembered that a significant rise in Hb will occur as the boy passes through puberty.

Hence, in HS patients with symptomatic anemia, lagging growth or troublesome left upper quadrant pain will derive impressive subjective and objective benefit from splenectomy. The costs will be a permanent increased risk of serious infection, a large increase in the risk of delayed adverse vascular events and loss of the vascular benefits of mild anemia. Patients with moderate anemia, especially children, should undergo careful observation and evaluation before splenectomy is advised.

#### **Data of splenectomies for haematological disorders in Children Hospital Lahore in the last ten years (2008-2018)**

Haematological disorders	Time period in years									
	2009	2010	2011	2012	2013	2014	2015	2016	2017	2018
Thalassemia major	02	03	01	03	02	01	-	04	-	02
Storage disorder	-	-	01	-	02	01	-	02	01	01
Hereditary spherocytosis	-	-	-	01	01	-	-	-	01	01

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## Haematology

Five things physicians and patients should question

by

Canadian Hematology Society



- 1 Don't give IVIG as first line treatment for patients with asymptomatic immune thrombocytopenia (ITP).**

Treatment for ITP is recommended for a platelet count less than  $30 \times 10^9/l$ . corticosteroids are considered first-line treatment, with the addition of IVIG reserved for severe ITP and bleeding. When a rapid rise in platelets is required, or when corticosteroids are contraindicated. There is no evidence of benefit of IVIG in combination with corticosteroids for first-line treatment of asymptomatic ITP. Unnecessary IVIG infusions can result in multiple adverse effects, including acute hemolytic or anaphylactic reactions, infections, thromboembolic events, and aseptic meningitis.
- 2 During interruption of warfarin anticoagulation for procedures, don't bridge with full-dose low molecular weight heparin (LMWH) or unfractionated heparin (UFH) unless the risk of thrombosis is high.**

Patients on warfarin with a low-risk for thrombotic events do not require bridging anticoagulation. If interruption is necessary warfarin can be stopped 5 days prior to a planned procedure and resumed when it is felt to be safe to do afterwards. Bridging with LMWH or UFH has been shown to cause excess bleeding when compared with no bridging and may ultimately delay resumption of warfarin. High-risk patients (e.g. mechanical mitral valve, venous thromboembolism within the last 3 months or atrial fibrillation with recent stroke/TIA) should be considered for bridging if the risk of thrombosis is higher than the risk of peri-procedural bleeding.
- 3 Don't order thrombophilia testing in women with early pregnancy loss.**

Early pregnancy losses are common amongst healthy women. Current guidelines do not support the routine screening of women with pregnancy loss for inherited thrombophilias. Moreover, there are recommendations against instituting thromboprophylaxis in women with inherited thrombophilia's wishing to achieve a successful term pregnancy. By performing testing for inherited thrombophilia's, patients may be unnecessarily exposed to the harms of thromboprophylaxis, inappropriately labeled with a disease-state, and may unnecessarily modify future plans for travel, pregnancy or surgery based on detection of an "asymptomatic" thrombophilia. Further, patients with negative testing may receive false reassurance.
- 4 Don't request a fine-needle aspirate (FNA) for the evaluation of suspected lymphoma.**

The diagnosis of lymphoma requires specimens with intact cellular architecture for accurate histopathologic and immunophenotypic classification. FNA is associated with a low sensitivity and potentially results in delays in lymphoma diagnosis. Although excisional biopsy is the gold standard for lymphoma diagnosis, depending on the lymph node location, excisional biopsy may be associated with complications and the need for general anesthesia. At a minimum, an imaging-guided core biopsy should be obtained to improve the accuracy and timeliness of lymphoma diagnosis.
- 5 Don't transfuse patients based solely on an arbitrary hemoglobin threshold.**

Decisions to transfuse should be based on assessment of an individual patient including their underlying cause of anemia. There is high quality evidence that demonstrates a lack of benefit and in some cases harm to patients transfused to achieve an arbitrary transfusion threshold. If necessary, transfuse only the minimum number of units required instead of a liberal transfusion strategy. Risks of red blood cells transfusions include allergy, fever, infections, volume overload and hemolysis.





## QUALITY MANUAL FOR LABORATORY

### Introduction

In order to protect the welfare of patients and laboratory personnel and to ensure that precise results are obtained and reported, the laboratory needs to maintain quality throughout its pre-analytical, analytical, and post-analytical path of workflow. A Quality Management Plan provides the infrastructure for maintaining quality in the laboratory.

The Quality Management proposition included in these guidelines is based on the 12 Quality System Essentials (QSE) model developed by CLSI and is mostly consistent with College of American Pathologist (CAP) accreditation and ISO 15189. The 12 QSEs are elemental for maintaining standard, safety, and efficiency throughout the laboratory's path of workflow.

### Description of the 12 QSEs:

**QSE 1 – Infrastructure:** Dedication from high-level managers is essential for the success of the overall laboratory quality program. This QSE describes how the regulatory structure and quality management system ensure that consumer's needs and regulatory requirements are met.

**QSE 2 – Acceptability and customer satisfaction:** This QSE describes how the laboratory evaluates and addresses its ability to meet the needs customers both internal and external.

**QSE 3 – System of safety of personnel:** This QSE describes how the expanse of the laboratory is maintained in a manner that guarantees systematic workflow, accurate test results, and personnel safety. It also describes how the laboratory supplies the materials and tutelage necessary to ensure the safety of all personnel.

**QSE 4 – Technical staff:** The most essential part of the laboratory's assets is its personnel. This QSE describes how personnel are organized and provided with the utensils necessary to perform testing so that reliable and accurate test results are acquired.

**QSE 5 – Purchasing & Inventory:** Availability of dependable and reliable test kits and stock is vital. This QSE describes how reagents and supplies are obtained, dispensed, and managed.

**QSE 6 – Equipment evaluation:** This QSE describes how equipment is chosen, stationed, identified, validated/verified, and maintained. This is done in order to ensure personnel safety and testing result rectitude.

**QSE 7 – Procedure workflow:** This QSE describes the tasks and techniques that are performed to ensure that the testing procedures are correctly carried out, the environment is appropriate for accurate testing, and the testing methods work as presumed to produce accurate and reliable results.

**QSE 8 - Documents and Records:** This QSE describes the processes for creating systematized documents and records and maintaining documents and records so that they are current and accurate, easily available to laboratory staff, and protected from destruction, decay, and unauthorized use.

**QSE 9 – Information Management:** This QSE describes how patient-related and testing information is managed in order to maintain the precision, reliability, privacy, and accessibility of the data.

**QSE 10 – Non Confirming Essential (NCE):** This QSE describes how nonconforming events (occurrences) are noted, looked into, resolved, and followed.

**QSE 11 – Assessment:** This QSE describes the internal and external assessments that are coordinated to assess the productiveness of the laboratory's quality management system.

**QSE 12 – Continuous Improvement:** This QSE describes elements of a process improvement program that recognizes and addresses chances for advancement and problems that impact patient care.





## **1. Infrastructure**

### **1.1 Vision & Mission**

The laboratory follows all external requirements and has an organizational structure that guarantees quality and meets consumer needs.

### **1.2 Objective**

This policy provides the effective demonstration of the laboratory's structure and quality management system as it is synchronized with other facility quality management programs.

### **1.3 Process Management**

The Laboratory Director is charged with the allotment of resources so that shrewd plans can be implemented.

The Laboratory manager is responsible for maintaining the organizational chart.

The Quality Manager is responsible for developing the laboratory quality management system and coordinating it with other facility quality management programs.

### **1.4 Quality Management System**

The laboratory quality management system will be established and connected with other facility quality management systems.

### **1.5 External QA**

A chart recognizing all Referral Laboratories is maintained.

### **1.6 Documentation of Quality Management**

The following procedures support this policy:

Executing a Laboratory Quality Management System

Organizational Chart Development and Maintenance

Recognition of Referral Laboratories

## **2. Acceptability & Customer Satisfaction**

### **2.1 Vision & Mission**

The laboratory has processes and measures for both internal and external customer service.

### **2.2 Objective**

This policy provides direction for the processes and procedures to effectively manage the laboratory's customer service.

### **2.3 Responsibility**

The Laboratory Director is responsible for the identification of the buyers.

The Laboratory Manager is at the helm for providing services and feedback mechanisms to the customer.

The Quality Manager is in charge of monitoring customer service and contentment.

The supervisors are take care to identify problems and provide solutions.



## **2.4 Services provided by the laboratory**

The laboratory has processes and procedures for existing and ongoing reviews of contracts to render its medical laboratory services to other services or facilities.

## **2.5 Customer demands and their identification**

The laboratory has identified internal and external customers, their demands and expectations and originates processes in order to meet these needs.

## **2.6 Mechanisms for customer feedback**

The laboratory has systems for determining customer satisfaction and managing grievances in order to recognize processes that are causing difficulties for the customers and initiate responses.

## **2.7 Referral for Process Improvement**

Information from customer complaints and satisfaction surveys are analyzed, problems are assessed and referred for process improvement.

## **2.8 Documentation**

The following procedures support this policy:

- i. Allocation of Laboratory Services (defined Turn-Around Times-TAT)
- ii. Recognizing and Managing Customer Needs
- iii. Managing Customer grievances
- iv. Monitoring Customer Contentment

## **3. Facility and Safety**

### **3.1 Vision & mission**

The laboratory facilities are planned, refurbished, used, and maintained to meet all essential requirements for well-being, productivity, and ergonomics. Laboratory practice make possible the safety of all employees and visitors.

### **3.2 Objective**

This policy provides direction for the processes and procedures to effectively manage the laboratory's facilities and safety practices.

### **3.3 Process Management**

The Laboratory Director is in charge of working with the parent organization to obtain excellent facilities and and is also responsible for laboratory safety.

The Laboratory Manager is responsible for conveying the needs of the laboratory through workflow analysis and for providing a secure workplace for all laboratory personnel.

The Laboratory Safety Officer is responsible for providing instructions to laboratory management and staff regarding safety matters and responsibilities.

The laboratory supervisors are responsible for training and compliance of staff to safety policies and general maintenance of the facility.





### **3.4 Allocation of facility space, design and renovation**

The laboratory work areas are planned such that the workload can be executed without compromising the quality of work or the well being of personnel or patients.

### **3.5 Safe working environment**

The laboratory provides a secure working environment and opportunities for each employee to follow the safety requirements by providing instructions and appropriate personal protective and other safety equipment.

### **3.6 Facility Maintenance**

The facility is frequently inspected and maintained as a secure and comfortable workspace.

### **3.7 Documentation**

The following procedures support this policy:

- i. Laboratory design
- ii. Security procedures, records, and audits
- iii. Safety training
- iv. Safety Equipment (PPEs) availability and Use

## **4. Personnel**

### **4.1 Vision & mission**

The laboratory collaborates with the Human Resources Department to recruit qualified personnel and ensure that they have the knowledge and can exhibit skills necessary to perform their duties.

### **4.2 Objective**

This policy provides direction for the processes and procedures to effectively manage the laboratory's staff.

### **4.3 Process management**

The Human Resources Department is responsible for the employment advertising, applicant screening, employee record maintenance, and Facility Orientation of new employees.

The Laboratory Director is in charge for recruiting, for selection process and performance review of all personnel reporting to this position.

The Laboratory Manager is responsible for interviews, hiring selection process, facility orientation, and performance reviews of all staff reporting to this position.

The laboratory supervisor have the duty for job-specific tutelage and competency, work period scheduling, and contributing to the performance reviews of staff reporting to this position.

The Quality Manager is at the helm for assisting with training of new employees.

### **4.4 Job particulars**

Job qualifications are specified in job descriptions for each laboratory position.

### **4.5 Personnel Qualifications**

Certification and education records are maintained for all members of the laboratory administration and staff.





#### **4.6 Training and orientation**

Employees are oriented to the organization and instructed in each duty as designated.

#### **4.7 Staff competency**

Employees are evaluated for expertise twice the first year and then annually in their job duties as assigned.

#### **4.8 Continuing Education**

Opportunities are provided for continuing education and professional development. These activities are documented by the employees.

#### **4.9 Documentation**

The following procedures support this policy:

- i. Maintaining Adequate Staff Resources
- ii. Job Descriptions
- iii. Personnel Qualifications
- iv. Employee Orientation and Training
- v. Evaluating Employee Competency
- vi. Continuing Education

### **5. Purchasing and Inventory**

#### **5.1 Vision & mission**

The laboratory buys and maintains an archive of equipment, supplies, and reagents used in the path of the laboratory workflow.

#### **5.2 Objective**

This policy provides direction for the processes and procedures to effectively work with the Purchasing Department to manage purchase and inventory processes.

#### **5.3 Process management**

The Laboratory Director assesses and authorizes contract of services or facilities.

The Purchasing Department is responsible for order processes, vendor payment, and delivery & storage equipment's.

The Laboratory Manager is responsible for Manufacturer recognition and selection and order processes.

The Laboratory Supervisors are responsible for inventory maintenance including collecting, storage and auditing of inventory.

#### **5.4 Vendor Selection**

The laboratory has a system for selecting vendors of equipment, supplies, and services.

#### **5.5 Purchasing Supplies and Reagents**

The laboratory works with the Purchasing Department to purchase supplies and reagents.

#### **5.6 Receiving, Inspecting, Storing, and Managing Supplies & Reagents.**

The laboratory has processes and procedures for collecting, scrutinizing, storing, and managing the inventory of supplies and reagents used in the path of workflow.



## **5.7 Documentation**

The following procedures support this policy:

- i. Identifying and Selecting Vendors
- ii. Purchasing Supplies and Reagents
- iii. Handling Supplies and Reagents

## **6. Equipment**

### **6.1 Vision & mission**

The laboratory keeps comprehensive documentation on each piece of equipment that is necessary in the processes across the laboratory's path of workflow.

### **6.2 Objective**

This policy provides direction for the processes and procedures to effectively manage the laboratory's equipment.

### **6.3 Process management**

The Laboratory Director is responsible for laboratory purchase decisions, executing an instrument verification and maintenance program, and documentation review.

The Purchasing Department is responsible for the actual purchasing process associated with instrument purchases.

The Laboratory Manager is in charge for instrument justification and comparative analyses.

The Laboratory Supervisors and Quality Manager are responsible for the validation, maintenance and mending once the instruments are on-site.

### **6.4 Complete equipment evaluation**

The laboratory buys and installs the equipment needed for producing quality results. A record of all laboratory equipment is maintained.

### **6.5 Lab instrumentation validity**

Validation studies are conducted and documented as required on laboratory instrumentation to include accuracy, precision, linearity, reportable reference ranges, sensitivity (as needed) and specificity (as needed).

### **6.6 Carryover**

Studies are executed to show instruments do not have any carryover.

### **6.7 Quality assessment of instrumentation**

Each instrument in use has a separate manual which includes all instructions and documentation generated during the life of the instrument and 2 years post retirement.

### **6.8 Discarding irreparable instruments**

The laboratory retires instruments that are no longer in use and has procedures for storage of these instruments and their records.





## **6.9 Documentation record**

The following procedures support this policy:

- i. Equipment Acquisition, Installation, Identification, and Inventory
- ii. Instrument Validation Studies
- iii. Method Comparisons
- iv. Carryover Studies
- v. Instrument and Equipment Preventative/Corrective Maintenance
- vi. Troubleshooting and Corrective Actions
- vii. Retiring Instruments

## **7. Procedure workflow**

### **7.1 Vision & mission**

The laboratory records and approves all processes in the pre-analytic, analytic, and post-analytic activities path of workflow.

### **7.2 Objective**

This policy provides direction for the processes and procedures necessary to make certain that testing procedures are accurately performed, the environment is suitable for reliable testing, testing methods work as expected to yield accurate results, and governmental and other regulations are met. It also provides regulations to effectively manage the laboratory's processes such that all impacts of the changes on customers and other processes are considered and dealt with accordingly.

### **7.3 Process control**

The Laboratory Director is in charge for ensuring process control is monitored for customer satisfaction.

The Laboratory Manager and supervisor are responsible for the development and validation of pre-analytical, analytical and post-analytical procedures.

The Laboratory Supervisors and Quality Manager are responsible for documenting test performance procedures and monitoring internal quality control.

### **7.4 System validity**

The laboratory records and endorses all processes in the path of workflow prior to implementation.

### **7.5 Sample handling**

Specimen handling processes in pre-analytic, analytic, and post-analytic activities are planned and endorsed to ensure they work as intended.

### **7.6 Procedure management**

All pre-analytic, analytic, and post-analytic activities utilize methods that have been authorized and have established and verified reportable ranges and reference intervals.

### **7.7 Internal Quality Control**

Process and statistical measurements are made to identify random and systematic causes of process variations.





## **7.8 Documentation record**

The following procedures support this policy:

- i. Process Identification and Validation
- ii. Specimen Management
- iii. Method Verification/Validation
- iv. Internal and External Quality Control

## **8. Documents and Records – Policy**

### **8.1 Vision & vision**

The laboratory has a process for recording the management's instructions to staff about the laboratory's work, what to do, how to do it, and how to document what happened when work was done.

### **8.2 Objective**

This policy provides direction for the processes and procedures to effectively manage the laboratory's documents and records.

### **8.3 Process management**

The Laboratory Director is responsible for reviewing/endorsing all documents before implementation and on biennial basis.

The Laboratory and Quality Managers are in charge for establishing and maintaining the document control system.

The Laboratory Supervisor are responsible for authoring, amending, dispensing, and retaining documents, forms, and records as needed.

Laboratory Staff are accountable for following document control policies and procedures as necessary to complete their work.

### **8.4 Documents**

The laboratory has a document control system to make certain that all documents in use are written in the approved format, reflect the current version, and are assessed and approved by the laboratory director at least biennial.

### **8.5 Procedures**

All procedures used in the laboratory will be recorded, appraised, and signed by the Laboratory Director or designee prior to implementation and at least biennial thereafter.

### **8.6 Records**

Records are created, amended, archived, and destroyed in a way that enables retrieval, prevents damage and unauthorized use, and maintains patient confidentiality.

### **8.7 Storage**

All laboratory records, inclusive of requisitions, patient results, and QC, QA, and maintenance logs, will be retained for at least three years.

### **8.8 Documentation**

The following procedure supports this policy:

- i. New Document design, Review, and Approval
- ii. Revision of Approved Existing Documents
- iii. Document Control
- iv. Records Review, Retention, Storage, Retrieval, and Destruction
- v. Records Modification



## **9. Information Management**

### **9.1 Vision & mission**

The laboratory controls how patient and laboratory information is received, reviewed, conveyed, and stored in both paper-based and electronic information systems.

### **9.2 Objective**

This policy provides direction for the processes and procedures to effectively manage laboratory-generated information.

### **9.3 Process management**

The Laboratory Director is in charge of making turnaround time decisions.

The Laboratory Manager is responsible for adherence to patient confidentiality and assessment of laboratory results.

The Laboratory Supervisor and Quality Manager are responsible for evaluation of laboratory results.

Information Technology department is responsible for technological processes in the management of electronic information.

### **9.4 Patient Confidentiality**

Processes are designed to ensure that patient information is kept exclusive and confidential.

### **9.5 Usage & accessibility of electronic information**

Electronic data is retrievable only to authorized personnel

### **9.6 Reporting of Results**

Results are reported accurately and within established turnaround times.

### **9.7 Results Modification**

Any results changed or modified are documented to show both the original and modified results, reason for change, name of person notified, and date and time of notification.

### **9.8 Reporting Delays**

Hold ups in testing or reporting results are relayed to important persons.

### **9.9 Result Reporting Changes**

Modification in test methodology or reference ranges are communicated to the ordering physicians.

### **9.10 Data Integrity and Storage**

Data is saved and used in a manner that maintains the integrity of electronic and paper-based data.

### **9.11 Documentation**

The following procedures support this policy:

- i. Patient Confidentiality
- ii. Accessing and Using Electronic Information
- iii. Reporting of Results
- iv. Results Modification
- v. Reporting Delays
- vi. Data Storage and Maintaining Data Integrity





## **10. Non Confirming Essential (NCE)**

### **10.1 Vision & mission**

The laboratory recognizes, registers, and investigates occurrences (non-conformances); classifies, analyzes, and trends the information they represent; performs remedial/corrective actions; and identifies the need for root cause analysis and process improvement.

### **10.2 Objective**

This policy provides direction for the processes and procedures to competently discern and resolve problems and to classify problems so that reparative actions aimed at removing root causes and enhancing processes can be planned and implemented.

### **10.3 Process management**

The Laboratory Director is responsible for reviewing the events and complaint resolution reports.

The Laboratory and Quality Managers are responsible for assembling and analyzing occurrence data.

The Laboratory Supervisor is responsible for problem inquiries and resolution. The Laboratory Supervisors are responsible for documentation of complaints and problem resolution.

### **10.4 Identifying Occurrences**

The laboratory has means to pinpoint, document, investigate, and respond to complaints from internal/external customers; recalls of materials, equipment, or software; and other nonconforming events.

### **10.5 Investigation and Response to Occurrences**

The laboratory has a procedure for recognizing, recording, and investigating occurrences and performing restorative and corrective actions in response to those nonconforming events.

### **10.6 Classifying and Analyzing Occurrence Information**

The laboratory has a procedure for classifying and analyzing occurrences, including trending information, so that the portions of the path of workflow with the most important patient-related and expensive problems can be recognized, amended, and referred for root cause analysis and process improvement.

### **10.7 Documentation**

The following procedures support this policy:

- i. Identifying and Documenting Occurrences
- ii. Remedial Actions and Investigation of Occurrences
- iii. Analyzing Occurrence Information and Referring for Root Cause Analysis and Process Improvement

## **11. Assessments**

### **11.1 Vision & mission**

The laboratory will undergo internal and external evaluations to determine the efficacy of the laboratory's quality management system.

### **11.2 Objective**

This policy provides direction for the processes and procedures to effectively manage Quality Assessment of the laboratory.





### **11.3 Process management**

The Laboratory Director is in charge for reviewing the Quality Assessment Report.

The Quality Manager is responsible for developing and administering a Quality Assessment Program.

The Laboratory Manager and Supervisor are responsible for carrying out the activities of the Quality Assessment Program.

### **11.4 Internal Quality Indicator Surveillance**

The laboratory has procedures to recognize the path of workflow and choose and observe Key Performance Indicators (KPI) that measure the performance of processes.

### **11.5 Internal Audits**

Internal process audits are performed for both Quality System Essentials and operations on a predetermined schedule.

### **11.6 External Quality Assurance**

Reviews provided by external organizations determine the laboratory's performance based on regulatory, accreditation, and predetermined values.

### **11.7 External Benchmark**

Laboratory performance is compared to self and others for ongoing improvement goals.

### **11.8 Quality Assessment Report**

A quality report is prepared and presented to the Quality Assurance Team and Laboratory Management on a regular basis.

### **11.9 Documentation**

The following procedures support this policy:

- i. Internal KPIs Surveillance
- ii. Internal Audits
- iii. External Quality Assurance
- iv. External Benchmarking
- v. Quality Assessment Reports

## **12. Continuous Improvement**

### **12.1 Vision & mission**

The laboratory participates in a defined process improvement program to recognize and review problems that impact relevant areas and outcomes of patient care.

### **12.2 Objective**

This policy provides direction for the processes and procedures to effectively recognize and address probable issues or areas of improvement within the laboratory.



### **12.3 Process management**

The Laboratory Director is responsible for reviewing Quality Improvement activities.

The Quality Manager is in charge of collection and presentation of data for performance improvement. The Quality Assurance team is responsible for carrying out the activities of the Performance Improvement program.

The Laboratory Supervisor is responsible for recording the data needed for monitoring performance.

### **12.4 External quality assessment**

Opportunities for improvement (OFIs) are identified from several sources. Laboratory personnel participate in Quality Improvement activities that deal with relevant areas and outcomes of patient care.

### **12.5 Preventive actions review**

A mechanism is in place to assess processes in order to recognize and avert possible non-conformances.

### **12.6 Corrective measures**

A defined approach is used for process improvement when mistakes are identified.

### **12.7 Evaluation of usefulness of actions taken**

The laboratory evaluates the usefulness of actions taken to enhance performance.

### **12.8 Documentation**

The following procedures support this policy:

- i. Recognizing Opportunities for Improvement
- ii. Quality Improvement
- iii. Quality Improvement Evaluation



Alhamd-o-Lillah, under the banner of PSH, we have started a long awaited dream of haematology registry for different diseases, Diamond Blackfan Anaemia, Fanconi's Anaemia, Glanzman Thrombasthenia, Bernard Soulier Syndrome and many more diseases. Following is the registry proforma for Fancon's Anaemia & Glanzman Thrombos Asthenia which will be shortly available on PSH website (www.psh.org.ok) and we will be pleased to accept the case entries from various Haematology Centers of Pakistan.

## Fanconi's Anaemia

Data Collection Proforma

MR Number\*  Lab Number\*  Age\*  Gender\*  Male  Female Date\*

Name\*  Phone\*   
 First Name and Last Name

Address\*

### History

Transfusion  Yes  No H/O Sibling Death  Yes  No H/O Drug Intake  Yes  No Medical  Yes  No Family  Yes  No S/Studies  Yes  No

### Clinical Signs and Symptoms

Pallor  Yes  No Jaundice  Yes  No Splenomegaly  Yes  No Lymphadenopathy  Yes  No Skeletal Anomalies

### Investigation

CBC & Peripheral Film  Retics  Bone Marrow & Trephine Biopsy   
 Abdominopelvic Ultrasound  Skeletal Survey  Chromosomal Breakage Analysis   
 Autoimmune Profile  Viral Serology

Submit

Print





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## Glauzman Thrombos Asthenia

Data Collection Proforma

MR Number\*  Lab Number\*  Age\*  Gender\*  Male  Female Date\*

Name\*  Phone\*   
First Name and Last Name

Address\*

### History

H/O Bleeding  Yes  No H/O Pit txs  Yes  No H/O Medication  Yes  No H/O Trauma/Surgery/Dental Extraction  Yes  No Family History  Yes  No

### Clinical Signs and Symptoms

Pallor  Yes  No Petectiac / Brvises  Yes  No Bleeding Sites  Yes  No

### Investigation

CBC & Peripheral Film  Coagulation Profile  PT/APTT/BT/TT   
 Plt Count & Morphology  Plt Aggregation Studies  Flowcytometry

Genetic Analysis

### Management

Drugs  DDAVP  Antifibrinolytic  Platelet Transfusion  Factor VII Concentrate



# **HAEMCON2019 KARACHI**

## **21<sup>ST</sup> ANNUAL PSH MEETING**



March 14<sup>th</sup> – 16<sup>th</sup>, 2019

**3<sup>rd</sup> ANNOUNCEMENT &  
CALL FOR ABSTRACTS**

**Last Date of Submission  
February 15, 2019**

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