

**GUIDEBOOK FOR THE MANAGEMENT  
OF  
HEMOPHILIA TREATMENT CENTRE**



**HEMOPHILIA**  
PATIENTS WELFARE SOCIETY  

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RAWALPINDI CHAPTER

**Haemophilia Patient Welfare Society**

1<sup>st</sup> floor , Thalassemia House, opposite Rawalpindi Medical College (RMC) , Tipu road Rawalpindi

# **GUIDEBOOK FOR THE MANAGEMENT OF HAEMOPHILIA TREATMENT CENTRE**

**Prepared by the Consultants & Staff of Haemophilia Treatment Centre (HTC)**

- |                        |  |
|------------------------|--|
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# GUIDEBOOK FOR THE MANAGEMENT OF HAEMOPHILIA TREATMENT CENTRE

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# Haemophilia Treatment Centre (HTC )

## Introduction

The Haemophilia Treatment Centre looks after patients with **Inherited** bleeding disorders.

- These are diseases which are due to an inherited ( passed on from parents to their children) deficiency of coagulation factors. This results in prolonged bleeding after cuts / injury. The commonest disorder is Haemophilia which is X-linked (mostly males are affected). von Willebrand's Disease (vWD) is the next common and has autosomal inheritance (both males and females are affected).

## Most Common

- Haemophilia A (FVIII deficiency)
- Haemophilia B (FIX deficiency)
- Von Willebrand's Disease (vWF deficiency)

## Other Coagulation factor deficiencies / Platelets Function Disorders

- FV
- FVII
- FX
- FXIII
- Glanzman's Thrombasthenia
- Bernard Soulier Syndrome
- Storage Pool Defect

## Working of HTC

The HTC works under the guidance of:

- Director
- Deputy Director

The Centre has a **full time staff, visiting consultants** and a **Medical Advisory Board (MAB Annex 2)** which is composed of doctors from different fields of Medicine who work closely with the HTC .

treatment is provided at **Haemophilia Friendly Hospitals (HFH Annex 1)**

**Indoor**

**Full time staff Working 08:30 to 15:00 hours Monday to Saturday**

- Medical Officer
- Coordinator / Nurse
- Data operator / nurse assistant
- Receptionist

**Visiting Staff    Working    Visits day & time per rota    Monday to Saturday**

- Consultants (3)
- Research Coordinator

**N.B all visiting staff volunteer at the HTC**

**Emergency Cover    24 hours    Monday to Sundays**

- Consultants per rota
- HTC Coordinator

The job descriptions of all the staff are clearly defined and they work in close liaison with office staff , Office manager & Office Supervisor

## **Registration at HTC**

New Patients with Inherited Bleeding disorders are registered at the HTC. They are received at the reception and follow our protocol for registration.

### **Procedure for registration**

- Patients with suspected inherited bleeding disorder/ already diagnosed , are checked by the Medical Officer (M/O)
- Detail medical history and physical examination is done and documented in a file.
- Investigations are advised (if not already done)

### **Blood test Requirement    : at AFIP, CMH Rawalpindi**

- Complete Coagulation Profile
- Factor level
- HBs Ag
- HCV
- Other tests like Blood CP, LFTs, etc if required.

### **Document Requirement:**

- B- Form / CNIC of Patient
- Parents CNICs
- 2 passport size pictures of Patient
- Home address or (Proof of residential documents if the patient is foreign national)
- Phone No. / Email

### **Charges (per policy)**

- Registration fee (Life time ) Rs: 1000
- Annual fee Rs: 1500
- Handling charges (on visit) Rs: 200

The patient is given a HTC registration number and is also registered with Haemophilia Foundation Pakistan (HFP)

#### **Advantages of registration:**

- Free check up by the MO
- Free consultation & check up by the consultants at HTC ,if required
- Free / subsidized treatment (as per Policy)
- Free Referrals to & consultations by our Medical Advisory Board (MAB) & relevant Doctors at Haemophilia Friendly Hospitals (HFH) ,if required
- Admission in HFH if required
- 24 hr on call service for emergencies
- Subsidized emergency care per policy
- Free physiotherapy sessions
- Free counseling
- Free training to prepare factors
- Free training to infuse factors
- Access and invitation to Awareness activities, Seminars , workshops , picnics etc with free refreshments.

**N.B if requirement for registration is incomplete, the patient is still checked by medical officer and given emergency treatment with advice to complete requirements and visit again**

#### **Care of registered patient, advice, treatment, follow up**

The patient reports to the HTC reception where entry is recorded. The patient's file is taken out , and is passed on to the Medical Officer . After history and complete checkup, treatment advised by the MO is provided to the patient by the HTC nursing coordinator. A record of the treatment is maintained in the patient's file / Computer.

#### **Consultant appointments**

All **NEW** patients and all those who need to be seen by the HTC consultant have their appointments arranged by the HTC Coordinator (according to consultant visit days per ROTA or request by the MO to be seen by a specific consultant).

#### **Emergency Care during working hours**

Patient who comes in emergency is given priority. Complete check-up is done by the Medical Officer. If further consultation is needed, then the senior doctor according to ROTA is

consulted for expert opinion and treatment is given. If referral or admission is required then patient is guided accordingly. All this to be documented in patient's file

### **Out of hours Emergency Care (when the HTC is closed)**

Only **Life threatening bleeds** (bleeding in head , throat , gut , iliopsoas & post delivery )are treated out of hours (according to Policy). The patient contacts the HTC Coordinator who informs the Consultant on call and according to their advice treatment is given. The HTC Coordinator opens the HTC if required and the patient is given treatment .Treatment plan and referral for admission in Haemophilia Friendly Hospitals (HFHs) is given. The treatment is also documented in patient's file with the name of the consultant who advised it.

### **Treatment of patient's admitted at HFH's**

Patients who are referred to HFH for admissions are also given plan for treatment and supported while in hospital. In case the hospital staff contacts the HTC for advice regarding uncontrolled bleeding in some patient, we advise treatment and management. Factors are also provided as per policy.

### **Treatment of patients from other HTC's**

HPWS Rwp has a proper reimbursement policy approved by all four Chapters. When a registered patient of other chapter comes to HPWS Rwp for treatment, he is provided free consultation & examination by our Medical Officer. Treatment (factor cover) given per requirement. Later on his chapter is approached for reimbursement of consumed factors

### **Prophylaxis in patients with haemophilia (to prevent bleeding)**

In developed countries Prophylaxis is routinely carried out in Haemophilia patients to prevent bleeding, improving joint health and preventing deformities. The dose of factor VIII or IX is 20 to 40 IU/kg body weight, given 2-3 times a week to keep factor level above 1% at all times. This is very expensive & not at all practical for our set up where patients are given on demand treatment ( when bleeding occurs)

### **Low Dose Prophylaxis (LDP) project**

A number of studies in developing countries have shown good results by using a lower dose of prophylaxis, Low Dose Prophylaxis”(LDP) . WFH is supporting LPD project in many countries including Pakistan.

we started the LDP project in patients of Haemophilia A on 17<sup>th</sup> April 2018 ( factor donation from WFH ).Fifteen patients of ages between 1 to 8 years are enrolled in this ongoing study .They are given factor VIII concentrates at a dose of 250 IU I/V on weekly basis. Higher doses are given to older patients according to their weight.

The physiotherapy department of Benazir Bhutto Hospital (BBH) is affiliated with us and is participating in this study under the guidance of orthopedics surgery department of BBH. They are providing the facility of 3 monthly assessments of these patients for joint health .

10 new patients will be added to this project this year

## **Synoviorthesis**

Bleeding into the joints (Haemarthrosis) is one of the most common complications in Haemophilia Patients. If not treated properly, repeated bleeding into joint can lead to chronic Haemophilic synovitis which in turn causes more bleeding. **Recurrent bleeding in a joint ,4 times in a 6 month period ,makes it a ‘target joint’.**

The treatment of these joints is either surgery , to remove the inflamed synovium or intra-articular injection of chemical or radioactive materials which causes sclerosis of synovial blood vessels thus stopping further bleeds. This is called Synoviorthesis.

We use Rifampicin for chemical Synoviorthesis which is simple and cost effective and has given good results at our centre

### **Referral for Synoviorthesis**

Patients with target joints are selected. They are referred to Orthopedic surgeon for examination and appointment for intra-articular injection. Injection Rifampicin 600 mg is provided by the HTC .Factor cover protocol for synoviorthesis is available at the HTC .The patient is followed up at the HTC. In case of repeat bleeds the procedure may be repeated at weekly or fortnightly intervals for up to 6 injections.

Orthopedic Surgeons for Synoviorthesis on our Panel are :

- Dr. Riaz Sheikh BBH Rawalpindi
- Dr. Khalid Aslam QIH Rawalpindi
- Dr. Noman Maqbool Attock Hospital Morgah Rawalpindi

## **Treatment of Hepatitis**



Some of our patients develop Hepatitis B or Hepatitis C as consequence of use of FFP or Cryoprecipitate. We recommend **Hepatitis B vaccination** to all our patients at the time of registration & have their Hepatitis B & C status checked. Thereafter these tests are repeated 6 monthly, and if the tests are Positive they are referred BBH Pathology department for further evaluation and free treatment (as per Govt. Policy).

- Free PCR test
- Free treatment of Hepatitis C
- Vaccination provision of Hepatitis B

Focal person is Dr. Nadeem Ikram haematologist in B.B.H Pathology Laboratory.

## **Surgeries, Pregnancy care & delivery**

**Surgery** : The patient is seen by the MO and if there is a need for surgery/ procedure, is reviewed by **the** HTC Consultant .The patient is then referred to the relevant surgeon with a written plan to cover the procedure / surgery

Protocols / SOPs for various planned surgeries like Major or Minor operations for patients with Haemophilia and VWD are available at the HTC. However these are modified according to the specific needs of the individual patients and given to them. Calculation of the dose of factor, FFPs etc. according to weight of the patient and timing of infusion for most effective results, are provided as well. All factor support is provided by the society per policy

Further more the Medical Officers / Consultants at the HTC are available for consultation and help during hospital admission (Occasionally hospital visit also) and after discharge till complete recovery occurs.

## **Care of Females with inherited bleeding disorders**

### **Menorrhagia**

Advice regarding heavy menstrual bleeding is given by our medical officer or consultants.

If need be, these patients are referred to Gynecologists at Haemophilia Friendly Hospitals mainly BBH, HFH & PIMS.

### **Pregnancy**

Pregnant patients need antenatal care with special attention to prevention of bleeding / abortion. For this they are booked at HFH Gynae / Obs. Department and also seen regularly at the HTC to keep a track of the progress of pregnancy. (any bleeding episodes, anemia, Psychological support etc.)

#### **Protocol for Pregnancy care**

- 1<sup>st</sup> trimester registration

- Regular antenatal checkups
- Regular investigations
- Plan about delivery ( normal delivery )
- Operation plan provision (C section)
- Regular follow up after delivery

### **Delivery / Caesarian Section (C-Section)**

The patients are given specific plan to cover normal vaginal delivery as well as C- Section. The HTC is fully involved in taking care of any bleeding episodes in these patients during and after delivery.

### **Delivery of infants with suspected inherited bleeding disorder**

This Protocol is available for care of the baby and is passed on to the Obstetric team along with the delivery plan with advice for blood tests to diagnose inherited bleeding disorders

**NOTE: There is no need to recommend c- section to these patients only because of their bleeding disorder. C- section should be done only if there is an obstetric indication, otherwise a normal delivery is possible under factor cover and some precautions.**

### **Prenatal diagnosis**

This is diagnosis of **Haemophilia** before birth. This procedure is very expensive, and presently only available at National Institute of Bleeding Disorders (NIBD) Karachi. If the patient or her family is interested in getting it done, they are referred to NIBD. ( Dr. Arshi Ph. No. 021-34228522 is the focal person ). The test is done during 1<sup>st</sup> trimester if the child is **male**.

### **Factor procurement Policy**

HPWS Rwp has an approved Factors Procurement Policy, no one (Patient , Member or employee) can change the rules of approved Policy.

The HPWS Rwp arranges the factors from various sources like WFH, (with the help of NMO), Project Share, and purchase through donations etc. **The treatment for Haemophilia patients is our top most priority.**

The factor cover to registered patients is provided at a nominal cost.

### **Factor storage, in store & treatment room**

HTC has a proper mechanism of factors storage according to WFH Guidelines

HPWS Rwp has a proper, separate storage room for factors, with facility of UPS and generator  
The HTC Coordinator **checks & records** the refrigerators temperature on daily basis (twice a day).

For routine daily consumption of factors a refrigerator is placed in treatment room. Limited number of factors are taken from main store and placed in the refrigerator. (According to average daily requirement

## Factor availability

The following Factors are available at the HTC:

- FVIII ( long acting and short acting)
- FIX ( long acting and short acting)
- VWF
- FVII
- FEIBA



FVIII



FIX



FEIBA



FVII



VWF

## Waste disposal

For waste disposal the standard Protocol is followed. Two bins, Red for infectious and blue for non-infectious waste are placed in treatment room. All the waste (infectious and non-infectious) is disposed off according to health & safety guidelines. This process is being done on alternate days. In this regard National Cleaner Production (NCP) is responsible for taking out the waste and dispose it off/ incinerate if required.

**Red bin** (for infectious waste) used for

- Butterfly needles
- Swabs
- Sticking
- Changing / removing dressing if necessary
- Syringes
- Gloves
- Needles cut by a cutter & disposed off in a sharps container with bleach in it

**Blue bin** (for Noninfectious waste) is be used for

- Empty vials
- Boxes / Factors packing
- Syringe / Butterfly covers
- Empty cartons
- Used papers etc.

### Training of patients to prepare & infuse factors

Majority of patients (grownups and adults) and parents are trained to prepare and infuse the factors , it is one of the most important tasks of HTC so that patients can infuse themselves at home if they have bleeds and cannot come to the HTC . It facilitates home infusions / emergency cover for factors.

#### Training is provided by HTC Coordinator:

- Reconstitution and preparation of Factor
- Mix the powder with distilled water and then roll the vial gently between the palms of hands slowly (Rotatory movement). During reconstitution of Factor , **DO NOT shake the vial vigorously.**
- **Infusion:** Factor is infused slowly and gradually intravenously, making sure that no factor remains in the tubing. The ideal time for infusion is not more than 100u/min in children & 3ml/min in adults. Butterfly needles are preferred. After infusion 5 minutes persistent pressure at the site of infusion is recommended.



**Patient infusing factor successfully**



**Dedicated father infusing factor without any difficulty**

### Monthly Reporting

The activities of the HTC are reported every month

## Haemophilia Treatment Centre (HTC) Reporting

Sr.#	Activity	Reporting	Responsibility
1.	Patient details	Details of all patients attending the HTC Breakdown of <ul style="list-style-type: none"> <li>• type of inherited bleeding disorder</li> <li>• type of bleeding</li> <li>• referrals</li> <li>• admissions</li> <li>• surgeries including circumcisions</li> <li>• deliveries</li> <li>• Procedures like synoviorthesis</li> <li>• deaths</li> </ul>	<b>Preparation</b> <b>HTC coordinator</b> <b>Checking</b> <b>MO</b>
2.	Parent /Patient training	<ul style="list-style-type: none"> <li>• Preparation of Factors</li> <li>• Infusion of Factors</li> </ul>	<b>HTC coordinator</b>
3.	Counselling	<ul style="list-style-type: none"> <li>• Awareness of bleeding disorders</li> <li>• Psychosocial</li> <li>• general</li> </ul>	<b>MO</b>
4.	Physiotherapy details	Details of patients attending physiotherapy Breakdown of treatments given to them	<b>Preparation</b> <b>HTC coordinator</b> <b>Checking</b> <b>MO</b>
3.	Factor utilization	Details of utilization of factors along with units 2.1 Purchased Factor Concentrates 2.2 Donated factor concentrates 2.3 cost of all factors	<b>Preparation</b> <b>HTC coordinator</b>
4	Factor stock	Available factor stock	<b>Preparation</b> <b>HTC coordinator</b>  <b>Checking</b> <b>MO</b>
5.	Patient success stories	Minimum 2 per month	<b>Preparation</b> <b>MO</b> <b>Soft copies</b> <b>Data operator</b>
6.	CME Seminars & conferences	Detail activity report	<b>Preparation</b> <b>Director / Deputy Director</b> <b>Soft copies</b> <b>Data operator</b>
7.	Health group activities	Activity report	<b>Preparation</b> <b>Group leader</b> <b>Soft copies</b> <b>Data operator</b>
8.	Awareness sessions	Activity report	<b>Preparation</b> <b>Incharge activity</b>

			<b>Soft copies</b> <b>Data operator</b>
<b>9.</b>	Research projects Papers/posters	Project details Papers under preparation Papers submitted for publication/ published Poster details / presentation	<b>Preparation</b> <b>Ist authors ,Director /Deputy Director</b> <b>Soft copies</b> <b>Data operator</b>
<b>10</b>	Photos	All HTC related activities files kept & updated	<b>Soft copies</b> <b>Data operator</b>
<b>11.</b>	National registry	update	<b>Data operator</b>
<b>12</b>	Monthly ROTA	For consultants & Staff	<b>Preparation</b> <b>HTC coordinator</b> Approval <b>Director / Deputy Director</b>

## **N.B Report to be submitted to Director/ Deputy Director HTC in the First week of each month**

### **CME activities at HTC**

The HTC is actively involved in continuing medical education (CME). These activities are routinely held at the centre. Young doctors as well as trainee Haematologists are given awareness about bleeding disorders and their management by seminars, case based discussions, practical demonstrations

The centre staff is responsible for carrying out these activities

### **Future Projects**

- Gene Therapy is the latest development in Haemophilia Care, in the developed countries. The HTC has started collaboration with Royal Free Hospital in London to enroll our patients in this program.
- Treatment of Patients with Inhibitors with Immune Tolerance Induction (ITI)
- Hepatitis C treatment of patients

### **Collaboration of HTC and HPWS Rwp**

HTC collaborates with the Society in all its Awareness sessions and other programs

### **Research**

The HTC encourages active research in the field of bleeding disorders and their management. The researcher applies to the Director HTC for permission to conduct research at Centre along with synopsis which is reviewed and approved by our Institutional Review Board (IRB Annex 3) A protocol for research (available at HTC) is to be followed by the researcher

## Annex 1

### Haemophilia Friendly Hospitals & their contact Doctors

Hospital	Name	Designation
Benazir Bhutto Hospital Rawalpindi	Dr. Nadeem Ikram	Consultant Haematologist
	Dr. Ranaa Zeeshan	Consultant Haematologist
	Dr. Riaz Sheikh	Consultant Orthopedic Surgeon
	Dr. Moazzma Ahmed	Senior Physiotherapist
	Dr. Ayesha Fareed	Consultant Psychiatrist
	Dr Humera	Consultant Gynaecologist
PIMS Hospital Islamabad	Dr. Zahida Ahmed	Consultant Dental surgeon
	Dr. Tazeen Anwaar	Medical officer Haemophilia Centre
	Dr. Riaz Khattak	Medical Officer Haemophilia Centre
	Dr. Bisma	Senior Physiotherapist
Holy Family Hospital Rawalpindi	Dr. Qasim Ali	Consultant Surgeon
	Dr. Atifa Shuaib	Consultant Haematologist
	Dr. Rizwana Chaudhry	Consultant Gynaecologist
Attock Refinery Hospital Morgah Rawalpindi	Dr. Noman Maqbool	Consultant Orthopedic Surgeon
AFIP/ AFBMTC Rawalpindi	Brig. Assad Abbasi	Consultant Haematologist
	Lt. Col. Helen Robert	Consultant Haematologist
	Maj. Saima Saad	Consultant Haematologist
	Brig Qamar un Nisa	Consultant Haematologist
	Col Mehreen	Consultant Haematologist
Quaid e Azam International Hospital Rawalpindi	Dr. Khalid Aslam	Consultant Orthopedic Surgeon
Shifa International Hospital Islamabad	Dr. Ayesha Junaid	Consultant Haematologist
Al-Shifa Eye Trust Rawalpindi	Dr. Sarah Zafar	Consultant Ophthalmologist

## Annex 2

### Medical Advisory Board

SR#	Name	Designation
1.	Prof.Tahira Zafar	Consultant Haematologist <b>Chairperson</b>
2.	Prof. Lubna Zafar	Consultant Haematologist <b>Deputy Chairperson</b>
3.	Dr. Nadeem Ikram	Consultant Haematologist <b>Member</b>
4.	Dr. Qasim Ali	Consultant Surgeon <b>Member</b>
5.	Dr. Sarah Zafar	Consultant Ophthalmologist <b>Member</b>
6.	Dr Khalid Aslam	Consultant Orthopaedic Surgeon <b>Member</b>
7.	Prof.Riaz Sheikh	Consultant Orthopaedic surgeon <b>Member</b>
8.	Prof. Nouman Maqbool	Consultant Orthopaedic Surgeon <b>Member</b>
9.	Prof. Naeem Zia	Consultant Surgeon <b>Member</b>
10.	Dr. Humaira Bilqees	Consultant Gynaecologist <b>Member</b>
11.	Dr. Irum Naeem	Consultant Gynaecologist & Sonologist <b>Member</b>
12.	Dr. Zahida Ahmed	Dental surgeon <b>Member</b>
13.	Dr. Ayesha farid	Consultant Psychiatrist <b>Member</b>
14.	Dr. Farkhanda Nazli	Consultant Rehab Medicine <b>Member</b>



## Annex 3

### Institutional Review Board (IRB)

SR#	Name	Designation	
1	Prof Tahira Zafar	Haematologist	<b>Chair Person</b>
2	Prof Lubna Zafar	Haematologist	<b>Member</b>
3	Dr Nadeem Ikram	Haematologist	<b>Member</b>
4	Munawar Sher Khan	Social Welfare	<b>Member</b>
5	Fazal Qayyum	Parent	<b>Member</b>
6	Mehr Amer Malik	Lawyer	<b>Member</b>
7	Bushra Suhaib	Lay person	<b>Member</b>

**N.B First three are Primary reviewers & form the Scientific Review committee**