

Spectrum of Patients at Haemophilia Treatment Centre, Children Hospital, PIMS, Islamabad.

Dr. Riaz Ul Haq Subhani

Dr. Tahira Zafar

Dr. Lubna Naseem

Prof. Khalid Hassan

Pakistan Institute of Medical Sciences

Islamabad

Introduction

- ★ Haemophilia is an X-linked congenital bleeding disorder.
- ★ 1 in 10,000 male births.
- ★ Deficiency of coagulation factor:
VIII (Haemophilia A) or
IX (Haemophilia B)
- ★ Haemophilia A is more common (80-85%)

Haemophilia is characterized by:

★ Prolonged Bleeding

1. Spontaneous
2. Traumatic

★ Types of bleedings

1. Joints
2. Soft tissues
3. CNS
4. External tissues
5. Traumatic

Joint Bleeding

- ★ Pain
- ★ Swelling
- ★ Immobility
- ★ Ankylosis
- ★ Chronic Arthritis



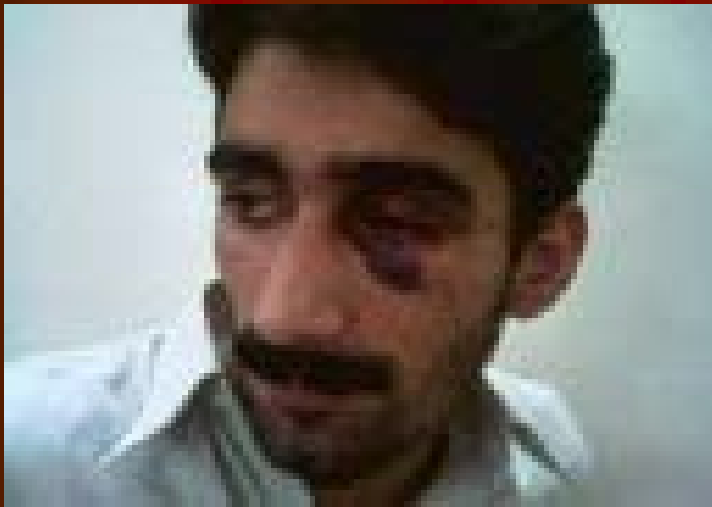
Joint Bleeding



Joint Bleeding



Peri-orbital Bleeding



Bruises



Presentation in a young child

- ★ Gum & tongue bleed
- ★ Bruises after fall
- ★ Bleedings into joints, soft tissues and muscles is seen more frequently in >2 years of age



Haemophilia - Lab Diagnosis

Haemophilia	PT	APTT	TT	CT	BT	<u>Mixing studies</u> Correction by adsorbed plasma	<u>Mixing studies</u> Correction by aged serum	Factor VIII assay	Factor IX assay
A	N	↑	N	↑	N	Corrected	Not Corrected	Deficient	Normal
B	N	↑	N	↑	N	Not Corrected	Corrected	Normal	Deficient

Objective

To find the:

- ★ Demographic pattern
- ★ Presentations of signs and symptoms
- ★ Frequency of Haemophilia A, B and other bleeding disorders
- ★ Severity of Haemophilia
- ★ Response to different treatments in patients presented at Haemophilia Centre, PIMS.

Materials & Methods

- ★ Patients referred to Haemophilia Centre PIMS were included in the study.
- ★ Diagnosis was established on the basis of:
 1. Normal PT
 2. Prolonged APTT
 3. Mixing studies (Correction of APTT by adsorbed plasma and not by aged serum)
 4. Factor VIII level assay

- ★ On the basis of factor level, hemophilia was categorized as mild, moderate and severe.
- ★ Treatment given & response was dependent on:
 1. *Type & site of bleeding*
 2. *Factors initiating bleeding*
 3. *Clinical characteristics of joint involvement*
 4. *Functional loss and disability*
- ★ Records were maintained.

Results

★ Total	163
★ Haemophilia A	104 (63.8 %)
★ Haemophilia B	13 (7.98 %)
★ Other bleeding disorders	46 (28.22 %)
★ Haemophilia A : B	8: 1

Breakup Of Cases With Inherited Bleeding Disorders

Disorder	No	% age
Haemophilia A	104	63.8
vWD	27	16.56
Haemophilia B	13	7.98
Glanzman Thrombasthenia	6	3.68
Factor X Deficiency	3	1.84
Factor V Deficiency	2	1.22
Factor XIII Deficiency	1	0.61
Factor XI Deficiency	1	0.61
Bernard Soulier Syndrome	1	0.61

Demographic Details

Gender

100% males

Demographic Details

Family history of Haemophilia	% age
Positive	56.41%
Negative	43.59%

Demographic Details

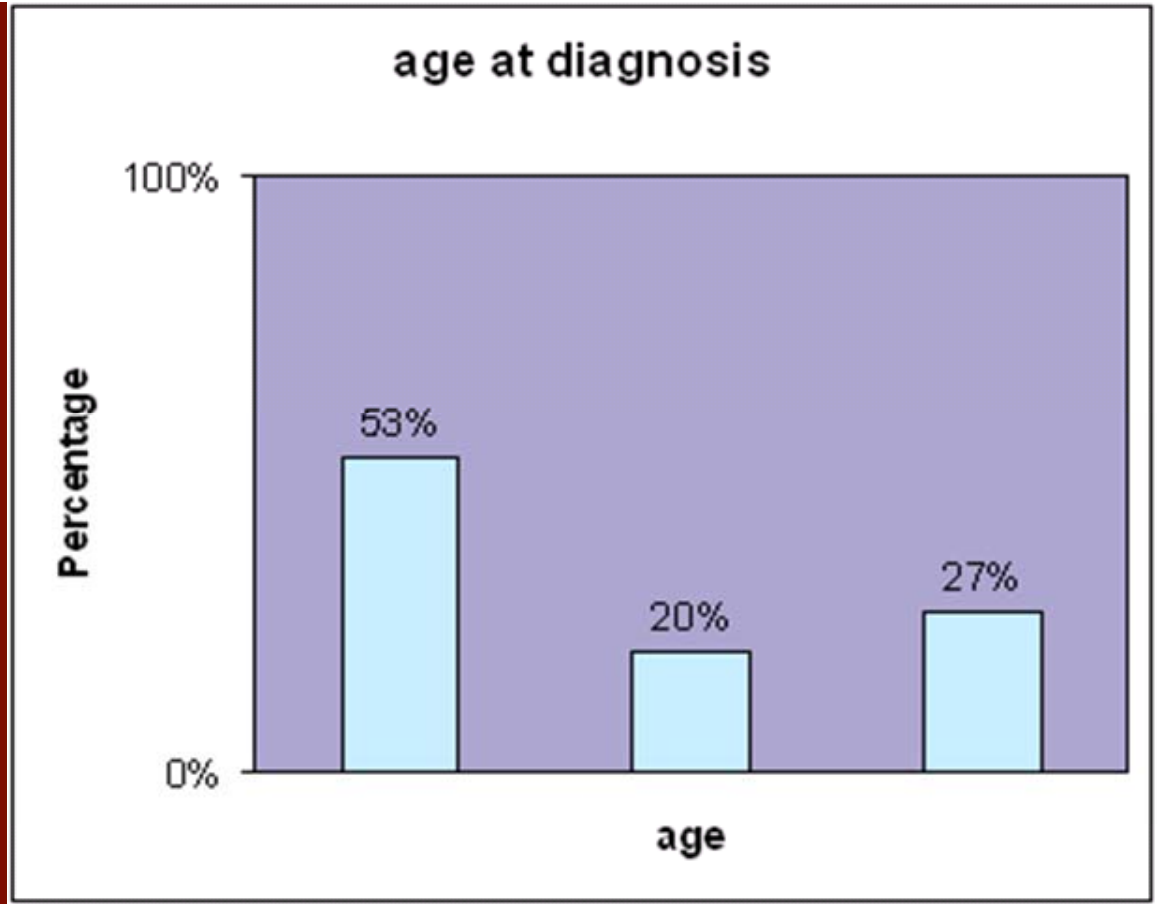
Rural / Urban division:

Rural	54.7%
Urban	45.3%

Demographic Details

Age

Age	% age
<2 yrs	11.82
3-10 yrs	37.89
11-20 yrs	43.46
21-30 yrs	2.56
>30 yrs	4.27



Up to 1 years:

62 (53%)

1 - 5 years:

23 (20%)

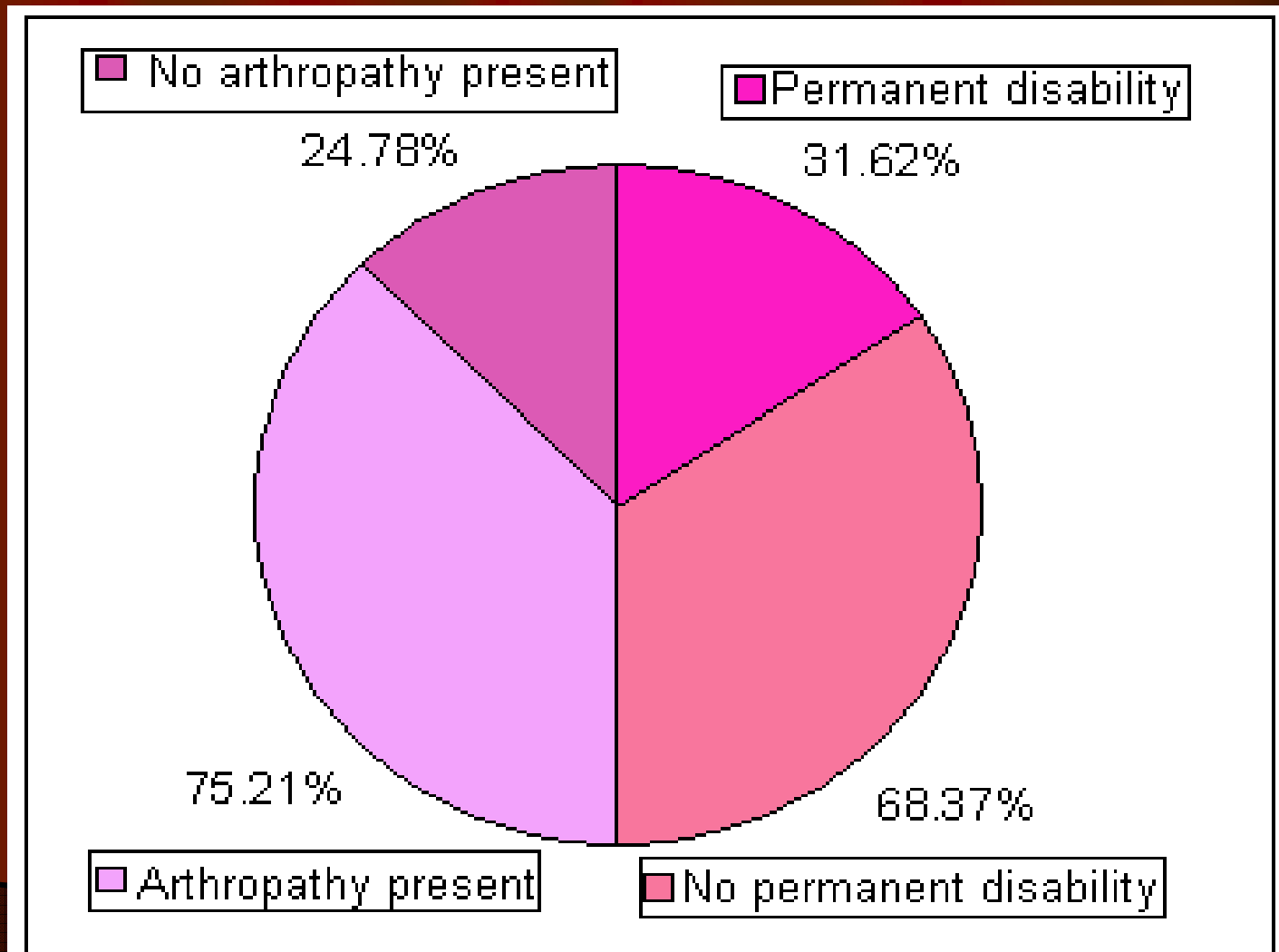
> 5 years:

32 (27%)

Haemophilia - Stratification on clinical severity

Severity	Bleeding Tendency	Number (%)
Severe (Factor levels: <1 U/dl)	Frequent spontaneous bleeding into joints, muscles and internal organs	65 (55.55%)
Moderate (Factor levels: 1-5 U/dl)	Some spontaneous bleeds; bleeding after minor trauma	22 (18.8%)
Mild (Factor levels: >6–30 U/dl)	Bleeding only after significant trauma or surgery	30 (25.64%)

Disability and Arthropathy



Bleeding pattern

Bleeding episodes/year	Number of patients (%)
Up to 3	57 (48.71%)
4 – 10	32 (27.35%)
11 - 20	16 (13.67%)
>20	12 (10.25%)

Mode of Bleeding:

Spontaneous bleeding (74.35%)	87/117
Bleeding after trauma (25.64%)	30/117

Haemophilia-Bleeding Episodes

Site	No of Episodes (%)
Knee joint	310 (47.83)
Ankle	90 (13.88)
Elbow	80 (12.34)
Bleeding gums/oral cavity/ Teeth	35 (5.39)
Shoulder	30 (4.62)
Toe	18 (2.77)
Hip	15 (2.31)
Wrist	11 (1.69)
Fingers	10 (1.54)
Urinary tract	9 (1.38)
Gastrointestinal tract	8 (1.23)

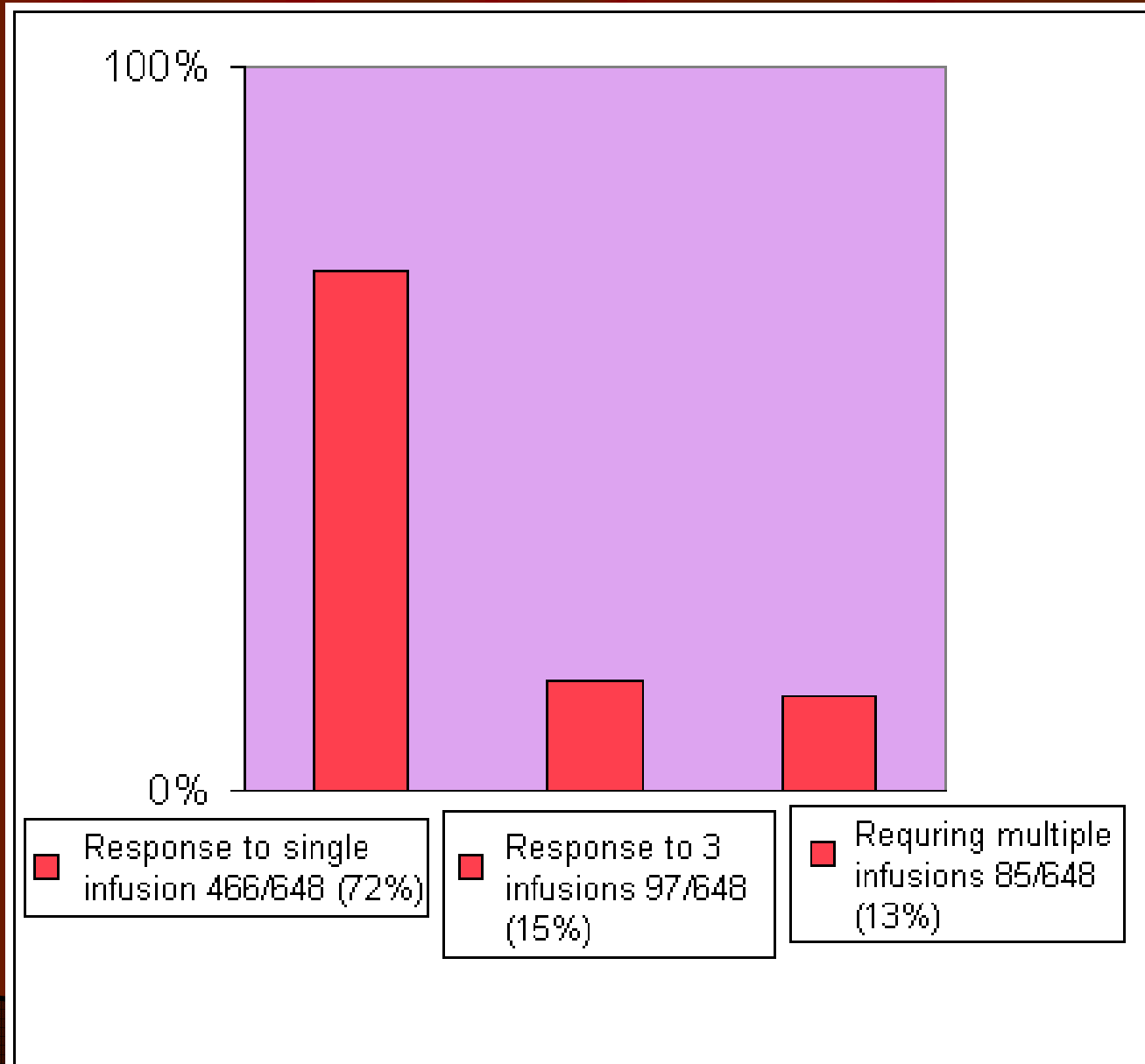
Haemophilia-Bleeding Episodes (cont.)

Site	No of Episodes (%)
Epistaxis	6 (0.92)
Intracranial	5 (0.77)
Post Operative	5 (0.77)
Haematoma Iliopsoas region	2 (0.3)
Ecchymotic spots	2 (0.3)
Haematemesis	2 (0.3)
Eye	1 (0.15)
Haematoma perineum	1 (0.15)
Haematoma scalp	1 (0.15)
Haematoma gluteal region	1 (0.15)

Treatment Given

Product used	Episode(%)
Factor Concentrates	492/648 (62%)
Fresh frozen plasma	98/648 (15%)
Cryoprecipitate	71/648 (11%)
DDAVP	19/648 (03%)
Combination	58/648 (09%)

Treatment response



Haemophilia Management

★ General management

- Avoid - Aspirin

Anti platelet agents

Intra muscular injections

- Treat bleeding episodes promptly

- ★ Lessens morbidity

- ★ Prevents complication

- Plan surgical procedures carefully

- Tranexamic Acid

**A view of seminars and
workshops at
Haemophilia Centre,
Pakistan Institute of
Medical Science,
Islamabad.**







THANKS

