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PREVALENCE OF HAEMOPHILIA IN A CHILDREN WITH JOINT INVOLVEMENT.

Pediatrics

Dr.Brajesh Kumar Senior Resident,Department of Pediatrics PMCH Patna.
Dr. Abhishek Kumar Senior Resident,Department of Pediatrics PMCH Patna.
Dr. Manoj Kumar Singh* Associate Prof. Department of Pediatrics PMCH Patna. *Corresponding Author

ABSTRACT

Background: Defciency of factor VIII (HemophiliaA) , factor IX (HemophiliaB) and von willebrands factor are the most frequent coagulation defects.The classification of HA provides a guidance to possible types of bleeding and the rate of occurrence of hemorrhagic episodes. The hemophiliac patients with severe form, can be presented with spontaneous bleeding and hemorrhage after minor trauma about 1-6 times in a month, including hemarthrosis and intramuscular hemorrhage., the patients usually experience bleeding after mild to moderate injuries, while HA patients with mild form may not be diagnosed for years and bleed after surgery or major trauma. About 70% of children who have a positive history of HA in the family are diagnosed at birth or after the first bleeding episode.

Material and Methods: Selected patients were under went for complete blood count,General blood picture, PT,APTT< Thrombin time, A complete medical history of the patient was taken with reference to the name, age, sex, address, occupation, history of present illness, duration illness, education of patient and parent. Previous history of treatment, past and personal history and socio-economic status were noted. Parents were explained the purpose of the study and the procedures involved.The Hemophilia Joint Health Score (HJHS) was used to assess joint health in children included in the study.

Conclusion: The prevalence of hemophilia and incidence of inhibitors in these patients is varies in different regions of india.

KEYWORDS

Haemarthrosis, Hemophilia A and B, joint disease, haemophilic arthropathy.

INTRODUCTION

Haemophilia is an X linked heritable coagulopathy with an overall prevalence of approximately 1 in 10,000 individuals. Hemophilia A and B are clinically indistinguishable congenital bleeding disorders caused by the X-linked recessive disorder due to deficiency of coagulation factor VIII (hemophilia A) or factor IX (hemophilia B). The frequency and severity of bleeding into tissues, organs, and joints is generally related to the severity of the deficiency of the respective factor. Hence, definitions of hemophilia severity have been assigned by coagulation factor activity (FA), with severe being <1%, moderate being 1% to 5%, and mild being >5% FA[i]. From 1998, the Centers for Disease Control and Prevention (CDC) is monitoring the people with bleeding disorders who receive care in federally supported hemophilia treatment centers (HTCs) through a public health surveillance system initially called the Universal Data Collection (UDC) system[ii].

Worldwide Hemophilia A (HA) is the most common coagulation factor deficiency. It affects all populations and its prevalence varies among different countries, but roughly prevalence is estimated to be of 3-20 cases per 100,000 population[iii]. The classification of HA provides a guidance to possible types of bleeding and the rate of occurrence of hemorrhagic episodes. The hemophiliac patients with severe form, can be presented with spontaneous bleeding and hemorrhage after minor trauma about 1-6 times in a month, including hemarthrosis and intramuscular hemorrhage. In moderate form, the patients usually experience bleeding after mild tooderate injuries while HA patients with mild form may not be diagnosed for years and bleed after surgery or major trauma. About 70% of children who have a positive history of HA in the family are diagnosed at birth or after the first bleeding episode. At the same time, children who have a negative family history are generally diagnosed in situations such as injection of a drug or vaccine, minor surgeries and they experience easy bruising after minor trauma.

Hemarthrosis is a common finding in hemophilia and is the hallmark of a severe form. Inflammation of the joint capsule, secondary to hematoma is an important cause of pain[v].

MATERIALAND METHODS:

The Prospective study was carried out in Patna medical college and hospital patna Bihar, to compare the severity of joint involvement with different demographic parameters in children suffering from hemophilia A

A complete medical history of the patient was taken with reference to the name, age, sex, address, occupation, history of present illness, duration illness, education of patient and parent. Previous history of treatment, past and personal history and socio- economic status were noted.Parents were explained the purpose of the study and the procedures involved.

The Haemophilia Joint Health Score (HJHS) was used to assess joint health in children included in the study.

OBSERVATIONS AND RESULTS:

A total of 51 patients were included in the study

Table 1: Age of diagnosis wise distribution of cases

Age of diagnosis	Percentage	N
>=12 months	54.90%	28
< 12 months	45.10%	23
Total	100%	51

Age of diagnosis >=12 months was observed in 28 (54.90%) of cases while in 23 (45.10%) cases age of diagnosis was < 12 months

Table 2: Distribution of cases according to severity of disease

Severity of disease	N
Mild	2(3.95%)
Moderate	4(7.84%)
Severe	45(88.23%)
Total	51(100%)

Severe disease was observed in 45 (88.23%) of cases, moderate in 4 (7.84%) and mild form was observed in 2 (3.95%).

Table 3: Individual Joint and Item wise score

Joint (Sum of score of all cases)								
Item	LE	RE	LK	RK	LA	RA	Total	%
Swelling	0	9	14	18	8	8	57	24.1
Duration	0	0	4	5	3	2	14	5.9
Muscle atrophy	0	0	2	4	1	1	8	3.4
Crepituson motion	1	7	10	12	4	3	37	15.6
Flexion loss	1	13	12	15	7	8	56	23.6
Extension loss	1	9	8	10	3	4	35	14.8
Joint pain	2	4	5	7	4	4	26	11.0

Strength	0	0	1		1	1	4	1.7
Total	5	42	56	1	31	31	237	100

*LE- Left elbow, RE-Right Elbow, LK- Left knee, RE - right Elbow, LA- Left Ankle, RA -Right Ankle

Table 4: Type of family wise distribution of cases

Type of family	Frequency	Percentage
Nuclear	42	82.4%
Joint	9	17.6%
Total	51	100%

Out of total 51 cases 42 (82.4%) were from the nuclear family while 9 (17.6%) were from the joint family

Table 5: BMI wise distribution of cases

BMI (Kg/m^2)	Frequency	Percentage
Underweight	13	25.5%
Normal Weight	25	49.0%
Overweight	5	9.8%
Obese	8	15.7%
Total	51	100%

As shown in above table nutritional status according to BMI indicate s that 25.5 %(13) cases were underweight,49% (35) cases were normal weight, 9.8% (5) cases were overweight and 15.7%(8) were obese.

Table 6: Family history of Haemophilia wise distribution of cases

Family history of haemophilia	Frequency	Percentage
Present	24	47.1%
Absent	27	52.9%
Total	51	100%

Out of 51 cases of Haemophilia 47.1% (74) cases had family history of Haemophilia while in remaining 52.9% (27) cases there was no family history of Haemophilia.

Table 7: Correlaion of HJHS with the type of Family

Type of Family	Frequency	HJHS mean	SD	P value (t test)
Nuclear	42	5.36	10.79	0.649
Joint	9	3.67	4.74	
Total	51	5.06	9.98	

Mean HJHS was higher in patients of nuclear family as compared to joint family.

Table 8: Distribution of cases according to distance from home to hospital

Distance from home	N	percentage
≤ 15 kms	38	74.5%
>15 kms	13	25.5%

As shown in above table out of 51 cases of haemophilia 74.5% (38) cases were residing 15 km or less from the hospital while remaining 25.5 %(13) cases were reading from more than 15 km away from the hospital.

Table 9: Correlation of HJHS with distance between home and hospital

Distance from home to hospital	Frequency	HJHS mean	SD	P value (t test)
≤ 15 Km	38	3.97	8.468	0.187
> 15 Km	13	8.23	13.386	
Total	51	5.06	9.98	

Mean HJHS was higher in cases residing more than 15 km away from the hospital (8.23 ±13.386) as compared to cases nearer to the hospital (3.97±8.468). However the difference was not statistically significant. So HJHS was not correlated with the distance from home to hospital.

Table 9: Number of bleeds wise distribution of cases

No .of bleeds	Frequency	Percentage
20 or less	31	60.8%
More than 20	20	39.2%
Total	51	100%

In the history of bleed till date it was observed that 31 (60.8%) cases had 20 or less bleeds till date while 20 (39.2%) had more than 20 bleeds till date.

Table 10: Correlation of HJHS with number of bleeds till date

Number of bleeds	Frequency	HJHS Mean	SD	P value (t test)
20 or less	31	2.26	5.209	0.011
More than 20	20	9.40	13.659	
Total	51	5.06	9.98	

It was observed that mean HJHS was higher in cases having bleeds more than 20 (9.40) as compared to cases having bleeds less than 20 (2.26). This difference was statistically significant (P=0.011). So HJHS and thus disability was higher in cases having more number of bleeds.

Table 11: Education wise distribution of cases

	Frequency	Percentage
Do not go to school	8	15.7%
Go to school	43	84.7%
Total	51	100%

As shown in above table out of 51 cases of Haemophilia, 15.7% (8) do not go to school while remaining 84.3%(43) go to school.

Table: 12 Correlation of HJHS with education of the cases

Education	Frequency	HJHS Mean	SD	P value (t test)
Did not go to school	8	5.13	8.36	
				0.984
Go to school	43	5.05	10.34	
Total	51	5.06	9.98	

As shown in the above table mean HJHS was slightly higher in cases not going to school (5.13) compared to cases going to school (5.05). However application of test indicated that this difference was statistically not significant (p>0.05), so HJHS was not correlated with the education of patient.

Table13: Distribution of cases according to father's education

Father's education	Frequency	Percentage
Primary or below	11	21.6%
Secondary or above	40	78.4%
Total	51	100%

Out of 51 cases Father of 21. 6%(11) cases were illiterate or had studied upto primary or below.Father of 78.4%(40) cases had studied up to secondary school or above that.

Table 14: Distribution of cases according to Mother's education

Mother's education	Frequency	Percentage
Primary or below	27	52.9%
Secondary or above	24	47.1%
Total	51	100%

Out of 51 cases, mother of 52.9%(27) cases were illiterate or had studied up to primary or below. Mother of 47.1%(24) cases had studied up to secondary School or above that.

Table 14: Income wise distribution of cases

Income per head per month (in Rs.)	Frequency	Percentage
<834	5	9.8%
835-1670	14	27.5%
1671-2784	20	39.2%
2785-5569	9	17.6%
>5570	3	5.9%
Total	51	100%

Above table shows distribution of cases according to their family income based on Modified Prasad classification. The highest number of cases were from middle income and upper middle income.

Table 13: Correlation of HJHS with BMI of the cases

BMI (Kg/m^2)	Frequency	HJHS Mean	SD
Underweight	13	7.00	9.94
Normal Weight	25	3.88	10.29
Overweight + obese	13	5.38	9.84
Total	51	5.06	9.98

Underweight Vs Normal weight -P value 0.376
(Overweight + Obese) Vs Normal weight -P value 0.668

DISCUSSION:

Comprehensive musculoskeletal assessment for monitoring joint health in haemophilia can be done by both physical assessment with Haemophilia Joint Health Score (HJHS) and assessment of self-reported function by Haemophilia Activities List (HAL). In the present study swelling and flexion loss was contributing highest among the all disability items taken into account for calculation of HJHS. It was also observed that almost half of the score was due to contribution of swelling and flexion loss. In a study by Ribeiro Tet al[vi]The HJHS item weights provide a sum to 1.0; the highest item was extension loss (0.139) followed by swelling (0.121), whereas the lowest was duration of swelling (0.057) followed by muscle atrophy (0.08).

Disability and Health, "structure and function" is a major component of an individual's outcome in health and disability. Surveillance of musculoskeletal changes is recognized to be essential for timely patient evaluation and management of the disease state.

Our study showed that Underweight Vs Normal weight - P value was 0.376 while (Overweight + Obese) Vs Normal weight -P value was 0.668, although not significant but higher HJHS (7.00) was observed in underweight. In the history of bleed till date it was observed that 31 (60.8%) cases had 20 or less bleeds till date while 20 (39.2%) had more than 20 bleeds till date. HJHS was higher in cases having bleeds more than 20 (9.40) as compared to cases having bleeds less than 20 (2.26). This difference was statistically significant (P=0.011). So HJHS and thus disability was higher in cases having more number of bleeds.

In a study by Feldman BM et al[x]. The mean age of the study group was 10.8 years. Sixty-eight percent were severe (93% of whom were treated with prophylaxis), 15% were moderate (24% treated with prophylaxis), and 17% were mild (3% treated with prophylaxis). The HJHS correlated moderately with the physician total joint score (rs=0.42, P<0.0001) and with overall arthropathy impact (rs=0.42, P<0.0001).

CONCLUSION:

HJHS shows construct validity and is more sensitive for mild arthropathy and can be used for studies in children with hemophilia. Dosing should be individuals and increased in the case of bleeding, prevention of bleeding episodes through early treatment will prevent accumulation of blood in the joint and the subsequent inflammation and potential haemophilic arthropathy.

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