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RESEARCH ARTICLE
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FREQUENCIES OF BLEEDING DISORDERS IN CHILDREN PRESENTING TO PEDIATRIC DEPARTMENT OF A TERTIARY CARE HOSPITAL

Arifa Jabeen[1*], Amna Begum[2], Asghar Ali Memon[3], Waheed Ali[4], Asma waheed[5]

[1*]Assistant professor Pediatrics Gambat Institute of medical and health sciences
[2]FCPS.MBBS Consultant Gynaecologist Assistant professor Pir Sayed Abdul Qadir Shah Jilani Institute of Medical Sciences Gambat
[3]Assistant Professor, department of community medicine, GMMMC, (SMBBMU) Larkana
[4]Cardiologist PG trainee in NICVD Karachi
[5]Gynecologist PG trainee in Shaikh Zaid women Hospital Larkana

***Corresponding Author:** Arifa Jabeen
*Assistant professor Pediatrics Gambat Institute of medical and health sciences
Email: arifabalouch@yahoo.com

Abstract:
Background: bleeding disorder are very common in children which occurs due to multiple factors.
Objective: the aim of this study was to find out the frequencies of bleeding disorders in children presenting to pediatric Department of a Tertiary Care Hospital**.**
Material and method: The present study was carried out at the department of Pediatrics Gambat Institute of medical and health sciences from January 2018 to June 2018 after taking approval from the research committee of the hospital. A total of 116 Children and neonates (day 1 to 12) years with a suspected bleeding tendency or a family or personal history of congenital bleeding disorders were included in the study, as confirmed by the relevant laboratory tests. Parents or legal guardians gave their informed permission. A thorough clinical examination and history were documented. All patients had baseline laboratory tests performed on the same day, including complete blood counts, bleeding times, prothrombine times, APTTs, fibrinogen, and blood groupings. A vascular wall defect was thought to be the source of the bleeding if the coagulation profile was determined to be normal. Schonlen Henock A skin biopsy confirmed purpura. The frequencies and percentages of each qualitative characteristic were examined. For quantitative factors such as age, the mean plus standard deviation was computed. Tables were used to display the findings. The statistical software SPSS version 16 for Windows was used to analyze all of the data.
Results: A total 117 individuals with bleeding disorders were examined in this study .the mean age of the study participants was 06.6884 ± 4.4823 (ranged day 1 up to 12) years. Out of the 56(47.86%) patients who had clotting factor defects, the etiology of coagulative disorders revealed that 17 (14.5%) of them had hemorrhagic disease of the newborn, 15 (12.8%) had diarrhea, 5 (4.2%) had liver parenchymal disease, 5 (4.2%) had DIC, 2 (1.7%) had biliary atresia, 6 (5.1%) had hemophilia "A," 3 (2.5%) had von Wille brand disease, and 3 (2.5%) had factor VII deficiency, respectively. 52 (44.4%) patients had bleeding from thrombocytopenia, 21 (17.9%) had aplastic anemia, 11 (9.4%) had idiopathic thrombocytopenic purpura , 9 (7.5%) had acute lymphocytic leukaemia, 7 (5.9%) had megaloblastic anemia, and 4 (3.4%) had infection-induced thrombocytopenia. Nine cases (7.6%) had vessel wall defects**.**

Conclusion: The current study concluded that among bleeding disorders the most common was clotting factor deficiencies, followed by platelet and vascular wall problems, respectively.

Key words. Bleeding disorders**,** clotting factor defects, vascular wall problems

Introduction

Children with bleeding disorders experience symptoms such as severe bruising, frequent or continuous nosebleeds, and joint bleeding because these illnesses impair the blood's capacity to clot. These disorders, which result from defects in primary or secondary hemostasis, can be inherited or acquired. Defects in primary or secondary hemostasis can cause bruises with or without prior trauma, although clotting factor defects are typically the cause of severe, visible bruises. The most common cause of petechiae is a platelet or blood vessel abnormality.[1] Doctors have been treating bleeding problems since the 16th century.[2] Although congenital bleeding disorders are common around the world, little is known about their frequency and other related factors in poor nations like Pakistan. The most prevalent of them are von Wille brand disease (vWD), hemophilia A (HA), and hemophilia B (HB). Because the degree of bleeding is correlated with the degree of factor deficit, X-linked recessive illnesses, such as HA and HB diseases, are categorized as moderate, severe, and mild based on the amount of factor in the plasma.[3] A vitamin K deficit causes hemorrhagic disease of neonates (HDN), an acquired illness.[4] Because of their limited foetal storage and trans placental transfer of vitamin K, newborns are susceptible to hemorrhagic diseases.[5] Vitamin K deficiency hemorrhage in the very first week of life is currently predicted to occur in 0.01% to 0.44% of newborns who do not get vitamin K prophylaxis.[6] Between 1 in 15,000 and 1 in 20,000 babies born who were exclusively breastfed but did not get enough vitamin K at delivery have bleeding issues.[7]India and the Middle East have been shown to have higher rates of platelet functioning abnormalities.[8] One in 5000 live male births has HA (FVIII deficiency), whereas one in 30,000 has HB (FIX deficiency).[9] On the other hand, vWF malfunction or deficiency is the most prevalent bleeding condition in females, with a frequency of 1 in 1000 or more.[10] There is currently no treatment for these illnesses. The cornerstone of care for these individuals is clotting factor replacement therapy. However, factor replacement treatment increases the patient's chance of contracting HIV, HBV, and HCV infections, among other transfusion-transmitted illnesses. It also causes the creation of alloantibodies, or inhibitors, against the deficient factor. The patient eventually becomes resistant to traditional factor replacement therapy as a result of these antibodies. Inhibitors appear 20% to 30% of the time in HA and 2% to 5% of the time in HB.[11] The current study was carried out to find out the Frequencies of Bleeding Disorders in Children Presenting to Pediatric Department of a Tertiary Care Hospital

Material and method

The present study was carried out at the department of Pediatrics Gambat Institute of medical and health sciences from January 2018 to June 2018 after taking approval from the research committee of the hospital. A total of 116 Children and neonates (day 1 to 12) years with a suspected bleeding tendency or a family or personal history of congenital bleeding disorders were included in the study, as confirmed by the relevant laboratory tests while individuals taking anticoagulant medication with immune thrombocytopenic purpura, disseminated intravascular coagulation , aplastic anemia were excluded. Parents or legal guardians gave their informed permission. A thorough clinical examination and history were documented. Additionally location of the bleeding were noted the kind and location of the bleeding, the treatment, including any surgery, that was employed. All patients had baseline laboratory tests performed on the same day, including complete blood counts, bleeding times, prothrombine times, APTTs, fibrinogen, and blood groupings. On the basis of the baseline findings, additional analysis was conducted. Adsorbed plasma and serum were used in correction studies if the platelet count & bleeding time was normal but the P.T. and APTT were prolonged. As needed, clotting factor testing were conducted. A vascular wall defect was thought to be the source of the bleeding if the coagulation profile was determined to be normal. Schonlen

Henock A skin biopsy confirmed purpura. The frequencies and percentages of each qualitative characteristic were examined. For quantitative factors such as age, the mean plus standard deviation was computed. Tables were used to display the findings. The statistical software SPSS version 16 for Windows was used to analyze all of the data.

Results

A total 117 individuals with bleeding disorders were examined in this study .the mean age of the study participants was 06.6884 ± 4.4823 (ranged day 1 up to 12) years. Out of the 56(47.86%) patients who had clotting factor defects, the etiology of coagulative disorders revealed that 17 (14.5%) of them had hemorrhagic disease of the newborn, 15 (12.8%) had diarrhea, 5 (4.2%) had liver parenchymal disease, 5 (4.2%) had DIC, 2 (1.7%) had biliary atresia, 6 (5.1%) had hemophilia "A," 3 (2.5%) had von Wille brand disease , and 3 (2.5%) had factor VII deficiency, respectively. 52 (44.4%) patients had bleeding from thrombocytopenia, 21 (17.9%) had aplastic anemia, 11 (9.4%) had idiopathic thrombocytopenic purpura , 9 (7.5%) had acute lymphocytic leukemia , 7 (5.9%) had megaloblastic anemia, and 4 (3.4%) had infection-induced thrombocytopenia. Nine cases (7.6%) had vessel wall defects as presented in **table 1.**

Table 1. The frequencies of different causes of bleeding issues in the study population N=117

Clotting factor disorders	**Frequency (%)**
HDN	17 (14.5%)
Diarrhea	15(12.8%)
disease of liver parenchyma	5(4.2%)
DIC	5%(4.2%)
Biliary atresia	2 (1.7%)
Hemophilia A	6 (5.1)%
vWD	3 (2.5%)
Deficiency of Factor VII	3 (2.5%)
Bleeding due to thrombocytopenia:	
Aplastic anemia	21 (17.9%)
ITP	11 (9.4%)
ALL	9 (7.5%)
Megaloblastic anemia	7(5.9%)
Thrombocytopenia due to infection	4(3.4%)
Vessel wall defects	9(7.5%)

HDN ;Hemorrhagic disease of newborn DIC ;Disseminated intravascular coagulation
vWD ;von Willebrand disease ,ITP ;Idiopathic thrombocytopenic purpura
ALL; Acute lymphocytic leukemia

Discussion

A wide range of uncommon genetic defects and illnesses known as inherited platelet functioning disorders can cause bleeding symptoms of different severity. According to reports, autosomal recessive genes stay hidden for generations within the family and only emerge (seen phenotypically in offspring) following fresh consanguineous marriages.[12] Many individuals with bleeding problems are either misdiagnosed or not identified at all because of inadequate diagnostic facilities and understanding, even in Pakistan's largest cities.[13] In the present study a total 117 individuals with bleeding disorders were examined in this study .the mean age of the study participants was 06.6884 ± 4.4823 (ranged day 1 up to 12) years. Similar findings were found in a local research conducted in Pakistan where the mean age was 5. 35 ± 3.7 years.[14] In the present study out of 117 individuals with bleeding disorders 56(47.86%) individuals had clotting factor defects, 52 (44.4%) had bleeding from thrombocytopenia and nine cases (7.6%) had vessel wall defects. In contrast, 81 of the participants (18.6%) in a local study exhibited platelet function abnormalities, and 273 cases (62.8%) had coagulation factor insufficiency. There were 81 more (18.6 percent) with vWF insufficiency. Of the 273 individuals with coagulation factor deficit, 218 (79%), had hereditary

deficiencies, and 53 (20.0%) had acquired deficiencies in the form of multiple factor insufficiency, primarily as a result of liver illness and vitamin K deficiency.[13] According to a study on rare coagulation deficiencies by Peyvandi et al. [15], in societies with high rates of consanguineous marriages, like southern India and Muslim countries, recessively inherited coagulation disorders are so common that they can outnumber the prevalence of conditions like haemophilia B, which is a significant clinical and social issue. In a cohort of 1475 individuals with congenital bleeding disorders, Ahmed et al. [16] found that the prevalence of platelet functional problems was higher (27.77%) than that of factor IX and other uncommon coagulation diseases. Major bleeding is possible in patients with thrombocytopenia brought on by a variety of primary bone marrow disorders and neoplastic conditions.[17] Aplastic anaemia was the most prevalent bleeding disease in our research, affecting the majority of patients.(18%) According to a local research conducted at the Paediatrics Department of PGMI/LRH, Peshawar, it was likewise the most prevalent condition, occurring in 20% of patients.[14] The same findings were reported by a different recent local research, which found that 20% of instances of aplastic anaemia occurred.[18]For more than 150 years, Hemorrhagic Diseases of the Newborn—now known as Vitamin K Insufficiency Bleeding Disorder—has been acknowledged as a clinical entity. According to the current study, hemorrhagic disease of the newborn (HDN), which affected 17 individuals (14.5%), was the second most prevalent condition after aplastic anaemia. Our results are integrated into both national and international research.[19] Idiopathic or immune thrombocytopenic purpura is one of the most prevalent children acquired bleeding diseases. In 9.4% of the instances in our research, it was reported. In a similar vein, a local investigation revealed that 15.7% of patients had ITP.[14]. a prospective research found that boys had agreater incidence of acute ITP than girls (54.8% vs. 45.2%).[20] in the present study 44.4% patients had bleeding from thrombocytopenia, 17.9% had aplastic anemia, 9.4% had idiopathic thrombocytopenic purpura (ITP), 9 (7.5%) had acute lymphocytic leukemia , 7 (5.9%) had megaloblastic anemia, and 4 (3.4%) had infection-induced thrombocytopenia. Nine cases (7.6%) had vessel wall defects. Similar results were seen in the study conducted by Karim et al.[21]

Conclusion

The current study concluded that among bleeding disorders the most common was clotting factor deficiencies, followed by platelet and vascular wall problems, respectively.

References

1. Medeiros D. Bleeding disorders. In: Case based pediatrics for medical students and residents. Hawaii: Department of Pediatrics, University of Hawaii John A. Burns School of Medicine. 2003.
2. Laffan MA, Lee CA. Inherited bleeding disorders. In: Hoffbrand AV, Catovsky D, Tuddenham EGD, editors. Postgraduate haematology. 5th ed. 2005. p. 825-41.
3. Hoyer LW. Haemophilia A. N Engl J Med 1994; 330: 38-47.
4. Stanojevic M. Controversies concerning the prevention of hemorrhagic disease newborns Paediatr Today 2008; 3: 11-23.
5. Shearer MJ. Vitamin K. Lancet 1995; 345: 229-34.
6. McNinch AW, Tripp JH. Haemorrhagic disease of the newborn in the British Isles: two year prospective study. BMJ 1991; 303: 1105-9.
7. Autret-Leca E, Jonville-Béra AP. Vitamin K in neonates: how to administer, when and to whom. Paediatr Drugs 2001; 3: 1-8.
8. Ahmed F, Kannan M, Ranjan R, Bajaj J, Choudhary VP, Saxena R. Inherited platelet function disorders verses other inherited bleeding disorders: an Indian overview. Thromb Res 2008; 121: 835-41.
9. Arun B, Kessler C. Inherited Bleeding Disorders: Haemostasis and Thrombosis. 4th ed. Philadelphia: Lippincott Williams and Wilkens; 2001.

10. Margaret ER, Christopher EW, Nigel SK. Congenital bleeding disorders. Hematology ASH. 2003; 559-74. Also available from URL://http://asheducationbook. hematologylibrary.org/cgi/reprint/2003/1/559.
11. Zafar T, Ikram N, Amanat S, Zafar A, Hassan K. Clinicohaematological spectrum of Haemophilia. J Rawal Med Coll 2006; 10: 54-60.
12.Peyvandi F, Cattaneo M, Inbal A, De Moerloose P, Spreafico M. Rare bleeding disorders. Haemophilia 2008; 14: 202-10.
13.Nazir K, Ahmed S, Kamran S, Anwar J. Frequency of bleeding disorders diagnosed at Armed Forces Institute of Pathology, Rawalpindi. Pak Armed Forces Med J 2011; 61: 387-90
14.Khan A, Aqeel M, Khan TA, Munir A. Pattern of hematological diseases in hospitalized paediatric patients based on bone marrow examination. J Postgrad Med Inst 2008; 22: 196-200
15.Peyvandi F, Duga S, Akhavan S, Mannucci PM. Rare coagulation deficiencies. Haemophilia 2002; 8: 308-21.
16.Ahmed F, Kannan M, Ranjan R, Bajaj J, Choudhary VP, Saxena R. Inherited platelet function disorders versus other inherited bleeding disorders: An Indian overview. Thromb Res 2008; 121: 835-41
17.Park YB, Lee JW, Cho BS, Min WS, Cheung DY, Kim JI, et al. Incidence and etiology of overt gastrointestinal bleeding in adult patients with aplastic anemia. Dig Dis Sci 2010; 55: 73-81
18. Shah M, Khattak IUD, Khattak ST, Ahad A, Mahsud MAJ. Medical conditions associated with prolonged activated partial thromboplastin time in Swat. Gomal J Med Sci 2011; 9: 15-8
19.Committee on Fetus and Newborn, Policy Statement. Controversies concerning vitamin K and the newborn. Pediatrics 2003; 112: 191-2.
20.Kühne T, Imbach P, Bolton-Maggs PH, Berchtold W, Blanchette V, Buchanan GR, et al. Newly diagnosed idiopathic thrombocytopenic purpura in childhood: an observational study. Lancet 2001; 358: 2122-5
21. Karim R, Zaman A, Tasneem K, Syed MA, Zehra M. Frequencies of Bleeding Disorders in Children Presenting To Pediatric Department of a Tertiary Care Hospital At Peshawar. KJMS. 2013 Jul;6(2):267.