

Effect of Inherited Bleeding Disorder on Some Physiological Parameters for Patients In Karbala City of Iraq

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ABSTRACT

Background: Severe inherited bleeding disorders are most common during early childhood. Hereditary bleeding disorder refers to a group of medical conditions that cause poor blood clotting and uncontrollable bleeding.

Objective: This study aimed to report the hematological changes that may occur in patients diseased with hemophilia to help the medical team know how the body responds to treatment, and consequently decrease complications and deaths.

Methods: The current study was conducted in Maternity and Pediatrics Hospital, Karbala, Iraq. The study included 100 samples of female patients infected with hemophilia and 100 samples of healthy female children within the age range of 1-18 years.

Results: The current study indicated total red blood cells count, hemoglobin, and packed cell volume that were significantly decreased in patients with hemophilia ($p \leq 0.05$) compared to healthy children. In contrast, total white blood cell count and erythrocyte sedimentation rate were increased significantly in patients with hemophilia compared to healthy children ($p \leq 0.05$).

Conclusion: Changes in hematological parameters for patients infected with hemophilia occur as a result of frequent bleeding, which is the primary sign of hemophilia.

Keywords: Children, Hemophilia, Total red blood cell counts, Hemoglobin.

INTRODUCTION

Hereditary bleeding disorder refers to a group of medical conditions that cause poor blood clotting and uncontrollable bleeding ⁽¹⁾. Coagulation factor deficits and platelet abnormalities are two types of hereditary bleeding diseases ⁽²⁾.

Hemophilia A, hemophilia B (Christmas disease), and von Willebrand's disease are the most common inherited coagulation deficiencies ^(2, 3).

Factor VIII deficit (Hemophilia A) accounts for 85% of instances, while Factor IX deficiency (Hemophilia B) accounts for 15% of cases ^(4, 5). Of note, in case a child inherits a factor deficiency from only one parent, the child will be a carrier of the disease, even if he or she does not show symptoms ^(3, 6).

Bernard Soliuer syndrome is a rare autosomal recessive bleeding illness characterized by a lack of platelet glycoprotein Ib (GpIb), the receptor for von Willbrand factor, which plays a role in clotting ^(7, 8). Severe inherited bleeding disorders are most common during early childhood. Circumcision bleeding, umbilical stump bleeding, cephalohematomas, and subgaleal hemorrhages after delivery are all common symptoms of bleeding diseases in healthy infants. As a result, these indicators should always be considered with a high level of skepticism ⁽⁹⁾.

MATERIALS AND METHODS

Study groups and blood samples collection

This study was conducted in the Maternity and Pediatrics Hospital, Karbala, Iraq.

The study groups included 100 patients diseased with hemophilia and 100 healthy children within the age range of 1-18 years. To test blood parameters, 5 ml of

blood samples were taken from the study groups, of which 2 ml were used for the erythrocyte sedimentation rate (ESR) test, and 3 ml were put in an anticoagulant tube.

Blood parameters tests

An automated hematoanalyzer was used to measure blood parameters (Sysmex xp300 B1269). Packed cell volume (PCV), hemoglobin concentration (Hb), total red blood cell count, and total white blood cell count were among the blood parameters examined.

Measurement of ESR

To conduct the study, a Westergren tube with a length of 300 mm (open on both ends) and a diameter of 2.5 mm was used. Then, 2 mL of blood and 0.5 mL of anticoagulant (sodium citrate) were added.

The mixture was pulled up to the zero mark into a Westergren tube, which was then positioned upright in a stand with a spring clip on top and rubber on the bottom. After an hour, the level of the top of the red cell column was measured ⁽¹⁰⁾.

Ethical Approval:

The study was approved by the Ethics Committee of Alsafwa University. Participants were informed about the study, experiments and publications.

This work has been carried out in accordance with The Code of Ethics of the World Medical Association in terms of "Declaration of Helsinki". The participants' parents signed informed written consents.

Statistical analysis

The results were presented as mean ± SE and subjected to a one-way analysis of variance using IBM SPSS Program (version 20). LSD post-hoc was utilized to specify the significant difference between means in the post hoc test ⁽¹¹⁾. P value ≤ 0.05 is considered significant.

RESULTS AND DISCUSSION

The obtained results of the current study indicated that packed RBCs volume, RBCs' count and hemoglobin were significantly lower in hemophilia patients than in their healthy counterparts (Table 1).

Table (1): Effect of hemophilia on red blood cells, hemoglobin concentration, and Packed cell volume (Means ± SE)

Parameters Groups	PCV value (%)	Hb g/dl	RBC ×10 ⁶ /cell/mm ³
Patient persons	A 24.70 ± 1.96	A 8.00 ± 1.00	A 1.50 ± 0.50
Healthy persons	B 43.30 ± 0.59	B 12.50 ± 0.50	B 4.50 ± 0.50

N=100, different letters in a column represent a significant difference at p ≤ 0. 05

Table (2) showed that the total WBCs count was significantly increased in patients' group compared to healthy children. The ESR means were significantly increased in patients' group compared to healthy individuals.

Table 2. Effect of hemophilia on white blood cells and ESR (Means ± SE)

Parameters Groups	WBCs ×10 ³ /mm ³	ESR mm/hr
Patient persons	A 11.00 ± 18.00	A 33.33 ± 1.54
Healthy persons	B 8.00 ± 2.00	B 5.87 ± 1.03

N=100, different letters in a column represent a significant difference at p ≤ 0. 05

DISCUSSION

The presence or absence of anemia or evidence of blood regeneration depends on the severity and frequency of bleeding. Basic hematological investigations have indicated nothing specific for common inherited bleeding diseases ⁽¹²⁾. The obtained results of the current study indicated that RBCs were significantly lower in hemophilia patients than in their healthy counterparts. Regarding RBCs' count, the

obtained results are consistent with a previous study ⁽¹³⁾ indicating that RBCs' count was lowered as a result of recurrent spontaneous bleeding and persistent bleeding, even after minor injuries. The severity of the disease determines the frequency of bleeding; in the severe form of the disease, recurrent bleeding can lead to anemia and various transfusion problems, whereas in the mild form of the disease, there is less bleeding, and hence fewer difficulties ^(3, 14).

Hb concentration means significantly decreased in patients with hemophilia, compared to healthy participants. This study agrees with another study ⁽¹⁵⁾ that revealed that hemoglobin concentrations decreased in patients with hemophilia due to recurrent unavoidable blood loss. The decrease in packed cell volume in the current study was due to the reduction in RBCs' count and Hb concentration due to the recurrent bleeding episodes, which is the most important manifestation of the disease ⁽¹⁶⁾.

For patients with hemophilia, the total WBCs' count was significantly increased compared to healthy children. Similarly, another study ⁽¹⁷⁾ indicated a highly significant leukocytosis in hemophilic patients with arthropathy. In the same vein, it was found that hemophilic arthropathy could be considered as the most critical complication of hemophilia, especially in its severe form, so as the WBCs count elevated due to the inflammation of the joint ⁽⁴⁾. In addition, hemophilic patients are prone to frequent infections due to their frequent hospitalization as well as the transfusion of plasma and cryoprecipitate. Moreover, bone marrow reacts to blood loss, and neutrophilia may accompany significant bleeding, as in other cases of posthemorrhagic anemia ⁽¹²⁾.

The ESR means significantly increased in patients infected with hemophilia, compared to healthy individuals. These results are consistent with another study revealing a highly significant increase in ESR in patients with hemophilic arthropathy ⁽¹⁷⁾. The findings of the current study were also supported by another study indicating that patients with hemophilic arthropathy have all the signs and symptoms of inflammation, including high WBCs and ESR ⁽¹⁸⁾.

CONCLUSION

In conclusion, changes in hematological parameters for patients infected with hemophilia occur as a result of frequent bleeding, which is the primary sign of hemophilia. Present study has been conducted in region of Iraq. Further studies are suggested for different regions.

Consent for Publication:

I confirm that all authors accepted the manuscript for submission.

Competing interests: None

Funding: None

Conflicts of Interest: The authors declared no conflicts of interest

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