Glanzmann's Thrombasthenia: Epistaxis Treatment and Outcomes in Children

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ABSTRACT

Background: Glanzmann's Thrombasthenia (GT) is a rare inherited blood clotting (coagulation) disorder characterized by the impaired function of specialized cells (platelets) which are essential for proper blood clotting. **Objectives:** The present study aimed to assess the management steps of epistaxis in children with GM disease and assess the clinical outcome and the success rate or field of management.

Methods: Data was taken via scanning all the patients' files in ENT clinic and 4 children with GM were identified as receiving therapy. Furthermore, data collected from each file were primary complaint needing a duration of hospitalization, intensive care unit (ICU) admission, coagulation management, hematologic results, medical therapy administration, blood products transfusion and the surgical intervention performed.

Results: With a total of 25 hospital presentations, the highest percentage of the present symptoms was for epistaxis with 68% [(17)], then 16% [(4)] for gingival bleeding and 8% for each of menorrhagia and hematemesis.

Conclusion and recommendation: Our study carried out the most effective technique that control epistaxis is Fresh Frozen (FFP) Plasma. The epistaxis in GT should be concern as sever threating life condition and should be managed with effective treatment.

Keywords: Glanzmann's Thrombasthenia, epistaxis, disorder.

INTRODUCTION

Glanzmann's Thrombasthenia (GT) is a rare inherited blood clotting (coagulation) disease categorized by the impaired function of specialized cells (platelets) which are important for appropriate blood clotting. Prolonged untreated or unsuccessfully treated hemorrhage can be life threatening [(2)]. Glanzmann thrombasthenia affects males and females at the same stage [(2)].

There was a study published in 2006; showed that its incidence has been increased in consanguinity populations [(9)]. Awidi reported that it was the second most frequent bleeding disorder in Jordan [(8)]. There is difficulty to be distinguished from other congenital platelet function defects on clinical ground alone [(10)]. It is a severe autosomal recessive inherited disease which is characterized by failure of platelet aggregation in response to adenosine diphosphate (ADP), epinephrine, collagen, and thrombin and by absent or diminished clot retraction [(1)]. The Symptoms are usually observed at birth (neonates) or even during infancy stage; where 300 cases have been stated [(2)].

It usually contains abnormal bleeding, which could be severe **[(2)].**

The most common clinical complained is epistaxis episodes and some of them may have sever bleeding after circumcision, surgery or dental extraction [(1)]. Also, may they have other clinical manifestations which included: easy bruising, purpura, gingival bleeding, menorrhagia and less frequently gastrointestinal bleeding, hematuria, hemarthrosis, muscle hematoma and central nervous system bleeding [(1)]. The Management of epistaxis usually with nasal packing or application of foam soaked in thrombin. It was stated that a recombinant aspect VIIa product was agreed to treat bleeding episodes or per operational managing when platelet transfusions through or even without antibodies to platelets are not operative. On the other hand, treatment that is considered as symptomatic and supportive as consistent dental repair is an important role to avoid bleeding from the gums. Also, hormonal rehabilitation can be used to suppress menstrual stages. On other hand, genetic counseling benefits people with GT and their families **[(2)].** GT is considered a severe hemorrhagic disorder; moreover, the prognosis is outstanding with careful supportive care [(9)].

OBJECTIVE

The present study aimed to assess the management steps of epistaxis in children with GM disease and assess the clinical outcome and the success rate or field of management.

MATERIALS AND METHODS

Data was taken via scanning all the patients' files in ENT clinic and 4 children with GM were identified as receiving therapy. Furthermore, data was collected from each file were primary

Received: 11/12/2017
Accepted: 21/12/2017
DOI: 10.12816/0044344

complaint needs a duration of hospitalization, intensive care unit (ICU) admission, coagulation management, hematologic results, medical therapy administration (i.e., antifibrinolytics, factor VII), blood products transfusion (i.e., platelets, packed red blood cells), and the surgical intervention performed. We divided any surgical intervention to categories according to the general nature of the intervention. However, procedures using nasal packing or any other materials as tamponade to stop nasal hemorrhage are considered.

Otherwise, it was classified according to their placement, either anterior, posterior, or both. A separate two categories were created for using factor VII and FFP. Also, there was category, such as surgical interventional. The failure or success was identified by the control of nasal hemorrhage and/or preventing the need for orotracheal intubation or use other maneuver. The small number of patients lead to preclude the analysis of statistical significance.

Inclusion criteria

- The age is younger than 14 years old.
- Genetic studies that confirm the GM disease.
- Follow up during treatment at KKH.

Exclusion criteria

- The age is elder than 14 years.
- Epistaxis's history with no genetics studies.

Ethical consideration

Ethical approval was obtained to perform a retrospective chart review of all patients with Glanzmann's thrombasthenia treated at King Khaled Hospital. **The study was done after approval of ethical board of Hail university.**

RESULTS

Our study included four cases of GM disease, who fulfilled the inclusion and exclusion criteria, with total of 25 hospital presentations. The highest percentage of the present symptoms was for epistaxis with 68% (17), then 16% (4) for gingival bleeding and 8% for each of menorrhagia (2) and hematemesis (2). Moreover, none of the patients presented with melena, purpura or bloody otorrhea (table. 1). Table (2) is showing the medical visits for hemorrhage in children with Glanzmann's thrombasthenia and demonstrating the degree of their severity was indicated by their high number of hospital admissions.

According to the management of epistaxis, we found that anterior nasal packing (ANP) procedure was applied 14 times, failure 11 times, success 3 times with failure rate of 78.57% which was considered to be the highest. Whilst, failure rate of FFP was 0%, which was used 9 times. Other procedures and their failure rate were shown in table (3). Moreover, table (4) showed that some of the admissions were just to receive FFP only or with Packed RBC. Actually, FFP was received 10 times; however, one of them was only FPP and 9 of them were after management filed.

TABLE (1): Bleeding in Children with Glanzmann's thrombasthenia.

Symptom	No. of Presentations	Frequency (%)
Epistaxis	17	68
Menorrhagia	2	8
Hematemesis	2	8
Melena	0	0
Gingival bleeding	4	16
Purpura	0	0
Bloody otorrhea	0	0

TABLE (2): Medical visits for hemorrhage in children with Glanzmann's thrombasthenia demonstrating their severity by their high number of hospital admissions.

Patient	Medical Visits for Any Form of Hemorrhage	Medical Visits for Epistaxis	No. of Hospital Admissions for Any Form of Hemorrhage	No. of Hospital Admissions for Epistaxis	Days Hospitalized	No. of ICU Admissions
1	10	8	10	8	5	0
2	9	6	9	5	3	1
3	4	2	4	2	3	1
4	2	1	2	1	2	0
Total	25	17	25	16	13	2

TABLE (3): Techniques used to control nasal Hemorrhage in children with Glanzmann's thrombasthenia and their rate of failure

Technique	No. of Procedures	No. of Failures	Rate of Failure (%)
Factor IIV	16	10	62.5%
Anterior packing	14	11	78.57%
posterior packing	1	0	0
FFB(Platelet)	9	0	0
Cautery with packing, either anterior or posterior	0	0	0
Total	40	21	52.5%

TABLE (4): Frequency of transfusion

FFP transfusion	10
Packed RBC transfusion	5
FFP &Packed RBC transfusion	3

DISCUSSION

Epistaxis is common in children, affecting 30% of children especially aged from 0 to 5 years old, 56% of children aged from 6 to 10 years old, and 64% of children 11 to 15 years old [11,12]. Most epistaxis is a self-limited, respond to symptomatic therapy, and requires no medical attention. Equally important, in children with GT, epistaxis is more frequent and severe and require multidisciplinary management [13,14]. In our study, four patients were diagnosed with Glanzmann's thrombasthenia, three were males and one was female and all of them were Saudi. Furthermore, epistaxis is the most common symptom with the rate of 68% (17), compared to **Bashawri *et al***.[10].

Other study conducted that epistaxis was 52% (16) [13]. On the other hand, there was another study reported (77%) epistaxis [15]. Other symptoms in our study, indicated the following; 16% (4) for gingival bleeding, 8% for each of menorrhagia and hematemesis and 0% for each melena, purpura and bloody otorrhea. While in **Bashawri *et al*.[10]** study, it was 11 (35.5%) for menorrhagia, gum bleeding 10 (32.3%), bruises 7 (22.6%), bleeding at circumcision 4 (13%), hemarthrosis 4 (13%), ecchymosis and petechial rash 5 (16.1%), gastrointestinal (GIT) bleeding 7 (23%), hematuria 2 (7%), delayed wound healing 1 (3%), bleeding with tooth eruption 1 (3%) and hemoptysis 1 (3%) [10]. While other study reported that gingival bleeding was highest presentation symptoms which was (81%) and easy bruising (55%), hematoma 14%, gastrointestinal bleeding 11%, post-dental extraction bleeding 14%, hematuria 3% and hemarthrosis 3%[15].

In our cohort study, the total medical visits of any hemorrhage and required admission were 25, which were 17 visits for epistaxis; a total of 13 days spent as inpatient (Table. 2). However, 16 visits from these 17 was required admission where 2 of them were admitted to ICU. In comparison with other study which was done in Minnesota, 52 visits for epistaxis of total medical visits which were 63[3]. Hospitalization was required in 47/63 of these visits for a total of 281 days spent as an inpatient, ICU admission was required in 20 of these hospitalizations [3].

There was no mortalities observed in patients. During period of analysis, we found the success technique that control epistaxis was fresh froze plasma (FFP); which was 100%. On the other hand, Minnesota's study reported that limited septoplasty technique success was 100% to control epistaxis [3]. Our study showed 52.5% as a total failure rate of all techniques that are used to control epistaxis, anterior packing was 78.57% which was the highest failure rate. Furthermore, VII factor was 62.5% of failure. Compared to Minnesota's results, 100% failure rate endovascular embolization which was the highest rate of failure then 91% of anterior and posterior packing with thrombotic agent and 50% of failure for each Collagen thrombotic agent only and Cautery with packing, either anterior or posterior [3].

CONCLUSION

Our study reported that the most effective technique that control epistaxis is Fresh Froze Plasma. The epistaxis in GT should be concern as

sever life threating condition and should be managed with effective treatment.

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