



CODEN [USA]: IAJPBB

ISSN: 2349-7750

**INDO AMERICAN JOURNAL OF  
PHARMACEUTICAL SCIENCES**<http://doi.org/10.5281/zenodo.1087450>Available online at: <http://www.iajps.com>

Case Report

**A CASE REPORT ON GLANZMANN'S THROMBASTHENIA**  
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*Glanzmann's thrombasthenia (GT) is a rare autosomal recessive disorder in which the platelet glycoprotein IIb/IIIa (GP IIb/IIIa) complex is either deficient or, dysfunctional. The incidence is about 1 in 1,000,000. It is more common in populations where marriage between blood relatives is common. The signs of GT occur early in life and include easy bruising, epistaxis and prolonged bleeding from minor injuries. Epistaxis, menorrhagia, postpartum bleeding and surgical bleeding can be life-threatening. Here we are presenting a case of 4 year old female child with recurrent epistaxis and gingival bleeding for past one year. Diagnosis associates prolonged bleeding time with absent platelet aggregation in response to all physiological stimuli except ristocetin, with normal platelet count and morphology. Coagulation tests such as prothrombin time and partial thromboplastin time are normal. The cure for the disease does not exist; the only effective therapy consists of transfusions of fresh platelets or platelet concentrates. With proper supportive care Glanzmann's thrombasthenia has a very good prognosis.*

**Keywords:** Glanzmann's thrombasthenia, Platelet disorder, Epistaxis, Glycoprotein IIb/IIIa (GP IIb/IIIa) complex, gingival bleeding, Platelet aggregation.

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Please cite this article in press as Annamol Joseph et al., *A Case Report on Glanzmann's Thrombasthenia*, Indo Am. J. P. Sci, 2017; 4(12).

**INTRODUCTION:**

Glanzmann's Thrombasthenia (GT) is a rare genetic platelet disorder, with an incidence of 1 in 1 million, in which the platelet glycoprotein IIb/IIIa (GP IIb/IIIa) complex is either deficient or dysfunctional [1]. It is characterized by prolonged bleeding time, diminished clot retraction [3] and usually seen in populations with an increased consanguinity with an autosomal recessive pattern of inheritance. The signs of GT occur early in life and include easy bruising, epistaxis and prolonged bleeding from minor injuries. Epistaxis, menorrhagia, postpartum bleeding and surgical bleeding can be life-threatening [2,4].

At a molecular level, GT is a heterogeneous condition with multiple deletions and mutations of the genes encoding the  $\alpha$ IIb $\beta$ 3 integrin. The genes of both these platelet glycoprotein  $\alpha$ IIb $\beta$ 3 complex are on chromosome 17. The ITGA2B gene code for the  $\alpha$ IIb and the ITGB3 codes for the  $\beta$ 3. A defect in this glycoprotein can lead to the bleeding disorder. The majority of the patients have a normal platelet size

and count. Platelet glycoprotein  $\alpha$ IIb $\beta$ 3 complex levels of <5 % leads to higher bleeding tendencies, called as Type 1, and 10-20 % levels of the platelet glycoprotein  $\alpha$ IIb $\beta$ 3 complex lead to mild to moderate bleeding events (Type 2) [5].

**CASE REPORT:**

A 4 year old baby girl was admitted in a paediatric ward at tertiary care hospital with the complaints of fever and headache for two days. During hospitalisation, one episode of epistaxis was found and based on the history taken from her parents she had a history of recurrent epistaxis and gum bleeding since 3 years of age. She also had history of prolonged bleeding from minor cuts and haematuria. She did not have history of bleeding from any other site and so far not received any transfusion. She is the first sibling born of second degree consanguineous marriage and there is no family history of any bleeding diathesis. On examination she was found to be pallor and stunted growth.

**Table No.1: Laboratory investigation chart**

SL.NO	Investigations Done	Lab Results			
		Day- 1	Day - 2	Day - 3	Day - 4
1	Haemoglobin (g/dl)	4.4	4.5	6.8	8.0
2	RBC ( $\times 10^{12}/L$ )	3.46	3.62	4.24	4.33
3	HCT (%)	20.1	21.3	27.8	29.7
4	WBC ( $\times 10^9/L$ )	3.9	5.4	8.3	11.9
5	Platelets ( $\times 10^9/L$ )	151	151	159	192
6	MCV (fl)	77.1			
7	Differential count (%)	N-86, L-10, E-3, B-1			
8	Bleeding time	>15 Mins			
9	Prothrombin time	Cont :10.3 secs PT :11.1 secs INR : 1.063			
10	aPTT	Cont : 30.4 secs PT : 24 secs			
11	TT	Cont : 14.5 secs PT : 12.5 secs			
12	Fibrinogen (mg/dl)	266			

Based on investigations she was found to have normal platelet count and morphology with a normal aPTT, PT, TT and fibrinogen levels. On aggregometry study RIPA (Ristocetin Induced Platelet Aggregation), there was an absent response with ADP, Epinephrine and Collagen but a normal response to Ristocetin 1.5mg/ml. The clot retraction was also present for this patient. The immunophenotyping of platelet surface glycoprotein receptor GpIIb/IIIa suggests marginally reduced CD41 and CD61 on flow cytometry.

#### TREATMENT GIVEN:

Patient was treated with T.Paracetamol 500mg TDS and nasal oxygen (4L/min) for three days. Since she had an episode of epistaxis, she was treated with adrenaline, Tranexamic acid nasal packing (anterior and posterior) and transfused to have one unit of platelet concentrate. She had transfused with whole blood on day 1 and 3 for her altered haematological values. Gradually the blood counts were increased.

Patient did not have any further bleeding episodes in next two days and was discharged with advice that the patient has to avoid any strenuous and physical activity that may lead to bleeding, she was also advised not to take any NSAIDs like Aspirin, to use soft bristle tooth brush and maintain oral hygiene. Prognosis was explained to her parents that as age advances the disease severity may decrease.

#### DISCUSSION:

GT is a hereditary platelet dysfunction due to quantitative or qualitative defect in the platelet glycoprotein  $\alpha$ IIb/ $\beta$ 3 the major integrin complex which leads to inability of platelet aggregation by attachment to fibrinogen, leading to non-formation of platelet plug, and thus excessive, apparently spontaneous bleeding. The common features of GT are bruising, epistaxis, gingival haemorrhage and menorrhagia. Bruising typically occurs after minor trauma [1,6].

Typical laboratory tests of patients with GT include a normal platelet count with prolonged bleeding time, a defective clot retraction and failure of platelets to aggregate in response to ADP, epinephrine, collagen, thrombin or arachidonic acid, 5hydroxytryptamine; but aggregation occurs normally with ristocetin; all of which stood true for this patient. Coagulation tests such as prothrombin time and partial thromboplastin time are normal. The findings of markedly prolonged bleeding time with typical aggregometry findings marginally reduced CD41 and CD61 are suggestive of GT. However because of good clot retraction she is probably a case of type 2 GT.

Here the patient is treated with packed cell transfusion and tranexamic acid nasal pack which helped to stop epistaxis. Advised to have good oral hygiene and regular dental visits to prevent gingival bleeding. Drugs that affect platelet function, such as NSAIDs or aspirin, should be avoided. Immunizations for hepatitis B should be given due to the infectious risks of frequent transfusion [7]. Treatment with Novo Seven RT, a recombinant coagulation factor VIIa, was newly approved by FDA for GT has been explained. This medication is indicated to treat bleeding episodes for who have a decreased or absent response to platelet transfusions.

#### ABBREVIATIONS

- GT – Glanzmann's Thrombasthenia
- MCV – Mean Corpuscular Volume
- N – Neutrophil
- L – Lymphocyte
- B – Basophil
- E – Eosinophil
- aPTT – Activated Partial Thromboplastin Time.
- TT – Thrombin Time
- RIPA – Ristocetin Induced Platelet Aggregation
- PT – Prothrombin Time
- ADP – Adenosine Diphosphate
- NSAIDS – Non Steroidal Anti-Inflammatory Drugs
- FDA – Food and Drug Administration

#### CONCLUSION:

The cure for the disease does not exist; the only effective therapy consists of transfusions of fresh platelets or platelet concentrates. Since Glanzmann's Thrombasthenia is a rare blood disorder, physician should aware about the significant clinical diagnosis of this disorder as compared with others. With proper supportive care GT has a very good prognosis.

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