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Glanzmann's Thrombasthenia

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Objectives: To study the clinical presentation and laboratory findings of Glanzmann's thrombasthenia (GT).

Methods: This retrospective study was carried out from January 1st 1983 to 31st December, 2003. The records of the coagulation laboratory, Hematology clinic and medical records department were reviewed. Clinical data and family history were recorded. Laboratory investigations done included; complete blood count (CBC) peripheral blood smear (PBS), bleeding time (BT), activated partial thromboplastin time (APTT), prothrombin time (PT), clot retraction, platelet aggregation and in some patients flow cytometric analysis of platelet glycoproteins was carried out.

Results: Thirty-one patients were diagnosed with Glanzmann's thromboasthemia. Seventeen were males and 14 were females. All were Saudi patients (most from eastern province and the southern part of the Kingdom) except for one Sudanese male patient. The mean age of patients was 26 ± 12.34 years. The oldest was 71 years and the youngest 20 days. Many patients had bleeding from more than one site. The clinical presentations are as follows: epistaxis 16 (52%), menorrhagia 11 (35.5%), gum bleeding 10 (32.3%), bruises 7 (22.6%),

bleeding at circumcision 4 (13%), hemoarthrosis 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%).

Thirty patients had a prolonged bleeding time, all 31 patients had normal APTT and PT. Thirteen patients (42%) had poor clot retraction, 12 (39%) no clot retraction and 6 (19%) had normal clot retraction. All patients had a marked reduction or absent aggregation to platelet agonists; adenosine diphosphate (ADP), collagen, epinephrine and arachidonic acid. Thirteen patients also had a reduction in aggregation with ristocetin. Flow cytometric analysis done on some patients showed a reduction in platelet membrane glycoproteins 41 and 62 (CD41, CD62).

Conclusion: In spite of the fact that GT is a rare disease worldwide, the situation might be different in our country. The spectrum of clinical presentation and complications in patients with GT appears to be wide, in addition to different platelet aggregation patterns i.e. reduction in aggregation with ristocetin. Still more studies are needed to clarify the pattern in Saudi patients and to raise awareness hoping to help in early recognition, presentation, and more appropriate

management. Extensive collaborated studies are needed to predict the true incidence of hereditary bleeding disorders including GT among the Saudi population, as well as a national registry for these disorders.