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Managing a Pregnant Woman with Glanzmann's Thrombasthenia: A Clinical Challenge.

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Introduction: Glanzmann's thrombasthenia is a rare bleeding disorder, in which platelets lack glycoprotein 11b/111a, but with normal morphology and count. It has an autosomal recessive pattern of inheritance or can be due to mutations. Parturition with this condition poses a risk of severe early and delayed postpartum hemorrhage and often necessitates hysterectomy. Bleeding is managed by platelet and blood transfusions. There are limited data on the management of pregnancy, delivery and postpartum bleeding in women with Glanzmann's thrombasthenia.

Case presentation: A 23 year old diagnosed patient of Glanzmann's thrombasthenia since childhood was referred for specialized management of her ongoing pregnancy and delivery. Her main symptoms since childhood were mucocutaneous bleeding and menorrhagia. Along with iron therapy she had regular blood transfusions due to anaemia. She was transferred from a district hospital at 21 weeks gestation with haemoglobin of 5.2 g/dl. Her haemoglobin was optimized with blood transfusions and was followed up until admission at 36week. She went in to spontaneous labour at 37 weeks and 5 days and delivered vaginally a 3.05kg baby. During labour, she was transfused with 12 units of apheresis platelets. She developed postpartum haemorrhage, with an estimated blood loss of 1200 ml. Her PPH was intensively managed with blood and platelet transfusion, syntocinon infusion, intravenous tranexamic acid and recombinant human coagulation Factor VIIa. In addition an intrauterine balloon tamponade was inserted which achieved a speedy reduction in blood loss. Over the next three days she was transfused with26units of platelets, blood and oral tranexamic acid.

Discussions: Minimal data are available with regards to the management of Glanzmann's thrombasthenia complicating pregnancy. Management of our patient's postpartum haemorrhage was a clinical challenge and involved multi therapeutic actions. In addition to the use of known modalities of treatment, the use of intrauterine balloon tamponade may be of additional benefit even in the absence of uterine atony as it also helps in endometrial balloon interface interaction, alterations of vascular flow patterns secondary to myometrial stretching and occlusive effect secondary to uterine wall attenuation.