EP.0807 obstetric haemorrhage in a 25-year-old primigravida with Noonan syndrome

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Objective

- To raise awareness of the increased bleeding risk associated with Noonan syndrome (NS), so that it can be prepared for and appropriately managed during the intrapartum period.

Case Report

- 25-year-old female with NS (diagnosed in childhood) and associated mild pulmonary stenosis. No other medical conditions, regular medications, or previous surgery.
- Antenatal care was provided by the multidisciplinary team, including anaesthetists and cardiologists. The plan included a FBC on admission, use of haemostatic medications, and that blood loss should be considered alongside the patient's short stature.
- Spontaneous labour occurred at term. It was a quick and uneventful labour until suspected fetal distress resulted in a kiwi ventouse delivery by easy lift out with an episiotomy, which led to a massive obstetric haemorrhage.
  - She was given 3x RBCs, 2x FFP and 2x platelets, an oxytocin infusion, tranexamic acid and syntocinon.
  - The wound was sutured and packed in the room and no further intervention was required.
- She recovered well with a satisfactory postnatal echo prior to discharge.
- Antenatally, a history of excessive bleeding was never reported, however, postnatally, the patient disclosed that it took a long time for cuts and grazes to stop bleeding and a history of menorrhagia.

References - 1. Artoni A, Selicorni A, Passamonti SM, Lecchi A, Bucciarelli P, Cerutti M, Cianci P, Gianniello F, Martinelli I. Hemostatic abnormalities in Noonan syndrome. Pediatrics. 2014 May;133(5):e1299-304. doi:10.1542/peds.2013-3251. PMID:24753526.

Discussion

- NS is an autosomal dominant condition which effects between one in 1000 to 2500.
- Not only are there characteristic features such as short stature, typical facial features, skeletal and cardiac abnormalities, it has also been found that approximately 40% of those with NS have bleeding diathesis (1).
  - The three main reasons for this are: clotting factor deficiencies, Von Willebrand disease and abnormal platelet count/ function.
  - In our case, investigations for bleeding diathesis had never been carried out. Her platelet counts, aggregation and coagulation profiles were always within normal range.

Conclusions

- It is important to recognise the association between NS and bleeding diathesis. Understanding the increased bleeding risk allows planning and improved intrapartum management.
- Screening for bleeding disorders should be carried out prenatally, however, optimising haemoglobin level, taking a bleeding history and involving haematologists are also essential to antenatal care.
- If a deficiency is identified, specific clotting factors can be ready in preparation of a bleed.

