FACTSHEET #8 BLOOD CLOTTING



The most common blood disorders are abnormalities of bleeding caused by coagulation defects.

A history of abnormal bruising or bleeding is common in people with Noonan syndrome, and some may have deficiencies of clotting factors in the coagulation pathway that leads to appropriate clotting and stopping blood loss. The reason for this is unclear.

A study of 72 people with Noonan syndrome found:

- 65% had a history of abnormal bruising or bleeding.
- 40% took longer for blood clots to form.
- 50% had a single deficiency in the coagulation pathway
- 17% had multiple deficiencies in the coagulation pathway

People with Noonan syndrome have also been found to have von-Willebrand disease - another condition where blood clots do not form correctly - and disorders related to platelets, which are cells in the blood which clump together at the start of clotting.

Although problems rarely occur, before an operation it is important that the surgical team is aware of the potential to bleed. This should lead to a careful coagulation scree before major surgery and in the case of minor day surgery the child may be kept overnight or at least until it is clear that any post operative bleeding has stopped.

REFERENCES/FURTHER READING

Sharland M, Patton MA, Talbot S, Chitolie A, Bevan DH. Coagulation-factor deficiencies and abnormal bleeding in Noonan's syndrome. Lancet. 1992 Jan 4;339(8784):19-21.

Linglart L, Gelb BD. Congenital heart defects in Noonan syndrome: Diagnosis, management, and treatment. Am J Med Genet C Semin Med Genet. 2020;184(1):73-80.