AHCDC Rare Inherited Bleeding Disorders Committee
Annual Report – May, 2019

Rare Inherited Bleeding Disorders (RIBD) Committee Members
Drs. John Wu and Margaret Rand (Co-Chairs), Victor Blanchette, Stephanie Cloutier, Sara Israels, Walter Kahr, Man-Chiu Poon, MacGregor Steele, Irwin Walker and Rochelle Winikoff (Nurse Representative: Ms. Vanessa Bouskill)

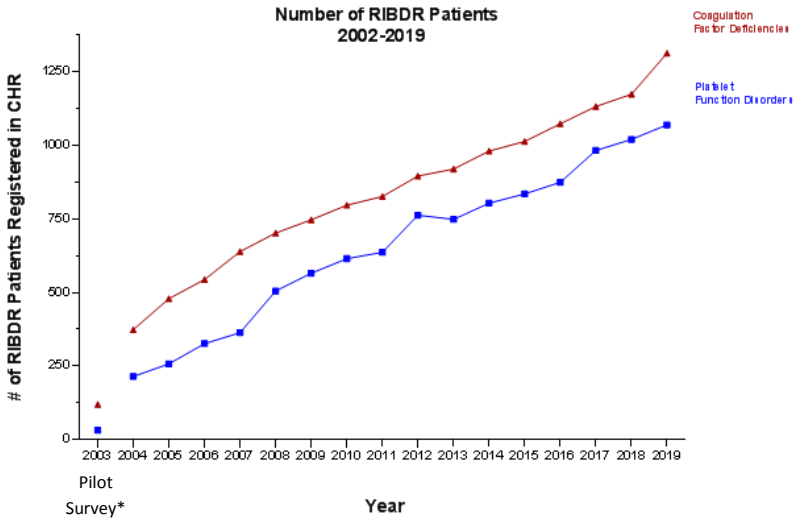
RIBD Data Manager
Ms. Taneya Hossain

2018-2019

The Rare Inherited Bleeding Disorders Registry (RIBDR), established in 2003, tracks the numbers of patients with **deficiencies of the rare coagulation factors** *(fibrinogen, factor II, factor V, factor VII, factor X, factor XI, factor XIII, combined deficiencies of factors V and VIII, or combined deficiencies of the vitamin K-dependent coagulation factors [factors II, VII, IX and X])* and with **platelet function disorders** *(including deficiencies of membrane glycoproteins, disorders of storage granules, and familial thrombocytopenias)*.

Thanks to the continued support of all the Centres across Canada, our national registry remains a tremendous success. As of April 2019, there are 2,474 patients registered, 1,393 with coagulation factor deficiencies and 1,081 with platelet function disorders (see figure below); this is an 8% increase over the 2018 numbers. Of the patients with coagulation factor deficiencies, 13% have dys- or hypo-fibrinogenemia; 1% has factor II deficiency; 7% have factor V deficiency; 34% have factor VII deficiency; 3% have factor X deficiency; 37% have factor XI deficiency; 4% have factor XIII deficiency; and 0.2% has combined factor V/factor VIII deficiency. Of the patients with platelet function disorders, 10% have deficiencies of membrane glycoproteins (mainly Glanzmann thrombasthenia and Bernard Soulier syndrome); 15% have disorders of storage granules; 10% have familial thrombocytopenia; and 65% are in the "other" category, with most having an undefined platelet disorder.

The Canadian Bleeding Disorders Registry (CBDR) required adjustment to capture the rare disorders; working with the CBDR Project Manager Arun Keepanasserril and his team and the Australian National Blood Authority in this past year, we now have all of the rare inherited bleeding disorders identified under diagnoses in the CBDR, to allow their registration and reporting as has been possible with the RIBDR. Moving forward, we will be reconciling RIBDR and CBDR numbers to ensure that the Canadian rare disorders become fully captured into the latter.



*Factor VII or XIII deficient patients and patients with Glanzmann thrombasthenia only