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ABSTRACT N. PO41

COMPARISON OF THE HAEMORRHAGIC PHENOTYPE BETWEEN FEMALE AND MALE PATIENTS AFFECTED BY AUTOSOMAL INHERITED BLEEDING DISORDERS IN A SINGLE-CENTRE COHORT.

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Introduction and Objective. Inherited bleeding disorders (IBDs) are a heterogeneous group of coagulation disorders characterised by significant variability in the frequency and severity of haemorrhagic symptoms, with either autosomal or sex-linked (X-linked) inheritance. Multicenter studies, able to gather large populations of patients but burdened by significant heterogeneity, highlighted gender differences in disease incidence and clinical phenotypes. In this study we focused on the autosomal IBDs, validating these findings and exploring gender disparity across haemorrhagic phenotypes in a homogeneous single-centre case series.

Material and Methods. We retrospectively analysed 148 patients with von Willebrand disease (vWD), rare bleeding disorders (RBDs) and inherited platelets function disorders (IPFDs), referred to the Simple Interdepartmental Structure "Presa in Carico Malattie Emorragiche e Tromboemboliche della Romagna, Cesena, Italy", and compared diagnosis characteristics [age at diagnosis, reason for referral, Bleeding Assessment Tool (BAT) at diagnosis (evaluated as International Society of Thrombosis and Haemostasis BAT, ISTH-BAT)] and haemorrhagic phenotypes (sex-related and non-sex-related spontaneous bleeding, post-surgical and post-traumatic bleeding) in male and female populations.

Results. Median age at diagnosis was higher in female population regardless of the type of IBD (30 years in female vs 18,3 years in male, $p<0.0001$), with similar reasons for referral to our center, family history being the most common one (56 patients, 42.7%). The percentage of abnormal ISTH-BAT at diagnosis was higher in males than in females [11% in male vs 3% in female, $p=0.044$, odds ratio (OR): 4.24, 95% confidence interval (CI): 1,177-14,69] (Figure 1). The most common type of non sex-specific bleedings were epistaxis and mucocutaneous bleeding (27.7% and 16.2%, respectively), which were more frequent in the female population (32% vs 23.3% and 20% vs 12.3% in male population, respectively). Conversely, post-traumatic bleedings were observed more often in male patients (20.5% vs 9.3% in females; $p=0.055$, OR: 2.513, 95%CI: 0.9589-6.582). Regarding sex-specific bleedings, heavy menstrual bleeding (HMB) was observed in about half of the female population (36 patients, 48%).

Conclusions. Our homogeneous case series provide a landscape of clinical characteristics of patients carrying autosomal IBDs. We were able confirm a younger age of diagnosis of autosomal IBDs in males when compared to females. This diagnostic delay may be due to an underestimation, in female population, of the severity of para-physiological bleedings, such as HMB, occurring in about half of our patients. The ISTH-BAT at diagnosis was higher in males; this finding could be explained by a higher frequency, in this group of patients, of post-traumatic bleedings.

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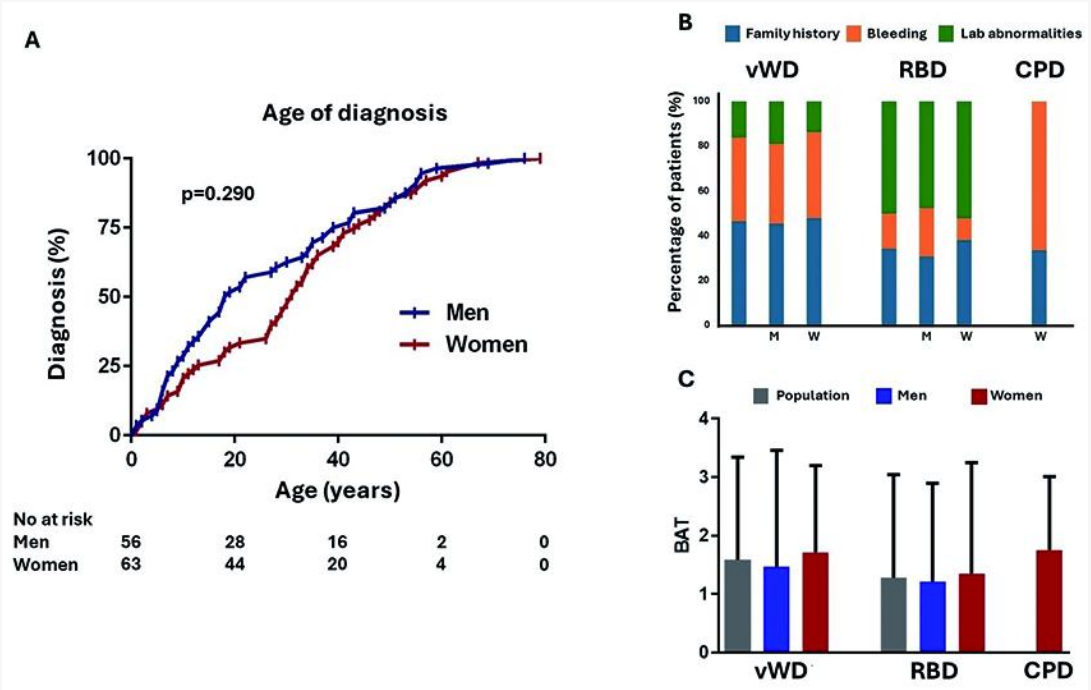

Characteristics at diagnosis. 1A Curve showing the correlation between number of diagnosis and age at diagnosis. 1B Histograms showing the reasons for referral in the different AIBDs (Autosomal Inherited Bleeding Disorders). 1C Median BAT for each AIBD.