A contribution to the debate about the possible different clinical severity between hemophilia A and B

We thank van Miert et al.1 for their comments on our paper.2 Our patient population consisted of patients consecutively enrolled according to regular check-up followup. As such, the temporal interval was the same for patients with hemophilia A and B since all the patients were routinely called for clinical and laboratory monitoring according to a pre-established program. During the period in question, only patients suffering from at least one joint bleed were enrolled. We agree that a larger population of patients with severe hemophilia B would be warranted to obtain more conclusive results as to the possible difference between the hemophilia A and B patient populations. Similarly, the three different categories (<10, 10-50, >50 joint bleeds) were pre-selected to stratify the risk since very few patients with hemophilia B would have been available using joint bleed as a continuous variable. Clearly, the use of prophylaxis rather than on-demand treatment could affect the evolution of arthropathy; however, the percentage of these treatments was similar for the two groups. Furthermore, all HB patients included in the paper presented with significant arthropathy in agreement with clinical and instrumental scores (World Federation of Hemophilia (WFH), Pettersson and ultrasound scores). Of most importance is the difference in pattern of serological and histological changes between hemophilia A and B, which suggests

that arthropathy evolution could be subject to different pathophysiological mechanisms. We agree with van Miert *et al.* when they say that our paper is a contribution to the debate on the possible different clinical severity between hemophilia A and B, and that larger patient populations are needed in order to draw any definitive conclusion.

Daniela Melchiorre, Silvia Linari, Marco Matucci-Cerinic, and Giancarlo Castaman²

'Department of Experimental and Clinical Medicine, Section of Internal Medicine, University of Florence, Rheumatology Unit, Careggi University Hospital; 'Center for Bleeding Disorders, Careggi University Hospital, Florence, Italy

Correspondence: daniela.melchiorre@unifi.it doi:10.3324/haematol.2016.150789

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