Dear Sir,

We read with interest the comments made by Castori [1], which present a further important contribution to the subject of hypermobility type Ehlers-Danlos syndrome (EDS) and joint hypermobility syndrome. Castori is correct in pointing out that the clinical scenarios described in our systematic review for the majority of the cases apply to the vascular-type EDS [2]. It is indeed a complicated field due to the large number of connective tissue disorders, often encompassing overlapping symptoms and clinical presentations and we appreciate to be given the opportunity to elaborate on our views regarding this subject.

As previously stated, the vascular type of EDS includes only approximately 5% of all patients with EDS [3]; however, this number may be an underestimation [4]. Patients with vascular-type EDS often face the most severe complications. Within this field of surgical complications in patients suffering from EDSs, we do agree that generalizations must be applied with caution due to the clinical variability and the lack of evidence in the form of (randomized) large scale studies. However, when we decided to describe the possible surgical complications and suggested relevant general precautions in spite of the low level of evidence, it was to draw attention to what a surgeon (and the patient) may face if a thorough preoperative diagnosis is not performed and necessary assistance is not available [2]. Our clinical everyday life shows that surgeons and physicians in general often are unaware of the possible complications related to EDS and how to prevent them.

Clinical decisions regarding this heterogeneous group of patients should always be made on a case-by-case basis as well as while bearing in mind that EDS is a systemic disease involving almost all organ systems. EDS is a complicated field including a variety of genetic disorders with multiple forms and clinical presentations. The table of general recommendations regarding surgical and anesthesiological considerations for the EDS hypermobility type is very helpful, and we will include it in our local department treatment guidelines.

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References