Position statement of the New Zealand Vascular Society on hypermobile Ehlers-Danlos Syndrome (HEDS).

HEDS is a diagnosis that should only be made by those who have specific expertise with this condition. There is no definitive genetic, radiological, or laboratory testing that provides a result which confidently diagnoses HEDS. The Baden score is the best diagnostic scoring system, but other important diagnoses need to be considered. The VSAoNZ only considers the diagnosis to be confirmed once the patient has been through a multidisciplinary team (MDT) process involving specialists in general medicine, gastroenterology, radiology and vascular surgery. A psychological assessment is an important part of the MDT process. This assessment can be provided by either a psychiatrist or a registered clinical psychologist. Other healthcare professionals including physiotherapists, dietitians and nurse specialists could also be important members of the MDT. The VSAoNZ acknowledges that there is evidence that patients being worked up for this diagnosis can suffer harm if they are not managed by engaged clinicians with access to the multidisciplinary expertise required. These patients should not be referred to a vascular surgeon who has not expressed an interest in this condition.

Once a patient has confirmed diagnosis of HEDS, the question remains whether these patients increasingly suffer from vascular compression disorders. There is not sufficient evidence yet in the literature to support this at this point. Specifically, there are not many groups in the world treating these patients. Professor Sandimann in Germany has published his data on this subject. (Sandimann et al 2021). This was an observational single centre study of the experience and management of vascular compression syndromes (including MALS). 169 patients underwent 196 operations for compression syndromes between 2010 and 2020. Of the 169 patients, 40 (23.7%) had a previously diagnosed hypermobility syndrome before referral to the centre, and 81 (47.9%) were diagnosed with a hypermobility syndrome. Of these 121 patients, a large proportion were reported to have multiple co-existing vascular compression syndromes including 19 patients who were identified as having four co-existing vascular compression syndromes. This is the only study located outside of sparse case reports that describes both an association between heritable connective tissue disorders and vascular compression syndromes, and a cohort of patients undergoing multiple simultaneous surgeries for multiple compression syndromes. The limitations of this study include the following:

1. The (primarily) retrospective approach to heritable connective tissue disorder diagnosis.

2. Short-term outcome reporting (noting that studies with longer-term reporting demonstrate variable but relatively high recurrence rates).
3. Whilst there is a relatively detailed description of the ultrasound technique, there is no detailed reporting of imaging findings.
4. There is no detailed reporting of the relationship between imaging and symptomatology, their relative weighting in surgical decision-making, and how a surgical decision to undertake more than one vascular release operation is made; in this study there are significant numbers of people reported to have three or four simultaneous vascular compression syndromes, but outside of this study there are only sparse case reports of individuals with simultaneous compression syndromes diagnosed.

At this point this surgery has not been part of the training of any New Zealand based vascular surgeon obtaining the FRACS in vascular surgery. Surgery for compression syndrome in patients with HEDS is not well established in the Vascular Surgical literature. On that basis there is not enough strong scientific evidence to support providing safe and clear guidelines at this time. This surgery however does involve anatomy and techniques most commonly performed by vascular surgeons in other clinical settings however that does not mean that expertise is there to provide care for patients with HEDS. It is agreed that vascular surgery would be the appropriate surgical specialty to be involved in an MDT setting, however because of the low level of scientific evidence to support the surgery it is imperative that the outcomes of the surgery are highly audited and follow up of these patients is fastidious.

The VSAoNZ position is that the care of these patients is best undertaken as part of MDT and that a national registry or database be established. The VSAoNZ will only endorse vascular surgeons undertaking the surgery if they meet all the above criteria including contributing all their patient's data to this national database.

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