Whilst Ehlers-Danlos Syndrome (EDS) is quite rare, with an occurrence of one per five thousand across all race and ethnic groups, and most osteopaths might never see a case, it is important to be able to recognise those factors that might affect the way in which the patient should be handled and the special precautions that must be taken.

The condition, which is an inherited connective tissue disorder characterised by skin hyperelasticity, tissue fragility, articular hypermobility, subcutaneous molluscid fibrous tumours, multiple calcified subcutaneous spherules. Additionally, sufferers may exhibit mitral valve prolapse, atrial tachycardias, various heart murmurs, chest pain, fragility of internal organs, accentuated kyphosis, reversed lordosis, scoliosis, spondylolisthesis, anterior vertebral body wedging, posterior vertebral body scalloping, congenital hip dislocation, periodontal disease, profuse scarring and in women, premature delivery.

Difficulties in Examination

The skin being easily damaged, any palpation must use the lightest possible touch and all mobility tests should be carried out with the minimum of interference to the patient to avoid traumatising the skin and underlying tissues. All examinations should be carried out very slowly and gently to avoid stressing already stressed joints. Because of the hypermobility, joints when being examined must not be taken through the full range of movement. Using the principles of minimum interference, osteopathic diagnosis may be accomplished by examining the quality of movement that exists within the normal physiological range. Under no circumstances should passive, full range mobility tests be used, as they may precipitate a secondary joint injury.

Normal reflex tests must also be carried out with caution due to the underlying vascular fragility. Excess force from a patella hammer must be avoided and care must be taken when holding an arm or leg in attempting to ascertain muscle strength.

Treatment

The avoidance of excessive force must be the guiding principle at all times. As hypermobility may be present in even a lesioned joint, an accurate osteopathic diagnosis must be made before any adjustment is carried out. It is also recommended that non-locking techniques be used rather than...
those techniques that require physiological locking. Also, high velocity, low amplitude techniques must not be used. As there is inherent hypermobility, all that is necessary to adjust any osteopathic joint lesion is to use positional techniques. In this way, the possibility of secondary trauma will be reduced.

Following any specific adjustment, the joint must be re-examined for any increase in hypermobility and any traumatically induced haemorrhage. If there is any sign of bleeding then cold packs should be applied to inhibit the local circulation and the patient re-examined the next day to assess the extent of the tissue damage. It is important under no circumstances should ice be used. Cold water packs with an initial temperature of no less that five degrees Celsius should be applied. In this way, there will be lessened possibility of embarrassment to the capillary circulation. In acute joint pain presentations, no less than five days, and preferably seven days, should elapse between any two treatments.

The secondary effects of osteopathic joint lesions, via the visceral-somatic and somatic-visceral reflexes must be borne in mind when treating patients with EDS. A visceral response is more likely due to the secondary involvement of the abnormal muscular tensions that will exist within the abdominal and other trunk musculature. It is important to advise the patient that if they have any abdominal pain, difficulty in breathing, changes in bladder or bowel function, to advise you immediately. This applies whether the reaction has occurred as a result of any treatment or in the normal course of the patient’s life.

The Problems of Hypermobility

Hypermobility presents specific problems to the osteopath. In the normally mobile patient, joint restriction is obvious and easily diagnosed. With a hypermobile joint, loss of range of movement and quality of movement may easily be masked by the normally excessive movement that the hypermobile joint presents. Thus, if there is either the known presence of Ehlers-Danlos Syndrome or the possibility of it’s being a factor, manipulative procedures should be withheld until a positive diagnosis is known. A definitive diagnosis may not be easy to establish but for Vascular, Kyphoscoliosis and Arthrochalasia (formerly types IV, VI and VII) laboratory tests are able to confirm or exclude a positive diagnosis. X-ray diagnosis may show the presence of calcified necrotic fat deposits around pressure points such as elbows, knees and heels. Vascular type (formerly type IV) may also confirmed by electron microscopy but if would be necessary to refer the patient and their family to a clinical geneticist for a confirmatory diagnosis.

In the juvenile patient, where joint laxity and hypermobility are the norm, as compared to the adult, extreme caution must be observed when abducting and adducting the hip joints, to avoid the risk of spontaneous dislocation. In the adult, the same precautions must also be observed, but the risk of dislocation is reduced. If a dislocation does occur, under no circumstances should a reduction be attempted but the patient MUST be referred to hospital even if the patient has learnt to self reduce their own dislocations.

Hypermobility in the lower limbs and feet is more problematical that in the upper girdle with the exception of the shoulder, where any excess mobility may result in damage to the axillary nerves and blood vessels. If there are signs of pronation in the feet, then the help of a podiatrist should be sought to fit the appropriate supports.

Spinal hypermobility may result in difficulty in carrying out specific joint manipulative procedures that require any form of physiological locking and therefore the most appropriate type of technique is the functional positional where there is no applied manipulative force applied. In applying any technique in the prone positions, support must be given to the abdomen and chest in order to prevent any pressure being transmitted via the chest wall.
Secondary Advise to Patients

Patients should be advised that flat shoes with a maximum of one-inch heel must be regarded as the norm. **Slippers are to be avoided as they do give any support to the structures of the feet.** Low chairs that do not provide any lumbar support are to be avoided, as are cars that do not have well fitting seats and properly positioned head restraints. Showers rather than baths should be taken to avoid the effects of excessive forward bending of the spine produced when lying in a bath. Also, the circulatory effects of hot baths together with the negative effects on the skin in terms of softening even more fragile skin are to be avoided. Highly alkaline soaps should not be used and, with the exception of those areas highly supplied with sweat glands, merely a mild cleansing agent is all that is needed.

Hydrotherapy, using either alternating hot and cold water or hot and cold packs may be used but, because of the underlying vascular fragility, the use of ice or ice packs, together with heat lamps or hot water bottles should be avoided. These are procedures which cause vascular insufficiency and result in secondary tissue damage. Hot packs should be no hotter than can be comfortably born by the patient and cold packs should be no colder than 5 degrees Celsius.

Any joint injury should be examined by a health care professional within a maximum of six hours of the trauma, to avoid the possibility of secondary complications of haematoma and scar tissue formation. Patients who have had repeated joint injuries are well advised to have regular, preventative maintenance treatments to ensure that all joints function as well as is possible and that repeated micro-traumatic injuries do not result in a diminution of full joint function.

*The views expressed are those of the author(s) and should not be construed to represent the opinions or policy of the Ehlers-Danlos Support UK or its Trustees*