

Orthopaedic surgery in hypermobile Ehlers-Danlos syndrome and hypermobility spectrum disorders

Mr Aresh Hashemi-Nejad, Consultant Orthopaedic Surgeon & Mr Ivor Vanhegan, Specialist Registrar in Trauma and Orthopaedic Surgery, Royal National Orthopaedic Hospital, Stanmore

Physiotherapy to strengthen muscles and stabilise the joints is the main form of management of hypermobile Ehlers-Danlos syndrome (hEDS) and the hypermobility spectrum disorders (HSD). A good physiotherapist will teach you techniques to protect your joints, in particular you should be advised to avoid pushing your joints past the normal range of motion. In other words, you need to learn to keep your movements within the same limits as someone who isn't hypermobile, as hyperextending your joints can cause long-term traumatic arthritis. Only after appropriate non-invasive treatment has been exhausted should you consider surgery.

Occasionally surgery is needed to treat fractures or, indeed, joint problems. Operating on the soft tissues of the body alone tends to offer only short-term relief, as the soft tissue can stretch again causing the original problem to recur.

Quite often surgery of the soft tissue is combined with bone surgery. In certain circumstances arthritic joints may be stiffened or even replaced. 'Arthrodesis' may be offered, which is stiffening of the joint using internal metal work. Joint replacements are performed when there is quite a lot of arthritis which is painful, the aim here is to provide pain relief and for you to regain function. Your surgeon and physiotherapist will discuss any special measures which will need to be taken concerning stability of the joint.

The idea of surgery should be approached in full consultation with your surgeon and the other medical professionals involved in your care. Before the operation, issues that will need to be addressed include pain management and assessment of cardiac, lung and possible bleeding disorders. If you are found to have mitral valve prolapse (a floppy heart valve) antibiotics may be required as this can increase the risk of infection settling on the valve. With regard to lung problems, you might have a restriction in lung capacity if you have a spinal curvature, and there may be fragility of the lining of the mouth and upper airway, as well as an increased risk of lung collapse. You should also be counselled on the increased risk of poor scar appearance, as delayed wound healing may result in widened or atrophic (sunken) scars.

At the time of the operation the surgical team will need to take your hEDS/HSD into consideration. For instance, care needs to be taken when transferring and positioning someone with hEDS/HSD on the operating table to protect the soft tissues and nerves. Intramuscular injections (injections administered directly into a muscle) are discouraged because they can cause excessive bruising. Particular attention should be paid when elevating limbs, for instance by applying a tourniquet (a tight bandage) so as to avoid injury or dislocation of the joint. During the operation the surgeon must be respectful of the fragility of your skin and soft tissues, limiting unnecessary retraction (pulling back of tissues to expose the part of the body to be operated on) or trauma to these structures. Attention also needs to be paid to the closure of wounds, with a multilayered method (where several layers of stitches are inserted) suggested to reduce the tension on the skin edges and improve wound healing and scar appearance. Wounds should preferably be closed with subcuticular stitches (stitches placed immediately below the skin surface to draw together the tissues beneath the skin) and use of steristrips (adhesive dressings which hold the edges of a wound together) which should remain undisturbed for

longer than usual to allow for the delay in healing time. Bleeding can also be a problem and occasionally surgeons do allow extra operating time to account for this.

Bone fixation (the implementation of implants to hold the bones in a more fixed position) should be rigid with appropriate internal metal work. Your surgeon should discuss with you whether subsequent removal of this metal work is necessary and if it will actually alter the long-term outcome if the metal work is left in place.

Pain control should be discussed with your anaesthetist and surgeon just before the operation. You probably already have a good idea of what works for you in terms of pain relief. Local anaesthetics do not work for everyone with hEDS/HSD. A good indication of whether the use of local anaesthetic would be successful is whether local anaesthetic nerve blocks at the dentist have worked on you in the past. Although doubt has been cast over the use of epidural anaesthetic techniques (where a local anaesthetic is injected into a space that surrounds the spinal cord) because of the increased risk of bleeding, these have been used safely and effectively in women with hEDS/HSD during childbirth.

Occasionally intravenous fluids (fluids administered directly into a vein) may not flow normally in people with hEDS/HSD and IV pumps (devices which control the rate of delivery of intravenous fluids) may be needed.

Most importantly, surgery is only successful if proper rehabilitation is undertaken. This needs to be tailored to suit you, with less aggressive range of motion than is standard procedure. In other words, whereas range of motion exercises usually help someone to move their joints as far as they will go in different directions, you will want to avoid stretching of repaired or reconstructed tissue, especially as you are less likely to develop stiffness anyway. It is also advisable to avoid excessive 'open kinetic chain exercises' where the affected limb is free to move and not in contact with the ground at all times. If an intensive rehabilitation programme is not available this will be difficult. You can initially deteriorate following surgery, particularly if advice has been given for you to spend time on crutches, so it is important to continue with the rehab to get the full benefit from the surgery.

Complication rates following surgery in people with hEDS/HSD have been reported to be higher than in the general population and surgical intervention should be considered only as a last resort. This specialist care should be approached in a multi-disciplinary fashion addressing the issues before, during and after the operation. Results can be good if the goals and expectations are discussed beforehand and a co-operative approach is taken to achieve the best outcome.

Peer reviewed by: Dr Hanadi Kazkaz, Consultant Rheumatologist, University College London Hospital

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