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Care planning for patients with this connective tissue disorder must take a holistic approach

Nursing care planning in Ehlers-Danlos syndrome

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Ehlers-Danlos syndrome (EDS) is the collective term for a group of heritable connective tissue disorders resulting from faulty collagen. There are five cardinal clinical signs:

- » Joint hypermobility;
- » Doughy skin;
- » Skin and tissue fragility;
- » Easy bruising;
- » Atrophic scarring (Beighton et al, 1998).

The symptoms patients display depend on the type of EDS they have; there are six main types. Ocular, dental, gastrointestinal, cardiovascular, urinary, hearing, orthopaedic and rheumatological problems may present in isolation or in any combination. The effects of the condition can be disabling and, in the vascular form, life-threatening (Hakim et al, 2010).

Patient presentation

Mary Jones*, aged 58 years, was diagnosed with EDS when she was 16. She presented wearing a cervical collar, wrist splints and a brace on the left knee. She was using one gutter crutch on her right side. Ms Jones' face was dysmorphic with slight ptosis of the left eye and swelling to the left cheek.

History

During Ms Jones' childhood, she would use bodily contortions to both amuse and alarm family and friends.

Apart from needing several attempts to resolve bilateral strabismus, her childhood health was fairly stable. Her tendencies to fall for no reason and experience frequent unexplained abdominal pain, instability in the feet and knees and increasingly severe lower back pain were all attributed to "growing pains".

At the age of 16, a right patellar subluxation that did not resolve itself led to Ms Jones being seen by a different consultant,

who recognised the underlying syndrome as EDS. Further assessment at this time led to the discovery that she had a missing lumbar vertebra. An orthopaedic consultant said in 1970 that it was surprising that Ms Jones had ever walked and warned she could be confined to a wheelchair by her mid-20s. A lumbar fusion prevented this.

During adulthood Ms Jones has undergone numerous operations to fuse areas of her lumbar and cervical spine, left thumb and both big toes. Other operations have been needed to resolve a patellar subluxation and ongoing temporomandibular joint (TMJ) difficulties; she had received a TMJ implant to the left side. Some of these procedures had to be repeated before the desired outcome was achieved.

Ms Jones had a trabeculated bladder, where the lining of the bladder is overdeveloped and becomes ridged. She had episodes of unexplained urinary retention, and also experienced symptoms of irritable bowel syndrome, with episodes of acute colitis.

She was referred for investigations into the ongoing slippage of the upper cervical spine and placement of a right TMJ implant.

Nursing care

Nursing care plans should take a holistic approach, making use of assessment data and nurses' clinical judgement, to provide a client-specific, realistic nursing outcome. For patients with EDS, many areas of their general health and ability to cope with life processes are affected and need to be considered in all planning. Often, sadly, patients' previous experiences are of a lack of both understanding and acceptance of their situation.

Nurses need to accept that those with EDS may truly be "expert" patients. They tend to know more about the condition and their needs than health professionals, and will often have developed ways of dealing with difficulties. The best way to help informed patients with EDS is to involve them in care planning. Nurses may also need to be ready to suspend disbelief, accepting patients' reporting of what is

5 key points

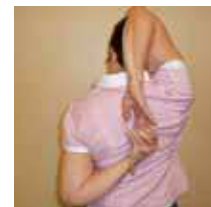
1 Ehlers-Danlos syndrome (EDS) has five cardinal clinical signs

2 The nursing care plan aims to take a holistic approach

3 Nurses need to accept that those with EDS may be "expert" patients

4 The best way to help informed patients is to involve them in care planning

5 Nurses should allow hospital patients to continue with self-help regimens



Joint hypermobility is a clinical sign of Ehlers-Danlos syndrome

happening to, or with, their bodies.

Chronic pain is often a feature of EDS (Hakim et al, 2010). This leads patients to adapt their lifestyle, environment and equipment; nurses should allow hospital patients to continue, as much as possible, with self-help regimens.

The use of clinical assessments, radiological and other results to inform the care plan needs to be done with extra care as the internal picture is often found to be more severe than results have suggested. **NT**

*The patient's name has been changed

• Ehlers-Danlos Support UK: www.ehlers-danlos.org

References

- Beighton P et al (1998) Ehlers-Danlos syndromes: revised nosology, Villefranche, 1997. Ehlers-Danlos National Foundation (USA) and Ehlers-Danlos Support Group (UK). *American Journal of Medical Genetics*; 77: 1, 31-37.
- Hakim A et al (2010) *Hypermobility, Fibromyalgia and Chronic Pain*. London: Churchill Livingstone.