Ehlers-Danlos Syndrome with Associated Bleeding Symptoms

Ehlers-Danlos (ELL ehrs  DANN lows) Syndrome (EDS) is a genetic (inherited) disorder that affects mainly the skin and joints. A person is born with EDS. Other family members may also have been born with it.

There are many types of EDS. People with some types of EDS may bruise or bleed easily. They often have:

- hyper-flexibility (the joints bend farther than the joints of most other people)
- delayed (slow) wound healing
- easy or severe bruising
- loose, unstable joints that may dislocate (come out of place) easily.

Dislocations most often happen in these joints:

- Shoulders
- Hips
- Knees
- Ankles
- Jaw

EDS can also be called Benign Joint Hypermobility Syndrome.

Diagnosis

Your child’s doctor will ask a lot of questions about how flexible your child is (how far he or she can bend). The doctor will also ask your child to try some simple exercises to check for “stretchiness.” For example, the doctor may ask your child to do the following:

- touch the thumb to the wrist of the same hand;
- touch the floor with the palms of the hands without bending the knees.

These tests are scored from 1 to 9 on a Beighton scale. This is a test that rates flexibility. The doctor may make a referral to the Genetics Department to help decide exactly what type of EDS best describes your child’s symptoms.

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Diagnosis, continued

Some types of Ehlers-Danlos Syndrome have bleeding symptoms. These may include:

- **nose bleeds** lasting longer than 10 minutes despite holding pressure on the nostrils
- **prolonged bleeding** with minor cuts, with bleeding lasting longer than 10 minutes, despite steady pressure against the wound
- **excessive bleeding** with minor surgery (such as a tooth extraction)
- **heavy periods** (menstrual bleeding).

Treatment

Depending on your child’s symptoms, the doctor may recommend desmopressin (Stimate®) nasal spray. This medicine works to lessen prolonged bleeding time. It often helps with bleeding symptoms in patients with EDS.

If the doctor prescribes Stimate®, he or she will give you detailed information on how to use this medicine. A nurse in the Hematology and Thrombosis Center will educate you about the medicine and show you how to use it. After this, your medicine can be kept at home. It can be used for bleeding episodes and for severe bruising if first aid does not help.

More Information

Your child’s medical team has recommended these resources for helpful information on EDS with bleeding symptoms: