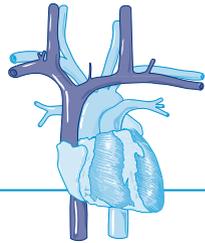


ARTERIAL RUPTURE IS THE MOST COMMON CAUSE OF SUDDEN DEATH.

- **Arterial or intestinal rupture commonly presents as acute abdominal or flank pain that can be diffuse or localized.**
- **Spontaneous arterial rupture is most likely to occur in a person's twenties or thirties, but can occur at any point in life.**
- **Mid-size arteries are commonly involved.**

Arterial, intestinal, or uterine fragility or rupture usually arise in EDS Vascular type, but should be investigated for any EDS type.

Specialized information about EDS Vascular type and the effects of EDS on the vascular system can be downloaded or viewed on-line at ednf.org/medical.



EHLERS-DANLOS NATIONAL FOUNDATION

This **Medical Resource Guide** brings you highlights of information found at ednf.org, available whenever and however you need it.

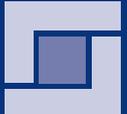
Supported by **Ehlers-Danlos National Foundation**, the **Professional Advisory Network** is a group of dedicated doctors and researchers (find out more on-line) who keep our comprehensive catalog of information for Health Care Professionals up-to-date and factual.

At ednf.org you can find important resources about EDS, from a quick summary for an anxious patient waiting in your office to in-depth articles and media.

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Medical Resource Guide


Ehlers-Danlos
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MARG

EHLERS-DANLOS SYNDROME

Collagen is the most abundant protein in the human body.

It provides the structural strength in most human tissue, including the heart and blood vessels, eyes and skin, cartilage and bone.

What happens when this basic building block is flawed?

What happens when muscles, ligaments, tendons, even large organs are built with structurally defective collagen?

There is systemic weakness, there is instability clearly evident throughout the body.

There is Ehlers-Danlos Syndrome.

Excessively mobile joints, chronic pain, skin softness and stretchiness, gastroenterological problems, easy bruising, scarring and bleeding can characterize Ehlers-Danlos Syndrome (EDS). As a group of genetic disorders of connective tissue, the estimated prevalence of EDS is 1 in 5,000. It is known to affect men and women of all racial and ethnic backgrounds. At least six types of EDS have been identified, and each type is thought to involve a unique defect in connective tissue, although not all of the genes responsible for causing EDS have been found.

Depending on the type of EDS, clinical manifestations vary; they may include joint hypermobility, chronic pain, skin hyperextensibility, poor wound healing with atrophic scars, easy bruising and generalized connective tissue fragility. The fragility can be found throughout the body, and is caused by alterations in either the synthesis or the structural integrity of either collagen or other molecules essential to connective tissue structure. Within each family the type of EDS runs true, but individual family members may vary in clinical severity and manifestations.



DENTAL MANIFESTATIONS OF EDS

A dental patient with Ehlers-Danlos will probably exhibit at least one, and frequently several, of the following indicators (many of these can also be found in other areas of the body):

Extra-Oral

- Scarring on the skin and forehead;
- TMJ (temporo-mandibular joint) noise, including clicking ± crackling (crepitation);
- TMJ locking/immobility;
- TMJ (and other joint) subluxations/dislocations;
- TMJ (and other joint) laxity;
- Skin hyperelasticity;
- Pain, chronic and widespread;
- Resistance to local anesthesia.

Intra-Oral

- Mucosa, including surrounding skin and blood vessels, may be noticeably fragile:
 - Sutures may not hold well;
 - Increased risk of excessive bleeding/hemorrhage.
- Fragile gingival tissue with:
 - Persistent inflammation ± hyperplasia;
 - Early-onset periodontitis.
- Teeth may be malformed ± translucent:
 - Under-developed enamel (hypoplasia);
 - Deep fissures;
 - Pulp stones;
 - Premature loss of deciduous and permanent teeth
- Tongue is usually very supple:
 - About 50% can touch the end of nose with tongue (+ Gorlin sign)
- Palate is abnormally high and arched.

CLINICAL CONSIDERATIONS

There is no real difficulty treating someone with EDS. Remember their entire body can be affected, including neck and back. No procedure can be considered routine, but precautions are relatively easy to take:



- For a person with Mitral Valve Prolapse:
 - Use prophylactic antibiotics.
- For any EDS or suspected EDS patient:
 - Take brief time-out rests every ten minutes;
 - Bite block may help prevent TMJ problems;
 - Reduce waiting times and length of visits;
 - Avoid neck/back stress wherever possible.
- When using local anesthesia:
 - EDS patient may not respond as expected.

Orthodontic

- Forces for orthodontic treatment should be lighter due to fragility of the periodontal ligament and to prevent damage to the buccal mucosa;
- Increased risk for mucosal ulceration related to bracket position;
- Increased risk for rapid migration/teeth mobility;
- Consider longer periods of retention.

Surgical and Endodontic

- Dental and maxillofacial surgery should be avoided if other options are available;
- Blood coagulation studies should be done before proceeding with any surgery;
- DDAVP® (desmopressin) may be indicated;
- Raise mucoperiosteal flap with care;
- Tie sutures, under slight tension, to larger-than-normal sections of tissue;
- Care must be taken on forces applied to gums and surrounding fragile tissues and structures;
- Possible poor wound healing after extractions:
 - Sutures may not hold well;
 - May need prefabricated acrylic plate over surgical site;
- Check for pulp stones and abnormal roots.