WHAT IS ELHERS DANLOS?

Ehlers–Danlos syndrome (**EDS**) is a rare inherited connective tissue disorder with different presentations that have been classified into several primary types including Hyperflexibility, Classic, and Vascular. There can be some symptoms overlap in various types. EDS is caused by a defect in the structure, production, or processing of collagen or proteins that interact with collagen, such as mutations in the COL5A or COL3A genes. The features of EDS were first described by Hippocrates in 400 BC. The syndrome is named after two physicians, Edvard Ehlers from Denmark, and Henri-Alexandre Danlos from France, who described it at the turn of the 20th century.

Collagen in connective tissue helps tissues resist deformation. It is an important contributor to the physical strength of skin, joints, muscles, ligaments, blood vessels and visceral organs; abnormal collagen renders these structures more elastic. Depending on the individual, the severity of the mutation can vary from mild to life-threatening. There is no cure. Treatment is supportive, including monitoring of the digestive, excretory and cardiovascular systems. Occupational/physical therapies, bracing, or corrective surgery may help with the frequent injuries and pain that tend to develop in certain types of EDS. Extra caution and special practices are advised to prevent permanent damage.

Three basic surgical problems arise due to EDS: the strength of the tissues is decreased, which makes the tissue less suitable for surgery; the fragility of the blood vessels can cause problems during surgery including hemorrhage ; and wound healing can be delayed or incomplete. Surgery with Ehlers-Danlos patients requires careful tissue handling and a longer immobilization afterwards. Studies have also shown that local anesthetics, arterial catheters and central venous catheters cause a higher risk in haematoma formation in patients with EDS.

Signs and symptoms vary widely based on which type of EDS the patient has; each symptom is based on faulty collagen. A partial list of signs, symptoms, and potential complications is:

- Hyper-flexible joints
- Joint instability; sprains, dislocation, subluxation & hyperextension
- Gait problems (eg Trendelenburg Lurch)
- Early onset osteoarthritis
- Chronic degenerative joint disease
- Deformity of the fingers
- Tearing of tendons or muscles
- Deformities of the spine (eg scoliosis)
- Myalgia (muscle pain)
- Arthralgia (joint pain
- Osgood-Schlatter Disease
- Fragile skin that tears easily
- Easy bruising
- Arterial rupture
- Valvular heart disease (eg mitral valve prolapse)
- Dilation /rupture of ascending aorta
- Sleep Apnea
- Gastroesophageal Reflux Disease
- Tremors
- Fibromyalgia
- Hernias
- Prolapse (anal, uterine, organ)
- Collapsed lung
- Nerve compression disorders (carpal tunnel syndrome, neuropathy)
- Insensitivity to local anesthetics
- Platelet aggregation failure
- Pregnancy complications (eg hemorrhage, uterine tearing, miscarriage)