Pathogenetic mechanisms underlying the Ehlers-Danlos syndromes

There is an additional genetic classification structure of the EDS into groups according to similarities in the way the responsible genes affect the body.

Group A: Disorders of collagen primary structure and collagen processing, comprised of cEDS, vEDS, aEDS, dEDS, and cvEDS.

Group B: Disorders of collagen folding and collagen crosslinking, comprised of kEDS-PLOD1 and kEDSS-FKB14.

Group C: Disorders of structure and function of the myomatrix, comprised of clEDS and mEDS.

Group D: Disorders of glycosaminoglycan biosynthesis, comprised of spEDS-B4GALT7, spEDS-b3GALT6, mcEDS-CHST14, and mcEDS-DSE.

Group E: Defects in complement pathway, comprised of pEDS.

Group F: Disorders of intracellular processes, comprised of spEDS-SLC39A13 and BCS.

Group G: Unresolved forms of EDS, comprised of hEDS.

Conditions no longer included in the EDS spectrum are occipital horn syndrome, fibronectin-deficient (EDS X), familial articular hypermobility (EDS XI), X-linked EDS with muscle hematoma (EDS V), and filamin A related EDS with periventricular nodular heterotopia.