# DERMAL ARCHITECTURE UNDER THE MICROSCOPE

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Tensile properties of skin are one major clinical criterion to recognize connective tissue disorders. There is a strong interdependance between tensile variables and the dermal microanatomy.


- Piérard GE, Lapière ChM. Microanatomy of the dermis in relation to relaxed skin tension lines and Langer's lines. Am J Dermatopathol 9, 219-224, 1987.
- Piérard GE, Masson P, Rodrigues L, Rogiers V, Berardesca E, Elsner P, Lévêque JL, Loden M, Marks R, Parra JL, Trevethan MA. EEMCO guidance to the in vivo assessment of tensile functional properties of the skin. Part 1: Relevance to the structures and ageing of the skin and subcutaneous tissues. Skin Pharmacol Appl Skin Physiol 12, 352-362, 1999.
- Henry F, Goffin V, Piérard-Franchimont C, Piérard GE. Mechanical properties of skin in Ehlers-Danlos syndrome, types I, II and III. Ped Dermatol 13, 464-467, 1996.


Connective tissue naevi represent models for understanding and classifying genetic and acquired connective tissue disorders at the microscopic level. Histochemical stains are mandatory to get an overall information about most of the connective tissue abnormalities.


- Piérard GE, Lapière ChM. Nevi of connective tissue. A reappraisal of their classification. Am J Dermatopathol 7, 325-333, 1985.
- Piérard GE, Piérard-Franchimont C, Ben Mosbah T, Al Rustom K. Aspects communs des hyperplasies conjonctives du syndrome de Protée et des collagénomes. Ann Dermatol Venereol 118, 788-790, 1991.
- Piérard GE. Sirius red polarization method is useful to visualize the organization of connective tissues but not the molecular composition of their fibrous polymers. Matrix Coll Rel Res 9, 69-72, 1989.


Ehlers-Danlos syndromes are characterized by prominent and typical alterations in the organization of the fibre networks in the dermis including collagen bundles and elastic fibres. In addition, the size and shape of collagen fibrils are altered.


- Vogel A, Holbrook KA, Steinmann B, Gitzelmann R, Byers PH. Abnormal collagen fibril structure in the gravis form (type I) of the Ehlers-Danlos syndrome. Lab Invest 40, 201-206, 1979.
- Piérard GE, Piérard-Franchimont C, Lapière ChM. Histopathology aid at the diagnosis of the Ehlers-Danlos syndrome gravis and mitis types. Int J Dermatol 22, 300-304, 1983.
- Piérard GE, Lê T, Piérard-Franchimont C, Lapière ChM. Morphometric study of cauliflower collagen fibrils in Ehlers-Danlos syndrome type I. Coll Rel Res 8, 453-457, 1988.
- Piérard GE, Lê T, Hermanns JF, Nusgens BV, Lapière ChM. Morphometric study of cauliflower collagen fibrils in dermatosparaxis of the calves. Coll Res Res 6, 481-492, 1986.

- Hausser I, Anton-Lamprecht I. Differential ultrastructural aberrations of collagen fibrils in Ehlers-Danlos syndrome types I-IV as a means of diagnostics and classification. Hum Genet 93, 394-407, 1994.
- Kobayasi T, Ullman S. Twisted collagen fibrils. Significance for diagnosis of Ehlers-Danlos syndrome. J Invest Dermatol 107, 266, 1995.
- Smith TL, Schwarze U, Goldstein J, Byers PH. Mutations in the COL3A1 gene result in the Ehlers-Danlos syndrome type IV and alterations in the size and distribution of the major collagen fibrils of the dermis. J Invest Dermatol 108, 241-247, 1997.
- Watson RB, Wallis GA, Holmes DF, Viljoen D, Bryers PH, Kadler KE. Ehlers-Danlos syndrome type VII B. Incomplete cleavage of the patient's abnormal type I collagen by N-proteinase resuls in the formation of rough-bordered collagen fibrils characteristic of the disorder. J Biol Chem 267, 25529-25534, 1992.
- Nusgens BV, Verellen-Dumoulin Ch, Hermanns-Lê T, de Paepe A, Nuytinck L, Piérard GE, Lapière ChM. Evidence for a relationship between Ehlers-Danlos type VIIc in humans and bovine dermatosparaxis. Nature Genet 1, 214-217, 1992.
- Piérard GE, Hermanns-Lê T, Arrese Estrada J, Piérard-Franchimont C, Lapière ChM. Structure of the dermis in type VIIc Ehlers-Danlos syndrome. Am J Dermatopathol 15, 127-132, 1993.


The current classification of Ehlers-Danlos syndromes disregards morphological alterations and thus may blur unrelated conditions


- Beighton P, de Paepe A, Steinmann B, Tsipouras P, Wenstrup RJ. Ehlers-Danlos syndromes : revised nosology, Villefranche, 1997. Am J Med Genet 77, 31-37, 1998.
- Burrows NP, Nicolls AC, Yates JRW, Gatward G, Sarathachandra P, Richards A, Pope FM. The genetic encoding collagen $\alpha$1 (V) (COL5A1) is linked to mixed Ehlers-Danlos syndrome type I/II. J Invest Dermatol 106, 1273-1276, 1996.
- De Paepe A, Nuytinck L, Hausser I, Anton-Lamprecht I, Naeyaert JM. Mutations in the COL5A1 gene are causal in the Ehlers-Danlos syndromes I and II. Am J Hum Genet 60, 547-554, 1997.
- Nuytinck L, Freund MM, Lagae LG, Piérard GE, Hermanns-Lê T, De Paepe A. Classical Ehlers-Danlos syndrome caused by a mutation in type I collagen. Am J Med Genet (in press).


The role of dermal dendrocytes is probably underrecognized in the control of the extracellular matrix structure.


- Piérard-FranchimontC, Fazaa B, Benzarti H, Kort R, Arrese JE, Nikkels AF, Kamoun MR, Piérard GE. Phenotypic heterogeneity in the fibroblast-like cells of the dermis. Giorn Int Dermatol Ped 6, 7-13, 1994.
- Arrese Estrada J, Piérard GE. Factor XIIIa-positive dendrocytes and the dermal microvascular unit. Dermatologica 180, 51-53, 1990.
- Piérard GE, Arrese Estrada J, Piérard-Franchimont C, Deleixhe-Mauhin F. Is there a link between dendrocytes, fibrosis and sclerosis ? Dermatologica 181, 264-265, 1990.

- Piérard-Franchimont C, Piérard GE, Hermanns-Lê T, Arrese Estrada J, Verloes A, Mulliez N. Dermatopathological aspects of restrictive dermopathy. J Pathol 167, 223-228, 1992.
- Herouy Y, Nockowski P, Schopf E, Norgauer J. Lipodermatosclerosis and the significance of proteolytic remodeling in the pathogenesis of venous ulceration. Int J Mol Med 3, 511-515, 1999.