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I have Ehlers-Danlos Syndrome Type ____, which causes defective connective tissue in my collagen. Please be sure to check carefully for the following:

- Epicanthal folds
- High myopia
- Keratoconus
- Blue sclera
- Lens subluxation
- Angioid Streaks
- Cataracts
- Dry eyes
- Glaucoma (with pachymetry for accuracy)
- Photophobia
- Retinal Detachments, holes, tears
- Strabismus
- Macular Degeneration
- Posterior Staphyloma
- Carotid-cavernous sinus fistulas
- Accommodative difficulties
- Dry eyes
- Diplopia, monocular or binocular
- Large phorias which may be symptomatic
- Blepharoclonus

Recommended Ocular Testing for the Patient With EDS

- Complete slit lamp exam with TBUT. Understand that I may be more prone to corneal dystrophies, dry eyes.
- Dilated fundus exam; fundus photography
- Ocular topography to rule out early keratoconus
- RTA may be needed – Retinal Thickness Analyzer
- Orbscan and/or pachymetry to check corneal thickness
- EDS patients are not good candidates for LASIK.
- Pupil testing (rule out APD), aperture measurements (check for ptosis), rule out blepharoclonus.
- ?? I may be more prone to recurrent corneal erosions ??
- I may be more prone to migraine episodes and/or aura without the migraine headache.
- I may be more prone to macular degeneration

For more information, please check into www.EDNF.org.

Articles

Title: Orbscan mapping in Ehlers-Danlos Syndrome

Authors: Pesudov, Konrad

Date: August, 2004

Publisher: Elsevier

Citation: Pesudovs, K. 2004 Orbscan mapping in Ehlers-Danlos syndrome, "Journal of Cataract and Refractive Surgery", Volume 30, No. 8, 1795-8

Abstract: A candidate for refractive surgery presented with classic (Type 1) Ehlers-Danlos syndrome (EDS). Clinical examination revealed blue sclera, limbus-limbus thinning, myopia, and astigmatism. Orbscan (Bausch & Lomb) pachymetry mapping provided a striking demonstration of the limbus-to-limbus thinning with a central corneal thickness of 360 microm in the right eye and 383 microm in the left eye and midperipheral corneal thickness ranging from 370 to 438 microm and 376 to 434 microm, respectively. Despite the theoretical biomechanical weakness from the thin cornea and defective collagen, regular surface topography was maintained without the development of keratoconus. Although all types of EDS remain a contraindication to laser refractive surgery, Orbscan mapping provides a valuable insight into corneal shape and thickness in this condition.

Title: Keratoconus and the Ehlers-Danlos Syndrome: a new aspect of keratoconus

Author: I. Robertson

Date: May, 1975

Publisher: Medical Journal of Australia

Abstract: Hypermobility of the joints is a frequent finding in patients who have keratoconus. Twenty-two of the 44 patients (50%) presenting in 1973 were found to have hypermobility of joints (mainly Ehlers-Danlos syndrome II or mitis type). The significance of this finding is discussed in relation to biochemical defects recently found in corneas with keratoconus. The hypothesis is presented that keratoconus is often a part of a generalized heritable disorder of connective tissue due to a biochemical defect causing a weakness in its structure.

Title: Joint Hypermobility in Keratoconus

Author: Woodward EG, Morris MT

Date: Oct, 1990

Publisher: Department of Optometry and Visual Science, City University, London, UK Ophthalmic Physiological Optometry

Abstract: There are several reports linking keratoconus and connective tissue disorders, such as Ehlers-Danlos syndrome, osteogenesis imperfecta and mitral valve prolapse, suggesting that keratoconus may be the result of a localized dysfunction in collagen metabolism. In view of this the incidence of hypermobility of the joints among a group of patients with keratoconus was compared with a normal (matched) control

group. Eighty-four patients were examined using the Beighton modification of the Carter and Wilkinson scoring system. A control group matched for sex, age and ethnic group was also assessed. No statistically significant difference between the groups was found for the trunk or knees; a difference was found for the metacarpo-phalyngeal and wrist joints. The findings support the theory that keratoconus is a localized manifestation of a mild connective tissue disorder. Also, it can be stated that patients with Keratoconus are five times more likely to show hypermobility of the metacarpo-phalyngeal and wrist joints.

Title: Corneal Topography in Ehlers-Danlos Syndrome

Authors: McDermott ML, Holladay J., Liu D, Puklin JE, Shin DH, Cowden JW

Date: Sept 1998

Publisher: Journal of Cataract and Refractive Surgery

Abstract: Purpose: To assess the use of corneal topography in conjunction with slitlamp biomicroscopy and retinoscopy to diagnose keratoconus in a large group of patients (72 eyes) with Ehlers-Danlos Syndrome (EDS). Result: In 72 eyes, no keratoconus was found using slitlamp biomicroscopy . No eye had an I-S value greater than 1.60 diopters, 2 eyes had a CCP greater than 46.50 D, and 2 eyes had a Q value less than -1.00. Conclusions: Slitlamp biomicroscopy of the cornea was unremarkable in all patients. Only 1 patient had Q values and profile difference maps that were mildly suggestive of keratoconus. Even after adding topography to the examination, it appears that keratoconus in a known population of patients with EDS remains rare.

Title: Blepharoclonus and Arnold-Chiari Malformation

Authors: Jacome DF

Date: August 2001

Publisher: Acta Neurol Scand

Abstract: Objective: Blepharoclonus (BLC) denotes large amplitude, involuntary tremors of the orbicularis oculi muscles, observed during gentle closure of the eyelids. BLC may follow major head trauma. Four patients with Arnold-Chiari malformation (ACM) and BLC are described. Conclusion: BLC is an underdiagnosed neuro-ophthalmological sign of ACM.

Title: Tretinoin Pre-Treatment but not Direct Treatment Shows a Beneficial Effect of Wound Healing in Diabetic Mice

Authors: Hans W, Ghermai U, Gossler A, Bittner M, Furst A, Jacob K, Mesterma S, Goppelt A

Date: September 2004

Publisher: European Society for Dermatological Research

Abstract: (condensed): There is circumstantial evidence that topical pre-treatment with tretinoin (all-trans retinoic acid) might improve the healing of human ulcers. Especially in diabetic patients with long lasting ulcers the delayed healing process is a persistent problem. We investigated and compared the effect of tretinoin pre-treatment and direct on impaired wounds in genetically diabetic mice. Wounds of db/db mice display delayed wound healing due to lessened wound contraction and delayed granulation tissue formation. We learned that the beneficial effect of tretinoin can be offset by the irritation caused by the retinoid and that when treatment and wounding start at the same time, positive effects, like thickening of skin, increase of collagen and wound contraction, set in too late to be beneficial in this wound healing model. In conclusion, the incisional wound healing model with pre-treatment of the skin of db/db mice can be used to investigate the effects on tensile strength of retinoids and their derivatives, of which those lacking the extremely irritating properties of tretinoin would get the most promising drug candidates for ulcers that are often observed in diabetic patients.

Title: Brittle Cornea Syndrome and its Delineation from the kyphoscoliotic type of Ehlers-Danlos Syndrome (EDS VI): report on 23 patients and review of the literature.

Authors: Al-Hussain H, Zeisberger SM, Huber PR, Giunta C, Steinmann B

Date: January 2004

Publisher: American Journal of Genetics

Abstract: The brittle cornea syndrome (BCS) is a generalized connective tissue disorder characterized by corneal rupture following only minor trauma, keratoconus or keratoglobus, blue sclerae, hyperelasticity of the skin without excessive fragility, and hypermobility of the joints. Because of the similarities between the BCS and the kyphoscoliotic type of the Ehlers-Danlos syndrome (EDS VI), both disorders tend to have been confounded. Here, we show that all of our BCS patients tested in this regard had biochemical findings reflective of normal activity, of lysyl hydroxylase, characteristically deficient in EDS VI, such as normal urinary total pyridinoline ratios and/or normal electrophoretic migration of collagen chains produced by dermal fibroblasts. The BCS is, therefore, an entity distinct from the kyphoscoliotic type of EDS, which has a much poorer prognosis.

Title: Effectiveness of short-contact topical tretinoin in promoting wound healing in db/db mice.

Authors: Toyama T, Ohura N, Kurita M, Momosawa A, Haril K

Date: 2006

Publisher: Taylor & Francis, Basingstoke, Royaume-uni
Scandinavian Journal of Plastic and Reconstructive Surgery and Hand Surgery.

Abstract: We have investigated the effects of short-contact topical application of tretinoin using 16 male db/db mice by creating two full-thickness wounds on the skin of the back. A 0.1% tretinoin aqueous gel was applied to one wound for five minutes daily for five successive days while only aqueous gel was applied to the other (control). The mean (SD) percentage surface area unhealed before and after treatment were 0.88 (0.3) and 0.64 (0.15). In tretinoin-treated mice and non-treated mice (controls), the mean (SD) thicknesses of granulation were 1,383 (697) micrometers and 683 (413 micrometers), the density of capillary vessels in granulation was 12.2 (5.5%) and 5.7(3.9%), respectively. Differences between the two groups were significant for each variable. Wound healing was accelerated with short-contact topical application of tretinoin in db/db mice.