

Patients Suffering from Ehlers-Danlos Syndrome type III Do Not Respond to Local Anesthetics

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The classical features of Ehlers-Danlos Syndrome type III (EDS) are hyperextensibility of the joints, hyperelasticity and fragility of the skin. Only minor visible changes of the skin and joint hypermobility makes this syndrome difficult to distinguish from the more common simple hypermobility. Ehlers-Danlos syndrome is claimed to be a rare syndrome with an incidence of 1/150,000. Since we initiated our research on this syndrome, we have found 4 families in an area with about 300,000 inhabitants. The syndrome, therefore, seems to be more common than assumed and the reason why the syndrome is not diagnosed can be due to the fact that the syndrome is diagnosed as hypermobility.

We have observed that local anesthesia has an insufficient effect in Ehlers-Danlos type III patients and that it is difficult to distinguish the Ehlers-Danlos type III syndrome from hypermobile patients diagnostically. In genetic advising and prognosis of the EDS patients, there is a need for new tools to separate them from hypermobile patients. We, therefore, investigated quantitatively if the Ehlers-Danlos type III patients objectively responded differently from hypermobile patients to cutaneous analgesia, and we sought to find out if these parameters could be used as a new test to discriminate between the two diseases.

Topical analgesics (EMLA cream) was applied to seven EDS patients, ten hypermobile patients, and to fifteen controls. The depth of the cutaneous analgesia was measured by sensory and pain threshold depths to controlled needle insertions. It should be easy to carry out the measurements

in the daily clinical situation. EMLA cream is commonly available and insertion of the needle can be done without the special equipment used in this study. Controls and hypermobiles did not differ in their response to cutaneous analgesia. The thresholds to cutaneous laser stimulation and the depth of analgesia increased significantly less in the Ehlers-Danlos patients, compared to the two other groups. In clinical practice, a needle insertion test can easily be applied to investigate if patients are responders or non-responders to local analgesics.

When the Ehlers-Danlos type III patients were biopsied in the hip region for skin biopsy, they all reported considerable pain although large doses (5 ml) of 1% lidocaine-epinephrine were infiltrated subcutaneously. When we asked them for more details, they reported that they had all previously experienced difficulties in obtaining a sufficient analgesia at the dentist, although they had been given substantial doses of local analgesics. Some of the women reported no pain alleviation of local analgesics when they were sutured after episiotomy. They were commonly characterized as hysterics. We have definitely proved that this is not the case.