

Local Multiple Injections of Lidocaine as a Very Efficient Treatment of Intractable Pains in Ehlers-Danlos Disease. The Dysproprioception Hypothesis

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Abstract

Ehlers-Danlos is an inherited connective tissue disease whose frequency is greatly underestimated due to its lack of knowledge by physicians (Tinkle et al., 2017). The symptomatology, the modalities of expression and the typology of pain very often direct to other diagnoses. The intractable nature of the pain leads to therapeutic „escalations” with negative effects. Multiple local injections of painful areas used often in a cohort of 5,700 patients, followed for 25 years, constitute a considerable new therapeutic contribution. It is easy to achieve and without local complications.

Keywords: Ehlers-Danlos; Chronic Pain; Intractable Pain; Local Injections of Lidocaine; Proprioception

Introduction

Ehlers-Danlos disease is commonly known through two signs: skin hyperlaxity and joint hypermobility that were highlighted by its first descriptors Tschernogobov in Moscow in 1881 [1] and Ehlers in Copenhagen in 1900 [2], both dermatologists. The presence of chronic and intractable diffuse pain is more recent [3] and is still too little known to doctors. Present in more than 90% of cases [4], they are nevertheless exceptionally attached by them to The Disease of Ehlers-Danlos, dragging patients into a very painful and dangerous medical wandering especially by iatrogenesis. Together with fatigue, they are the two main causes of disability and social exclusions that are often experienced by patients who are affected [5-8]. Their unusual expressions confuse the patient who has trouble local treatments and local injections of Lidocaine.

Materials and Methods

Population studied

Our experience with Ehlers-Danlos disease is based on a cohort of more than 5,700 patients, children and adults, followed for 25 years of which about 50% have been treated with lidocaine injections at least once. Here we describe our observation and the techniques used for the lidocaine injections. The diagnosis was made based on the clinical criteria of Brighton [3] initially highlighted by Rodney Grahame to which we supplemented a recent study on 849 cases versus 820 healthy subjects [9]. Diffuse and chronic pain, often intractable, is included in the list of 9 selected signs, of which 5 are enough to make the diagnosis with a e describing them, and the doctor who does not know how to interpret them. This is due to the presence of a major infringement of proprioception. It is also this dysproprioception that explains the effectiveness of sensitivity of 99.6% and a specificity of 98% [10,11]. Other signs are motor proprioceptive disorder, joint instability including sensations of joint „cracking” expression of subluxation, a thin, fragile skin not protecting against the sensations of

electric shocks on contact with static electricity, joint hypermobility, a hemorrhagic tendency with frequent bruising, hyperacusis, gastric reflux. These symptoms align with the current New York classification for HSD/EDSh 2017 [12-14].

The multiple pains of Ehlers-Danlos disease

They have several characteristics that are evocative [15]:

1. They are diffuse, present in the three dimensions of body perception: internal, external and environmental. To the question „where does it hurt?“ the answer is „everywhere“. These are the muscles (myalgias) and their insertions (tendonitis), ligaments and joint capsules (arthralgias), skin and mucous membranes (gingiva, mouth, nasal pits, trachea, oesophagus, stomach, intestine, rectum, bladder, vagina), meninges (migraines), veins, ribs, teeth, hair, nails, skin muscles of the face and neck. Because of a dysproprioceptive syndrome, the location of pain is often made more difficult due to projections into other areas, surprising and misleading for the physician. Thus, the pain caused by aggressive vesicular lithiasis for a thinned mucous membrane, radiates not in the right shoulder but in the lower dorsal region to the right [15].
2. Pain is permanent. To the question „when do you feel like this?“, the answer is „all the time.“ Patients mention a permanent painful background with very violent seizures especially after trauma [16].
3. Their expression is surprising, unusual for doctors. Patients describe diffuse tingling, sensations of electric shocks, crushing, tearing, stabbing, red iron embedded in the skin, chest, skull, or of having been beaten. They are accompanied, during extremely painful sensations, often by imminent death.
4. Imaging (X-rays, RMI, scanners) is negative. This contrast of normal imagery and particularly intense pain is evocative of Ehlers-Danlos disease but is most often interpreted as being of psychic origin.
5. They are aggravated by movement, posture (standing or sitting), dislocations, violent shocks not necessarily very important (fall, road accident) with generalized tissue shake, meteorology (cold) hormonal variations in women (puberty, menstrual cycle, pregnancy) [6,17].
6. They resist many painkillers, including morphinics, which aggravate them. The treatments that appear best suited are: intermittent oxygen therapy [18,19] on migraines and fatigue in particular [18], Nefopam, nonsteroidal anti-inflammatory drugs under gastric protection [20], anti-dystonics [21], Baclofen and, above all, local treatments: TENS, heat, cryotherapy, physiotherapy [22], patches of anti-inflammatory or painkillers and, in particular, local injections of Lidocaine.

The method of local injections of lidocaine

The injected product is Lidocaine 10 Mg for 10 ml. For children we use 5 mg for 10 ml. The amount injected depends on the structure injected: a few drops are enough from a tendon, or on contact with a rib, half a millimetre or a millimetre are needed for a muscle. Local cortisone injection, associated with Lidocaine, is responsible in our observation for an increase in pain in the following days without positively affecting the results. We thus stopped adding cortisone ever since the first injections. Something that is particularly observed in Ehlers-Danlos Syndrome.

The choice of the needle is important: it should be as fine as possible. Subcutaneous needles provide access to most areas to be treated. For deeper areas, intra-muscle needles are the right ones. The skin and subcutaneous tissues are very easy to pierce because of their thin and fragile structure. Skin contact through the needle and its penetration can be painful. The prior use of a lidocaine patch may then be indicated. The pressure of the injected liquid often triggers an unpleasant sensation often with a type of burn.

Injection sites should be identified by precise and careful palpation to avoid worsening pain. It is guided by the patient who often knows how to identify the painful points and areas on his own. Only the distinctly painful points should be injected. The most commonly affected places are: neck muscles (high and body insertions of trapezoids and sterno-cleido-mastoids, deltoids, shoulder blade fixing muscles, frontal muscles, chewing muscles, epicondyles, the anatomical snuffbox, the first interosseous space. The xyphoid region („Ehlers point“), the lower edge of the ribs, the sacroiliac joints, the insertions of the thigh, knee and ankle muscles (Figure 1-7).



Figure 1: Injections at the temporo-mandibular joint.



Figure 2: Injection into the left trapezius.



Figure 3: Injection near the temporomandibular joint.



Figure 4: Injection to upper insertion of the sartorius muscle.



Figure 5: Injection on the "Ehlers Point" (xiphoid).



Figure 6: Injection into the anatomical snuffbox.



Figure 7: Injection points at the back.

Several injections, up to 15, can be performed in the same treatment sequence. We have always avoided injecting more than one 10ml bottle in a single session. The general tolerance is usually excellent. There are sometimes feelings of generalized ill-being that quickly give way in an elongated position after a brief resting phase.

A feeling of weariness, which can be interpreted as a consequence of the body's painful perception suppression and the resulting relief, can be observed. It is exceptional that injections cause increased pain. They must then be discontinued immediately. Due to the excessive fragility of the vessels, a bruise or hematoma may occur in the injection area. But we have never observed, on several thousand patients, the slightest skin lesion, necrosis for example, despite the thinness and disorders of the vascularization of the skin especially at the extremities (the feet and hands are cold). It should also be noted that these patients never make bedsores.

Results

If successful, in the majority of cases, the analgesic effect is very rapid, almost immediate, and is accompanied by a change in the perception of the area of the body concerned where the pain makes way, in case of success to the sensations of position and ease of more physiological and less or more painful movements. It is possible to turn your head freely, sit without pain, walk more easily, take objects and handle them, raise your arms and put on the sleeves of clothing more easily. Patients report seeing their bodies as 'lighter', less 'tied in a knot'. These positive effects can be transient from a few hours to a few days, but in most cases, they last from several weeks to several months. In some of them, the repetition of the injection sessions is accompanied by a decrease in the intensity of the painful phenomena from one session to the next. It becomes possible to space them. The success of the injections does not exclude the use of other analgesic treatments in particular oxygen therapy and proprioceptive orthotic devices [23,24]. Their association reinforces their effectiveness. It is more difficult to use this treatment in children because of the phobia of injections, but some of those we have followed appreciate them and call them 'magic bites'.

Sometimes after the disappearance of the pain patients describe "odd" sensations that preoccupy them. They do not feel the painful sensations that they are accustomed to anymore and these sensations are replaced by new unfamiliar ones. This new body sensation must be relearned which in some instances may take some time.

Discussion

Ehlers-Danlos is responsible for remarkable pain due to its resistance to treatment. A simple innocuous gesture can achieve spectacular results where other treatments fail or get incomplete results. The question of this effectiveness questions the very mechanism of pain

in this unusual and misunderstood pathology that is Ehlers-Danlos disease. One of the explanations in this disease of proprioception is, possibly, the temporary suppression of painful signals more easily perceived by the brain in favour of sensations that express more the body of the person. This mechanism could explain the effects of this treatment outside of Ehlers-Danlos disease as shown by some authors in myofascial syndromes [25].

Conclusion

A simple, risk-free, easy-to-achieve, local injection of Lidocaine has enabled us to achieve rapid and lasting results on the particularly intense, disabling and intractable pains of an inherited connective tissue disease whose frequency is largely underestimated: Ehlers-Danlos. Beyond the interest of the patients concerned, this new contribution opens new perspectives for understanding the interactions between the body and the brain.

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