

CORONA VIRUS / COVID19. Practical advice for people with

Ehlers-Danlos (inherited connective tissue disease or collagen)

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TO WHOM IT MAY CONCERN

These recommendations refer to patients who, despite their numbers, are rarely diagnosed or only after a painful and dangerous medical pilgrimage, on average about 20 years. The diagnosis is made, without a specific marker, but with a clinical criterion. These symptoms are easy to highlight, validated by international publications: **diffuse pain, severe fatigue, difficulties in controlling movements** (clumsiness, involuntary movements, pseudo-paralysis), joint instability (pseudo sprains, dislocations, joint blocks, joint crunches), **thinness and fragility of the skin** (excessive laxity, apparent veins, stretch marks, difficult healing, less protection against static electricity), diffuse joint **hyperlaxity**, hemorrhages, **hyperacusis** (being bothered by noise, feeling of uproar in the presence of several interlocutors, "musical sound", perception of feeble sounds and ultrasounds, better than dogs), **gastroesophageal reflux**. **FIVE of these signs** are sufficient for diagnosis. **The hypermobility** (more than hypermobility) that is **common**, is not necessary for the **diagnosis**, contrary to a general statement, it can even be replaced by **retractions** (shortening) of the muscles and their tendons (knees, ankles, more rarely, elbows and fingers). The pains that are very recurrent, make their evaluation random.

In general, if one refers to the complaints of the affected person, they go something like this: "everything hurts", "she hits herself easily", "her joints creak" and "she bleeds from many sources". Evidence of similar signs in the family provides proof that it is a hereditary disease, transmission to all children of a person (man or woman), who is affected. Evidence, which is very difficult to provide by means of a biological genetic test, in the current state of biomedical knowledge.

People with Ehlers-Danlos: they are a fragile population

Ehlers-Danlos patients have a special susceptibility to infections (Ears, nose and throat, bronchi, bladder, skin ...)

The excessive "flexibility" (hypermobility) of their ribs exposes them to painful muscular limitations (especially the diaphragm), responsible for respiratory distress, which is very frequent during exertion (climbing stairs).

Fluids may go by a false route, it is common with the simple flow of saliva or during feeding, producing bronchial floods, that cause cough and respiratory discomfort and sometimes pulmonary atelectasis.

People with emphysema are particularly vulnerable to pneumothorax (spontaneous rupture of the lung). Pleural effusion may occur.

The hyper-reactivity of the bronchial tissue with hypersecretions, the flaccidity of the bronchi, too soft (dilated), explains the possibility of increased secretions and bronchial dilation with superinfections. Direct damage to the bronchi or lungs by the virus, by decreasing the exchange of oxygen between inhaled air and blood (the alveolar-capillary barrier), according to Professor Philippe Juvin, from the Georges Pompidou University Hospital, can only have dire consequences.

What is known about the coronavirus COVID19 should make patients with Ehlers-Danlos disease be considered to be particularly vulnerable to this new viral disease. Therefore, they must be monitored with special attention and benefit, with priority of the therapeutic devices specially developed for viral disease (oxygen therapy in particular).

The complications of viral infection are to be feared: they worsen Ehlers-Danlos disease, as borreliosis that causes Lyme disease, has been observed in the past, under similar circumstances, after another infection.

Preventing people from going to their workplaces seems particularly justified in the case of Ehlers-Danlos, to protect these patients.

Practical tips

The results of temperature measurement in these patients, can be biased in both directions, due to their dysregulation in Ehlers-Danlos disease, due to the existence of dysautonomia (vasovagal alteration).

Some nasopharyngeal or respiratory disorders may be due to Ehlers-Danlos and should not be confused as caused by coronaviruses.

These people must be particularly vigilant, to avoid contamination, they must wear a mask in case that they are forced to go out of home, wash their hands very frequently, with a hydroalcoholic solution or soap, avoid direct contact with other people, sneeze or cough in a disposable handkerchief, leaving the shoes outside the house or apartment and taking off the used clothes outside, when entering the house.

They must drink water regularly.

It is desirable to disinfect the upper respiratory tract with sprays or gargles of antiseptic products, several times a day, especially in case of inflammation.

They must follow all effective treatments for the treatment of this disease, especially compressive garments and oxygen therapy (do 30-minute sessions, whenever possible)

A minimum of one hour of physical activity should be maintained a day, for the maintenance of proprioception, despite confinement, with self-rehabilitation exercises and exercises at home. Walk, if there is no period of confinement in your commune.

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