COVID-19 and ectodermal dysplasias. Recommendations are necessary.

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Dear Editor,

The term ectodermal dysplasias (EDs) refers to a heterogeneous group of rare congenital conditions affecting the normal development and/or homeostasis of two or more ectodermal derivatives including skin, teeth, hair, nails, and eccrine glands.[1-3] Hypohidrotic ectodermal dysplasia is estimated to affect at least 1/5,000–10,000 newborns.[1] X-linked hypohidrotic ectodermal dysplasia (XLHED; OMIM 305100),[3] is the most common subtype of EDs, with an incidence of 1/50,000–100,000 males.[1-3] XLHED is characterized by a clinical triad of hypotrichosis, hypo-, oligo- or anodontia, and hypo- or anhidrosis.[2,3]

XLHED is associated with the *EDA* gene located at (Xq12-q13.1), leading to loss or dysfunction of the signaling protein EDA (Wohlfart et al., 2020), a critical signaling unit involved in the interaction between the ectoderm and the mesoderm.[1-3] The dominant (OMIM 129490) and recessive (OMIM 614941) subtypes involve the *EDAR* (2q13), and *EDARADD* (1q42.3) genes, respectively. Moreover, X-linked anhidrotic ectodermal dysplasia with immunodeficiency (XL-EDA-ID, OMIM 300291) is a primary immunodeficiency disease. This is due to the mutation of *IKBKG* that encodes NF-κB (nuclear factor-kappa B essential modulator).[4,5]

Mutations in this gene can cause others disorders, including incontinentia pigmenti (OMIM 308300) and hypohidrotic ectodermal dysplasia with immunodeficiency associated with osteopetrosis and lymphoedema (OMIM 300301).[6] Clinical expressions of the disease are characterized by abnormal teeth, hypohidrosis, sparse hair, and the immunological defects of impaired antibody response to polysaccharides, hypogammaglobulinemia, and impaired natural killer cell cytotoxicity. Patients are susceptible to infections with pyogenic bacteria, mycobacteria, parasites, viruses, and fungi.[4]

The deficient development of other eccrine glands results in recurrent respiratory infections.[2] The presence of abnormal cilia and glands explains the pooling of secretions in the nasal cavities. Allergic rhinitis often worsen nasal symptoms and predispose to sinonasal infections. In order to reduce the nasal symptoms and infections, it is necessary to humidify the environment where patients normally live and to periodically and gently remove nasal crusting. Nasal saline solutions and nasal douches with sodium bicarbonate have demonstrated to be helpful for the purpose, as well as, topical therapy with vaseline-based antibiotic ointments have been proposed when an acute infection occurs.[6]

Given the clinical problems in some patients with EDs and the emergence of SARS-CoV-2, a new corona virus, responsible for the pandemic named Coronavirus Disease 2019 (COVID-19), which causes severe acute respiratory syndromes with a significant morbidity and mortality,[7] it is essential to mention some recommendations for patients affected by EDs. Firstly, social isolation can limit exposure to the contagion including relatives or associates living in the same home. Routine, non-urgent hospital visits should be postponed and telemedicine plays an important role in delivering clinical care.

Hand washes and disinfection with 70% alcoholic solutions are part of the preventive measures when dealing with patients in dermatology. Enveloped viruses like SARS-CoV-2 respond better to ethanol than propanol. The addition of other antimicrobial compounds to 70% alcoholic hand disinfections does not increase efficacy but may increase the risk of irritant contact dermatitis. Alcoholic hand disinfectants preserve the epidermal barrier better than hand washes with soap.[7] The use of face masks may also be necessary.

The global ED patient organizations came together in 2007 to form the International Ectodermal Dysplasia Network https://edinetwork.org/ providing a leading role in patient advice and support to ED patients and their families. The global registry of patients with EDs affected by COVID-19, their prognosis and clinical evolution will be necessary to study the presentation of the

infection in patients who have this genetic condition and thus be able to establish management recommendations.

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