COMMENTARY

Heart Failure in Epidermolysis Bullosa

Disfunción cardiaca en la epidermolisis ampollosa

The term inherited epidermolysis bullosa (EB) describes a group of genetic disorders in which a protein defect causes skin fragility and subsequent blistering. It is a rare disease, with a prevalence of just 8 cases per million population. It can affect deeper layers, and patients may also develop complications in other organs, caused not by the genetic defect itself but by its consequences. One such complication is dilated cardiomyopathy, which needs to be contemplated in patients with EB, not so much because of its frequency (which is low), but because of its severity and the important role of early diagnosis and treatment in prognosis.

The pathogenesis of dilated cardiomyopathy in patients with EB is unknown. It has been associated with numerous factors, such as deficiencies in certain micronutrients (iron, thiamine, selenium, carnitine) and iron overload, and it may be multifactorial. Very few case series of dilated cardiomyopathy in EB have been reported. In this issue of Actas Dermosifiliogrías, Batalla et al. present an interesting article that reports on the prevalence of this serious complication in a series of patients with EB seen at a referral hospital over a period of 30 years. The condition was most common, with a prevalence of 15%, in patients with recessive dystrophic EB, and this greater frequency has also been reported in other series. The authors point to possible causes of dilated cardiomyopathy in EB and also outline a follow-up plan aimed at early detection and treatment. Considering the low prevalence of both EB and dilated cardiomyopathy, more prospective, multicenter studies are needed to further define this association.

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