DYSTROPHIC EPIDERMOLYSIS BULLOSA IN ALGERIA: A CASE REPORT AND REHABILITATION APPROACH

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Background

Dystrophic Epidermolysis bullosa (DEB) is a rare genetic disease characterized by skin fragility with blistering that occurs spontaneously or following minor trauma such as mild pressure or friction. Children with DEB have a range of disabilities. The most severely affected may suffer from scarring, fibrosis, and contractures affecting any part of the body. Pain can be both acute and chronic, independence in activities of daily living can be significantly limited, and quality of life can be affected. There is currently no cure for DEB, but symptomatic therapies can be provided through a multidisciplinary team(MDT) approach where possible. Physical and rehabilitation medicine (PRM) can be an integral part of MDT. This poster highlights the interest of PRM in the management of deficiencies and disabilities related to this disease.

Case report

We present the case of M K, a 4-year-old child, admitted for the management of recessive dystrophic epidermolysis bullosa. Our initial assessment found: • uncooperative child, malnourished with growth retardation • bullous lesions with generalized scabs, global amyotrophy, • bilateral syndactyly due to synechiae • delayed psychomotor development. EBDASI score at 45/ 88, QOLEB score at 52/ 68 During his stay, the pediatric surgery team performed an esophageal dilation, the child was put on intravenous gentamicin course . Also, the child benefited from physiotherapy and occupational therapy sessions. After 3 months, we noted good wound healing. The child walks without technical aids, this functional progress is reflected by an improvement in the EBDASI scores (28/ 88) and the QOLEB score (31/ 68).

Conclusion

The management of DEB is multidisciplinary, rehabilitation by its means including occupational therapy, physiotherapy and equipment are an integral part. MPR management must be early and continuous in order to maximize functional autonomy and improve the quality of life for these patients.

Keywords: Epidermolysis Bullosa, EBDASI, QOLEB, Algeria