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Research Institution:	Eyeness AG, Switzerland
Project Title:	Eye care for children and adults with Epidermolysis Bullosa: Evidence-based guideline
Research Area:	Clinical Practice Guidelines
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Project Abstract

The extreme skin is fragile nature of Epidermolysis bullosa (EB), can also affect the eye layers. Eye problems for people living with EB may vary to any degree, where it is commoner in junctional and dystrophic types of EB, but more in their more severe forms, than in EB simplex. They are prone to corneal blisters or erosions due to the loosening of the skin cell layer bindings which are vital for corneal epithelium (this is the transparent layer on the surface of the eye cornea) [Fig 1]. This recurrent ocular surface lesions and erosions result in severe eye pain, light sensitivity, vision fluctuations, tearing, blepharospasm which leads to chronic inflammation and severe infection risks to the cornea and the adjunct conjunctiva. In chronic cases sight threatening scarring occurs leading very often to partial or, in very severe cases, to complete blindness.

Debra International identified this priority need for the development of a clinical practice guideline on eye care of EB since 2016. Furthermore, corneal erosions is one of a number of emergency situations may occur in the context of EB (Mellerio et al. 2020). In addition they recognised that whilst general management principles may apply, specific considerations are essential in managing EB to avoid undue trauma or damage to delicate tissues (Mellerio et al. 2020).

Fortunately, a variety of established clinical therapeutic options are available depending on the individual degree of the severity.

Clinical practice guidelines will help practitioners around the world to diagnose the severity, to evaluate the actual need for interventions and to choose from the most appropriate options for therapy.

Blog post written about project for website

Clinical practice guidelines (CPGs) are recommendations which have been developed by experts in a disease area to guide clinicians in the treatment of specific conditions. CPGs aim to improve the quality of care received by patients and ensure they are treated appropriately. EB is a rare disease and therefore can be unknown to medical staff, as they may not have come across it before. It is important that CPG's are developed for various procedures and treatments associated with EB to ensure clinicians do not unintentionally cause discomfort or harm to the individual.

DEBRA Ireland recognises the importance of CPGs and has previously collaborated with many international colleagues to fund the creation of CPGs in many indications related to EB. DEBRA Ireland is currently co-funding the creation of two CPGs, one for the treatment of oesophageal strictures and the other for eye care in EB. Below you will find descriptions of both CPGs, which DEBRA Ireland is hopeful will improve the quality of life and level of care the EB community currently receive in these areas, both in Ireland and worldwide. A full list of CPGs for EB is on the [DEBRA International website](#).

Eye care CPG

The fragility of the skin associated with EB can also affect the layers of the eye. Eye problems for those who live with EB can range from mild to severe but can affect all EB types. Those with EB are prone to corneal blisters or erosion of the cornea due to loosening of the skin layers. Eye issues such as this can lead to severe eye pain, light sensitivity, tearing and chronic inflammation of the eye area, amongst many other symptoms. Complete blindness can result in very severe cases. The development of this CPG is being led by Dr Baertschi based in Eyeness AG, Bern, Switzerland. The CPG is being designed to guide clinicians in the most effective ways to care for eyes in adults and children who have EB.

Quotes we have from Researchers

None

Researcher (s) Bio

None