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EYE:

Use of therapeutic Bandage Contact Lenses in Epidermolysis Bullosa

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Introduction

Due to a mutation on the collagen VII-Gen, which is part of the adhesion-complex of the corneal epithelial cells, between **14-35%** (Tong et al. 1999; Gans 1988) of all eyes in EB patients are affected with painful corneal blisters, corneal scarring, refractive irregularities and permanent reduced vision.





Recent best practice

"The condition and severity varies between individuals and over time."

"Calming and pain management are essential."



AUGEN

1. Einleitung

Probleme mit den Augen können in unterschiedlicher Häufigkeit bei den meisten Formen von EB auftreten. Betroffen sein können die Augenlider, die Bindehaut und die Hornhaut. Probleme in diesem Bereich sind besonders unangenehm und oft recht schmerzhaft. Dieser Abschnitt zeigt Ihnen Wege, wie die Häufigkeit von Augenproblemen reduziert werden kann und was zu tun ist, wenn sie auftreten. Darüber hinaus bekommen Sie hier einige Hinweise, die beim Anfertigen einer Brille zu beachten sind.



- Blasen und Verletzungen im Augenbereich können an Augenlidern, Bindehaut und Hornhaut auftreten.
- Häufig bei RDEB, immer wieder bei DDEB,
- Ruhigstellung und Schmerzlinderung sind die wichtigsten Erstmaßnahmen.
 - Erstmaßnahmen,
 - Vorbeugung durch regelmäßige Anwendung von Tränenersatzmitteln sowie Augensalben reduziert die Häufigkeit von Problemen.
 - · Alle Maßnahmen sind durch Kontrollen beim Augenarzt zu begleiten!

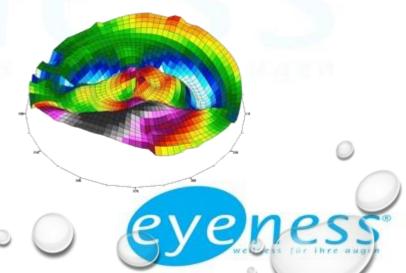






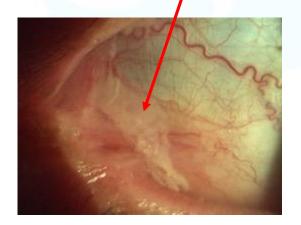
- Chronic eye pain
- Inflamed cornea and conjunctiva (red eyes)
- Light sensitivity "Photophobia"
- Tearing "Epiphora"
- Reduced, distorted and fluctuating vision





Long term complications

- Permanent corneal and/or conjunctival scars due to healing process
 (e.g. symblepharon, corneal scarring)
- Frequent medical/surgical interventions needed
- Chronic visual impairment due to massive neovascularization
- Isolation from society or working place
- Eye drops, gel or ointment only for less severe cases applicable

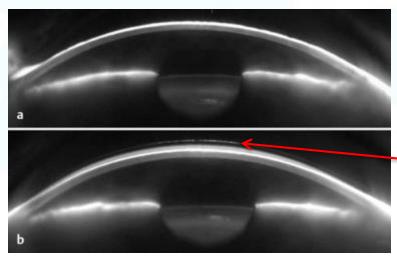






Method

Fitting of therapeutic and refractive effective soft or hard bandage contact lenses as ocular surface protection layer (since 2006).



Without contact lens

With protecting contact lens





EBEye Questionnaire (Feel free to optimize the naming, smileys or colors)

This Questionnaire was created to better understand your actual Eye Pain and Vision Situation and over time. Please indicate your personal feelings in the provided Scale.

Pain	Never			sometimes				Always			
Pain						5					10
No pain Morst pain Pain											
Intensity								7		9	10
Excellent			variable					Extreme bad			
	0	1	2	3	4	5	6	7	8	9	10
	<u></u>		sometimes Always							Always	
Tearing	Never										

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Method

EBEye Questionnaire





Method Bandage Contact Lens Fitting Procedure

Therapeutic Contact Lens Fitting Suggestions for EB Patients

with recurrent Corneal Blisters/Erosions, Pain Sensation and/or Epiphora

- 1. EBEye Questionnaire
- 2. Free Visual Acuity / Corrected Visual Acuity with current Eye Glasses
- 3. Autorefractor or Sciascopy (only if Photophobia is not to severe)
- 4. Ocular Topography or Autokeratometer
- 5. Slit Lamp / Biomicroscope Evaluation (Documented by Photography)
- 6. Fluorescein Vital Dye Stain (Documented by Photography)
- 7. Wearing Modality Decision (Extended/Constant Wear or Daily Wear)
- 8. Contact Lens Material Decision (Silicone-Hydrogel or Scieral Lens)
- 9. Basecurve, Over all Diameter and Contact Lens Power Calculation
- 10.Insertion of Trial Contact Lenses with unpreserved Saline Solution. After 5-15 Minutes
- 11. Check Trial Contact Lenses for spontaneous Comfort, Centration, Fit and Movement/Tear Exchange
- 12. Educate proper Contact Lens Handling and Hygiene (for Daily Wear with unpreserved Peroxid System)
- 13. Provide written Handling and Hygiene Instruction (Brochure) with **Emergency Contact Information**
- 14.Plan Follow-Up Visits depending the Wearing Modality
- 15. Record Progression by Photography and EBEyes Questionnaire
- 16. Educate your colleagues about the Possibility of Therapeutic Contact Lenses for EB Patients

Created for DEBAA International and sure-shity Dr. Michael Baertschi, Eyersen AG, Saitzerland; September 2008.

Suggested Soft and Hard Contact Lens Materials for EB Patients

Fundamentals:

- · Highest possible oxygen (Gas) permeability for extended or constant wear (EW 24h/7d/30d) up to 30 days and 29 nights. Ideal Dk/t > 100.
- · Lowest protein and oily deposits possible to reduce "biofilm" on lens surface over time and to maintain good eye health.
- · Best possible fit and lowest dehydration properties to maintain highest possible long-term comfort and ideal post-lens tear film exchange.

Silicon-Hydrogel (soft) contact lens brands with FDA extended/constant wear (EW) and therapeutic use approval:

- . Biofinity (COOPER Vision, UK) spheric, toric and multifocal, monthly lens for EW
- . Air Optix "Night&Day" (ALCON, USA) spheric, 2 different basecurves available, smaller over all diameter for smaller eyes/lid aperture, monthly lens for EW
- Acuvue Oasys (JOHNSON & JOHNSON, USA), spheric, toric and multifocal, 2 different basecurves available, weekly (!) lens for EW

Rigid Gas Permeable (hard) contact lens brands for Scleral Contact Lenses, only Daily Wear recommended even all of them have a FDA EW approval !:

- Optimum Extra/Extreme (CONTAMAC, UK) spheric, toric and multifocal
- XO/XO2 (BOSTON, USA) spheric, toric and multifocal
- HDS 100 (PARAGON, USA) spheric, toric and multifocal

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Results

"Immediate" elimination of (eye) pain sensation in minutes or hours.

"Whitening" of the eyes in days due to permanent reduction of ocular inflammation.





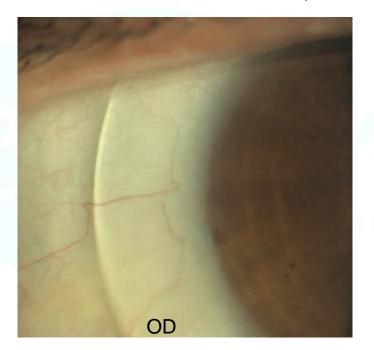
"Soft" lens example : Silicone-Hydrogel Contact Lenses





Results

Status after 13 years of contact lens wear





"Hard" lens example: Scleral Contact Lenses



Results

Rehabilitation of permanent and stable vision in a few weeks

Continuous support of the healing process of the corneal epithelium over months/years.

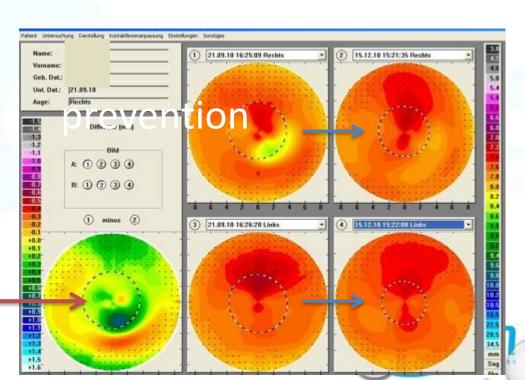
"Permanent" restoration of the irregular corneal surface.

Comparison:

OD 1 -> 2

OS 3 -> 4

Differential map: OD 1 -> 2 before and after treatment



Summarize

Bandage CL are serving as a protective layer between the ocular surface and the environment. In this role they are reducing the superficial stress to the ocular surface; e.g. less friction from the eye lids, less dehydration from environmental air/dryness and less burden from environmental foreign bodies like dust, pollen and microbes. All this:

- => lead to less acute or recurrent corneal erosions or lesions,
- => result in less necessity of wound healing processes,
- => results in less chronic ocular inflammation (basic clinical strategy of "inflammation inhibition")
- => results in normalizing the basal corneal epithelial cell reproduction and migration,
- => and therefore, reduces the importance of high oxygen demand,
- => As a final result bandage CL calm down the stressful physiological and pathological ocular surface situation.
- In addition, a more intact, stronger and more stable corneal and conjunctival epithelium support reducing the risk of microbial infections due to an intact first-line defense system. Despite the discussed reduced tear film exchange behind the bandage contact lens.
- All these advantages of bandage contact lenses are supporting reduced corneal scarring and fibrosis, are preventing vision disorders e.g. due to neovascularization and scarring before they occur, are eliminating itching and severe eye pain almost immediately and are increasing the quality of life of EB patients in a permanent way/manner.
- And do you know what ? It is completely easy to do so and a cheap therapy as well.

Bandage CL are a clinically proven, successful symptom-relief-therapy to prevent secondary inflammatory and fibrotic reactions of the ocular surface of EB patient eyes.

Conclusion

Therapeutic bandage Contact Lenses are an easy, quick and effective method to eliminate pain sensation, restore vision and ocular surface regularity on a "permanent" base.









 Protecting contact lenses increases the quality of life of EB patients significantly, quick, permanent and effective.

• Schützende Kontaktlinsen heben die Lebensqualität von EB PatientenInnen markant, rasch, permanent und effektiv.

• Lentille de contact protective améliorer la qualité de vie des patients EB marqué, vite, permanent et effective.



