

Challenges of Fitting Scleral Lenses on a Patient with Blepharophimosis

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Clinical Overview of BPES:

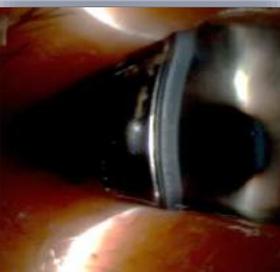
Blepharophimosis, Ptosis, and Epicanthus Inversus Syndrome (BPES)

BPES is an inherited condition that mainly affects development of the eyelids. The individuals affected by the condition have narrowing of the eyelids, droopy eyelids, and an outward fold of the lower eyelid. Due to these eyelid abnormalities, the eyelids cannot open completely, and vision may be limited resulting in obstruction amblyopia.

The inheritance pattern of BPES is Autosomal Dominant and the prevalence of BPES is unknown. Only one gene is known to cause BPES and that is FOXL2. This gene controls the production of the FOXL2 protein, which is involved in the development of the muscles in the eyelid as well as the growth and development of ovarian cells. Mutations in the FOXL2 gene result in the signs and symptoms described above.

Case History	
Chief complaint:	Patient presents for scleral lens fitting referred by a local corneal specialist. Patient reports halos, glare, and extremely dry eyes OU which started 10+ years prior to initial office visit.
Medical Hx:	Acoustic neuroma treated with radiation
Ocular Hx:	Irregular astigmatism OU, blepharophimosis OU, s/p multiple Blepharoplasty OU, Jones tube OS, Strabismus corrective surgery, MRSA infection OD secondary to Jones tube, corneal ulcer secondary to exposure keratitis, allergic reactions to preservative (BAK) in eye drops.
Ocular medications:	PF Refresh Optive drops, Celluvisc ointment several times per day.

Clinical Findings	OD	OS
Presenting VA cc:	20/100+ OD	20/100+ OS
Manifest:	OD: + 4.00- 2.00 x 146 VA 20/80+ ADD: +2.50	OS: + 0.50-1.00 x 060 VA 20/50+ ADD: +2.50
-Ocular Adnexa	OD: clear	OS: clear
-Lids/Lashes	OD: incomplete lid closure	OS: incomplete lid closure
-Bulbar Conjunctiva	OD: injected grade 3+	OS: injected grade 3+
-Palpebral Conjunctiva	OD: injected grade 3+	OS: symblepharon in nasal canthus, chronic injection
-Tear Meniscus	OD: poor lid-globe congruity secondary to ectropion	OS: poor lid-globe congruity-spilling tears
-Cornea	OD: neovascularization 360 and arcus senilis 360	OS: thick salzmann nodule, neovascularization 360 and arcus senilis 360
-Anterior Chamber	OD: deep and quiet	OS: deep and quiet
-Iris	OD: flat/intact	OS: flat/intact
-Lens	OD: grade 2 nuclear sclerotic cataract	OS: grade 2 nuclear sclerotic cataract

Diagnostic Lenses	OD	OS
T1:	Blanchard One Fit 2.0 BC: 8.20 Pwr: -0.50 Diam: 14.90 Over refraction: +3.50-1.00x165 20/50+  Evaluation: No impingement, minimal clearance 75 microns New order: smaller diameter and increased sagittal depth	Blanchard One Fit 2.0 BC: 8.20 Pwr: -0.50 Diam: 14.90 Over refraction: +3.50-3.00x120 20/50+ (flexure)  Evaluation: No impingement OU, touch OS over nodule, flexure due to tight eyelid OS, minimal clearance OU New order: steepen BC OU, smaller diameter OU increase CT OS
T2:	Final Lens: BC: 7.90 Pwr: +0.87 Diam 14.60 VA: 20/40 Evaluation: Good fit, no impingement, clearance 150 microns after 30 min+ settle time. Patient reports great comfort and clear vision.	Zenlens with microvault over Salzmann nodule (nodule parameters- 3mm width, 400microns depth at axis 135, 6mm decentered from center of lens) BC: 7.85 Pwr: +0.50 Diam: 14.80 Evaluation: Able to get the lens in, touching nodule, edge of lens touching symblepharon in nasal canthus causing discomfort, return the lens
T3:	 Patient can comfortably wear his scleral lens for 12+ hours and is thrilled to eliminate his dependency for artificial tears and lubricating ointments.	Blanchard Oblate fit 2.0 BC: 7.90 Pwr: +0.87 Diam: 14.60 Evaluation: The lens does not vault over nasal limbus, slight touch of nodule apex clearance 100microns initial centrally After 20min: The lens was difficult to remove, not clearing nodule, irritating eyelid margin upon insertion due to small palpebral fissure, return the lens. After extensive trials, it was determined that a scleral lens was not indicated for patient's left eye due to high risk of complication and difficulty of handling. Patient prefers spectacles over sclerals for cosmetic reasons Final Spectacle RX: PALs OD: plano Add: +2.25 20/40+ OS: +0.50-0.75 x 160 Add: +2.25 20/50+



Conclusions:

Scleral lenses can be utilized for patients with BPES. BPES is often associated with high amounts of irregular astigmatism and exposure keratitis due to severe restriction of eyelid function.

The scleral lens fitting process requires a balance between a large enough lens to meet the HVID requirements as well as small enough to allow insertion despite extremely small palpebral fissures.

