Abstract:
A patient with pellucid marginal degeneration (PMD) with unilateral Intacs, presents with mid-day fogging OD, discomfort in soft toric lenses OS, and moderate itchiness OS>OD.

Case History
38 year old East Indian female, lawyer
I. Chief complaint: mid-day fogging with OD scleral lens, discomfort with OS soft toric lens, itchiness and mucous discharge OS>OD
II. Ocular history: pellucid marginal degeneration (PMD) OU x 2004
   A. Attempted corneal gas permeable lenses without success due to failure to adapt
   B. Intacs OD x 2005
      1. Soft toric contact lenses OU worn after Intacs
      2. Corneal crosslinking (CXL) OU recommended by ophthalmologist in 2014, but patient declined
   C. Chronic allergic conjunctivitis
III. Medical history: unremarkable
IV. Medication: Alaway BID OU

Pertinent findings
I. Slit lamp exam
   A. OD: 2 intrastromal rings with clear center and well centered in deep stroma, mild apical thinning and steepening
   B. OS: Faint Fleischer’s ring, mild inferior thinning
   C. OU: Moderate fine papillary response on palpebral conjunctiva, mucous in tear film
II. BCSVA:
   A. OD: -3.75-3.00x090  20/100
   B. OS: -4.00-2.50x088  20/50
   C. Significant shadowing OU
III. Special testing:
   A. Tomography Oculus Pentacam
      1. OD: 49.4@93.7/54.2@3.7, moderately advanced central corneal ectasia
         a) No topography or tomography information pre-Intacs
         b) Comparison of tomography taken from 2015 shows increase in corneal steepening within 1 year
      2. OS: 41.1@96.0/46.5@6.0, inferior pellucid marginal degeneration
         a) Stable compared to 2015 tomography
   B. Anterior OCT by Optovue
   C. Pachymetry: Increased central thinning OD since 2015, stable inferior thinning OS
   D. Anterior segment photos

Differential diagnosis
I. Keratoconus
II. Keratoglobus
III. Terrien’s Marginal Degeneration
Diagnosis and discussion

Pellucid marginal degeneration (PMD) is a noninflammatory, progressive corneal disease characterized as bilateral peripheral corneal thinning. Typically, thinning occurs inferiorly from 4 to 8 o’clock, causing steepening superior to the thinned area and ectasia. There have been reports of patients with both keratoconus and PMD features, suggesting that PMD may be a variant of keratoconus.

Intacs is a reversible surgical treatment for PMD meant to flatten the cornea, increase contact lens tolerance, and mildly improve uncorrected visual acuity. It involves inserting two PMMA semicircular segments into the deep stroma, while maintaining a clear central optical zone with no loss of tissue. Corneal crosslinking is often done in addition to Intacs to provide the best outcome by strengthening the cornea to attempt to slow or halt progression of corneal ectasias. The current patient presented with mild to moderate pellucid marginal degeneration with Intacs in her right eye and had elected not to have corneal crosslinking done.

Treatment and management

Presenting lenses:
OD: Zenlens Oblate/9.00/16.0/+1.00DS/4.700SAG/+100 limbal curve zone (LCZ)/ steep-1 APS 20/25+1
Fit: 250um central clearance, thin inferior nasal clearance (30um), thin limbal vault, good EL, (−) blanching or impingement. Mild debris in tear reservoir. Deposits on surface of lens. 1 vial of celluvisc inserted into lens prior to insertion
OS: Biofinity Toric/8.7/14.5/-3.00-2.25x100 20/25+1
Fit: Good coverage, centration, and movement. 20 degree left rotation

Re-order OD lens with increased SAG, Fit OS into scleral lens.

CL 1
OD: Zenlens Oblate/9.00/16.0/+0.50DS/4.800SAG/+100 LCZ/steep-1 APS 20/20-1
Fit: 350um central clearance, thin limbal vault, good edge lift, (−) blanching or impingement, debris in tear reservoir
SOR: -0.50 NI
OS: Zenlens Prolate/7.10/16.0/-7.50DS/4.800SAG/+70 LCZ/std APS 20/30
SOR: -1.00 20/25+2
Fit: 300um central clearance, thin but adequate limbal vault, excessive edge lift inferior, (−) blanching or impingement, debris in tear reservoir, front surface deposits

Re-ordered OD lens with decreased SAG (to decrease debris build up and fogging) and increased LCZ, re-ordered OS lens with new power and toric periphery (to decrease debris buildup).

CL 2
OD: Zenlens Oblate/9.00/16.0/+0.50DS/4.700SAG/+130 LCZ/steep-1 APS 20/25-3
Fit: Harsh bearing over inferior nasal cone, adequate limbal vault, mild edge lift nasal and temporal, (−) blanching or impingement
SOR: +0.50 20/25-2
OS: Zenlens Prolate/7.10/16.0/-8.25DS/4.800SAG/+70 LCZ/steep-2 APS 20/25+2
SOR: -0.75 20/20
Fit: 300 central clearance, adequate midperipheral vault, superior limbal bearing, mild edge lift nasal and temporal, (−) blanching or impingement, toric markers at 7 and 2 o’clock
Re-order OD lens with steeper BC (to decrease midperipheral clearance in attempts to decrease fogging), decreased LCZ (to decrease debris build up and fogging), new power, and increase SAG (to increase central clearance). Re-order OS lens with new power and increase LCZ (due to limbal bearing).

CL 3
OD: Zenlens Oblate/8.44/16.0/-1.50DS/4.900SAG/+100 LCZ/-1 steep APS 20/25+2
Fit: 365um central clearance, good midperipheral and limbal clearance, good edge lift, (-) blanching or impingement, mild debris in tear film (asymptomatic)
OS: Zenlens Prolate/7.10/16.0/-9.00DS/4.800SAG/+120 LCZ/ std/2 steep APS 20/20+2
Fit: 410um central clearance, good midperipheral and limbal clearance, mild superior nasal limbal bearing without NaFl staining, good edge lift, (-) blanching or impingement, moderate debris in tear film (asymptomatic)

Finalized scleral lenses OU. Patient was prescribed Pataday qDay OU, which reduced all itching and irritation OU, and reduced mid-day fogging. She also continued to use 1 vial of celluvisc in each lens prior to insertion. She was advised to discontinue showering in her lenses and to try to stay away from fumes whenever she was in the kitchen to decrease fogging of her vision. These recommended lifestyle changes allowed the patient to have increased comfort and she was able to wear both scleral lenses for a majority of the day before experiencing mild foggy vision.

Optical coherence tomography (OCT): showed adequate central, midperipheral, and limbal clearance OU. Good edge alignment OU.

Patient experienced good comfort, fit, and visual outcome with the newly fitted scleral lenses OU, while maintaining good ocular health. She reported significantly less shadowing in vision with the lenses as compared to her spectacles. The Intacs eye (OD) was more challenging, requiring multiple parameter adjustments in comparison to her OS. The patient is to return to our clinic in 3 months to re-evaluate the fit of the lenses and her ocular health. She mentioned apprehension with additional surgical procedures, including corneal crosslinking, and may revisit this idea at a later date.

Conclusion:
While Intacs may decrease corneal irregularities and improve unaided visual acuity, it has not been proven to slow or halt progression of corneal ectasias, as shown by the current patient. When the procedure is done in adjunct with CXL there may be a better outcome and may slow progression of the disease. Therefore, Intacs with CXL can be a good option for patients with pellucid marginal degeneration who cannot tolerate contact lenses. Corneal transplants would be another surgical option for patients who have more advanced corneal ectasias and is reserved for those who have severe scarring or extremely poor visual capabilities.

Clinical pearls
1. Intacs may improve CL tolerance, but may also make CL fits more difficult. Therefore, all non-invasive options should be considered prior to any surgical intervention.
2. Be open to using advanced technology to improve CL fit, such as an OCT (for both clearance and edge lift), specular microscope, pachymeter, and/or tomography.
3. Always confirm the contact lens parameters to make sure that they match what was ordered, as errors from the manufacturer can occur.
4. Achieving an acceptable fit includes optimizing the tearfilm by managing associated conditions, such as ocular allergy and consideration of lifestyle environment.
References: