

## INTRODUCTION

Axenfeld-Rieger syndrome (ARS) is a group of genetic disorders most significantly recognized for its anterior segment dysgenesis of the eye. Corneal integrity is a principal concern for patients with a history of ARS presenting for a contact lens evaluation. This case report evaluates the efficacy of a piggyback system to improve centration of a reverse geometry gas permeable lens and preserve corneal health post penetrating keratoplasty in a patient with Axenfeld-Rieger Syndrome.

## CASE REPORT

A 42 YO WF with Axenfeld-Rieger syndrome and a history of cataract extraction OU, three failed corneal transplants OD, and one successful transplant OS presents for a contact lens fit OS.

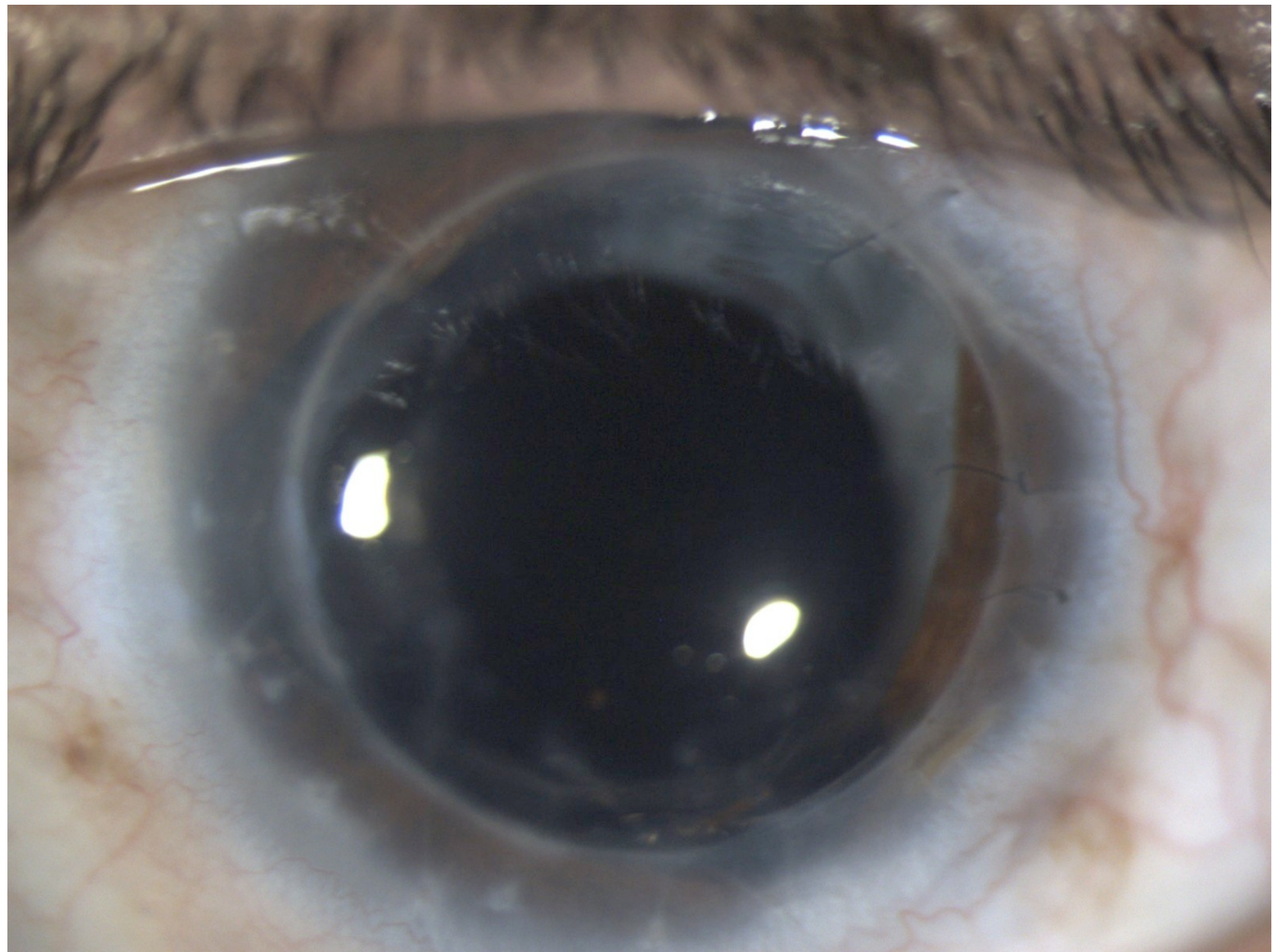


Figure 1: Appearance of OS s/p PKP

**Ocular History:** The patient has a history of Axonfeld-Rieger leading to corneal dystrophy requiring corneal transplants OU. Patient had three failed penetrating keratoplasties OD and one successful penetrating keratoplasty OS. Patient has severe stage glaucoma OU and has a history of tube shunts OU. The patient is managed with topical IOP lowering drops: Latanoprost QHS OU, Combigan BID OU, Azopt BID OU. The patient has a history of cataract extraction OU with PC IOL OU. The patient presents inquiring about possibility of improvement of vision OS with a contact lens.

**Habitual VA** (with spectacles): HM OD, 20/80 OS  
Current glasses: -2.25 -1.00 x 80 OD, OS

### Remarkable Exam Findings:

- Fixed, dilated pupil OU
- Iris deformities
- Constricted CVF OU
- Tube shunts OD 11 o'clock, tube shunt OS 2 o'clock
- PC IOL in good position OU
- Adequate host-graft junction OU s/p PKP, pigmented endothelium OU
- Flat, pale optic disc OU with a C/D of 0.95H/0.99V OD, 0.95H/0.90V OS

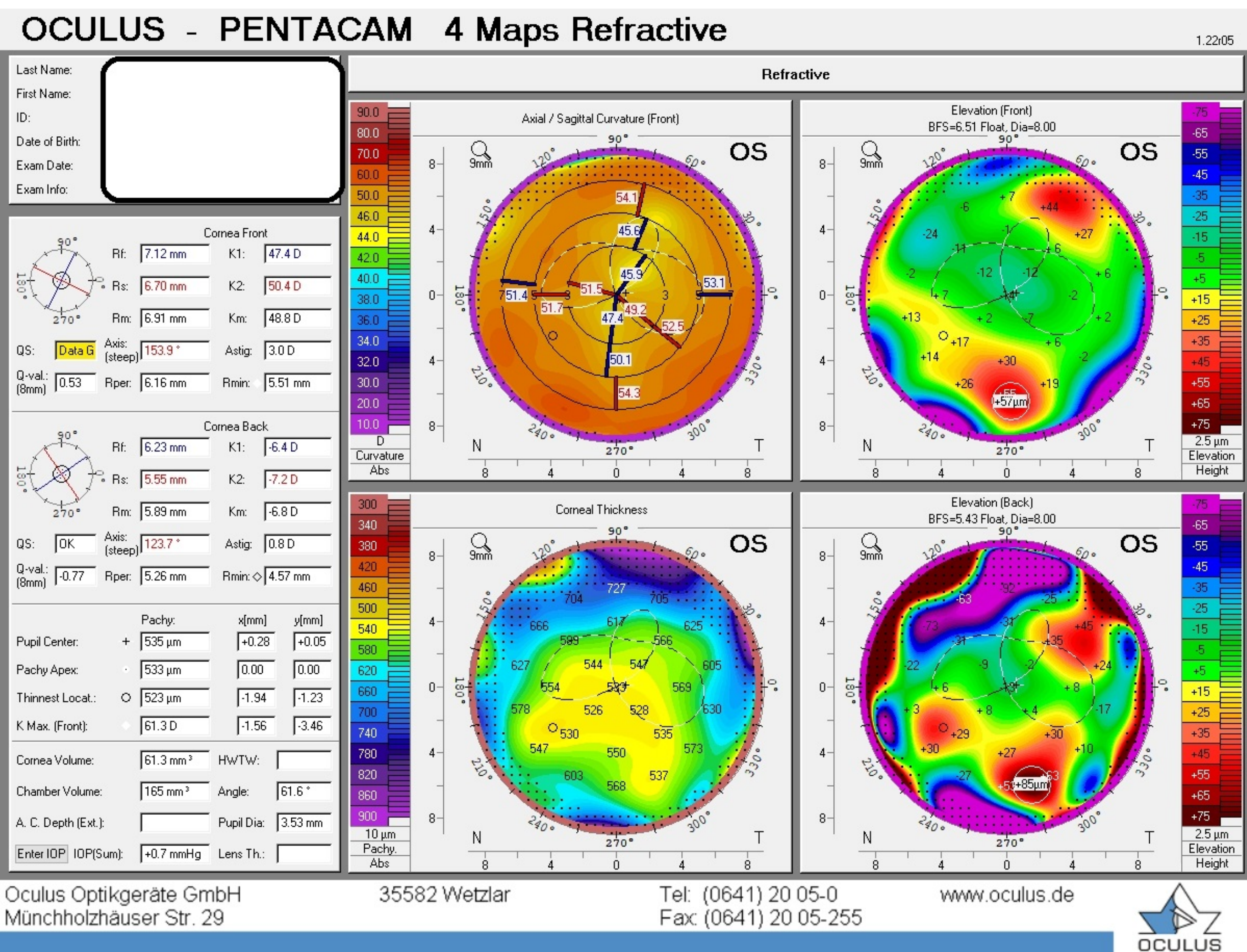


Figure 2: Topography OS s/p PKP

**Lens Fitting:** Based on topography, specifically the elevation map of the left eye, a scleral lens seemed like the best lens option initially however multiple factors such as the patient's history of penetrating keratoplasty and glaucoma had to be considered. Due to the fragility of the patient's corneal health and the diagnosis of glaucoma, scleral lenses were avoided to ensure optimal oxygen permeability and reduce the likelihood of IOP increase. A spherical corneal gas permeable (GP) lens was first trialed on the left eye.

Power	BC	OAD	2 C-R/W	3 C-R/W	4 C-R/W	5 C-R/W
-8.00	6.75	9.6	7.25/0.30	8.25/0.30	9.75/0.30	11.75/0.40

However, a spherical GP did not provide adequate fit and centration. Based on the previous spherical lens fit and elevation map from corneal topography, a reverse geometry lens in larger diameter was the ideal solution. A reverse geometry GP lens was then trialed on the left eye.

Power	BC	OAD	2 C-R/W	3 C-R/W	4 C-R/W	5 C-R/W
-4.50	6.70	10.00	6.40/0.60	7.5/0.20	8.65/0.20	10.65/0.20

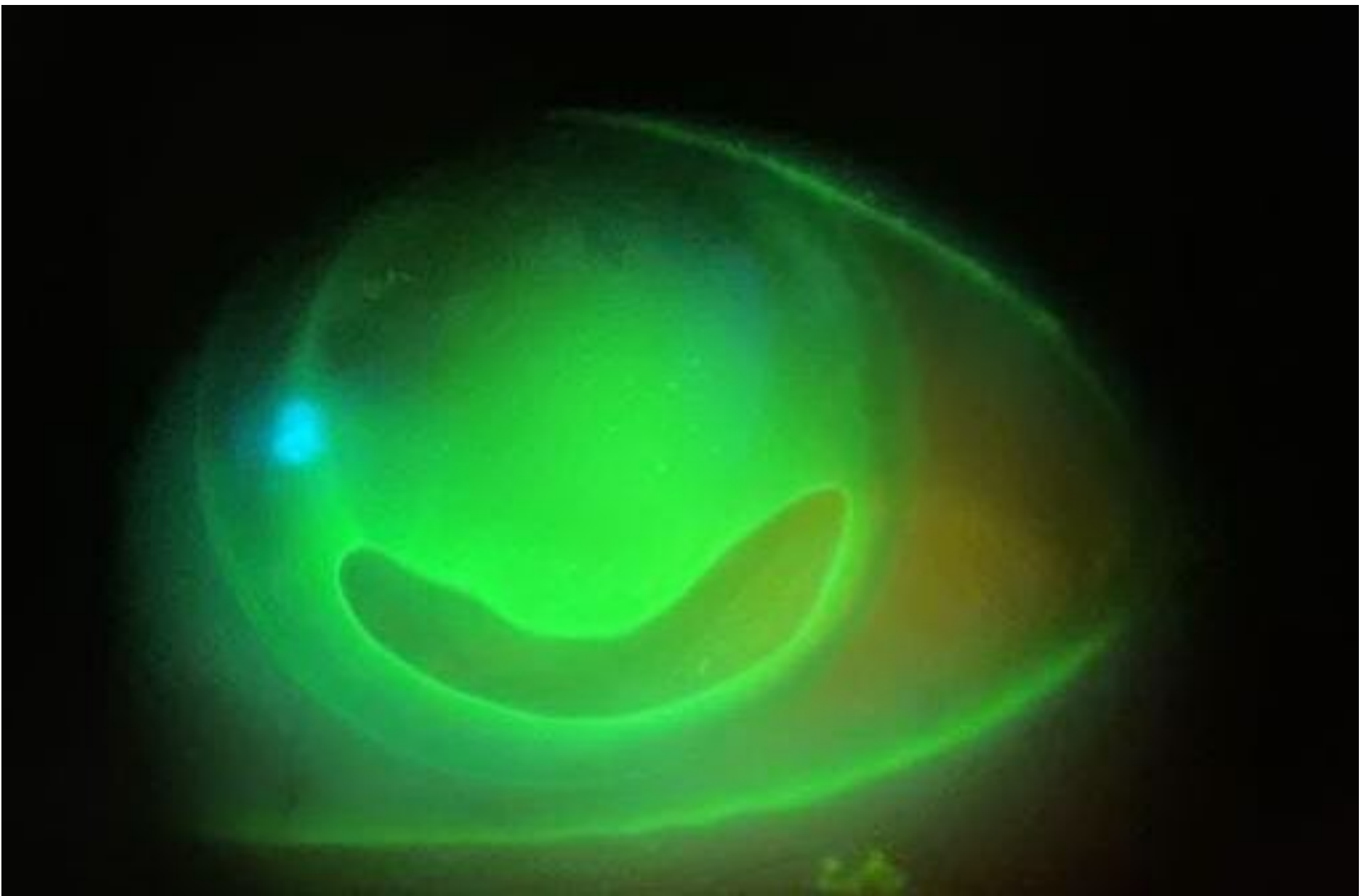


Figure 3: OS fit with reverse geometry GP lens

However, the reverse geometry GP lens had minimal movement and was decentered inferiorly with poor fit. A silicon hydrogel lens was used in conjunction with the GP lens to create a piggyback system (PBS) to improve patient comfort and centration of the GP lens.

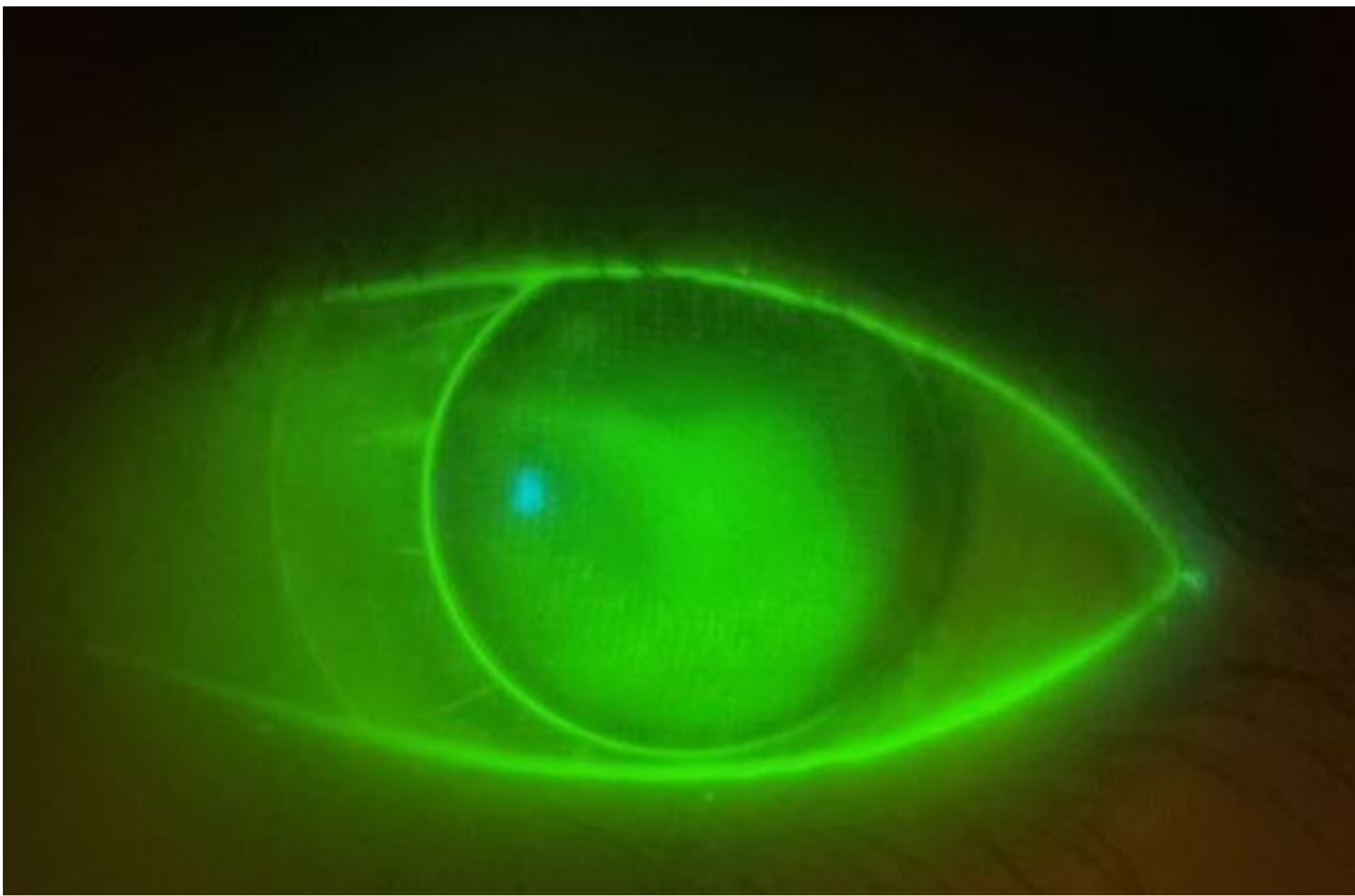


Figure 4: OS corrected with a PBS

The patient subsequently reported difficulty with removal of the soft contact lens and was refit with a silicone hydrogel colored lens for ease of handling. The patient achieved 20/25+1 vision in the left eye with the PBS and no longer had issues with insertion and removal of the soft lens. Fitting this patient with a PBS was an optimal solution to address issues with decentration, poor lens fit, and preservation of corneal health post penetrating keratoplasty. By adding a colored soft contact lens, the patient was easily able to visualize the SCL for insertion and removal. The importance of preserving corneal integrity was imperative in this case report as the patient relied only on the contact lens wearing eye for vision.

## DISCUSSION

Although ARS is known to be a rare condition with a prevalence around 1 in 2 million, the characteristic association of anterior segment dysgenesis prompts caution when fitting contact lenses. Patients with ARS typically have anteriorly displaced Schwalbe's line with iris and pupillary anomalies. Further complication may lead to corneal dystrophy, requiring corneal transplantation to preserve vision. Secondary glaucoma is a significant concern as 50% of patients with ARS develop glaucoma as a result of angle dysgenesis, often requiring surgical intervention. It is imperative to understand the typical sequelae associated with ARS as all factors must be evaluated for successful contact lens fitting. In a patient with ARS with confounding factors such as glaucoma and corneal transplantation, a piggyback system was found to be most favorable clinically.

## CONCLUSIONS

Post corneal transplant patients who have critical contact lens demands, but are deemed unfit for scleral lens wear, have limited possibilities. A PBS was a compelling option considering all underlying factors in this case report. The colored soft contact lens in combination with a reverse geometry corneal gas permeable lens found to be an acceptable option to provide improved comfort, fit, easy of lens handling, and life changing improvement in vision.