



## A CASE REPORT ON BILATERAL CONGENITAL ANIRIDIA WITH SUBLUXATION OF LENS

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**ABSTRACT** Aniridia is a rare genetic disorder with variable degree of hypoplasia or absence of iris. Here report a case of 20 year old female presented with gradual progressive loss of vision in both eyes with photophobia. On examination there was absence of iris and lens subluxated superiorly in both eyes. Parsplana vitrectomy with parsplana lensectomy done and SFIOL implanted.

**KEYWORDS** : Aniridia, Subluxation, Lens.

### BILATERAL CONGENITAL ANIRIDIA WITH SUBLUXATION OF LENS

Aniridia is a rare genetic disorder which consists of variable degree of hypoplasia or absence of iris.<sup>(1)</sup> May present from birth or later in the life. May associate with other ocular abnormalities. INCIDENCE ranges from 1:40,000 to 1:1,00,000.<sup>(2)</sup>

#### CASE REPORT:

A 20 year old female presented with complaints of gradual progressive loss of vision in both eyes and photophobia since 10 days. Difficulty in reading.

No complaints except glare in the past.  
Absence of iris since birth.



Family history : Nil significant.

figure -1

General examination

Patient is coherent and conscious

Temperature: 98.4F

Pulse Rate: 86beats per minute

Respiratory Rate 14per minute

Central Nervous System: No focal deficit.

Cardiovascular System: S1,S2 heard, no murmurs.

Respiratory System: Normal vesicular breath sounds heard.

At the time presentation visual acuity is counting fingers 1meter in right eye and counting fingers in left eye

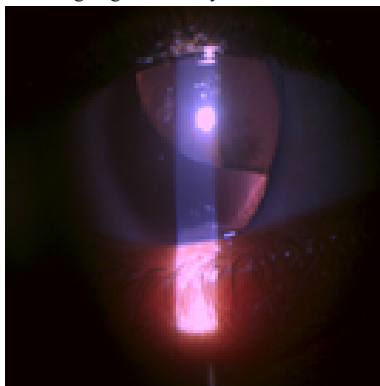


figure-2 right eye  
98.4F

Right eye

Vision cf 1m with +10d 6/60

Cornea : pigment on endothelium

Anterior chamber: superiorly subluxated lens

Iris and pupil : aniridia

Lens : superiorly subluxated with stretched zonules

Fundus : absent foveal reflex

**LEFT EYE**

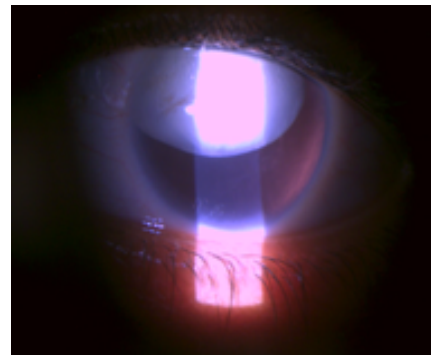


figure-3 left eye

Vision :cf 1/2m with +10d 6/36partial

Cornea :pigment on endothelium

Anterior chamber: superior subluxated lens

Iris and pupil : aniridia

Lens : cataractous and superiorly subluxated with stretched zonules

Fundus :absent foveal reflex

Iop measured with applanation tonometry : 32mmhg in both eyes.

On gonioscopy rudimentary iris stump seen.

Optical coherence tomography showed foveal hypoplasia in both eyes.

Diagnosis of congenital aniridia with superior subluxated lens with foveal hypoplasia made.

#### TREATMENT

Brinzolamide eye drops

Iop measured with applanation tonometer 18mmhg both eyes

Parsplana vitrectomy with pars plana lensectomy with scleral fixated lens done for left eye followed by right eye. Best corrected visual acuity 6/60 in both eyes.

#### DISCUSSION

Aniridia is rare congenital genetic disorder with variable degree of hypoplasia of iris or complete absence. Associated with other ocular manifestations like cataract, subluxated lens,

Corneal defects like aniridia associated keratopathy(aak)(3)

Ocular hypertension and glaucoma

Optic nerve hypoplasia.

Foveal hypoplasia.

Refractive errors and strabismus

Cataract surgery in aniridia patients is associated with increased risk of intra operative complications due to weak zonules.

3 phenotypes are identified

1 isolated aniridia without systemic involvement

2. WAGR syndrome

### 3.gilliespie syndrome

Most commonly inherited as autosomal dominant with pax6 mutation. Aniridia diagnosed by clinical examination.

Implantation of colored iris diaphragm lens following cataract surgery to improve postoperative outcomes by reducing glare, photophobia .Spectrum of corneal diseases treated with artificial tears, autologous serum and limbal stem cell transplantation. All patients with should be screened for glaucoma.Glaucoma treatment includes anti glaucoma drugs, trabeculectomy , glaucoma drainage devices.

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