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# ADIE'S TONIC PUPIL

Muhammad Arfan[1], Seskoati Prayitnaningsih[1], Wino Vrieda Vierlia[1]
[1].Department of Ophthalmology, Faculty of Medicine, Universitas Brawijaya,
Saiful Anwar General Hospital, Malang, Indonesia
Correspondence: *Muhammad Arfan, dr.arfan17@gmail.com*

**ABSTRACT**
**Background:** Adie's tonic pupil is a rare neuro-ophthalmological disorder which mostly presents in young female patient. Adie's tonic pupil is resulted from parasympathetic denervation of the ciliary ganglion. It is characterized by a dilated pupil with decreased response to light but preserved constriction response to accommodation, vermiform movements of the pupillary border, and hypersensitivity to pharmacologic constricting agent.
**Case Presentation:** A case report of Adie's tonic pupil in a young female patient. A 37-year-old woman came with complaint of different size of her pupils, accidentally found while performing medical check up. There were no signs of inflammation, blurred vision, or any other abnormality. Ophthalmological examination obtained visual acuity 6/6 on both eyes. Anterior segment examination revealed that left pupil was bigger than the fellow eye and reacted poorly to direct light stimulus. After one drop of dilute pilocarpine 0.1% given to the affected eye, the pupil did constrict. There was no abnormality found on the posterior segment.
**Conclusion**:The diagnose of Adie's tonic pupil was made through history taking, physical and medical examination.
**Keywords**:**:** Adie's Tonic pupil, cholinergic super sensitivity, Unilateral mydriasis.

## BACKGROUND

Adie Tonic pupil is a rare neurological disorder that causes one or both pupils dilate abnormally with impaired response to light. The disease is caused by denervation of the post ganglionic parasympathetic supply to the pupillary sphincter and ciliary muscle. Adie's tonic pupil can be caused by ocular and orbital mechanism after surgery, trauma, laser procedures, infection, inflammation or ischemia. This disorder usually occurs in young women that involves mostly one eye only. Fellow eye involvement usually develops within months or years afterwards. Damage to the ciliary ganglion or short ciliary nerve (post ganglionic parasympathetic nerve injury) produces tonic pupil, which is characterized by a poor reaction to light, sectoral paralysis of the iris sphincter, accommodative paresis, denervation of cholinergic sensitivity, and a strong pupillary response to tonic, near vision, followed by slow dilation.[1-3]

Patients with pupillary tonic may have accommodative symptoms or photophobia but most of them complaint no symptoms and the anisocoria may be noticed by the family of friends.[4-5] Anisocoria more than 1 mm with sectoral paralysis on the slit-lamp examination is the specific sign of Adie's tonic pupil. [2,4-7]

The diagnosis can be confirmed by clinical examination and topical confirmation with a low dose topical pilocarpine test. Clinical examination will often show sectoral paresis of movement of the iris sphincter or iris vermiform. Topical pilocarpine 0.1% can constrict the affected pupil, due to the super sensitivity of cholinergic denervation. Normal pupils usually will not constrict with a low-dose of pilocarpine.The use of reading glasses may be used to improve eye vision problems. A low dose of pilocarpine can be given to the affected pupil as well. [6-7]

We reported the rare case of a young woman with Adie's Tonic pupil on the left

eye. Establishing the diagnose of unilateral mydriasis is needed to rule out the other possible emergency causes.

## CASE PRESENTATION

A 37-year-old female patient was referred from the nearest hospital because of a difference in pupil size of both eyes while doing a routine medical checkup. The patient did not complain any blurred vision, pain or redness and no double vision. No history of eye or head trauma and no other systemic disorders.A tonic pupil can be caused by local ocular or orbital processes such as surgery, trauma, laser procedure, infection (eg, herpes zoster virus, herpes simplex virus, syphilis, botulism), inflammation, or ischemia.

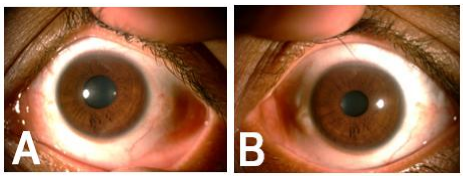
The vital signs were normal with normal body posture. General neurologic status showed no weakness and no reduced nerve sensations in all four limbs noted. Examination of the multiple cranial nerves also showed no abnormalities.


**Picture 1. A. More visible anisocoria in bright light condition. B. Anisocoria in dim light.**

The ophthalmological status examination revealed 6/6 visual acuity on both eyes. Anterior segment examination showed bigger left pupil size for about 5 mm with poorly pupillary reflex on light stimulus. The anisocoria was bigger in bright room condition. The left eye also revealed of segmental paralysis on the iris in slit lamp examination. Other parts of the eye remained normal. The fellow eye showed no abnormality.

Both Eye's position and ocular motilities were normal. Color vision and contrast sensitivity examination showed no impairment. The posterior segment of both eyes remained normal.

The laboratory examination found no abnormalities. A drop of 0.1% topical pilocarpine was administered to the affected eye and resulted in constriction of the pupil.




**Picture 2. A. Left eye picture before 0.1% pilocarpine administration.B. Left eye picture 1 hour after administration of pilocarpine.**

The patient was then diagnosed with Adie's tonic pupil based on physical and medical examination. Regular follow up with no specific treatment to monitor the progressivity were applied to the patient.

**Picture 3. Normal nine gaze eye position and ocular motility on both eyes**

## DISCUSSION

Anisocoria is the asymmetry of the efferent signal to the iris muscles resulting in a difference in the size of the two pupils, which may be either a physiologic or pathologic condition. Physiological anisocoria is quite common as a normal variant. Anisocoria of the same condition in dim and bright light indicates that the pupillary sphincter and dilator muscles are functioning well. If the reflex and pupillary dilation are symmetrical and consistent between the eyes, it may be a physiological anisocoria. Careful examination of pupillary reaction to light

and near stimuli, the difference in anisocoria in light and dark, and attention to distinctive associated signs and symptoms facilitate differentiating the abnormalities in pupil size and response to stimuli. If the light reaction is poor in both eyes but the near reaction is intact, the patient has bilateral light-near dissociation of the pupils. Argyll Robertson pupils are small and irregular and are characterized by light-near dissociation, variable iris atrophy, and normal afferent visual function. They are classically described with neurosyphilis, and the lesion is within the rostral midbrain and pretectal oculomotor light reflex fibers on the dorsal side of the Edinger-Westphal nucleus. Anisocoria worse in bright light is an abnormal condition. Abnormal dilated pupil can result from paralysis of the oculomotor nerve, Adie's tonic pupil, pharmacological mydriasis, angle closure glaucoma or trauma. Careful slit-lamp biomicroscopy of the iris should be performed in all patients with anisocoria to exclude structural iris abnormalities or damage. Abnormalities of the iris are a common cause of anisocoria. False-positive pharmacologic testing may result in patients with structural abnormalities of the iris that prevent dilation or constriction to pharmacologic agents. In these cases, it may be necessary to test the integrity of the pupil dilation or constriction. The typical presentation of the tonic pupil is isolated anisocoria that is greater in light. Patients often present with acute awareness of the dilated pupil. The clinical features of tonic pupil are Poor pupillary light reaction, segmental palsy of the sphincter, Tonic pupillary in near response with light-near dissociation, Cholinergic super sensitivity of the denervated muscles.[3-4,10]

Adie's tonic pupilis a neurological condition of an unknown origin. The pathophysiology of Adie's tonic pupil is damage to the ciliary ganglion. After damage to the ciliary ganglion, aberrant regeneration of fibers originally destined for the ciliary body now innervate the iris sphincter. A tonic pupil can be caused by local ocular or orbital processes such as surgery, trauma, laser procedure, infection (e.g., herpes zoster virus, herpes simplex virus, syphilis, botulism), inflammation, or ischemia. With prevalence 2 cases per 1000 population, Cases of Adie's tonic pupils are mostly idiopathic, occur in young women, and accompanied by complaints of near vision disturbances, photophobia, asymmetric pupils and often complain of difficulty reading due to accommodative paresis.[3-4,9-13]

Denervation of the iris sphincter is particularly sensitive to low-dose of parasympathomimetic agents such as topical pilocarpine. With the tonic pupil, the iris sphincter and ciliary muscles become supersensitive to acetylcholine, and thus when they are stimulated their response is strong and tonic and their relaxation is slow and sustained. Pharmacologic testing with low-dose pilocarpine (0.125%) may demonstrate cholinergic super sensitivity in the tonic pupil (more miotic response than the fellow eye).[3-4] The patient was later given the topical 0.1% pilocarpine to rule out the other possible causes. On the evaluation after 60 minutes of topical pilocarpine administration, the affected pupil did constrict. It may be suggested that the abnormality of the pupil is caused by Adie's tonic pupil. With the tonic pupil, the iris sphincter and ciliary muscles become supersensitive to acetylcholine, and thus when they are stimulated their response is strong and tonic and their relaxation is slow and sustained. The diagnosis of a tonic pupil can usually be made on clinical grounds alone. Once the diagnosis of the Adie's tonic pupil is confirmed clinically and/or pharmacologically, no neuroimaging examination are required[3-4]

This disorder usually resolves spontaneously within months of onset and

currently there is no specific treatment for Adie's tonic pupil. The treatment of Adie's tonic pupil is usually reassurance alone. Unequal bifocal reading aids or a unilateral frosted bifocal segment may be needed for patients with accommodative paresis. The use of topical low-dose pilocarpine has been suggested by some clinician for Adie's syndrome, but may precipitate ciliary spasm, induce myopia, cause browache, or worsen anisocoria due to miosis.[3-4]

## CONCLUSION

The diagnose of Adie's tonic pupil was established through history taking, physical and medical examination by administration of a low dose topical pilocarpine. Most cases require no treatment but regular follow up to monitor the progressivity.

## REFERENCES


1. Mannis MJ. *Kanski's Clinical Ophthalmology*: A Systematic Approach. LWW; 2016.Chapter 19; p810-1
2. Sirang, Z., & Jeeva, I. (2019).Holmes-Adie syndrome. *Ophthalmology Journal*, *4*, 44-5.
3. *American Academy of Opthalmology*, 2016. Neuro-Opthalmology: Basic and Clinical Science Course 2016-2017. Section 5; : Chapter 10; p. 198-9
4. Smith, Stacy, Andrew G. Lee, and Paul W. Brazis. *Clinical pathways in neuro-ophthalmology: an evidence-based approach*. Thieme, 2019.
5. Sinharoy, U., Chakraborty, D. P., Saha, S. P., & Mukherjee, J. (2016). Isolated dilated pupil: Is it adie'spupil?.*Archives of Medicine and Health Sciences*, *4*(2), 233.
6. Choudhury, E., Almarzouqi, S. J., Morgan, M. L., & Lee, A. G. (2016). Adie's Pupil (Tonic Pupil), Pharmacologic Testing. *Encyclopedia of Ophthalmology*, 1-2.
7. Iannetti, P., Spalice, A., Iannetti, L., Verrotti, A., &Parisi, P. (2009). Residual and persistent Adie's pupil after pediatric ophthalmoplegic migraine. *Pediatric neurology*, *41*(3), 204-206.
8. Martinelli, P. (2000). Holmes-Adie syndrome. *The Lancet*, *356*(9243), 1760-1.
9. Sandhya R, Sarda V. Unilateral mydriasis with cholinergic super sensitivity: a diagnostic dilemma--a case series report. Journal of Evolution of Medical and Dental Sciences. 2014 May 12;3(19):5334-4.
10. Kennedy SA, Noble J, Wong AM. Examining the pupils. CMAJ. 2013 Jun 11;185(9):E424.
11. Venkataraman A, Panda BB, Sirka C. Adie's Tonic Pupil in Systemic Sclerosis: A Rare Association. Case reports in ophthalmological medicine. 2015;2015.
12. Kanzaria HK, Farzan N, Coralic Z. Adie's tonic pupil. Western Journal of Emergency Medicine: Integrating Emergency Care with Population Health. 2012;13(6).
13. Moeller JJ, Maxner CE. The dilated pupil: an update. Current neurology and neuroscience reports. 2007 Sep 1;7(5):417-22.