



# RELATIVE AFFERENT PUPILLARY DEFECT



**Eye Learn**  
All about the Eye

Dr. Krati Gupta  
Dr. Saurabh Deshmukh

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## RELATIVE AFFERENT PUPILLARY DEFECT

1. Causes, grading and clinical importance of relative afferent pupillary defect. (5) J2016

### Afferent pupillary defect

**Absolute afferent pupillary defect** (amaurotic pupil) is caused by a complete optic nerve lesion and is characterized by:

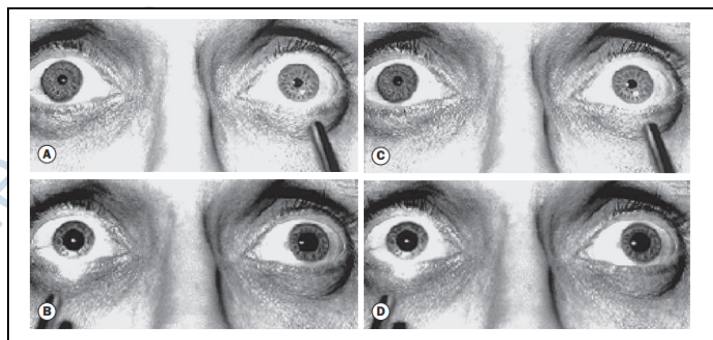
1. The involved eye is completely blind (i.e. no light perception).
2. Both pupils are equal in size.
3. When the affected eye is stimulated by light neither pupil reacts.
4. When the normal eye is stimulated both pupils react normally.
5. The near reflex is normal in both eyes.

### Relative afferent pupillary defect

- A relative pupillary defect (Marcus Gunn pupil) is caused by an incomplete optic nerve lesion or severe retinal disease, but never by a dense cataract.
- There is marked difference in the disease process in both the eyes which leads to an assessment of the “relativity”.
- The clinical features are those of an amaurotic pupil but more subtle.
- Thus the pupils respond weakly to stimulation of the diseased eye and briskly to that of the normal eye.
- The difference between the pupillary reactions of the two eyes is highlighted by the ‘swinging flashlight test’ in which a light source is alternatively switched from one eye to the other and back, thus stimulating each eye in rapid succession.

➤ A right relative defect is characterized by the following:

1. When the normal left eye is stimulated, both pupils constrict (A)
2. When the light is swung to the diseased right eye, the stimulus delivered to the constriction mechanism is reduced and both pupils dilate instead of constricting (B)
3. When the normal left eye is again stimulated, both pupils constrict once more (C)
4. When the diseased right eye is stimulated, both pupils dilate (D)



- It should be remembered that in afferent (sensory) lesions, the pupils are equal in size; anisocoria (asymmetrical pupil diameter) implies disease of the efferent (motor) nerve or the iris itself.
- As this is essentially an afferent defect, the presence of the consensual light reaction ensures that there is no anisocoria. The visual acuity need not necessarily correlate with the extent of an RAPD.

### Grading of RAPD

#### A. The “swinging flashlight test”

- It is a popular and an established way for identifying an RAPD.
- In this test, a strong, steady light is used.
- The light is shone into one eye, quickly switched to the other and is repeated back and forth.
- Since light in one pupil causes both pupils to constrict, quickly switching from one eye to the other will give a “relative” indication of the functioning of each eye and optic nerve.
- If both eyes are equally dysfunctional, no “relative” defect would be found.
- The results of the test can be graded as follows (as per the I-V system of Bell et al)

Grade I	initial weak constriction with greater redilatation
Grade II	initial stall and greater redilatation
Grade III	immediate pupillary dilatation
Grade IV	immediate pupillary dilatation following 6 seconds of illumination
Grade V	immediate pupillary dilatation with no secondary constriction at all

- Certain standard conditions have been identified for applying this grading.
- The patient fixes on a 6/60 Snellen letter at 6 meters in a darkened room.
- An indirect ophthalmoscope head set (standardized as for the luminance measurements) is used as the light source.
- A swinging flashlight test is then carried out with each pupil illuminated for 3 seconds.

- However subjective grading leads to discrepancies between clinicians, is difficult to quantify, and limits its use in diagnosis.

**B. Neutral density filters (NDF)** can be used to measure the relative afferent defect.

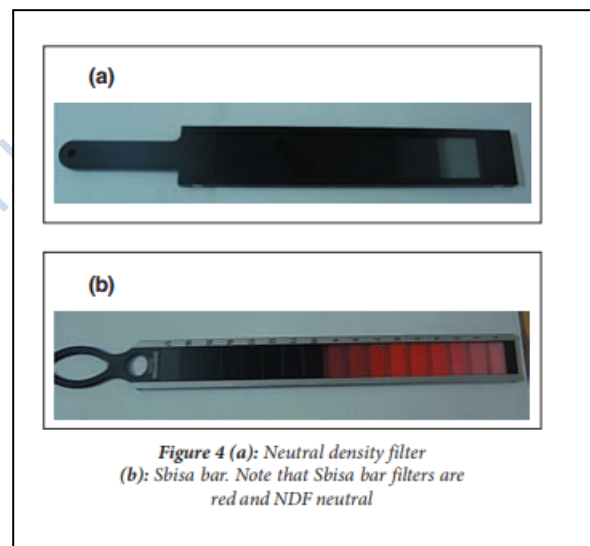
1. Graded denomination of the filter (NDF) is inserted in front of the better eye and the RAPD is assessed.
2. The density of the filters is measured in log units ranging from 0.3 to 3.0 in steps of 0.3
3. Beginning with the lowest 0.3 log<sub>10</sub> unit, the swinging flashlight test is repeated
4. The filters, placed in front of the normal eye, decrease the intensity of the light stimulus.
5. If an RAPD is still detected, increasingly stronger filters are placed over the normal eye until an RAPD disappears
6. At this balance point, the light input from the normal eye with the filter matches the light input from the abnormal eye. It implies the amount of afferent input that has to be decreased in the normal, unaffected eye so that it equals the input in the diseased eye.
7. The RAPD is quantified by the strength of the neutral-density filter needed over the normal eye to reach the balance point. That measurement of the NDF in log units which results in equalization of the pupillary reflexes gives the numerical magnitude of the RAPD.
8. Thus it numerically indicates the extent of pupillomotor deficit in the diseased eye with afferent defect, meaning thereby that the diseased eye which requires 0.9 log units of NDF in front of the better eye to neutralize the RAPD has poorer conduction than a diseased eye where the pupillary reflexes equalize with 0.5 log unit NDF in front of the better eye.

**C. Crossed polarized filter**

1. Crossed polarised filters (CPF) produce exponential attenuation of light and can be used in the same way as NDF to assess RAPD
2. Limitation with the use of NDF and CPF is their scarce availability

**D. Sbisa or Bagolini filter bar**

1. It is similar to NDF, and is used commonly by orthoptists to assess density of suppression in strabismus.
2. It consists of 17 sequential red filters.
3. Sbisa bar filters are red and NDF neutral,
4. It is the apparent brightness not color that determines pupil response
5. Sbisa bars are generally much easier to obtain than NDFs.



**Conditions not leading to a Relative Afferent Pupillary Defect**

<b>Refractive Error</b>	<b>Conditions with an Efferent Pupillary Defect</b> Third Cranial Nerve Palsy Adie's Pupil Horner's Syndrome
<b>Media Opacity</b> Cataract, Corneal scar, Hyphema, vitreous hemorrhage	
<b>Conditions which are typically bilaterally symmetrical will not show an RAPD</b> Bilateral retinitis pigmentosa Bilateral nutritional or metabolic optic neuropathies.	<b>Mild retinal diseases</b> Mild background diabetic retinopathy Central serous choroidopathy Non-ischemic vein occlusions Mild macular degeneration
	<b>Strabismus</b>
	<b>Previous eye surgery</b>

**Light near dissociation and the Argyll Robertson pupil:**

- The classical example of light near dissociation is the Argyll Robertson's pupil (ARP) classically observed in the now historical neurosyphilis, is postulated to be due to lesions in the intercalated neuron.
- In 1869, Douglas Argyll Robertson was the first to describe several patients with neurosyphilis whose frequently bilaterally miotic, and irregular pupils reacted poorly to light with a normal near response.
- These pupils in addition were highly recalcitrant to pharmacological dilatation.
- Generalized iris atrophy with loss of the normal pattern of the iris was seen in association with such pupils.



- The classical ARP was considered to be pathognomonic of tertiary syphilis.
- Optic neuropathy and RAPD do not cause anisocoria. Although the affected pupil reacts poorly, it does not dilate because the consensual input from the normal eye equalizes the pupil size.

**Conditions leading to a relative afferent pupillary defect include:**

<b>A. Optic nerve disorders</b>	<b>B. Ischemic retinal disease</b>
1. Unilateral optic neuropathies(mcc)	1. Ischemic ocular disease (Ocular ischemic syndrome): This usually arises from obstruction of the ophthalmic or carotid artery on one side. RAPD on affected side, subsequent to i. CRVO, CRAO, BRVO, BRAO, ii. Diabetic retinopathy Sickle cell retinopathy
2. Optic neuritis	
3. Ischemic optic neuropathies(GCA)	2. Retinal detachment: i. An RAPD can often be seen if the macula is detached ii. If at least two quadrants of retina are detached.
4. Glaucoma(1 eye>)	
5. Traumatic optic neuropathy	3. Severe macular degeneration: i. If unilateral and severe, an RAPD can be seen. ii. Usually the visual acuity would be less than 20/400.
6. Optic nerve tumor  i. Glioma ii. Meningioma iii. Sphenoid wing meningioma iv. Pituitary lesions	
7. Orbital disease i. Thyroid related orbitopathy (compression from enlarged extraocular muscles in the orbit) ii. Orbital tumors, iii. Vascular malformations	4. Intraocular tumor: lead to an RAPD if severe. i. Retinal and choroidal tumors including ii. Melanoma, retinoblastoma, and metastatic lesion
8. Radiation optic nerve damage	
9. Leber's optic neuropathy(B/L)	5. Retinal infection: can lead to an RAPD if extensive i. Cytomegalovirus, ii. Herpes simplex, iii. Other causes of retinitis
10. Optic nerve infections or inflammation i. Cryptococcus ii. Sarcoidosis iii. Lyme disease	<b>C. Amblyopia</b> If severe, can lead to a relative afferent pupillary defect. Usually the visual acuity would be 20/400, or worse.
11. Optic atrophy status/ post papilledema	<b>D. Cerebral vascular disease:</b>
12. Surgical damage to the optic nerve i. Retrobulbar anesthesia ii. Damage following orbital hemorrhage related to eye, iii. Orbital, sinus, or plastic surgery iv. Damage following neurosurgical procedures such as pituitary tumor resection v. Damage related to migration of an orbital plate after surgery to correct a blow-out fracture	i. Usually, it is an optic nerve disorder that leads to an RAPD, rather than an optic tract or visual cortex disorder. ii. However, there tends to be a higher percentage of crossed vs. uncrossed nerve fibers at the optic chiasm. iii. Thus, in a patient with a homonymous hemianopia from an optic tract disorder, an RAPD could be seen in the eye with the temporal visual field defect. iv. The nasal retina serves the temporal visual field, and these are the fibers that would cross at the chiasm

**Step wise approach to a patient of RAPD**

- i. Knowing whether the patient is aware of changes in vision.
- ii. If so, are these changes acute or gradual? An acute optic neuropathy or retinal detachment would be associated with sudden vision loss, while gradual vision loss might support the existence of a compressive lesion.
- iii. Eliciting significant ocular and medical history.
- iv. Does the patient have a history of vascular disease, cancer, auto-immune disease, recent infections or trauma?
- v. Carefully examining the patient while paying special attention to the retina and optic nerve.
- vi. Optic nerve findings can be subtle or nonexistent.
- vii. For instance, a relatively normal appearing nerve might be present in a retrobulbar Optic Neuritis.
- viii. The need to refer for further testing depends on the case.
- ix. If the patient's history and retinal examination do not offer an obvious explanation for the RAPD, one must assume that a condition affecting the optic nerve or optic tract is causing it.
- x. An immediate visual field and color vision test should be performed.
- xi. Depending on the condition, the patient may need to be referred for emergent neuro-imaging and laboratory testing .

- xii. For example, an individual who presents with an RAPD as a result of swollen optic nerve and a history of leukemia, would require emergent imaging and radiation treatment if the swollen nerve represented a leukemic infiltrative optic neuropathy.
- xiii. Computed tomography (CT Scan) and Magnetic resonance imaging (MRI) are valuable tools for diagnosis of optic nerve disorders (especially optic nerve tumours and traumatic optic neuropathy) and in orbital diseases causing compressive damage to optic nerve.

### **Characteristics of the Relative Afferent Pupillary Defect**

- i. A RAPD is an index of significant optic nerve or retinal disease, when there is a difference in the disease process between the two eyes.
- ii. If both eyes have severe but equal disease, there will be no RAPD.
- iii. Thus, a “bilateral” RAPD does not exist. Severe disease in one eye, causing a RAPD, will not result in anisocoria.
- iv. The pupil of the diseased eye will be equal in size to that of the other eye due to the consensual light reaction (unless the iris itself is diseased or unreactive).
- v. One functioning pupil is sufficient to determine the presence of an RAPD, (due to the presence of consensual light reflex).
- vi. The visual acuity does not necessarily correlate with an RAPD.
- vii. Some conditions lead to a marked reduction of visual acuity with an RAPD, while others spare the central vision.
- viii. Usually, extensive loss of peripheral vision is associated with an RAPD.

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