

Argyll Robertson Pupil

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The Argyll Robertson pupil, a miotic pupil that responds to an accommodation effort but fails to react to direct light, has been described in medical literature for more than a century. This pupillary reaction (**Sidebar**) is a simple way to confirm the integrity of the optic pathways and is a marker of such neurological disorders as neurosyphilis, neurosarcoidosis, and multiple sclerosis. The test requires no special equipment, holds great diagnostic yield, and takes less time to elicit than is needed to pronounce the epnym.

HISTORIC PERSPECTIVE

Douglas Argyll Robertson was born in Edinburgh, Scotland, in 1837. Robertson obtained his medical degree from St. Andrews University (Edinburgh, Scotland) in 1857. He studied ophthalmology in Prague, Czechoslovakia, and worked in Berlin, Germany, with the German ophthalmologist Von Graefe, the preeminent ophthalmologist of that era.¹

Robertson's first landmark contribution to ophthalmology came in early 1863, when he reported the ocular effects of the Calabar bean.² The active agent of the Calabar bean is physostigmine, a cholinesterase inhibitor. Robertson showed the antagonistic property of the Calabar bean to atropine, and this agent became the first effective medication to treat glaucoma.

Robertson first described the Argyll Robertson pupil in a case report of a patient with spinal disease in 1863.³ Ten months after he published his first case report, Robertson published four more similar cases. Although the absence of pupillary light response in patients with spinal disease had been reported previously,⁴ Robertson was the first to realize that the pupils still reacted to near stimuli. This dissociation between light and accommodation reflexes had been reported by Von Graefe in 1856 and even earlier by Italian psychiatrist Vincenzo Chiarugi (1759–1820) in 1793,⁵ but their reports were primarily unnoticed.

Robertson believed the responsible lesion could be found in the cervical spinal cord, and he termed the

ARGYLL ROBERTSON PUPIL

Definition: Miotic, irregular pupil that does not react to light but responds to accommodation, which indicates a lesion in the rostral midbrain.

Elicitation: To check for pupillary reflex, lights should be switched off in the examination room. A flashlight is shone in one eye at a time and both the direct and consensual reflexes (ie, reflex in other pupil) are observed. The light is then switched on, and after a few minutes pass for the patient to adjust to the ambient light, the patient is asked to look straight at a distant object. Either the examiner's finger or the tip of a pen is placed about 4 to 6 inches in front of the eyes at the level of the bridge of the nose and patient is asked to switch gaze from the distant object to the object in front of the patient. Pupillary response to this near focus is noted.

Normal response: Pupils are regular and constrict promptly and equally to accommodation and to direct and indirect light.

Positive response: Brisk pupillary constriction occurs after accommodation. Pupils do not respond to light.

disorder *spinal miosis*. Almost three decades after Robertson's description of spinal miosis, such disorders as tabes dorsalis, general paresis, and central nervous system lues were finally linked and recognized to represent the spectrum of the single entity, neurosyphilis. The Argyll Robertson pupil then became generally

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accepted as a pathognomonic sign of neurosyphilis, and this sign has since been identified with other central nervous system diseases as well.

PHYSIOLOGY

Light Reflex

From an inverted image on the light-sensitive cells of the retina, impulses pass through the optic nerve to the optic tract via the optic chiasma. The fibers partially cross at the chiasma. The optic tracts then pass to the lateral geniculate body; some fibers pass to the midbrain as the afferent limb of the pupillary light reflex. From the midbrain, information is relayed to the parasympathetic preganglionic neurons located in the Edinger-Westphal nucleus. The efferent fibers from the Edinger-Westphal nucleus pass through the ciliary ganglion and supply the sphincter pupillae muscles. The partial crossing of the optic nerve fibers at the optic chiasma explains the consensual reflex. Also, optic tract fibers pass through the pretectal nuclei and are then redistributed to both sides.

Accommodation Reflex

A blurred image of a near object is formed on the visual cortex. Cortical connections are then relayed through the frontal cortex to the oculomotor nerves that control the medial rectus muscles, causing the eye to converge. Fibers from the visual cortex are also relayed through the temporal lobe to efferent parasympathetic fibers originating in the ciliary ganglion. This causes ciliary muscle contraction, which increases lens convexity and brings the near object into focus on the central retina. Concomitantly, pupillary constriction occurs, mediated by oculomotor parasympathetic outflow, which enhances optic resolution.

Pathophysiology

In an Argyll Robertson pupil, the pupil's better response to accommodation than to light stimuli occurs because the lesion involves the more dorsally located fibers that subserve the pupil's response to light. The lesion spares the more ventrally located fibers that subserve the pupil reaction to near stimuli.

CLINICAL PRESENTATION

Typical Argyll Robertson pupils are small and irregular and react to accommodation but not to light. Initially, the pupil's response to light may only be sluggish, but the accommodation reflex is always more pro-

nounced and the light reflex eventually disappears. The exact site of the lesion is debated. The lesion is generally believed to be in the rostral midbrain proximal to the oculomotor nuclei. In some patients, magnetic resonance imaging studies have localized the lesion to the Edinger-Westphal nuclei.

The Argyll Robertson pupil has become a rare diagnostic sign of neurosyphilis. If neurosyphilis is suspected, examination may reveal clues such as ptosis; ataxia (positive Romberg's test); tremors of the mouth, tongue, outstretched hands, and whole body; diminished or absent tendon reflexes; or impaired vibratory and joint position sense. Severe cases of neurosyphilis may even demonstrate aortic incompetence, gastric crisis, and Charcot's arthropathy. Finally, Argyll Robertson pupils in the patient with neurosyphilis are invariably bilateral. However, with the declining incidence of neurosyphilis, the Argyll Robertson pupil is increasingly likely to indicate another disorder, such as sarcoidosis, multiple sclerosis, and, occasionally, diabetes mellitus.⁶

DIFFERENTIAL DIAGNOSIS

Argyll Robertson pupil should not be confused with Adie's pupil, which may yield a similar result on cursory examination. In Adie's pupil, which is caused by ciliary ganglion destruction, the reaction to light is absent or greatly diminished when tested in the routine examination; however, Adie's pupil does react slowly with prolonged maximal stimulation. Furthermore, once the Adie's pupil reacts to accommodation, the pupil tends to remain tonically constricted and dilates very slowly. HP

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