ARGYLL ROBERTSON PUPILS TRUE AND FALSE

A POST-GRADUATE LECTURE DELIVERED AT THE ROYAL LONDON OPHTHALMIC HOSPITAL

BY

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The true Argyll Robertson phenomenon is an infallible sign of syphilis; yet pupils that react on accommodation but not to light, the usual definition of an Argyll Robertson pupil, are met with in many conditions unrelated to syphilis. If both of these statements are true there must be something wrong with the usual definition. There is. It defines nothing; hence the prevailing confusion. I tell you that the sign is infallible. Others give you a long list of conditions in which it has been observed, and you do not know what to believe. I am afraid it is the same with much your teachers tell you. The chief reason for these discrepancies is failure on our part to agree upon a definition of the terms we employ; sometimes we are using different names for the same thing; in this instance we are applying the same name—the Argyll Robertson pupil—to a number of different things; often enough we are using names for nothing at all.

NEED FOR ACCURATE DEFINITION

If we are to understand one another, then, we must decide what it is we ourselves are thinking about, and what we expect others to think about when we use the term Argyll Robertson pupil; we must agree upon a definition. I give you a choice. You may define it as one that reacts on accommodation but not to light. If that is your definition, the Argyll Robertson pupil has been found not only in various forms of syphilis of the nervous system, but also in non-syphilitic congenital cerebral defects, cerebral haemorrhage and thrombosis, tumour, myelitis, arterio-sclerotic and senile dementias, internal hydrocephalus, meningitis, disseminated sclerosis, trauma, syringomyelia, progressive muscular atrophy, polio-encephalitis, encephalitis lethargica, Friedreich's disease, chronic alcoholism, diabetes, nicotine and carbon disulphide poisoning, and in various other conditions of unknown origin. I could add to this list, but I think it is long enough to show you that your sign is hardly unequivocal. It has about as much value for differential diagnosis as fever or a headache. On the other hand, if you care to base your definition on the description of the man after whom the sign is named—surely it is not fair to his name to do otherwise—you possess yourself of the most valuable of neurological signs, and one that is perhaps nearer to being unequivocal than any other in medicine.

ARGYLL ROBERTSON'S PAPER

After this introduction you may wish to know exactly what Argyll Robertson did describe. In a short, unimportant-looking paper, published in the *Edinburgh Medical Journal* in 1869, with the title "Four cases of spinal miosis, with remarks on the action of light on the pupil," he says he observed that, although the retina was quite sensitive and the pupils contracted at once during the act of accommodation for near objects, an alteration, however, in the amount of light admitted to the eye did not influence its size, and that slow and only partial dilatation ensued on the application of a strong atropine solution. If you study this statement you will see that it says much more than is implied in the usual short definition.

Time has proved that he wrought better than he knew; pupils that conform in every particular to his description are peculiar to syphilis; if they differ in any way they are no longer certain evidence of syphilis. Let us study the essential features in detail. The sign was found in patients with "spinal miosis." There is no agreement on what constitutes miosis; let us define it as a pupil with a diameter not exceeding 2½ mm. The typical Argyll Robertson pupil is small, often very small; I advise you to retain this feature in your definition, and to be very careful in forming your opinion if the pupil is not definitely on the small side. When it is inactive to light, but not small, you will find, as a rule, that the reaction on convergence is also defective; it is then an incompletely fixed pupil, not an Argyll Robertson pupil. The patient may still be a syphilitic, but there are many other causes of an incompletely fixed pupil. The sign was present in both eyes; we know now that it may be unilateral with the same significance. If it is genuine, the Argyll Robertson pupil is always the smaller one. Again I advise caution; many unilateral Argyll Robertson pupils that have been reported were false; a pupil on one side that does not react to light but reacts on convergence is found in various non-syphilitic conditions.

The retina was "quite sensitive"—that is, vision was good. The pupil of many a blind eye reacts on convergence but not to light, but that does not make it an Argyll Robertson pupil. Further, it may react consensually; the Argyll Robertson pupil never does. I mention this because one of you told me recently that the commonest cause of an Argyll Robertson pupil is blindness. The sign is often associated with tabetic optic atrophy; in these cases the light reflex is lost, perhaps without exception, however slight the visual failure from this cause may be. If vision is defective from some other cause—for example, optic atrophy from pressure by a pituitary tumour—some reaction to light persists so long as any vision remains, and may even do so after vision is lost. In such a case the pupil reacts very sluggishly to light but well on convergence; but it is not an incomplete Argyll Robertson pupil. You may not diagnose an Argyll Robertson pupil with complete blindness, and it is unsafe to diagnose an incomplete Argyll Robertson pupil if vision is defective. The cause of the eye signs must be sought for elsewhere.

Reaction on Convergence

Typically the true Argyll Robertson pupil contracts at once and fully, often excessively, on convergence. Thereafter it dilates again promptly. I must insist on these essential features; their neglect has led to many errors. We shall return to them when we come to consider partially fixed, myotonic, and other spurious Argyll Robertson pupils. For the moment I content myself by advising you to examine the reaction on convergence very carefully; if it is defective or peculiar in any way the pupil is not a true Argyll Robertson pupil, and you are not justified in concluding from the eye signs alone that your patient has had syphilis.

Variations in Illumination

The amount of light admitted to the eye does not influence the size of the pupils. Using ordinary bedside methods they do not contract when the illumination is increased, nor do they dilate when the eye is shaded. This is another feature that often serves to distinguish the true from the false. The Argyll Robertson pupil remains the same size for days, weeks, months on end, whatever the amount of light that falls upon it; false Argyll Robertson pupils often vary in size from time to time, or dilate when shaded, and may then react briskly to light.

Effect of Mydriatics

Mydriatics cause "slow and only partial dilatation"; whereas pupils that react on convergence but not to light may often be proved false in that they dilate rapidly and completely under the influence of atropine or cocaine.

True Argyll Robertson Pupil

Small pupils, constant in size, unaltered by light or shade, contracting promptly and fully on convergence, dilating again promptly when the effort to converge is relaxed, and dilating slowly and imperfectly to mydriatics —these are the essential features. The pupils may be irregular or unequal ; the lids may droop or be retracted. If these are the ocular signs, and these alone, the pupils by our definition are true Argyll Robertson pupils, and they indicate syphilitic infection of the nervous system. I need hardly say that syphilis may produce many other pupillary and ocular signs ; my point is that nothing else is peculiar to syphilis.

Partial Argyll Robertson Pupil

This is firm ground. Let us see if we may proceed further with safety. If you find that the response to light, direct and consensual, in one or both eyes is certainly pathologically slow or incomplete, and the other criteria are fulfilled, this " partial Argyll Robertson sign " has the same value as the complete phenomenon, but only if vision is good.

Pupil in Encephalitis Lethargica

Some alleged exceptions to the rule that the sign is certain evidence of syphilis may now be considered. It has been stated so frequently that it may be caused by encephalitis lethargica that you have come to believe it. During the last eight years I have made a careful *ad hoc* examination in one hundred and fifty-seven cases, and the numbers given by others who have made the same investigation reach a total of many hundreds ; no critical observer has ever seen a true Argyll Robertson pupil, and if it occurs at all it must be extremely rare.

Pupil in Quadrigeminal Tumours

The pupillary signs produced by tumours of the quadrigeminal region are important for discussions on the site of the lesion responsible for the Argyll Robertson phenomenon, but they can hardly be confused with the true Argyll Robertson sign as I have defined it. The pupils are rarely small, they may vary in size under observation, dilate in the dark, react after a sojourn in a dark room, dilate well with mydriatics, react imperfectly on convergence, or present some other distinguishing feature. As a rule, there is an associated conjugate palsy of upward ocular movement, papilloedema is present, and other symptoms point to the existence of a tumour.

Among a large number of others, diabetes, disseminated sclerosis, syringomyelia, chronic alcoholism, and cunningly placed vascular, inflammatory, and traumatic lesions, have been named as possible causes. All these things are possible, but you may safely ignore them. Of the recorded cases that I have looked up not one bears scrutiny.

FIXED PUPILS

Pupils that react neither to light, direct or consensual, nor on convergence, are said to be fixed. The name is unfortunate. They are not fixed like the pupil of a glass eye ; indeed the normal contraction that occurs on forcible closure of the lids is often excessive and unusually easy to observe in pupils with loss of the reactions to light and convergence. Here again you see the disadvantage of using ill-defined expressions. If your records are to be of any value you must put down exactly what the pupil does in response to the various stimuli. If you write " the pupil is fixed " I do not know what the pupil does any more than I do if you say it is an Argyll Robertson pupil. This by the way. Sometimes in fixed pupils the defects are partial ; when this is so the reaction to light is usually more affected than the reaction on convergence, and the findings are easily mistaken for the Argyll Robertson sign.

The causes of partially fixed pupils are as numerous and as dissimilar as the causes of supranuclear, nuclear, and infranuclear lesions of the third nerve ; syphilis is merely one of them. I said that the reaction to light is more affected than the reaction on convergence ; almost always the fact that the reaction on convergence is also defective will tell you that you are dealing with an incompletely fixed and not an Argyll Robertson pupil. In a small but important group of cases, however, where the signs are due to a partial or recovering lesion of the third nerve, most often traumatic, the reaction to light is completely lost, while the reaction on convergence is normal (traumatic pseudo-Argyll Robertson pupil of Axenfeld). These false Argyll Robertson pupils are usually unilateral, they vary in size from time to time, and they usually dilate promptly and fully under the influence of mydriatics.

PUPILLOTONIA

The myotonic pupil is frequently mistaken for an Argyll Robertson pupil, because it does not react to light with the ordinary tests, but does react on convergence. It is distinguished by its peculiar behaviour during convergence. It is not very rare. I have seen ten cases during the last year, and have read descriptions of about fifty others. You should know about it, because it gives rise to an unfounded suspicion of syphilis, especially when, as sometimes happens, the abnormal pupils are associated with loss of the tendon-jerks. I have seen six patients with this combination of signs, and have found descriptions of eleven isolated cases in the journals ; in almost every instance a diagnosis of syphilis of the nervous system had been wrongly made. The myotonic pupil is most often unilateral ; it is then almost always larger than its fellow ; it may be oval, with the long axis horizontal or vertical ; it is often large, sometimes quite small, but never miotic. When ordinary bedside methods are used the reaction to light, direct and consensual, appears to be completely, or almost completely, abolished; after a sojourn in a dark room, however, the pupil dilates, and on subsequent exposure to diffuse daylight contracts very slowly again ; this contraction may proceed until the pupil becomes considerably smaller than it was before it dilated in the dark ; thereafter it dilates slowly again to its original size. Even after a long stay in a dark room the pupil may not contract to the usual short illumination with a pocket torch ; a long exposure may be necessary— in two of my cases I had to expose the eyes for several minutes to bright light at the window on a sunny day, before the size of the pupil changed appreciably.

During the act of accommodation for a near object contraction of the pupil, sometimes after a short delay, proceeds slowly through a range often greatly in excess of the normal ; the larger abnormal pupil may then become smaller than its normal fellow. In all the cases that I have examined, and in a large majority of those reported by others, the pupil remained small for some seconds up to several minutes after the act of convergence had ceased ; rarely it begins to dilate again at once ; in both cases dilatation proceeds at an even slower rate than did contraction, so that many minutes may elapse before the pupil regains its usual size. Sometimes accommodation is affected in the same way ; it is noticed most often during relaxation ; after a near object is looked at some seconds elapse before distant objects become clear. I will not burden you with more details about this interesting phenomenon, but I must emphasize the points on which it differs from the true Argyll Robertson sign. It is most often unilateral ; the pupils vary in size from time to time ; they are never miotic ; the reaction to light, though absent to ordinary tests, is not really abolished ; the pupil dilates in the dark ; the reaction on convergence is peculiar ; accommodation, though normal in range, is

sometimes slow ; and prompt and full dilatation occurs with mydriatics.

The cause of this phenomenon is unknown, and it is probably a manifestation of a disorder *sui generis*. Certainly the pupillary reactions differ essentially from those with which we are most familiar—namely, the Argyll Robertson phenomenon, fixed pupils, and ophthalmoplegia interna. The Argyll Robertson pupil is usually bilateral, the myotonic pupil unilateral ; the first is typically small, the latter large ; the one is always syphilitic in origin, the other never. Similar considerations distinguish it from fixed pupils and ophthalmoplegia interna. The iris of the myotonic pupil is not paralysed, and if the stimulus is intense enough or sufficiently prolonged the pupil contracts through a range wider than the normal ; accommodation, too, may be slow, but the range is normal. The terms " internal ophthalmoplegia " and " iridoplegia " that have been used in the titles of papers dealing with myotonic pupils are inapplicable and confusing.

Mr. Foster Moore, to whom I am greatly indebted, has seen about twenty patients with myotonic pupils, and has allowed me to examine some of them. He applied the term " non-luetic Argyll Robertson pupil " to them, but is not satisfied with it. According to the view I am expressing here it is a contradiction in terms ; moreover, there are many other non-luetic spurious Argyll Robertson pupils than the one he has described. As we must use linguistic shorthand, " myotonic " pupil is perhaps the best name, but you should think of the word myotonic as being in inverted commas, because the reaction is not the same as the myotonic reaction in Thomsen's disease. Its importance, whatever you choose to call it, is that it gives rise to an unfounded suspicion of syphilis.

I might mention other non-luetic conditions in which the pupils react on convergence but not to light, but I think I have said enough to convince you that pupils conforming to this definition occur in many dissimilar disorders, and that this definition is of little value ; certainly it does not define what Argyll Robertson described. I hope, too, that your own experience will prove to you that the true Argyll Robertson pupil, as I have defined it, is, as near as may be in an imperfect world, an infallible sign of syphilis of the nervous system.

COMPLETE ERADICATION OF THE THYROGLOSSAL TRACT

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(With Special Plate)

In an adult it is usually a simple matter to tell at a glance if a thyroglossal fistula is an old-standing one. Fistulas originating during infancy—they are rarely congenital—have their orifices situated low down in the neck ; furthermore, the crescentic appearance of an old fistula is characteristic (Fig. 1, Special Plate).

ANATOMICAL CONSIDERATIONS

The thyroglossal tract extends from the isthmus of the thyroid gland to the foramen caecum. The ramifications of a fistula connected with this tract can often be demonstrated by injecting lipiodol prior to a lateral radiogram, and the duct can sometimes be shown to pass right into the back of the tongue (Fig. 2, Special Plate). Unless the entire tract is removed, secreting epithelium is left behind, and after a short interval the fistula reappears (Fig. A). In the case of thyroglossal cysts the story of incomplete extirpation of the tract is much the same, for simple removal of the cyst is followed by a fistula which continues to discharge. In a review of thirty cases[1] of thyroglossal fistula I found that seventeen followed an operation for a thyroglossal cyst.

OPERATIVE PROCEDURES

Sistrunk's method of ensuring that the whole tract is removed is an excellent one, which should be looked upon as the standard operation for dealing with these otherwise troublesome cases. It is only by completely eradicating the whole tract in every case of thyroglossal

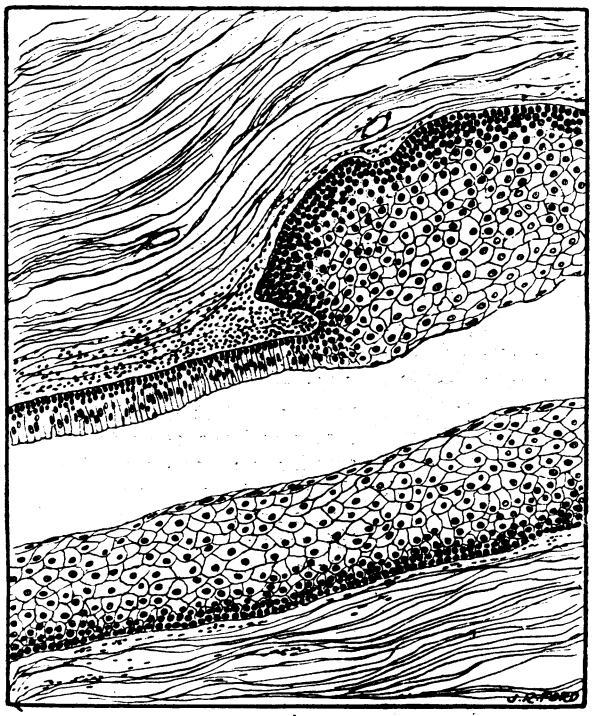

FIG. A.—Section of a thyroglossal fistula in the region of the hyoid bone, showing variations in the type of epithelium which lines the tract. (Section by Dr. P. O. Ellison.)

cyst as well as in every thyroglossal fistula that a perfect end-result may be confidently expected. The details of the operation are as follows. Through an elliptical transverse incision about the orifice, the tract is dissected up to the level of the hyoid bone. There is nothing to disturb the even tenor of the dissection up to this point. The body of the hyoid bone is then cleared on either side and divided on the left and the right so as to free a little more than a quarter of an inch of the centre of the bone, to which the tract is attached. Incidentally, even in a child, the hyoid is a tough little bone, and full-sized bone-cutting forceps are necessary to divide it cleanly. The wedge out of the hyoid is lifted up and the thyrohyoid membrane displayed. Dissection is now continued in the direction of the foramen caecum, but this time without attempting to isolate the duct (Fig. B). The tissues are cored through, allowing about a third of an inch on every side of the duct. The foramen caecum lies upwards and backwards at an angle of 45 degrees from the body of the hyoid bone (Fig. C), and it is along this line that we proceed until the cavity of the mouth is reached. This completes the dissection, and one lifts away *en bloc* the skin about the orifice of the tract ; the tract as far as the hyoid bone ; the centre of the body of the hyoid bone ; a disk of muscle from the mylohyoid muscle ; a disk of muscle from the geniohyoid muscle ; a tubular portion of the very heart of the geniohyoglossi muscles, and the foramen caecum.

Within this vermiform length of tissues, a seemingly sorry prize for so much labour, lies every secreting cell of the thyroglossal tract. Nothing short of this radical extirpation will guard against recurrence.