Anophthalmos and microphthalmos in a chick / by E. Treacher Collins and J. Herbert Parsons.

PLATE XIII.

Illustrates Messrs. E. Treacher Collins and J. Herbert Parsons' paper on Anophthalmos and Microphthalmos in a Chick.

FIG. 1 shows the microscopical appearances of a vertical section through the chick's head traversing both orbits, under a low power. A malformed microphthalmic eye is shown in the left orbit. Deep in the right orbit is a ring of hyaline cartilage with some irregularly pigmented tissue, like that of the choroid, in its interior.

FIG. 2 shows the left microphthalmic eye under a higher power. The adhesion of the lens to the posterior surface of the cornea is well depicted, also the extension of the upper part of the iris and ciliary body round the posterior surface of the lens, and the arrested development of the iris below.

FIG. 3 shows the ring of hyaline cartilage from the right orbit more highly magnified, and the character of the tissue contained within it.

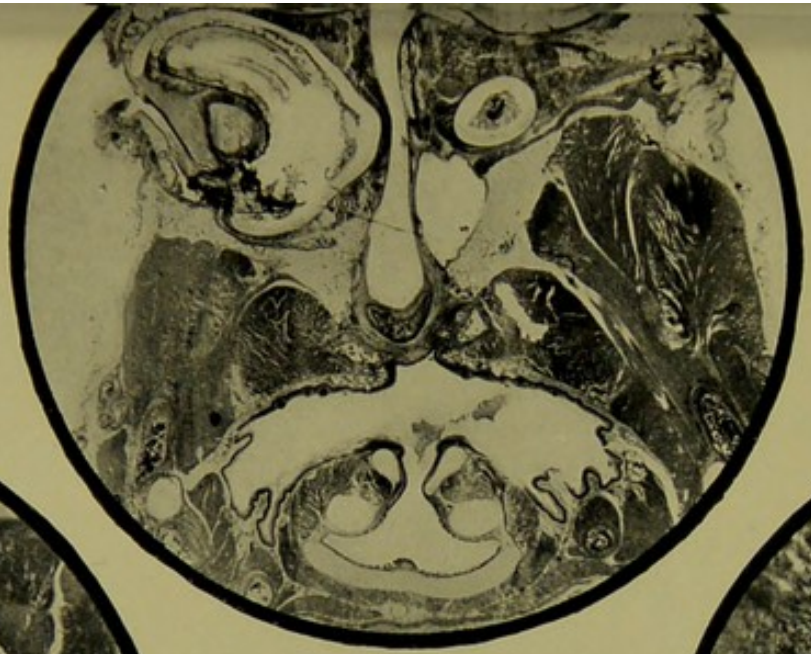


Fig. 1.

Fig. 2.

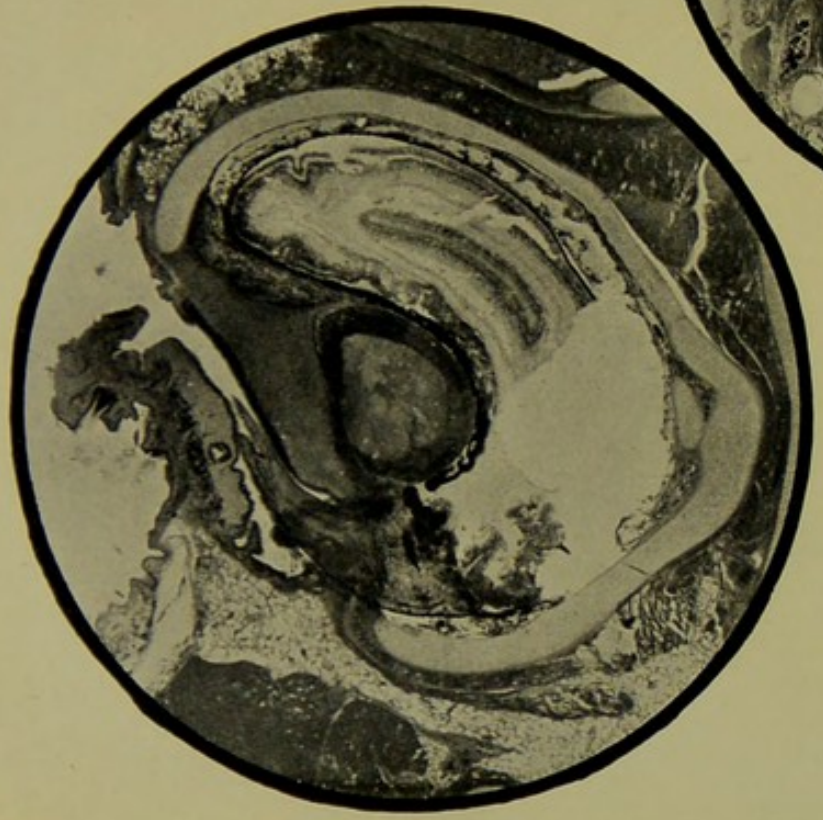
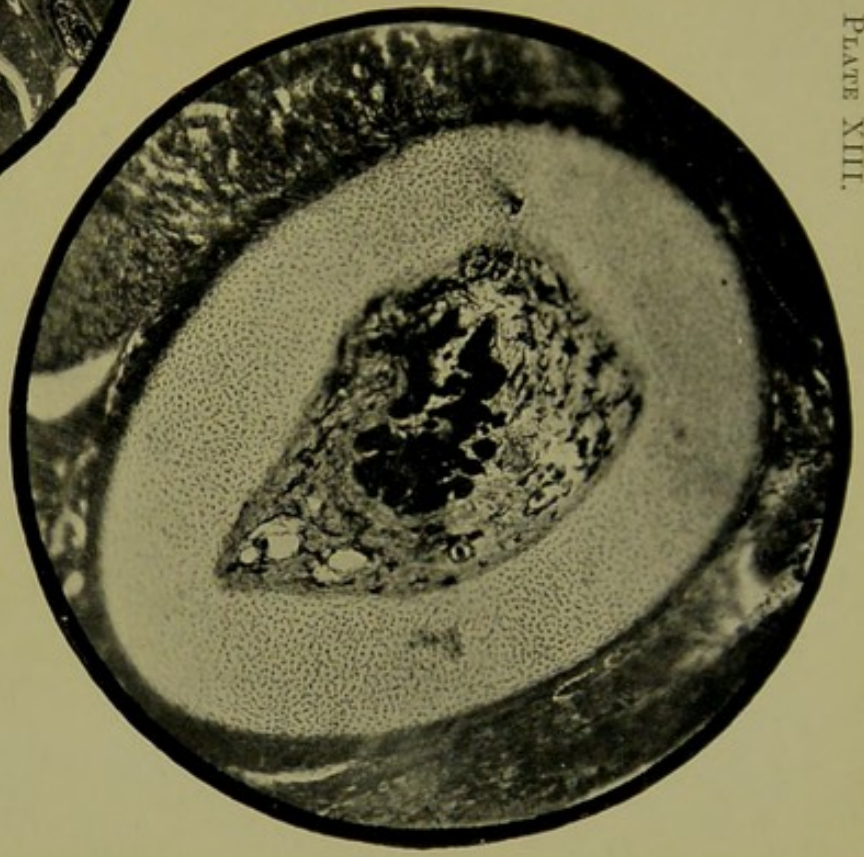


Fig. 3.



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Anophthalmos and microphthalmos in a chick.

By E. Treacher Collins and J. Herbert Parsons.

(With Plate XIII.)

The chicken which forms the subject of these notes was bred from an Indian game-cock and a Dorking hen. The same fowls have been bred from for the past two years. None of their previous progeny have presented any malformations.

The chick was hatched in an incubator; it was quite healthy, but born blind. It could not see to find its food, but could eat and drink quite well "when put to it." It was killed when four days old, and its head sent for examination.

After the removal of some of the feathers, the eyelids on each side were seen to be well formed. On separation of them, a small eyeball with a clear bright spot in it representing the cornea could be seen on the left side. On the right side no eyeball could be made out.

The head of the chick, after being hardened in formol, was decalcified and embedded in celloidin. A series of sections were then cut through it vertically so as to pass across both orbits. All the sections traversing the orbits were preserved, and a large number of them from different levels stained and mounted.

Microscopical examination of these sections show that on the right side no optic nerve, retina, or lens are present. Some hyaline cartilage, situated a little depth from the surface in the form of a complete ring, is seen in some of the sections (Plate XIII, figs. 1 and 3). The width of the cartilage composing this ring is about that of the hyaline cartilage in the sclerotic of the left eye, and like it has externally a small amount of fibrous tissue. Outside this fibrous tissue is striated muscle and adipose tissue.

In some sections the hyaline cartilage is not a complete ring ; it has a gap in one side through which fibrous tissue similar to that on the external surface passes. In others the cartilage is arranged in two semi-lunes, there being breaks in its continuity filled with fibrous tissue both on its deep and superficial surfaces.

The interior of the ring of cartilage is filled with tissue resembling that of the choroid somewhat condensed, fibrous tissue, blood-vessels, and nerves, like the ciliary nerves, being seen. There are also delicate, fine, branching, pigmented cells, and in the centre some much denser pigmented tissue in the form of irregular dots and twisted branching lines. A careful comparison of this latter pigmented tissue with that contained in the choroid of the left eye shows that it is of the same character though somewhat more condensed, and that it is not pigmented epithelium of retinal origin, which it might possibly at first be taken for. There is no tissue in the interior of the ring or anywhere in its vicinity resembling retina or optic nerve.

The eyelids and conjunctival sac on this side appear well developed. In those sections where the ring of cartilage above described approaches nearest the surface, it is separated from the epithelium of the conjunctiva only by a small amount of dense fibrous tissue and some loose adipose tissue. Nowhere can any down-growth of epithelium or anything representing the lens be distinguished. On the left side the eyelids and conjunctival sac appear normal. The cornea is small and the anterior part of the eyeball much flattened. The sclerotic is seen to be formed in the normal way of a cup of hyaline cartilage, becoming thicker at the posterior pole and having a little fibrous tissue external to it. There is a wide gap in the posterior part of the cup through which the optic nerve passes. On one side of the nerve there is a considerable band of fibrous tissue between it and the cartilage. Anteriorly the cup of cartilage is seen to end in rounded extremities a short distance from the corneal margin.

Normal laminated epithelium covers the anterior surface of the cornea; there is no anterior limiting membrane (Bowman's layer). The substantia propria of the cornea appears imperfectly developed. It is largely composed of spindle-shaped cells; the nuclei of the cells in it are much closer together than in normal corneæ. There is no posterior limiting membrane (Descemet's membrane).

The lens is in immediate apposition with the posterior surface of the cornea, its hyaline capsule being attached to the substantia propria for a portion of its extent. Above, nothing intervenes between these structures; below, some vascular tissue, of the nature of the stroma of the iris, separates them for a short distance (fig. 2).

The choroid appears normal and ends anteriorly in the ciliary body, which is much distorted. The striated fibres of Crampton's muscle can be seen on each side; its anterior part is pressed into close contact with the sides of the lens.

At the lower part there is hardly any iris. The pigment epithelium of the ciliary body and unpigmented layer of cells forming the pars ciliaris retinæ are much plicated, and terminate in contact with the lower border of the lens. As above mentioned, a small amount of tissue like the stroma of the iris has insinuated itself for a short distance between the cornea and lens.

At the upper part of the eye the anterior part of the ciliary body and what represents the iris turn backwards behind the lens, lying in close contact with its posterior surface for more than two-thirds of its extent. Behind the upper part of the lens there is, from before backwards, tissue like the stroma of the iris, pigment epithelium, and imperfectly formed retina, *i. e.*, retina in which its several layers have not become differentiated. Further down on the posterior surface of the lens the stroma of the iris ceases, and the pigment epithelium, much rucked, lies in direct contact with the lens capsule. The tissue-like undifferentiated retina passes into a single layer of unpigmented cells which continue up to the pupillary margin.

The iris from above extends so far down on the back

of the lens that its pupillary margin is quite at the lower border.

The lens is very imperfectly formed. A hyaline capsule completely surrounds it. At the anterior pole a single layer of flattened cells lines the capsule. A short distance away from the anterior pole the lining cells gradually begin to lengthen out. They are longest and thinnest of all about the equator. They diminish slightly in length on the posterior capsule, but even there are of considerable size. The surface formed by these lining cells towards the interior of the lens gives rise to a very well-defined line. The space left within this line is filled with what looks like coagulated albuminous material with scattered, large, squamous-like, epithelial cells in it.

The retina at the posterior part of the eye is fully developed, presenting its normal arrangement of layers.

It will be gathered from the above detailed description of this specimen that on the right side there was absolute non-development of all portions of the eye formed from neural epiblast, *viz.*, optic nerve, retina, and pigment-epithelial layer. Further, that though all the neural epiblastic elements were absent, some imperfect formation of the subsidiary structures developed from mesoblast (choroid and sclerotic) had taken place.

These structures of mesoblastic origin formed a nodule to which the extra-ocular muscles were attached, and which, therefore, might have presented the appearance of a moving stump at the back of the orbit.

The question of where to draw the line between anophthalmos and microphthalmos has been much discussed. In cases where at first it appears that the eye is congenitally absent, by more careful examination a small hard nodule is frequently discovered. Sometimes it is so small that it can only be discovered when the child is kept quiet by an anæsthetic and the finger passed between the lids to the back of the orbit. The existence of such nodules have led some to suppose that in these cases the eye has become shrunken by intra-

ocular inflammation occurring *in utero*. Others have considered that where a nodule is present the case is not really one of anophthalmos, but only a very high degree of microphthalmos.

In a paper on anophthalmos in the *Royal London Ophthalmic Hospital Reports*, vol. xi, p. 429, 1887, one of us (E. T. C.) gave a fairly complete review of cases recorded under that name up to that date.

The notes of nine cases were tabulated in which a *post-mortem* examination had been made. Not in any of these cases was an optic nerve found to enter the orbit; in one it ended in a cone at the optic foramen, in another in a fibrous filament, and in five the chiasma was absent. In one case one olfactory lobe was absent, and in another both olfactory lobes and one of the cerebral hemispheres. This is worthy of note because these structures, like the primary optic vesicles, are expansions of the anterior primary encephalic vesicle.

At the end of this paper a further epitome of the literature of the subject of anophthalmos up to the present time will be found.

In the cases of Van Duyse, Bietti, and Spiller, there referred to, it will be seen that the optic nerves, chiasma, and optic tracts were all absent.

So far as we have been able to ascertain, there seems no definite case recorded in which a microscopical examination of the orbit was made and where the mesoblastic structures of the eye were found to be entirely absent; whilst there are several (Michel, Sgrosso, Van Duyse, and probably Bietti) in which, as in the chick the subject of this paper, there was a body in the orbit composed only of structures of mesoblastic origin. From the appearance of the sections through the orbit in this chick, and the complete absence of all round-celled inflammatory exudation, it is evident that the condition of the right eye is not one of phthisis bulbi from inflammation before birth.

The essential element of an eye is a nervous mechanism which serves to receive visual sensations for transmission

to the brain. All the other tissues connected with and surrounding such a mechanism are merely subsidiary. When, therefore, there is a complete failure in the development of this essential nervous mechanism, it would seem fair to speak of the condition as one of anophthalmos, even though there may have been some formation, from mesoblastic tissue, of subsidiary structures sufficient to produce a moving nodule at the back of the orbit.

When, however, the essential nervous mechanism has been formed, no matter how imperfectly, and the eye is below the normal dimensions, the case would be one of microphthalmos.

In this chick which we have examined, we should then describe the condition on the right side as one of true anophthalmos, notwithstanding the presence of the small nodule of tissue like sclerotic and choroid. This distinction, which we think is the only true one which can be drawn between these two conditions, is, unfortunately, one which can only be determined with certainty by a microscopical examination.

On the left side in this chick a microphthalmic eye was present, and, as will be gathered from the description of it which has been given, there has been a failure in the development of the anterior chamber. The lens was in contact and adherent, for a portion of its extent, to the back of the cornea. There was no round-celled infiltration or other appearances which would lead us to suppose that this adhesion was inflammatory in origin. It appears to have been simply an instance of arrested development.

It is of considerable interest in showing the altered arrangement in the iris to which it has given rise. It was suggested by Manz, as a probable explanation of the failure in the development of the iris in cases of aniridia, that that structure was prevented from growing inwards by an unusually strong adhesion of cornea and lens before the formation of the anterior chamber.

In this specimen an abnormal adhesion of cornea and lens has persisted until after birth. We find in it that the upper part of the iris, prevented from growing inwards in front of the lens, has turned backwards behind it, the iris having thus come to line the posterior surface of the lens for a considerable portion of its extent. Below, the growth of the iris forwards has become arrested, it being represented only by a little of the stroma between the lens and cornea where they are not quite in contact, the pigment epithelium of the ciliary body terminating at the lower border of the lens.

Summary of Literature on Anophthalmos.

Haab (1) carefully investigated a case of anophthalmos in a man, æt. 27 years. The chiasma was absent, the optic tracts were very small, and the corpus geniculatum externum was absent. The corpora quadrigemina were normal. All the extrinsic eye muscles, with their nerves, were normal in size and shape. The L. eye measured 3·5 mm. by 2·5 mm. The very small optic nerve entered behind. Cornea, iris, ciliary body, and lens were absent. The sclera contained choroidal elements, a few rods and cones and pigment-epithelial cells (no nerve-fibres), and vitreous. There was no lamina vitrea, but there were colloid bodies surrounded by pigmented epithelium. The R. eye was globular, 4 mm.—4·5 mm. in diameter. It contained more retinal elements, especially rods and cones and outer nuclear layer; there were no ganglion cells or nerve-fibres, and no colloid bodies. There were no nerve-fibres in the optic nerve.

Nieden (2) describes a case of anophthalmos cyclopica; but there is no microscopic examination.

Strösse (3) reports a case of unilateral anophthalmos in a calf. Careful examination (? microscopic) of the orbital tissues failed to disclose any eye or optic nerve.

Hilbert (4) reports bilateral anophthalmos in a girl, æt. 9 years. There was no anatomical examination. He (5) also

reports a bilateral anophthalmos in which there was catarrhal conjunctivitis at birth. This is of importance in consideration of the possibility of anophthalmos being due to the intra-uterine phthisis bulbi. On the R. side there was no sign of an eye ; on the L. a grey, pigmented globe, 2 mm. in diameter.

Huth (6) records bilateral anophthalmos, without anatomical examination.

Fromaget (7) records a case of R. anophthalmos with congenital cyst in the lower lid, and L. microphthalmos. The cyst (only) was carefully examined.

Harlan (8) reports a similar case of anophthalmos with cyst. No microscopic examination could be made of the orbital contents. No eye or optic nerve could be made out.

Stuffler (9) reports bilateral anophthalmos in an otherwise normal new-born child. There was no anatomical examination.

Sgrosso (10) examined a pig in which one eye was apparently absent, but microscopic examination revealed sclera, ciliary body, and traces of choroid covered by a dermoid with cartilage, sweat and sebaceous glands, bone, etc. There was a coloboma of the optic nerve in the other eye.

E. von Hippel (11) examined bacteriologically a case of R. microphthalmos and L. anophthalmos in which pus was present in the conjunctival sac at birth. Pneumococci, which were not virulent as regards rabbits, were present. The patient had a brother with bilateral anophthalmos, another with hydrocephalus, and a sister with microphthalmos, and congenital opacities of the cornea. Von Hippel reviews the whole subject in this paper, concluding in favour of intra-uterine inflammation as the usual cause. Hoppe (12) arrived at the same conclusion.

Van Duyse (13) gives a very careful account of a bilateral anophthalmos. The optic nerves, chiasma, tracts, and external geniculate body were absent. The extrinsic eye muscles were present. The eyes were extremely

small, and consisted of a fibrous envelope, containing branched pigmented cells and blood-vessels. There were no epiblastic tissues present (corneal epithelium, lens, retina, optic nerve).

Fromaget (14) reports another case of anophthalmos with congenital cyst in 1900. There was no anatomical examination.

Bietti (15) gives an excellent description of a bilateral anophthalmos, and a critical discussion of the causes. In a boy, æt. 15 days, the palpebral fissure each side measured 0·5 cm.; there was intense conjunctival catarrh. The child died when 15 months old. *Post mortem*, the convolutions of the brain were good, including the occipital lobe; corpora mammillaria present; no infundibulum or hypophysis; chiasma, optic nerves, tracts, and external geniculate bodies absent; pulvinar and ant. corp. quadrigemina smaller than normal; internal geniculate body normal. All the cranial nerves were present except the second. The fourth and sixth were small. The optic foramen was occupied by a filament, proved microscopically to be an artery. The extrinsic muscles were present in the orbits. The eyes were microscopic. The R. eye was a nodule 3 mm. ant.-post. by 1·5 mm. vert. Another nodule posteriorly was the ciliary ganglion. The bulb consisted of a fibrous capsule with a pigmented centre; in the latter a more deeply pigmented crescent, with the concavity looking forwards, probably represented retinal epithelium consisting of angular pigmented cells. The other cells were uveal—oval or spindle-shaped. The L. eye measured 1 mm. by 600 μ, and consisted of sclera containing remnants of uvea and retinal pigment.

Brose (16) reports a R. anophthalmos and L. microphthalmos without anatomical examination.

Zimmermann (17) records a case of unilateral anophthalmos. *Post-mortem*: no L. olfactory nerve or bulb; L. optic nerve one fifth size of normal R.; L. third nerve half normal size; other nerves normal. The eye consisted of a small mass of fibrous tissue, the size of a pea, contain-

ing a small amount of black pigment. There was no microscopic examination.

Spiller (18) found complete absence of the visual system in an adult idiot, æt. 22 years. The palpebral fissures were extremely small, and the orbits contained only fibrous tissue (macroscopic). No eyeballs could be seen. There were no optic foramina, nerves, chiasma, or tracts; no external geniculate bodies. The pulvinars and ant. corp. quad. were apparently normal. The occipital lobes were small, the cuneus ill developed, and the calcarine fissure short. The ocular nerves were well developed, except both sixths.

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