Congenital defects of the iris and glaucoma / by E. Treacher Collins.

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By E. Treacher Collins.

(With Plate V.)

The opportunity for the microscopical examination of eyes with congenital absence of the iris but seldom occurs. I am much indebted to Mr. Nettleship for kindly giving me the specimens from the following case.

Case 1. *Congenital absence of both irides, and anterior polar opacities of both lenses.*—David B—, æt. 7 months, was brought to Mr. Nettleship at St. Thomas's Hospital in October, 1889, by his mother, who was herself a patient for her eyes, having congenital absence of both irides and leucoma of the right cornea following ulceration, together with a staphylomatous condition of the sclerotic. Her right eye was ultimately excised; a full description of the pathological condition of it is given in the 'Ophthalmic Review,' vol. x, p. 103.

The following is the note which was made as to the condition of the child's eyes:—"Child evidently sees light, but does not take notice of things. Absence of irides. There are concentric rings of opacity in each lens; they resemble lamellar cataracts. The edge of the lens is clearly seen attached by the suspensory ligament. Examination by focal light unsatisfactory."

The child subsequently died on April 3rd, 1892, when three years old, at St. Thomas's Hospital, as the result of a burn. Both eyes were removed post mortem and hardened in Müller's fluid.

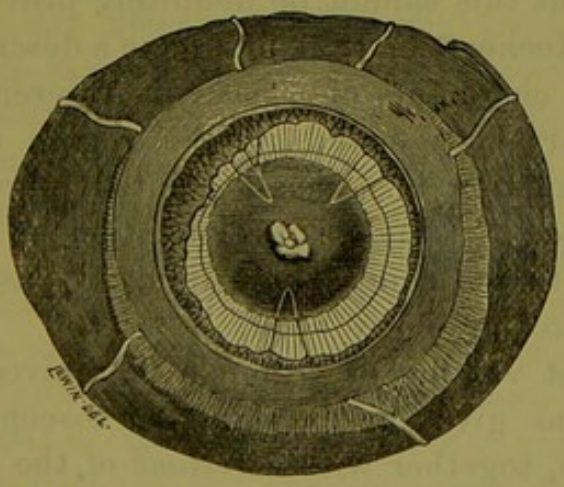
Pathological examination of the left eye.—Antero-posteriorly it measured 19 mm. vertically, and laterally 20 mm. The cornea measured 10·5 mm. vertically, 11·5 mm. laterally. The eyeball was frozen, and opened by an antero-posterior vertical section. The ciliary body is seen to end in a rudimentary iris on both sides at the seat of section; above it measures 1 mm. in length, but is much smaller below. The lens is small, it measures 4 mm. antero-posteriorly and 7 mm. laterally; it is in position; there is a raised opacity at its anterior pole. The vitreous has a good consistency. The retina is *in situ* but slightly creased.

Microscopical appearances.—The corneal tissue shows nothing abnormal; there is no break in the continuity of either Bowman's or Descemet's membrane; the latter ends in the ligamentum pectinatum in the usual way. The whole tissue of the iris is more condensed than usual. Below, the tip of it does not extend beyond the termination of Descemet's membrane in the ligamentum pectinatum. No indication of the sphincter muscle in it can be made out. The uveal pigment at its tip ends in a double fold. One section shows a tag of membrane prolonged from its anterior surface and floating free, a remnant of the pupillary membrane (Plate V, fig. 1). In some of the sections a delicate adhesion passes between the extreme root of the iris and the ligamentum pectinatum. This by no means blocks the filtration area; it seems as though the separation of the anterior surface of the iris from the cornea at its extreme root had been incomplete. Above the iris is longer, and its tip extends just beyond a line drawn backwards on a level with the termination of Bowman's membrane. A small rudiment of a sphincter muscle is here discernible. The uveal pigment extends beyond the stroma of the iris, ending in a double fold (Plate V, fig. 2). There is a more pronounced band of adhesion between the root of the iris on this side, than above; it goes to the inner portion of the ligamentum pectinatum, not up into the extreme periphery of the

anterior chamber. The ciliary muscle is well developed, but the ciliary processes are small: they have their usual direction inwards. The anterior perforating arteries and the large circular arteries of the iris are present.

At the anterior pole of the lens there has been some subcapsular proliferation of the lining epithelial cells. The capsule is raised and rucked over them, and there is slight excavation of the lens substance beneath them. Amongst the proliferated cells is substance of a hyaline structure and also some calcareous deposit. Besides the changes at the anterior pole of the lens, there are scattered irregularly throughout it, small patches of a granular character lying in broken spaces amongst its fibres. There are also numerous small round bodies which stain deeply with logwood, and look like large micrococci. I have met with similar bodies once before in a lens with a lamellar cataract. I think they must be shrunken nuclei of the lens fibres which have failed to completely disappear. They are about half the size of the nuclei in the epithelial cells lining the capsule.

Pathological examination of the right eye.—It measured



Anterior half of right eye from which the cornea and sclerotic have been removed, exposing an elementary iris, tags of pupillary membrane, lens with an anterior polar opacity, and fibres of the suspensary ligament.

20 mm. in all three diameters; the cornea measured 10·5 mm. vertically and 11 mm. laterally. It was opened by

an equatorial section, after which the sclerotic and cornea were peeled off the front half. In doing this it was noticed that the cornea was abnormally adherent at its periphery.

A narrow rim of iris is exposed in its entire circumference ; this is broadest on the outer side, where it measures 2 mm. in length. Above it measures ·75 mm., and it is slightly broader there than below. On the inner side it is narrowest. Throughout its whole circumference the uveal pigment shows at what corresponds to its pupillary border. The ciliary muscle appears well developed. Some delicate tags, remnants of the pupillary membrane, can be seen passing from the anterior surface of the iris to the lens capsule. The lens measures 7 mm. transversely. At its anterior pole is a raised white opacity. The fibres of the suspensory ligament are very distinctly seen, passing between the ciliary body and the margin of the lens. A little distance from the lens, running concentric with its margin and crossed transversely by the fibres of the suspensory ligament, is a very well-defined line ; it may be the limit of the capsule of the lens away from which the lens substance has shrunken in process of hardening, but this cannot be definitely proved.

H. Pagenstecher* in 1871 first gave a description of the microscopical characters of a case of irideremia. In his case the ciliary body ended in the region of the ligamentum pectinatum in a small pointed, pigmented, and vascular process ; and a prolongation of Descemet's membrane with the endothelium lining it passed beneath this process to the ciliary body.

In the last volume of Graefe's 'Archives,' 1891, G. Rindfleisch has given an excellent microscopical description of a case, together with a *résumé* of the literature of the subject. His case was that of a woman aged fifty-one, who presented the typical symptoms of irideremia in both eyes. She had nystagmus, some amblyopia, some opacities of the corneæ and lenses, the latter being

* 'Klin. f. Augenheilk.,' Bd. ix, p. 425.

displaced upwards. Microscopically there was found to be considerable encroachment of the episcleral tissue on the cornea, some hyaline excrescences on Bowman's membrane, and a triradiate deposit of pigment on Descemet's membrane. At the lower sclero-corneal margin was the scar of a perforation. A rudimentary iris was present, but its sphincter muscle was completely absent. It was closely approximated to the cornea, the angle of the chamber appearing pointed. The ciliary body was badly developed, and its processes were directed backwards.

Any theory, which in the case I have described might be suggested to account for the arrested development of the iris, should also explain the cause of the anterior polar opacity of the lens, of the abnormal adhesion at the root of the iris to the ligamentum pectinatum, and the folding of the uveal pigment at the tip of the iris. That which has been put forward by Manz may, I think, be made to explain all these abnormalities. To describe it, it is necessary to briefly refer to the mode of development of the iris. Before the iris is formed, there exists between the posterior surface of the cornea and the anterior capsule of the lens the anterior portion of the fibro-vascular sheath. This receives its blood-supply partly from the ciliary arteries aud partly from those in the posterior fibro-vascular sheath, which is prolonged forwards around the sides of the lens to join it. The cornea, anterior fibro-vascular sheath, and lens, lie in close contact with one another.

The iris in growing forwards has to insinuate itself between the cornea and anterior fibro-vascular sheath on the one side and the lens on the other (Plate V, fig. 6), pushing the prolongation from the posterior fibro-vascular sheath in front of it. The anterior fibro-vascular sheath subsequently becomes the pupillary membrane.

If we suppose some abnormal adhesion to occur between these structures, viz. the cornea, anterior fibro-vascular sheath, and lens capsule, or some delay in their separation, then we can understand how a mechanical obstruction

would be introduced to the growth of the iris forwards. The opacity at the anterior pole of the lens is evidence of abnormal contact between the lens and cornea, for exactly similar opacities are produced when they have been brought into contact as the result of ulceration of the latter.

The abnormal tags of adhesion between the root of the iris and ligamentum pectinatum, is part of the imperfect separation of the anterior fibro-vascular sheath from the cornea. The puckering of the uveal pigment at the tip of the iris is due to the arrested ingrowth of the termination of the secondary optic vesicle, which consequently becomes indented.

Rindfleisch agrees with Manz that the arrest in the development of the iris is due to the contact of the lens and cornea. He would, however, explain this in his case by intra-uterine inflammation, beginning in the choroid, extending forwards, and causing perforation near the sclero-corneal margin and escape of aqueous.

In my specimens no sign of a perforation is to be seen either in this region or in the cornea. The sclero-corneal margin is a very unusual position for a perforation to occur in, and it seems unlikely that a bilateral affection, as irideremia usually is, would be caused in this way. It is, moreover, unnecessary to have a perforation for the lens to come in contact with the cornea, for at the time the iris is developing these structures are in apposition, the anterior fibro-vascular sheath alone intervening: there is then no anterior chamber and no aqueous.

Regarding cases of irideremia from a clinical standpoint alone, we should be inclined to think that it was impossible for them to become the subjects of glaucoma; for we fail to see how, when the iris is absent, the filtration area can become blocked. Microscopical examination of these cases, however, seems to show that they are really predisposed to glaucoma, for not only do we find that the ciliary body terminates in a rudimentary iris, which if pushed forwards is quite sufficient to block the whole of the posterior surface of the ligamentum pectinatum, but

that between this rudimentary iris and the ligamentum pectinatum there are abnormal adhesions. This was so in the eyes examined by Pagenstecher and Rindfleisch, and in those I here record.

It is of considerable interest that the mother of Case 1, who also had irideremia, had increased tension secondary to ulceration of the cornea and intra-ocular hæmorrhage. As I have already stated, I have described in full the minute anatomy of her eye in the 'Ophthalmic Review.' In it the rudimentary iris was pushed forwards, and was intimately adherent to the cornea in the region of the ligamentum pectinatum.

The next case I have to record also bears on this point.

Case 2.—*Congenital coloboma of iris and lens outwards; absolute glaucoma.*—Charles W—, æt. 36, was shown by Mr. Lang at the Ophthalmological Society on October 17th, 1889,* as a case of chronic glaucoma, with congenital coloboma of the iris and lens outwards.

He was admitted to the Moorfields Hospital again on April 27th, 1892. He stated that during the last ten days his right eye had been very painful, so much so that he had been unable to get any sleep. During the last eighteen months his left eye had also become quite blind. There was much ciliary congestion of the right. The cornea was steamy. No details of the fundus could be seen in it, and the tension was increased. It was excised the following day.

Pathological examination.—The eyeball measures antero-posteriorly 22·75 mm., laterally 22·5 mm., and vertically 22 mm. The cornea measures 9·5 mm. laterally and 8·5 mm. vertically.

Opened after hardening in Müller's fluid, and freezing by an antero-posterior horizontal section.

The iris on the outer side is seen to be absent, but the ciliary muscle and processes are present. On the inner side the root of the iris is in contact with the posterior

* 'Trans. Ophth. Soc.,' vol. x, p. 106.

surface of the cornea, blocking the filtration area. The lens lies nearer the ciliary processes on the inner side than on the outer. A notch in the outer border of the lens is distinctly seen. The vitreous is of good consistency. The retina is in position, but slightly rucked just behind the the ora serrata. The optic disc is deeply cupped.

Microscopical appearances.—The stroma of the iris on the inner side is much atrophied; there is marked ectropion of the uveal pigment at its pupillary border. The sphincter muscle, which appears unusually broad, also turns up at its tip.

The ciliary muscle and processes on the inner side are much atrophied: the latter are directed straight inwards towards the side of the lens.

On the side of the coloboma of the iris the point where Descemet's membrane commences to break up into the fibres of the ligamentum pectinatum is clearly defined. The canal of Schlemm is not so easily made out, as its two walls are closely pressed together. The ciliary body here ends in a rounded knob, which is in contact with the back of the cornea. The termination of this knob, however, does not reach as far as the end of Descemet's membrane, so that the whole of the posterior surface of the ligamentum pectinatum is not blocked by it (Plate V, fig. 4). The uveal pigment on the back of this termination of the ciliary body is composed of two layers united at the tip, the posterior being much thicker than the anterior. Just where these end on the posterior surface of the ligamentum pectinatum is a little accumulation of fibres and cells; from it the endothelial lining of Descemet's membrane passes inwards, and a layer of cells outwards beneath the pigment on the termination of the ciliary body.

The ciliary muscle on this side of the eye is very thin, thinner than on the other. The ciliary processes are small, and in the centre of the coloboma the anterior ones slant at a very acute angle backwards (Plate V, fig. 3). A little to one side of the coloboma they have their usual direction.

The lens is broader on the outer than on the inner side; in some sections it has also a slight depression inwards there at its equator, and the cells which line the capsule terminate at a point considerably anterior to what they usually do. The nucleated zone on the outer side is as much in front of the equator of the lens as it is usually behind it.

The nerve-fibre layer of the retina is extremely atrophied, and its blood-vessels dilated. The lamina cribrosa is depressed backwards and the nerve-fibres atrophied down to it, the blood-vessels standing out prominently on its surface.

This case presents several points for consideration. First, the unusual position of the coloboma in the iris, viz. outwards. It is frequently—and, I think, wrongly—assumed that a congenital deficiency of the iris must necessarily bear some relation to the ocular cleft. As I have just shown, it seems likely that the whole iris may be arrested in its development by some abnormal adhesion or late separation of the cornea and lens. If this abnormal adhesion or late separation existed in one particular part only of the surface of the lens, then it would be possible for the iris to develop in the normal way everywhere except opposite the adhesion, and a localised congenital deficiency would be produced. Such a deficiency might occur in any direction, or possibly in two directions, which would account for cases of two colobomata in one eye. The layer of unpigmented cells on the posterior surface of the rudimentary iris, continuous on one side with the endothelium of Descemet's membrane, and on the other reaching as far as the ciliary body, is somewhat similar to what Pagenstecher has described in his case of irideremia, as a prolongation from Descemet's membrane behind the iris. I would suggest that possibly this layer is a remnant of the prolongation forwards of the posterior fibro-vascular sheath.

It is interesting to find that in the sections which pass across the notch in the lens, the ciliary processes are directed backwards. If the notch is, as I believe, due to

some defect in development of the suspensory ligament at that point,* then we should expect them to assume this direction, not being held forwards by it.

In Rindfleisch's case of irideremia, where there was congenital ectopia of the lens, a condition also due to a defect in the suspensory ligament, the ciliary process pointed backwards.

That primary glaucoma should come on in an eye with a congenital coloboma of the iris, seems at first to offer an unsurmountable objection to the theory, which attributes the increased tension to the blocking of the filtration area of the cornea by the root of the iris. We fail to see why an iridectomy should relieve glaucoma if, when there is already a coloboma of the iris, the tension can become increased.

In this case in the situation of the coloboma, the ciliary body, as in the case of irideremia, ended in a rudimentary iris, or rather in a process which lay intimately adherent to the outer half of the ligamentum pectinatum. From the arrangement of this process, with a layer of unpigmented cells behind it, I should think that it had never become separated from the cornea, rather than that it had recently become pushed forwards into contact with it; so that there had always been obstruction to the exit of fluids in this situation, and that the onset of the glaucoma was determined by the root of the iris in the rest of its circumference becoming pushed forwards.

In my article in the 'Ophthalmic Review,' before referred to, I described a case of traumatic aniridia with increased tension. In that eye the lens as well as the whole iris had partially escaped through a corneal wound. The lens capsule remained adherent to the corneal cicatrix. The advanced position it had thus taken up resulted in the dragging forwards of the ciliary processes, and the blocking of the filtration area by the most anterior of them. This next case is a very similar one.

* See article "On the Development and Abnormalities of the Zonule of Zuin," 'Ophth. Hosp. Reports,' vol. xiii, p. 81.

Case 3. *Traumatic aniridia; adhesion of lens capsule to cornea; glaucoma.*—William L—, æt. 57, was admitted to the Moorfields Hospital under the care of Mr. Gunn, on August 23rd, 1892. Eight and a half months previously his right eye had been injured by a piece of a punching press, which, breaking off, flew up and struck it. The blow was a severe one and fractured the lower orbital margin; the sight of the eye was immediately lost. At the time of the accident he was wearing spectacles, and he thinks it possible that a piece of the glass may have entered the eye. It has been painful at times ever since the injury, and much worse the last two months.

On examination an irregular triradiate vascular scar was seen in the lower part of the cornea. The lens, which appeared dislocated into the anterior chamber, was grey and opaque. He could only distinguish light from dark and his projection was defective, especially on the inner side. The tension was + 1.

There were no signs of sympathetic mischief in the other eye. The day following admission his right eye was excised.

Pathological examination.—There is a broad adhesion of the lens, which is opaque and partly absorbed to the cicatrix in the lower part of the cornea. No iris can be seen; it appears to have been completely torn away from its attachment at the ciliary body, and to have escaped from the eye. The vitreous is shrunken and completely detached posteriorly. The retina is in position; the optic disc deeply cupped and excavated.

Microscopical appearances.—Sections through the scar in the cornea showed thickening of the anterior epithelium overlying it, a break in the continuity of Bowman's and Descemet's membranes, and a mass of irregular fibrous tissue with new vessels and groups of round cells in it. This fibrous tissue projects through the gap in Descemet's membrane posteriorly; the anterior capsule of the lens which has been perforated comes forwards from both sides and is incorporated in it. The lens fibres are much broken; they protrude through the break in the

capsule, and are in contact with the cornea. The iris above has been torn away almost at its extreme root; a small piece of tissue at the anterior extremity of the ciliary body with uveal pigment on it, which has been left, is drawn forwards by the adhesion of the lens capsule to the cornea into contact with the ligamentum pectinatum at the angle of the anterior chamber. It blocks about the outer half of the filtration area. Below, the whole of the filtration area is closed by the anterior part of the ciliary body, which is similarly drawn forwards by the lens capsule and suspensory ligament (Plate V, fig. 5).

(*October 20th*, 1892.)

DESCRIPTION OF PLATE V,

Illustrating Mr. E. Treacher Collins's paper on Congenital Defects of the Iris and Glaucoma.

FIG. 1.—Section of the upper part of the rudimentary iris of the left eye in Case 1, showing a tag of adhesion at the periphery of the iris to the back of the cornea and the uveal pigment extending beyond its free extremity.

FIG. 2.—Section of the lower part of the iris of the same eye, less well developed than above, and with a persistent tag of pupillary membrane at its free extremity. The uveal pigment on its posterior surface ends in a double fold, and a tag of adhesion passes between its root and the back of the cornea.

FIG. 3.—Section from the region of the coloboma of the iris in Case 2, showing the ciliary processes sloping backwards.

FIG. 4.—The same section as Fig. 3, more highly magnified. It shows the mode of termination of the ciliary body in the region of the coloboma, and its relation to the ligamentum pectinatum.

FIG. 5.—Section of one angle of the anterior chamber in Case 3, showing the iris absent and the ciliary processes drawn forwards, the most anterior of them blocking the posterior surface of the ligamentum pectinatum.

FIG. 6.—Section of the front half of a fœtal eye at the fourth month, showing the lens in contact with the cornea, and the iris insinuating itself between them.

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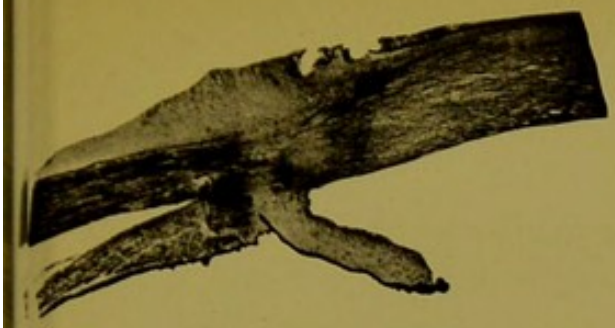

Fig. 1.

Fig. 2.

Fig. 3.

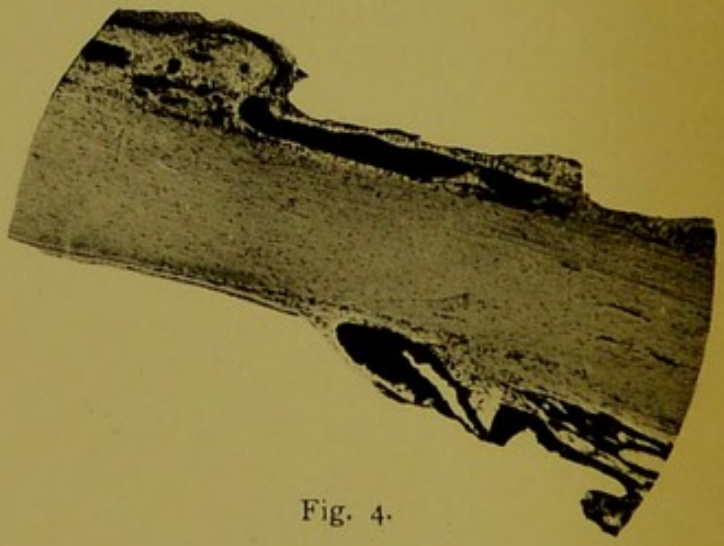

Fig. 4.

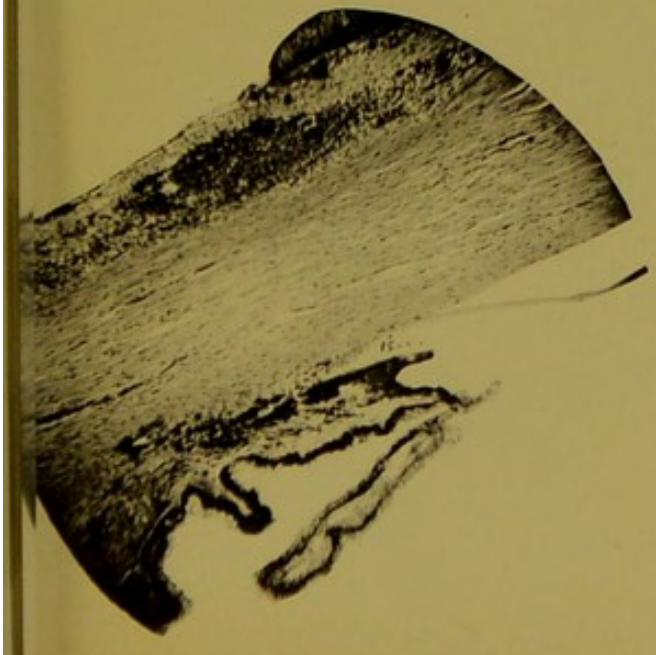

Fig. 5.

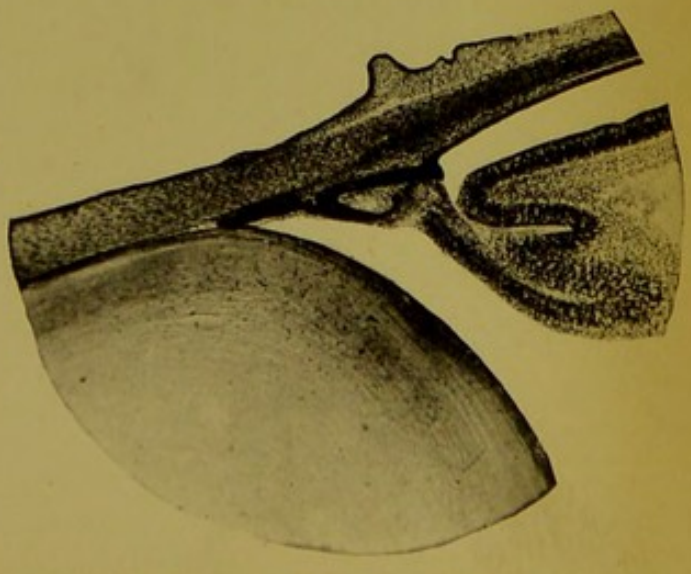

Fig. 6.

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