Coloured Areas in the Sclerotic

COLOURED AREAS IN THE SCLEROTIC

BY

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ABERDEEN

The occurrence of coloured areas in the sclerotic in some cases of melanosis of the eye is well known. The colour of these areas has been variously described as violet, purple, slaty-grey or brown. Bourquin in his comprehensive paper, in 1917, says that the scanty and contradictory accounts of the course of melanosis do not allow us to decide whether the sclerotic spots are progressive or stationary, and that most authors have seen their patients only once, and they tell us nothing as to whether the spots increase in size, or whether new ones are added. In expressing this opinion, Bourquin had only five cases under consideration, those of Collins, Coats, Fleischer's two cases and one of his own. The case described by Cerise and Chéné is mentioned but is not considered by him in his discussion of the stationary or progressive character of this condition. In 1911, Cerise and Chéné showed a patient with an eye affected for three months, following a difficult confinement. The sclerotic presented irregular slate-coloured spots more or less pigmented. During these three months the pigmented spots became larger. Rochon-Duvigneaud and Dupuy-Dutemps were of opinion that the sclerotic pigmentation was congenital. Since 1917, Hildesheimer in 1920 showed a case of congenital melanosis of conjunctiva and sclerotic in which the pigmentation had gradually increased. The Friedenwalds in 1925 found the condition unchanged in a girl, aged nine years, whom they had observed for four years. The sclerotics showed bluish, almost purple discoloration. In 1913, Dr. Fridenburg made a coloured drawing of the eye of Davis' case, a boy, aged 11 years, with two slate-coloured patches in his right eye; in 1927 he saw the case again and found practically no change. The cases which Bourquin had under consideration were as follows. (1) A case of melanosis of the uvea and sclera, with areas of brown pigmentation in the sclerotic described by Collins in which the melanosis had not altered from the time he had seen it ten years previously. (2) A case reported by Coats in which the family practitioner saw a small spot on the sclerotic shortly after birth, at four years of age the spot was larger, at fifteen years of age the pigmentation of sclerotic was much greater. Two cases of Fleischer. (3) The first case, a female, aged 3 years 6 months at first examination, showed no considerable change in seven years. (4) In the second case, a female, aged two years when first seen, pigmentation of the sclerotic had advanced in two years. (5) Bourquin's case, a female,
aged 12 years, in which the spots remained unchanged for two years.

In view of these conflicting results and the dearth of reliable observations a few cases seen by me in the years 1910 to 1925 were recently re-examined. By comparing the appearances with those seen at the first examination it was hoped to determine whether the coloured areas were stationary or progressive, whether they enlarge, increase in number, or change colour as does the iris in early life, and if so at what period the change occurs.

Of nine cases sought for six were found, one had died, one is abroad, and one could not be traced. At the time of the first examination a hasty sketch, and in case No. 7 a coloured drawing of the eye was made, which in conjunction with descriptive notes has been of use when comparing the then appearances of the sclerotic with those at the present time. The eyes were not originally examined with the purpose of comparing their appearance at a later date, but the data recorded at the first examination are sufficiently accurate to show whether any considerable change had occurred in the interval.

The notes of the cases preceded by a short précis of each are as follows.

The first case has the coloured sclerotic patches limited to one eye, which has a darker brown iris than the other eye and a more deeply pigmented fundus. With the exception of one area there is nothing to suggest that the purple patches altered in extent, number, or colour from the age of 7 to 18 years. In the darker iris which has a uniform appearance, except at the pupillary part, the usual surface markings are absent. Pigment spots are present in the skin on several parts of the body.

Case 1. April 16, 1921. Eva G., aged 7 years, a difference in the appearance of her eyes first noticed two years after birth and attributed to a fall down stairs.

Examination of eyes: Ps. equal, contract to light, eye movements full, uncertain nystagmus on extreme lateral movements of the eyes, no ptosis, T.N. Right eye: sclerotic has no purple areas, iris dark brown, fundus dark, no choroidal vessels definitely seen, refraction H. Left eye: sclerotic has extensive purple areas on three-fourths of the ciliary region, down and out they are very faint, anterior edge of each area is well defined and situated at a distance of from 2 to 4 mm. from corneal margin, posterior boundary of each area is visible but not well defined as it merges into the normal colour of the sclerotic, conjunctival vessels are conspicuous and moveable over the areas; iris very dark brown showing little distinction from the black pupil: fundus darker than right fundus, much reflection from retina which has a grey appearance; refraction H. Hair of head medium brown with tinge of red, eyebrows dark brown, eyelashes very dark brown; skin, medium shade of colour, has a number of pigmented spots on several parts of the body. Patient is fifth born in a sibship of six. No relative is known to have the eye condition that she presents. Father’s iris blue, mother’s brown. Father says that the children all took after their mother as regards eye colour.

Re-examined March 1, 1932, occupation printer’s feeder, health good. Right eye: sclerotic has no purple areas; iris brown with normal surface markings; V. 6/6 HM. 0°50 D., 1 J. Left eye: sclerotic purple areas have not altered in distance from corneal margin, excepting possibly an area at inner part which now nearly reaches corneal limbus, their posterior boundaries are visible, oculur conjunctival vessels appear as on first examination; iris, much darker brown than in other eye, surface uniform from absence of surface markings, except in pupillary region, a few minute brown elevations present at outer part of iris and near pupil, V. 6/6, not fully, with −0°25 D. =6/6, 1 J., field of vision full; Ps. equal, contract to light, no nystagmus; skin not dark.
COLOURED AREAS IN THE SCLEROTIC 673

The second case has unilateral violet sclerotic patches in the eye with darker brown iris and fundus. During a period of more than 20 years, from 1911, when she was 3 years of age, to 1932, the violet colour appears to have extended, for at the first examination the coloured area was separated from the cornea by normal white sclerotic, whereas at the second examination it had reached the corneal margin both above and below. Support is given to the accuracy of these observations by the mother’s statement, given quite voluntarily, that the affected areas had become darker and had extended. Numerous minute brown smooth elevations cover the surface of the darker iris.

Case 2. April 29, 1911. Jessie R., aged 3 years, diagnosis cyanosis bulbii.* Her father noticed a mark on lower part of sclerotic shortly after birth and thinks the purple marks on left sclerotic have become more conspicuous and the brown of left iris has got darker since birth. He cannot say there was any difference between the right and left iris at birth. EXAMINATION OF EYES: Right eye: sclerotic normal; iris dark brown; fundus normal, refraction H. Left eye: sclerotic has a diffuse violet area separated from cornea by white normal sclerotic, normal coloured sclerotic visible beyond posterior boundary of the violet area; iris diffuse black-brown; fundus much darker than the other; refraction H. no moles anywhere on the skin.

RE-EXAMINED February 16, 1932. Occupation ‘cellist. Hair of head, eyebrows and eyelashes dark brown, skin darker than that of mother and sister; Fs. contract to light. Right eye: sclerotic normal; iris brown with normal surface markings, no nodules; fundus, choroidal vessels visible. V. 6/6, HM. 0*75 D., 1 J. Left eye: sclerotic, violet areas surrounding the cornea extend to its periphery above and below, no part of the violet areas extends backwards to the equator of the eyeball; iris dark brown almost black and difficult to distinguish from pupil, on its surface are numerous minute smooth brown elevations seen best with the aid of a binocular loupe when she looks downwards. These elevations are present all over the iris surface; fundus darker than in other eye, no choroidal vessels seen, V. 6/6, HM. 0*75 D., 1 J., field of vision full. Father’s iris blue, mother’s iris brown. Jessie R. is sixth born in a sibship of seven. The first born, male, died aged 8 years, had blue eyes, no violet patches. The second, male, had brown eyes. The third, female, died, aged 7 years, had dark auburn hair, no violet areas. The fourth, female, died at 11 months, had blue eyes, no violet patches. The fifth, male, died, aged 1½ years, had brown eyes. The sixth, Jessie R. The seventh, Betty R., aged 20 years, iris brown.

The third case, has a unilateral purple and brown coloured area in the sclerotic nearly completely surrounding the cornea and extending to its margin. When comparing a sketch of the eye made in 1920, at the age of 30, with the appearance seen in 1932, some extension of the coloured area downwards and nasallywards is suggested, but it cannot be definitely stated that an increase has occurred, no measurements having been taken. The light colour of the iris of this eye is to be noted, as a pale iris is unusual in this condition.

Case 3. September 8, 1920. Bella M., aged 20 years, domestic servant, complained of headache over left eye. EXAMINATION OF EYES: Right eye: sclerotic no note: iris pale green; fundus normal; refraction, after H. and C. drops, V. with +1'25 D. = 6/6; T.N. Left eye: sclerotic at ciliary region shows pigmentation in form of purple and light brown areas with unpigmented parts between. They extend to corneal margin except at inner part, conspicuous conjunctival vessels over the purple and brown patches above and below the cornea; iris pale green; fundus much darker than the right fundus and general colour of background is grown than that in other eye; refraction, after H. and C. drops, V. with + 0'25 D. sph. = 6/6 T.N.

RE-EXAMINATION February 24, 1932. She is married and has a daughter. Right eye: sclerotic normal; iris pale green; refraction, V. 6/6, HM. 0*75 D. Left eye: sclerotic, purple area which nearly completely surrounded the cornea in 1920 appears from comparison with a sketch made at the time to have extended downwards and nasallywards; conjunctival vessels as seen at first examination; iris

*A term used by Liebreich.
pale olive green above and blue below (examined in good day light), surface markings normal, no small brown nodules present, upper part of this iris is just darker than the pale green of the right iris; refraction, V. with +0'75 D. cyl. = 6/6, Ps., equal, contract to light; skin is blonde, hair, eyebrows and eyelashes brown, not dark. No relative known to have purple patches on sclerotic. Father has blue eyes, mother dark brown eyes. She is first born in a sibship of three. Two brothers have brown eyes. Her infant child has blue eyes.

The fourth case has unilateral purple sclerotic areas, and a darker brown iris and fundus in the affected eye. No alteration occurred in the appearance of the sclerotic between August 19, 1925, when her age was 45 and February 20, 1932.

Case 4. August 19, 1925. Annie C., aged 45 years, had difficulty in reading. Examination of Eyes: Right eye: sclerotic has purple areas in ciliary region extending from corneal margin upwards and two small circular purple areas near inner margin of cornea, in none of these is the colour pronounced; iris very dark brown; fundus darker than in other eye; refraction, V. 6/6, HM. 0'75 D. Left eye: sclerotic, no purple patches; iris brown with blue-green peripheral; fundus normal; refraction, V. 6/6, HM. 0'75 D.; Ps., equal, contract to light; T.N. Re-examined February 20, 1932. Right eye: no change in appearance of the purple patches, sclerotic above the patches is yellower than usual, none of the patches is so far as she can see, extend backwards so far as she can see, extending from corneal margin upwards and two small circular purple areas near inner margin of cornea, in none of these is the colour pronounced; iris very dark brown; fundus darker than in other eye; refraction, V. 6/6, HM. 0'75 D. Left eye: sclerotic normal; iris surface markings have same appearance as in right iris; fundus no change; V. 6/24, with +1'25 D. = 6/6; Ps., equal, contract to light, no signs of old iritis in either eye. Hair, eyebrows and eyelashes brown; skin rather dark, she mentioned a small dark spot on her back. No relative known to have coloured patches in the sclerotic. Her father had grey eyes and mother brown eyes. She is third born in a sibship of five. The first born, male, grey eyes; second born, male; fourth born, female; and fifth born, female, eyes dark; she, Annie C., has five children. First born, female, brown eyes, has two male children, the elder has brown eyes, the younger grey eyes; second born, female, has grey eyes; third born, female, has blue eyes; these two and the fourth born, female, are married with no offspring; fifth born, male, has dark eyes.

In the fifth case the coloured part of the sclerotic formed a nearly complete zone about 4 mm. in breadth around the cornea of one eye. The brown iris and fundus were darker in the affected eye. She had disseminated sclerosis, was examined only once and died two years later.

Case 5. June 21, 1921. Margaret G., paperbag maker, aged 25 years, subject of disseminated sclerosis. Examination of Eyes: Right eye: sclerotic in anterior part has two purple areas, one of which forms a zone nearly completely surrounding the cornea with a breadth in most parts of about 4 mm. and reaching the corneal margin at its upper, outer and lower parts. The purple colour is deeper above the cornea and in one portion of the zone is most intense along sclero-corneal margin and at the part furthest from cornea. The sketch suggests a collar round the cornea open at inner side, with an isolated purple area situated above and separated from it by normal sclerotic; iris dark brown; fundus very much darker than that of left eye, no choroidal vessels visible, optic disc normal, refraction, V. = 6/6 nearly fully, not improved by lenses. Left eye: sclerotic has no purple patches; iris blue-grey with much yellow in stroma; fundus, choroidal vessels are clearly seen, optic disc obviously too pale, edge very sharply defined, retinal vessels not contracted; V. = 6/12 not fully, has central scotoma for red, both fields show some peripheral contraction, especially left one; Ps. equal, contract to light; lateral nystagmus in lateral positions of eyes. Hair of head, eyebrows and eyelashes very dark brown. She knows of no one with an eye resembling her own right eye. No Re-examination. February 17, 1932, her father's sister said that Margaret G. died in 1923, that she was a legitimate child without brothers or sisters, and that her father had blue eyes, her mother's eye colour she did not know.
The sixth case has several violet spots in sclerotic distributed around both corneae. Most of the spots are small and have not altered in appearance from 1919, when she was 17 years of age, to 1932. The iris has the usual surface markings and equal depth of brown in both eyes. A daughter has a dark grey patch in ciliary region of sclerotic.

Case 6. December 5, 1919. Eliza R., aged 17 years, clerkess, complains of difficulty in seeing. Examination of Eyes: Right eye and left eye: sclerotic, in each eye are a number of small and one or two rather large lilac and pale violet areas around the cornea, sketches show seven of these in each eye, none of them extends to sclero-corneal margin; iris rich brown; fundus normal, after H. and C. drops R.V. with −1.50 D. = 6/6, L.V. with −1.25 D. = 6/6. Hair medium shade of brown, complexion blonde. Re-examined February 17, 1932. She is married and has two daughters. Right eye and left eye: sclerotic, no change detected in number or size of the violet spots; iris brown with usual surface markings, no small brown elevations present; Ps., equal, contract to light; eyelashes, eyebrows and hair brown; no moles or naevi in skin. In the right eye of her first born child is a faint dark grey patch in sclerotic situated in ciliary region down-out from corneal margin. Both eyes of this child have epicanthus, and iris is brown. The younger daughter has blue iris.

The seventh case. A carefully executed coloured drawing made by Mr. J. B. Souter at the time of the first examination and since preserved from the light in a closed cabinet is available for comparison at subsequent examinations. In respect of colour, therefore, it is the most reliable case of the series. The sclerotic patches had changed colour when seen in 1932, nearly 22 years after the first examination. They were definitely darker than in 1910, when she was 9 years of age. Also another change had apparently occurred for a small circular violet patch, not shown in the drawing, was situated near the cornea. It is, of course, possible that this had been overlooked and not drawn. In reply to a letter on the subject sent to the artist he says "I should be inclined to say that the spot must be of later development because the drawings were done very carefully under your supervision, and I do not think anything then visible could have escaped your notice." The refraction of the affected eye, at first hypermetropic, became myopic whilst that of the other eye was hypermetropic and remained so. The affection is unilateral, with iris and fundus more deeply pigmented in the eye with melanosis.

Case 7. May 14, 1910. Rina B., aged 9 years, school-girl. Examination of Eyes: Right eye: eyeball normal, conjunctiva free of pigment; iris dark brown; fundus dark; refraction, V. 6/6 HM. 0·50 D., 1 J. Left eye: sclerotic, purple or violet areas in ciliary region, with conspicuous blood vessels in front of them. These areas are nearly homogeneous, but are darkest at part nearest the cornea, with a quite sharply defined anterior margin. The largest area is above and extends to corneal edge, other areas are separated from the corneal margin by 1 to 3 mm. of normal sclerotic, none of the areas extends so far back as to prevent the posterior margin being seen, this fades imperceptibly into normal sclerotic; iris very dark brown, much darker than right iris, no dilated vessels in it; fundus generally shows rather less red than does right fundus, especially noticeable at periphery, and has a slate coloured appearance, optic disc looks very red, retinal vessels normal, refraction, V. 6/6, HM. 0·25 D., 1 J. Corneae of equal size, Ps., equal, contract to light, T.N. Hair dark brown, eyebrows black, eyelashes brown. Skin contains numerous pigmented brown spots, one of 2·5 cm. at right outer canthus. According to the mother, purple areas on left eye have been present since birth. Re-examined February 3 and 20, 1932. Occupation, clerkess, unmarried. Right eye: sclerotic no coloured patches; iris has usual surface markings, fundus very dark, V. with −0·50 D. sph. = 6/6. Left eye: sclerotic violet coloured areas were compared with a coloured drawing of them made in 1910. They have become darker, more marked in day light than in artificial light; a small circular violet patch situated close to and down-in from cornea does not appear in the drawing, apart from this no new violet areas and no extension of old ones have appeared, blood vessels over the violet areas are now more obvious; iris surface markings as in right iris, no minute brown elevations, fundus very dark, much greyer than in other eye; refraction, V. 6/24 with −0·50 D. sph. = 6/6, 1 J.; Ps., equal, contract to
Case No. 7.

The upper two drawings are of the right and left eyes. Both lower drawings are of the left eye. The illustrations made from the coloured drawings, fail to show the much lighter colour of the right iris. Some of the characters of the coloured sclerotic areas are shown, but obviously not their violet or purple colour.

light; dark complexion. She is the first born in a sibship of three. The second born, male, has blue eyes, and the third born, male, has brown eyes. Father's iris brown, hair black; mother's iris blue-grey, no "black" blood in father's or mother's relatives, no consanguinity.

The eighth case has bilateral small purple patches. Depth of pigmentation of the brown iris and fundus is the same in both eyes. Brown pigment spots are present in the ocular conjunctiva of the right eye.

Case 8. December 7, 1914. Andrew N., aged 15 years, motor-mechanic. Attended for conjunctivitis. Examination of Eyes: Right eye: sclerotic has a small purple patch a short distance above corneal margin, and another 2 mm. in diameter down-in from corneal margin, two dark brown pigment spots in conjunctiva close to corneal margin below; iris dark brown; fundus dark with much reflection from retina; V. 6/6 nearly fully, no H.M. Left eye: sclerotic has pale purple patches, one patch about 8 mm. in length and 2 mm. wide is situated up-in and near corneal margin, two much smaller ones are placed up-out and down-in
COLOURED AREAS IN THE SCLEROTIC

respectively: iris dark brown; fundus dark with much reflection from retina; V. 6/6, no H.M.; Ps. equal, contract to light; eye movements full; hair very dark nearly black; no moles on skin. No Re-examination: on February 16, 1932, it was ascertained that he was working abroad and had no trouble with his eyes. He is the second born in a sibship of three, all males. His mother previous to marriage had a son and daughter by another man. Parents not consanguineous. Mother’s iris blue, father’s dark brown.

The ninth case has a unilateral very large purple area above in the anterior part of the sclerotic. The ocular conjunctiva is pigmented and forms a black oval on the upper part of the coloured sclerotic area. There is no difference in depth of pigmentation in the upper and lower parts of the fundus, which is dark in both eyes. The iris of the eye with melanosis is not appreciably darker than the iris of the other eye. Brown pigment spots are present in the skin on several parts of the body.

Case 9. June 23, 1912. Alexander D., aged 14 years, farm servant. Examination of eyes: Right eye: sclerotic has large single purple area occupying most of its anterior upper part and extending to within 2 5 mm. of corneal margin, posterior to this area sclerotic has normal appearance; an extension of the area upwards and outwards, however, cannot be followed to white sclerotic even when upper lid is completely retracted and patient looks down; pigmented conjunctiva obscures much of upper part of the area and forms a black oval about 2 cm. in length and 3 to 6 mm. in breadth with long axis horizontal, conjunctival vessels seen in front of the pigment; iris, dull greenish brown, not darker than that of other eye; fundus dark, no difference between pigmentation of upper and lower parts: V. 6/6, HM 0’50 D. Left eye: sclerotic has no purple patches and conjunctiva is not pigmented; iris dull greenish brown; fundus dark; V. 6/9 not fully, with −0’50 D. cyl. = 6/6. R.P. rather smaller than L.P., both contract to light, eye movements full, no strabismus. He is a strong healthy boy with brown hair, eyebrows and eyelashes, complexion rosy. Skin fair, has a number of isolated brown pigment spots in various parts of the body. He is first born in a sibship of five. The others, two brothers and two sisters, in that order of birth, have no birth marks anywhere. Father and mother also free from naevi and moles. The blackness on upper part of right eye was first noticed by his mother three years ago. No Re-examination, 1932. No trace of him could be found after extensive enquiry.

From the accounts given of the six re-examined cases of melanosis of the eye it is evident that no great change has taken place in any of them. In two of the cases there is definitely no change. In one case, No. 7, the violet patches became distinctly darker and a violet spot seen on second examination was not present in a coloured drawing made at the first examination. In another case, No. 2, the violet coloured area which at first examination had been separated from the cornea by normal white sclerotic was found at the second examination to be touching the cornea. In case No. 1, a purple area nearly reaches the corneal margin which at first examination was at least 2 mm. from it. In case No. 3, a suspected extension of the violet area backwards is much more doubtful. The ages when first examined of the two cases showing definite changes, Nos. 7 and 2, were 9 and 3 years respectively, and the ages of the other four, Nos. 1, 3, 4, and 6, were 7, 20, 17, and 45 years. The intervals of time between the two examinations in the former cases were 21 and 20 years, and in the latter cases, 11, 11, 6, and 12 years. It would be of interest...
to know whether greater changes would be found in cases examined in the first three years of life.

The characters of the coloured sclerotic patches in the present series are as follows. Their colour is described as purple, violet, and lilac; slaty-blue would be a good description of some of them. In the majority of cases the patches are limited to one eye. In both bilateral cases Nos. 6 and 8, the coloured areas are small as in three of Selter's bilateral cases, but the area may be large as in the Friedenwalds' case. Bilateral incidence, though unusual, is not rare for Coats mentions cases recorded by Paget, Chaillous and Coover, and cases have been published by Kestenbaum and the Friedenwalds, two cases each by Bourquin and Jablonski, and four cases by Selter. In some eyes of the present series a single patch is present, in others several occur. These are situated in the ciliary region. In some cases, Nos. 1, 6, 8 and 9, normal sclerotic separates them from the cornea, as in cases seen by Talko, Schaumberg, Behr and Bourquin (first case). In other cases, Nos. 2, 3, 4, 5, 7, the coloured areas reach the corneal margin at one or more places and in some they extend along nearly its whole circumference, as in cases reported by Coats, Fleischer and Bourquin (third case). The size of the coloured areas is very variable. Some occupy a large part of the sclerotic, others are small and in some cases measure only 1 or 2 mm. The posterior limit of a patch is usually readily seen though it tends to become faint as it merges into normal sclerotic and contrasts with the better defined anterior margin as in cases Nos. 1 and 7. In case No. 5, however, the posterior margin of the coloured area in places is as well defined as the anterior margin. In all cases with one exception the whole extent of each coloured area is visible. In case No. 9 the coloured area extends in one direction so far backwards that normal sclerotic cannot be seen beyond it. In this connection it may be noted that in excised eyes pigmented areas have been found at the posterior part of the sclerotic. Talko (1869) found in both eyes of a case of meningitis whitish slate-grey pigment spots confined to the posterior part of the sclerotic from equator to optic nerve. The black colouring of the sclerotic was not due to thinning but to pigmentation of its most superficial layers. Hulke (1860) found in an excised eye, with melanotic cancer of choroid, the sclerotic mottled with purple spots that were numerous in the ciliary region and around the posterior pole. Reese (1925) examined microscopically an eye with melanosis oculi which had pigment spots over the entire scleral surface and contained a melanotic sarcoma of choroid. Doherty (1927) examined a glaucomatous negro eye with melanosis oculi. Macroscopically the excised eye in its anterior third presented a diffuse collar of pigment, the posterior third showed isolated black blotches.
Coloured Areas in the Sclerotic

In some cases in this series besides abnormal sclerotic appearances other conditions were present of sufficient interest to mention. The iris in case No. 3 had a light colour which must be very unusual, for Bourquin found that not one of more than 60 cases of melanosis bulbi had a light coloured iris—blue, green, grey—in association with sclerotic spots. A comparison of Streiff’s case with this one is of interest. In his case the iris was pale olive green at nasal part and dark brown at temporal part. Faint violet-grey spots were present in the sclerotic only in the region adjoining the brown part of the iris. In the present case both irides were pale, and though the upper part of the iris in the affected eye was a shade darker than the rest of the iris the coloured sclerotic area almost completely surrounded the cornea. Another interesting condition is seen in case No. 2, in which the numerous minute brown elevations that cover the surface of the left iris are comparable with those described by Coats and Treacher Collins in 1912. Since their cases were published a number of others showing similar appearances have been reported by Langdon, Kuboki, Streiff, the Friedenwalds, Fleischer, Gjessing, Bourquin, and Larsen.

Associated pigmentary conditions of the conjunctiva and of the skin are not uncommon in melanosis bulbi. The former occurs markedly in case No. 9, in which a large black oval appears in the ocular conjunctiva, and in a minor degree in case No. 8. Pigmented spots were present in the skin in cases Nos. 1, 7 and 9. There was no evidence of disease in any of the eyes with melanosis excepting in case No. 5 with disseminated sclerosis.

The refraction of the eye with melanosis, according to Bourquin, presents nothing special. In the present series, four of the eleven eyes with melanosis have a low degree of myopia. The remaining seven eyes are hypermetropic or emmetropic. In case No. 7 the refraction of the eye with melanosis changed from hypermetropia to myopia whilst in the other eye there was no such change. Jablonski in a case of congenital melanosis of the sclerotic found the more melanotic eye by far the more myopic. On the other hand a case of Fleischer’s was emmetropic in the pigmented eye and myopic in the other.

Few hereditary cases have been recorded. Jablonski saw blue-black discoloration of both sclerotics in two sisters. In Bourquin’s first case, a male, aged 44 years, the son and father were also affected, melanosis bulbi thus appearing in three consecutive generations. Selter recorded sclerotic patches in two generations, a father, his daughter and two sons. The sons were twins, aged three years. In case No. 6 of the present series a mother with melanosis of both eyes has a daughter with a dark grey patch in the ciliary region of one eye. In none of the other families is there any suggestion of inheritance of sclerotic patches. The information,
however, is limited to statements received from patients and their parents. In none of the sibships is there more than one individual with coloured areas in the sclerotic. The order of birth of the affected individuals is known in six cases. Case No. 1 is fifth in a sibship of six, case No. 2 is sixth in a sibship of six, cases Nos. 3 and 7 first in sibships of three, case No. 8 second in a sibship of three, and case No. 9 is the first of five. In five instances the colour of the iris of both parents is known and in four of these the iris of one parent is blue and of the other brown, in the fifth instance the iris is grey in one parent and brown in the other.

In conclusion, of the six cases with coloured areas in the sclerotic which have been re-examined after a period of years it can be confidently stated that the appearances have not changed in two cases, but that definite changes have occurred in other two cases, in one as regards colour, in the other as regards increase of area. In the remaining two cases the evidence is not sufficient to warrant any statement that there has, or has not, been any change. The results suggest that further observations are needed and that measurements are necessary on first examination in order to detect subsequent minor changes. Information is particularly required regarding what occurs during the first two or three years of life.

REFERENCES

1. Behr.—Diss., Heidelberg, 1903. Quoted by Bourquin.
4. Chaillous.—Quoted by Terson.