Two Cases of Congenital Night-Blindness.

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In the recent communication of my paper on a "Fourth Sensation in Colour Vision," in April last, attention was called to the case of night-blind eyes as throwing light on the question of the functions of the rods and cones in the retina in regard to colour and colourless vision. I cited two cases of congenital stationary night-blindness, which, through the kindness of Mr. Nettleship, were brought to my laboratory for examination in view of certain researches in which Prof. (now Colonel) Watson and myself were together interested. As Colonel Watson has been long at the Front in France, and as he conducted a principal part of the only partially completed examination, I hesitated to give the details without his collaboration. I have now obtained his acquiescence in my request that I should communicate the results obtained to the Royal Society. I now do so. As a matter of fact, I had worked out the observations before my colleague left me for the Front some 2½ years ago, and these results I have put in the following communication:

I may say that any other form of night-blindness than those to which the late Mr. E. Nettleship introduced us would have been useless for the investigation on which we were engaged, as, if it were not so, night-blindness due to disease might call in question some of the deductions to be made. Mr. Nettleship's investigations of the family to which they belonged, and its history, left no doubt that we were dealing in our two cases with genuine cases of congenital stationary types.

The subject of congenital night-blindness (sometimes called moon-blindness) has been left with several obscure points unexplained, and the present communication, it is hoped, may throw light on some of them.

The late Mr. E. Nettleship collected a large number of pedigrees in which the characteristics of night-blindness are shown. His papers on the subject contain, it is believed, nearly everything that is worth knowing. The papers were published in the 'Royal London Ophthalmic Hospital Reports.'* He has amply proved that congenital night-blindness is hereditary. He commenced with the work that the late Florent Cunier† published in 1838 on a family in

* See vol. 17, Part III, and vol. 27 of the 'Ophthalmic Society's Transactions,' besides others.

† Fl. Cunier, 'Médecin Militaire,' "Histoire d'une Héméracopie Héréditaire depuis Deux Siècles dans une Famille de Vendemair près Montpellier."

which night-blindness existed. Cunier gave long genealogical pedigrees with the names, domiciles, and dates of birth and marriages of the members. The total number of persons in seven generations was 629, of whom 56 were affected by night-blindness. By subsequent researches Mr. Nettleship was able to enlarge the pedigree, which as it stands now reaches 2121 persons, 1001 males and 960 females (of the remainder, sex not known), of which 72 males and 62 females, and one (sex not stated) are known to have been night-blind, and all of whom sprang from a common ancestor, Nougaret by name, who lived about 1600.

This is only one of the several family pedigrees which Mr. Nettleship gives, but all show that there is heredity in congenital night-blindness. There is a great distinction between the night-blind and the congenital monochromatic vision, of which cases are rare. The one can see in daylight all the spectrum colours as ordinary normal vision does, the other does not. A quotation from one of Mr. Nettleship's papers will give an idea of how congenital night-blindness manifests itself. After a description of the retina and refraction, he says: "(The patient) is extremely night-blind, but can see some of the brighter stars, and can do well by bright moonlight and artificial light. On a moonless night has a great difficulty in finding his way, cannot see the street before him, but guides himself in keeping to the middle of the road, and looking up and recognising the sharp and black line of the house tops against the sky.

"A poor man, vine labourer, slow and awkward by training, but intelligent and most graphic in his description if allowed time to find words and gestures. In walking up and down the partially darkened room" (in which he was examined), "he would put his hands out and sometimes stop altogether; indeed, he had the aspect of one blindfolded. He was quite unable to see the fingers or even the back of the hands at 0·3 metre with an illumination that was ample for a normal person. He had been like this as long as he could remember (he was 46 years old), but had, nevertheless, been obliged to serve five years in the (French) Army. His sight is not getting worse.

"This man's son, 16 years old, is as night-blind as his father."

In the case of the monochromatic vision to which I have alluded, they all saw well in low illumination, and but moderately in bright daylight. These cases have almost exactly opposite kind of vision. For this reason a physical examination of the vision of the night-blind seems desirable, and though this examination was not completed in some points such as colour field, it is believed that we have indications of marked differences in the perception of colours and light when tested by the spectrum method.

The clinical aspects of night-blindness, of course, my colleague and myself are unable to discuss. It suffices to say, I think, that these two cases fulfilled the object we had in view, viz., to examine two of the congenital cases which (the late) Mr. Nettleship included in one of his pedigrees.

The first patient, Mr. E., a clergyman, came to the laboratory on February 25, 1913, when he was examined in the writer's apparatus and presence for his extinction of colour from the red to the blue of the arc spectrum.

The luminosity of the D line coming through slits to the screen was the standard of the intensity of the luminosity of the different rays of the spectrum. This was gradually diminished by an annulus placed in the path of the ray until he just saw no light in the illuminated small square in the darkened camera attached to the spectroscopic apparatus. He said that all light had vanished when the colour was extinguished. He repeated the observations in a reverse manner, noting the advent from darkness of the spectrum colour.

This series of observations was repeated in a second apparatus in which the source of light was a Nernst lamp, the thread of which was rendered incandescent by an ampère of current from a battery of 100 volts; the current was kept constant. Similar observations were made by this apparatus and recorded, and afterwards converted to the arc scale. Finally, the luminosity curve of his spectrum was taken and compared at the time with that of my colleague.

In his observations he invariably said that, when the extinction was noted by the reduction of the said intensity, whenever the colour was gone all the light had also gone. In other words, the same reduction in intensity of the light was the threshold for both light and colour. In the paper, "The Threshold of Vision for Different Coloured Lights,"* the same identity of extinctions of light and colour at the fovea of Class I retina is to be noted, as given by all observers, though the extinction of light outside the fovea is nearly 0·0001 times less than it is for colour at the fovea.

Comparing the loss of colour for my own eye (Class II) when the intensity of the spectrum D is one candle-foot, the colour is extinguished at an intensity of about 0·0016 candle-foot, and the extinction of light at about 0·000035 candle-foot intensity. For "E." the extinction of both light and colour in both cases takes place at about 0·0015–6. This indicates that the extinction of the feeble white light (or, as I have called it, of the fourth colourless sensation) is dependent on some other retinal perception that the normal eye possesses beyond that possessed by the night-blind. The same is probably the cause of the difference in the fovea of No. I retina compared with that of No. II

* 'Phil. Trans.,' A, vol. 216.

retina. If the rods and cones fill the places in reference to the light sensations which have been allotted to them, as stated in my last communication on the fourth sensation, then there is an absence of sensitive rods in the whole retinæ of the night-blind. The same remarks may apply to the observer B. and his measures, though they differ slightly in shape for those of E., but not more so than do the curves of various observers in the 'Philosophical Transactions' paper just referred to.

On May 20, 1913, B. came for his examination at the colour laboratory. He is a clergyman, and is, we believe, a cousin of E. His examination was conducted on the same lines as that of E., the arc light alone being used. In addition, he was made to match the colour of the D sodium line with mixed colours of thallium and lithium blue, in what is called the anomaloscope, a very useful instrument, though only partially indicative of any defect in colour vision.

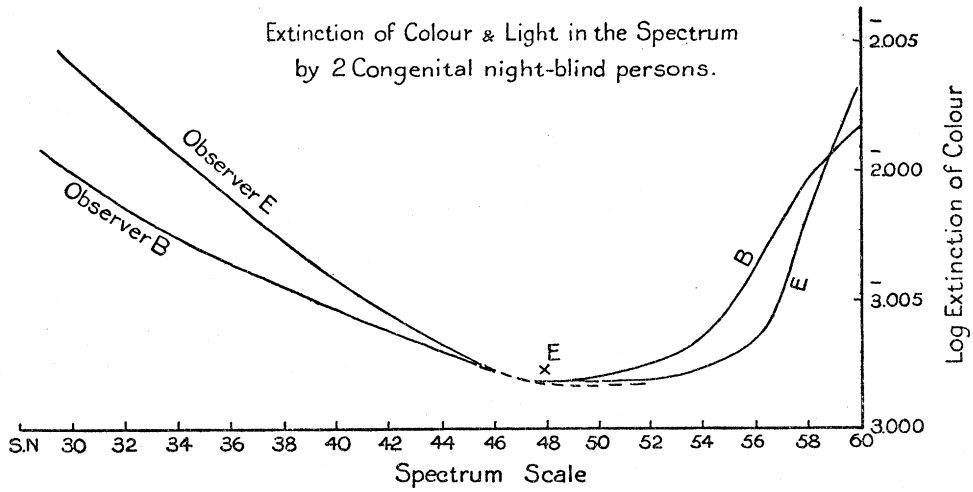
[In Colonel Watson's and my paper, jointly published in the 'Philosophical Transactions,' it was shown that there seemed to be two classes of retina: one in which only the colours of the spectrum were recognised in the spectrum and no colourless rays affected the fovea, and the other class in which both colour and the feeble colourless rays were equally effective. As we soon gathered the retinæ of the two patients at any part apparently were not stimulated by the colourless rays, we determined to compare them with the No. I class of retina, for which purpose we had to reduce the arc results into the Nernst coloured spectrum. This was done, and the night-blinds' extinction of colour coincided practically with the threshold of No. I retina at the fovea.]

It should be mentioned that Mr. Nettleship divides congenital night-blindness into two classes, both showing the same night-blindness. The one is myopic, but the other apparently normal, as far as refraction is concerned. The two observers appear to be in the first division.

The night-blind luminosity curves, taken with a luminous spectrum of the arc, compared with that of the normal eye, show that the intensity of colour is the same for both. Allowing for the difficulties of the shadow test, and a certain error in observation which is found in the flicker test, the curves of luminosity of both may be said to be the same. The luminosity curves can be best compared by making the night-blind measures the numerator and the normal measure the denominator of a fraction. The following are the fractions for E. and B.:—

S.S.N.	58.	56.	54.	52.	50.	48.	46.	44.	42.	40.	38.	36.	34.
$\frac{E}{N}$	1·12	1·12	1·06	1·00	1·00	0·98	1·00	0·92	0·90	0·93	0·96	1·02	1·05
$\frac{B}{N}$	0·97	1·00	1·00	1·02	1·02	1·00	1·03	1·08	1·08	1·08	1·10	0·94	1·05

Allowance has to be made for a difference in pigmentation of the foveal region. It has been shown in my recent communication on the fourth sensation that the added luminosity of the sensation of this fourth sensation is



negligible when the spectrum is of normal brightness (say when the D ray is of $\frac{1}{2}$ candle-power brightness at foot distance from the screen). These measures show that the luminosity of such a spectrum is, within the limits of error, the same in both cases.

Annulus 315. Observer E.

S.S.N.	Corrected annulus reading.	Absorption factor.	Product of 2 and 3.	Or	Reduction to one candle foot + $\bar{1}$·155 (log.)	Limit of vision.
60	66	−0·01170	−0·772	$\bar{1}$·228	$\bar{2}$·383	0·0242
58	106	−0·01184	−1·255	$\bar{2}$·745	$\bar{3}$·900	0·0079
56	143	−0·01197	−1·712	$\bar{2}$·288	$\bar{3}$·440	0·00276
54	158	−0·01212	−1·915	$\bar{2}$·085	$\bar{3}$·240	0·00174
52	153	−0·01229	−1·979	$\bar{2}$·021	$\bar{3}$·176	0·00150
50	124	−0·01244	−1·966	$\bar{2}$·033	$\bar{3}$·188	0·00154
48	136	−0·01259	−1·926	$\bar{2}$·074	$\bar{3}$·229	0·00170
46	144	−0·01273	−1·833	$\bar{2}$·167	$\bar{3}$·322	0·00210
44	136	−0·01288	−1·752	$\bar{2}$·248	$\bar{3}$·403	0·00253
42	126	−0·01302	−1·641	$\bar{2}$·359	$\bar{3}$·514	0·00374
40	115	−0·01318	−1·516	$\bar{2}$·484	$\bar{3}$·639	0·00436
38	105	−0·01334	−1·401	$\bar{2}$·599	$\bar{3}$·754	0·00560
36	91	−0·01354	−1·232	$\bar{2}$·768	$\bar{3}$·923	0·00838
34	78	−0·01374	−1·072	$\bar{2}$·928	$\bar{2}$·083	0·0121
32	66	−0·01390	−0·917	$\bar{1}$·088	$\bar{2}$·238	0·0173

Annulus 315. Observer B.

S.S.N.	Corrected annulus reading.	Obstruction factor.	Product of 2 and 3.	Or	Reduction of D to one candle foot + $\bar{1}$·155 (log.)	Limit of vision.
60	73	−0·01170	−1·088	$\bar{2}$·912	$\bar{2}$·097	0·00125
58	102	−0·01184	−1·208	$\bar{2}$·792	$\bar{3}$·947	0·00807
56	135	−0·01197	−1·156	$\bar{2}$·484	$\bar{3}$ 639	0·00436
54	147	−0·01212	−1·782	$\bar{2}$·218	$\bar{3}$·373	0·00236
52	155	−0·01229	−1·905	$\bar{2}$·095	$\bar{3}$·250	0·00175
50	157	−0·01244	−1·953	$\bar{2}$·047	$\bar{3}$·202	0·00159
48	157	−0·01259	−1·972	$\bar{2}$·028	$\bar{3}$·183	0·00152
46	152	−0·01273	−1·935	$\bar{2}$·065	$\bar{3}$·220	0·00166
44	144	−0·01288	−1·855	$\bar{2}$·145	$\bar{3}$·300	0·00200
42	136	−0·01302	−1·771	$\bar{2}$·229	$\bar{3}$·384	0·00242
40	128	−0·01318	−1·687	$\bar{2}$·313	$\bar{3}$·468	0·00294
38	120	−0·01334	−1·601	$\bar{2}$·394	$\bar{3}$·549	0·00354
36	112	−0·01354	−1·516	$\bar{2}$·484	$\bar{3}$·639	0·00436
34	103	−0·01374	−1·415	$\bar{2}$·585	$\bar{3}$·740	0·00550
32	93	−0·01390	−1·293	$\bar{2}$·707	$\bar{3}$·862	0·00727

B, 61·3; Li, 59·8; C, 58·1; D, 50·6; E, 39·8; *b*, 37·7; F, 30·05; Blue Li, 22·8; G, 11·2. The above give the positions of Fraunhofer lines on the (arc) spectrum scale.