THE SYNDROME OF VISUAL ALEXIA WITH COLOUR AGNOSIA.

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CASES showing alexia which is not part of a general visual agnosia or of a speech disorder (letter or word blindness, cécité verbale pure) are uncommon and deserve careful study for a variety of reasons. The conception of word blindness as loss of visual memories due to a localized cerebral lesion (Hinshelwood, 1917) has been challenged by later investigators who aimed at a more general analysis of impairment of cerebral functions. The application of the principles of Gestalt psychology has introduced new aspects into the study of word blindness. Investigations into acquired word blindness, apart from their psychological and neuro-pathological interest, are of value for the understanding of congenital word blindness which has been found to be a condition of considerable practical importance. It was the investigation of cases of acquired word blindness which led Hinshelwood to the first full description of the congenital disability.

Word blindness is, as a rule, associated with a disability termed "colour agnosia." The nature of that symptom and its relationship to visual agnosia on the one hand, and to aphasia on the other, have been the subject of controversy over many years. At first it was regarded as an aphasic symptom, later as a form of agnosia. More recently it has again been understood to be caused by the impairment of the same basic function which underlies amnesic aphasia.

The following two cases might contribute to the elucidation of some of the problems outlined above. The first case was under observation for ten months, while in the second case the syndrome was present for two weeks only.

CASE I.—Mr. J. A.—, aged 63, retired engineer, was admitted as a voluntary patient on 23.iii.46. His wife gave the following history :

The patient had been healthy physically and mentally until twenty months prior to his admission. He was of a cheerful disposition and was regarded as a man of good intelligence. He had left school at the age of I_4 and had no special training in mathematics. His main interests, apart from his work, were sports and politics. He used to take two daily papers and read them thoroughly. He also read engineering books. He had no difficulty in distinguishing colours before his illness. He used to be an excellent writer. In June, 1944, he complained of a sudden feeling of giddiness. There was no loss of consciousness; his right arm was weak and numb for a few days. Since that attack he had been unable to read. His memory was bad for some months after the attack, but improved gradually. Apart from an occasional difficulty in remembering names, his speech was unimpaired. During the year preceding his admission he had become very irritable and suspicious. He accused his wife of being unfaithful to him and trying to rob him. He complained about insomnia and irritability.

On admission the patient did not show any conspicuous mood or behaviour disorder. When asked why he had come to hospital he referred to the attack in June, 1944, and to his insomnia. Throughout his stay in hospital he never mentioned his suspicions and when questioned either denied or refused to discuss them. His behaviour while under observation was entirely normal.

Physical examination.—The patient was in good general health. His heart was slightly enlarged to the left and the second aortic sound was accentuated. B.P. 170/105. The peripheral arteries were rigid and tortuous, and the fundi showed signs of arteriosclerosis.

Nervous system.—There was a right-sided homonymous hemianopia with intact perimacular fields. The left visual fields were normal for white and colours (green, blue and red were tested). Visual acuity § with correction. Those findings were verified by the consultant ophthalmic surgeon, Dr. Fraser. There was no disturbance of gazing. No disturbance of sensation. Stereognosia intact. Serological and cytological examination of the blood were negative. Urine negative.

Throughout the period of observation, which extended over more than ten months, the patient was most co-operative and submitted willingly to testing for any length of time. He regarded the various tests as methods of rehabilitation. It was always easy to attract his attention and he never spontaneously complained of getting tired, although his performance deteriorated when an examination extended over more than half an hour.

The patient was correctly orientated for person, space and time. His general knowledge and knowledge of current events were fair. He sometimes failed when asked the names of leading political personalities, but he could tell their ranks and the parties they belonged to, as well as their most conspicuous personality features. When their names were offered to him among others he picked them out correctly. Recall was moderately impaired. When tested he often failed to remember correctly a date and an address after more than five minutes, but he would remember the nature of the item he was supposed to recall, even after several hours. He always knew the date correctly and he was always able to state when he was admitted and how long he had been in hospital. The use of numbers was unimpaired. He could carry out multiplication, such as 27×12 , without difficulty.

When asked to enumerate objects belonging to a certain category, such as capital cities, flowers, tools, furniture, birds, he usually succeeded, with some effort, in naming six to ten examples. His comments showed that in carrying out those tests he imagined a concrete situation, such as a map when enumerating the capital cities, a garden when naming flowers, a workshop when naming tools, etc. He had no difficulty in ordering objects according to certain qualities, such as material, shape, size, etc.

Orientation on his own body and on other persons' bodies was correct. No finger agnosia. Right-left orientation unimpaired.

In the Mill Hill Vocabulary Test he scored Grade 3 (average intelligence) in both definitions and synonyms. He obtained the same grade in Raven's Progressive Matrices. These results showed that neither his vocabulary nor his intelligence could have suffered severely and that his spatial orientation was unimpaired.

Speech.—The patient spoke fluently and with good articulation. In conversation his speech appeared entirely unimpaired and there was no noticeable poverty of nouns. His understanding of spoken language was perfect. Naming of objects, was, as a rule, correct. Only on rare occasions, especially when he was tired, he had difficulty in naming an object, e.g. in a series of tests he successively named handkerchief, wallet, ring and keys, pin, watch, neck-tie, matches, knife, inkwell, cigarette case, torch, radiator, comb, stamps, banknote correctly, but failed to name a desk-lamp ("they use it in offices, an electric....."). His ability to name colours was greatly impaired (see below).

Writing.—The patient was able to write letters and words dictated to him correctly and without difficulty. Writing of figures of any size and of decimals was unimpaired. Spelling was, as a rule, correct. However, when called to write long words he would sometimes repeat or leave out a letter or a syllable. He made similar mistakes in writing sentences, and he had a tendency to repeat letters, syllables and sometimes words for which his inability to read what he had written was obviously responsible. He used to write long letters home daily, describing his experiences in hospital. His letters showed a complete lack of punctuation, except for an occasional dash. He wrote capitals only at the beginning of his letters, which were not divided into paragraphs, and at the beginning of names of persons and animals. Apart from repetitions and occasional omissions the grammar was correct. The following lines taken from one of his letters shows the type of mistakes he made:

' Dear Flo-

"Just a few lines hoping you are keeping well trusting you are keeping well I am picking up very an improving and before long will be quite well again and Gill how is she getting on wait till I see her she is a good dog though she has a few dirty or should I say rude habits anyway look after her the weather here has been its old self again."

Copying of single letters, short words and numerals was correct. In copying long words he seemed to lose the thread easily and attempted in vain to read what he had written. The same applied to the copying of sentences. Short sentences consisting of words with one or two syllables were, as a rule, copied correctly. It was obvious that he wrote copying letter by letter and he often spelled aloud when copying. Although he would read most letters individually without difficulty he tended to copy "slavishly," e.g. he used to imitate unusual forms of the "s" written by the examiner and when copying a word written in capital letters or letters separated by a certain space he always followed the presented pattern meticulously.

Constructional ability was undisturbed. He could copy and, at request, lay without model patterns of matches, blocks or mosaics. His performance in the Kohs test was normal. He could draw geometrical figures and objects, such as a house, table, bird, hand, hammer with and without pattern. The perspective was correct and the professional touch was unmistakable.

Visual functions.—The patient was not aware of his hemianopia, although he sometimes knocked into objects which were in his right visual field. He was conscious of his alexia. He did not spontaneously refer to his disability of naming colours, but appreciated it when tested. When asked whether, apart from his alexia, there was anything wrong with his vision he used to complain about it being not clear, or blurred, or blotchy, and tended to blame this for his reading difficulties. He stated that for about a week following the sudden onset of his illness print, but not other objects, had appeared upside down. That symptom has passed completely and he did not remember that there had ever been a period when he saw letters sloping. Stationary or moving objects were recognized correctly. He enjoyed attending the cinema and could follow the film without difficulty. He never failed to describe and recognize pictures of objects and scenes correctly. He was able to pick out on request one of a large number of objects or pictures of objects spread before him. He also could recognize and name geometrical patterns. When shown a drawing of a face with a part missing he could fill it in. His visual imagery was unimpaired, but only as far as the shape of objects was concerned. His colour was imagery impaired (see below). There was no disturbance of spatial orientation. He found his way about the ward and in the hospital grounds without difficulty. He judged distances well and localized the position of objects in space correctly. The appreciation of perspective was unimpaired. The estimation of absolute and relative length and size was normal. He could connect two dots with a straight line and halve that line accurately, provided its right extremity did not fall into his right visual field. His topographical memory was good and he could draw a sketch of the ward with the correct position of his bed. He could read the time from a watch and he could set the hands of a watch to a given time.

Reading.—Single letters were, with some exceptions, recognized correctly. He often failed in recognizing letters of an angular shape, such as "K" (which he often read as "X"), or "N" or "X" (read as "K" or "I"), or "F" (often mistaken for "I"). Less frequently he mistook letters of circular shape for similar ones, such as "D" for "O," "P" for "Q," "Q" for "G," etc. Often the mistakes were of the mirror type, e.g. "b" was read as "d" and vice versa.

Drawing of cross hatching marks across the letters did not noticeably interfere with his performance. There was no indication that in reading he availed himself of other than visual aids, such as "writing movements" with his hands or head. If letters were put before him in wrong position in space he could read them as well as those presented in the correct position. The shorter the time of exposition, the more frequent were the mistakes, but he did not show any type of mistake on short exposition which did not appear otherwise.

Tachystoscopic examinations were carried out together with Dr. Pickford at the Psychological Department of Glasgow University. Single letters were exposed for one-fifth and one-tenth of a second. When coloured letters were exposed the performance did not suffer noticeably. However, when the patient was requested to read the letter and also to note the colour he made many more mistakes, and several times he failed to identify either when the time of exposure was below half a second.

When requested to pick out certain letters from a set of block letters or from a printed word he succeeded as a rule very well. Foreign letters, e.g. Greek, were recognized as such. In trying to identify wooden block letters stereognostically he made similar mistakes, as in reading visually.

When letters were written on his skin with his eyes closed he usually recognized the circular and bent shaped, such as B, R, S, D, O; sometimes he mistook them for similar ones, e.g. G for O, C for J, J for P. He could identify the simple angular letters L, T, V, drawn on his skin, but he failed with the more complicated ones, such as F, M, H.

Punctuation marks were not recognized at all, irrespective of whether they were presented within the context of sentences or isolated. He described them correctly as dots or geometrical figures, but they had no symbolic meaning for him

Reading of words.—The patient could read words only with the help of spelling. Having spelled the word aloud or to himself he would say it correctly. Only very short and common words consisting of not more than three letters he would occasionally read without spelling. In spelling longer or unusual words he lost the thread easily and he often gave up after the second or third letter. When trying to read a long word he would drop letters or read them incorrectly, even if they did not belong to the types of letters which he found it difficult to identify singly. When his attention was drawn to a mistake he sometimes rectified it.

Extracts from examination records :

"Extracts from examination feededs: "Evening" (handwritten): "e, v, e, n, i, n, g—evening." "Robert" (handwritten): "r, o, m, e, r, t." When the "m" was ques-tioned he said, "It's a 'b' I think." "Horse": "h, o, r, s, e—horse." "Tempest" (print): "t, e, m, n, e, s." Tompest "print): "t, e, m, n, e, s."

Tomato, sand, table, air, soil were spelled and read correctly. "Chestnut" (print): "You see n, u, t at the end. The first letter seems to bother me. I should know it; I have seen it. Is it not a 'w'? I might be a 'c'." He now spelt and read the word correctly.

He often declared spontaneously that he had mis-read a letter, but was not always able to choose the correct letter when it was said among others, e.g. "Hips" (print): "That's not 'n'? (Points to the first letter.) "n, i, p, but I think I got the first letter wrong." He did not pick the "H" when it was offered among other letters.

Sometimes he would grasp the approximate meaning of the word, or name the category to which it belonged, but he was unable to read it; for instance, "elder-berries": "Something belonging to fruit, but I cannot put it together."

In attempting to read short sentences he failed in the same way as he did in reading words, but he made spelling mistakes in reading short words which he would spell correctly when they were presented alone. For instance, when the sentence "It is rather cool to-night "was presented he read "It is r, a, t, p, e, r, c, o, a, t t, o, b, i."

When incorrectly spelt words which were not too long were presented he was, as a rule, able to rectify the mistake, e.g. in "Hause" he replaced the "a" by "o"; in windowe he struck out the "e."

When a letter or two were left out in the word he often succeeded in filling in XCIV. 4

1948]

the missing letters when told their number, e.g. "s-n" he completed to "sin"; "h—se" to "house"; "Mo-day" to "Monday," etc.

Reading of Arabic figures including decimals was unimpaired. He could read numbers such as 45,360,471 correctly without hesitation. He was quite unable to read Roman figures beyond IV. It was obvious from his mistakes that he set about reading them in the same way as Arabic figures, e.g. XXVII " Ten thousand, ten hundred and fifty-three."

Musical notes had always been unknown to him.

COLOUR VISION AND RECOGNITION OF COLOURS.

The patient very frequently failed in naming colours correctly, irrespective of the setting in which they were presented. This disability became more obvious when he was tired and when he had to describe multi-coloured objects or pictures. He was not colour-blind. He could distinguish yellow and blue, and green and red, but he frequently failed to name them correctly. In the Ishihara Test his responses were those of a subject with normal colour vision although he often failed in naming the colours of the dots constituting the numerals and the background ; e.g. in Plate 1 he read the number "12" consisting of bright red dots on a background of blue dots correctly, but he described the red dots as yellow and the blue ones as green. In Plate 2 he named the dark and bright green of the background correctly, but he described the various shades of red and yellow constituting the figure 8 as "pinkish." Sometimes the colour of the background seemed to influence the naming of the dots constituting the numerals. The absence of colour-blindness was confirmed by Dr. Pickford, who repeated the Ishihara Test and also carried out colorimetric examinations and the Holmgren Wool Test.

In naming colours he failed with some more often than with others, but there was none which he would invariably name correctly. He could always distinguish shades of brightness and never failed in recognizing the identity of colours in different objects. His performance varied a great deal. In the same series of tests he would give the right name of a colour, and five minutes later make gross mistakes when it was presented again. Even when he chose the correct name by the very often was uncertain and frequently qualified his description by the suffix "ish." White was described as "cream " or as "whitish," black as a rule as "blackish" and sometimes as "dark blue." Sometimes, when he mis-named a colour, certain features of the colour presented such as brightness, appeared in the wrong name. For instance, a bright green was called "pinkish." When told that he had mis-named a colour he would, as a rule, admit this, but only rarely was he able to rectify his mistake, occasionally he insisted on his description and admitted his error only when the colour, the name of which he had chosen by mistake, was immediately presented to him.

The patient frequently characterized a colour by relating it to an object, the colour of which was familiar to him. Brown or red objects were often called "stone coloured," grey objects "slate-coloured," etc. On other occasions the correct name was enforced by reference to concrete objects.

Extracts from examination records :

Extracts from examination records: Brown wallet: "Brownish as far as I can see." Blue stamp: "Blue." Black button: "Not quite black, but it is near it." Maroon-coloured wallet: "Reddish. It might be green." A week later he said, when shown the same article, "It is green, it might be blue." (Is this the colour of grass?). "There's a difference in shade, but if you take the average I would say yes." (This is red.) "No." Now a green square was shown to him and he named the colour correctly. When the wallet was presented again immediately afterwards he said "You are right. It flashed on me as green.

Red tie : "I can't remember. It doesn't seem to be red."

Blue ink : "Blue or green."

Buff coloured case book cover with dark red back and green string : He describes the buff as " yellow," the dark red as " reddish " and was unable to find a name for the green.

When asked to name the colours of the carpet in the office he described orange as "stone-colour" and blue as "green." He was able to match the green of the examiner's socks with an identical colour on the carpet.

50

1948]

Coloured pictures :---

Baby in green tub: Correct.

Blue shoe : "Green." Grey pail : "A slatish colour." Orange-red pail : "Salmon colour."

Red pail with blue and white stripes : The red is named correctly, the blue as "green," the white as "cream" and the black background as "blackish." Green book-cover: "Greenish." Orange book-cover: "That would baffle me." Yellow book-cover: "This is a colour I am beat on."

On a picture post-card he described the pink correctly, the green as "blue," the yellow as "straw colour," the blue of the sea as "green." The blue of the sky was very frequently described as "green," but he usually rectified this when a real green was presented immediately afterwards. On other occasions he would describe green and blue correctly. The description of mixed and intermediate colours he found most difficult. Purple was described as " reddish, or on the brown side."

Coloured skeins :---

White: "Cream" or "whitish" or "whitish cream."

Light brown : "A biscuit colour."

Yellow : Correct.

Light green he calls "light blue."

Light blue: "A light blue." He now spontaneously calls the light green "a yellowish colour."

Dark green : "Black with bluish colour."

Red and black were, on that occasion, described correctly.

The patient made similar mistakes in naming coloured squares, or the surfaces of the Kohs Blocks. The size of the coloured surfaces made no marked difference to the quality of his performances.

When requested to point out certain colours in a multi-coloured object or picture he did this, as a rule, correctly, but often failed in differentiating green and blue. He made the same mistake in Holmgren's wool sorting test. He was aware of this difficulty. In the sorting test he was rather uncertain in separating red and brown. Even if he did not sort colours incorrectly he tended, in that test, to leave out those which were not very similar to the one which he had chosen first, or which had been handed to him. He chose according to likeness rather than to category of colour. He was able, however, to sort colours according to their brightness.

When requested to pick out coloured skeins to match the colours of an object named by the examiner, he succeeded as a rule, but not without considerable hesitation. In this test, too, differentiation between green and blue was the most difficult. Which colour is a pillar box? "It would be a blue with a red—Post Office red." (Pt. picks a brown-red skein first, then a bright red.) Grass?: Pt. picks light green. Sky?: Picks dark green. Blood?: Correct. Tomato?: He hesitates, points to purple, then to orange, adding that the latter was more likely to be correct.

The patient was requested to enumerate objects of certain colours named by the examiner. His performance was poor. He mentioned trees and grass as green objects ; Post Office red, danger lights, certain tram-cars, as the only representatives of red objects. He found it equally difficult to name objects to match colours shown to him.

Reactions to wrong colours.-The patient was shown drawings from a children's painting book in which some sketches were painted by the examiner in wrong colours. He invariably rejected a wrong colour and characterized it as unreal, although he was, as a rule, unable to name it correctly. Red frog: "This is not the right colour." What is the right colour?: "It might be darkish brown." Green dog: "You never see them with that colour. It's blue, it's a toy colour." Black basket with purple fruit: "The basket might be right, but the colour of the fruit is too dark." He was shown drawings of three horses painted red, brown and green respectively. He pointed to the brown as the only correct one; about the others he said "Kiddian might do it like that" the others he said "Kiddies might do it like that."

Colour imagery .-- In describing colours of objects from memory the patient often failed. Not infrequently he appeared to describe a colour of the real object as it is seen in every day life rather than as it ought to be, e.g. when describing a goose as "greyish to whitish." On other occasions he failed absurdly. In his imagery, too, he showed a tendency to qualify his descriptions of colours. His imagery of forms of objects was intact.

Extract from examination record : Colour of blood—correct ; Grass—correct ; —aven—"I bit of grey in green"; Swan—" different colours. Some are green, others whitish"; Raspberry—" reddish"; Frog—" blue-grey or brown"; Goose—" greyish to whitish, according to age"; Sunflower—" yellow, and white in the centre." Cigar—correct ; Roses—" red, blue, green or pink." On the following day he described blood as " stone-coloured."

Progress notes.-The patient was discharged on 4.ix.46, but had to be re-admitted two months later owing to his paranoid behaviour at home. His condition did not change materially during the period of observation which had extended over ten months at the time of the conclusion of this article. However, certain changes in the relative degree of his symptoms took place. While the word blindness remained entirely unaltered, the colour agnosia showed a definite improvement. During the second half of the observation period mistakes in naming colours became less frequent and the patient did not use the qualifying suffix ' 'ish ''as often as previously. The most constant of his errors was still the confusion between green and blue. In recent months his colour imagery was noticeably improved, but he would still make characteristic mistakes occasionally, e.g. on 10.ii.46 he described from his memory the colour of a lion as "greyish" and that of a crow as "varying from black to grey," while the colours of ten other objects were named correctly in the same series of tests.

CASE 2.-Mr. A. J-, aged 68, retired bank clerk, was admitted as a voluntary patient on 3.ii.47. According to his relatives he had been in good health until December, 1946, when he became depressed and agitated, blamed himself for having led a wicked life and expressed fear of punishment. Sleep and appetite were poor. On admission his physical condition was found to be very satisfactory for his age. He did not show signs of arteriosclerosis. Blood-pressure 120/65. There were no signs or symptoms of an organic nervous disorder. The reflexes were normal. Patient was severely depressed and agitated; he expressed ideas of unworthiness, blamed himself for having masturbated in his younger years, and for not having offered to marry a girl whom he met thirty-five years ago. He complained about insomnia and frontal headaches. There were no signs of intellectual impairment. Orientation was intact. General knowledge and knowledge of current events were very good. He carried out arithmetical tests promptly and accurately and could recall a date and an address after half an hour. The diagnosis was agitated depression in senio. In view of the severe depression, electric convulsion treatment was instituted.

The patient had four treatments between 8 and 15 February, following which the depression subsided completely.

On 12 March the patient complained that he was unable to read, although he could recognize single letters. He said that his vision was blurred. The patient presented the syndrome right-sided homonomous hemianopia, word blindness and colour agnosia. He had no difficulty in naming objects, nor did he show any other features of dysphasia. Spontaneous writing of words and figures as well as writing on dictation were unimpaired. The patient used capital letters and punctuation marks correctly. When copying, he showed, like Case 1, a marked tendency to imitating peculiarities of the presented patterns. Constructional ability was unimpaired and he could draw simple geometrical figures correctly. Orientation in space and on his own body was unimpaired.

The patient was unaware of his hemianopia. He complained about his vision in the same way as Case 1. Acuity of vision was § (corr.).

Reading.—Single letters of any type were, as a rule, recognized correctly. In reading the patient did not avail himself of non-visual aids. Occasionally he failed in recognizing a letter of angular and rather complex pattern, such as "W" or " E." Tachystoscopic examinations were not carried out. In trying to recognize letters written on his skin, similar mistakes occurred as in visual reading. Arabic figures of any size, with and without decimals, were read correctly. The patient was able to carry out sizable multiplications (such as 244×7) and other oral arithmetical tests of a similar range accurately. On paper he could operate correctly with much higher figures. In trying to read Roman figures he failed in exactly the same way as Case I, being unable to recognize figures beyond IV. In attempting to read words he behaved in the same way as did Case I. He could recognize spelling mistakes and fill in missing letters if the word consisted of not more than three syllables.

Colour vision and recognition of colours.—The patient did not show signs of defective colour vision. In the Ishihara test his responses were normal, although he very often failed in naming the colours of the dots correctly. He made mistakes in naming and sorting colours. He was less inconsistent in his mistakes than Case I. Certain misnomers occurred more frequently than others. Green was hardly ever named correctly. The lighter shades of green were frequently called "yellow" or "yellowish" and sometimes "bluish." The darker shades were as a rule called "blue." Red was sometimes called "brown." A dark brown was often called "black" and sometimes "purple." Similar mistakes were made in describing colours of objects presented to him. Like Case I, he often qualified the colour named by appending the suffix "ish." When requested to sort skeins of coloured wool he failed in differentiating blue from green, and light green from yellow.

When tested for colour imagery he made mistakes almost identical with those of Case I. In a series of tests he named the colours of a raspberry, a cigar, grass, crow, correctly. A goose was described as "greyish," a lion as "doubtful greyish," an elephant as "blackish to brownish," a frog as "brownish," a daffodil as "vellowish."

When requested to enumerate objects of a certain colour from memory his performance was poor; for instance, when asked to enumerate red objects he named "red ink, red pencils, red dresses, red frocks, red paint." When pressed to mention objects which were red by nature he only named poppies. He found it generally difficult to list objects belonging to a certain category and, like Case I, had to imagine real situations when trying to do so.

Progress notes.—The patient presented the syndrome described above for only two weeks. On 26 March he complained about being confused. Clinical examination revealed a severe nominal aphasia with marked paraphasias. Understanding of spoken language was slightly impaired. In writing he showed the same type of paraphasia as in speaking. He was unable to read and to name colours. As a rule he did not even attempt to do so. He was very distressed about his disabilities and gave up immediately he found a task difficult to perform. His condition deteriorated rapidly. On 25 March he was unable to express himself and the only residue of his speech was the recurrent utterance, "I shall be all right," with which he responded to questions put to him. A right-sided hemiparesis developed, more marked on the arm than on the leg. On 27 March the right extremities were completely paralysed and the patient gradually lapsed into unconsciousness. He died on 29.iii.47. Permission for post-mortem examination was not granted.

COMMENT.

The patients presented the syndrome of alexia with colour agnosia, which was associated with a right-sided hemianopia. Only a small number of cases have been reported in which this syndrome was not complicated by marked aphasia and disorientation in space. Although typical in many respects, the two cases showed some uncommon features which deserve comment. The alexia could be described as word-blindness with partial letter-blindness. In the majority of cases of word-blindness the recognition of letters is not entirely unimpaired, but little is known about the type of mistakes which word-blind patients tend to make in reading letters. The partial letter-blindness in the two cases presented above was typical of the visual alexia (Pötzl, 1928) in cases with occipital lesions. In those cases the errors in the recognition of letters are visually conditioned, i.e. they can be explained by incomplete visual comprehension of the shapes of the letters. This type of mistake distinguishes the visual (occipital) alexia from the parietal or temporal in which the paraphasic element can be traced.

It is worthy of interest that the type of letters which Case I frequently, and Case 2 sometimes, failed to recognize was the same which patients with impairment of epicritic cutaneous sensibility find it difficult to identify when they are written on the skin. The present writer (1927), in a study on the tactile perception of movement on the body surface, has found that patients with disturbance of superficial sensation fail in comprehending angular geometrical figures written on their skin much earlier than in the recognition of circular ones. The similarity between this type of impairment of tactile perception and the errors of partial letter-blindness is of interest as it suggests a more intimate relationship between perception and recognition than is often assumed.

Word-blindness lends itself even more than other disabilities of higher cerebral functions to interpretation in terms of Gestalt psychology. There can be no doubt that the word-blind patient has lost the ability to conceive the word as a unit and that he tries to overcome that disability by proceeding piecemeal, reading letter by letter. His mistakes in reading letters can also be interpreted as the expression of an inability to comprehend certain configurations as wholes. It is not a coincidence that the patients found it most difficult to identify letters of angular shapes which are more complex than the circular ones, and, as configurations, lack completeness. However, it is well to remember that the ability to comprehend organized wholes is not a function which can be isolated from the process of perception. This has again been stressed recently by Goldstein (1943), who introduced Gestalt principles into the analysis of cerebral symptoms.

These patients, like other cases of "pure word-blindness," read words by spelling them. By attempting to read sentences or long words by that method they failed badly and appeared almost completely letter-blind. This is an example of a breakdown of a mechanism of compensation by the help of which the patient attempts to circumvent his disability. The difficulty of the task produced what Goldstein called a catastrophic situation.

Writing is, as a rule, unimpaired in cases of word-blindness. The writing in Case I showed certain abnormal features. They were, however, not typical of any known form of agraphia, but could be attributed to the alexia. The occasional omissions and repetitions of letters and syllables can be explained by the patient's inability to read his own writing. The failure to use capital letters at the beginning of sentences and the absence of punctuation were probably due to the same cause. The patient wrote as he spoke. The use of capital letters at the beginning of sentences and the application of punctuation marks are late acquisitions in the learning of written language. In wordblindness the visual comprehension of sentences as organized wholes is lost and this is obviously the cause of this patient's inability to use the demarcating symbols. However, it remains to explain why a word-blind person, even if he be unable to recognize punctuation marks, should not apply them in writing in the same way as he writes words that he cannot read. Possibly the length of the period over which the alexia has been in existence, i.e. the time of the disuse of punctuation marks, is the decisive factor. The patient, while trying to overcome his word-blindness by spelling single words, had long given up any attempt to read sentences, and punctuation marks have ceased to be of any use to him from the moment he has become alectic. Disuse of that group of symbols, which are the latest acquisitions in the process of learning to write, and the use of which remains precarious even in many normal subjects, may be responsible for the inability of some word-blind patients to recognize and to use punctuation marks. In Case 2, where the alexia existed only over a short period, the use of, and the ability to read punctuation marks were intact. It must also be taken into consideration that punctuation marks form a group of symbols in the acquisition of which acoustic aid, which plays such a great part in learning to read and to write, is almost negligible. This is probably one of the reasons why the acquisition of their correct use is difficult and why this ability is particularly vulnerable in cases of reading and writing disorders. The present author is not aware of any special observations on the use of punctuation in congenital word-blindness reported in the literature. It should be expected that in those cases the acquisition of the correct use of punctuation would be found to be more difficult than normally.

An interesting feature in both cases was the intactness of the ability to read Arabic numerals of any size, while the ability to read Roman figures beyond IV had been lost. One of Pötzl's (1928) cases showed exactly the same symptom. The patients described here attempted to read Roman figures in the same way as Arabic numerals. The only explanation for this discrepancy is again late acquisition and disuse.

The patients' ability to name colours was impaired, although they were not colour-blind in the usual meaning of the term. Wilbrandt (1887) described that symptom as amnesic colour-blindness and regarded it as an aphasic disorder. Lewandowsky (1908) attributed it to a dissociation of the conception of colours from that of objects. Sittig (1921) was the first to note that those patients often failed in sorting colours, which proved that the symptoms could not be of the nature of an amnesic aphasia for colour names. In his view that disability was the result of a combination of visual and aphasic disorders, the difficulty in sorting colours being due to the visual-agnostic component. Gelb and Goldstein (1918), rejecting all previous theories, expressed the opinion that the disability was the result of an impairment of " categorical behaviour." Patients in whom this basic function is affected behave and think in a primitive manner, i.e. according to the concrete situations in which they find themselves. They tend to name and sort objects according to their likeness, and to the way in which they fit into concrete situations, rather than as representatives of certain categories. The authors found the same impairment of categorical thinking in a variety of symptoms due to cortical lesions, particularly in amnesic (nominal) aphasia. This is not the place to enter into a discussion of this theory, the general applicability of which has been widely debated (Mayer-Gross and Guttmann, 1936, a.o.). The cases presented above showed

1948]

a variety of features similar to those on which Gelb and Goldstein's conceptions were based. In sorting colours the patients tended to choose more according to likeness than to categories. When enumerating objects belonging to the category they imagined concrete situations. In naming individual colours Case I often referred to concrete objects of the same colour. However, it is open to doubt whether the impairment of categorical behaviour which can be demonstrated in certain cases of colour agnosia is really the basic disorder underlying that disability, or if it is not, as Pötzl (1928) suggested, secondary to a disturbance of visual functions which still requires elucidation. The cases reported here showed certain features which it would be difficult to fit into the above theory. Although they were not colour-blind in the narrow sense, their references to their visual impressions in general, and the qualifying terms (" bluish, yellowish," etc.) which they used in describing colours in particular, suggested that colour perception was not quite intact. It is, however, still impossible to define in what way it deviated from normal. It is of interest that there are certain features which colour agnosia shares with defective colour vision. The differentiation between green and blue which is invariably impaired in colour agnosia is known to be difficult in anomalous trichomatic subjects whose colour vision is constitutionally inferior. Some errors which the patients presented above and others reported in the literature made in naming colours were reminiscent of the errors of colour defective persons, who, as Wright (1946) has pointed out, describe at various times the same stimulus by several different names, or call different stimuli by the same name. White and black are, as a rule, recognized in colour agnosia, as well as in defective colour vision. Their description as " whitish " or " creamish " and " blackish " suggests some abnormality of perception. Pötzl (1928) has assumed that lesions of the cortical fields adjoining the area striata tend to modify the functions of the latter and the observations reported above lend weight to that theory. One of the results of that modifying influence seems to be an interference with the established correlations which exist between colours and their names.

The two cases demonstrate that the difficulty in naming colours is not part of an aphasic disorder. Case 1 showed only a very slight disability in naming objects at a time when the naming of colours was gravely impaired, Case 2 none at all. The cases described by Brain (1941) and A. Adler (1944) in which colour agnosia was observed without any signs of aphasia demonstrate equally clearly that the symptom is independent of a disorder of speech. Such observations argue against Nielsen's (1947) contention that colour agnosia is an aphasic disorder, the impairment being one of symbolization. Pötzl (1928), in describing the symptom as aphasic, made it clear that he regarded it as a basically agnostic disorder resulting in a difficulty of naming.

It is noteworthy that in Case I the colour agnosia improved while the wordblindness remained unaltered. The improvement of the former might have been due to the development of mechanisms of compensation in the brain. Frequent testing probably stimulated such a process. The fact that the wordblindness did not show the same improvement suggests a functional independence of the two symptoms of each other. The regular combination of visual alexia with colour agnosia cannot be explained on psychological or physiological grounds. It is obviously due to an intimate anatomical relationship of the cerebral structures, the lesions of which are responsible for the symptoms. Congenital word-blindness, which has a different pathology, is not associated with a disability of naming colours.

The clinical observations presented cannot contribute directly to the pathology of the syndrome which was obviously the result of a localized vascular lesion in both cases. Since Déjerine (1892), pure word-blindness has been associated with lesions of the left gyrus angularis, of its connections with the visual area and of the splenium corporis callosi. More recently cases have been reported in which the lesions were localized in the lingual and fusiform gyri of the left hemisphere and the validity of Déjerine's localization has been questioned, especially for cases in which word-blindness was not associated with agraphia and disorientation in space (Bouman, 1928; Pötzl, 1928; Brouwer, 1932; Kleist, 1934; Weisenburg and McBride, 1935). It is worthy of note that in the case reported by Hinshelwood, MacPhail and Ferguson (1904) there was extensive destruction of the left fusiform and lingual gyri, while the angular gyrus was not directly affected. For this reason Pötzl classified the word-blindness of the type described in this article as occipital alexia. At the time when the pathology of pure word-blindness was first studied much less was known about the sympatomatology of angular lesions than is known to-day. It is therefore of interest to note that pure word-blindness has never been found associated with Gerstmann's syndrome which results from lesions of the left angular gyrus. This argues, from the clinical point of view, against the assumption that damage to that area or its immediate neighbourhood is responsible for pure word-blindness.

SUMMARY.

Two cases have been reported which presented the syndrome of visual alexia with colour agnosia in unusual purity. The alexia was of the nature of a wordblindness with partial letter-blindness. The types of errors which the patients made in reading letters have been analysed. Certain abnormal features in the writing of Case I have been shown to be related to his alexia. The patient did not write capital letters at the beginning of sentences and he was unable to read punctuation marks and to apply them in writing. The significance of those disabilities has been considered.

The various theories on colour agnosia have been discussed. Certain features which colour agnosia has in common with defective colour vision have been pointed out. During the period of observation the colour agnosia in Case I improved considerably while the alexia remained unaltered.

The cerebral localization of the lesion causing the syndrome has been discussed.

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1948]

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58