PROSOPAGNOSIA

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"And what is the nature of this knowledge or recollection? I mean to ask, Whether a person, who having seen or heard or in any way perceived anything, knows not only that, but has a conception of something else which is the subject, not of the same but of some other kind of knowledge, may not be fairly said to recollect that of which he has the conception?"

"And when the recollection is derived from like things, then another consideration is sure to arise, which is, Whether the likeness in any degree falls short or not of that which is recollected?" "The Philosophy of Plato" Phaedo (the Jowett translation).

Does visual agnosia exist in a partial or isolated form, in which certain qualities only are affected, as opposed to generalized visual agnosia? Many workers cast doubt on this concept, maintaining that partial visual agnosia is no more than a combination of defects in vision, memory, and orientation, appearing together.

The clinical elucidation of partial visual agnosia is likely to be affected by the patient's intellectual capacity, his mental state at the time of examination, and his ability to cooperate without being influenced either by fatigue or by suggestion on the part of the examiner.

Critchley (1953), one of the leading workers in this field, states critically that "it would be dangerous to claim that a particular visual agnostic defect can occur in isolation".

There are some who consider visual agnosia as a patho-physiological optical problem derived from an organic lesion in the visual pathways, more especially the visual centres. Others maintain that visual agnosia is merely a mental disturbance which prevents the patient from evaluating properly and integrating into the general mental visual picture the various optical impressions, thus affecting the level of awareness.

Between these two extreme views is the more moderate one which holds that a physiological optical disturbance is associated with mental upset. Bay (1952, 1953) subscribes to this view, saying that visual agnosia is not in itself a specific cerebral disturbance but one where visual pathway defects are allied to mental disturbances. (Jung, 1954, who appraised Bay's book Agnosia and Lability of Function stated "the author wrote a book about agnosia in order to show that agnosia is a scientific phantom"). Bay described the disorders as fine defects in the visual system in the form of disturbances in sensation time, in adaptation time, in visual acuity, and in brightness discrimination.

Ettlinger (1956) rejected Bay's contentions. After analysing 30 cases of head injury, he showed that some patients had neither field nor perceptual defects, others had field but not perceptual defects, and only in eight of the 30 patients were field and perceptual defects found together. It is true that visual agnosia is frequently associated with homonymous hemianopia, but despite this there are cases of hemianopia without gnostic defects.

There is a special type of visual agnosia called prosopagnosia which means inability to recognize and identify known faces. The number of cases in this condition described is small. Hécaen, Angelergues, Bernhardt, and Chiarelli (1957) collected 25 cases described in the literature since 1932.

In addition to the prosopagnosia, all the patients except two showed various psychosensory disturbances as well, such as unilateral agnosia of space, constructive apraxia, loss of topographical memory of alexia, acalculia, and in a few, optical disorders.

The case history that follows is unusual in that we were able to follow up the dynamic development of the symptoms of which prosopagnosia was a marked feature. In the early stages of the illness, topographical disorientation, slight simultanagnosia and achromatopsia were also found.

Case Report

A.G., a man, aged 64, was formerly a merchant, and now is a clerk. He has two grown up children. He has completed high school, speaks, reads, and writes five languages, and is right handed. He was well till five years ago, when he woke up suddenly from his sleep one night because of severe dizziness. He got out of bed and immediately fell to the floor, where he remained unconscious for some minutes. The following morning he felt completely well and carried on with his usual work.
further details are available concerning this episode. His family did not notice any particular change in him. Two years later, when aged 61, he was again awakened at night by severe dizziness and a feeling of pressure in the head. He lost consciousness for some minutes and on recovering, suffered from headache for the next 24 hours. He felt quite well two days later and returned to work: there were no paralyses, no speech disturbance, no visual defect, and no loss of orientation.

The present illness began five days before his admission to hospital. The patient woke up with severe frontal headache which lessened somewhat during the next three hours. He had an appointment at noon of the same day and caught a bus in order to get there but on the way his headache increased and he decided to go home instead. He alighted from his bus as he had to change his route and on his way to another bus stop, on a road well known to him, he felt all of a sudden that the whole area appeared strange. He knew where he was and could not account for it. He managed to cross the road—he knew he had to do that—and boarded the correct bus in order to reach home. He paid his fare and received a ticket. The route, obviously well known to him, appeared strange and for a time he thought he had caught the wrong bus. He became anxious, gave the driver his address and asked him to see that he reached home should anything happen to him meanwhile. When he thought that he had reached his stop he alighted, but not because he recognized the surroundings, which appeared unfamiliar to him, but because he estimated that sufficient time had elapsed in order to arrive at it. He remembered these details clearly. He also remembered that while on the bus he found it difficult to distinguish men from women passengers. After getting off the bus, “I stood in the street and asked passers by how to get to my home. I gave them my address and carried on in the direction indicated.” After walking for about five minutes, the approximate time needed to reach his home, he could not find it. He again turned to passers by who told him to cross the road as the required number was on the opposite side of the street. “I eventually reached home, and though I knew it was my home, it seemed strange to me.” He opened the door, entered the flat which he knew was his, but the room, furniture, and pictures appeared different.

“The first thing I did was to go to the bathroom to wash. I looked in the mirror and saw a strange face. I put a cold compress on my forehead and as far as I remember, lay down for about three hours. Perhaps I slept. When I woke up I walked about the flat and noticed that the landscapes on the wall seemed covered by mist. I didn’t pay any attention to the portraits.”

The patient’s wife was overseas at this particular time and his son lived elsewhere. He decided to go to the post office in order to telegraph his family.

“The road seemed completely strange to me though I knew it well. I asked directions from people in the street a number of times but felt I couldn’t carry on. I turned back to see my doctor who lives opposite me and who has known me for years. I saw in my mind’s eye the doctor’s house, the steps leading to his flat, but I couldn’t find it. I wandered up and down the stairs in a few houses till I saw the doctor’s plate on one of the doors. A woman opened the door but I didn’t know who she was till I heard her say ‘Come in Mr. G.’. I then realized that she was the doctor’s wife. The doctor examined me and advised me to go to hospital. I remember that he measured my blood pressure and gave me a prescription. As I knew I was in Dr. A’s surgery I didn’t pay any attention to his face and didn’t notice anything wrong in particular. It was already dark and I reached home with difficulty.”

The events of the next two days are not clear. The patient said he had headaches and noticed for the first time that he did not see well with his left eye. He remembered reading the paper, but with difficulty. He understood what he read and could recall the items later.

The next day his son came to visit him. The patient did not recognize him and only realized who it was when he spoke. He returned to Dr. A. together with his son, and in his own words “I wouldn’t have recognized the doctor if I hadn’t known where I was going”.

He was referred to an eye specialist and on the way to the latter’s consulting rooms found that the streets and buildings appeared strange and unfamiliar. He remembered being examined by the eye specialist for more than half an hour, an instrument was used (apparently a perimeter), and he was asked to read. He had no difficulty in recalling these details but could not remember the doctor’s face nor anything else about him that would help in describing him, such as his height, colour of hair, or facial features.

Till his admission to hospital 24 hours later he remained in the care of his son, which was a great relief to him as he was still disoriented. Though on the whole his flat still appeared unfamiliar to him the colour of the walls and the pictures on the walls partially regained their former clarity.

The drive to the hospital, along a route well known to him, appeared strange. “I knew from memory what to expect along the way—the various turnings, buildings, and landmarks, but when I passed these points they seemed different to what I had imagined.”

The patient was admitted during the afternoon and was examined within a few hours. He was orientated for time and place. He complained of headache and difficulty in seeing objects, especially those to his left. In particular, he found difficulty in appreciating the facial features and expressions of people he had known for some time. The same thing was noticed in the ward. The patient was in constant contact with three doctors but for the first few days he was unable to distinguish one from the other. Later he reported that he had found a way out of the difficulty. “One is very tall, another has a moustache and wears glasses, and the third has neither glasses nor a moustache.” He learnt to distinguish the doctors by their voices. “As soon as I hear your voice I know who you are and can tell you apart from your colleagues.” During his first few days in hospital it took some minutes to recognize the various voices. He was
also unable to recognize the nurses. "They all looked alike" and usually he addressed them wrongly. He learnt to overcome this problem by associating a particular nurse with her duty shift. Six days after his admission he accosted a visiting doctor in his ward and spoke to him as if he were his own doctor. When asked about this later he said "but they both wear glasses".

His son visited him every day and he only recognized him when he sat down next to him and spoke, but after a few days he learnt to recognize him by his walk as well. With regard to other relations, he was able to distinguish them after some minutes of conversation. The patient reported that frequently during conversation, a face familiar to him suddenly lost its clarity.

"If three people I know sit opposite me, after talking for a few minutes their faces very often become blurred and lose their individual characteristics and I no longer know to whom I am talking. I have to concentrate very hard in order to tell them apart and this gives me a headache."

During the early part of the patient's period in hospital he was referred back to the eye specialist who had examined him originally. The doctor remarked that he knew him as he had examined him a few days previously.

The patient replied "I beg your pardon, but you are mistaken. I'm seeing you for the first time and you have never examined me before". During the examination the patient exclaimed "I am very sorry. Yes, yes, I know you now, you did examine me before because that same instrument (perimeter) is in exactly the same position on your table as it was before, the last time I was here". During this period there was no spatial disorientation. The patient knew the layout of the beds, the rooms, lavatory, dining-room, doctors' room, and hospital grounds. In walking about the grounds he could describe the various flowers he saw, could differentiate their colours, but regarding the sun and heavens, he repeated time and again that they were blurred.

Neurological Examination.—A full examination was made on August 6, 1957.


Ears.—Vestibular and cochlear function normal.

Limbs.—Power, sensation, and reflexes were normal, but ataxia was detected. Abdominal reflexes were normal, and pathological reflexes were noted. Gait was normal.

Other clinical findings were: Blood pressure 160/95 mm. Hg, E.S.R. 25/50, blood picture normal. The heart showed left ventricular hypertrophy radiologically. An electrocardiogram was normal, and the lungs appeared normal.

An electroencephalogram was within normal limits. Lumbar puncture showed cells 4/3, protein 51 mg. per 100 ml. mastix negative.

Wassermann and Kahn reactions were negative.

The basal metabolic rate was —2.

Patient's Behaviour in Hospital.—He felt well and was emotionally stable. He had normal contact with his surroundings and showed the same interest in hospital life as other patients. His only and repeated complaints were severe headaches of varying intensity and inability to recognize faces whether those of other patients or of visitors. He tried to distinguish people by other characteristics such as walk, dress, hair style, and so on. He had no insight into his condition.

Psychosensory Examination.—The delivery, rate, and intonation of speech was normal. Articles shown him were named correctly and described in detail without error. Size of object or distance from the eye made no difference.

All commands were carried out correctly and without delay.

The patient read without difficulty printed and written texts both in Hebrew and Latin script. His diction and locution were satisfactory. He could tell the time and move the watch to any desired hour.

He wrote easily and correctly both in Hebrew (from right to left) and Latin (left to right) scripts. There was no difference between spontaneous writing and writing to dictation.

When asked to cross out certain letters in the script given him he did so without difficulty and without making any mistakes. In sentences where words or letters were missing, he completed the text correctly.

Calculations, both easy and hard, were performed without difficulty whether by memory or on paper.

When adding, the figures were aligned correctly beside one another.

Simple geometric patterns such as a square, oblong, triangle, or star were drawn properly on the blackboard.

Faces were drawn in detail but in rather primitive fashion. However, the patient admitted to never being able to draw faces well.

Animal pictures were distinguished correctly even if part of the picture was covered.

Pictures showing national costumes, scenes, maps, emblems, stamps, road signs, and so on, were correctly interpreted.

When shown pictures depicting action, for example, a policeman directing traffic, the patient was unable to indicate the meaning or intent of the picture. He saw a number of cars, recognized the policeman, mentioned a child crossing the road, but could not grasp the theme intended. When pressed, he said "something is obviously happening here but I don't know what". Similar difficulties were encountered when shown other pictures depicting action or motivation, or others with allegorical or humorous indications. As time went on his interpretation of pictures improved, but the time taken for this was not in proportion to the difficulties of the test nor were his answers given with certainty. Sometimes he was non-committal and gave more than one answer to the same problem. Four weeks after admission, picture interpretation became normal in all respects.

In the early days of his stay in hospital he went to the cinema, but left the hall after a few minutes. "I saw people but couldn't understand what was going on" he said. One month later he had no difficulties whatsoever when watching the screen.
No disturbances in praxis were noted at any time. The disorders in topographical memory passed off gradually within a few days of his admission to hospital. While in hospital no disturbance in colour sense was noted, but apparently it did exist before that time, for “everything I saw appeared grey and the sky always seemed cloudy. The sun looks dirty to me”.

Revisualization of colours did not suffer at any time nor was there any disturbance in body schema awareness. He differentiated between right and left and between his own and other bodies. Finger gnosia was normal and there was no disturbance in time estimation.

The main defect was inability to recognize faces, whether living or from pictures. He did not recognize pictures of famous personalities that he would be expected to know such as portraits of Hitler, Stalin, Khrushchev, or Ben Gurion. If given a picture upside down he corrected the position but still was unable to say who it was. Caricatures also had no meaning for him. There was no difference whether the portrait was full face or profile. When he was shown a picture of Kosciuszko, a Polish national hero, which he could not but know, he recognized it immediately and said “every child knows this—he has such a typical cap that one can recognize him immediately”.

When asked to describe certain parts of the face, such as the mouth or ears, he did so without difficulty. Revisualization of people known to him was normal; the same applied to buildings or objects which he had previously seen.

In hospital he read the paper, took an interest in daily events, and showed no objective evidence of mental deterioration.

Neurological examination 10 days after his admission showed a field defect now limited to the upper left quadrants, a normal E.E.G., and a blood pressure of 170/110 mm. Hg. The patient refused pneumoencephalography and was subsequently discharged remaining under weekly supervision in the out-patient department for the next 11 months.

According to his wife, the patient returned to his former post and carried on without encountering any particular difficulties. He travelled about alone and was completely orientated. The difficulty in recognizing faces persisted but to a lesser degree and he still found it hard at times to tell who was greeting him in the street. Three months after the onset of the illness a neighbour whom he had known for years asked him a question. He replied that he did not know her but as soon as he recognized her voice he begged her pardon and explained that he was not wearing his glasses and could not see clearly in the twilight.
An interesting episode occurred after he had left hospital, when the doctor who had treated him in the neurological ward met him in the street. The doctor walked alongside him for nearly five minutes. The patient looked at him frequently but did not recognize him. Finally the doctor asked him how he was. The patient stopped and said "Your face is so familiar" and paused. Then he continued, "I recognize you now—in hospital you don't wear a hat."

His wife stated that when they were expecting guests, he always asked beforehand who they were. Four months after the onset of the illness he still had difficulty in recognizing faces. He said, "I see people and not individuals, but as soon as they speak or make some movement I know who's who." As time passed on and with repeated examinations this difficulty in recognizing faces was the only defect found persisting.

Summary of Clinical Findings

1. In the early stages of the illness prosopagnosia was associated with topographical disorientation, simultanagnosia, and achromatopsia, all of which passed off gradually. Six months after the onset of the illness the prosopagnosia had declined to a great extent.

2. No intellectual deterioration was noted at any time nor was any change seen in his critical faculties or memory.

3. The only neurological finding early on was homonymous hemianopia which soon shrank to a left upper quadrant defect. The last examination performed on September 9, 1958, disclosed that the hemianopic defect had shrunk still further (Fig. 1d). The boundaries of the peripheral fields are now normal. There is a dense left upper quadrant paracentral scotoma of a type which indicates a lesion of the lower lip of the calcarine cortex.

Discussion

From a review of the literature it appears that pure prosopagnosia is rarely found as an isolated phenomenon. It is usually associated with other findings, such as object agnosia, aletic and apractic disturbances, and optic disorders. Our case resembles that described by Pallis in 1955. The latter's patient suffered from mitral stenosis and developed an embolus in the right posterior cerebral artery, which was later confirmed by vertebral arteriography. This patient showed prosopagnosia and achromatopsia. Topographical memory and facial revisualization were preserved as in our case. An additional finding was a noncongruous homonymous sector defect in the left upper quadrant and absolute scotoma in each field on both sides of the vertical meridian.

There is also some similarity to Hoff and Pötzl's patient, described in 1937, who showed prosopagnosia together with colour disturbance, and to a lesser extent object agnosia and spatial disorientation.

Klein and Stack (1953) reported on a patient, who, after a vascular lesion, developed prosopagnosia with inability to interpret pictures, disturbance in topographical memory, inability to identify colours, and also had a left upper quadrantic field defect.

In the same year Alajouanine and his co-workers published a report of progressive cerebral atrophy in a man of 56, with transient left hemianopsia, who three and one half months later developed prosopagnosia (Alajouanine, Lhermitte, Sabouraud, and de Ribeaucourt (1953). The patient found it difficult to distinguish male from female faces and to estimate age by observing a face. In addition this patient was unable to recognize colours and suffered from simultanagnosia.

Between the years 1947 and 1955 Faust collected seven cases of cerebral trauma which were clinically similar to the picture of prosopagnosia described above, six of which also showed alexial disturbances (Faust, 1951, 1955).

Hécaen's two patients (1956) were operated on by Penfield in order to remove epileptogenic foci in the right parieto-occipital area. Post-operative prosopagnosia developed, together with other gnostic and apractic disturbances (Hécaen, Penfield, Bertrand, and Malmo, 1956).

In other fairly similar cases reported the appreciation of prosopagnosia was difficult owing to the presence of other signs such as optic, memory, visual memory disturbances, and general intellectual deterioration. Heidenhain's (1927) cases belong to this group. One of his patients suffered from malacia of both occipital lobes following embolic infarction could recognize neither his wife's nor his sister's face and could only differentiate them by their voices. But this patient also suffered from alexia, complete colour blindness, and paracentral scotoma in both upper field quadrants.

Jossmann's patient (1929) also showed a rich pathology. Following encephalitis this patient was unconscious for some days. On recovery she was unable to recognize faces. She was also unable to differentiate animals at rest but could do so when they moved. In addition to this prosopagnosia she also showed object agnosia, transient alexia, disturbance in perspective vision, and simultanagnosia.

Perhaps Hoff and Pötzl's case (1937) showed the most severe prosopagnosia. The patient did not recognize his own face in the mirror, suffered from severe colour disturbance, from spatial disorientation, disturbance in re-visualization, and optical reception. The pathological lesions in this case were "apoplectic" foci in both temporo-occipital lobes.

In 1947, Bodamer published three cases of prosopagnosia following traumatic lesions. It was Bodamer who introduced the term prosopagnosia.
In a detailed work he tried to explain this rare disturbance but in his cases too the prosopagnosia was superimposed on severe brain damage. In the first case, Uffz. S., a penetrating bullet caused a depressed fracture in the lower part of the left parietal bone and in the right occipital bone. The patient was blind for some weeks following the injury, and on partial recovery remained with a left upper quadrantic field defect, with metamorphopsia, object agnosia, disturbances in orientation, and simultaneous agnosia.

The second case was more severe. After a serious head injury Lt. H. A. lost consciousness. Examination later revealed left hemiparesis, reduced sensation over the left half of the body, and concentric bilateral hemianopsia. Psychosensory disturbances included a left-right orientation disorder, optical disturbances, difficulty in recognizing colours, and simultaneous agnosia. Concentration was also affected.

The third case, O. Geffr. B., had a fracture in the left occipital bone with severe brain damage. He showed signs of metamorphopsia, parts of pictures were blurred and distorted, and right hemianopsia and dyslexia were also found.

These three cases form the basis on which Bodamer tried to build his thesis. According to this author, prosopagnosia represents a severe regression in the gnostic sphere affecting the old and deep levels of perception. He states that the infant recognizes facial expression before recognizing the object. This is opposed to the view that maintains that the infant recognizes the object first. Recognition of the object is associated with high level optico-gnostic function. Bodamer sees in facial recognition an Urphenomenon, and, according to Monokaw's theory, a time recorded memory, Chronogene Engramierung. Following Kaila who examined the effects of faces on children’s behaviour, he is of the opinion that the area around the eyes is the part which engages the infant’s attention first. Bodamer calls this eye area the “ocula”. In cases of prosopagnosia he thinks that the patient’s attention is concentrated on these parts and cannot be withdrawn.

Bodamer’s patient, H.A., said “apart from the eyes, the rest of the face is blurred. I don’t see the facial characteristics of living people. The most obvious thing to me is the eye”. Bodamer’s theory, which is basically similar to Pötzl’s, is unacceptable as it stands despite its originality and phylogenetic approach. All his cases suffered severe brain injury which probably affected brain function as a whole, and because of this detailed analysis is extremely difficult. We paid special attention to this point when examining our patient, who was able to define every part of the face, whether living or from a photograph. As he improved his facial recognition became more prompt, and at no time did we consider visual function to be disturbed.

Bodamer’s basic tenet that facial recognition is associated with the Urphenomenon is also open to criticism. It seems clear that the developing infant recognizes the mother’s face as an object and not as an expression because first optical impressions are directed towards the woman’s face attending him, holding him, or suckling him. At the same time the rest of the woman’s body is not in the infant’s field of vision and the direct facial stimuli will crowd out any lesser peripheral ones.

Recognition of facial expression depends on a number of stimuli, visual, auditory, and tactile. The child learns from experience; in the early stages he is attracted by toys which have no particular shape but which are prettily coloured or produce tones. At a later stage he plays with dolls or animals, and still later the faces of these dolls or animals take on a meaning.

As opposed to the inability to recognize facial expressions whether living or still life, our patient’s ability to visualize a face he had known previously was intact. He was also able to recall facial changes of people he knew with the passage of years. The same facility applied to places, and in common with Pallis’ patient, he said, “In my mind’s eye I know exactly where places are, what they look like and the squares and streets which surround them.” Our patient did not show any signs of Charcot-Wilbrand’s syndrome.

In seeking for yet another explanation, Goldstein’s theory is also unsatisfactory, that any defined lesion in the brain creates a “disturbance of integrity” or general disturbance of behaviour. There was no personality disturbance in our patient nor did his behaviour change after his illness began. During the nine months he remained under observation no change in his character was noted. There were no signs of catastrophic reaction, of perseveration, nor of a depressive reaction (anosodiaphoria) to his illness (Critchley, 1957).

The patient did not try to suppress knowledge of his illness. There were none of the characteristic signs described by Goldstein such as inability to distinguish between figure and background. The opposite is true: he tried everything in order to overcome his disabilities. Only one specific quality suffered and that was the expression which appeared separated from the object.

In 1949, Faust suggested the term pseudo-agnosia to explain the cause of prosopagnosia together with lowering of mental activity and mild visual disturbances. Here apparently the visual stimuli suffered, but again pseudo-agnosia does not apply in our case, where intellectual function particularly was
unaffected. It is difficult to consider prosopagnosia as a sign of Funktionswandel. It is not easy to accept reduction or lability of visual function lasting for such a long time without other aspects of perception being affected.

There is no doubt that the degree of attention varies in patients with gnocistic disturbances, due to various factors, mostly affective ones. Attention may become extremely limited. But despite this one cannot look upon these perceptual variations as explanations for prosopagnosia itself.

In the early stages of our patient’s illness he was unable to grasp the meaning or intent of a picture though he could describe and evaluate each individual aspect of it clearly. This lack of interpretation could not be ascribed to lack of attention or fatigue. There are also no grounds for supposing that there was visual or mental fixation on a particular part of the picture. Similar to his inability to see pictures “alive” was his inability to obtain a living or vital impression of faces of people with whom he came into contact. The ability to elucidate the individuality of what he saw was affected.

It may be supposed that at a certain stage of his illness facial features were identical to him and he could not differentiate between male and female ones. The faces looked like similar objects to him and had no special characteristics. He lost the ability to penetrate into and feel the hidden trait of a face. This, in our opinion, is not connected with the optic or perceptual sphere, and there was no memory defect. The ability to analyse and synthesize facial peculiarities, the active and creative process related to recognition, was disturbed.

Dementia is not a likely cause of prosopagnosia, simultanagnosia, or similar disorders.

The question is, To what extent a right parieto-occipital lesion by itself may explain the prosopagnosia? It may be seen from the history that our patient was stricken twice some years ago by short attacks of loss of consciousness, probably caused by small thrombotic processes, which do not leave any recognizable sequelae. The last sudden cerebral insult caused a left hemianopsia together with psychosensory disturbances of the highest degree.

Any attempt to connect these signs with the patient’s premorbid personality will be valueless.

Though the patient disclosed unasked, during the early part of his illness, that he did not see colours clearly, we were unable to detect this objectively at any time. This subjective disturbance had already disappeared by the time the hemianopic field defect became quadriant. Probably the right occipital lobe was damaged, and it is more likely that this dysfunction is closer to achromatopsia than to agnosia for colours.

At the onset of the illness topographical memory was disturbed to an acute degree. The disturbance passed off within five or six days, but while it lasted, though the patient’s pictorial memory for topography was retained, he was unable to activate these stored memories when needed or when faced with the immediate problem of which way to turn. It was as if optical impressions were unable to activate the memory, or a put another way, as if orientation activity were upset. Sense of time was not at all affected and our patient used time sense in order to cancel out other deficiencies.

Some patients of this type suffer from a change in dream content. Our patient did not dream at all.

Regarding localization, our patient does not offer much more than other publications about patients of this type. One can say generally that a lesion in the parieto-occipital area of the brain, on the right more often than on the left, may cause a disturbance in the ability to recognize facial features. In the 25 cases collected by Hécaen between 1932 and 1957, the lesion was found on the right in 18, on the left in one, and no clear localization was established in three. As in most of the cases, our patient showed a left field defect affecting mainly the upper quadrants. There was no disturbance in speech, reading, or writing.

All the above facts lead us to suppose that the non-dominant hemisphere is mainly responsible for the development of prosopagnosia. Despite the fact that all the signs observed over the course of six months point towards a right-sided occipito-parietal lesion, it is highly likely that the two previous cerebral attacks caused bilateral damage to the brain. This opinion is supported by Dr. Ritchie Russell (personal communication) who states, “I should doubt myself whether this condition ever occurs in a lesion affecting one side of the brain only, as otherwise it would occur much more commonly than it does.”

It appears that the occipito-parietal region, in addition to powers of integration and synthesis, is endowed with special creative functions. Possibly when the latter functions are disturbed, high level qualities such as recognition of faces and meanings and expressions are affected.

Summary
A patient is presented who showed prosopagnosia, simultanagnosia, topographic disorientation, and achrmatopsia. All these signs, excepting the first one, disappeared entirely after a short time. The
prosopagnosia persisted to a lesser degree over the course of a year during which time the patient remained under observation. At no time was mental deterioration noted. The only neurological finding apart from the above was a left upper quadrantic visual field defect. The literature pertaining to prosopagnosia is reviewed and theories are put forward to explain the phenomenon, which is related to dysfunction of the right occipito-parietal lobe.

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REFERENCES