



Neuropsychologia 42 (2004) 633-638

www.elsevier.com/locate/neuropsychologia

Review

Patient Schn: has Goldstein and Gelb's case withstood the test of time?

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Received 8 August 2003; received in revised form 16 October 2003; accepted 21 October 2003

Abstract

The current manuscript takes a critical look at the case of Goldstein and Gelb's patient, Schn, reported to be the first well-defined example of apperceptive visual agnosia. While doubts have been cast on the validity of the original investigations, we propose that perhaps the case of Schn should be reclassified as an example of integrative agnosia. Be that as it may, what is not in doubt is that the case of Schn has had a lasting impact on the development of neuropsychological theorem.

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Keywords: Patient; Neuropsychology; Visual agnosia

1. Introduction

One of the most thoroughly discussed and controversial cases in the history of neuropsychology is that of Goldstein and Gelb's patient, Schn (Ellis, 1938; Goldstein & Gelb, 1918). Over the course of repeated examinations, Schn displayed a staggering number of neuropsychological impairments that included alexia, form agnosia, loss of movement vision, loss of visual imagery, tactile agnosia, loss of body schema, loss of position sense, acalculia and loss of abstract reasoning. For the purposes of this review, we will concentrate on Schn's reported visual agnosia, described by Goldstein and Gelb as a "serious incapacity to grasp purely visual presentations". Goldstein and Gelb proposed that Schn was unable to organize local feature elements into larger, more coherent "wholes". The case of Schn has played a significant role in most discussions of visual agnosia and has contributed to the refinement of Gestalt psychological theory of visual perception. Before we begin to discuss Schn's case, however, we will start with an overview of visual agnosia so as to be able to place Schn's visual disorder in context.

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2. Visual agnosia

A major obstacle to understanding object recognition is that we perform it so rapidly and efficiently that the outcome belies the underlying complexity. One approach to discovering the processes that mediate object recognition is to study the performance of individuals who have recognition impairments. Visual agnosia is a disorder of visual recognition, in which a person cannot arrive at the meaning of some or all categories of visual stimuli, despite normal or near-normal visual perception and intact alertness, intelligence, and language. Despite the visual recognition problems associated with agnosia, there is normal recognition of objects through modalities other than vision (touch, audition, verbal definition or description of its function), which indicates that the deficit is not simply a difficulty in retrieving names or in accessing the necessary semantic information. In short, the problem is in accessing meaning from visual input.

Lissauer (1890) was the first to classify visual agnosia into two broad categories: "apperceptive mindblindness" and "associative mindblindness". Using Lissauer's classification, a person with apperceptive agnosia is impaired at constructing a perceptual representation from vision and, subsequently, is unable to copy, match or identify a drawing. In contrast, a person with associative agnosia is one who cannot use the derived perceptual representation to access stored knowledge of the object's functions and associations but is able to copy and match the drawing even though

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unable to identify it. In sum, apperceptive agnosia arises from the failure to derive a coherent percept whereas associative agnosia arises from the failure to gain access to meaning from the well-derived percept (note that the extent to which the percept is really normal is the subject of much current debate (Delvenne, Seron, Coyette, & Rossion, in press; Farah, 1990)).

Several authors have further fractionated Lissauer's original distinction to reflect the sub-processes involved at the different processing stages of object recognition. Neuropsychological accounts by Humphreys and Riddoch (1987a) as well as by Warrington (1986), and computational accounts such as that of Marr (1982), have sought to extend Lissauer's dichotomy because of the growing understanding that visual object recognition comprises a number of distinct steps, not captured by the simple dichotomy. Another reason for further differentiation of the underlying processes comes from more fine-grained neuropsychological analysis (Humphreys & Riddoch, 1987a). These developments have forced a refinement of our understanding of visual processing and the types of breakdown that are possible after brain damage (Behrmann & Kimchi, 2003b; Humphreys & Riddoch, 2001; Riddoch & Humphreys, 2003). Despite the simplicity of Lissauer's dichotomy and its clear inadequacy, it still provides a coarse framework that has proved useful in describing agnosia.

3. Apperceptive agnosia

Individuals with apperceptive agnosia have profound difficulty recognizing and naming line drawings and photographs, as well as three-dimensional objects; their ability to recognize, copy, or match simple shapes is also impaired. However, their elementary visual functions such as acuity, brightness discrimination, and color vision are relatively preserved, along with reasonable sensory and semantic memory functioning in the visual domain (Farah, 1990). The fundamental deficit involves an inability to process features, such that they are not fully available for developing a percept of the overall structure of an object.

Apperceptive agnosia corresponds to the breakdown at the stage where the sensory features of the stimulus are processed and its structural description is achieved—a relatively early stage of the visual recognition system. The neurological damage in apperceptive agnosia tends to be diffuse and widespread and can involve damage to the posterior regions of the cerebral hemispheres, involving occipital, parietal, or posterior temporal regions bilaterally (Davidoff & Warrington, 1999; Farah, 1999; Habib & Sirigu, 1987; Humphreys, 1999; Humphreys & Riddoch, 1987a; Riddoch, Humphreys, Gannon, Blott, & Jones, 1999; Warrington, 1985, 1986). Goldstein and Gelb (1918) considered Schn to be the first well-defined example of apperceptive visual agnosia.

3.1. Johann Schneider (Schn)

On 4 June 1915, Johann Schneider (Schn), who was 24 years old, was wounded by mine-splinters while serving as a musketeer in the German army. He received two wounds to the back of the head: one apparently reached the exposed brain, of which, however, only indirect pulsations were visible and the other was less deep, located in the left parietal-occipital junction region. He was unconscious for 4 days. The head wounds diminished during the course of the next month and were closed in the middle of the next month. At the end of December, Schn complained of a tense feeling in the occiput and there was a palpable elevation at the location of the first scar. On December 29, 1915 a small iron splinter was removed from this location (Goldstein & Gelb, 1918). In their summary of the case report, Goldstein and Gelb (1918) added that the X-ray still showed some small splinters in the brain.

Bay, Lauenstein and Cibis (1949) came to a different conclusion concerning the location of the mine splinters and the depths of the wounds. They reported that there were many iron splinters in the soft parts of the left half of the skull and face but that all of them proved to be outside of the skull. However, on tangential radioscopy a minor depression of the skull in the region of the first wound was revealed. The depression may have been the remainder of the extracted splinter but no evidence of a penetrating skull wound was found.

Given the diagnostic limitations at the time of Schn's injury, the overall extent of his brain damage is unclear. There is convincing evidence, however, that Schn did suffer substantial brain damage. His symptoms were evocative of elevated brain pressure (slowing of pulse, jerk of both legs, then transient headache, vertigo and postural imbalance) and Goldstein and Gelb (1918) reported that a lumbar puncture in 1917 showed elevated pressure of liquor. An EEG was said to show a slight general alteration like in diffuse brain injury but with no local focus (Jung, 1949).

In February 1916, Schn was transferred to the Hospital for brain injury in Frankfurt, where he was first examined by Kurt Goldstein and Adhemar Gelb. While Schn's visual acuity was reported to be intact, his visual perception was not normal and his disorder was classified by Goldstein and Gelb as "psychic blindness". Schn was said to have lost any visual experience of form. Goldstein and Gelb believed Schn saw only color patches, recognizing their approximate size and location in space but could not integrate them into unitary, "whole" concepts. They claimed Schn's failure came from a disruption of perceptual grouping processes that led to a serious incapacity to comprehend purely visual presentations. They felt that Schn had profound difficulties in organizing local feature elements (e.g. edges and line segments) into larger, more coherent "wholes"—a failure in the formation of a visual Gestalt. This led to the Gestalt-like theory that agnosic patients are able to recognize individual features, but cannot synthesize a whole concept from the parts. This theory was popular because it had several strong supporting points: one is the frequency with which it explains otherwise puzzling clinical findings and the second concerns the utilization of nonfocal, but wide-spread cerebral abnormality. Interestingly, this explanation of visual agnosia has enjoyed somewhat of a renaissance recently under the label 'integrative agnosia', as we describe towards the end of this paper.

Goldstein and Gelb proposed that Schn had developed a compensation for his severe visual deficit that allowed him to recognize some simple geometric shapes and read by means of "tracing" the visually seen contours by hand or head movements. The visual contours, which lacked meaning for him, were transformed via a kinaesthetic process, into another spatial modality which then permitted him to make inferences and deductions of object form (Landis, Graves, Benson, & Hebben, 1982). Because of the unusual efficiency of this compensation, the true nature of his deficit became apparent only during explicit examinations of visual functions (Goldenberg, 2003). For example, it took Schn 6 s to identify a triangle from repeated tachistoscopic presentations; during that time he made tracing movements of the head.

Goldstein and Gelb reported that when Schn used this kinaesthetic aid while reading, he "wrote" with his hand what his eyes scanned. Schn traced each letter of every word with his finger and wrote the letters one over the other, while tracing them with his head-movements. While Schn was able to read, longer words sometimes required as much as 10 s and his reading broke down when words were presented tachistoscopically. Furthermore, if Schn was prevented from moving his head or body, he was unable to read visually-presented words. In summary, Goldstein and Gelb's theory was that Schn lacked any visual experience of form (or Gestalt) but that he compensated for this deficit by tracing visually presented forms with movements of either the head or the fingers, eventually recognizing the form by kinaesthetic feedback. This method of compensation has also been reported by other investigators of visual agnosia as being a useful strategic method for obtaining information about visual displays (Benson & Greenberg, 1969).

As is evident from this short review of Schn, there are two major issues raised by the case. The first concerns the authenticity of Schn's symptoms and the second concerns the classification of his visual agnosia as being of the apperceptive type. We deal with each in turn.

4. How genuine were Schn's impairments?

While the case of Schn has significantly influenced the visual agnosia literature, it has come to be suspected of being unreal (Teuber, 1966). Indeed, some authors have suggested that the symptoms were either hysterical or grossly exaggerated (Bay, 1953) and, consequently, have doubted the reality of the reported compensation by kinaesthetic

mediation. Instead, these critics have suggested that Schn was actually using his vision and thus was not agnosic (Bay et al., 1949; Jung, 1949). In fact, on their first investigation of Schn, Goldstein and Gelb commented on the fact that apart from increased fatigue after reading, Schn did not spontaneously complain of any visual deficiencies, nor was he visually handicapped in his daily life. Goldstein and Gelb were initially surprised at how well Schn could interact in the world. For example, Schn was able to describe the content of scenes in paintings, which casts doubt on Goldstein and Gelb's claim that Schn could only see "formless dots". Furthermore, Schn's claim to have no visual imagery is contradicted by the fact that he could actually draw "unusually well" from memory (Goldenberg, 2003). In a recent review, Goldenberg (2003) suggests that Goldstein and Gelb were too eager for an all-embracing theory of the human mind and its reaction to brain damage. Schn became a valid and comprehensive illustration of their ideas about the influence of brain damage on perception and reasoning. Goldenberg claims Goldstein and Gelb invented fantastic embellishments about Schn and, in turn, Schn learned how to be an ideal case study.

Schn's reported use of kinaesthetic feedback has been particularly scrutinized. Critics of Gelb and Goldstein, who examined Schn years later, found his tracing movements rather showy and theatrical and doubted that the patient had apperceptive agnosia. For example, Jung (1949) noted that Schn was able to see and recognize most objects and seemed to switch into his tracing routine only when performing tests for psychologists. Furthermore, Bay (Bay, 1953; Bay et al., 1949) found that the head movements made by Schn had no relationship to the letters, syllables and words read in either their number or direction. While this certainly brings into question Goldstein and Gelb's claim, it should be noted that other patients, with similar visual impairments, have spontaneously adopted the same type of tracing strategy, which makes it unlikely that the tracing was purely for show.

In fact, a striking feature of apperceptive agnosia is that many patients spontaneously use quite laborious and time-consuming tracing strategies of the hand or head to aid in the recognition of visual objects (Benson & Greenberg, 1969; Campion, 1987; Landis et al., 1982). These strategies, although helpful, may not always produce an accurate result, as one requires a reasonably good visual image in the first place for the purposes of tracing. Landis et al. (1982) documented a visual form-agnosic, Patient X, who had very similar deficits to Schn and also used a kinaesthetic "writing cue" to aid in visual recognition. Both patients lacked the ability to discriminate visual patterns in the normal manner but both used kinaesthetic tracing. Landis felt their observations strongly imply that the kinaesthetic tracings of X and, by inference, those of Schn, served as genuine compensation for damage rather than being "hysterical" or "learned exaggerations". In contrast to Schn, however, Patient X apparently used the same strategy for cueing himself in naming from verbally presented definitions suggesting that the strategy served lexical retrieval rather than visual analysis (Goldenberg, 2003). Whether or not, then, the apparent compensatory behaviors in patient X and Schn are indeed equivalent remains unclear.

5. Classification of Schn's agnosia

Goldstein and Gelb's (1918) report has remained a citation classic in neuropsychology, likely attributable to the careful observation and comprehensive description of the impairments suffered by Schn. Of particular interest in light of some current advances in the study of visual agnosia is Schn's particular pattern of visual symptoms. Although Schn was classified as suffering from 'apperceptive agnosia', his profile may fit the more recent definition of 'integrative agnosia'.

Patients with integrative agnosia appear to have available to them the basic features or elements in a display but are unable to integrate all aspects into a meaningful whole. HJA, the first patient to whom the label 'integrative agnosia' was given and described by Riddoch and colleagues, performs normally on visual matching tasks and can copy images reasonably well. HJA also performs well on a search task when identifying a target that does not require a combination of elements (for example, differentiating '/' from '|') but performs poorly when required to bind visual elements in a spatially parallel fashion across a field containing multiple stimuli, such as searching for an upright 'T' among misoriented 'T's (Humphreys, 1999; Humphreys, Cinel, Wolfe, Olson, & Klempen, 2000; Humphreys & Riddoch, 1987b; Humphreys, Riddoch, Quinlan, Price, & Donnelly, 1992). The failure of HJA and other integrative agnosic patients to integrate elements occurs equally with displays of twoand three-dimensional stimuli and to black-and-white and chromatic displays although, in some cases, the presence of depth, color and surface cues may be of some assistance to the patients in segmenting the display (Chainay & Humphreys, 2001; Farah, 1990; Humphreys et al., 1994; Jankowiak, Kinsbourne, Shalev, & Bachman, 1992).

These problems in integrating components into a unified whole are most clearly demonstrated when there are multiple items present, such that there is competition in assigning elements between shapes (De Renzi & Lucchelli, 1994; Kartsounis & Warrington, 1991; Riddoch & Humphreys, 1987; Riddoch et al., 1999). For example, many of the patients with integrative agnosia are more impaired at identifying items that overlap one another compared with the same items presented in isolation. Interestingly and counterintuitively, in some patients, the presence of local information may even reduce the efficiency of visual recognition; in contrast with normal perceivers, some patients with integrative agnosia identified silhouettes better than line drawings, whose internal details apparently led to incorrect segmentation (Butter & Trobe, 1994; Humphreys et al., 1992; Lawson & Humphreys, 1999; Riddoch & Humphreys, 1987). The silhouette advantage is thought to arise from the reduced need to segment and integrate elemental features relative to the line drawings. Note that there is less information available in the silhouette too so any competition is reduced.

A further key feature of integrative agnosia is the failure to carry out figure-ground segregation; patient FGP, for example, cannot even determine the presence of a 'X' when it is superimposed on a noisy background (Kartsounis & Warrington, 1991). Finally and critically for our purposes, these agnosic patients seem to be impaired at grouping, as is Schn; for example, patient NM was impaired at detecting the presence of a target letter when it was defined by multiple oriented line segments in a display with distractors of different orientations (Ricci, Vaishnavi, & Chatterjee, 1999). The same was true when the target was defined by color, luminance or motion features, relative to the distractors (Marstrand, Gerlach, Udesen, & Gade, 2000). Behrmann and Kimchi (2003a,b) have recently conducted several experiments documenting the failure of two patients with integrative agnosia to derive a coherent whole from elemental components. For example, in contrast with normal subjects who identify the 'forest' before the 'trees' in Navon-style hierarchical displays (such as H constructed of little Ss), these patients identify the local components first. The rapid and efficient access to the local information impedes the patients' ability to gain access subsequently to the global information. This local-first pattern or local advantage also appears to be correlated with the severity of the agnosic disorder.

Although Goldstein and Gelb did not deliver watertight empirical evidence, their description of Schn suggests that he fits this pattern of agnosia. Schn's perceptual problems reflect an impairment in integrating local form elements into more holistic shapes and his "piecemeal" approach to object identification appears to reflect a problem in grouping local form information. This description of Schn was most timely for Goldstein and Gelb, given the zeitgeist at that time of Gestalt psychology. Indeed, the existence of an individual who showed a breakdown in Gestalt processing after brain damage, served to reinforce that era's ideas about visual perceptual organization.

The reclassification of Schn as an integrative agnosic is particularly relevant right now as there have been several recent cases categorized as being of this integrative type. Ideas about Gestalt processing and visual perceptual organization seem to be enjoying a revival to some extent. This might be attributable to the recent interest in perceptual organization in a number of related disciplines. For example, there are several neurophysiologists who are recording activity at the single unit level while awake, behaving monkeys perform curve tracing, figure-ground and boundary assignment tasks, and these studies have shed light on the neural mechanisms that might underlie the organization of visual input in the early stages of the visual system (Lamme & Roelfsema, 2000; Zhou, Friedman, & von der Heydt, 2000). In parallel, visual psychophysicists have devoted

much energy to carefully documenting the behavioral and functional properties of perceptual organization (Palmer, 1999, 2002). Sandwiched between these two groups, are the neuropsychologists who have documented cases of integrative agnosia, like Schn, and have served as a bridge between the behavioral and single-unit contributions to Gestalt processing.

6. Conclusions

Gestalt psychologists, like Goldstein and Gelb, were the first to investigate the relationship between the perception of the whole and that of its constituent parts. They proposed that perceptual organization is achieved by grouping elements together by virtue of certain properties that are present in the image. These Gestalt views on perceptual organization have been widely accepted as identifying crucial phenomena of perception. In fact, there are now several studies which appeal to Goldstein and Gelb's Gestalt-like theory in explaining visual agnosia (Behrmann & Kimchi, 2003b; Kartsounis & Warrington, 1991; Piccini, Lauro-Grotta, Michela Del Viva, & Burr, 2003; Ricci et al., 1999). The patients in these studies, like Schn, are unable to integrate low-level elements into global structure—a Gestalt foundation for perceptual organization. Instead, these patients operate in a "piecemeal" manner to identify objects. The integration of local part information with holistic shape seems to break down in these patients.

The almost universal applicability of Goldstein and Gelb's theory might be considered a major defect: when strikingly dissimilar cases are "explained" by some defect in Gestalt formation, the theory lacks specificity (Benson & Greenberg, 1969). One can certainly make the argument, as Goldenberg (2003) has recently, that if a case report is considered to be classic only if later papers have confirmed and expanded its findings, case Schn does not merit inclusion in this collection of classic cases. To that end, the fact that case Schn is still cited in papers on visual agnosia may be the result of the inaccessibility, length and (for non-German speaking readers) incomprehensibility of the original papers.

Nevertheless, the case of Schn still might be considered a classic if one considers its influence on the development of ideas not only in neuropsychology but also in general psychology and philosophy. The breakdown approach highlighted by Goldstein and Gelb has provided important insights into the mechanisms involved in normal object recognition. Perhaps most important is that studies of patients with visual object agnosia have constrained our theories of object recognition and, in turn, these theories have guided our investigation of these interesting and illuminating deficits. In conclusion, while there will always remain some doubt as to the validity of all of Goldstein and Gelb's claims about Schn, what is not in doubt is that this case has played a major role in guiding investigations of visual agnosia and object recognition.

Acknowledgements

We would like to thank Dr. Georg Goldenberg for reviewing this manuscript and providing us with valuable additional details about Schn's head injuries.

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