Visual agnosia with bilateral temporoparietal brain lesions in a child with autistic disorder: a case study

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A 2-year-old boy meeting the criteria for autistic disorder was diagnosed 2 years later with a visual agnosia characterised by a combination of certain aspects of associative and apperceptive agnosia. MRI then revealed a severe encephalomalacia of the right temporal lobe and bilateral temporoparietal areas. This association is discussed in terms of a clinical and aetiological relation between autistic disorder and visual agnosia.

Autistic disorder is a pervasive developmental disorder characterised by marked deficits in the areas of social interaction, communication, and imaginative play, as well as by the presence of a restricted range of interests. Among the social interaction diagnostic criteria for autistic disorder, the DSM-IV (American Psychiatric Association 1994) lists impairment in the use of non-verbal behaviours, such as eye-to-eye gaze, facial expression, body posture, and gestures to regulate social interaction. Empirical investigation has confirmed that face perception is qualitatively different in people with autistic disorder. Recognition of inverted faces is easier for people with autism than for people without autism (Tantam et al. 1989). In addition, people with autistic disorder show difficulty in face matching and memorising (Bouche and Lewis 1992), and in facial emotion recognition (Bormann-Kischkel et al. 1995). In the area of repetitive behaviours and restricted interests, several symptoms of autistic disorder subsumed under the DSM-IV item 3d (preoccupation with parts of objects), suggest irregular visual perception. These are incessant visual fixation with spinning objects and with surfaces, textures, and lights, combined with an absence of visual exploration of other aspects of objects (Ornitz 1988).

People with autistic disorder exhibit atypical visual behaviours towards people and objects. However, their visual perception, up to and including the formation of a representation of retinal information in occipital areas of the brain, is generally considered to be intact (Frith and Baron-Cohen 1987). This conclusion is based on strong performances in certain visuo-spatial tasks (Tymchuk et al. 1977) and normal ability to categorise visually presented objects (Tager-Flusberg 1985). Consequently, symptoms in the perceptual domain are not held as a primary diagnostic criterion in current models for autistic disorder, but instead are believed to stem from deficits in higher cognitive processes, particularly the ability to plan mental operations (Ozonoff et al. 1991), and to infer mental states in other people from facial cues (Baron-Cohen et al. 1995). This view, however, is not without its opponents (Frith et al. 1994). Yet a few cases of prosopagnosia, i.e. a visual perceptual deficit specific to facial cues, have been reported in patients with various pervasive developmental disorders (Kracke 1994, Jambaqué et al., unpublished data).

This paper presents the first case combining autistic disorder, visual agnosia for faces and objects, and a brain lesion congruent with visual agnosia.

Case report
The subject was born weighing 4140 g and with a head circumference of 35 cm after an uncomplicated pregnancy. His Apgar
score was 9 at 1 and 5 minutes. There was no family history of psychiatric illness. At 1 month, he presented with moderate symptomatology of viral meningitis, but no immediate sequelae were observed. The parents first became concerned when, at the age of about 6 months, he began to gaze upwards for a number of seconds and to present a converging strabismus. They also noticed an absence of facial reaction when leaving and returning to him. Hand-flapping at 6 months and prolonged visual fixation at the end of the first year were also cause for concern. He sat at 4 months and walked at 12 months. He uttered his first words at 18 months and his first two-word phrases before 24 months. He is right-handed.

A clinical diagnosis of high-functioning autistic disorder was made at 2 years 10 months and at 3 years 1 month based on an absence of social and symbolic play, an absence of gaze in reciprocal interaction, social avoidance, restricted interest in objects, and excessive orientation towards noises and reflections of light. Most of his verbal interaction consisted of stereotyped questions, pronoun reversal, and verbal rituals. At 3 years 5 months he was not yet able to eat or dress alone and did not jump or kick a ball. He could, however, move about in familiar surroundings without colliding with objects. A diagnosis of autistic disorder was formally established at 5 years, based on information provided by the Autism Diagnostic Interview-Revised (ADI-R) (Lord et al. 1994) about his behaviour around the times of his 2nd, 3rd, 4th, and 5th birthdays. This was performed separately on the same occasions, by the subject’s mother and an educational psychologist (SM) in charge of the child during this period of time. The information provided sufficient evidence for a diagnosis of autistic disorder according to the DSM-IV criteria in all three diagnostic areas of the ADI-R between 2 and 4 years, but fell below the more conservative ADI-R cutoff for autistic disorder in the social area at 5 years (Table 1). However, he continued to meet the DSM-IV criteria for autistic disorder at 5 years (i.e. 1a, 1b, 2b, 2c, 2d, 3a). as assessed independently through behavioural observation by two trained clinicians (LM, JPP). This course is not exceptional, as an improvement in social symptoms characterise people with autism and without mental deficiency (Piven et al. 1996).

COURSE
At age 3 years 6 months, the subject entered a day-treatment centre to receive therapy aimed at stimulating perceptual, motor, cognitive, and affective areas. It was observed after a few weeks that he could recognise people only after hearing their voices. He also held objects close to his eyes, moving them from side to side while keeping his head still. As a result of these observations, he was found to have visual agnosia. The treatment protocol (Mineau and Mottron, forthcoming) was modified in consequence, by using the repetitive questions he asked about his special interests to teach him to name his physical and social surroundings and to relate auditory information with visual information. An improvement in social and communicative behaviour was subsequently noted. Motor delay and self-care skills began to normalise after the age of 4 years. His spontaneous activity and discourse, however, continued to be pervaded by a special interest in roads and bridges. After the day-treatment centre, he was sent to a regular school with special assistance for children with visual impairment.

PSYCHOLOGICAL ASSESSMENT
The subject was assessed between the ages of 4 years 2 months and 4 years 7 months. He obtained a verbal IQ of 97 (Wechsler Primary and Pre-School Scale of Intelligence-Revised; WPPSI-R) and a global IQ of 108 on the William's Intelligence Test for children with defective vision (William). Memory assessment was average to above average. In short-term memory tasks he performed more than 1 year above his chronological age. Semantic memory was within his chronological age range, with a mental age of 4 years on the information subset of the WPPSI-R.

As people with autistic disorder are typically delayed in their ability to infer the mental states of others (Happé and

Table I: Course of DSM-IV symptoms for diagnosis of autistic disorder according to ADI-R scores

<table>
<thead>
<tr>
<th>DSM-IV criteria</th>
<th>Age (y)</th>
<th>6</th>
<th>7</th>
<th>8</th>
<th>9</th>
<th>10</th>
<th>11</th>
<th>12</th>
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<tbody>
<tr>
<td>Social interaction</td>
<td></td>
<td>(a) Failure to use non-verbal behaviours to regulate social interaction (/6)</td>
<td>6</td>
<td>6</td>
<td>3</td>
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<td></td>
<td></td>
<td>(b) Failure to develop peer relationships (/10)</td>
<td>7</td>
<td>7</td>
<td>4</td>
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<td></td>
<td></td>
<td>(c) Lack of shared enjoyment (/6)</td>
<td>6</td>
<td>6</td>
<td>3</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td>(d) Lack of socioemotional reciprocity (/10)</td>
<td>5</td>
<td>5</td>
<td>3</td>
<td></td>
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<tr>
<td></td>
<td>Total (cutoff=10)</td>
<td>24</td>
<td>23</td>
<td>13</td>
<td>8</td>
<td></td>
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<tr>
<td>Communication</td>
<td></td>
<td>(a) Delay in spoken language and failure to compensate through gesture (/8)</td>
<td>8</td>
<td>6</td>
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<td></td>
<td></td>
<td>(b) Lack of varied spontaneous make-believe or social imitative play (/6)</td>
<td>6</td>
<td>6</td>
<td>3</td>
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<td></td>
<td></td>
<td>(c) Relative failure to initiate or sustain conversational interchange (/4)</td>
<td>4</td>
<td>4</td>
<td>2</td>
<td>2</td>
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<td></td>
<td></td>
<td>(d) Stereotyped, repetitive or disynchratic speech (/8)</td>
<td>4</td>
<td>7</td>
<td>4</td>
<td>3</td>
<td></td>
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<td></td>
<td>Total (cutoff=8)</td>
<td>22</td>
<td>21</td>
<td>15</td>
<td>9</td>
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<tr>
<td>Repetitive behaviours and stereotyped patterns</td>
<td></td>
<td>(a) Encompassing preoccupation or circumscribed pattern of interest (/4)</td>
<td>4</td>
<td>4</td>
<td>3</td>
<td>3</td>
<td></td>
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<td></td>
<td></td>
<td>(b) Compulsive adherence to non-functional routines or rituals (/4)</td>
<td>0</td>
<td>2</td>
<td>2</td>
<td>3</td>
<td></td>
<td></td>
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<td></td>
<td></td>
<td>(c) Stereotyped and repetitive motor mannerisms (/2)</td>
<td>2</td>
<td></td>
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<td></td>
<td></td>
<td>(d) Preoccupation with parts of objects (/2)</td>
<td>2</td>
<td>2</td>
<td>1</td>
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<td></td>
<td>Total (cutoff=3)</td>
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<td>6</td>
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*Value above which an area is considered sufficiently impaired for the diagnosis of...
Frith 1995), the subject was tested for his reaction to strange stories (Happe 1994). Strange stories are narratives based on irony, figurative language, or lies, which autistic people have significant and consistent difficulties understanding. The subject failed to grasp 8 out of 6 stories, compared with a control subject of the same intelligence, age, and sex, who performed successfully on 3 out of 6 stories. In all, he performed approximately at his developmental and chronological level in tasks that did not involve a visual component or did not require him to infer the mental states of others. Consequently, none of the specific impairments he presented in the visual modality could be explained by mental deficiency or developmental delay.

NEUROPSYCHOLOGICAL ASSESSMENT OF VISUAL AGNOSIA

Low-level perception was assessed by a neuro-ophthalmological examination at 4 years 6 months of age. In the visual acuity test, the fixation pattern was centred, steady and held in each eye. Although formal colour-vision testing was impossible, he could identify colours in natural settings. Visual fields assessed by confrontation were normal in front of each eye, but this method does not preclude subtle deficits. Pupils reacted normally to light and accommodation, with no relative afferent pupillary defect. Ocular motility, stereo vision, pursuit, saccades, and optokinetic nystagmus were all normal. A slit-lamp examination was unremarkable. Ophthalmoscopy after pupillary dilation revealed normal optic discs, macular, retinal vessels, and periphery.

According to current models of high-level visual perception, the processing of visual stimuli relies on at least three different systems: (1) construction of perceptual representation at the cortical level, (2) matching this representation with stored visual and verbal representations, and (3) matching of these stored representations with semantic memory (Humphreys and Bruce 1989). These various processes were assessed between 3 years 8 months and 4 years 6 months of age by means of matching, decision-making, recognition, and naming tasks, and requests for information concerning objects and faces. Construction of perceptual representations of faces was severely impaired. He performed at the level of chance (12 out of 24) in a same–different judgement task involving paired line drawings of faces and obtained only 10 out of 15 correct answers in a task that consisted in determining whether the line drawing of a face was scrambled or not. In a decision-making task measuring familiarity, he recognised the correct face in only 2 out of 10 pairs of photographs. Face-naming was also severely impaired. When presented with photographs of a familiar face paired with photographs of an unfamiliar face, he identified the familiar face in only 1 out of 12 trials. Error analysis showed that he mistook children for adults, men for women, and white people for black people.

On measures of object recognition, the subject was successful in making same–different judgements involving pairs of simple geometric shapes and in naming these forms. This ability has been noted in two other cases of developmental visual agnosia (Gordon 1998). However, his performance was significantly impaired with more complex objects. In 40 pictures depicting an equal number of foods, animals, vehicles, and clothes, he successfully named 25% of all the objects, of which 60% were vehicles, 30% were animals, 10% were foods and 0% were clothes. He also demonstrated a striking dissociation between visual and tactile modalities. When 20 objects were presented to him either visually without tactile access or tactually without visual access (in a bag), he obtained 55% correct responses in the visual modality and 80% correct responses in the tactile modality. Twenty-five percent of the objects were recognised only in the tactile modality and 5% only in the visual modality. This suggests that he used threedimensional cues, as his performance is more efficient with real objects than with photographs.

The subject provided, on average, two correct items of information on 10 familiar people, of whom he could name only one. In the object domain, he performed successfully on a task assessing knowledge of perceptual and functional properties by means of questions whose responses required access to semantic memory and therefore did not rely solely on rote memory. His efficient performance on tasks measuring semantic memory for people and objects precludes memory impairment or lack of knowledge as explanations for his recognition difficulties. These findings, combined with his normal peripheral vision, provide support for a diagnosis of visual agnosia. Together with the visuo-tactile dissociation, this suggests that he is able to construct a semantic base of knowledge based mainly on linguistic input and to access this base through nonvisual modalities. His profile is one that combines certain features of associative agnosia (his visual pursuit and fixation are normal, and he does not behave like a blind person) and of apperceptive agnosia (he cannot match or copy objects).

CEREBRAL IMAGERY

MRI was performed at 4 years of age on a SGR 2 tesla System (Elekta Limited®, Haifa, Israel). Following a sagittal T1-weighted spin-echo sequence, T1 and T2 sequences were obtained in the axial and coronal planes with 3 mm slices at 1 mm gaps. Lesions were accurately localised with the help of an MRI atlas (Damasio 1995).

The right hemisphere was the most severely affected, exhibiting multiple foci of encephalomalacia. There was complete lateral destruction of the temporal lobe (Figs 1, 2). Only the posterior third of the superior temporal gyrus was spared. On the medial surface, the amygdala, the hippocampus, and the anterior half of the fusiform gyrus showed extensive parenchymal loss. The right fornix was not identified. No lesion was noted on the occipital medial surface: the cuneus, the calcarine area, the lingual gyrus, the posterior part of the fusiform gyrus, and the retrosplenial area were normal as was the parasagittal part of the parietal cortex, including the precuneus and the paracentral lobule. The posterior part of the corpus callosum and the splenium were moderately atrophic. In the posterolateral cortex, foci of encephalomalacia affected the whole angular gyrus and most of the supramarginal gyrus. The superior parietal lobule and the external occipital gyri were normal. The left hemisphere was less involved. Foci of parenchymal loss were noted in the temporoparieto-occipital junction. The anterior part of the fusiform gyrus, the amygdala, the whole hippocampus, and the lingual gyrus were normal. Lesions were visible in the posterior parts of the temporal gyri (T2, T3, T4), the parieto-occipital junction and most of the angular gyrus, and extended posteriorly to involve the lateral occipital gyri (Figs 3, 4). Similar lesions have been shown to produce visual agnosia in adults (Alexander and Albert 1983, Damasio et al. 1983, Farah 1990). The etiology of this lesion is unknown. The localisation and extensiveness of the subject's neuroradiological lesions
are analogous to those resulting from herpes encephalitis. However, this explanation is hardly compatible with the medical history of the subject as the meningitis with which he presented at 1 month of age was mild and involved only a few days of hospitalisation. A more plausible explanation would be a prenatal agenesis of several cerebral territories due to a deficit in vascularisation.

**Discussion**

The subject’s case provides a particularly clear example of autistic disorder with bilateral temporo-occipital lesions and visual agnosia. The discussion will focus on the possible causal relation between autistic disorder and temporo-occipital lesions, on the one hand, and autistic disorder and visual agnosia, on the other.

**AUTISTIC DISORDER AND TEMPORAL LOBE LESIONS**

Anatomical or functional abnormalities of the temporo-occipital region have been discovered in a small but increasing number of cases of autistic disorder. In the restricted number of cases where abnormalities exist, this region appears to be most frequently implicated, although such abnormalities may appear in other regions or may be absent (Jones and Kerwin 1990, White and Rosenbloom 1992, Berthier et al. 1993).

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**Figure 1:** Axial MRI cut.

**Figure 2:** Mapping of lesions in the axial plan (-20°) on the corresponding Damasio Atlas picture nr 120.

AG=angular gyrus; Cun=cuneus; IN=insula; pin=pineal gland; RS=rostroplenial area; SPL=superior parietal lobule; STG=superior temporal gyrus; Tpole=temporal pole; f=basal forebrain; thalamus.
Additional arguments to this effect are provided by functional neuroimagery, where decreased or abnormal regional cerebral blood flow has been observed, with or without lesions, in some subjects suffering from autistic disorder, Asperger's disorder, and West syndrome with autistic symptoms (Ozbayrak et al. 1991, George et al. 1992, Jambaqué et al. 1993, Garreau et al. 1994, McKelvey et al. 1995). An association between temporo-occipital lesions and pervasive developmental disorder has also been reported within the framework of various medical or surgical conditions. These include mesial left temporal structures destroyed by an oligodendroglioma in a picture of autistic disorder (Hoon and Reiss 1992); autistic disorder after herpes encephalitis with temporal lesions (DeLong et al. 1981; Gillberg 1986, 1991); and temporal tubers frequently associated with autistic disorder, in tuberous sclerosis (Jambaqué et al. 1991, Deonna et al. 1993, Calderon Gonzalez et al. 1994, Bolton and Griffith 1997). The wide variety of etiologies responsible for temporo-occipital lesions suggest that it is localisation, and not etiology, that is relevant.

The age at which neurological damage occurs may also play a role in the etiology of autistic disorder. Schiavetto and colleagues (1996) describe the case of a girl with associative visual agnosia resulting from complete destruction of the temporal lobe by herpes encephalitis at age 9 years. She did not, however, present any developmental impairment consistent with autistic disorder. Differences in the age at which the damage...
occurs may therefore explain the presence or absence of autistic disorder.

AUTISTIC DISORDER AND VISUAL AGNOSIA

The subject's clinical picture suggests three possible relations between autistic disorder and visual agnosia. First, a diagnosis of autistic disorder could be rejected and his symptoms attributed to visual impairment. For example, his abnormal gaze behaviour could be accounted for by failure to construct perceptual representations of faces instead of by autistic gaze avoidance. Such an explanation might account for why certain researchers have described more favourable outcomes for autistic clinical pictures associated with a visual symptomatology than is the case for autism 'sine materia' (Goodman and Ashby 1990; Jambaqué et al. 1993). Two arguments, however, undermine this interpretation. First, as is common in individuals with autistic disorder, the subject is impaired in his ability to infer the mental states of others. Second, it seems arbitrary to separate individuals whose autistic symptoms are caused by neurological lesions from those whose symptoms are coincident with lesions, given that the definition of autistic disorder is at present purely clinical.

A second interpretation of the relationship between autistic disorder and visual agnosia is that it is temporal lesions, and not visual agnosia, that are causally linked with autistic disorder. Consistent with this hypothesis, the rare combination of visual agnosia and autistic disorder would result from the same pathological insult, in cases where this insult is extensive enough to include the tempo-occipital areas. This may explain why visual agnosia has been described only in a restricted number of cases of autistic disorder with functional or anatomical temporal lesions.

A third, more speculative position is that people with autistic disorder present with a visual agnosia in which object and face recognition is qualitatively modified (Mottron and Belleville 1997). This stance is congruent with the temporal model for autistic disorder proposed by Damasio and Maurer (1978) and Hetzer and Griffin (1981) (see also Bachevalier 1994, for a review). Within this model, autism is seen as a developmental Klüver-Bucy syndrome, which results from the destruction of mesial temporal lobes and involves visual agnosia. The deficits that people with autistic disorder display in face perception, as well as their abnormalities in constructing visual representations (Mottton and Belleville 1993) support this position. Similarities in the cognitive profile of people with autistic disorder and visual agnosia are also remarkable. People with autistic disorder generally present superior performances on the 'block design' subtest of the Wechsler Adult Intelligence Scale (Shah and Frith 1993), as do some individuals with visual agnosia (McConachie 1976; Greer et al. 1989).

Conclusion

According to Kracke (1994) no case of developmental prosopagnosia without features of pervasive developmental disorder has ever been reported. Nevertheless, several cases seem to contradict this position (McConachie 1976; 1995; Young and Ellis 1989; De Haan and Campbell 1991; Martins et al. 1993). The difficulty in resolving this difference in view stems as much from the absence of an in-depth assessment of visual agnosia in the autistic disorder literature as from the lack of any assessment of autistic disorder in the literature on developmental agnosia.

The discovery of a visual agnosia in a child with autistic disorder significantly changes the manner in which such a child is cared for (Mottton and Mottron, forthcoming). Whereas our subject's social deficits were originally understood as resulting from 'social avoidance', the discovery of impairments in visual recognition led to the causal implication of a perceptual deficit. With this change of focus, verbal communication was enhanced, and he was taught to identify his peers during social interaction through the recognition of local cues he was able to recognize. In addition, he was oriented towards the blind system of special education, especially for reading. In conclusion, this subject's case shows that autistic disorder in general may be associated with a visual agnosia. It also suggests that a diagnosis of autistic disorder may justify cerebral imaging or a neuropsychological investigation focused on visual agnosia, as important consequences in rehabilitation may result from this diagnosis.

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References


