Visuospatial deficits
and visual agnosia

Masud Husain

Dept Experimental Psychology & Nuffield Dept Clinical Neurosciences, University of Oxford
Visuomotor deficits

Often associated with right posterior parietal (dorsal visual stream) damage or dysfunction

- **Visual disorientation** often with visual mislocalization and gaze apraxia
- **Constructional apraxia**
- **Spatial working memory deficits**
- **Optic ataxia** | associated with right (or left) superior parietal lesions
- **Visual extinction** | associated with right (or left) parietal lesions
- **Neglect syndrome** | more severe and long-lasting with right inferior parietal lesions
- **Topographical disorientation** | of the egocentric variety (see later)

*In contrast, left parietal damage is often associated with language dysfunction, verbal working memory deficits, dyscalculia and limb apraxia*
Visual disorientation

Gordon Holmes (1918)
When asked to touch an object in front of him he would grope hopelessly.

He could not count coins set before him. He had difficulty in seeing more than one item at a time and would bump into objects.

Emphasized a disorder of visual space perception

Gordon Holmes (1918)

Visual disorientation
Bálint’s syndrome

Bálint put greater emphasis on inattention and visually guided misreaching

- Patient could no longer judge where things were. Felt unsafe to cross roads.
- Looked straight ahead, unaware of objects on either side (bilateral inattention)
- Bálint called it “psychic paralysis of gaze” (effectively a gaze apraxia)
- Could only report one object at a time (simultagnosia)
- Misreached to visual objects (optic ataxia)
- Post-mortem: large bilateral strokes involving parietal lobes

Bálint (1917)
Holmes’ case series from 1918

Remarkable similarity to Bálint’s case although Holmes emphasized disorder of space perception

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Oculomotor apraxia</th>
<th>Misreaching Disorders of attention</th>
<th>Space perception disorders</th>
<th>Run into things</th>
<th>Topographical disorientation</th>
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<tbody>
<tr>
<td>1</td>
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<tr>
<td>2</td>
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<td>Right neglect</td>
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<td>Left neglect</td>
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+=presence of symptoms. Patient 7 was reported by Holmes and Horrax (1919). From gunshot injuries through both parietal lobes.

Some refer to this constellation as Bálint-Holmes syndrome
Dot counting

Still used as a simple test of visuospatial ability, e.g. in Addenbrooke’s Cognitive Examination
Other tests of spatial localisation

*Position discrimination* | *Number location* | *Both from Warrington’s VOSP battery*
Optic ataxia

Misreaching to peripheral visual targets
Constructional apraxia

*Difficulty in perceiving spatial relationships – tested by asking patient to copy a figure*

Rey-Osterreith Figure  Patient A’s copy  Patient B’s copy
Constructional apraxia

Difficulty in perceiving spatial relationships – tested by asking patient to copy a block model

Block construction
Spatial working memory

*Traditionally tested using the Corsi blocks task – remember increasing sequences of spatial locations*

- Examiner taps out a sequence of locations
- Patient has to reproduce this
- Obtain a measure of spatial span or how many locations can patient maintain
So what underlies visual disorientation?

Egocentric perceptual mislocalization, impaired attention, spatial working memory or visuomotor control?

- Even simple dot counting requires spatial perceptual localization, attention, memory and eye movement control

- Would a deficit in egocentric localization be sufficient?

- Is there a deficit in ‘spatial remapping’ (remapping locations across eye, head and body movements so there is spatial constancy)?

Russell et al (2010) Brain
Egocentric localisation

*Mapping objects with respect to the body* | *Requires convergence of different types of sensory input*

![Diagram of egocentric localisation](image)
Egocentric localisation

*Mapping objects with respect to the body | Requires convergence of different types of sensory input*

If I know:

- where an object is with respect to direction of gaze (retinotopic or eye-centred location)
- which direction the eye is pointing with respect to the head and
- where the head is with respect to the trunk

I can calculate the object’s position with respect to the body.

The position of the hand with respect to the body can also be computed from proprioceptive inputs from the arm, so I can reach accurately even to the remembered location of an object.
**Allocentric localisation**

*Mapping objects relative to each other’s location | Requires viewpoint independent representations*
Egocentric to allocentric transformation

Hypothesized to require lateral parietal to hippocampal inputs, via medial parietal regions
Topographical disorientation
There are different ways to get lost!

- Egocentric disorientation
  Associated with right posterior parietal lesions

- Heading disorientation
  Observed after retrosplenial / posterior cingulate lesions

- Landmark agnosia
  Associated with parahippocampal place area (PPA) lesions

- Allocentric disorientation
  Hippocampal damage?

Epstein & Vass (2013) Brain
Visual agnosias

*Often associated with temporal lobe damage or dysfunction*

- Apperceptive visual agnosia
- Associative visual agnosia
- Prosopagnosia
- **Topographical disorientation** | some types of disorientation
- Cerebral achromatopsia
- Akinetopsia
Visual agnosia

*Difficulty in recognizing visual objects*

*Note that a patient with anomia may also respond in a similar way, but they can describe what the object is used for, whereas a patient with agnosia cannot.*
Lissauer’s (1890) two-stage framework

For understanding disorders of object processing

Object

\[ \text{apperception} \]

\textit{Form stable perceptual representation}

\[ \text{association} \]

\textit{Access stored knowledge}
Elizabeth Warrington’s view on apperception

What it must be like to see the world with apperceptive agnosia
Test of early visual processing | apperceptive agnosia

Incomplete letters from Warrington’s VOSP battery
Lissauer’s (1890) two-stage framework

For understanding disorders of object processing

Object

apperception

Form stable perceptual representation

association

Access stored knowledge
Copying in apperceptive agnosia

Poor because perception is poor
Shape matching in apperceptive agnosia

*Poor because perception is poor*

Figure 3.
The shape matching ability of apperceptive agnostic patients. On the left is a set of rectangles matched for overall area, which were presented pairwise to Mr. S. to be judged same or different in shape. He was unable to discriminate all but the most distinctive, and made errors even with these. On the right are a set of rows containing a target shape (left) and a set of four choices to be matched with the target shape. Mr. S.'s answers are marked.
Copying in associative agnosia

*Can be good although slow because perception is present but stripped of meaning*
Patient DF

*Perhaps the most famous visual agnosic*

- She was unable to identify shapes but able to recognize objects by colour
- Could not identify edges, line orientation or figure from ground – she has visual form agnosia (a type of apperceptive agnosia)
- Poor direct copying
Patient DF

*But she could shape her hand correctly to grasp different objects*

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**Preserved ability to “grasp shape” in D.F.**

Pebble like objects (Blake’s shapes) used to test robotic grasping abilities

Curvature of different parts of the object to be taken into account

D.F. grasping not very different from controls

**Ability to negotiate obstacles**

Unfamiliar environment with obstacles of different height

As in controls, D.F. was able to step over the obstacles leaving just enough clearance to ensure that the foot does not touch the obstacle

**Matching and posting task**


Matching: turn card to match the orientation of the slot

Posting: reach out and “post” the card into the slot

D.F. undistinguishable from controls in the posting task only

**How do we know that D.F. symptoms are not due to general “poor” vision that makes it impossible for her to process detailed aspects of shape but leaves enough visual processing ability to carry out generalised actions towards objects?**

Double dissociations can be used to rule out the possibility that deficits in a particular task are just related to task difficulty

Need for evidence from another syndrome presenting opposite patterns of impaired and spared functions

**Optic ataxia as part of “Balint’s syndrome”**

Therefore, specific problems in translating vision into action

Inability to reach for visible objects with right hand

Deficit could not be purely perceptual since patient could reach for objects using left hand

Ability to reach for specific body parts on request

Deficit cannot be purely motor as reaching can be achieved using tactile or proprioceptive information

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**Balint (1909)**

Patient with bilateral damage to parietal (and dorsal occipital) cortex

Holmes (1918) considered the deficit as mainly spatial, i.e., inability to use egocentric co-ordinates to locate objects

Symptoms can vary according to the specific damage suffered by different similar patients, e.g., in addition to reaching problems, inability to shift gaze, inability to focus attention on more than one part of a visual scene.
Patient DF

*She could also ‘post’ a letter through different slot orientations correctly*
Patient DF

Bran damage in DF aligns with area LOC (lateral occipital complex)
fMRI studies reveal object, face and place regions

In human ventral visual stream

Grill-Spector Current Opinion Neurobiology 2003 &
One conceptual framework for object recognition

And its breakdown in visual agnosias

A hierarchical, bottom-up model
Patient HJA

A case of ‘integrative’ agnosia
Patient HJA
A case of ‘integrative’ agnosia

- Good identification of elementary shapes
- Very accurate copy of drawings and objects

Copy of an etching of St Paul’s cathedral

Riddoch & Humphreys *Brain* 1987
Patient HJA

A case of ‘integrative’ agnosia

• Good identification of elementary shapes
• Very accurate copy of drawings and objects
• Good semantic memory as indexed by drawing from memory
Patient HJA

A case of ‘integrative’ agnosia

- Good identification of elementary shapes
- Very accurate copy of drawings and objects
- Good semantic memory as indexed by drawing from memory
- Deficit in integrating single features of a stimulus in a coherent fashion
Patient HJA

Difficulties with parsing briefly presented overlapping figures

Whereas associative agnosics can do this

*Even though they might not identify each object*
Framework for understanding object recognition

*From retinal image to object identity in the ventral visual stream*

Image → Local computation of contours → Integration of contours into shapes → Computing object ‘identity’

- *In early visual cortex (V1) - intact in HJA*
- *Intermediate visual areas*
- *Inferotemporal cortex*
One framework for object recognition

And its breakdown in visual agnosias

A hierarchical, bottom-up model

Integrative agnosia

Semantic dementia

Humphreys & Riddoch Cognitive Neuropsychology 2006
Behrmann’s classification for visual agnosias

*But note that now degenerative conditions are far more common causes than strokes*

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<th>Type of Agnosia</th>
<th>Cause</th>
<th>Clinical Manifestation</th>
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<td>Apperceptive agnosia</td>
<td>Stroke, anoxia, carbon monoxide poisoning affecting occipital, parietal, or posterior temporal regions bilaterally</td>
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<td>Integrative agnosia</td>
<td>Extensive extrastriate damage bilaterally or just to the right hemisphere</td>
<td>Able to copy and match stimuli, may even be able to provide verbal description of aspects of input but still fails to recognize object</td>
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<td>Associative agnosia</td>
<td>Both types: usually bilateral infarction of the posterior cerebral arteries but unilateral temporo-occipital damage may suffice</td>
<td>Fails to access stored representation of object structural description</td>
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<td>Impaired access to structural knowledge</td>
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Framework for understanding object recognition

*From retinal image to object identity in the ventral visual stream*

Image

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<td>Integration of contours into shapes</td>
<td>Intermediate visual areas</td>
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<td>Computing object ‘identity’</td>
<td>Inferotemporal cortex → Semantics</td>
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Temporal pole
Semantic dementia

Progressive loss of understanding of meaning

Semantic dementia patients can repeat the word “ostrich” but have difficulty pointing to it. They lack the conceptual knowledge of what an ostrich is.

Semantic dementia

Imaging demonstrates atrophy of left temporal pole – considered by some to be a ‘semantic hub’
Anatomy of achromatopsia and prosopagnosia

Involvement of regions specialized for colour and face processing
Akinetopsia | Motion blindness

Very rare following focal lesions – requires bilateral V5 / MT dysfunction

Poured fluid ‘frozen’ like a glacier, or like viewing the world with strobe lighting
Structure from motion

*Biological motion examples show how sparse motion information can be decoded as an object*
Structure from motion

Sparse motion information can be decoded as an object
Case History
Middle aged man with visual symptoms

- 59 year old right-handed man with a progressive history
- 7 years ago: Frustrated with difficulty reading. “Letters jumbled”
- Seen by several opticians & ophthalmologists: no cause found
- 5 years ago at daughter’s wedding: Daughter realized he was afraid of treading on her gown
- Wouldn’t eat at wedding for fear of making a mess
- Prior to admission: Hit by a car moving slowly
- No visual hallucinations

Copying

Revealed severe constructional apraxia
Describing a scene

Demonstrated simultagnosia

Described parts of the picture piecemeal but had no grasp of what the entire scene depicted
Eye movements while asked to view a scene

Shows he neglects to inspect left and right sides of space
Case History

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- **Examination** | No Parkinsonism. Occasional myoclonic jerks. Optic ataxia – with both arms in both visual fields. Simultagnosia
- **Investigations** | MRI brain abnormal
MRI brain

Bilateral posterior atrophy
Diagnosis: Posterior cortical atrophy (PCA)

A clinical diagnosis which most commonly is found to be due to Alzheimer’s disease at post mortem

Progressive condition with predilection for occipital, parietal and temporal regions with both dorsal and ventral stream deficits

- Often presents to neurologists many years after symptom-onset
- May have been seen by ophthalmologists or psychiatrists previously
- Elements of Bálint’s syndrome
- Visual agnosia
- Memory affected long after visual symptoms
- Many cases have Alzheimer pathology
Posterior cortical atrophy

Associated with progressive atrophy of posterior visual regions

Crutch et al. (2012) *Lancet Neurology*
Case History

Middle aged man with visual symptoms

• 55 year old right-handed bus driver
• Retired because of accidents while driving
• Reported seeing shadowy figures moving beside him
• Sometimes well-formed like people, but often vague
• Never with auditory hallucinations

- **Examination** | Mild reduction of arm swing bilaterally on walking. Neuropsychological assessment demonstrated profound deficits in executive function and visuospatial abilities, as well as fluctuating attention.
Copying

*Revealed constructional apraxia*
Diagnosis: Dementia with Lewy bodies (DLB)

A clinical diagnosis which most commonly is found to be associated with Lewy body pathology

Progressive condition associated with

- Fluctuating attention
- Visual hallucinations
- Visuospatial deficits
- Executive dysfunction
- May not have signs of Parkinsonism at presentation but these often develop
- Probably the same condition as Parkinson’s disease dementia (in the latter motor signs of Parkinsonism precede cognitive changes)
- Both conditions can show marked improvements on cholinesterase inhibitors
- NB: Hallmark of Parkinson’s disease is Lewy body pathology
**Reading**

*Core text is available online on SOLO*

- For overviews: parts of **Chapters 4, 5 and 14** cover ventral and dorsal visual stream and deficits associated with damage to them

- See also separate reading list