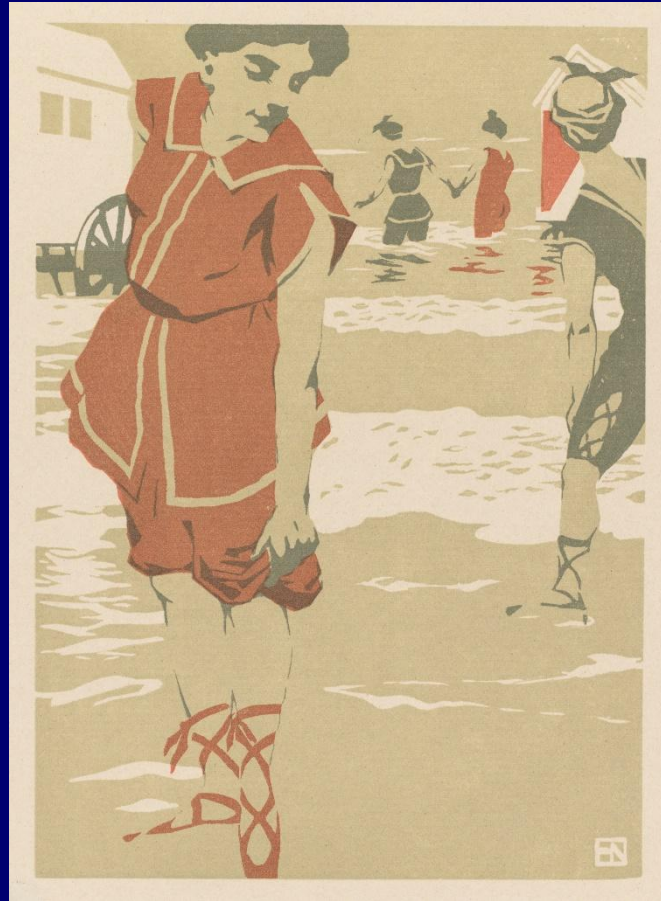


# Disorders of Body Representation



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October 20, 2022

# Objectives

- Identify the types of body representation disorders that can occur
- Describe the various systems the brain uses to create a mental representation of the body
- Discuss how body representation disorders may relate to psychiatric disorders

# What are body representation deficits (BRDs)?

- Impairments in the mental representation of one's own body
- Can occur due to stroke or other focal brain lesion (trauma, tumor)
- Can occur due to neurodegenerative disease
  - This talk focuses on post-stroke BRDs, as best understood form
- Episodic conditions (seizures, migraines) can cause transient, reversible BRDs
- Postulated mechanism: disruption of neural systems subserving the mental representation of the body
- Do disturbances in the brain's representation of the play a role in anorexia nervosa, bulimia nervosa, body dysmorphic disorder, somatic delusions, and control delusions?
  - Possible, but unknown at this time



Seated Figure, Olmec, 12th–9th century B.C.

# Clinical manifestations of BRDs

- Types of impaired body representation include:
  - Misperception of body size or shape
  - Misjudgment of body part location and motion
  - Impaired knowledge of body structures
  - Maladaptive emotions or judgements directed towards the body
    - Or combinations of these
- Deficits in one or more these functions can produce various clinical presentations
- No universally accepted list of conditions that qualify as BRDs



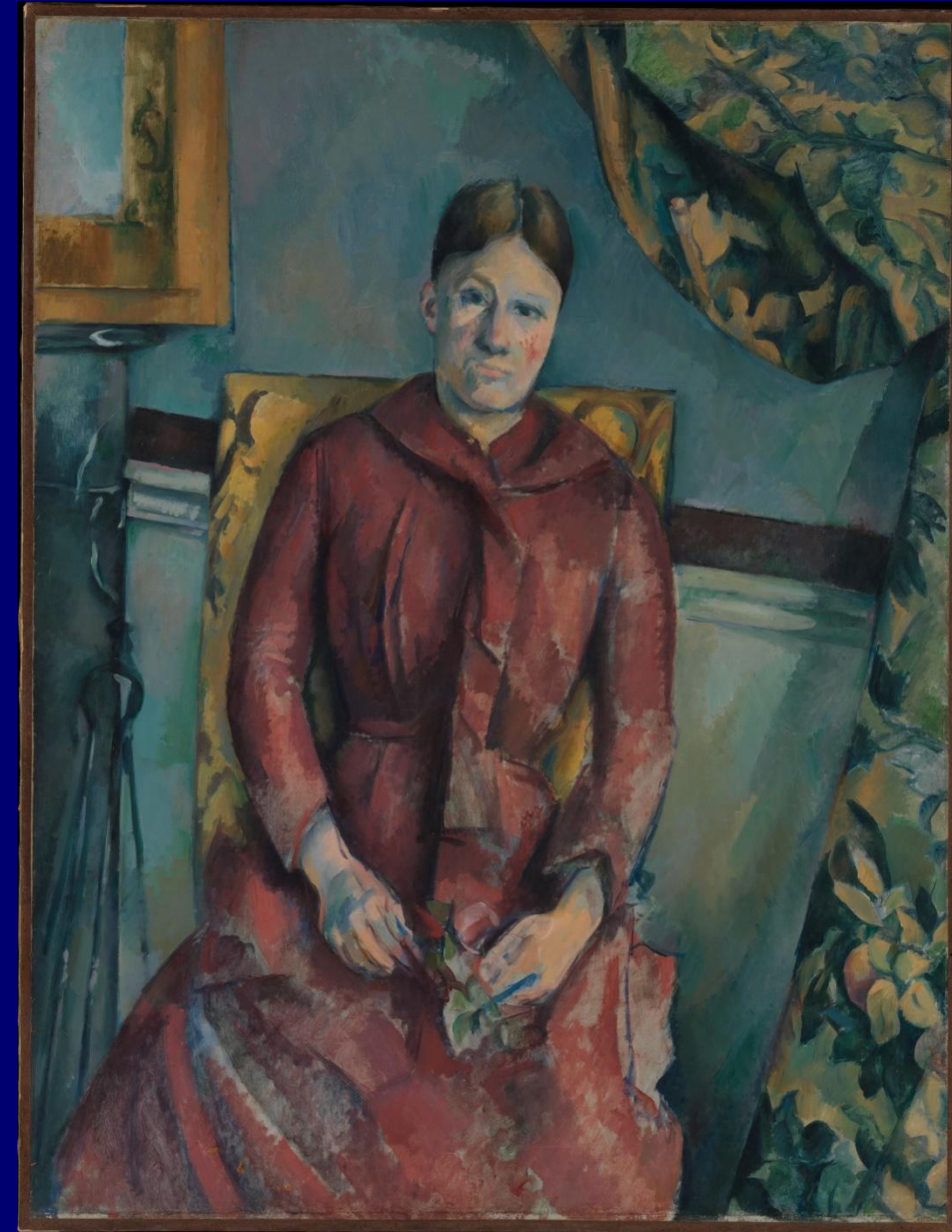
"Emma Van Name", Joshua Johnson, ca. 1805

## Specific BRDs

Altered sense of limb ownership	
Asomatognosia	The sense that one's limbs do not belong to oneself
Somatoparaphrenia	The sense that one's limbs belong to someone else
Alteration of body emotions	Feeling that one's limbs are strange or transformed, and/or hatred for one's limbs
Supernumerary phantom limb	The perception that one has developed new, extra limbs
Alien limb syndrome	The sense that one's limb acts of its own accord or contrary to one's wishes
Personal neglect	Failure to attend to one side of one's own body
Altered perception of body size / orientation	
Microsomatognosia	Underestimation of the size of one's own body/limbs
Macrosomatognosia	Overestimation of the size of one's own body/limbs
Body midline deviation	Perception that one's midsagittal plane is tilted or shifted
Somatotopagnosia	Inability to locate body parts
Autotopagnosia	Inability to locate the parts of one's own body
Heterotopagnosia	Inability to locate the parts of someone else's body
Altered motor function awareness	
Anosognosia for plegia	Lack of awareness of weakness or of the functional consequences of weakness
Illusory limb movements	The feeling that one's paralyzed limb is moving
Body-part-specific anomia	Selective inability to name body parts

# BRDs: a distinct category of cognitive disorders

- Symptoms not due to weakness or sensory loss
- Patients with BRDs also often have other cognitive impairments, such as extrapersonal (hemispacial) neglect, aphasia, or apraxia
  - But, BRDs can occur in the absence of any of these other cognitive deficits
- The false and fixed nature of BRDs may appear delusional, but usually these patients do not show any evidence of psychosis regarding non-body topics



"Madame Cézanne in a Red Dress", Paul Cézanne, 1888–90

# BRDs are common, but poorly understood and often overlooked

- Much less investigated and much less well-understood than other post-stroke cognitive deficits such as aphasia or extrapersonal neglect
- Often mistakenly believed to be rare
  - Symptoms of BRDs may seem very odd, so patients may not volunteer these experiences due to fear of being thought "crazy"
  - Clinicians rarely ask about them
  - Few validated assessment tools
- Some studies have found that BRDs typically resolve after the acute post-stroke period, but some reports describe BRDs lasting for years—so don't always spontaneously resolve



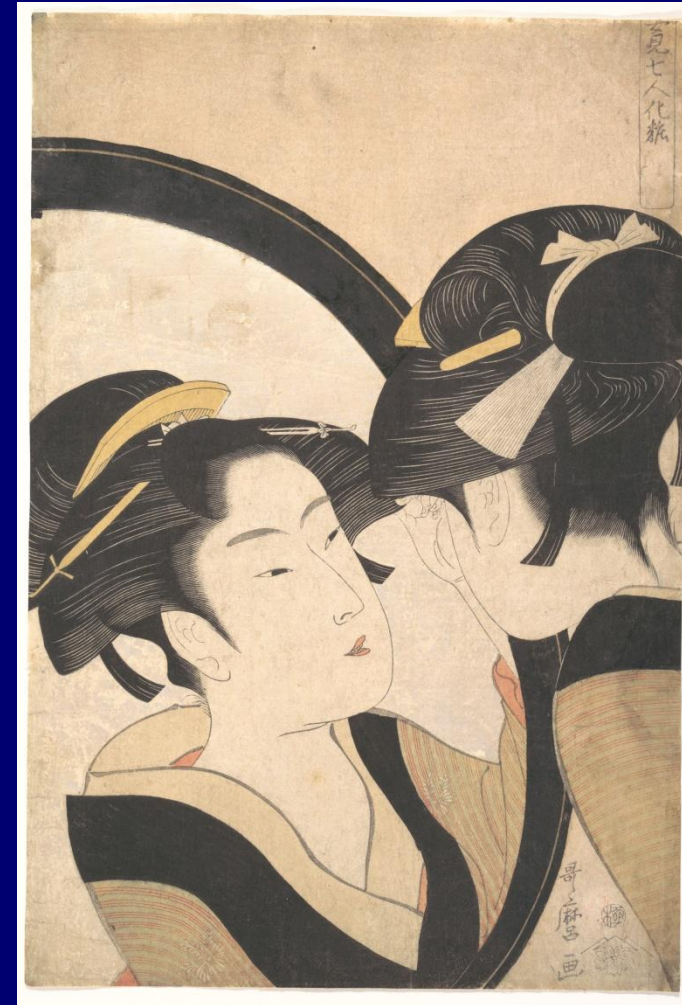
"Main Street of the Yoshiwara on a Starlight Night",  
Utagawa Kunisada II, 1852–64

## Post-stroke BRDs are common

	Population	Reported prevalence (%)
Altered sense of limb ownership	Right hemisphere stroke	15 - 100
Alteration of body emotions	Right hemisphere stroke	4 - 38
	Left hemisphere stroke without significant aphasia	23
Supernumerary phantom limb	Any stroke	< 2
Alien limb syndrome	ACA stroke (right or left)	44 - 100
Personal neglect	Right hemisphere stroke	17 - 96
	Left hemisphere stroke	52
Longitudinal body axis rotation	Right MCA stroke	60
	Left MCA stroke	25
Anosognosia for plegia	Right hemisphere stroke	15 - 58
	Left hemisphere stroke	2 - 14
Illusory limb movements	Right hemisphere stroke	4 - 55
	Left hemisphere stroke without severe aphasia	<5 - 22
Body-part-specific anomia	Cerebrovascular lesions (right or left) with left language dominance	7

# How does the brain represent the body?

- Several different systems may interact to produce our mental representations of our own bodies
- Body semantics: names and functions of body parts and the accessories associated with them
  - “This is my foot; I use it for walking; it can wear a shoe”
- Body structural description: knowledge of the shape, size, and spatial relationships of body parts
  - “My foot is at the end of my leg”
- Body schema: awareness the location and position of body parts as they move in space
  - Awareness of how to position and move one’s arm and wrist to serve a tennis ball
- Body emotions: emotions and value judgements about the body
  - “I hate my left arm; it feels weird and I want to cut it off”



“Naniwa Okita Admiring Herself in a Mirror”,  
Kitagawa Utamaro, ca. 1790–95

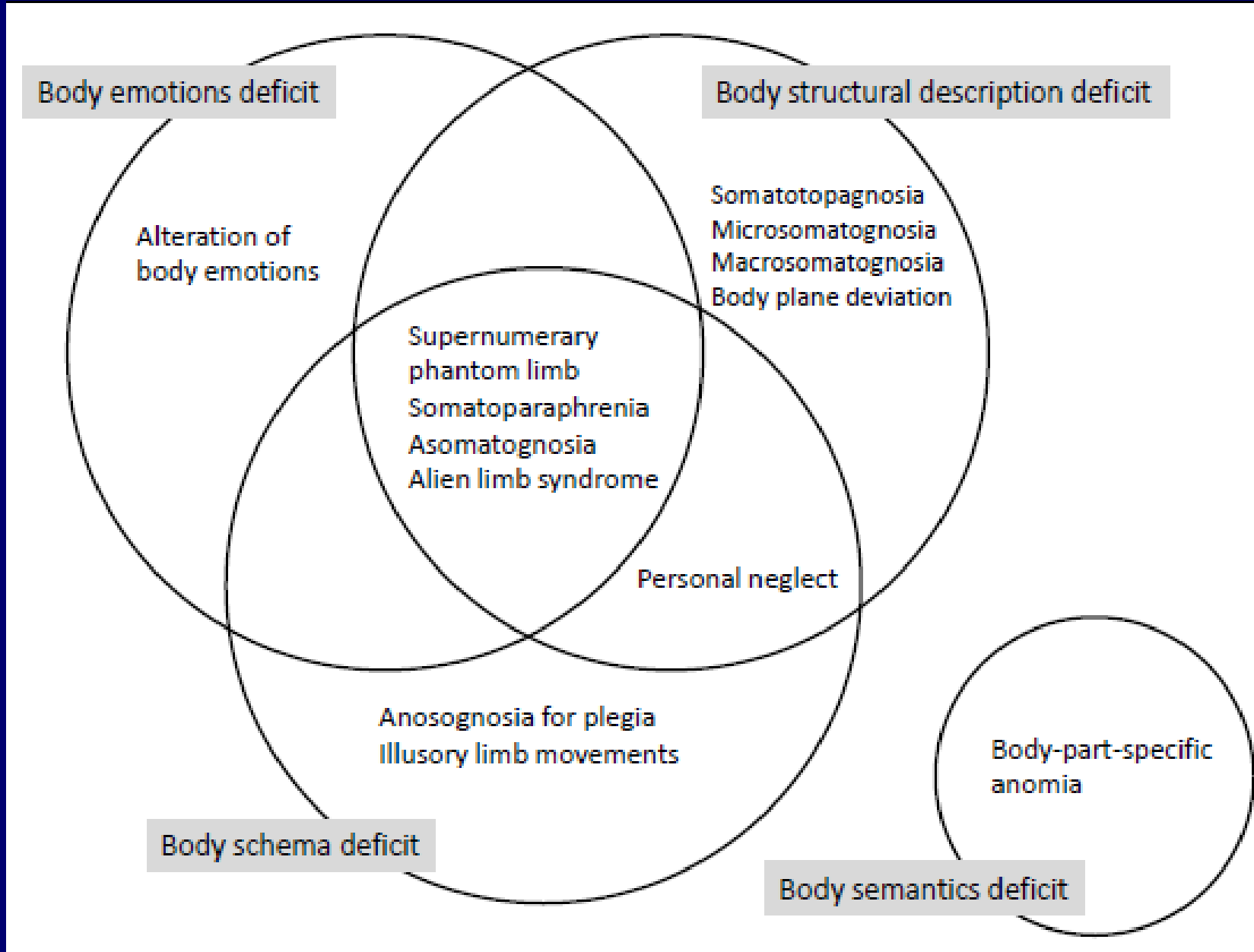
# Damage to body representation systems may produce BRDs

- Body semantics, body structural description, body schema, and body emotions usually work together to produce an integrated, accurate representation of one's own body
- Structural or functional damage to different brain regions can produce isolated impairment in any one of these systems, without necessarily involving the others
  - Experiments in patients with stroke show that these systems are independent and can be independently impaired
  - Some patients may have deficits in more than one of these systems



Figure of a Woman, Egypt, 5th century

# Body representation systems and BRDs





## Specific Body Representation Deficits

Seated Figure, Mexico, 3rd–8th century

# Altered sense of limb ownership (ASLO)

- Encompasses both asomatognosia and somatoparaphrenia
- Usually occurs with left limbs, following right hemisphere stroke
- Asomatognosia: the sense that one's limbs do not belong to oneself
  - Patients say they don't know who the limb belongs to; when asked directly if the limb is theirs, they say no
- Somatoparaphrenia, the sense that one's limbs belong someone else
  - Patients disavow ownership AND attribute limb to someone else: e.g., their spouse, another family member, or a cadaver
- Asomatognosia and somatoparaphrenia likely exist on a continuum, with somatoparaphrenia a more severe form of ASLO



Statuette of Phrixos, Roman, 1st century BC

- Some patients report there are animals inside the limb, or that the limb detaches from them and runs around the room of its own accord
- Patients may describe their limb as “fake” or “dead”
  - One patient reported that his affected hand was actually an armrest
- Less severe forms of ASLO: patients recognize their limbs are theirs, but:
  - Left limbs seem strange, changed, or transformed somehow;
  - Personification: giving their limb a name or talking about it as a separate entity
    - One patient called her left arm by her deceased sister’s arm



“Dutchwoman with Leopard:, Utagawa Yoshimori , 1860

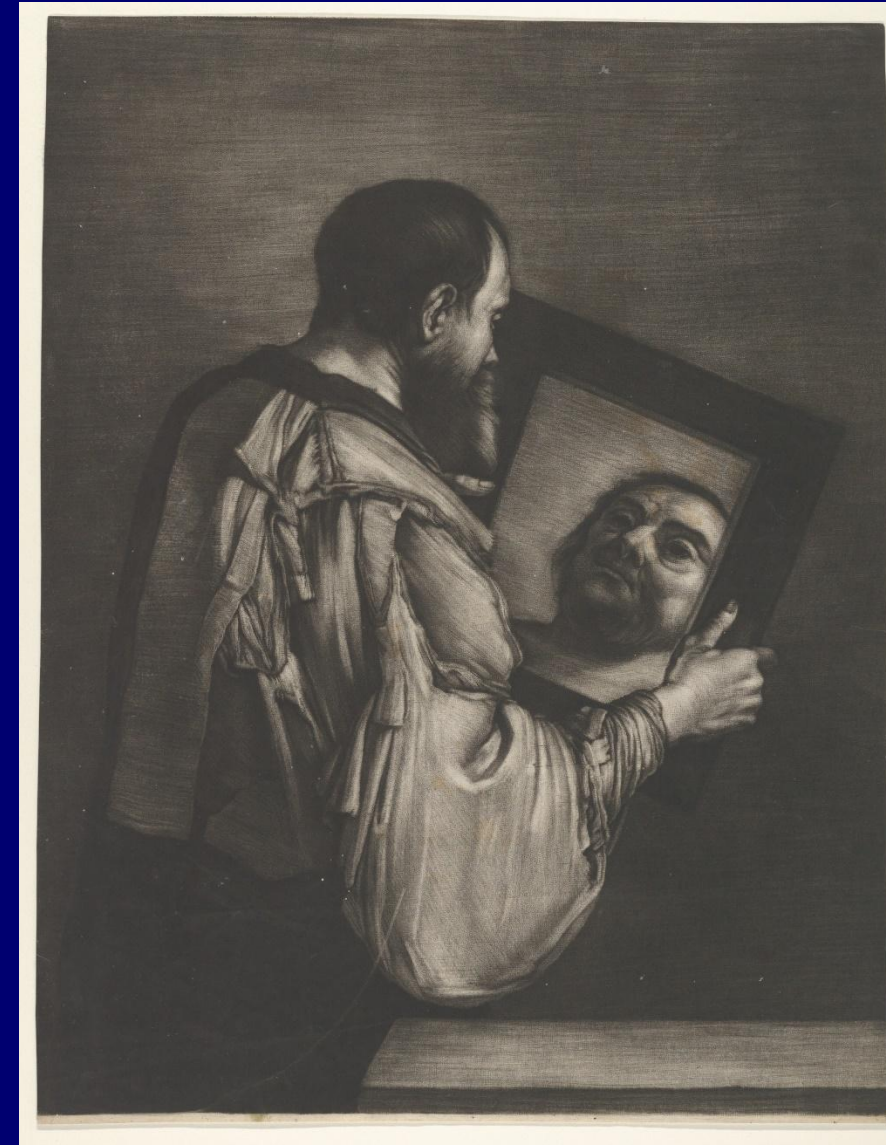


Standing Male Figure, Mexico,  
5th–10th century

- Can temporarily talk some patients with ASLO out of their false beliefs regarding limb ownership, but most maintain their disavowal with delusional intensity.
  - If asked to visually trace the location of their limb and see that it is attached to their body, patients generally admit that the limb is attached to them, and often agree that it seems strange that a limb not belonging to them is attached to their trunk, but still insist that the limb is not theirs
- However, patients with ASLO usually do NOT show any other psychotic features
  - Can reason correctly about topics unrelated to their body or their stroke

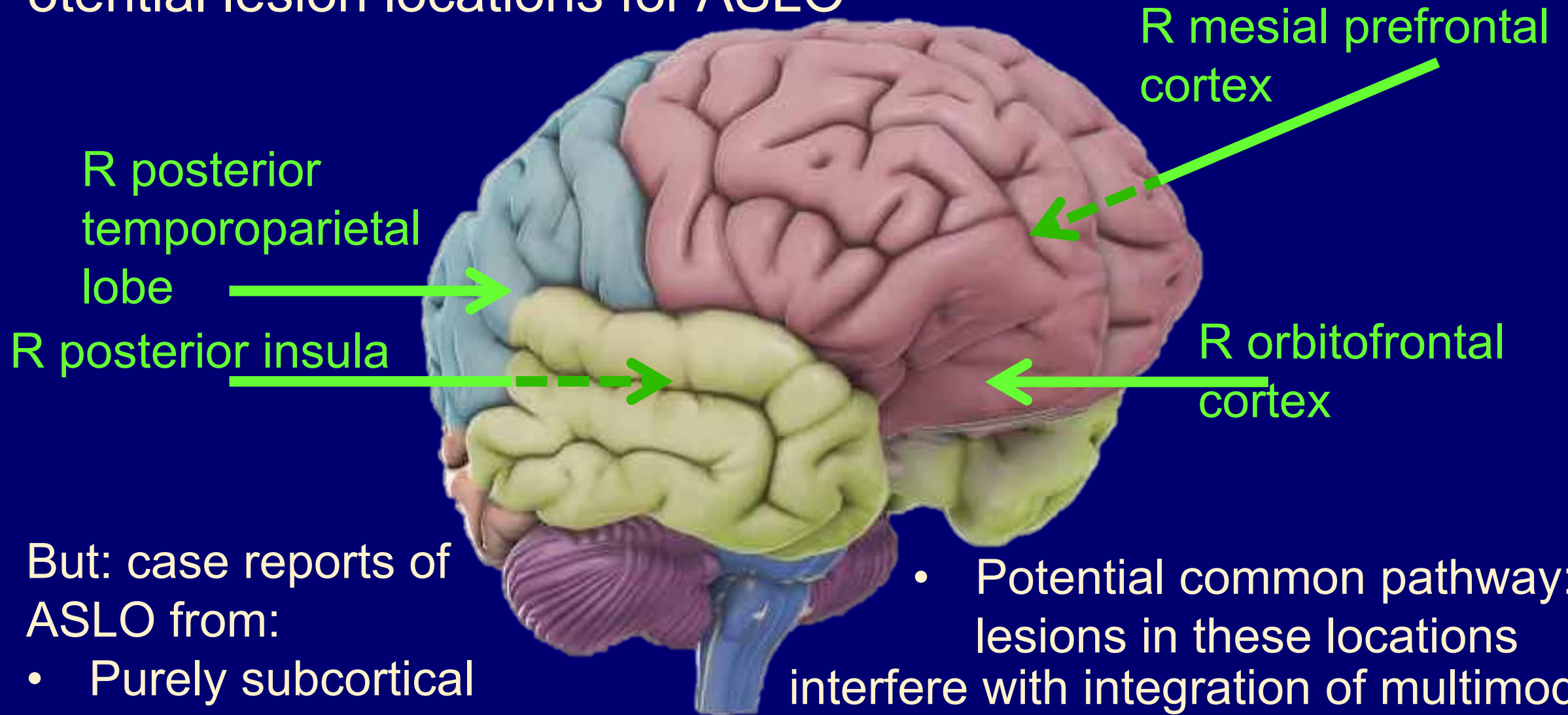
# Timecourse and resolution of ASLO

- Usually lasts days to weeks following stroke
  - Can fluctuate during that time
- In some cases persists years after the stroke
- Patients whose frank ASLO has resolved may still disavow ownership of their left-sided limbs when looking in a mirror, even if they know their limbs are theirs at other times
- Suggests that mental representation of their own bodies did not fully normalize
  - May have compensated for the brain damage that caused ASLO, but the mirror input conflicts with the body representation produced by the compensatory mechanisms and causes confusion



"Socrates Looking in a Mirror", Bernard Vaillant, 17th century

# Potential lesion locations for ASLO



- But: case reports of ASLO from:
  - Purely subcortical stroke
  - L-sided lesions in R-handed patients

- Potential common pathway: lesions in these locations interfere with integration of multimodal sensory and motor information needed to produce an accurate mental representation of the body

# Alteration of body emotions (ABE)

- Even in the absence of other BRDs, patients with stroke may experience striking and distinct changes in their emotions regarding their contralesional limbs
  - Limbs feel strange, or feel transformed or changed somehow (sometimes considered a form of ASLO), in a way that goes beyond the changes associated with weakness, sensory loss, or ataxia



- Misoplegia: hatred of contralesional limbs
  - Some individuals go so far as to say they would like to cut them off, and in extreme cases may even try to do so
- ABE fairly common, but rarely asked about or investigated
  - Lack of data makes it difficult to determine neuroanatomic localization

# Supernumerary phantom limb

- Supernumerary phantom limb: the sense that you have developed an extra limb, resulting in three limbs on the affected side
  - Most commonly occur on left side of body, following a right hemisphere stroke
    - Occasionally R-handed patients develop R-sided phantoms following L-sided CVA
  - Patients may perceive either a whole extra arm, leg, or both, or just an extra portion
  - Some patients report that the extra limb just appeared
  - Others give elaborate explanations: ie, someone else attached the extra limb
  - Extra phantom limbs may be consistently present, or may vanish and return

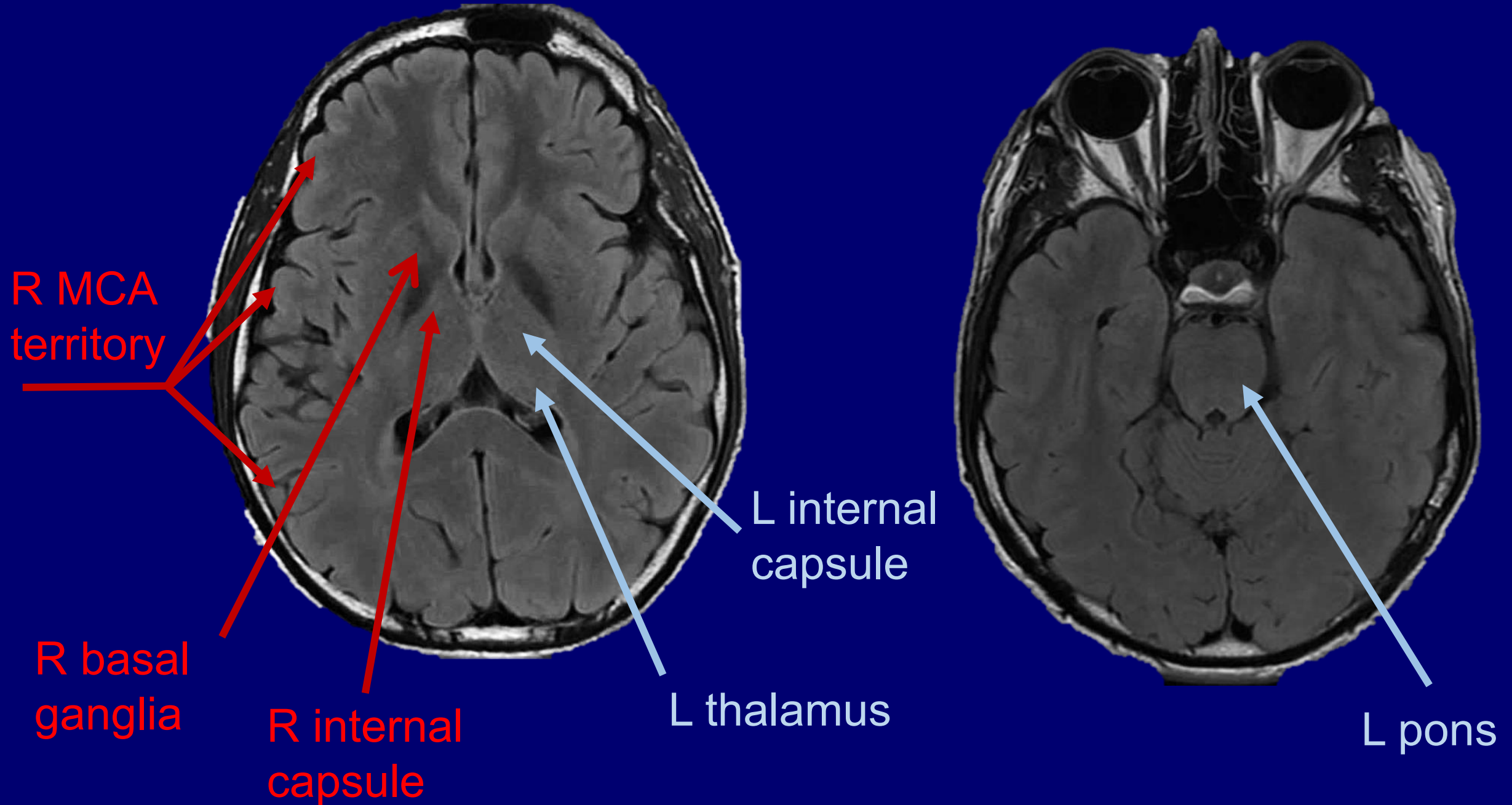


Right Arm of a Statuette, Roman, 1st - 2nd century

- Some patients retain insight that they do not really have a new extra limb, but others do not
- Some patients both see and feel their extra limbs, but others only feel them
- Subthreshold forms of supernumerary phantom limb may also exist: ie, transient misperceptions of the position or movements of an actual limb
  - May be more common than sustained sensation of extra phantom limb
- Supernumerary phantom limbs can begin immediately following stroke, or after a delay (up to five months later in the literature)
- Supernumerary phantom limbs may resolve following the acute phase of stroke, or last for over a year

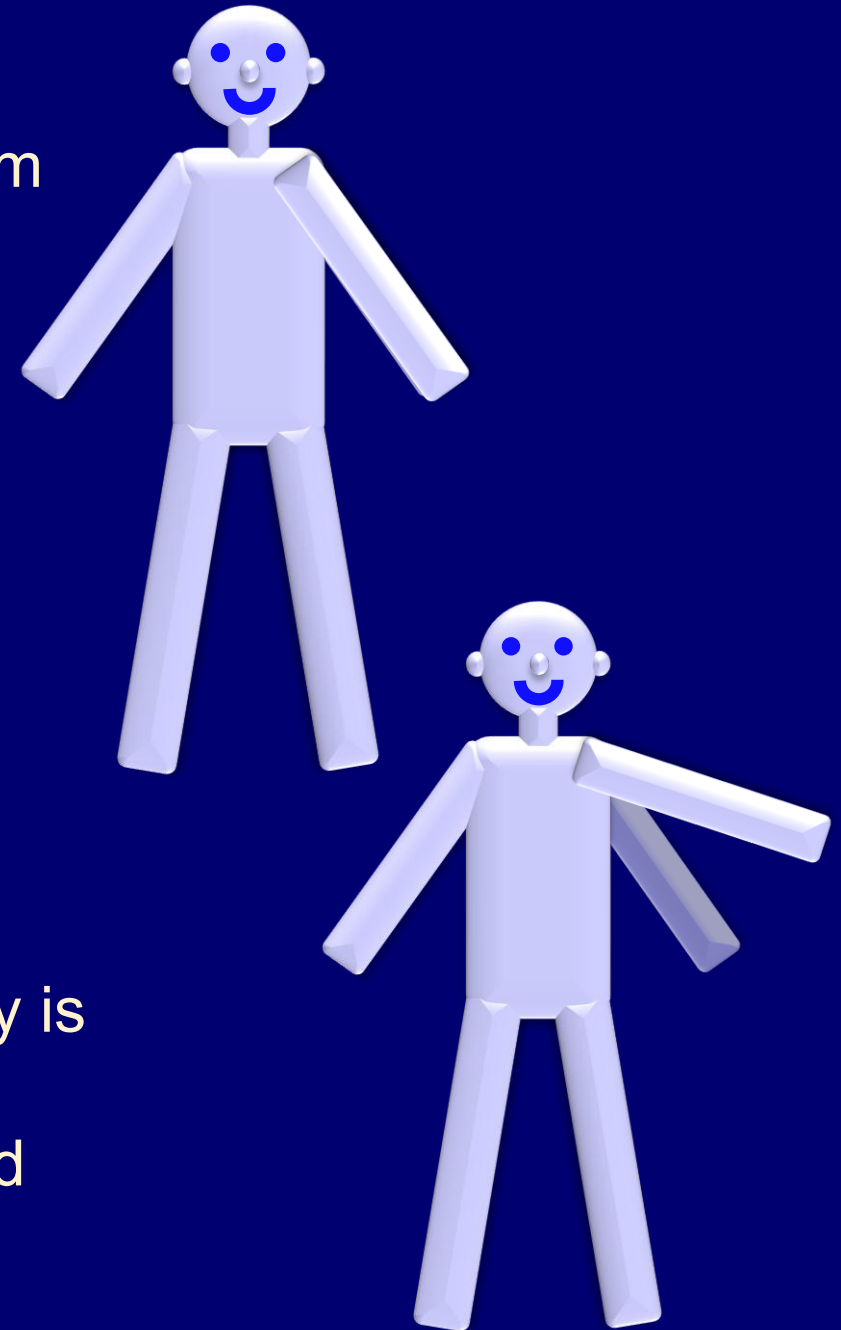


# Structures associated with **left-sided limb** and right-sided limb phantoms



## Etiology of supernumerary phantom limbs remains unknown

- One theory: supernumerary phantom limbs derive from a mismatch between planned and experienced motor activity
- Usually, when we attempt to move our limbs, we experience:
  - Motor planning which directs the movement
  - Sensory feedback which notifies us the move was successfully completed and updates our representation of limb position.
- When these 2 systems are disconnected, patients may experience a discrepancy between where they intended to move their limb and where their limb really is
- The brain may resolve this discrepancy by generating the perception of one limb in the intended location and one limb in the actual location, giving rise to a supernumerary phantom limb



# Alien limb syndrome

- Alien limb syndrome: the sense that one of your limbs does things of its own accord, without you willing the actions
- Often called “alien hand”, but can occur in lower extremity as well
- Manifestations of alien limb syndrome:
  - Stimulus-bound reaching behaviors: patient grabs at objects in the environment without wanting to
  - Intermanual conflict: the alien limb reaches over to interfere with tasks performed by the other limb, such as unbuttoning a shirt the "good" hand is trying to button



“Clasped Hands of Robert and Elizabeth Barrett Browning”,  
Harriet Goodhue Hosmer, 1853

- Purposeful but unintended acts: alien limb engages in unwilled behaviors such as writing with a pencil, (unwanted) public masturbation, or attacking the patient

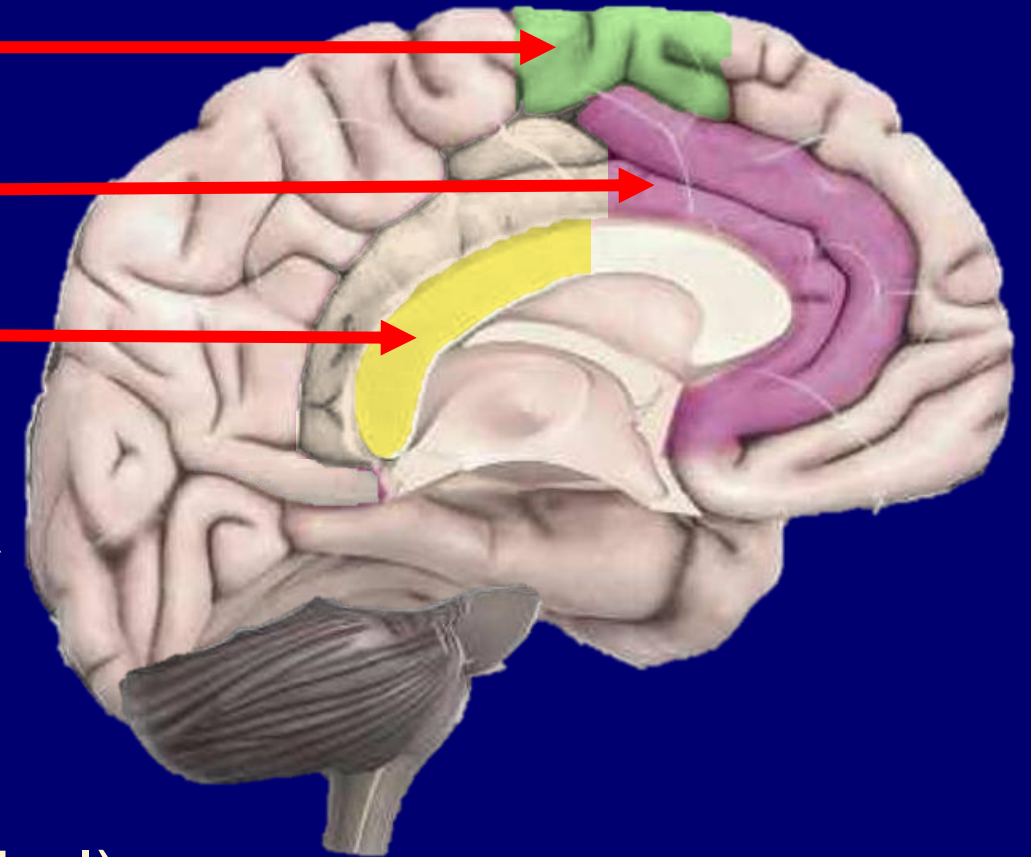
- Duration of alien limb syndrome: few hours to 3 years or longer
- Anatomy: can occur following either a left-sided or a right-sided lesion, but typically involves midline structures
  - Strongly associated with ACA stroke

R or L supplementary motor area

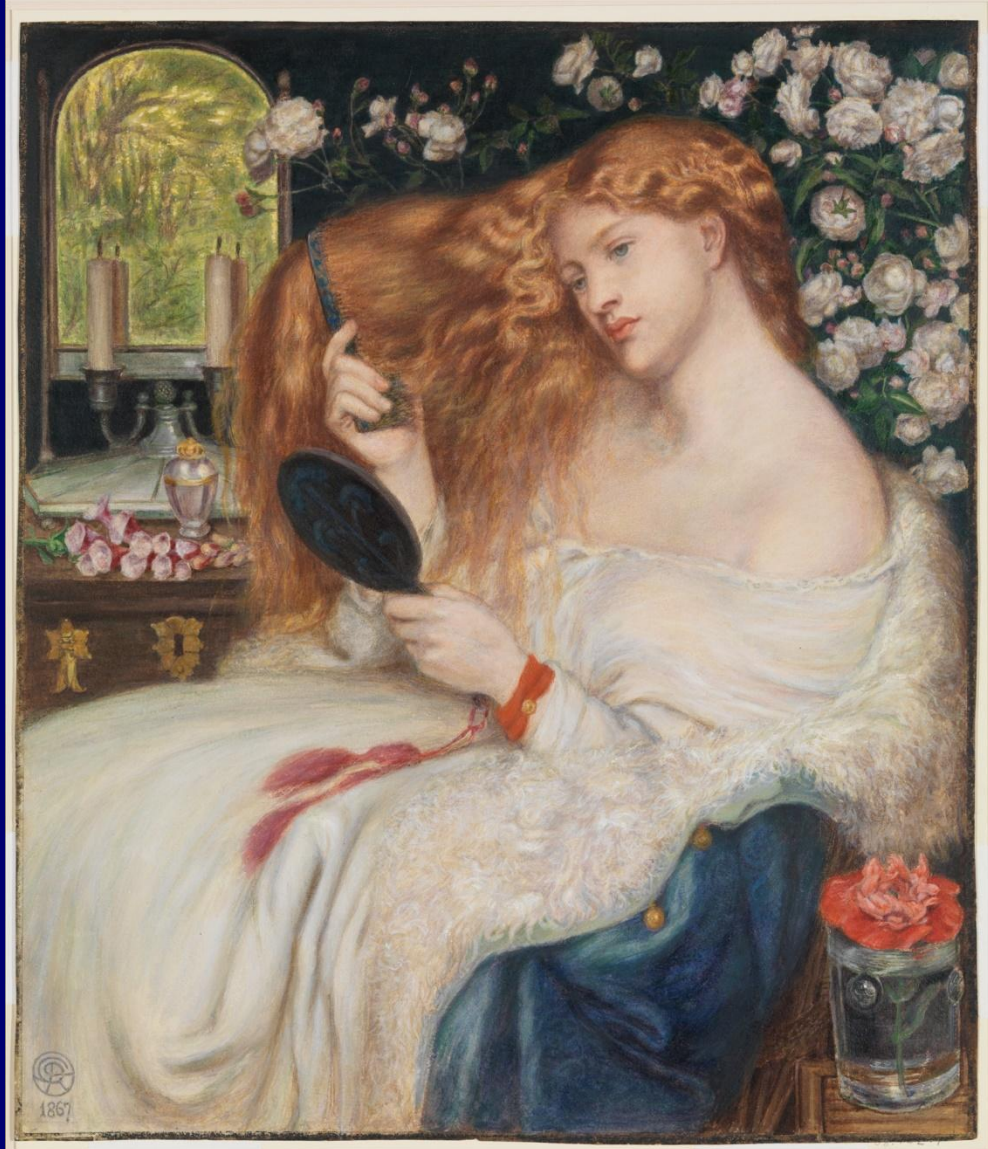
R or L anterior cingulate cortex

R or L posterior corpus callosum

- Potential etiologies:
  - Disrupted communication between R and L hemispheres, impairing ability for both hands to work together
- Motor activity planned in R (nonverbal) hemisphere not communicated to L (verbal) hemisphere, so activity perceived as unwilled



# Personal Neglect



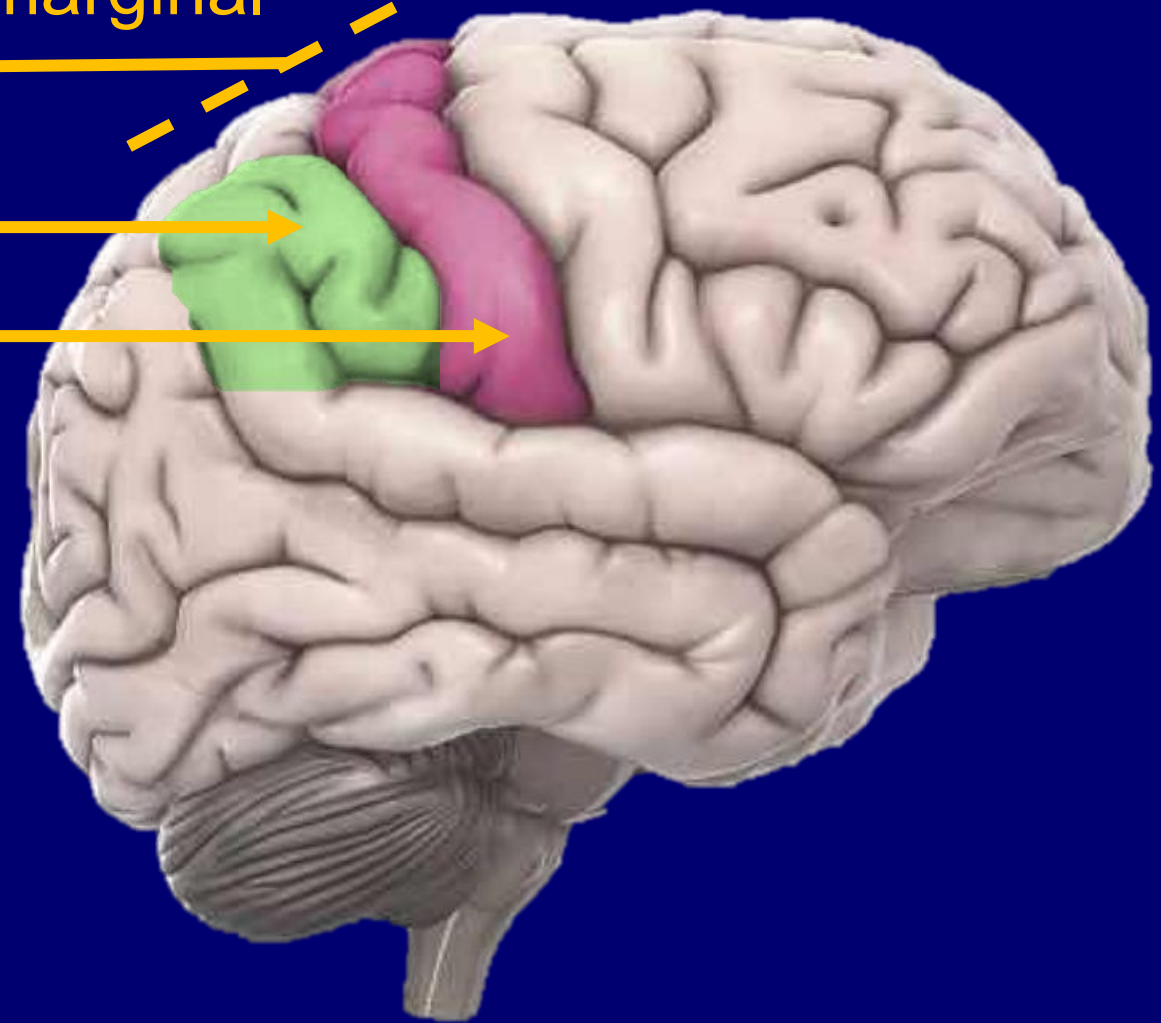
- Failure to attend to one side of your body—usually the left side, following a right hemisphere lesion
- May fail to dress /groom left side of body
- Personal neglect and extrapersonal neglect often occur together, but can have personal without extrapersonal neglect (25% of cases) or extrapersonal without personal neglect (20% of cases)
  - Brain regions involved in personal vs extrapersonal neglect are different
  - Distinct system for body attention?
  - Personal and extrapersonal neglect may co-occur merely because involved regions are both in R MCA territory, so could both be affected by single CVA

# Personal neglect: anatomy and possible etiology

Deep white matter medial to R supramarginal gyrus and R postcentral gyrus

R supramarginal gyrus

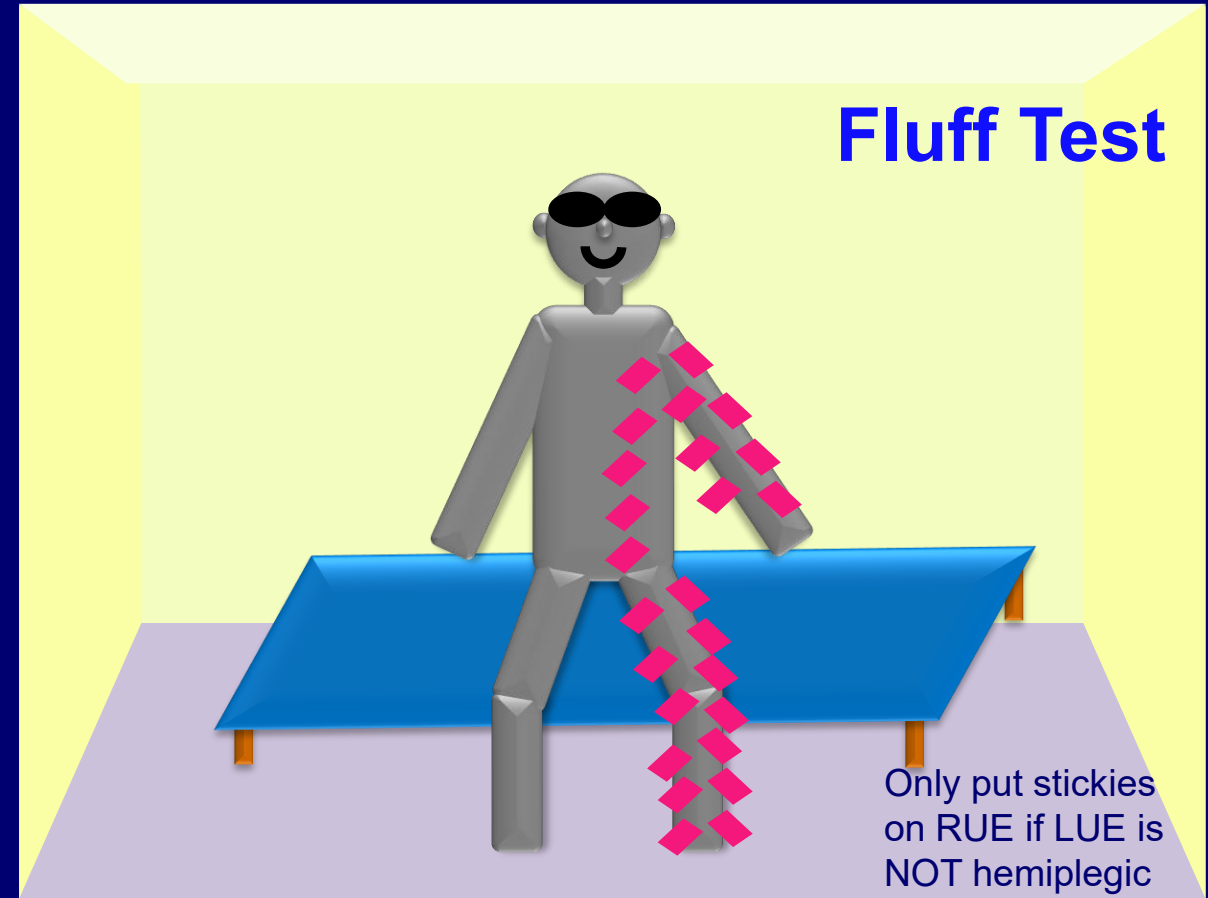
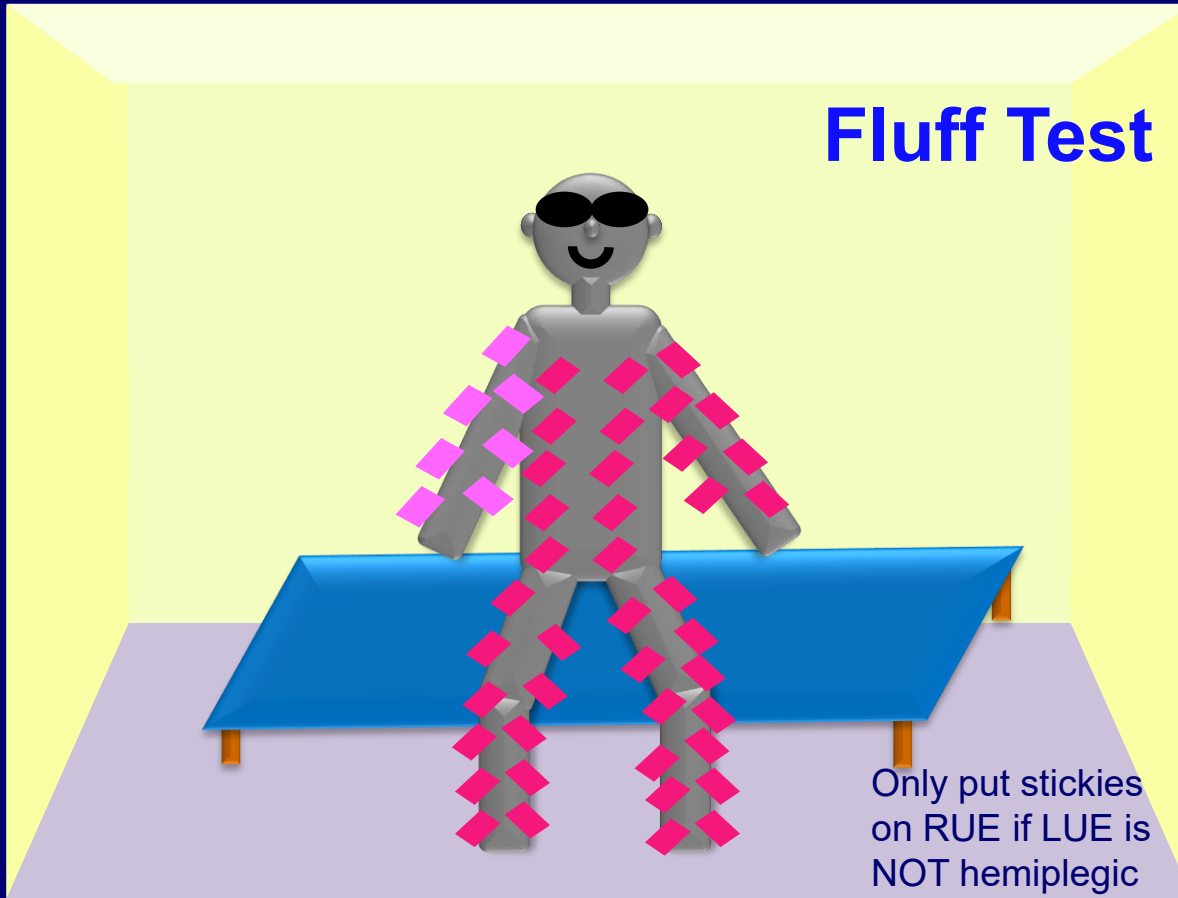
R postcentral gyrus



- May result from disconnecting regions for somatosensory function from those that represent where the body is in space
  - Thus impairing ability to attend to the left side of the body

# Assessment of personal neglect

- One of the few BRDs with validated assessment measures



- Fluff Test and Comb and Razor / Compact Test each assess different aspects of personal neglect
- May show impairment on one and not the other; doing both increases sensitivity

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# Altered perception of body size and orientation

- Altered perception of body size
  - Microsomatognosia: the sense that body parts on one side have become smaller
    - Potentially related to personal neglect?
      - Neglected things appear smaller
      - But reported to occur with R insula lesion (not known to be associated with neglect)
  - Macrosomatognosia: the sense that body parts on one side have become larger
- Described in a few case reports, no systematic study
  - Prevalence / neuroanatomy unclear
  - Sometimes considered form of ASLO



“Mary, Queen of Scots, with Her Son, James”, Style of 16th-century British Painter, painted 19th century

- Altered perception of body orientation
  - Patients with R hemisphere CVA and extrapersonal neglect may mislocalize their midsagittal plane, perceiving it to the right of its true location
    - Unknown if form of personal neglect or separate phenomenon
  - Longitudinal body axis rotation: perception that the vertical axis of your body is tilted to the side
    - May be more common in R hemisphere CVA, but can occur in L hemisphere CVA as well
    - Can occur in patients who accurately perceive environmental vertical



Madame Jerome Bonaparte, attributed to Thomas Sully, ca. 1805–10

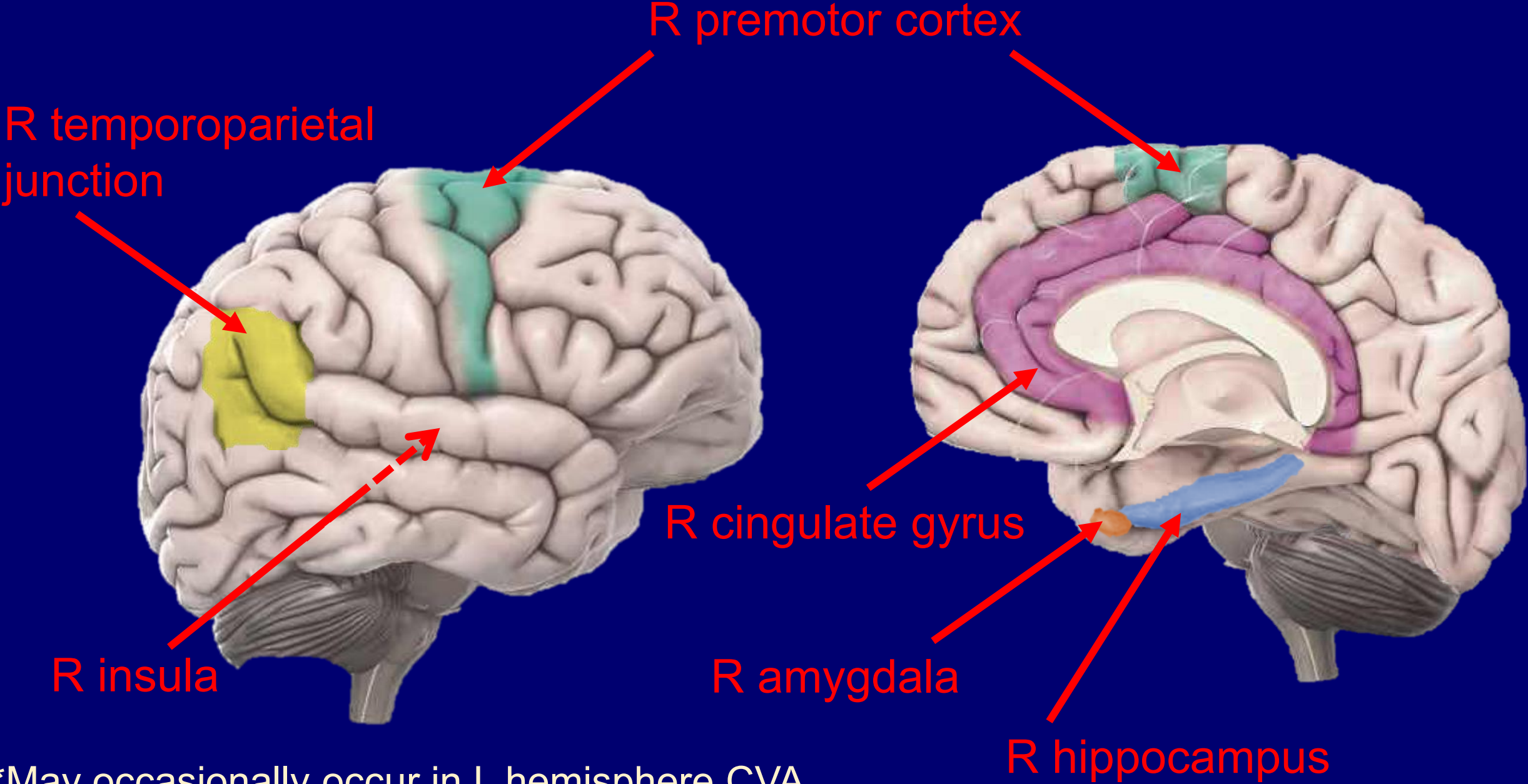
# Altered motor function awareness

- Anosognosia for plegia
  - Unawareness of weakness
  - Or, patients may acknowledge they have weakness but are unaware of the functional consequences of it
    - e.g., admit to lower extremity paralysis but deny difficulty walking
    - May agree that other people with same deficit couldn't walk
- Illusory limb movements: feeling one's paralyzed limbs moving
- Anosognosia for plegia and illusory limb movements both involve erroneous appraisal of one's own motor activity
  - Some patients experience only one or the other, showing that the 2 conditions can dissociate and are thus distinct entities



"Dancer Onstage", Edgar Degas, ca. 1877

# Anosognosia for plegia: neuroanatomy



\*May occasionally occur in L hemisphere CVA

# Anosognosia for plegia: potential etiologies

- One theory: arises out of a failure to attempt movement and thus generate feedback about the movement's success or lack thereof
  - How do we know if we can move our limbs or not?
    - By trying to move them and receiving sensory feedback as to whether they actually moved
  - Anosognosia and motor neglect (failure to attempt movement with the contralesional side) both strongly associated with R hemisphere CVA
  - If patients with anosognosia for plegia also have motor neglect, may never actually try to move L side
    - Thus no discordance between expected result and actual result of their (nonexistent) attempted movements
      - So patients fail to discover their weakness.



Standing Smiling Figure, Remojadas,  
7th–9th century

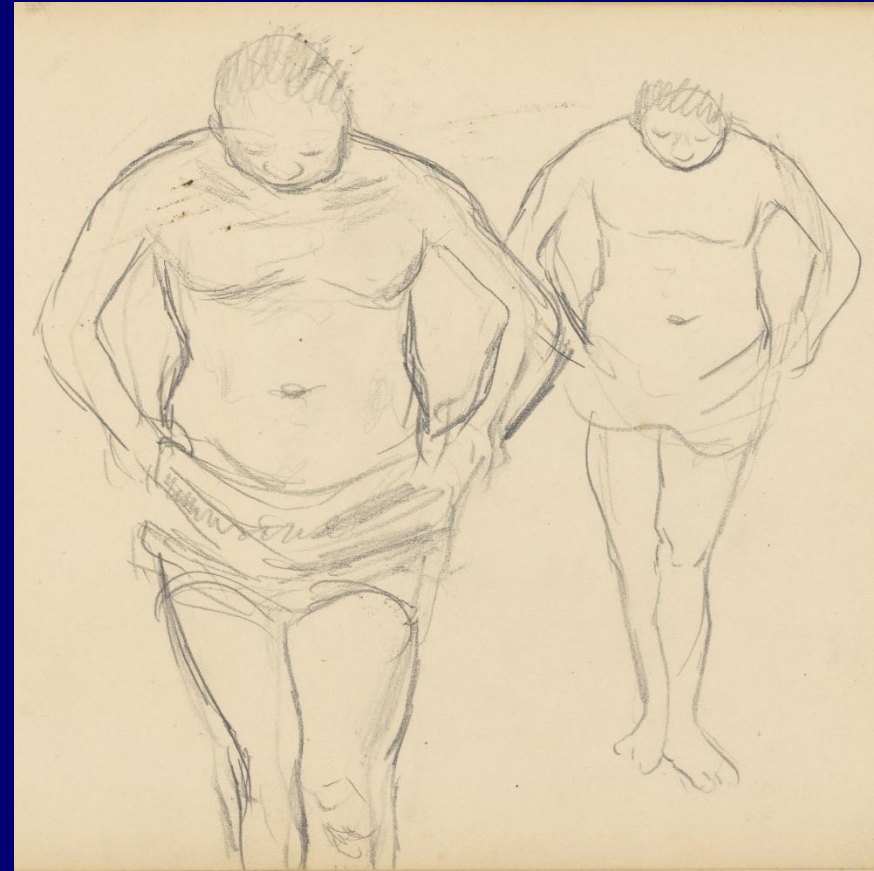
- Another theory: overlap between anosognosia for plegia and ASLO
  - Up to 92% of patients with anosognosia for plegia also have ASLO
  - Ability to know if we can move our limbs is closely linked to ability to know our limbs belong to us
    - We expect to know when we intend to move our own limbs
    - We don't expect to know if someone else intends to move their limb unless that person tells us
    - We don't expect someone else's limb to move when we will a movement
  - Thus, individuals with anosognosia for plegia may notice there is no movement in a limb, but not interpret this lack of movement as weakness: logically, not seeing someone else's limb move does not imply that they are paralyzed



“In the Studio”, Alfred Stevens, 1888

# Somatotopagnosia

- Inability to locate body parts
  - Autotopagnosia: inability to locate your own body parts
  - Heterotopagnosia: inability to locate someone else's body parts
  - Or both (the most common form)
- NOT a language problem
  - If examiner points at a body parts and asks patient to name it, patient can usually do so correctly
  - If examiner names a body part and asks patient to point to it, patient can't do so
  - Deficit still present when tested nonverbally
  - Patients may correctly describe the location of body parts, but still be unable to actually find them



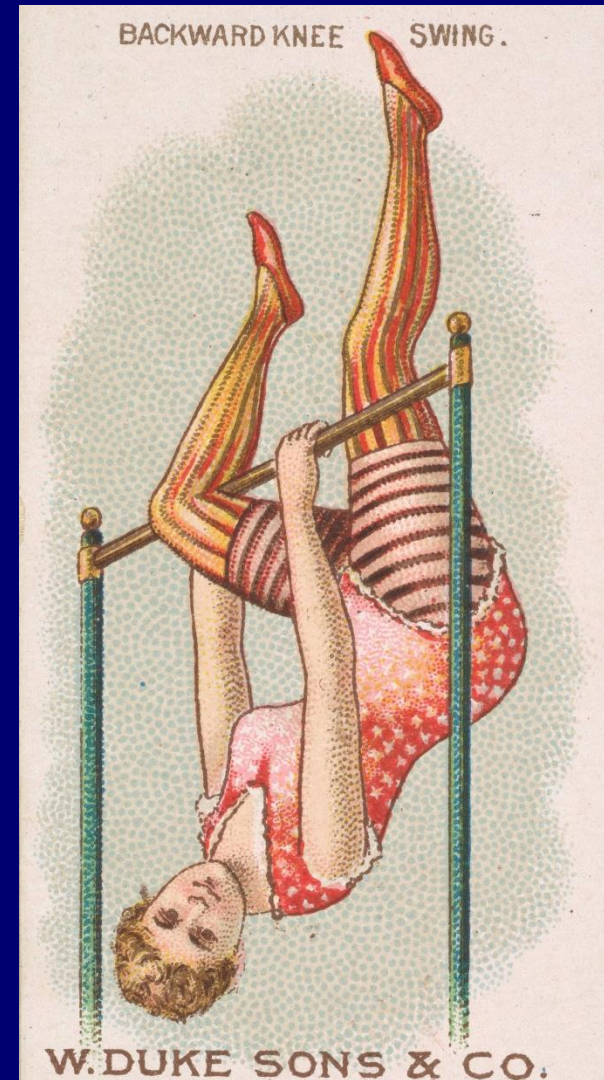
Copies of Cézanne's Bathers, Edgar Degas, about 1877

- Somatotopagnosia = loss of the mental representation of body part locations, NOT anomia
- But: even with nonverbal test administration, body parts whose names are lower frequency words are the body parts most difficult for patients with somatotopagnosia to identify
  - e.g.: Patients worse at pointing to their hip than their hair
  - “Hip” is lower frequency word than “hair”
- Theories:
  - Body parts which people explicitly think about less often—and thus whose names occur less frequently in ordinary discourse—become less firmly embedded in the explicit topographic body representation map
  - These body parts are less useful when thinking explicitly about our bodies, so we refer to them less often when speaking



“A Woman at Her Toilet Seated before a Mirror, Having Her Hair Combed by a Kameyui”, Kitagawa Utamaro, undated

- Errors in somatotopagnosia typically not random
- Most common error: pointing to an incorrect nearby body part
  - e.g.: pointing to the hip when asked to point to the knee)
  - Suggests that somatotopagnosia results from damage to body structural description causing patients to “misplace” their body parts, rather than a general destruction of body representation
- Somatotopagnosia usually results from a left hemisphere lesion, but deficit is bilateral
  - Patients equally impaired at identifying left side and right side body parts
- May be associated with left parietal lesions, but other locations also reported, and neuroanatomic basis still unclear
- Standardized assesment for somatotopagnosia exists



“Backward Knee Swing”, from the Gymnastic Exercises series (N77) for Duke brand cigarettes, Date:1887

- Somatotopagnosia is a loss of explicit knowledge about body part locations
  - Patients with isolated somatotopagnosia do not show deficits in praxis or other tasks which require them to move their body parts to accomplish a goal.
  - Patients with apraxia do not typically have difficulty identifying the locations of their body parts.
  - This dissociation supports the idea that explicit and implicit body representations are distinct—can have one impaired while the other is intact



L'Arlésienne: Madame Joseph-Michel Ginoux, Vincent van Gogh, 1888–89



- A human-body-part-specific deficit, not a general difficulty in identifying parts of a whole
  - Patients with somatotopagnosia can correctly locate parts of nonliving objects or body parts of nonhuman animals
- Suggests that knowledge of how specific body part locations fit into an overall body structure is a specific cognitive domain, which can be selectively damaged by a lesion in brain areas subserving this function.
  - Dissociation between autotopagnosia and heterotopagnosia implies a division between systems involved in processing the structural description of one's own body and that of someone else's

# Body-part-specific anomia

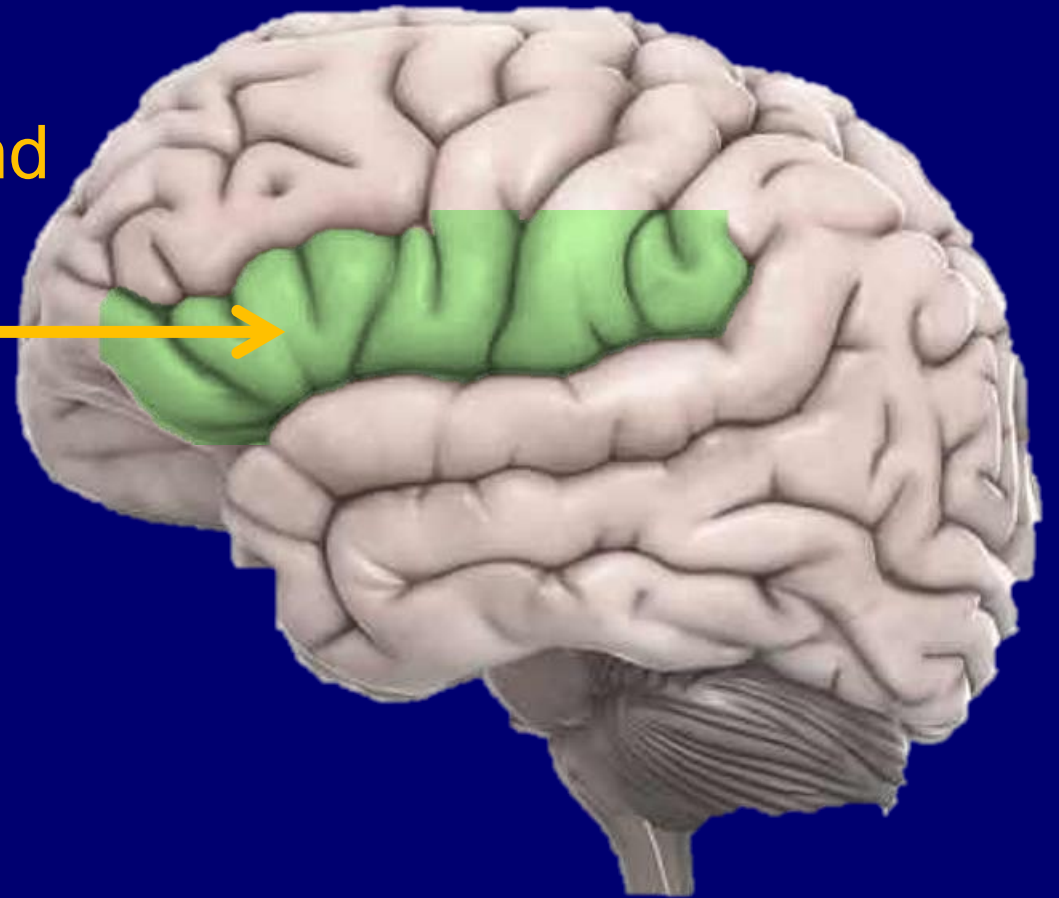


- Selective or disproportionate impairment of body part naming
- Patients typically do not show any evidence of other body representation disorders
- Category-specific anomias can occur for various classes of items
  - Existence of body-part-specific anomia—and the selective sparing of body part names in other cases—supports for the concept of a separate and dissociable body semantic representation.

# Body-part-specific anomia: neuroanatomy

Regions associated in some studies:

Left inferior frontal operculum and  
left anterior/inferior parietal  
operculum





## Body Representation Deficits: Significance and Treatment

# Do BRDs affect stroke outcomes?

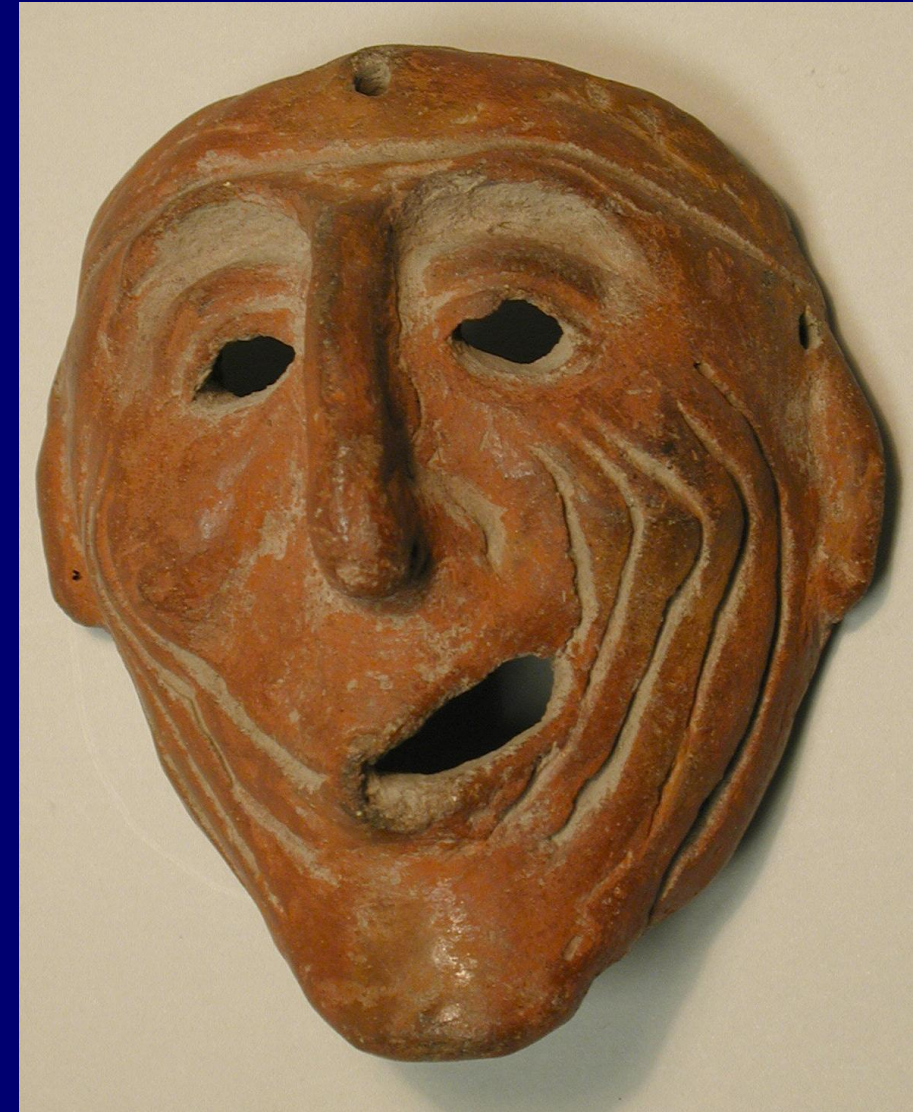
- Surprisingly, we don't know. Hardly anyone has looked.
- Since BRDs alter a person's sense of his/her own body, BRDs may potentially present challenges to effective self-care, caregiving by others, rehabilitation participation, and quality of life



Female figure, Inca,  
15th–early 16th century

- Most rehabilitation aimed at re-learning how to use body parts whose function was affected by the stroke
  - Plausible that BRDs could affect this learning process.
- 2 studies of body structural description impairment and rehab outcomes had contradictory results: one found worse dressing apraxia, the other no effect
  - Possibly confounded by other cognitive deficits
- 1 study found personal neglect did not worsen rehab outcomes
  - But, patients all had extrapersonal neglect, which was treated; so unclear if translates to patients with personal neglect only

- Most research on the impact of post-stroke body representation changes is psychosocial
- Mostly has not correlated findings with specific lesion locations
  - Narrative studies interviewing stroke survivors found that many felt alienated from their bodies and felt they could no longer rely on their bodies
  - Changes accompanied by an altered sense of self and feelings of distress.
  - One study in young adults with stroke found that individuals with left hemisphere strokes showed more negative body attitudes than those with right hemisphere strokes



Mask, Tlatilco, 12th–9th century B.C.

- Future studies of BRDs should assess impact on stroke recovery, post-stroke depression, and quality of life
  - If BRDs adversely affect stroke outcomes, clinicians could target patients with these deficits with more intensive rehabilitation interventions and identify opportunities for new therapeutic innovations
  - On the other hand, if BRDs do not influence outcomes, clinicians could reassure patients and their families that despite the often bizarre nature of these symptoms, they should not affect recovery
- No conclusively proven treatments for BRDs yet exist
  - Case reports and small case series—as well as evidence from other post-stroke disorders—offer some novel approaches worthy of further study

- Mirror therapy
  - One case series of two patients: mirror therapy temporarily reversed somatoparaphrenia
  - Multiple randomized controlled trials have found that mirror therapy for post-stroke hemiparesis improves motor function
    - Mechanism by which mirror therapy improves hemiparesis is not known
    - One possibility: mirror therapy helps normalize patients' body representations and this plays a role in recovery
  - When children see themselves in the mirror, this helps them construct their body representations; potentially, the same process could help adults repair body representations damaged by stroke

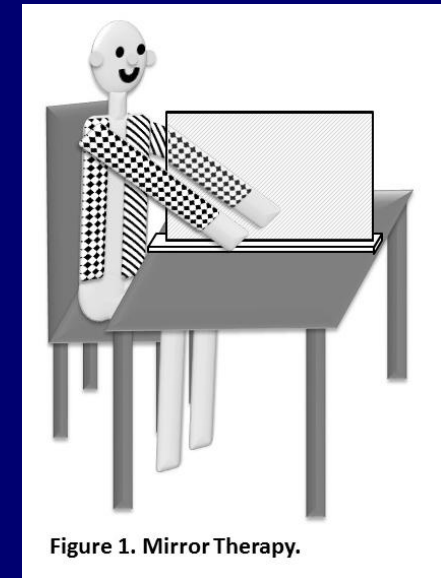
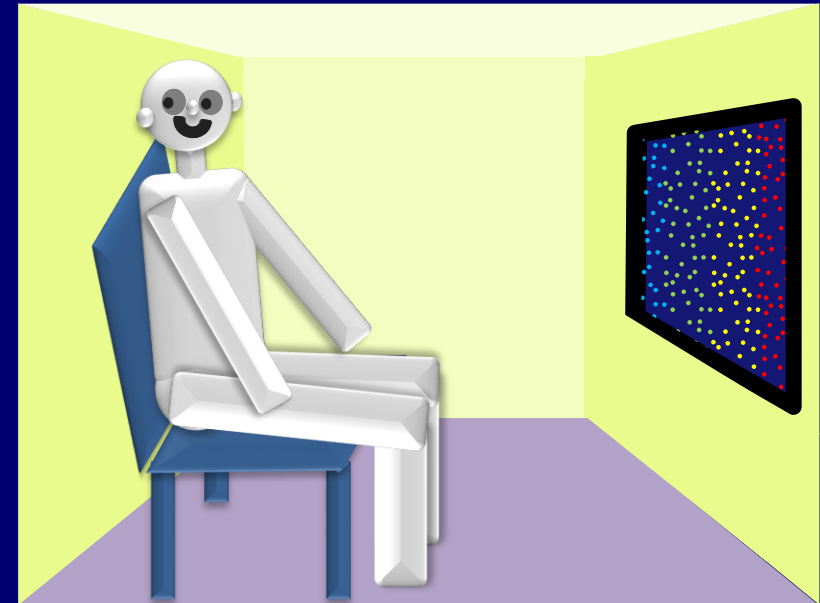
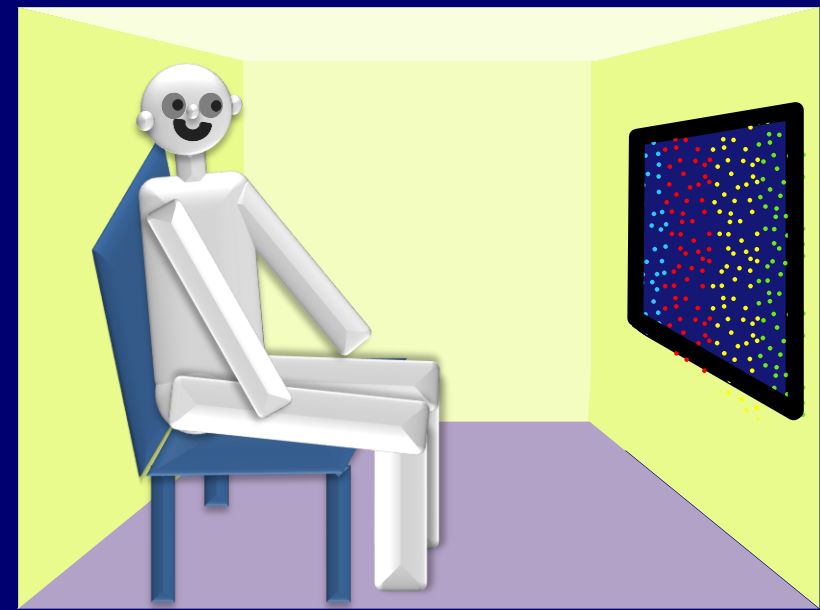


Figure 1. Mirror Therapy.

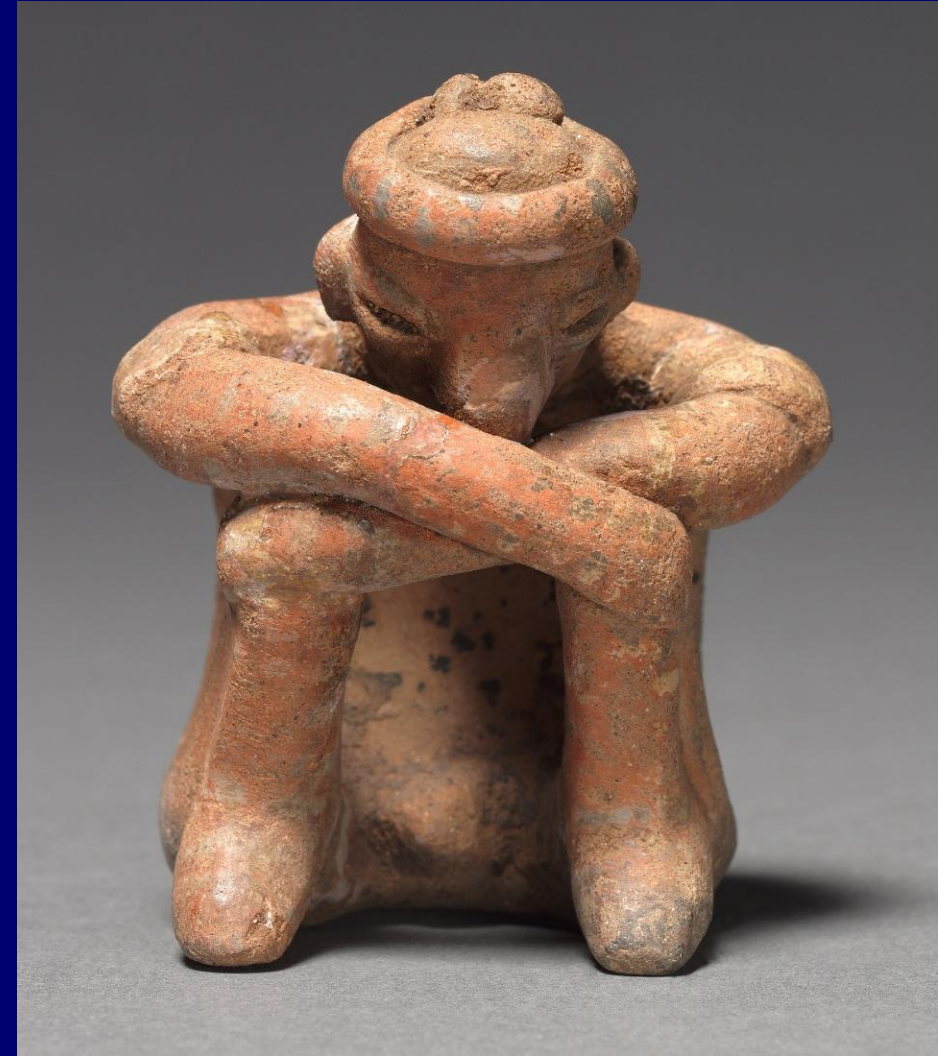
- Vestibular stimulation: case reports/small case series
  - Contralesional cold calorics (and ipsilesional warm calorics in one case) produced transient improvements in ASLO, anosognosia for plegia, personal neglect, and body midline shift
  - Improvement in body midline shift with contralesional neck vibration and contralesionally-directed optokinetic stimulation
  - Both contralesional cold calorics and optokinetic stimulation generate contralesional smooth pursuit eye movements
  - May activate brain regions in the impaired hemisphere
  - If newly activated systems include those involved in body representation, could ameliorate BRDs





# Body Representation Deficits and Primary Psychiatric Disorders

- Primary psychopathology of anorexia nervosa and bulimia nervosa:
  - Misperception of body size
  - Alteration of body emotions
  - Denial of deficits
- Body dysmorphic disorder
- Control delusions
- Somatic delusions



- AN:
  - Disturbance in the way in which one's body weight or shape is experienced, undue influence of body weight or shape on self-evaluation, or persistent lack of recognition of the seriousness of the current low body weight
    - Patients think they're obese even when emaciated
    - Form of macrosomatognosia?



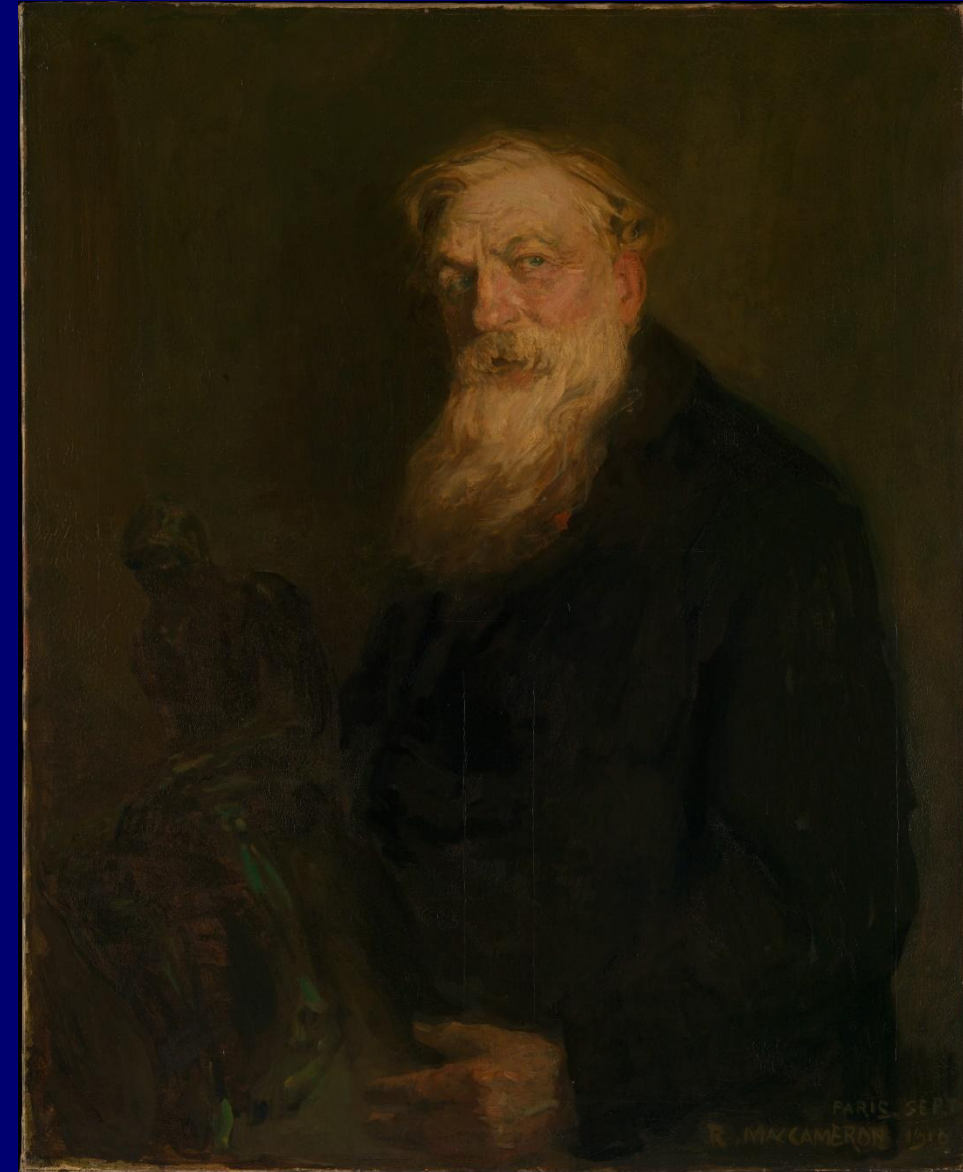
- AN:
  - Disturbance in the way in which one's body weight or shape is experienced, undue influence of body weight or shape on self-evaluation, or persistent lack of recognition of the seriousness of the current low body weight
- BN:
  - Self-evaluation is unduly influenced by body shape and weight
  - Form of alteration of body emotions?



- AN:
  - Disturbance in the way in which one's body weight or shape is experienced, undue influence of body weight or shape on self-evaluation, or **persistent lack of recognition of the seriousness of the current low body weight**
  - Form of body-related anosognosia for deficits/anosodiaphoria for deficits?

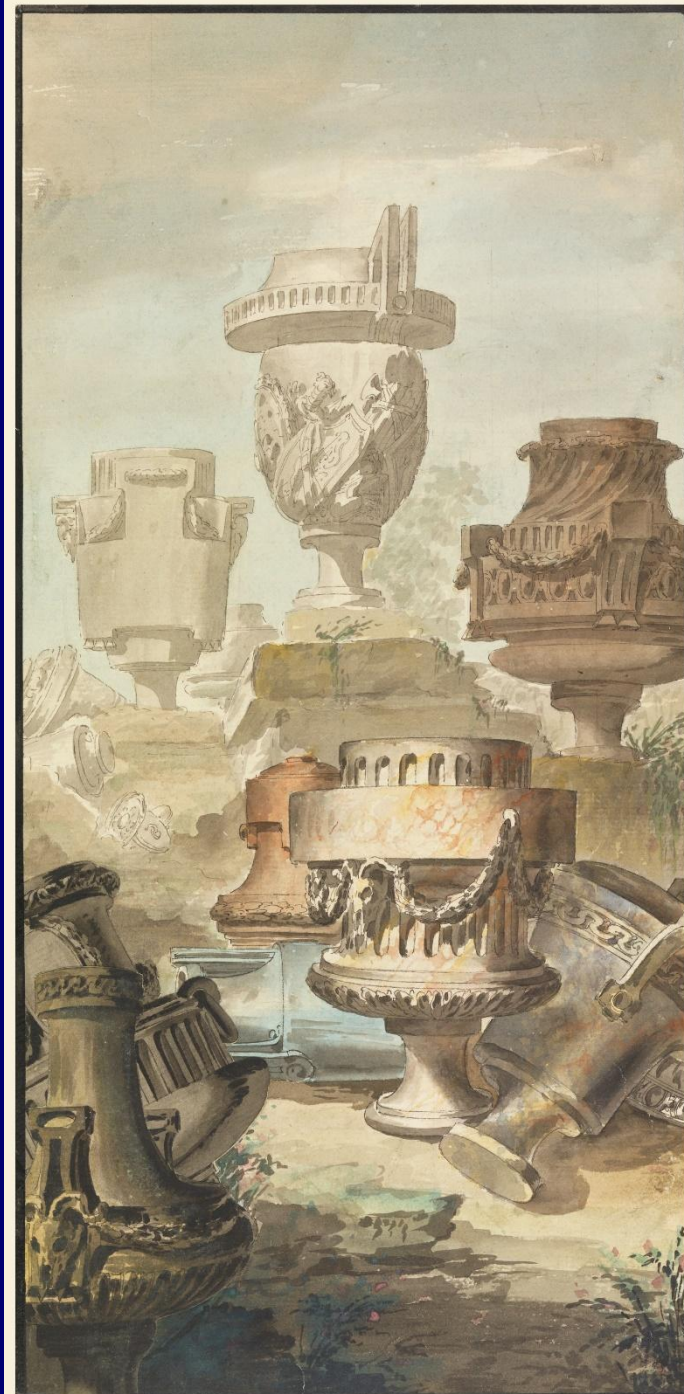


- BDD:
  - Preoccupation with one or more perceived defects or flaws in physical appearance that are not observable or appear slight to others
    - Form of alteration of body emotions?
    - Related to microsomatognosia/microsomatognosia?
      - “My nose is huge and everyone is staring at it”
      - “I have to grow a beard because people will be so freaked out by my weak chin”



## Somatic delusions:

- Koro
  - Genitals (usually penis) shrinking or retracting into abdomen
  - Form of microsomatognosia?
- Cotard delusion
  - Belief you are dead
    - Extreme form of altered body emotions, altered body structural description, and altered body schema?



- Control / passivity delusions in schizophrenia:
  - Patient thinks outside force is controlling his/her movements or actions
    - One theory:
      - Neural pathways involved in *executing* motor movements and neural pathways involved in *monitoring* motor movements become disconnected from each other
      - ie, monitoring system notices “Hey! My arm moved!” and is alarmed because it didn’t get the message “I am going to move my arm now”
      - So perceived as external force initiating the movement
  - Related to brain mechanisms involved in alien limb syndrome?



Questions?

