

# Behavioral and personality changes associated with structural changes

Geoffrey K Aguirre



These slides were created by GK Aguirre and may be distributed and used for instructional purposes freely.

For updated versions, please visit the website:

<http://cfn.upenn.edu/aguirre>

# Topics

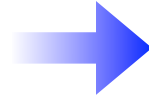
- The big picture
- Lobar and hemispheric phenomena
  - Disorders of language
  - Apraxias
  - Disorders of awareness (incl. neglect)
  - Agnosias
  - Frontal syndromes
- Cognitive effects of head trauma

# The big picture

Left



Language  
Praxis

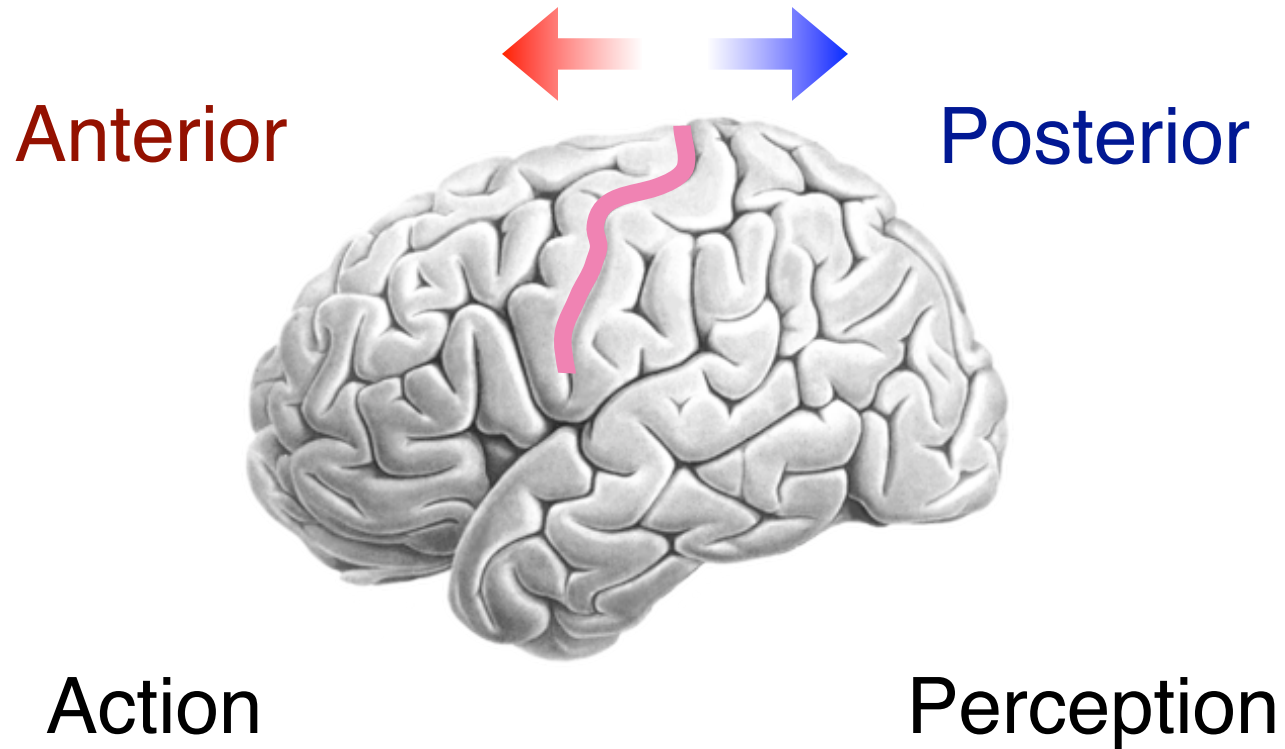


Right



Prosody  
Spatial representation  
Attention

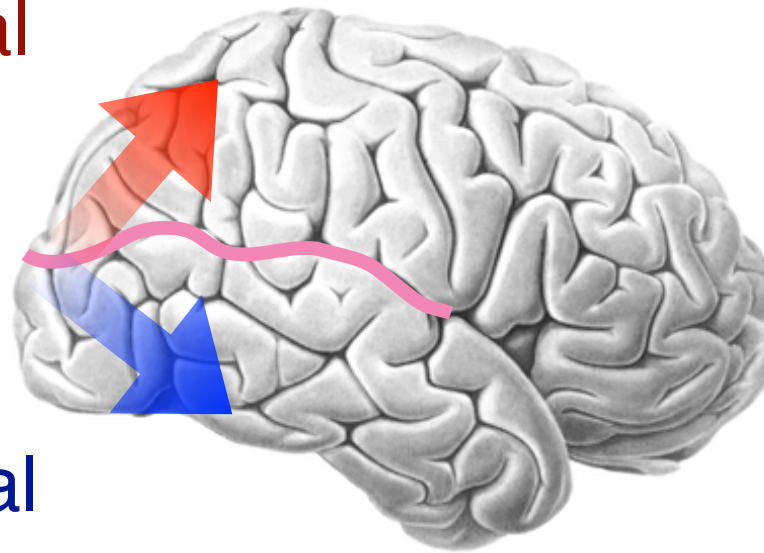
# The big picture



# The big picture

Dorsal

Where



Ventral

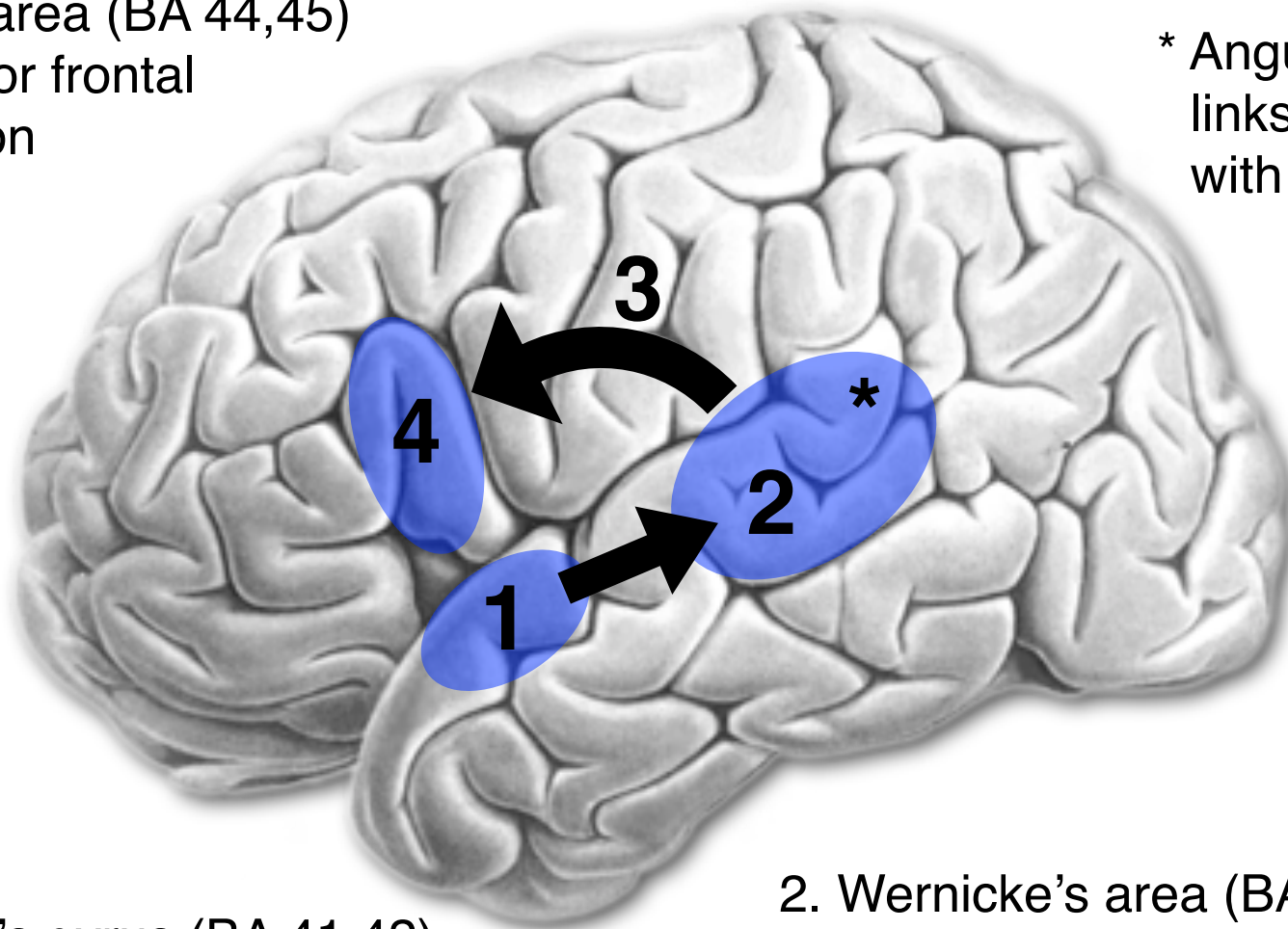
What

# Language anatomy

3. Arcuate fasciculus

4. Broca's area (BA 44,45)  
left inferior frontal  
production

\* Angular gyrus  
links written word  
with language



1. Heschel's gyrus (BA 41,42)  
1° auditory cortex  
tonotopic organization

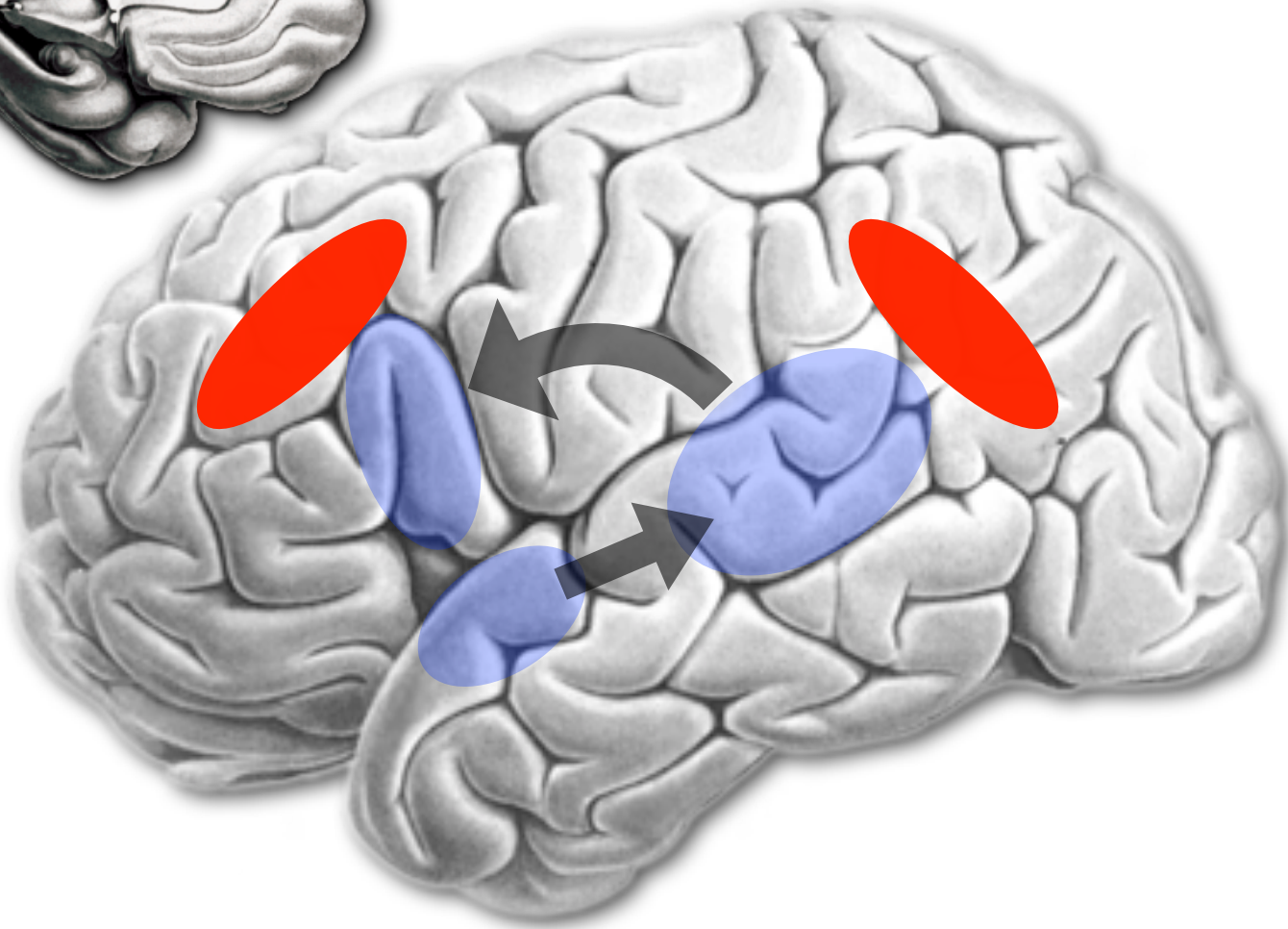
2. Wernicke's area (BA 21, 39)  
(superior temporal gyrus)  
comprehension, L/R asymmetry

# Aphasias

	<u>Fluent</u>	<u>Comp.</u>	<u>Repeat</u>	
Global	No	No	No	large dom. hemi.
Broca's	No	Yes	No	inf. frontal gyus
Wernicke's	Yes	No	No	post. sup. temporal gyus
Conduction	Yes	Yes	No	arcuate fasciculus



# Transcortical aphasia lesion sites (spare perisylvian cortex)



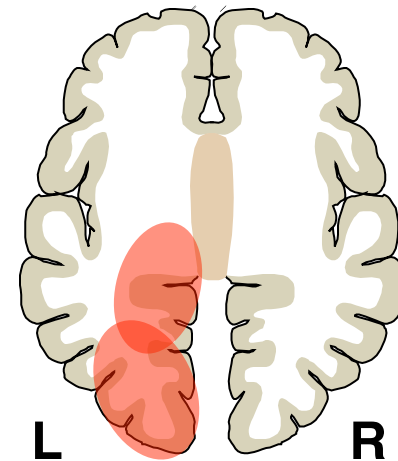
# Aphasias

	<u>Fluent</u>	<u>Comp.</u>	<u>Repeat</u>	
Global	No	No	No	large dom. hemi.
Broca's	No	Yes	No	inf. frontal gyus
Transcortical motor	No	Yes	Yes	mesial frontal, SMA, ACA
Transcortical mixed	No	No	Yes	both transcortical areas
Wernicke's	Yes	No	No	post. sup. temporal gyus
Transcortical sens.	Yes	No	Yes	temporal-parietal jxn.
Conduction	Yes	Yes	No	arcuate fasciculus

# Alexia and agraphia

## Alexia without agraphia -

- Impaired word reading with intact writing (letter reading may be OK)
- Disconnection of right hemisphere visual areas from left hemisphere language areas
- Associated with RHH, color anomia



Left PCA, including forceps major of CC

## Gerstmann's syndrome -

- Agraphia ( $\pm$  alexia), acalculia, finger agnosia, R/L confusion
- A more general "body schema disturbance" can be seen (autopagnosia)



Left angular gyrus (inferior parietal)

# Apraxia

An acquired deficit in learned or skilled movements in the presence of intact strength and sensation

Most cases of apraxia, even when the left hand is clumsy, result from left hemisphere lesions

How to test: have patient pantomime actions to imitation *and* verbal command

There are several types of apraxia -- ideomotor apraxia is the variety most discussed.

Can be seen after focal lesions, but also degenerative disorders, particularly cortico-basilar ganglionic degeneration (CBGD)

# Anatomy for praxis

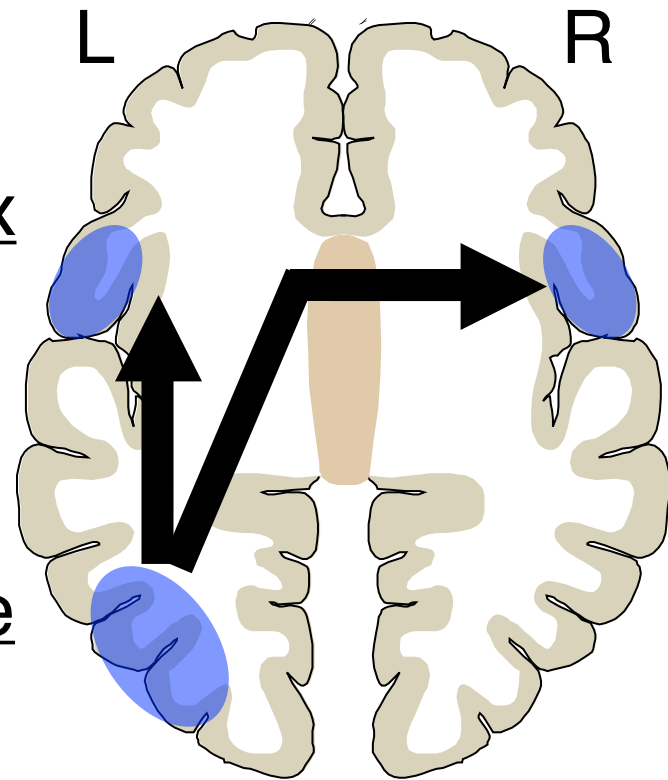


pre-motor cortex

implementation of code in motor terms

inferior parietal lobule

spatial kinesthetic code for actions



# Ideomotor apraxia

## Lesion effects

### 1. Corpus Callosum

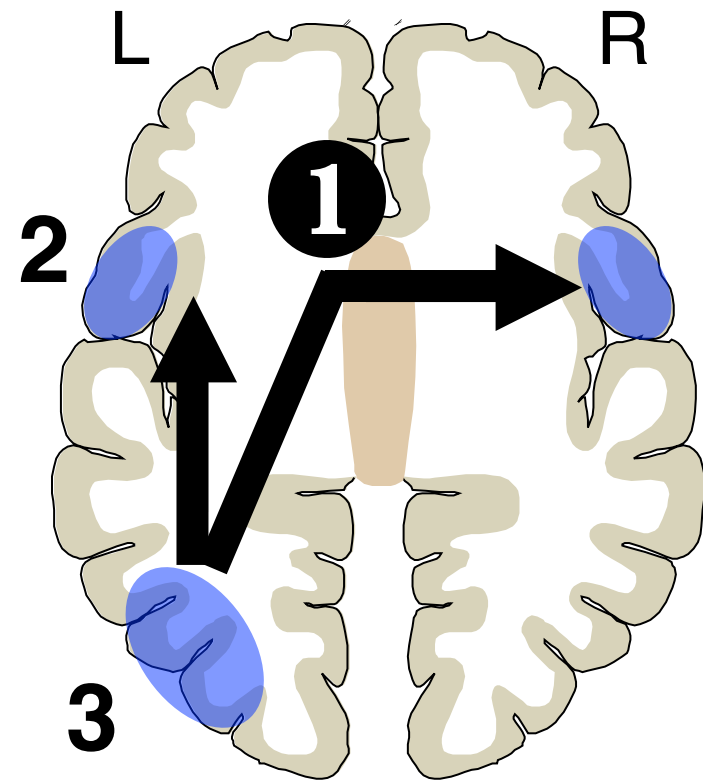
Clumsy left hand, particularly for verbal instructions. Caused by disconnection of right motor area from left sided praxis input

### 2. Pre-motor cortex

Contralateral apraxia, still able to discriminate between poorly and well-performed movements

### 3. Inferior parietal lobule

Apraxia in both limbs. Unable to discriminate between or identify movements of others



# Disorders of attention / neglect

## Attentional neglect -

- Decreased awareness of stimuli contralateral to lesion
- Degree of extinction related to stimulus salience, modality, proximal vs. distal, and location in hemispace



Right inferior parietal  
(also pulvinar, basal ganglia, cingulate)

## Intentional neglect -

- Decreased tendency to act towards stimuli or hemispace contralateral to lesion



Right dorsolateral frontal

# Disorders of attention / neglect

## Balint's syndrome -

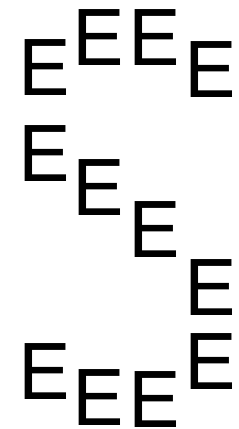
- Simultagnosia (can't see forest for trees)  
Will see only the individual letters of Navon figure
- Optic ataxia (can't reach for visual targets)
- Ocular apraxia (can't direct gaze)



Bilateral parietal  
(AD, CBGD, CJD, sagittal  
sinus thrombosis)

## Other right parietal syndromes -

- Impaired sarcasm - related to loss of prosody
- Topographagnosia - difficulty finding way in locomotor environment
- Dressing apraxia - not a true apraxia, but difficulty orienting limbs to clothing



Navon figure



# Visual agnosias

An inability to recognize a visual stimulus, despite intact function of lower-level vision

H. Lissauer's formulation (1890)

## Apperceptive agnosia



deficit of intermediate vision



deficit of object systems



- Intact acuity, but unable to copy drawings
- diffuse damage (CO poisoning)

- Can copy figures but cannot recognize them
- bilateral PCA strokes

# “Focal” agnosias

## Prosopagnosia -

- Unable to process internal facial features
- Patient can usually discern age, gender and emotion
- May have similar problems with other categories



Bilateral (or only right) temporal lesion

## Achromatopsia -

- Contra-lesional color blindness
- Usually with superior quadrantanopsia
- Ventral occipital lesion (areas V4 and V3/V2)

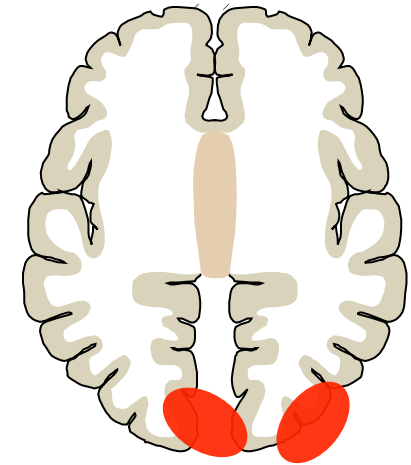


Think of it as: loss of color vision for half of visual field, plus a superior quadrantanopsia

# Other visual disturbances

## Anton's syndrome -

- Cortical blindness with denial of blindness (bilateral occipital pole lesions)



## Palinopsia -

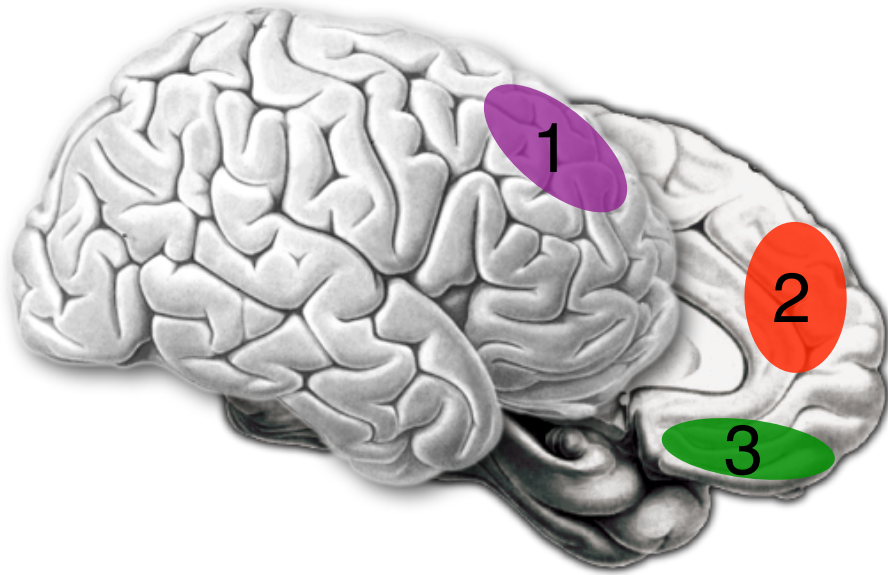
- Persistence of visual sensations
- Caused by occipito-temporal seizures and migraines



## Release hallucinations -

- Caused by loss of visual input (either ocular disease or V1 lesion)
- Extra-striate, categorical visual areas (for face, place, and object perception) interpret input noise as people, animals, and landscapes.
- Typically non-threatening, with intact insight
- In elderly people with ocular disease, this is Charles Bonnet syndrome
- “Hemianopic hallucinations” within the blind field following a V1 lesion

# Frontal syndromes



## 1. Dorsolateral (disorganized) -

- perseveration, persistence, stimulus bound (utilization behavior)
- depression, impaired digit span
- midline falx meningioma

## 2. Mesial-frontal (akinetic-mute) -

- minimal spontaneous action/speech, incontinence, abulia
- can follow ruptured ACA aneurysm

## 3. Orbito-frontal (disinhibited) -

- impulsive, emotionally labile
- inappropriate jocularity
- hypersexual

# Pseudo-bulbar affect (PBA)



## Symptoms -

- Exaggerated emotional responses (laughter, crying) with lability
- Incongruous mood and affect
- Upsetting to the patient, who is aware of the loss of control

## Causes -

- Common feature of many degenerative disorders (ALS, Alzheimer's, Parkinson's, multiple sclerosis)
- Seen after diffuse brain injury from trauma or stroke
- Mildly associated with orbito-frontal damage

## Treatment -

- amitriptyline
- dextromethorphan / quinidine sulfate 20/10 mg (Nuedexta<sup>®</sup>) BID
  - Reduced episodes by half in patients with ALS and MS

# Cognitive effects of head trauma

- Severe traumatic brain injury (TBI) is associated with orbito-frontal, temporal and occipital pole lesions
- A variety of focal cognitive impairments can result from local damage, hemorrhage and contusion
- Diffuse axonal injury (DAI) is the shearing damage to axons within white matter, producing “sub-cortical” cognitive impairments: poor concentration, fatiguability
- Post-traumatic amnesia is a common finding, and the duration of PTA provides prognostic information:

Duration of PTA predicts chance of permanent cognitive impairment

cognitive deficit	<u>&lt; 1 hour</u>	<u>&lt; 7 days</u>	<u>&gt; 7 days</u>
none	65%	18%	17%
mild	45%	21%	34%
severe	16%	12%	72%

# Mild concussion

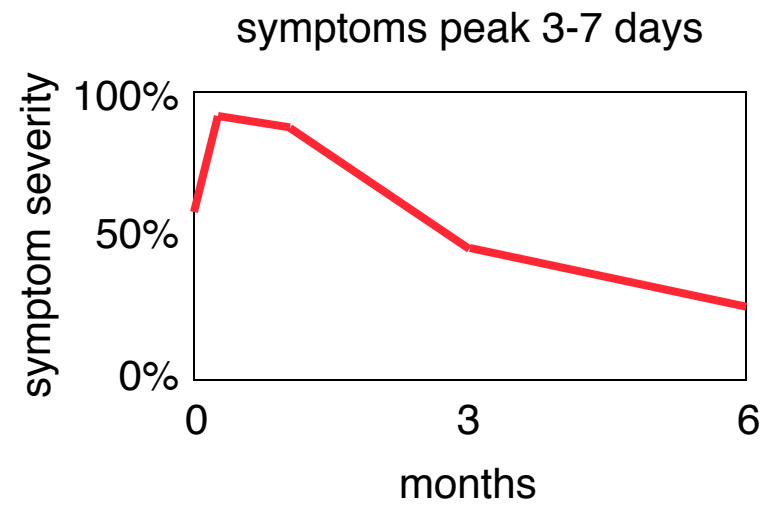
Immediate, transient, trauma-induced alteration in mental status without evidence of structural damage

## Acute symptoms -

- A mild concussion has no loss of consciousness, and less than 30 minutes of post-impact confusion and amnesia
- poor concentration, slurred speech, incoordination, emotionality

## Chronic symptoms -

- Symptoms peak after 3-7 days, persist for months
- Headache, vertigo, poor concentration, fatigability, irritability
- Depression and anxiety also more common post injury



# Delirium

17% of delirium cases are specifically drug induced

Top offenders -

- Meperidine - produces a toxic metabolite (Normeperidine) which is a hallucinogen
- benzodiazepines
- anticholinergics

Delirium is generally caused by an excess of dopamine or a decrease in acetylcholine (e.g., morphine increases DA release)

Other agents -

- $\beta$ -blockers - can cause depression, delirium
- Digoxin - unusual to have encephalopathy without other signs of toxicity
- Steroids - rarely associated with reversible dementia
- $\beta$ -lactams (i.e., penicillin, cephalosporins) and quinolones (i.e., ciprofloxacin)

Associated with time-on-bypass for post-CABG patients



# APPENDIX

Review aphasia descriptions

# Language terminology

Syntactic: relating to the order or structure of language

Semantic: relating to the meaning of words and language

Prosody: physical quality of speech that signifies stress and tone

Paraphasia: erroneous word substitution

semantic paraphasia: substitute meaning (chair → table)

literal paraphasia: substitute sound (house → blouse)

Irregular: a word that violates grapheme → phoneme rules (e.g. yacht)

Non-word: a word that does not exist in the lexicon

# Aphasias

	<u>Fluent</u>	<u>Comp.</u>	<u>Repeat</u>	
Global	No	No	No	large dom. hemi.
Broca's	No	Yes	No	inf. frontal gyus
Transcortical motor	No	Yes	Yes	mesial frontal, SMA, ACA
Transcortical mixed	No	No	Yes	both transcortical areas
Wernicke's	Yes	No	No	post. sup. temporal gyus
Transcortical sens.	Yes	No	Yes	temporal-parietal jxn.
Conduction	Yes	Yes	No	arcuate fasciculus
Pure word deafness	Yes	Yes	No	B. sup. temporal gyrus
Anomic	Yes	Yes	Yes	dementia, ant. temporal

# Aphasia description

Broca's: effortful, disordered grammar, literal paraphasic errors

(house → blouse)

Wernicke's: fluent, empty content, neologisms, semantic paraphasic errors (chair → table)

Conduction: poor repetition, hesitation, decreased auditory short-term memory

Transcortical motor: echolalia, imitation block, improves with dopamine agonists

Transcortical sensory: fluent, circumlocutions, poor comprehension but good repetition

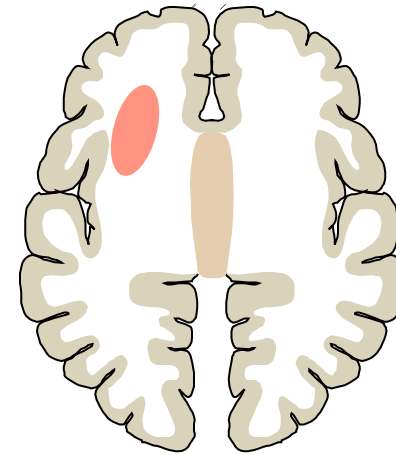
# Aphasia description

Pure word deafness: A selective impairment in understanding spoken language (intact reading and language production).  
Caused by bilateral damage to the superior temporal lobes

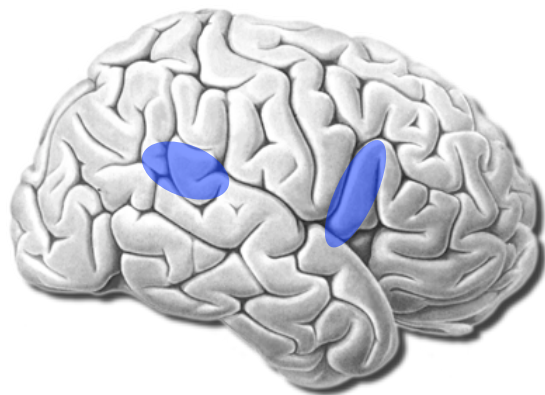
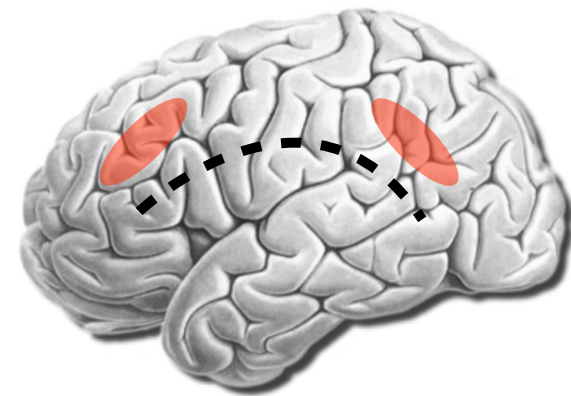
Anomic aphasia: Isolated impairment in word finding. Not associated with a focal lesion, but instead is a common feature of dementing illnesses

# Aphasia pearls

Aphemia (mutism) can result from a lesion that undercuts the white matter of Broca's area. Ability to write is spared.



Watershed ischemia generally results in transcortical aphasias, leaving repetition intact



Impairments in prosody (rhythm and inflection of speech) may accompany homologous lesions in the right temporo-parietal and frontal lobes (receptive and expressive prosody, respectively)

# Acquired dyslexias

## Deep dyslexia -

- Semantic paraphasic errors (e.g., tartan → kilt)
- Cannot read non-words
- Concrete words better than abstract
- “Right hemisphere” reading



Extensive L hemisphere damage

## Surface dyslexia -

- Regularize irregular words (read *sew* as *sue*)
- A common finding in the semantic dementia variant of Fronto-Temporal Dementia (FTD)



Dementia  
(poorly localized)

# “Focal” apraxias

## Limb-kinetic apraxia -

- Difficulty with precise, independent finger movements following pyramidal tract lesions

## Gaze apraxia -

- Disordered eye movements seen in Balint’s syndrome

## Apraxia of eyelid opening -

- Difficulty opening the eyes despite normal consciousness and strength
- Associated with right parietal lesions

## Bucco-facial -

- Impaired tongue / lip actions (whistle, blow out match, kiss)
- Often accompanies Broca’s aphasia



Left inferior-frontal (area 44)



# Pseudo-apraxias

Gait “apraxia” (Brun’s ataxia) -

- Magnetic gait of normal pressure hydrocephalus

Constructional “apraxia” -

- Disorganized copying complex figures (intersecting pentagons, Rey-Osterreith) from frontal or parietal lesions



Right parietal

Dressing “apraxia” -

- Difficulty with spatial arrangement of limbs and clothing

# Other brain-behavior relationships

Alien hand syndrome (disconnection) -

- limb (usually left hand) engages in purposeful movement the patient does not will
- Follows anterior callosal lesion

Alien hand syndrome (CBGD) -

- limb assumes postures and positions without the patient's awareness

Geschwind-Waxman syndrome -

- hyposexuality, hypergraphia, hyperreligiosity
- interictal phenomenon with temporal lobe epilepsy

Kluver-Bucy syndrome -

- hypersexuality, emotional placidity, hyperorality
- bilateral amygdala lesions

# Other brain-behavior relationships

Depression -

- associated with left frontal lesions

Schizophrenia -

- associated with enlarged lateral and third ventricles

Multiple sclerosis -

- impairments in “executive functioning” and slowed processing speed, similar to that seen with dorsolateral frontal lesions