

PSY 448 Clinical Neuropsychology

Basic Neuropathology & Neuropsychological Dysfunctions

Classes of Neuropathology

based Kolb & Whishaw (1990, 1996, 2003, 2015)

The list below is a representative, but *not complete* listing of the kinds of neuropathology which often come to the attention of neuropsychologists.

A. Cerebrovascular Disorders

1a. Cerebral vascular accident (CVA) = "Stroke"

- Infarct = area of dead or dying neural tissue arising from blood supply obstruction
- Hypertension is major risk factor for stroke

Note: In looking at CT and MRI scans, the images are usually reversed, that is, the right side of the brain is shown on the left side of the image and *vice versa*.

Last revised: Sept 22, 2023

1b. **Cerebral Ischemia** (= insufficiency of blood in brain; 87% of strokes)

- **Thrombosis** = plug or clot at place of formation
- Embolism = clot or plug of coagulated blood from elsewhere
- Cerebral arteriosclerosis = narrowing of arteries; inflammation
- Cerebral vascular insufficiency or transient ischemic attack (TIA)
- 1c. **Transient Ischemic Attacks (TIAs)** = brief ischemic episodes (generally < 1 hr)
 - · without lasting tissue damage
 - associated with atherosclerotic disease (artery plaque) and thromobotic microemboli

passing without causing permanent damage

- 2. **Hemorrhagic Stoke** = bleeding into the substance of the brain (10-20% of strokes)
 - Most often result of hypertension
 - mortality rate of 35-52% within 30 days
 - · Often subcortical, i.e., basal

• video of stroke in progress (2015)

- <u>Subdural Hematoma</u> (<u>Medscape</u>)
- Arteriovenuous malformations (AVMs; Mayo Clinic)

ganglia, thalamus, subcortical white matter

- Associated with aneurysms (dilation or ballooning of wall of blood vessel) in 5-10% of all strokes
- Arteriovenous malformations (AVMs) = tangled masses of arteries and veins which grow slowly after birth (1% of all strokes)

 Brain AVMs (U Toronto Brain Vascular Malformation Study Group)

B. Traumatic Brain Injury (TBI)

- 1. **Open-Head Injury** = Penetrating head wound; exposure of brain substance
 - · Danger of infection

2. Closed-Head Injury

- blow = coup + rebound on other side = contracoup
- twisting and shearing
- Disconnection Syndrome = two sides of brain cannot communicate
- hemorrhage -> trapped bloodhematoma
- swelling + collection of fluid = edema

Mild TBI (ca. 80-85% of injuries)

- Any loss of consciousness (< 30 min or Glasgow Coma Scale of 13-15 if > 30 min.) and/or
- any loss of memory for events before/after (#> 24 hr.) and/or
- any alteration in mental state (e.g., feeling dazed, disoriented) and/or
- focal neurological deficit

Moderate TBI: Injury to both gray and white matter (8-10% of injuries)

Severe: <10% of injuries

PBS Nova: Coma

 Dr. Jamshid Ghajar discusses new ways of treating patients in coma and reducing longterm brain damage.

C. Epilepsy (Seizure Disorder)

- = "episodic disturbance of behavior or perception arising from hyperexcitability and hypersynchronous discharge of nerve cells in the brain" (Lezak et al, 2012, p. 242)
- = recurrent, unprovoked seizures
 - Cause may either be symptomatic (cause known) or idiopathic (cause unknown)
 - Simple = no alteration of consciousness or only one mode of expression (movement, psychic, etc.) versus
 - Complex = alteration of consciousness
 - 1% of US population in general but 3% by age 75
 - Carries a lot of social stigma in world and, even, US
- 1. **Partial** (**"Focal"**) = begin locally and then spread
 - Motor (Jacksonian seizure)
 - Complex partial ("temporal lobe" or "psychomotor" seizures
- Complex partial seizure (YouTube, 1'23")
- 2. **Generalized** = bilaterally symmetrical seizures without local onset
 - Absence attack ("petit mal" seizures): nonconvulsive
 - Tonic-clonic ("grand mal" seizures): convulsive with major motor manifestations
- Absence Seizure (YouTube, 2'11")
- Tonic-clonic Seizure (YouTube, 1'25")
- **D. Tumors** (= mass of new tissue that persists and grows independently of its surroundings and without any physiological use)
- 1. **Gliomas** = arising from glial cells and infiltrating the brain substance
 - Astrocytomas (arising from astrocytes)
 - Glioblastomas (= Grade 4/High grade astrocytomas; quite deadly)

Malignant Astrocytoma (Brain-Surgery.com)

Glioblastoma Multiforme (Wikipedia)

- 2. **Meningiomas** = growths attached to the **meninges**; encapsulated (less serious)
- 3. CNS Lymphoma
 - malignant cancer cells form in the lymph tissue of the brain and/or spinal cord
 - associated in last 2 decades with weakened immune systems, e.g., HIV
 - rolativoly rare but increasing

Brain Stereotactic Radiosurgery

- <u>Ganma Knife</u>
 <u>Radiosurgery</u> (Mayo Clinic)
 - Image of Treatment Setting

• relatively rare but increasing rates even in those with intact immune systems

4. Secondary (Metastatic) Tumor

= growth arriving from elsewhere in the body (often lung)

E. Infections

• cause "encephalitis" (a "swelling of the brain")

1. Bacterial

- Meningitis
- Brain abscesses

2. Viral

- Neurotropic (found in neurons) viruses = rabies, poliomyelitis
- Pantropic (found "everywhere") viruses = herpes simplex, mump

PBS Nova - <u>Killer Disease on</u> Campus

- Bacterial meningitis is rare, but college freshman are 4-6 times more likely to get it than the general population
- Amy's Story

F. Dementing, Subcortical and Other Progressive Degenerative Disorders

CD = Cortical dementia; SCD = Subcortical dementia; DMD = Demylelinating disease

1. Mild Cognitive Impairment (MCI) [CD]

- Cognitive impairment "greater than expected for age and education without any obvious etiology, but not sufficiently severe to warrant a diagnosis of dementia" (Lezak et al., 2012, p. 249-250)
- 5-12% of older people
- Often precedes appearance of dementia, but not always.

2. Alzheimer's Disease (AD) [CD]

 "Inexorable progressive degenerative nerve cell changes within the cerebral hemispheres with

PBS Frontline: Pop

 Documentary portrait of 87-year-old, Hy Meyerowitz, by his son and grandson as all

concomitant progressive global deterioration of intellect and personality" (Lezak et al., 2012, p. 252)

- neurofibrils

 (neurofibrillary tangles)
 involving tau proteins
- Amyloid plaques involving Amyloid-Beta
- 65-70% of all dementia patients have an AD diagnosis.

three take a two-week car trip to Florida.

Alzheimer's Association

Alzheimer's Disease
Education and Referral
Center (National Institute on Aging)

2b. Frontotemporal Lobar Degeneration (FTLD)

- Insidious onset & slow progression
- Atrophy of frontal & temporal lobes
- Onset between 40 and 65 years old
- About 20% of all dementias
- Formerly called "Pick's disease"

2c. Dementia with Lewy Bodies (DLB)

- Progressive dementia with deficits in attention, executive function, visuospatial ability, visual hallucinations, fluctuations in cognitive functioning.
- Lewy bodies (alpha-synuclein protein) deposits in cortex and subcortical areas.
- Only identified in 1970s since it shared similar symptoms to AD & Parkinson's disease.

Susan Schneider Williams (2016) "The terrorist inside my husband's brain" (NEUROROLOGY).

 In a special editorial in the medical journal, Neurology, the widow of Robin Williams tells the story of her husband's experience with Lewy Body dementia, the condition which led to his suicide in 2014

Neuropathological findings in Williams' autopsy report

3. **Creutzfeldt-Jakob's Disease** (human spongiform encephalapathy) [CD]

- generalized atrophy believed to be caused by "prions" (protinaceous infectious particles; protein particles without a nucleic acid genome)
- · Incidence 1 in 1 million

4. Parkinson's Disease [SCD]

loss of striatal dopamine due

Prion Diseases (CDC Atlanta)

Human Kuru, New Guinea, 1963 (YouTube, 2'11")

PBS Nova: <u>The Brain Eaters</u> ("Mad Cow" disease & CJD)

to degeneration of the **substantia nigra** in the midbrain

 Onset usually in 50s (rare before 30) with prevalence of 1.5 to 2 per 1000 in Western nations.

5. **Huntington's Chorea** [SCD] (8-10 in 100,000 in Western countries)

 degeneration of basal ganglia, frontal cortex, & corpus callosum due to genetic abnormality

6. Multiple Sclerosis [DMD]

- demyelination of patches in cerebrum leads to abnormal neural activity
- broad range of symptoms: weakness, stiffness, gait problems, visual impairments (e.g., double vision), bladder & bowel irregularities, sexual dysfunction, fatigue (esp. in later part of day as body temperature rises
- varying course of disease: relapsing-remitting, benign (very infrequent attacks),

primary progressive, malignant progressive (very rare)

 Life-span is not significantly impaired: mean survival = 35-43 years.

7. HIV Infection & AIDS

 AIDS Dementia Complex [Mixed] in < 10% of HIV infected patients taking meds (60% of those not on antiretroviral therapy & protease inhibitors)

G. Toxic Syndromes

e.g., damage from drinking alcohol, street drugs, and environmental & industrial neurotoxins (solvents, pesticides, metals such as lead poisoning, formaldehyde, carbon monoxide poisoning, etc.)

Woman with Huntington's Chorea (YouTube, 5'02")

<u>Huntington's Disease</u> (CBS Sunday Morning; YouTube, 7'55")

<u>Living with MS-Dan Powell</u> (YouTube, 4"48")

Korsakoff's Syndrome (sequel to/result of Wernicke's encephalopathy)

 atrophy of mammillary bodies and medial thalamus from chronic excessive alcohol (ETOH) consumption which precluded vitamin B1 (thiamine) in diet

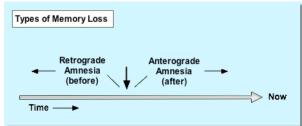
Forms of Neuropsychological Dysfunction

based on notes from Lezak (1995), Larner (2008), & Regan (2016)

- A. Cognition (= the information-handling aspect of behavior)
 - **1. Receptive Functions** (= selection, acquisition, classification, and integration of "information" broadly conceived)

Problems with

- sensory reception (impairment in one or more sensory modalities)
- perceptual functions (inability to process a continued stream of sensations via awareness, recognition, discrimination, patterning, etc.)
- Agnosias (disorders of recognition stemming from perceptual interpretive failure; visual, auditory, tactile agnosia)
 - Prosopagnosia (inability to recognize faces)
 - Alexia (inability to process words)
 - Finger agnosia (inability to identify which finger has been stimulated)
 - Akinetopsia (= visual motion blindness; very rare)
- 2. Memory & Learning (= storage & recall of "information" broadly conceived)



Problems with

- working memory:
 here & now manipulation of visual & auditory materials
- learning new declarative information & procedural skills (anterograde amnesia)
- retrieval of long-term declarative information (retrograde amnesia including both episodic or autobiographical and semantic information)
- confabulation: making up stories to cover over memory deficits
- **3. Thinking** (= the mental organization & reorganization of information)
 - Dyscalculia (inability to compute numbers)

- Impaired reasoning & judgment
- Impaired concept formation
- Loss of abstraction & generalizing
- Inability to order objects
- Poor organizing or deficient planning
- **4. Expressive Functions** (= the abilities through which information is communicated

or acted upon.) Including speaking, drawing, writing, manipulating, physical gesture, facial expression, movement

- Apraxia (= disturbance of purposeful expressive function) ["can't do']
- Constructional dyspraxia (= disturbance in assembling, building, drawing) Two- and three-dimensional dyspraxia
- Aphasia (=defects of symbol formulation)
 - Expressive (dysfluency; Broca's/motor/syntactic aphasia)
 - Memory/retrieval (anomic/semiotic aphasia)
 - Programming Sequences (conduction aphasia)
 - Comprehension (= fluent, garbled/jargon speech; Wernicke's/fluent aphasia)
 - Global
- **5. Mental Activity Variables** (= characteristics that concern the efficiency of mental processes)
 - Lowered Attention, Concentration, Tracking
 - Visual neglect
 - Disturbances of consciousness: drowsiness, somnolence, stupor, coma
 - · Disturbed speed of processing

B. Emotionality: Feelings & Motivation

- Emotional dulling
- Diminution of anxiety (blandness or mild euphoria)
- Decreased social sensitivity
- Heightened anxiety
- Depressed mood
- Irritability, restlessness, low frustration tolerance
- Apathy
- Increased/decreased sexuality

C. Executive Functions (= capacities which permit a person to engage successfully

in independent, purposive, self-serving behavior)

- Anergia (= loss/lack of initiation of activity)
- Defective planning & prioritizing of activities & goals; difficulty in sequencing tasks
- Loss of emotional self-control (see B. above)
- Loss of goal-direction & problems with sustained attention
- Metacognition: ability to evaluate how well one has accomplished or evaluated what they have done
- Overall organization, lack of, i.e., tracking information & materials of daily life and work by means of planners, lists, bins, etc.
- Impulsivity; perseveration (= inability to stop a behavior); lack of flexibility in behavior

- Time management difficulties
- Working memory: problems in keeping track mentally important information as task is being accomplished

REFERENCES

Kolb, B., & Whishaw, I. Q. (2003). Fundamentals of human neuropsychology (5th ed.). New York: Worth Publishers.

Larner, A. J. (2008). Neuropsychological neurology: The neurocognitive impairments of neurological disorders. New York: Cambridge University Press.

Lezak, M. D. (1995). Neuropsychological assessment (3rd ed.). New York: Oxford University Press.

Lezak, M. D., Howieson, D. B., Bigler, E. D., & Tranel, D. (2012). *Neuropsychological assessment* (5th ed.). New York: Oxford University Press.

Regan, T. M. (2016). *Understanding autism in adults and aging adults: Improving diagnosis and quality of life.* USA: Indiego Publishing LLC.

This page was first posted September 23, 2003.