TASP *A partner on the path to student success*

Texas Association of School Psychologists

The cognitive and psychosocial implications of epilepsy for students, families, and teachers.

Presented by Robb Matthews, PhD, LSSP, HSPP, NCSP
November 6, 2021

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Learning Objectives

- Participants will recognize epilepsy specific terms and their application.
- Participants will be able to classify seizures by type and location.
- Participants will be able to consider important cognitive and psychosocial factors in their conceptualization and planning for individuals with epilepsy.

2

Public Service Announcement

My goal for this presentation includes:

- Developing a working model of epilepsy and sequelae that will allow for efficient evaluation and intervention decisions.

My goal for this presentation DOES NOT include:

- Memorization of minutia that will not support the goal outlined above
  - Visit scholar.google.com to search for more epilepsy and sequelae information.
- I always try to operationalize Albert Einstein's axiom "make things as simple as possible, but no simpler"

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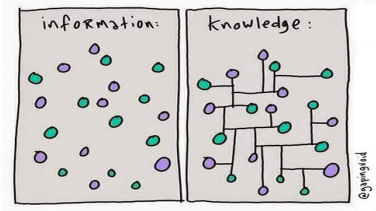
Public Service Announcement Summary



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Seizures in Children

- Seizures represent a physically and emotionally challenging circumstances for children and their families.
- Seizures occur in reaction to diverse causes or circumstances and their effects vary based on internal and/or external factors.
  - fever (i.e., febrile)
  - lack of oxygen (i.e., hypoxic)
  - head trauma
  - illnesses (e.g., hypocalcemia, hyponatremia, CNS infection)
  - genetic factors
  - brain tumors
  - macro- or microscopic structural brain differences

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Seizures in children

- The heterogeneity of the disease does not lend itself to a specific "epilepsy battery" or "go to" set of supports, accommodations, or goals.
- A general knowledge of the specific type and severity of the epilepsy syndrome as well as potential effects of the needed treatment regimen will allow for planning around the student's specific needs.

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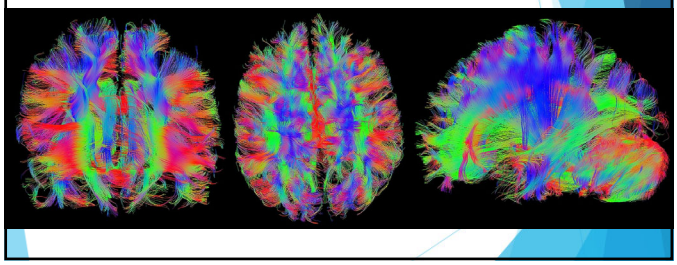

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Seizures in children

- Seizures result from dysregulated neuronal activation and manifest heterogenous neurobehavioral changes.
- There are ≈ 30 different seizure types (CDC, 2018).
- In some instances, provoked seizures are relatively common and unlikely to result in enduring developmental differences, neurocognitive deficits, or the need for long-term treatment.
  - Febrile seizures (<15-minutes with fever>101˚F) are estimated to occur in up to 1 in 25 children (National Institute of Neurological Disorders and Stroke, n.d.) between 6- and 60-months old.
- Although the event can clearly be traumatic and result in lasting parental concern, a single febrile seizure is not usually pathological in nature and is not considered evidence of epilepsy or other disease process.

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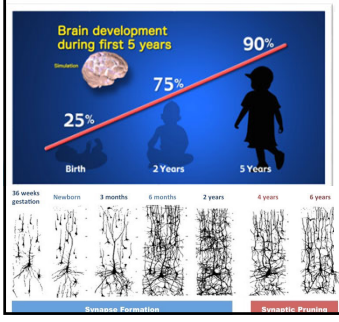

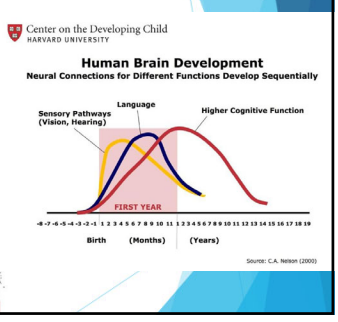

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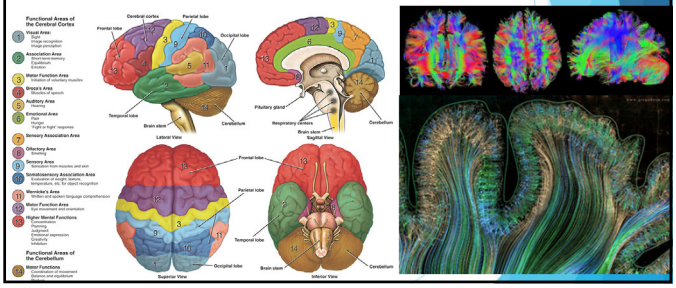

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Seizures in children

- Epilepsy is the most common neurological disease in children and even with improvements in our understanding of the human brain, our understanding of epilepsy remains functional in nature (Fisher et al, 2014).
- Some conditions associated with epilepsy include:
  - Genetic differences (e.g., Down syndrome, Prader-Willi syndrome)*.
  - Developmental differences (e.g., hypocalcemia, hydrocephalus, hypoxia, grey matter heterotopia, ASD).
  - Health factors (e.g., stroke, Central Nervous System (CNS) infections)
  - Head Injuries**.
  - Brain Tumors.
- Aaberg and colleagues (2017) suggest that in children with epilepsy worldwide, up to 65% of the causes remain undefined.

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Seizures in children

- Epilepsy is a **complex brain disease** that causes seizures (Sokka et al, 2017; Fisher et al, 2017, Fisher et al, 2014).
  - May consist of multiple unprovoked seizures (i.e., ≥2-events >24-hours apart).
  - May consist of 1 unprovoked or reflex seizure with a probability (≥60%) of additional seizures occurring over the next 10 years.
  - May consist of an epilepsy syndrome.
- Westerveld (2020) characterizes the nature of the disease as a neuronal dysfunction resulting from an imbalance in inhibitory and excitatory functions.

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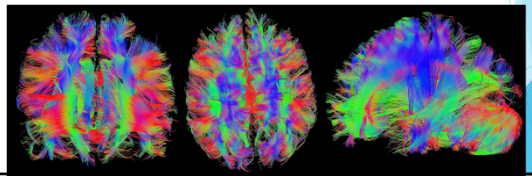
Seizures in children

- While much of the neurologically focused epilepsy research has focused on the characteristics of a particular type of epilepsy, the change in conceptualization to a **brain network disease** reflects current knowledge of the brain as an integrated system rather than a conglomeration of specialized functions.



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Seizures in children - Epilepsy in the US

- Zack and Kobau (2017) found in 2015 ≈1.2% of the US population were identified with active epilepsy (95% CI = 1.1-1.4).
  - 470,000 US children (≤ 17-years old).
  - 3 million US adults (≥ 18- years old).
- Designated as the costliest (US $9,103.25 annually) and 2nd most common chronic disease in children (diabetes; epilepsy; hypertension; food allergies; asthma; Miller et al, 2016).
- Epilepsy syndromes - clusters of signs and symptoms that typically occur together and may include seizure type, underlying cause, age of onset, severity, chronicity, and prognosis when evident.

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Behavioral Seizure Terminology

- Atonic - loss of muscle tone, formerly referred to as drop attacks.
- Tonic - sustained focal stiffening of the body, arm, or leg.
- Clonic - focal rhythmic jerking of the body, arm, or leg.
- Tonic-clonic - a convulsion, formerly referred to as grand mal.
- Myoclonic - irregular, brief focal muscle contractions.
- Epileptic spasms -focal flexion or extension of arms and flexion of trunk.
- Automatism - purposeless repetitive motor activity (e.g., lip-smacking).
- Behavior arrest - termination of movement with unresponsiveness.
- Absence “ahb-sahnce” - short periods of lack of awareness or responsiveness.
- Electrographic-only seizures - seizures with no behavioral component.

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Behavioral Seizure Terminology

- Cognitive manifestations - individual reports or exhibits changes/deficits in language, mathematics, thinking, or other higher cortical functions.
- Autonomic manifestations – individual experiences can vary widely and may include gastrointestinal symptoms, thermoreception changes, flushing, goosebumps, palpitations, sexual arousal, respiratory changes, and/or other sensations.
- Emotional symptoms – individual may experience include anxiousness, irritability, aggression, paranoia, elation, and/or other changes.
- Status epilepticus - a neurological emergency characterized by tonic-clonic seizures lasting 30 or more minutes or intermittent seizures occurring within the same timeline that do not return to baseline.

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ILAE 2017 Classification of Seizure Types Expanded Version [1]

Focal Onset

Aware | Impaired Awareness

Motor Onset
automatisms
atonic [2]
clonic
epileptic spasms [2]
hyperkinetic
myoclonic
tonic

Non-Motor Onset
autonomic
behavior arrest
cognitive
emotional
sensory

focal to bilateral tonic-clonic

Generalized Onset

Motor
tonic-clonic
clonic
tonic
myoclonic
myoclonic-tonic-clonic
myoclonic-atonic
atonic
epileptic spasms

Non-Motor (absence)
typical
atypical
myoclonic
eyelid myoclonia

Unknown Onset

Motor
tonic-clonic
epileptic spasms

Non-Motor
behavior arrest

Unclassified [3]

[1] Definitions, other seizure types and descriptors are listed in the accompanying paper and glossary of terms

[2] Degree of awareness usually is not specified

[3] Due to inadequate information or inability to place in other categories

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OLD TERMS VS. NEW TERMS FOR SEIZURES

The International League Against Epilepsy has revised the names we give paticular seizures. The Epilepsy Foundation of America published a short list of the newer and former names we typically gave specific seizures. Epilepsy Education Everywhere has created a meme of that list to help the public learn the revised terms.

OLD TERMS	NEW TERMS
COMPLEX PARTIAL	FOCAL IMPAIRED AWARENESS
PSYCHOMOTOR	FOCAL IMPAIRED AWARENESS
LIMBIC	FOCAL IMPAIRED AWARENESS
FOCAL MOTOR	FOCAL MOTOR AWARE *OR* IMPAIRED AWARENESS
FOCAL SENSORY	FOCAL SENSORY AWARE *OR* IMPAIRED AWARENESS
SIMPLE PARTIAL	FOCAL AWARE

OLD TERMS	NEW TERMS
ABSENCE	GENERALIZED ABSENCE
ATONIC OR DROP ATTACK	FOCAL *OR* GENERALIZED ATONIC
GRAND MAL	GENERALIZED *OR* UNKNOWN ONSET TONIC CLONIC
INFANTILE SPASMS	FOCAL, GENERALIZED, UNKNOWN ONSET EPILEPTIC SPASMS
MYOCLONIC	FOCAL *OR* GENERALIZED MYOCLONIC
PETIT MAL	GENERALIZED ABSENCE
TONIC CLONIC	GENERALIZED *OR* UNKNOWN ONSET TONIC CLONIC
TONIC OR DROP ATTACK	FOCAL *OR* GENERALIZED TONIC

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Seizure Conceptualization

- **Idiopathic** – "unknown" may be due to genetic causes or a disease.
- **Cryptogenic** – thought to be caused by developmental lesions too small to see on neuroimaging.
- **Symptomatic** – having an identified lesion thought to be etiologically related to the seizure disorder.
  - Having an identified causal factor (e.g., genetic, structural, metabolic, infectious) and an abnormal electroencephalographic (EEG) pattern increases the risk of subsequent seizures and decreases the likelihood of seizure control (Aaberg et al., 2018).

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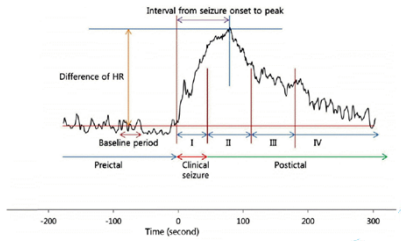
Seizure Event Staging



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Seizures in children

- Consciousness - refers to the degree of responsiveness or level of awareness of a patient during a seizure. These are safety factors.
  - Responsiveness – child's ability to carry out commands during a seizure or their degree of contact with the environment.
  - Awareness - child's degree of contact with, and memory for, events that occur during a seizure. Typically used in place of consciousness as a descriptor.

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Seizures in children

- Psychogenic Non-Epilepsy Seizures (PNES) – **NOT EPILEPSY**
  - Usually witnessed by someone else.
  - Awareness of surrounding (e.g., following directions, talking).
  - Movements begin slowly then escalate, are asynchronous, and come and go.
  - Crying or shrieking in middle or at end of seizure.
  - Duration much longer than typical seizure.

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Treatments

- Contemporary AEDs (≤30-years) only suppress seizures, they do not affect the underlying disease (Chen et al., 2018; Kellogg & Meador, 2017). There has been no significant improvement in rate of seizure control with newer generation AEDs (Chen et al, 2017).
- Factors associated with reduced outcomes:
  - a high number of seizures in the year prior to regimen initiation.
  - a family history of epilepsy in first-degree relatives.
  - previous brain injury.
  - a history of smoking.
  - previous recreational drug use.
  - the presence of psychiatric comorbidity.

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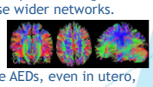
Treatments

- AEDs impact targeted symptoms as well as other neurocognitive and physiological processes (i.e., side effects).
  - Although seizures may be associated with a particular aspect of the brain (e.g., temporal lobe), it is likely that changes in the brain are distributed over wider brain networks (Hermann et al., 2010; Reed et al., 2020) and treatments are similarly likely to impact the cognitive and physiological functions associated with those wider networks.
  - Polypharmacy – intensifies the effects.



- Some research suggests exposure to more aggressive AEDs, even in utero, may result in broad differences in CNS development (Kellogg & Meador, 2017); however, these findings are not consistent across all AEDs (Arican et al., 2020).

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Treatments

AED Side effects may reflect the wider networks impacted by epilepsy

Cognitive
- Decreased attention and concentration
- Learning difficulties
- Reduced cognitive and fine motor efficiencies
- Somnolence
- Slurred speech

Behavioral
- Hyperactivity
- Irritability
- Sleep disturbance
- Mood changes
- Decreased Appetite

Physiological
- Diarrhea
- Double vision
- Weight gain
- Gum dysplasia
- Hirsuitism (male pattern hair growth)
- Rash
- Anemia
- Liver damage

Stopping an AED regimen without medical supervision can result in the recurrence of seizures and those seizures may be less responsive to medication in the future.

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Treatments - Epilepsy Syndromes

- Lennox-Gastaut syndrome
  - Occurs over the lifespan and is pharmacoresistant.
  - Characterized by more than one type of generalized seizure, including drop seizures for at least-six months and results in a slow spike-and-wave pattern on EEG.
  - Some evidence that it is the interaction between AED and CBD that results in the improved seizure control.
- Dravet syndrome
  - Occurs in typically developing children before about 15-months, is likely to be life-long, and is pharmacoresistant (De Liso et al., 2016).
  - Frequently begins with prolonged febrile seizures then progresses into other seizure types over time.
  - Includes status epilepticus and generally results in developmental disruption, gait disorders, psychiatric conditions, and sleep disorders (Nabbout & Thiele, 2020).

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Treatments

- If AEDs do maintain adequate control other options include:
  - Surgical resection of misbehaving parenchyma.
    - Thermal laser ablation of the specific parenchyma may lessen the broader side effects associated with typical resections (Westerveld, 2020)
  - Implantable stimulators (e.g., Vagal Nerve Stimulator (VNS), Responsive Neurostimulation (RNS), or Deep Brain Stimulator (DBS)).
  - Dietary Therapies (e.g., ketogenic).
  - Cannabinoids may be an effective treatment addition to standard AEDs for individuals with epilepsy syndromes; however, controlled studies of naturally occurring cannabinoids have inconsistent results and make specific recommendations unrealistic (Nabbout & Theile, 2020).

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Life Effects

- Further risks result from other chronic conditions associated with epilepsy, such as Intellectual Disability (ID), Autism Spectrum Disorder (ASD), and/or specific genetic conditions.
- Research also suggests the type and duration of active epilepsy seems to be related to differences in intellectual functioning than other factors (Lopes et al., 2014, Westerveld, 2020).
- More significant impacts on intellectual functioning may be associated with genetically based neurodevelopmental differences and the degree of variance may reflect the degree of neuronal dysfunction (Aaberg et al., 2018, Riccio et al., 2010, Wolf et al., 2015).

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Life Effects

- Epilepsy is a disease that may persist throughout the lifespan dependent on the type of epilepsy and the underlying causal factors (Westerveld, 2020).
- Given that epilepsy results from changes in the central nervous system, these risks for meeting typical developmental accomplishments should not be unexpected.
- Epilepsy is associated with decreases in academic achievement, social success, vocational attainment, and Quality of Life (QoL).

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Life Effects - Educational Planning

- Transition planning should begin in early adolescence/middle school and is likely to increase in importance as the complexity of student's presentation or the number of risk factors increases.
- One area of high importance is preparation for self-management of epilepsy medications to facilitate practice and mastery.
  - Even if the child or adolescent cannot manage the complex aspects of the medication regimen independently, understanding the importance of consistent medication management and regular communication about their symptoms should be emphasized.

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Educational Planning – Safety and Support

- Frequency of seizures. Is there a pattern to monitor?
- Identify preictal symptoms and plan for safety.
- Ictal Procedures
  - Plan for maintaining safety during a seizure event occurs (e.g., student's awareness level, environmental risks).
  - Plan for emergency intervention(s) if indicated.
- Identifying appropriate expectations during the postictal phase.
  - How do we support a return to baseline?

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Evaluation Planning

- When a seizure has occurred near a scheduled evaluation session and the seizure impact is mild, it may be reasonable to wait 30- to 60-minutes before beginning/resuming the evaluation.
- When a seizure has occurred near a scheduled evaluation session and the impact is more widespread or severe, such as a generalized convulsive event, it is reasonable to wait 24-hours before beginning/resuming the evaluation.
- Research suggests that IEDs (Interictal Epileptiform Discharges) occur in or around the seizure onset zone and may result in spurious evaluation results (Reed et al., 2020). There is no specific timeline for allowing postictal symptoms to remit; however, the goal of gathering reliable data should inform scheduling decisions.

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Evaluation Planning

Research - Transitive Cognitive Impairment

- Reed and colleagues investigated how seizures impact memory.
  - Measured electrical activity in the hippocampus while using a visual recognition task.
  - Results suggested interictal epileptiform discharges (IEDs), temporarily changed the firing of individual cells in the hippocampus.
  - Journal of Neuroscience 15 January 2020, 40 (3) 682-693; DOI: https://doi.org/10.1523/JNEUROSCI.1380-19.2019

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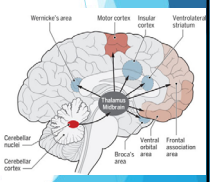
Evaluation Planning - Attention

- Changes in the thalamo-frontocortical network are likely to impact several aspects of higher-level brain functions.
  - Attention deficits, impulsiveness, working memory deficits, and planning inefficiencies have been associated with epilepsy-related changes in this circuit and may be progressive in nature (Wolf et al., 2015, Westerveld, 2020).
  - Children with absence epilepsy show impairments in aspects of the underlying attention network (Westerveld, 2020).



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Evaluation Planning - Attention

- Changes in the thalamo-frontocortical network are likely to impact several aspects of higher-level brain functions.
  - Moschetta & Valente (2013) found attention and verbal memory difficulties were also associated with social adjustment difficulties in individuals with temporal lobe epilepsy.
  - Decreased inhibitory control, mood consistency, and decreased sensitivity to other's thoughts/opinions are also frequently associated with Juvenile Myoclonic Epilepsy (JME) (Wolf et al., 2015).

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Evaluation Planning - Attention

- Changes in the thalamo-frontocortical network are likely to impact several aspects of higher-level brain functions.
  - Patterns of inattention, hyperactivity, and impulsivity in epilepsy frequently meet criteria for ADHD.
  - Research suggests that medications commonly used to treat idiopathic ADHD are likely to be an effective for children comorbid epilepsy and ADHD and TYPICALLY DO NOT lower the seizure threshold (Auvin et al, 2018, Westerveld,2020).

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Evaluation Planning - Learning & Memory

- Research suggests that learning and memory differences are a common area of difficulty across most types of epilepsy and that the mechanisms underlying these differences are not well understood (Bell, 2012, Kernan et al., 2012, Reed et al., 2020, Ung et al., 2017).
  - Differences may span across auditory and visuospatial aspects of learning and recall.
  - Changes in the hippocampus are common in adolescence and extend into adulthood (Westerveld, 2020).
  - Correlated with diverse microstructural changes in the bilateral temporal lobes and related ipsilateral subcortical structures employed to efficiently transfer information (Keller et al., 2012, Reed et al., 2020).

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Evaluation Planning – Learning & Memory

- Petitet and colleagues (2018) attribute broad learning efficiency to adaptability of the CNS.
  - Adaptation is a fundamental aspect of the CNS that is evident across neurocognitive domains (Lodhi & Agrawal, 2012; Petitet et al., 2018).
  - Reduced adaptability in a brain network disease likely have decreased learning efficiency, error correction, and/or generalization.
  - The impact of a systemic brain disease on learning and recall may result in difficulties mastering broad aspects of information and behavior over the lifespan.

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Evaluation Planning - Learning & Memory

- Keller and colleagues (2012) found broad differences in adults with left temporal lobe epilepsy and associated hippocampal sclerosis (TLEhs).
  - The changes seem to be related to the duration of the disease rather than age at onset or a history of febrile seizures.
  - Some research (Berg et al., 2008) suggests TLEhs may become evident by adolescence.
  - Although they may not be immediately evident in younger students, a careful consideration of the functions associated with these networks is likely worthwhile as both a baseline and for future planning.

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Evaluation Planning - Learning & Memory

- Research has also identified several learning and recall patterns associated with specific types of epilepsy.
  - Ung and colleagues (2017) found that IEDs occurring outside the seizure onset zone were more impactful on learning and recall than those occurring within the zone.
    - This finding suggests that the seizure onset zone tissue is dysfunctional and the IEDs disrupt the function of underlying healthy tissue as well.
  - Reed and colleagues (2020) found that IEDs in the hippocampi (particularly the right) impacted familiar recall accuracy, but not novel learning efficiency.
    - This research suggests that inhibitory neurons seemed more effected by IEDs than excitatory ones.

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Evaluation Planning - Language

- Westerveld (2020) argues that onset of seizures during specific periods of language development may exacerbate epilepsy's impact on language development.
- Keller and colleagues (2012) and Lodhi and Agrawal (2012) submit that more pronounced language learning, comprehension, and use deficits are associated with the broader changes in those with dominant-side temporal lobe epilepsy.
- Lah and Smith (2015) also found that language and language-based learning did not show significant improvement 1-year after temporal lobe resection.

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Evaluation Planning - Psychosocial

- Emotional and behavioral dysregulation may also be associated with the epilepsy disease process, with greater difficulties being associated with more difficult to control epilepsy syndromes (Auvin et al., 2018).
  - Wolf and colleagues (2015) found that psychiatric disorders in individuals with epilepsy was a strong predictor of pharmacoresistance and medication nonadherence.
  - Emotional and behavioral symptoms representing peri-ictal indicators, medication related effects, or discrete psychiatric conditions and/or some combination of those factors.

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Evaluation Planning - Psychosocial

- The psychosocial prognosis of individuals with epilepsy may also be impacted by psychiatric comorbidities; however, the specific nature of the relationship remains unclear (Wolf et al., 2015).
  - Wolf and colleagues (2015) suggest that individuals with greater degrees of neuronal dysfunction will evidence increased severity of epilepsy symptoms and psychiatric problems.
  - Higher incidences of mood and anxiety disorders are evident throughout the epilepsy literature. Research suggests that 1 in 3 individuals with epilepsy is likely to develop emotional and/or behavioral conditions (Mula, et al., 2020).
    - SSRIs DO NOT lower the seizure threshold.
  - Lodhi and Agrawal (2012) also suggest risk factors for suicidal ideation or actions is higher in those with epilepsy.

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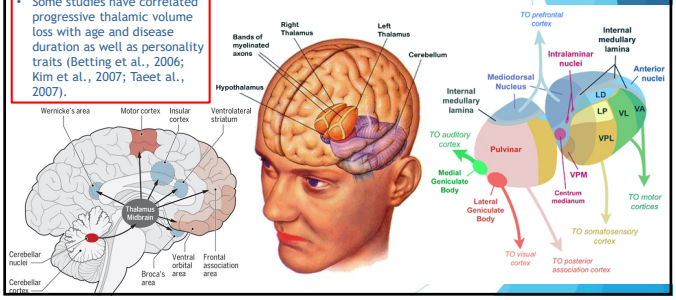
- Some studies have correlated progressive thalamic volume loss with age and disease duration as well as personality traits (Betting et al., 2006; Kim et al., 2007; Taeet al., 2007).



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Psychosocial Intervention Planning

- History of seizures in embarrassing circumstances (e.g., in a social circumstance).
- Social limitations related to seizures (e.g., restrictions from typical activities like sports or driving).
- Fears or worry related to seizure occurrence (e.g., loss of bladder control in the classroom).
- The need for behavioral control procedures (e.g., turning in earlier than peers, avoiding triggers, maintaining medication intervals).

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Psychosocial & Transition Planning

- There is a high likelihood that students with more complex presentations will carry the effects of epilepsy into adulthood. Thus, the focus of planning should move from primarily educational in early grades toward preparing for post-graduation circumstances like higher education intentions/needs, independence in adulthood, and/or potential vocational directions.
- One area of high importance is preparation for self-management of epilepsy medications to facilitate practice and mastery.
  - Even if the child or adolescent cannot manage the complex aspects of the medication regimen independently, understanding the importance of consistent medication management and regular communication about their symptoms should be emphasized.

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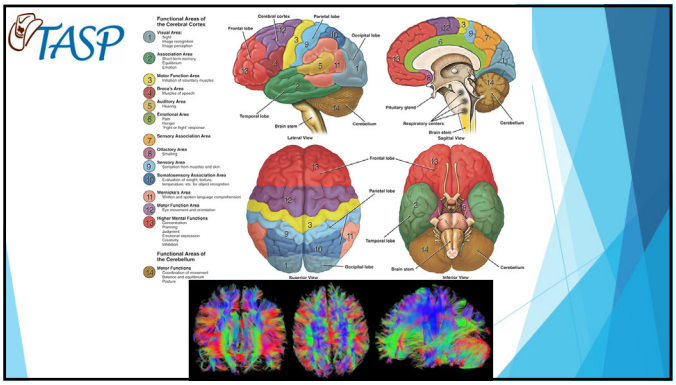

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