SA17- Providing PA/LTC to Adults With Intellectual and Developmental Disabilities

Saturday, March 24
2:30 PM- 3:30 PM

Session Description

This session will provide a clinical introduction to caring for adults with intellectual and developmental disabilities in PA/LTC settings, based on literature review and the presenter’s 25 years of experience as a clinician, medical educator, and researcher in developmental disabilities medicine. The content material presented will be applied to four case scenarios which exemplify the breadth and complexity of health care in this population. Participants will develop and share proposals for a quality improvement initiative aiming to improve care of this population, tailored to the facilities where they practice.

Learning Objectives

- Describe how the etiologic diagnosis for the developmental disability guides clinical care of this population.
- Apply a literature-based, culturally-sensitive model of health care to persons with IDD in PA/LTC.
- Outline key elements of a quality improvement initiative addressing the health care for persons with IDD in facilities where they practice.

Presenter(s): Carl Tyler, Jr., MD, MS, CMD

Presenter(s) Disclosures: All speakers have reported they have no relevant financial relationships to disclose
Providing Post-Acute and Long-Term Care to Adults with Intellectual and Developmental Disabilities

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Cleveland Clinic Lerner College of Medicine
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Speaker Disclosures
Dr. Tyler has disclosed that he has no relevant financial relationship(s).

Learning Objectives
By the end of the session, participants will be able to:
1. Describe how the etiologic diagnosis for the developmental disability guides clinical care of this population.
2. Apply a literature-based, culturally-sensitive model of health care to persons with IDD in PA/LTC
3. Outline key elements of a quality improvement initiative addressing the health care for persons with IDD in facilities where they practice.

My Background
• Primary care of persons deinstitutionalized from developmental centers in Ohio in the late 1980s
• Student of the cumulative wisdom from persons with IDD, their families & caregivers, and disabilities professionals
• Education and training about health and health care in this population
  - Self-advocates, DSPs, QDDPs, DD nurses, medical students, resident physicians, practicing physicians
  - Researcher- clinical care, practice-based research, health disparities, EMRs and large administrative datasets
• Medical consultant for state crisis prevention network
• Regional health system level approaches to improving care

Case 1: Tony, age 52, with Down syndrome recently admitted to Assisted Living Facility
Tony is age 52 and has Down syndrome and moderate intellectual disability. He and his mother simultaneously entered an Assisted Living Facility (ALF) when she began falling frequently and was found to have early dementia. They now share a suite in the facility. Previously, Tony had lived with his mother all his life in the family home.
Since the move, Tony has regressed and is no longer brushing his teeth, showering, or changing his clothes without prompting; all of these he used to do independently, without prompts.
Prior to admission to ALF, Tony had not seen a doctor for years. How would you assess and intervene with Tony?

Case 2: Lucinda, age 67, with cerebral palsy, post-operative hip fracture, in Post-Acute Care
Lucinda has hemi-paretic cerebral palsy, mild intellectual disability and previously well-controlled epilepsy. She was living in a supported apartment complex with visitation by a direct support professional for two hours every morning and every evening. One week ago, she fell at her day program and fractured her hip. Following operative repair, she is undergoing rehabilitation in the Post-Acute Care unit where you practice.
Upon arrival, Lucinda has a tonic-clinic seizure. Upon return to consciousness, she is complaining that her repaired hip hurts. Describe your plan for immediate and longer-term post-acute care.
Case 3: Darren, age 48, with Down syndrome and dementia newly admitted to Intermediate Care Facility

Darren is age 48 was admitted to the Immediate Care Facility with diagnosis of Down syndrome complicated by early-onset dementia. His father had died suddenly from a stroke 5 years ago, and his mother died from an acute myocardial infarction 2 years ago. He lived with a brother who reluctantly assumed his care following mother’s death.

What initial assessment would you conduct upon his arrival? The nursing and direct care staff have no previous experience with residents with Down syndrome. How would you counsel the staff to optimize Darren’s adjustment to his new residence?

Case 4: Michael, age 30, with autism, epilepsy and severe intellectual disability in respite care

Michael lives with his single mother Lena. He has diagnosis of autism, epilepsy and severe intellectual disability. His mother is arranging weekend respite care at the long-term care facility where you are medical director. What would you want to know about Michael? How can you work with your staff to prepare to provide respite care at your facility for adults with developmental disabilities?

Critical Understandings

- Etiologic Diagnosis
- Baseline Adaptive Functions
- Communication Abilities & Supports
- Service & Support Network
- Preferences, Interests, Triggers, Comforts
- Mental & Behavioral Health History

What Is IDD?

What Is IDD?

A severe, chronic disability of a person which:

A. is attributable to a mental or physical impairment or combination of mental and physical impairments;
B. is manifested before the person attains the age twenty-two;
C. is likely to continue indefinitely;
D. results in substantial functional limitations in three or more of the following areas of major life activity:
   i. Self-care
   ii. Receptive and expressive language
   iii. Learning
   iv. Mobility
   v. Self-direction
   vi. Capacity for independent living
   vii. Economic sufficiency
E. reflects the person's need for a combination and sequence of special interdisciplinary, or generic care, treatment, or other services which are life-long or extended duration and are individually planned and co-coordinated.

Changes in Describing IDD

- R Word: Spread the Word to End the Word:
  - "Intellectual disability" has replaced the pejorative term "mental retardation"
  - http://www.r-word.org/
- Less emphasis on IQ test results, more on functional capabilities & supports necessary to maximize capabilities
- "Person-first" language:
  - "The individual/adult/elder/man/woman with Down syndrome" NOT "The Down syndrome"
Epidemiology

- An estimated 641,000 adults with IDD age 60 and older were residing in the United States in 2000 (Heller, 2002)
- This group will increase threefold (approximately 2 M) by 2020. (Janicki, 2002)
- Presently over 200,000 person in US with Down syndrome age over 55
- Health issues associated with aging occur at earlier ages and a higher rates than general population (Heller 2013)
  - Mobility limitations; osteoporosis/osteomalacia; sensory impairments; dental problems; obesity/underweight; HTN; DM; dementia; depression

Aging Trajectories

- Vary according to etiology & phenotypic features of DD; genetic & social- environmental factors; lifestyle
- Shorter life expectancies associated with specific DD syndromes are not representative for most persons with IDD
  - Average life expectancy for persons with Down syndrome (DS) is now in the 60s
  - Average life expectancy for community dwelling persons with IDD, not DS, is in the mid-70s
- Aging cohorts of persons with IDD greatly influenced by previous institutional residences; opportunities for healthy living; health care

Who with IDD Has Decreased Life Expectancy?

Individuals with:
- Cerebral palsy w/ severe motor & functional impairments
- Epilepsy with refractory seizures
- Chronic upper respiratory infections
- Heart conditions
- Infections
- Reduced mobility
- Dependency in eating and toileting
- Severe to profound levels of ID

Causes of Mortality: Respiratory Disease

- Leading cause of death
- Recurrent aspiration events, untreated asthma may lead to irreversible broncho-constriction
- Increased risk for pneumonia due to immobility, recurrent aspiration
- Restrictive lung disease due to kyphoscoliosis, obesity

Causes of Mortality: Cardiovascular Diseases

- Second most common cause of death
- Cardiovascular disease risk factors similar to general population
- Recognition of acute coronary syndromes difficult because of communication impairments
- Heart failure due to
  - Unrecognized sleep apnea
  - Congential heart diseases
  - Acquired valvular heart conditions
  - Untreated hypertension

Causes of Mortality: Cancer

- Cause of death in about 10% of persons with IDD
- Disparities in cancer screening practices
- Fewer tobacco and alcohol-associated malignancies
- Significant risks for breast, colorectal cancer
- Less risk for prostate cancer, cervical cancer
- Increased risk of some gastrointestinal cancers
  - Chronic GERD –Barrett’s esophagus – esophageal carcinoma
  - Chronic constipation may ↑ risk of colorectal cancer
  - Gallbladder cancer
  - Hepatocellular carcinoma due to chronic hepatitis B
- Specific cancer profiles associated with some genetic conditions
  - Down syndrome associated with ↑ risk of leukemia, testicular cancer; but ↓ risk of other solid tumors such as breast cancer
  - As genetic etiologies of DD syndromes are clarified, syndrome-specific cancer risks will be clarified
Other Facets of Aging & IDD

- Aging and specific syndromes
- Behavioral and mental health co-morbidities
- Age-associated conditions
- Chronic diseases
- Pharmacotherapy

Influences on Aging Process

- Lack of physical activity
- Obesity
- Smoking
- Access to health care
- Poor nutrition
- Poor dental care
- Poor hygiene
- Poverty
- Abuse
- Violence
- Inadequate social networks
- Inadequate education

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CLINICAL APPROACH TO ELDERS WITH IDD

Principles of geriatric medicine
- Common geriatric syndromes
- Under-recognized health problems
- Pharmacotherapy
- Decline in Adaptive Functioning

Principles of geriatric medicine
- Disease and illness often present in atypical ways
  - Developmentally-determined behavioral expressions of distress
  - Signs and symptoms influenced by longstanding spasticity, structural and functional impairments
  - Influence of disease & illness on long-standing neurobehavioral issues
  - Self-injurious behavior; physical aggression; stereotypes
  - Multiple organ system dysfunctions, chronic diseases, acute illness, nutritional deficiencies, and psychosocial stressors operate together to cause impairments
  - Successful aging is not just about physical health
  - Under-recognition of importance and influence of social relationships, changes in environment on health and behavior

Principles of Geriatric Medicine

- As individuals age, they become more dissimilar from each other
  - Even more heterogeneity given baseline differences in health and function in persons with IDD
- An abrupt decline in function should always be assumed to be a disease or illness, not due to aging
  - Diagnostic overshadowing
  - Misattribution of any decline in adaptive functioning to dementia
- The rate of decline with aging can be modified by lifestyle modifications and treatment of known illness
  - Diagnostic omissions
  - Failure to provide healthy lifestyle choices & modifications

Common Geriatric Syndromes

- Dementia
- Delirium
- Urinary Incontinence
- Falls
- Dizziness
- Syncope
- Sleep Problems
- Hearing Impairment
- Visual Impairment
- Osteoporosis
- Osteomalacia
- Malnutrition
- Feeding Problems
- Pressure Ulcers
Under-Recognized Health Problems

- CAD
- CHF
- PAD
- COPD
- Osteoporosis
- Osteoarthritis
- Spinal Stenosis
- GERD

- Podiatric Problems
- Dental Caries
- Periodontal Disease
- Malnutrition
- Dehydration
- Anxiety disorders
- Depression

Pharmacotherapy

- Polypharmacy
- “The prescribing cascade”
- Long term chronic pharmacotherapies
- Side effects of psychoactive medications
  - Drug-induced movement disorders
  - Adverse neurocognitive effects

High-risk Medications

- All psychotropic drugs
- Antiepileptic drugs
- Anticholinergic drugs
- Cardiovascular drugs

Factors Contributing to Decline in Adaptive Functioning

- Visual impairment
- Hearing impairment
- Adverse medication effect
- Depression
- Adjustment disorders
- Grief
- Suboptimal epilepsy management
- Unrecognized pain
- Malnutrition
- Unrecognized cardiac disease
- Unrecognized pulmonary disease

Preventive Health Care

- Guidelines: concurrence & discordance
- Individualized decision-making
- Syndrome-specific health risks
- Secondary conditions

Health Promotion

- Individual beliefs & attitudes
- Informal networks
- Formal networks
- Meaning of activity
- Family, direct care staff, health care providers
- Motivation
- Access
End of Life Care

- Planning
- Health care decisions
- Advance Directives
- Bereavement
- Pain management

Summary: Clinical Approach to Aging Adult with IDD

- Begin clinical thought process with the etiologic diagnosis for the DD
  - Implications regarding health risks, co-morbid conditions, secondary disabilities
- Scan medication list
  - Consider likely adverse drug effects, apparent errors, drug-drug interactions, drug-disease interactions
- Assume undiagnosed disease and/or undertreated disease
- Assume problematic behaviors may be in part a sign of physical distress (pain) or disease

Avoiding Common Clinical Errors

- Optimize diagnosis
- Optimize treatment
- Evaluate for common mental health conditions
- Evaluate for medical causes of neurobehavioral disorders
- Scrutinize pharmacotherapy

Focus #1: Optimize diagnosis

Consider common & medically serious conditions and risks that are often unrecognized:

- Spinal cord compression
- Recurrent aspiration
- Heart disease- CAD, HF
- Chronic lung disease
- Seizures

Consult: Spinal Cord Compression

- Presenting issues: 67-year-old female with recurrent psychiatric hospitalizations for self-injurious behavior; physical and verbal aggression; sudden decline in ADLs (mobility, self-feeding)
- Problem List:
  - Medical: Cervical spinal stenosis C4-5; Recurrent UTI; Iron-deficiency Anemia; Constipation with fecal impaction; Xerodermatitis; Dysphagia; Hypothyroidism; Rosacea
  - Neuro-behavioral: Severe intellectual disability; Epilepsy (GTC); Bipolar disorder; Psychosis NOS; Tardive dyskinesia; Neurodermatitis
- Changes in Adaptive Functioning: Used to walk independently, now requires wheelchair; Used to feed herself, now requiring full assistance with eating most days; cannot hold spoon

Spinal Cord Compression: Recognition

- Risk factors: Congenital defects of base of brain and spine anatomy; cerebral palsy; Down syndrome; degenerative arthritis of spine; scoliosis; osteoporosis; cancer
- Symptoms: Severe neck, back, or limb pain; weakness in arms or legs; loss of feeling in limbs; change in gait pattern; swallowing difficulty; (new) urinary or fecal incontinence
- Signs: Loss of strength in limbs; change in deep tendon reflexes; posturing of neck; loss of sensation; loss of adaptive functions
Spinal Stenosis - Imaging

Spinal Cord Compression: Evaluation & Treatment

- Evaluation:
  - Thorough review of changes in strength, coordination, adaptive functioning
  - Adequate neurological examination
  - MRI imaging of spine (Plain X rays of spine not good enough!)

- Treatment:
  - Usually requires surgery
  - Sometimes “steroids” or radiation therapy (if cancer) may temporize the condition

Spinal Cord Compression: Take Home Messages

- Risk factors for & conditions causing compression of the spinal cord are common in this population
- Delayed recognition & treatment of SCC can lead to irreversible loss of function or death
- Clinicians often fail to appreciate the medical implications of loss of strength or significant changes in adaptive functioning
- Patients with alarming symptoms often receive an inadequate neurological examination & inadequate imaging of the spine.

Consult: Recurrent Aspiration

- 66-year-old with mild intellectual disability and schizophrenia; psychiatric condition stable for 15 years, with sudden decompensation requiring several hospitalizations for both medical and for psychiatric indications

- Problem List:
  - Medical: Myasthenia gravis; constipation; hypertriaglyceridemia; hypothyroidism; osteoporosis; iron deficiency anemia; seizure disorder; sideroblastic anemia; muscular degeneration; osteoarthritis; urinary incontinence
  - Neuro-behavioral: Paranoid schizophrenia; mild intellectual disability “with behavior problems;” depression

- Medications: Levothyroxine; Pantoprazole; Miralax; Mestinon; Simvastatin; Bisacodyl; Alendronate; Os-cal; Cyanocobalamin; Enablex; Colace; Ferrous sulfate; Gemfibrozil; Omeprazole

- Psychoactive medications: Risperidone; Benztropine; Depakote; Carbamazepine

Abnormal CT Chest: “Bilateral patchy areas of groundglass opacity, greatest at left upper lobe. Course interstitial densities at the lung apices and lung bases, probably fibrotic in nature.”

Aspiration Events: Recognition

- Risk Factors: Dysphagia, dependency in feeding, quadriplegia, severe scoliosis, GERD, rumination. Medications: psychoactives, AEDs, muscle relaxants, anti-spasmodics, anticholinergics

- Signs & Symptoms
  - For Dysphagia: Mealtime coughing, gurgling, gagging, agitation; drooling, spitting out food; tongue thrusting, pocketing of food; prolonged feeding times; food refusal; weight loss
  - For Recurrent Aspiration Events: Recurrent fevers, episodes of rapid breathing & low pulse oximetry readings, chronic cough, chest pain, wheezing, recurrent pneumonia

Aspiration Events: Evaluation

- Observation
  - Mealtime behaviors

- Examination
  - General alertness, speech quality & clarity
  - Oral cavity (Physicians don’t look carefully in the mouth!)
  - Bedside swallowing evaluation
  - Chest examination

- Review
  - Medications that dry up, alter consciousness, or slow reflexes
  - Previous studies (MBS, CXR, CT chest)
  - Hospitalizations related to infections or breathing problems
Aspiration Events: Take Home Messages

- Aspiration events are common & are often unrecognized, especially if they don’t lead to hospitalization
- Swallowing function is dynamic & can be transiently affected by acute illness & medications
- Clinicians are often falsely reassured by a single normal MBS
- Clinicians often ignore abnormalities of CXR or CT chest that indicate recurrent aspiration
- Recurrent aspiration can lead to irreversible changes in lung structure & function

Focus # 2: Optimize treatment

Review the adequacy of treatment of these common conditions that are frequently diagnosed but are often sub-optimally treated

- Constipation
- GERD
- Epilepsy
- Osteoporosis

Consult: Constipation

- Presenting Issues: 67-year-old female with recurrent psychiatric hospitalizations for self-injurious behavior; physical and verbal aggression; sudden decline in ADLs (mobility, self-feeding)
- Problem List:
  - Medical: Cervical spinal stenosis C4-5; Recurrent UTI; Iron-deficiency Anemia; Constipation with fecal impaction; Xerodermatitis; Dysphagia; Hypothyroidism; Rosacea
  - Neurobehavioral: Severe intellectual disability; Epilepsy (GTC); Bipolar disorder; Psychosis NOS; Tardive dyskinesia; Neurodermatitis
- Medications:
  - Medical: Omeprazole; Ascorbic Acid; Multivitamin; Docusate 100 mg BID; Ferrous sulfate; Provera q 3 months; Synthroid; Lactulose 30 cc TID; Benadryl Psychoactive: Depakote 500 mg QID; Clozaril 50 mg TID
- She was recently hospitalized for constipation requiring enemas and laxatives.

Constipation: Recognition

- Risk Factors: Cerebral palsy; medications for seizures, mental health conditions, pain, bladder control, cardiovascular conditions, or spasticity; immobility, hypotonia; more severe ID; DS, WS, Cri-du-chat
- Signs & Symptoms: Abdominal pain; straining at defecation; prolonged toileting; hemorrhoids with bleeding; poor appetite; vomiting; acid reflux; weight loss; SIB; irritability; sleeplessness; restlessness; urinary incontinence; fecal incontinence

Abdominal Radiography
Constipation: Evaluation

- Constipation is poorly detected on physical examination
  - Abdominal examination is an unreliable indicator
  - Clinicians often misinterpret absence of stool in rectal vault as indication that constipation is absent
- Radiologists often fail to comment about or grade severity of fecal accumulation evident on abdominal plain film X rays and abdominal CT scans.
- Clinicians often ignore radiology comments about fecal accumulation even when reported
- Therapeutic response to laxatives is best indicator that constipation was problematic

Constipation: Take-Home Messages

- Maintain a high index of suspicion for constipation causing or contributing to any behavioral, nutritional, or gastrointestinal complaint
- Use therapeutic response to laxatives, not physical examination or bowel record, as most reliable indicator for symptomatic constipation
- Clinicians tend to under-recognize constipation as a problem or they will give laxatives short-term without changing daily regimen

Constul: Gastroesophageal Reflux Disease

Presenting Issues: 61-year-old male with Down syndrome, severe intellectual disability, with frequent complaints of headaches, stomachaches, and ear aches, who engages in physical and verbal aggression, self-injurious behavior, and property destruction.

Problem List:
- Medical: Hypothyroidism, Hearing impairment, Constipation
- Neuro-behavioral: Severe intellectual disability, Phonoconstructive disorder, ADHA, oppositional disorder

Past Social History: Reported abuse and neglect as a child; self-reported abuse with hot water and by confinement to locked room. History of foster care.

Medications:
- Medical: Levothyroxine 0.125 mg, Senna 8.6 mg QHS
- Psychoactive: Benztropine mesylate 0.5 mg TID, Depakote sprinkles 1500 mg QHS, Risperidone 4 mg QHS

GERD: Recognition

- Definition: “Troublesome” symptoms or complications caused by reflux of stomach contents into the esophagus
- Esophageal syndromes:
  - Symptoms: Typical reflux syndrome, reflux chest pain
  - Injuries: Reflux esophagitis, stricture, Barrett’s esophagus, adenocarcinoma. Obesity is risk factor for esophageal CA and GERD
- Extra-esophageal syndromes:
  - Reflux cough, laryngitis, asthma, dental erosions
  - Possible: Sinusitis, pulmonary fibrosis, pharyngitis, recurrent otitis media
  - Acid reflux may trigger angina in patients with CAD
- Prevalence:
  - General population: 14% overall, 10% nocturnal sx
  - IDD: 50% institutionalized populations

GERD: Evaluation

- Assessment by signs or symptoms suggestive of complications requiring more intensive investigations
- Decision regarding empiric trial PPI vs. diagnostic testing
- EGD identifies complications of stricture, Barrett’s esophagus, or adenocarcinoma
- Most patients with GERD have normal EGD
- Findings on EGD often do not correlate with symptom severity
- Common co-morbidities
  - Impaired salivation
  - Dysphagia
  - Esophageal dysmotility
  - Gastroparesis
  - Duodeno-gastroesophageal (bile acid) reflux
EGD: Diagnostic Testing

- 24 hour pH probes or reflux scan can document reflux if EGD & manometry are normal but diagnosis still suspected and there is need to confirm it
- In tube-fed individuals, food dye coloring can be mixed with tube feeding and patient observed for appearance of dye-colored food in mouth
- Obtain CBC, iron studies, stool occult blood testing to screen for GI blood loss, determine need for colonoscopy in addition to EGD
- If co-morbid dysphagia/aspiration suspected, obtain clinical swallowing evaluation with modified barium swallow (MBS)
- If GERD unresponsive to standard therapies, consider esophageal manometry

GERD-Management

- Behavioral
  - Weight loss: if overweight or obese
    - Mealtime: Timing: At least 3 hours before bedtime/lying down
    - Foods: Avoid caffeinated drinks, fatty foods, chocolate, peppermint, citrus. Promote high fiber, high protein foods.
    - Eating: Avoid overeating, rapid eating
    - Sleeping: Elevate head of bed at least 6 inches
    - Treatment-resistant: Right-inclined, side-lying position during mealtimes
    - Thickening of liquid to pudding consistency
    - Separate times for food and liquid ingestion

GERD-Management-Medications

- Initial therapy:
  - Proton-pump inhibitors (PPIs) most effective RX
  - All PPIs have similar efficacy at standard dosage
  - Administer 1 hour before meals
- Eradication of Helicobacter pylori no longer thought to improve or worsen GERD
- Maintenance Therapy:
  - PPIs maintain remission more than H2RAs
  - Addition of H2RA @HS to daily PPI decreased nocturnal acid
  - Less than daily dosing of PPIs not recommended as maintenance therapy in patients with history of erosive esophagitis

Focus #3: Mental/Behavioral Health

Look for evidence of these common mental health conditions that are often unrecognized and untreated
- PTSD
- Mood Disorder
- OCD
- Anxiety
- Dementia

Consult: PTSD

- Presenting topics: 61-year-old female with profound intellectual disability and autism; recent series of hospitalizations for recurrent pancreatitis with weight loss. Longstanding behavioral issues of sleeplessness, physical aggression, and rectal digging. Recurrent hospitalizations for lethargy, alternating with persistent screaming and early morning awakening with sleeplessness.

PTSD-Background

- Trauma-related anxiety disorder
- Often cyclical and progressive
- Compromise biological, psychological, social, spiritual functioning
- General population prevalence: 5-10%
- Prevalence in IDD population: unknown

She was reportedly burned with cigarettes, and sustained a fractured arm, due to abuse.
### PTSD- Diagnosis-Adapted Criteria

**Criteria**
- Traumatic event or threat
- Response-fear, helplessness, horror
- Recollections
- Recurrent dream of event
- Avoid activities, places, or people that arouse recollection

**Adaptation to IDD**
- Range of potential events
- Disorganized or agitated behavior, especially with severe/profound ID
- Act out or SIB
- Dreams w/o content
- Trauma-specific enactments appear psychotic
- May be interpreted as non-compliance

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### PTSD-Evaluation & Treatment

- Thorough medical evaluation because of high medical co-morbidity
- Investigate the nature, circumstances, contexts of trigger events
- Recognize co-existing mental health disorders
- Avoid inappropriate use of antipsychotic and other medications
- Change environment to eliminate frightening cues
- Training & support of caregivers to respond appropriately
- Cognitive-behavioral therapy
  - Exposure therapy
  - Imaginary rehearsal therapy

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### Focus # 4: Mental/Behavioral Health

Evaluate for medical causes of neurobehavioral disorders
- Self-injurious behavior
- Aggression
- Stereotypy
- Decline in Adaptive Behavior

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### Case Study: Aggressive Behavior

60 y/o male with impulsive aggressive behavior

**Problem List:**
- Medical: Cerebral Palsy; Hypothyroidism; Constipation
- Neuro-behavioral: Severe intellectual disability; Epilepsy; Impulse Control Disorder; Anxiety

**Behaviors:**
- Picking skin; grinding teeth; sudden aggression; left arm twitches; pelvic gyrations
- Possible focal seizures
- Repetitive arm twitching despite triple AEDs
- Behavioral disturbances may occur before, during, or after temporal lobe seizures
- Video EEG

**Medications:**
- Medical: Levothyroxine 100 mcg; Calcium 500 /Vitamin D 250 Units QD; Colace 60 mg
- Psychotropic: Divalproex 750 mg BID & 625 mg at noon; Carbamazepine 200mg BID; Gabapentin 1200 mg TID; Seroquel 50 mg TID

**Past Surgical History:** Ventriculo-peritoneal shunt
**Past Medical History:** Hyponatremia
**Imaging:** CT brain: moderate ventriculomegaly. Right porencephaly. Right anterior temporal encephalomalacia. Old infarctions basal ganglia.

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### Case Study: Aggressive Behavior

- Focal brain pathology: right temporal lobe injury
- Anxiety
  - Skin picking and bruxism
- Benztropine may paradoxically worsen disruptive behaviors when used to treat anxiety in persons with brain injury and/or ID
- Video EEG
- Possible Tardive Dyskinesia
  - Pelvic gyrations suggest possible tardive dyskinesia vs. features of temporal lobe sx
- Syndrome-specific surveillance: common conditions co-morbid with CP
  - Constipation; degenerative joint disease; dysphagia; incomplete bladder emptying; spinal stenosis; spinal spondylosis.

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### Decline in Adaptive Functioning (Example: Individual with Down syndrome)

**Document extent and course of changes in adaptive functions**

**Perform** comprehensive medical assessment as follows:

**Review of medications**
- Focus: Psychotropic, anti-epileptic, anti-cholinergic, most recently introduced medications

**Screen for mental health issues**
- Focus: Depression, anxiety, trauma, grief, loneliness, boredom, disordered sleep

**Assess social environment and networks**
- Focus: Changes in home, work, peer and family relationships; changes in work/day program, home and recreational environments

**Review chronic diseases**
- Focus: Review clinical status and treatment of known chronic diseases (e.g., congenital heart disease, obstructive sleep apnea, swallowing disorder, hypothyroidism, celiac disease)
### Decline in Adaptive Functioning (Example: Individual with Down syndrome)

**Review Down syndrome-specific screenings**
- Focus: Utilize Down syndrome Preventive Healthcare Checklist or similar published guideline to update deficient screenings

**Consider undiagnosed occult disease**
- Focus: Cardiac and pulmonary diseases, caries and periodontal disease, hypothyroidism, vitamin B<sub>12</sub> deficiency, H pylori gastritis, gastrointestinal ulcer or GERD, constipation, sleep disorder, epilepsy

**Perform nutritional screen**
- Focus: Underweight, obesity, specific nutritional deficiencies (iron, vitamin B<sub>12</sub>, folate, vitamin D)

**Consider unrecognized pain**
- Focus: Occult fracture, osteoarthritis, degenerative spinal disease, osteoporosis, gout, neuropathies

If above assessments are unrevealing, perform focused assessment for dementia

### Focus # 5: Pharmacotherapy

Maintain constant vigilance for potential dangers and errors related to medications
- Serotonin syndrome
- Neuroleptic malignant syndrome
- Drug-induced movement disorders
- Lactulose
- Anticholinergics
- Bladder Agents
- Antiepileptic Drugs
- Co-administration Errors

#### Serotonin Syndrome

- Defined as a constellation of clinical findings attributed to excessive serotonin activity in the brain, typically induced by a combination of psychoactive medications that affect brain serotonin levels
- Clinical features:
  - Anxiety, agitation, delirium, restlessness, disorientation, easily startled
  - Autonomic nervous system:
    - Increased sweating, heart rate, temperature, vomiting, diarrhea
  - Neuromotor hyperactivity:
    - Tremor, muscle rigidity, myoclonus, hyperreflexia, Babinski sign especially affecting legs

#### Anti-Epileptic Drugs

- AEDs commonly affecting cognition
  - Primidone, Phenobarbital, Phenytoin, Topiramate
- AEDs commonly associated with weight gain
  - Valproate, Gabapentin, Pregabalin
- AEDs commonly associated with weight loss
  - Topiramate, Zonisamide
- AEDs associated with low bone mineral density
  - Phenytoin, Phenobarbital, Primidone, Carbamazepine, Valproate

### Critical Understandings

- Etiologic Diagnosis
- Baseline Adaptive Functions
- Communication Abilities & Supports
- Service & Support Network
- Preferences, Interests, Triggers, Comforts
- Mental & Behavioral Health History

### Communication

Caring for adults with IDD who have dementia:
- Prepare for non-verbal communication methods
- Altering communication style and keen observation skills are critical
- Overemphasis on gestures, expressions and use of objects can be highly valuable
- Preparing staff through training and reminders of a broad observance (e.g. person’s hand motions, difference in expression from baseline/normal, etc.)
- These may apply to IDD individuals who do not bear the diagnosis of dementia

*Guidelines for structuring community care and supports for people with intellectual disabilities affected by dementia*. National Task Group on Intellectual Disabilities and Dementia Practice.
Case 1: Tony, age 52, with Down syndrome recently admitted to Assisted Living Facility

Tony is age 52 and has Down syndrome and moderate intellectual disability. He and his mother simultaneously entered an Assisted Living Facility (ALF) when she began falling frequently and was found to have early dementia. They now share a suite in the facility. Previously, Tony had lived with his mother all his life in the family home.

Since the move, Tony has regressed and is no longer brushing his teeth, showering, or changing his clothes without prompting; all of these he used to do independently, without prompts.

Prior to admission to ALF, Tony had not seen a doctor for years. How would you assess and intervene with Tony?

Case 2: Lucinda, age 67, with cerebral palsy, post-operative hip fracture, in Post-Acute Care

Lucinda has hemi-paretic cerebral palsy, mild intellectual disability and previously well-controlled epilepsy. She was living in a supported apartment complex with visitation by a direct support professional for two hours every morning and every evening. One week ago, she fell at her day program and fractured her hip. Following operative repair, she is undergoing rehabilitation in the Post-Acute Care unit where you practice.

Upon arrival, Lucinda has a tonic-clinic seizure. Upon return to consciousness, she is complaining that her repaired hip hurts. Describe your plan for immediate and longer-term post-acute care.

Case 3: Darren, age 48, with Down syndrome and dementia newly admitted to Intermediate Care Facility

Darren is age 48 was admitted to the Intermediate Care Facility with diagnosis of Down syndrome complicated by early-onset dementia. His father had died suddenly from a stroke 5 years ago, and his mother died from an acute myocardial infarction 2 years ago. He lived with a brother who reluctantly assumed his care following mother’s death.

What initial assessment would you conduct upon his arrival? The nursing and direct care staff have no previous experience with residents with Down syndrome. How would you counsel the staff to optimize Darren’s adjustment to his new residence?

Case 4: Michael, age 30, with autism, epilepsy and severe intellectual disability

Michael lives with his single mother Lena. He has diagnosis of autism, epilepsy and severe intellectual disability. His mother is arranging weekend respite care at the long-term care facility where you are medical director. What would you want to know about Michael? How can you work with your staff to prepare to provide respite care at your facility for adults with developmental disabilities?

Resources

Local Resources
- Local county board of DD
- Local residential and other service providers, advocacy organizations

National Organizations
- AANMD = American Academy of Developmental Medicine and Dentistry [http://aamnd.org/]
- RRTCDD = Rehabilitation Research and Training Center on Developmental Disabilities and Health [http://www.rrtcdd.org/]
- The Arc – National advocacy organization & state and local chapters [http://www.thearc.org/]

Reference Materials
- Genetic Testing Registry: Medical genetics information service [https://www.ncbi.nlm.nih.gov/igt/]

Thank You

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