Managing Developmental Disabilities and Mental Health Conditions in Primary Care: A Quality Indicator Approach

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Objectives

Define developmental disabilities.

Describe the Regional Center system, clients, and services

Describe presentations of people with developmental disabilities and mental health conditions

Discuss recognition, evaluation and treatment using a quality indicator tool

Answer questions

Developmental disability definition

People with intellectual disability (ID), cerebral palsy (CP), autism, epilepsy & conditions similar to intellectual disability

and

significant limitations in 3 or more areas of life functioning:

learning, expressive and receptive language, mobility, self-care, self-direction, economic self-sufficiency, capacity for independent living

Manifested before 18 yrs of age

Developmental disabilities: epidemiology

– Poverty	1 in 6 children
– Developmental disabilities	1 in 10-20
– Asthma	1 in 12
– Intellectual disability (MR)	1 in 33-62
– Epilepsy	1 in 100
– Infant mortality rate	1 in156 live births
– Autism spectrum d/o	1 in 150-350
– Cerebral palsy	1 in 200-300
– Childhood cancer (all)	1 in 333
– Down syndrome	1 in 700
– Orthotopic liver transplant	1 in 160,000 children
– Chronic granulomatous dz	1 in 4-5 million

What is the Westside Regional Center?

- State and federally funded, independent, non-profit, community-based organization
- Mandated by law to provide services, case management and care for individuals with developmental disabilities from birth to death
- Serves over 7000 clients in western Los Angeles county

Regional Center Clients

2 Groups:

1. Children <3 years old (Early Start) at risk for developmental delay or with developmental delay

2. Children and adults ≥3 years old with a “developmental disability”

Autism Spectrum Disorder

- DSM-V Definition: **pervasive** developmental disorder, present in early childhood, consisting of impairment in

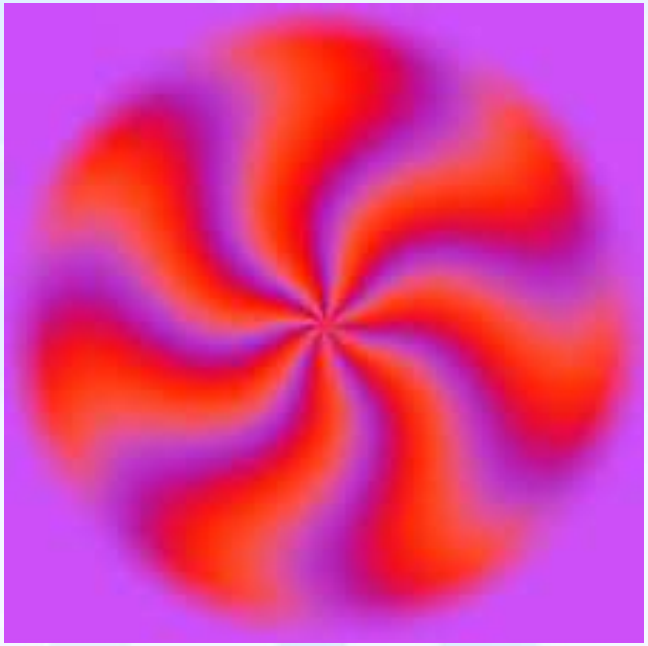
  (1) social communication and social interaction

  (2) restricted, repetitive and stereotyped patterns of behavior, interests, and activities

- Across contexts
- Not accounted for by general developmental delays

RRBI: Sensory Processing

- **Deficits in detecting, modulating, interpreting and responding appropriately to sensory input**
- **Visual**
- **Auditory**
- **Tactile**
- **Taste/smell**
- **Position/movement**



RRBI: Sensory Processing

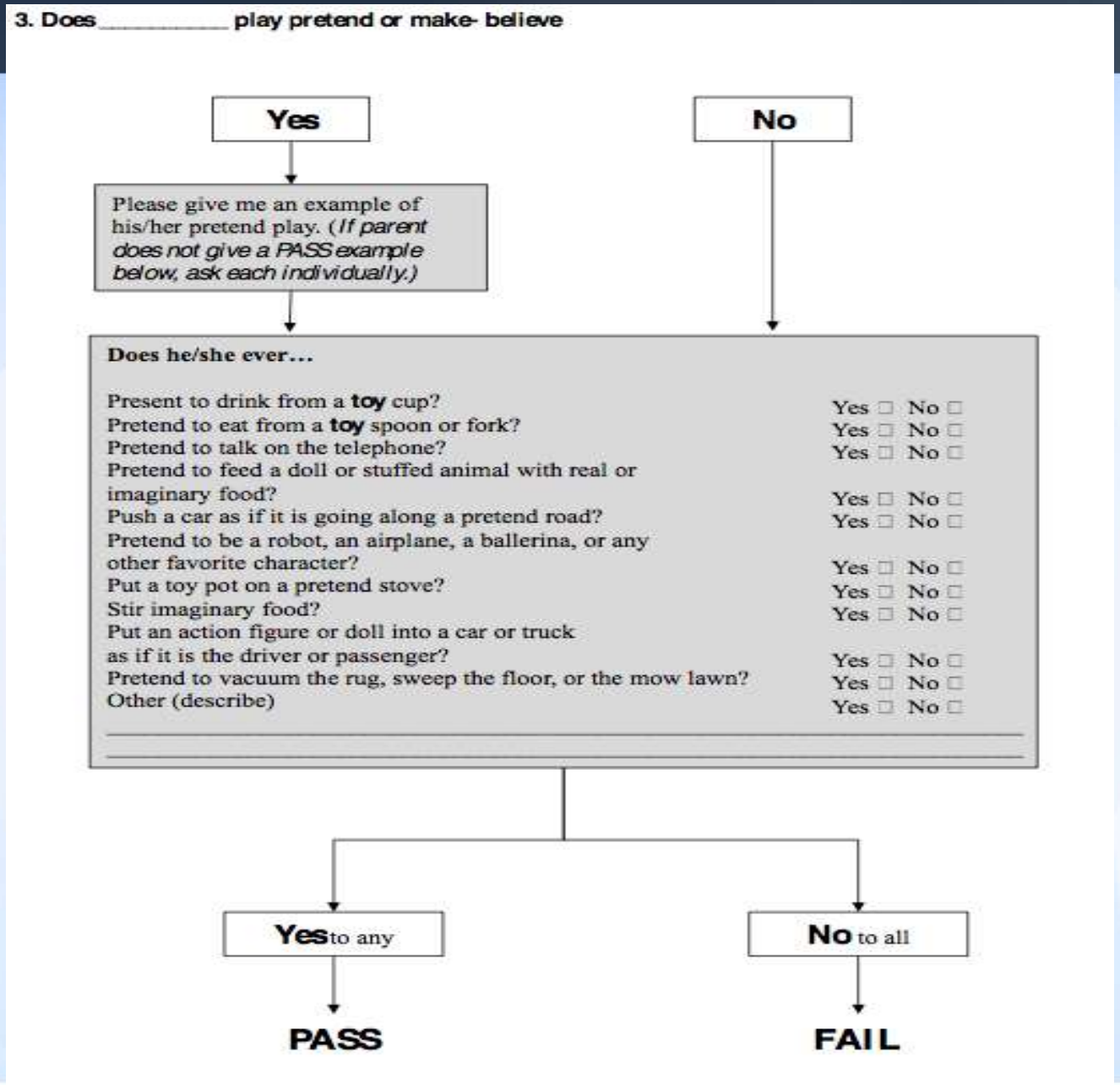
- Covering ears at restaurants, with loud music, at birthdays
- Bouncing on barricades or against other children while standing in line
- Restricted diet to less than 10 foods of only soft texture
- Refusal to go in sandbox, step on grass, have shoes off, tags must be taken off clothes
- Insensitivity to heat, cold or pain

Screening for ASD and RED FLAGS

- Screen for ASD at 18 mo & 24 or 30 mo (AAP, 2007; CDC 2014)
- Tools: all perform better at 24-36 mo than 18-24 mo
  - PDD-ST (PPV = 0.06)
  - M-CHAT (PPV = 0.06)
  - M-CHAT-R + Follow-up interview (PPV = 0.57 for DD or ASD)

- RED FLAGS: Immediate referral
  - no babbling or pointing or other gesture by 12 mo
  - no single words by 16 months
  - no 2-word (non-echolalic) phrases by 24m
  - loss of language or social skills at any age (20% of children with ASD have regression) (Child Neurology Society 2004, NICHD 2009, CDC 2014)

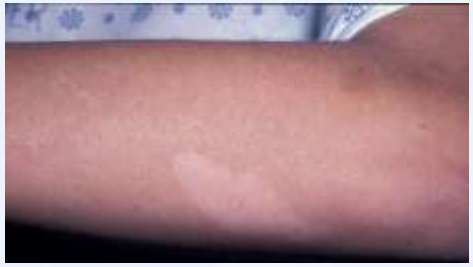
M-CHAT-R Follow-up Interview



ASD Initial Evaluation/ Work-up

- Multi-disciplinary assessment: multiple professions observing child and parents on multiple occasions

- Thorough H & P, including pre/post natal hx, FH, development, regression, head circ, growth, neuro, skin



- Audiology testing
- Labs:
  - **Lead, High resolution chromosomes, Fragile X testing Comparative genetic hybridization (CGH)**
- Imaging: autism + ID or + FH = MRI brain (AAN, CNS 2005)

ASD Treatment Strategies

- **Early Intervention**
- **Behavioral programs:**
  - **ABA (DTT, Pivotal Response)**
  - **FloorTime (DIR)**
  - **RDI, TEACH**
- **Speech therapy, OT**
- **Highly structured programs, social skills training**

Adjuncts:

- Medications: Risperidone (FDA-approved Rx)
- Psychiatrists use other antipsychotics, stimulants, clonidine, AEDs, SSRIs but scant evidence and no long term safety

ASD Follow-up and Prognosis

- Repeat cognitive and multi-disciplinary assessment at 5-6 yrs if initial dx at ≤ 3 yr
  - **20-40% children dx'd at ≤ 3 yr will not have ASD dx at 7 yr**
  - **Cognitive dx (ID) often "deferred" until 5-6 yr but needed for treatment planning and prognosis**
  - **Individual Educational Plan (IEP) rec's**
- To evaluate therapies:
  - Address when beginning, document behaviors & fxning b/f and a/f, use objective measures, have therapeutic endpoint
- Chronic disorder
  - Majority of children with autistic disorder continue to require significant supports as adults

Intellectual disability: classification

- Intellectual disability ≠ developmental delays
  - May use DD early on when IQ tests not available or predictive but once IQ and adaptive known, use ID

- Levels of Intellectual disability (old):

IQ 50-55 < ~70 - Mild intellectual disability (85%)

IQ 35-40 < 50-55 - Moderate intellectual disability (10%)

IQ 20-25 < 35-40 - Severe intellectual disability (4%)

IQ < 20-25 - Profound intellectual disability (1%)

Reflect IQ measurement error of ~5 pts. (DSM IV-TR)

Intellectual disability: Functioning and Prognosis

- Mild ID: ~ 6th grade academics, self-supporting, social and vocational skills, may need guidance especially under stress
- Moderate ID: ~2nd grade academics, learn to travel independently in familiar places, unskilled or semiskilled work under supervision, adapt to community life, usually supervised
- Severe ID: may learn to talk during academic yrs, elementary self-care skills, closely supervised group homes or family settings
- Profound ID: most have identified etiology, considerable impairment, constant aid and supervision (DSM IV TR)

Intellectual disability: treatments

- Learning supports, special education
- Speech therapy, occupational therapy, physical therapy, behavioral services
- Independent/supported living services
- Social supports
- Job training/coaching, housing, aging services

- Co-morbid neurological and mental disorders:
  - **Seizures, cerebral palsy**
  - **Child: ADHD, PDD/ASD, stereotypic mvmt d/o**
  - **Adult: depression, anxiety, OCD, PTSD**
  - **Syndrome assoctns: Down: dementia in 50s; fragile X: ADHD, social phobia; Prader Willi: hyperphagia, OCD; Williams: anxiety, ADHD**

When You Say "RETARD" Someone HURTS.

Trauma and Abuse

60-80% of people with developmental disabilities have been physically or sexually abused.

Clinical Etiquette for Developmental Disabilities

1. Do you sit at eye level?
2. Do you speak directly to the patient instead of a companion or interpreter?
3. Do you introduce yourself and shake hands?
4. Do you wait expectantly for any response to a question? Even up to 90 seconds?
5. Do you wait for people to finish even if they have difficulty speaking or if they are using an augmentative communication device and they are slow?
7. Do you treat adults as adults? Do you use shorter sentences, ask questions that require short answers, use more concrete language, and pair words with visuals such as gestures or pictures? Do you use baby talk or sweet names (hunny, sweetie)?
8. Do you treat wheelchairs and guide dogs as extensions of a person's body, not leaning on them or touching them without permission?
9. Do you wait until offers of help are accepted and listen or ask for instructions?
10. Do you tap a person with a hearing difficulty on the shoulder or wave a hand to get their attention, then speak slowly and clearly to see if the person can read lips, using a normal volume?

The Other Dual Diagnosis: Mental Health

- 20-30% of people with developmental disabilities have co-morbid mental health conditions
- Under-diagnosed and over-treated!
- Diagnostic overshadowing
- Chemical sedation, especially in group settings

Tools for Assessing Quality of Mental Health in Developmental Disabilities

- Access: how difficult to get into care?
- Assessment: how adequate are the evaluations to determine if mental health disorder is present?
- Treatment: how appropriate are the treatment modalities used to improve relevant mental health outcomes?

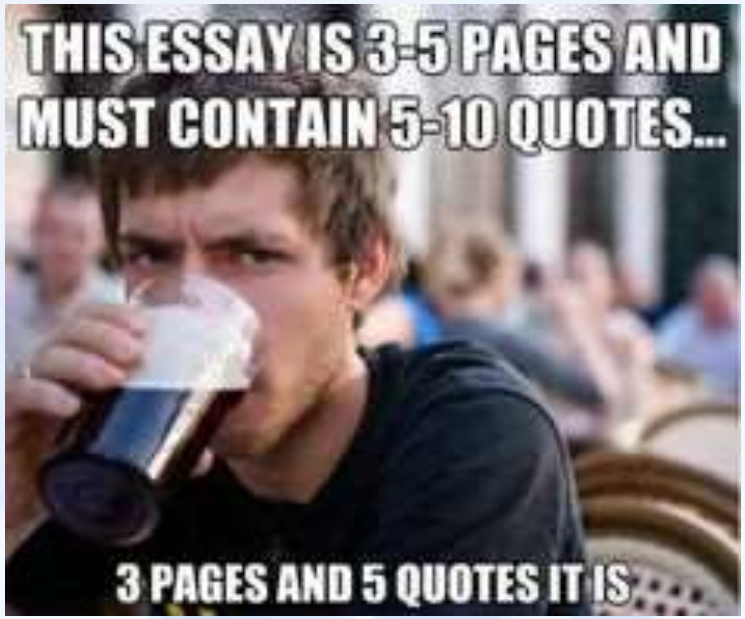
Mental Health Assessment Quality Indicators

Did you document…

- ☐ means of communication?
- ☐ strengths AND difficulties in functioning?
- ☐ psychosocial stressors?
- ☐ impact of the mental health symptom on life functioning?
- ☐ prior history of mental health care?
- ☐ prior strategies/approaches to help deal with mental health and behavioral problems?
- ☐ environmental changes?
- ☐ history of prior ST, OT, PT, behavior?
- ☐ prior psychotropic medications?
- ☐ medical, developmental, medication history?
- ☐ abuse history (physical, sexual)?
- ☐ aggression? SIB? SI/SA?

Mental Health Assessment Quality Indicators

- ☐ a mental status examination?
- ☐ a physical examination?
- ☐ a follow-up plan?
-
- ☐ Did the primary provider or mental health professional provide a treatment goal?
- Bare Minimum!



Mental Health Treatment Quality Indicators

- ☐ At least one treatment goal?
- ☐ progress in meeting that goal?
- ☐ A recommendation for an evidence-based psychotherapy?
- ☐ A provision of at least one evidence-based psychotherapy session? OR
- ☐ A clinical rationale for not recommending an evidence-based psychotherapy?

For psychotropic meds:

- ☐ target symptom(s) that medication is supposed to address?
- ☐ informed consent
- ☐ monitoring: med rationale, name, frequency, dosing, side effects, follow-up?
- ☐ If 2 antipsychotics, give a rationale for both or tapering?
- ☐ Baseline and every monitoring visit: BMI, labs, EPS?
- ☐ If med stopped, reason for d/c?

Cerebral Palsy: definition

- Non-progressive **motor** impairment present from early in life
  - **Associated with abnormal tone & reflexes, spasticity, involuntary movements &/or ataxia** (Trauner 2008)
- Descriptive term: NOT A DIAGNOSIS!
- Does NOT imply level of cognitive function
  - **Only 30-50% of those with CP have any ID/MR** (Odding, 2006; Russman, 2004)

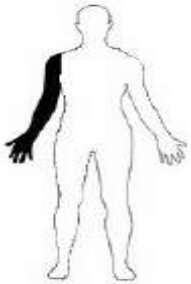
Cerebral Palsy: Classification

Spasticity = velocity-dependent increase in tone, spastic CP is 70% of CP

Hemiplegia
One side of the body is affected. The arm is usually more involved than the leg

Diplegia
All four limbs are affected. Both legs are more affected than the arms.

Quadriplegia
All four limbs are involved.



Monoplegia
Only one limb is affected, usually an arm.

Triplegia
Three limbs are involved, usually both arms and a leg.

Cerebral Palsy: Classification

- **Dyskinetic CP** 15-20%, (athetoid CP, choreoathetoid CP, and dystonic CP) - extrapyramidal signs characterized by abnormal movements, hypertonia
- **Ataxic CP** – <10%, cerebellar involvement, with weakness, incoordination, wide-based gait, imbalance
- **Hypotonic CP** – ? rare, truncal and extremity hypotonia with hyperreflexia and persistent primitive reflexes
- **Mixed CP** - no single tone predominating; typically mix of spastic and dyskinetic components

Cerebral Palsy: Red Flags

- AAP: Evaluate and refer for motor delays at 9 month visit
- RED FLAGS for Motor delays:
  - Not rolling normally by 6 mo
  - Not sitting without support by 7 mo
  - Stiffness or floppiness
  - Not crawling by 12 mo
  - Not walking by 16 mo
  - Any early hand dominance (<2 y)
  - Toe walking at 2 yr or ≥50% of time or with extensor Babinski

Cerebral Palsy: Evaluation/ Work-up

- Exam:
  - hypertonia, hypotonia, contractures, “spastic catch”
  - Reflexes: hyperreflexia due to UMN lesion
  - Persistent primitive reflexes (ATNR, Moro)
  - Gait: circumduction
  - Cerebellar: ataxia

- Imaging: MRI of brain & ± spinal cord
- Labs: if MRI normal, metabolic and genetic studies; if spastic hemiplegia, coagulation studies

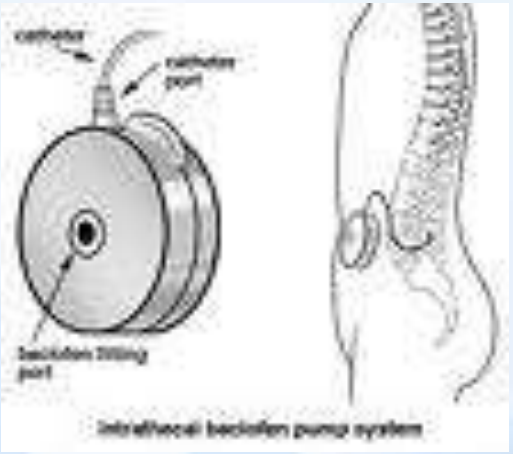
Cerebral Palsy: Treatment

- Therapies: physical, occupational, and speech
  - Comprehensive PT not showing global benefit
  - Specific Rx with efficacy:
    - Strength training for muscle strength, intensive UE for bimanual
    - PT more effective in short-term, intense intermittent rehab
    - Passive stretching: review 7 studies, limited evidence that ↑ROM or ↓ spasticity(Pin 2006)

- Mobility aids (AFOs, braces, crutches, standers, walkers, wheelchairs)

Cerebral Palsy: Treatment

- Medications:
  - Baclofen po
  - Botox: efficacy for spastic equinus, hip adductors, class II for UE
    - (AAN 2008)
- Baclofen intrathecal pump
- Surgery: tendon lengthening, dorsal rhizotomy



intrathecal baclofen pump system

Cerebral Palsy: Follow-up & Prognosis

- CP dx should be deferred until ≥ 2 yr & reassess at 7 y
  - 50% of children with CP and 66% of children with spastic diplegia "outgrew" CP by 7 yrs. Others w/o full motor signs of CP until 1-2 yrs.

    (Collaborative Perinatal Project)
- Spastic quadriplegia:
  - 25% no impairment, 50% moderately impaired but function is satisfactory, 25% non-ambulatory

Cerebral Palsy: Prognosis

- If sit by 2, ambulatory by 3 yrs, much better prognosis
- Co-morbidity:30% have seizures, 30-50% have ID
- Causes of M & M: contractures, fractures due to osteopenia, scoliosis/kyphosis, hip dislocation, aspiration pneumonia, FTT/malnutrition/constipation

Epilepsy

- Definition: d/o characterized by ≥ 2 unprovoked seizures
- Causes: abnormal electrical discharges of brain cells; sometimes genetic syndromes
- Treatments: medications (anti-epileptic drugs), ketogenic diet, vagal nerve stimulator, surgery
- Note: 2/3 of epilepsy can be controlled with medications; few people with epilepsy have MR

Collaboration with WRC

- Who to refer:
  - any red flags
  - any developmental concerns
  - any significant dx CP, seizure d/o, autism, MR, syndromes with associated delays
- What to expect from us:
  - Letter stating eligibility and services
  - Request for medical records initial and yearly
  - Calls re: pt issues, hospitalizations, services
  - Specialty consultation reports
- How to collaborate better?

Westside Regional Center Contact Info

- Main number: 310-258-4000
- Referrals:
  - Early Start: Claudia Osorio, Intake Coordinator, 310-258-4096, FAX: 310-258-0571
  - Over 3: Monica
- Health and Medical
  - Aga Spatzier, MPH, Wellness Coordinator

  310-258-4254
  - Alicia Bazzano, M.D., Ph.D., Chief Physician

  310-258-4213, aliciab@westsiderc.org

Welcome to our world!