

Approach to Primary Care in Special Needs Populations

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COI/Disclosure Information

*57th Annual Family Medicine Seminar
July 31 - August 3, 2014*

- I have no financial relationships to disclose

Objectives

- Common behavioral presentations of pain in a nonverbal patient with I/DD.
- Secondary conditions commonly seen in the patient with spastic quadriplegic cerebral palsy.
- Complications of management of behavior with AED's or psychotropic medications.
- Resources in New Mexico which can assist practitioners and caregivers of individuals with Intellectual and Developmental Disabilities.

Overview

- Adults with Intellectual/Developmental Disabilities (I/DD) can have different patterns of illness and complex interactions among comorbidities
- More difficulty accessing primary care than does the general population.
- Review the general, physical, behavioral, and mental health recommendations for adults with I/DD, especially for those conditions not screened for by routine health assessments of the general population.
- Atypical manifestations of pain and distress in adults with DD and long-term use of antipsychotic medications to address behavioral issues.

Common Behavioral Presentation of Pain in a Nonverbal patient with I/DD

- Agitation
- Irritability
- Screaming/yelling
- Weight loss
- Sleep changes
- Aggression
- Self Injurious Behavior (SIB)
- Withdrawal
- Elopement
- Changes in Eating- Hyperphagia or Decreased appetite

Case 1

- Self injurious and aggressive behavior in non verbal adult

Cerebral Palsy



Goals of Spasticity Management- The PCP Perspective

- Improve Function
 - Activities of Daily Living
 - Mobility
 - Ease of Care by Caregivers
 - Sleep
 - Overall Functional Independence
- Prevent Medical and Orthopedic Complications
- Prevent Deformity and Contractures
- Prevent Development of Pressure Areas
- To Reduce Pain

Medical Complications of Spasticity/Immobility

- Scoliosis
- Hip Dysplasia
- Contractures
- Cervical spinal disorders
- Pressure areas/Skin Breakdown
- Osteopenia/Osteoporosis/Fractures
- Falls
- Constipation
- Neurogenic Bowel/Bladder
- Swallowing Disorders/Dysphagia
- Difficulty Maintaining Ideal Body Weight/ Nutrition
- Difficulties with activities of daily living
 - bathing
 - dressing
 - eating
 - toileting
 - maintaining hygiene
- Poor sleep
 - Sleep Apnea
 - Pain/Discomfort
 - Necessity of being Turned by another person every 2 hours is disruptive to sleep
- Depression

Prevent Medical and Orthopedic Complications

Musculoskeletal

Metabolic

Oral Motor

Nutritional

Gastrointestinal

Respiratory

Integumentary

Urologic

Gynecologic

Sleep

Neurologic

Psychosocial

Musculoskeletal- Neuromuscular Scoliosis

- Irregular spinal curvature due to abnormalities of the myoneural (muscle-nerve) pathways.
- Generally most severe in nonambulatory patients.
- Curve progression is much more frequent than idiopathic scoliosis
- Progression continues into adulthood.
- Bracing does not prevent progression of the spinal curvature.

Untreated Scoliosis-Complications

- Cardiopulmonary Complications
 - Respiratory compromise
 - Heart problems- especially with curves over 100 degrees
- Gastrointestinal Complications
 - Reflux
 - Constipation
 - Disrupted anatomy of the internal organs
- Positioning Complications
 - Pressure points
 - Wheelchair
- Functional Implications
 - Use of hands
 - Positioning for safe Feeding
- Skin Integrity
- Pain
 - Degenerative disk or arthritic

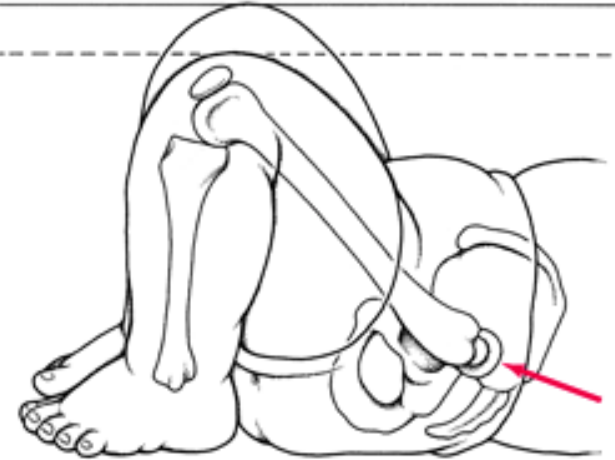
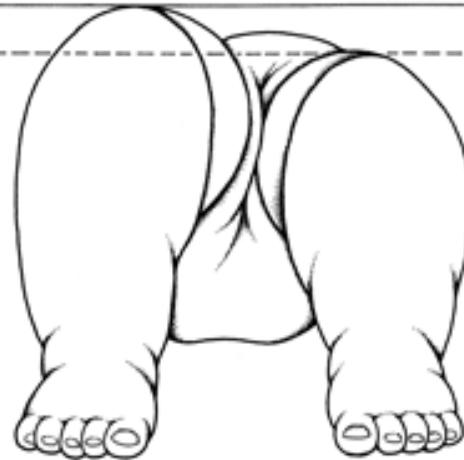
Untreated Scoliosis



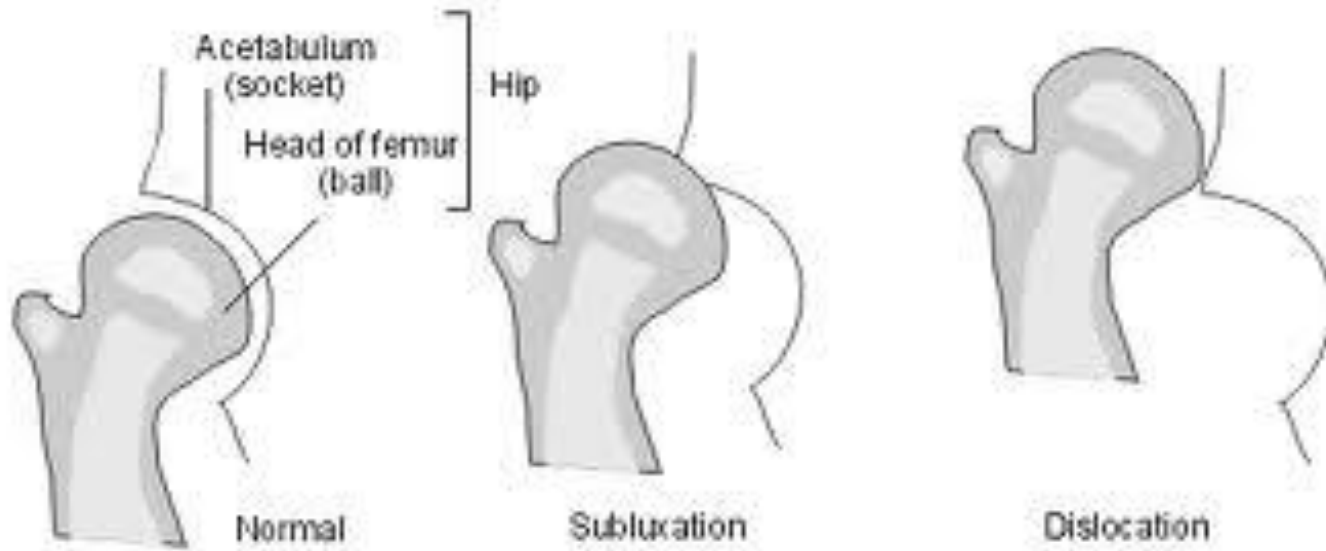
Musculoskeletal- Hip Dysplasia

- Hips are normal at birth
- Progressive hip subluxation occurs in up to 50% of children with spastic quadriparesis.
- Strong tone in hip adductor and flexors leads to scissoring and predisposes to hip subluxation and dislocation
 - dislocation is typically posterior and superior
- In time dysplastic and erosive changes in the cartilage of the femoral head can develop and lead to pain

Hip Dysplasia Physical Exam- Positive Galeazzi Sign



Hip Dysplasia



Osteoporosis Risk Factors

General Population

- Age over 50
- Female
- Menopause
- Family History
- Medications
- Low Body Weight/Being Small and Thin
- Broken Bones or Height Loss
- Not Enough Calcium and Vitamin D
- Too Much Protein, Sodium, Soda and Caffeine.
- Inactive Lifestyle
- Smoking
- Excessive Alcohol

Patients with Cerebral Palsy

- Immobilization
- Non Weight Bearing
- Low weight/Underweight
- Hormonal Issues
 - Menopause
 - Depo Provera
- Nutrition/Feeding Issues
- Low Serum Vitamin D levels
 - Seizure Medications
 - Lack of sun exposure

Nutritional/Oral Motor

Aspiration Risk

- Poor Oral Motor Skills
- Oral Dysphagia
- Pharyngeal Dysphagia
- Esophageal Dysphagia
- Positioning Difficulties
- Needing to be fed by others
- Tube Feeding

Chronic Aspiration can lead to:

- Recurrent Respiratory Infections
- Asthma/Wheezing
- Chronic Lung disease
- Pulmonary Fibrosis
- Cor Pulmonale
- Difficulty Maintaining Weight

Gastrointestinal

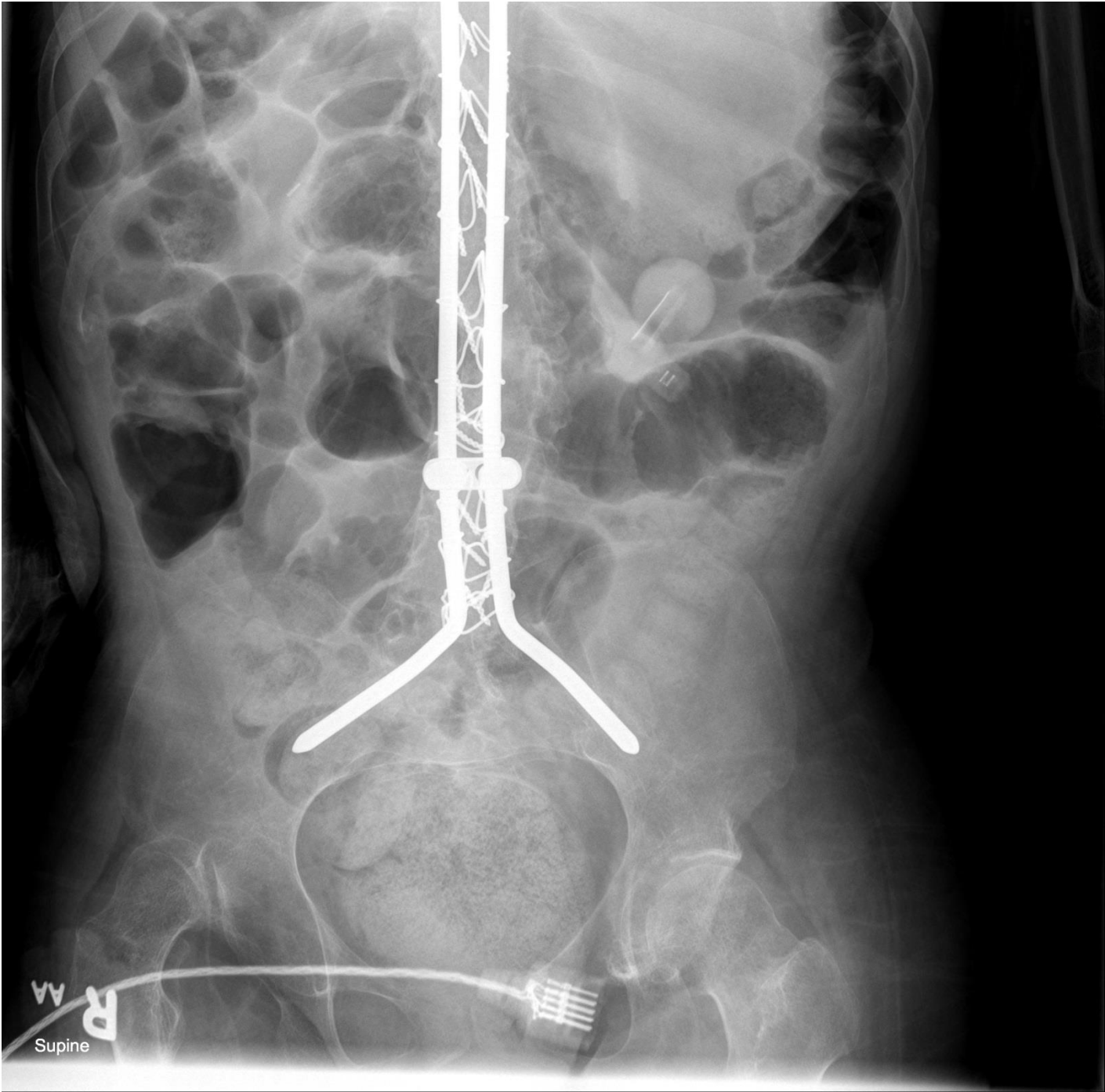
Gastroesophageal Reflux

- Frequent regurgitation or vomiting, especially after meals
- Coughing or wheezing
- Arching
- Self-injurious or aggressive behavior
- Screaming, Crying or Irritability especially after eating or at night
- Refusal to eat, at all or in limited amounts
- Failure to maintain weight

Gastrointestinal

Constipation

- Neurogenic Bowel
- Decreased mobility
- Slow gastrointestinal transit
- Inadequate fiber intake
- Stasis of stool, bloating, and impaction
- Worsens GERD



Drooling

Anterior Drooling

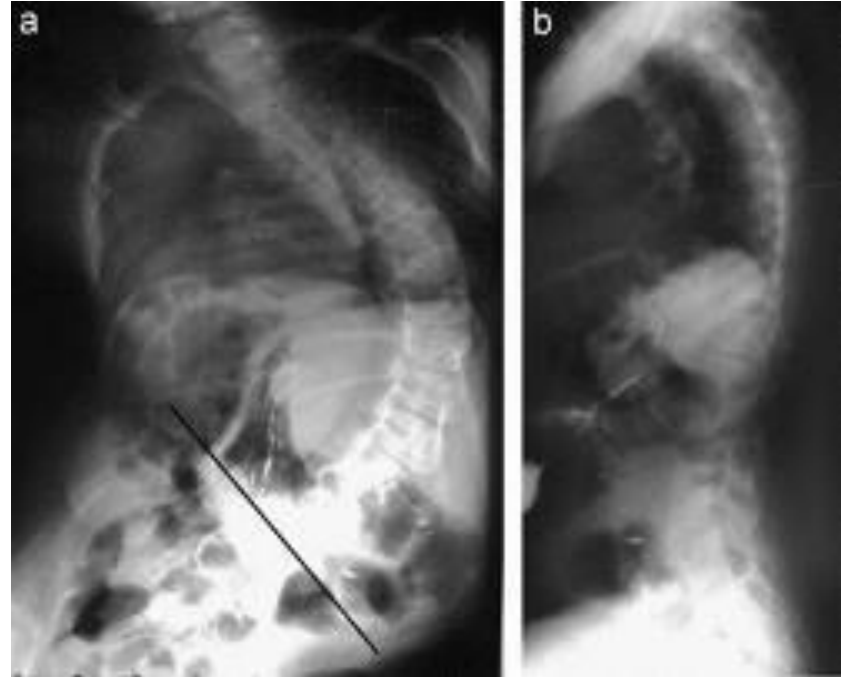
- Functional, social, psychological, and clinical implications
- Perioral skin breakdown and infections
- In severe cases of drooling, dehydration may even become a problem
- Social embarrassment may limit interaction with peers,
- May lead to isolation

Posterior drooling

- Problems with breathing, coughing, gagging, vomiting
- Aspiration
- Asthma
- Recurrent respiratory infections/pneumonia

Respiratory

- Scoliosis
- Aspiration
- Poor Chest Wall Motion
- Diaphragmatic Weakness
- Weak Cough
- Chronic Lung Disease
- Lung disease of Prematurity

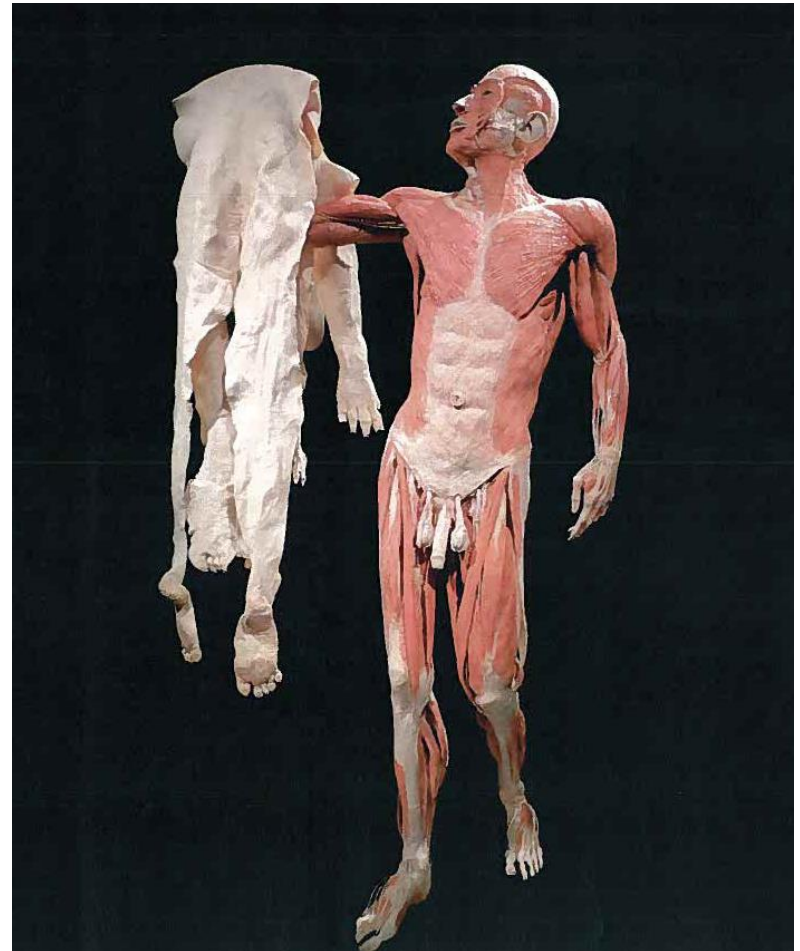


Integumentary

Prevent Skin Breakdown

Look at areas of greatest risk on the skin

- Bony prominences
- Skin contact: *anything that touches the skin*
 - Braces, TED hose, Bi-PAP masks, tubes, O2 tubing, NG tubing, heel/elbow foot protectors, Foley catheter, I.V. tubing and hubs, jewelry etc.
- If it is covered – uncover and inspect site
- Turn the person in order to do thorough head to toe skin assessment



Gynecologic

Menstrual Management

- Depo Provera
- Oral Contraceptives
- Analgesics
- Local Pain Relief- Massage, Warm Packs

Menopause

Medical Considerations

- Bone Density
- Risk of Blood Clots
- Menstrual Pain
- PMS
- Seizures

Personal Hygiene and Perineal Care



Neurogenic Bladder

- 80% with spastic hyperreflexive type neurogenic bladder on urodynamic testing
- Tendency for urinary retention and hyporeflexia in the adult over 30 years old
- New onset incontinence- consider:
 - Urinary retention (constipation)
 - Cervical spinal stenosis urinary tract infection
 - B12 deficiency
 - Seizures

Neurologic

- Seizures
 - Antiepileptic medications
 - Ketogenic Diet
 - Vagal Nerve Stimulator
- Shunt management
- Spasticity Management
 - Oral Antispasticity Medications
 - BoTox
 - Baclofen Pump
- Dystonia Management
 - Oral Meds
 - BoTox
- Sensory Deficits
 - Hearing- Sensorineural, conductive
 - Vision- Strabismus, Cortical Visual Impairment

Sleep

- Interrupted Sleep
- Pain
- Needing to be Repositioned
- Diaper Changes
- Craniofacial deformities
- Sleep Apnea
 - Obstructive
 - Central



CranioFacial

Normal



Mild



Moderate



Severe



Sleep Apnea

Obstructive

- Craniofacial Anomalies
- Laryngomalacia
- Reduced Upper Airway muscle tone
- Central Hypotonia
- Medication
- Body position (supine)

Central

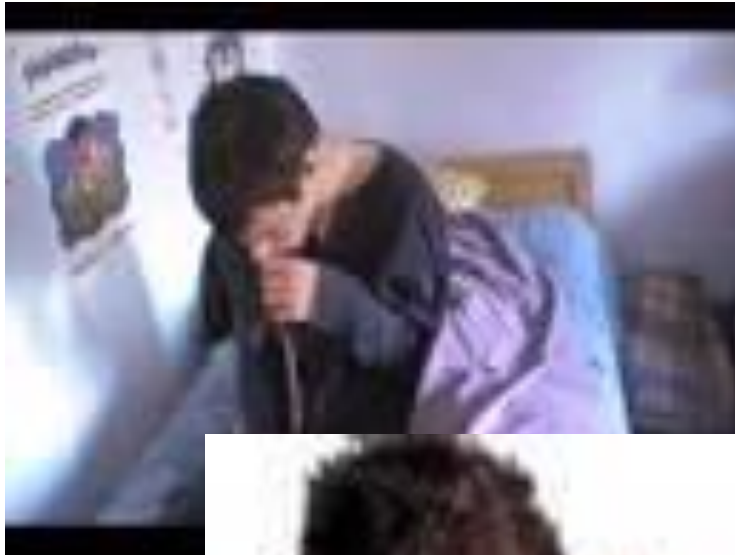
- Brainstem abnormalities
- Medications
- Normal variability of breathing in REM w/ exaggerated degree of desaturation
- Seizures (overt, subtle)

Depression

Signs of depression

- Change in Personality
- Increased anger, irritability, moodiness, aggression, self injury
- Change in appetite
- Change in sleep: difficulty falling asleep, staying asleep, or excessive sleeping
- Loss of energy, lethargy
- Loss of interest in friends, play, activities
- Low self-esteem, self-deprecating and negative talk

Problem Behaviors



Problem Behaviors

- Over 30% of people with ID have a comorbid psychiatric disorder
- Many patients with I/DD are on multiple medications aimed at managing Problem behaviors
- Polypharmacy is Common
- Side Effects and Interactions difficult to sort out-particularly in Non verbal or minimally verbal patient

“Behavior Management” in the I/DD population

- Assessment of target behavior/symptoms
- Rule out medical/environmental/psychosocial
- Initiation of treatment-
 - Start low, go slow, increase over weeks to months
- Change only one thing at a time
- Assessment of out-come and adverse effects,
- Follow-up
- Possibility of discontinuation of treatment
 - Go slow- taper over weeks
 - Change only one medication at a time

Psychotropic Medications- Side Effects

- Weight gain
- Metabolic abnormalities
 - glucose tolerance
 - lipid metabolism
 - prolactin secretion
- Cardiac conduction problems
- Dysphagia/feeding difficulties
- Bowel dysfunction
- Involuntary Movements
- Akathisia
- Acute Dystonia
- Extrapyrarnidal Symptoms
- Tardive Dyskinesia
- NMS

Antiepileptic Medication- Side Effects

- **Hyponatremia** (oxcarbazepine, carbamazepine)
- **Hepatotoxicity** (carbamazepine, phenytoin, valproic acid)
- **Blood dyscrasias** (phenytoin, carbamazepine, lamotrigine)
- **Vitamin D deficiency** (phenytoin, phenobarbital)
- **Hyperammonemia** (valproic acid)
- **Gingival hyperplasia** (phenytoin)
- **Osteoporosis** (phenytoin)
- **Cerebellar atrophy** (phenytoin)
- **Vitamin K deficiency** (phenytoin)
- **Carnitine deficiency** (valproic acid)
- **Folate Deficiency/Megaloblastic anemia** (phenytoin, phenobarbital)

Key Points

- Avoid Polypharmacy
- Try to stabilize the patient on a minimum number of medications prescribed at the lowest possible dose.
- Withdraw one medication at a time.
- Withdraw medication slowly.
- Allow time (sometimes a few weeks) after withdrawing one medication and before starting to withdraw another.

NEW MEXICO PREVENTIVE SCREENING GUIDELINES

- Adapted from US Preventive Services Task Force (USPSTF) “The Guide to Clinical Preventative Services”
- Adapted For Adults with Intellectual/Developmental Disabilities
- Some Syndrome specific recommendations
- Collaborative project by Continuum of Care (CoC) and Transdisciplinary Evaluation and Support Clinic (TEASC)
- Available electronically on COC website
- *<http://coc-cmstest.health.unm.edu/resources/guidelines.html>*

Some New Mexico Resources

- UNM TEASC Project
- Adult Special Needs Clinic
- UNM Continuum of Care Project
- Adult Cerebral Palsy Clinic
- DDMI Clinics
- SAFE Feeding Clinic
- DOH DDSD
- NM DOH Specialty Seating Clinic

Transdisciplinary Evaluation and Support Clinic (TEASC)

- Comprehensive, whole-person evaluations for people with developmental disability provided statewide
- Adult Special Needs Clinic, provided bi-monthly in Albuquerque
- Community-based physician consultations, provider support, technical assistance.
- Contact: Toni Benton, MD through the PALs Line 272-2000 or Liz Donsbach 505-272-5158
- *fcm.unm.edu/programs/teasc*

Adult Special Needs Clinic (ASNC)

- Team members include family medicine physicians, psychiatrists, neurologists, systems experts, clinical pharmacists, nurses, clinical dental hygienist, neuropsychologists, a variety of medical students/residents/interns.
- Offers whole-person, team evaluations for persons with developmental disabilities
- Meets twice monthly at the Family Practice Clinic on the UNM North Campus in Albuquerque
- Contact: Liz Donsbach 505-272-5158
or Patricia Beery 505-272-2579

fcm.unm.edu/programs/teasc

Referral Issues

TEASC/ASNC sees adults with developmental disabilities for a variety of issues, including:

- Complex behavioral concerns that may result from medical and/or psychiatric conditions
- Assistance in understanding and navigating available adult support systems
- Complex medical pictures that could benefit from transdisciplinary team evaluation

Continuum Of Care (COC)

- COC offers a wide range of services to help support individuals with disabilities or chronic illness, their families and those that support them.
- Has established a network of medical professionals at UNM and around the state with expertise in developmental disabilities who are available for consultation.
- To request consultation, call Main: (505) 925-2350; Fax: (505) 925-2389; or toll free 1-877-684-5259
- *coc.unm.edu*

Adult Cerebral Palsy Clinic

- Comprehensive evaluations include psychosocial assessments, full medical and neurological examinations, and facilitation of specialist referrals as necessary.
 - Family Medicine
 - Neurology
 - Social Work
 - Nursing
- Referrals include spasticity management, functional decline, falls, pain, agitation, unexplained weight loss, behavior changes.
- Contact: Vera Asplund, RN -505-925-2386

DDMI Clinics

- Collaboration with local psychiatrists and primary care
- Clinics for people who have co-occurring developmental disabilities and mental illnesses (DDMI).
- TEASC and Continuum of Care experts work with psychiatrists, families and IDT teams to provide services for individuals with I/DD who present with complex behavioral needs
- Clinical Sites statewide in Taos, Shiprock, Farmington, Roswell, Las Cruces, Silver City
- Contact Alya Reeve, MD, MPH, at 505-925-2395
or Eula Michaels- administrator at 505-925-2350

NM SAFE Program (Supports and Assessment for Feeding and Eating)

- Multidisciplinary team feeding evaluations of children and adults with developmental disabilities, for the purpose of improving health and preventing aspiration.
- The SAFE team includes a registered dietitian, physical therapist, physician and speech pathologist with expertise in swallowing disorders.
- Contact: Deirdre Muldoon, SLP
- <http://cdd.unm.edu/nmsafe>

NM DDSD Specialty Seating Clinic

- Based in Albuquerque with a team that travels around the state
- Custom fitted wheelchairs, molded seat formed specifically to the patients needs
- Physical Therapist evaluates and designs the seat for optimal function, safety, skin integrity
- Ideal option for patients who have significant skeletal deformities, contractures and compromised skin integrity
- Contact: 1-800-283-8415
- <http://nmhealth.org/about/ddsd/csb/swe>

NM DOH Developmental Disabilities Supports Division(DDSD) Clinical Services Bureau

- Provides information and referral services to people with disabilities and their families who are seeking help locating the right resources in their communities.
- Oversees various Medicaid home-and community-based waiver programs that are designed to help people with disabilities live as independently as possible.
- Contact: (505) 841-2948 Toll free 1-800-283-8415
- <http://nmhealth.org/about/ddsd/csb/>