Who Are People with Disabilities?

Anyone of any age can have a disability. People of all races and ethnicities can have disabilities. People with disabilities live throughout the United States, in towns, cities, and rural areas.

People with disabilities go to school and attend places of worship. They also vote, marry, have children, work, and play. To do all these things, people with disabilities need health care and health programs for the same reasons anyone else does—to stay well, active, and a part of the community.

Anyone can have a disability:
- An infant can be born with hearing loss.
- A child can become paralyzed by an injury from a car crash.
- A young adult can have depression or another mental illness.
- A woman in her early 30s can have multiple sclerosis.
- A man in midlife can develop type 2 diabetes.
- An older adult can lose her sight from glaucoma.

Different kinds of disabilities affect people in different ways. And the same disability can affect each person differently.
Several revisions have led to what is now found in 29 U.S.C.A. §706:

- “Individual with a disability means . . . Any person who (i) has a physical or mental impairment which substantially limits one or more of such person’s major life activities, (ii) has a record of such impairment, or (iii) is regarded as having such an impairment.”

“Severe Disability” includes:

- Amputation, arthritis, autism, blindness, burn injury, cancer, cerebral palsy, cystic fibrosis, deafness, head injury, heart disease, hemiplegia, hemophilia, respiratory or pulmonary dysfunction, intellectual disability, mental illness, multiple sclerosis, muscular dystrophy, musculoskeletal disorders, . . .

- Neurological disorders (including stroke and epilepsy), paraplegia, quadriplegia and other spinal conditions, sickle cell anemia, specific learning disabilities, end-stage renal disease, or another disability or combination of disabilities determined . . . to cause substantial functional limitation.”
The Special Patient

- How many persons are affected?
  - 19.3% are considered to be disabled
  - 54 million disabled U.S. citizens
  - 1.09 million disabled Marylanders

Based on 2002 U.S. Census

Dental Care For The Disabled Barriers to Care

- Financial
- Lack of trained personnel
- Lack of support for training
- Lack of recognition of the importance of oral health
- Difficulties in physical access

Stiefel, DJ. 2002

The Special Patient

- The Cost of Care
  - Healthcare for the disabled is Medicaid’s most costly function
  - Although PSHCN make up only 14% of Medicaid recipients, they consume about 37% of Medicaid expenditures
  - Medicare now pays for virtually all renal dialysis in the U.S.
Disability Prevalence
Arthritis or rheumatism (17.5%)*
Back or spine problems (16.5%)
Cardiovascular and Heart Disease (7.8%)
Lung or respiratory problems (4.7%)
Deafness or hearing problems (4.4%)
Limb or extremity stiffness (4.2%)
Mental or emotional problems (3.7%)
Diabetes (3.4%)
Stroke (2.8%)
Blindness or vision problems (3.4%)
Broken bone or fracture (2.1%)
Intellectual disability (mental retardation)** (2.0%)
Cancer (1.9%)
High blood pressure (1.7%)
Head or spinal cord injury (1.1%)

Dental Care For The Disabled
- Why is dental care for the disabled so limited?
  - Money! - or the lack of it!
  - Dental services are expensive
    - Disabled persons have lower employment and lower incomes than the general population
    - Medicaid provides only limited dental services and many dentists refuse to participate in Medicaid because reimbursement is so low

- The common thread is dependency
- Young special needs children, adult disabled and the frail elderly require assistance:
  - Transportation
  - How and where dental care will be obtained
  - Benefit from routine oral hygiene and preventive care
  - Correlate to poverty and inadequate health coverage
Dental Care For The Disabled

- Why is dental care for the disabled so limited?
  - Lack of willing and able providers
  - Fear and apprehension of Dental personnel
  - Fear of causing harm to the patient
  - Fear of lack of ability to provide competent care in view of the patient’s disabling condition
  - Fear of professional liability
  - Fear of personal injury from some patients

Dental Care for the Disabled

- 2005 HRSA “Dental and interdisciplinary workforce approaches to oral health for vulnerable and SN populations”
- “people who suffer from both health and social liabilities experience the least access to care, highest levels of treatment difficulty, the greatest disease burden, and the most intense consequences of poor oral health”

SPECIAL CARE DENTISTRY WILL INCLUDE A VARIETY OF PATIENTS WITH UNIQUE AND NEEDS.

William Osler, MD.
“Treat the patient with the disease not the disease the patient has”
Special Care Patients
Examples
- Patients receiving radiation
- End stage renal disease
- Cardiovascular disease patients
- Bleeding and clotting disorders
- Psychiatric disorders
- Disabled veterans, and war injuries
- Developmental disabilities

Summarizing Challenges
- Acceptance of treatment e.g. mobility, comprehension, tolerance, or cooperation
- Response to treatment and ability to receive treatment: e.g. need for antibiotics, drug adjustment or unfavorable tissue response (bleeding)
- Risk for disease: e.g. xerostomia, inability to perform OH
- Intraoral complications: e.g. muscular dysfunction, swallowing, limited opening.

THE FIVE ESSENTIAL CONCEPTS OF DEVELOPMENTAL MEDICINE
- Neurodevelopmental Dysfunction
- Underlying Neurodevelopmental Disorder
- Complications of Neurodevelopmental Dysfunction
- Health Consequences
- Syndrome Specific Conditions
NEURODEVELOPMENTAL DISABILITIES

Intellectual Disability
Down Syndrome
Autism
Cerebral Palsy

Intellectual Disability: An Overview

- A wide range of developmental conditions classified as a disorder by virtue of a significant cognitive impairment (below-average intelligence) coupled with deficits in adaptive functioning.
- Approximately 1-3% of all people in the United States
- There are many different causes for intellectual disability, and huge variations in degree of impairment and presence or absence of a wide range of behavioral, emotional, or medical problems.

Intellectual Disability: DSM IV Axis II

- A. Significantly below average intellectual functioning; an IQ of approximately 70 or below on an individually administered IQ test.
- B. Concurrent deficits or impairments in present adaptive functioning (i.e., the person's effectiveness in meeting the standards expected for his or her age by his or her cultural group) in at least two of the following areas: communication, self-care, home living, social/interpersonal skills, work, leisure, health, and safety.
- C. The onset is before age 18.
Subclassification of ID: DSM IV

- **Mild**: IQ 50-55 to 70
- **Moderate**: IQ 35-40 to 50-55
- **Severe**: IQ 20-25 to 35-40
- **Profound**: IQ below 20-25

- **Intellectual Disability, Severity Unspecified**
  - When there is strong presumption of ID, but the person’s intelligence is untestable.

Severity of Cognitive Deficit
Relative proportions (Volkmar & Dykens, 2002)

![Pie chart showing the distribution of severity levels.](chart.png)

- 85%
- 1%
- 4%
- 10%

Prevalence of ID: Basic Facts

- 3% of the population has an IQ two or more standard deviations below the mean.
- 2-3% of school-age children are classified as ID
  - Because they have poor adaptation to the academic demands of school.
- Only about 1% of the adult population is actually classified as ID.
  - Because they have adequate adaptive skills to live independently in the community.
30-70% of people with ID have co-morbid psychiatric disorder.

ID greatly complicates the process of psychiatric diagnosis, because of difficulty in communication and in conceptualizing symptoms and their antecedent context.

“Diagnostic Overshadowing”
- A common mistaken tendency to regard all abnormal behavior in persons with ID as due to the ID
- Leads to failure to recognize, properly diagnose, and treat psychiatric disorders in this population.

Normal distribution of standardized IQ tests theoretically indicates that about 3% of people should have IQ’s below 70.

IQ below 70 is an arbitrary definition of the cognitive deficit in ID.

Biological causes have been identified for an ever-growing percentage of cases of mental retardation within the 3% lower extreme of the normal distribution of IQ’s.

Exogenous:
- Psychosocial adversity: extreme poverty, severely disrupted childhood, lack of educational opportunity

Exogenous: biologically mediated
- Prenatal
- Perinatal
- Prenatal
Prenatal Etiologies

- Prenatal
  - Fetal alcohol syndrome
  - Infection (rubella, toxoplasmosis, cytomegalovirus, herpes simplex, hepatitis, HIV)
  - Substance use
  - Teratogens, medicine side effects
  - Environmental toxins
  - Poor maternal diet

Perinatal and Postnatal Etiologies

- Perinatal
  - Perinatal adversity or poor care
  - Prematurity
  - Increased salvage of very premature infants, who develop with cognitive and learning disabilities
- Postnatal
  - Malnutrition and other dietary deficiencies (iodine)
  - Extreme early neglect

Genetic Etiologies

- More than 750 known genetic causes
  - Inherited single gene disorders
    - Mutations
    - Deletions
    - Chromosomal aneuploidy, heteroploidy
    - Chromosomal translocations
    - Non-inherited chromosomal microdeletions
- Probably many polygenic causes
Genetically Based Neurobehavioral Phenotypes

- Complex disorders of behavior and cognition with known genetic etiologies that can be reliably diagnosed by laboratory tests.
  - Many have associated medical problems, anatomic malformations, or dysmorphic features.
- While these conditions are IQ lowering, not all people who have them are intellectually disabled.
- The number of genetic neurobehavioral disability phenotypes being recognized and diagnosed is growing.

Examples of Common Neurobehavioral Phenotypes: 1

- Fragile X Syndrome (in males)
  - Single gene triplicate repeat mutation, X chromosome
  - ID
  - Hyperactivity, gaze aversion, shyness, and social anxiety
- Down syndrome
  - Trisomy 21 (non-disjunction during gamete formation)
  - ID, with relative strength in visuospatial skills, and impaired expressive language
  - ADHD and Oppositional Defiant Disorder common

Examples of Common Neurobehavioral Phenotypes: 2

- Velocardiofacial Syndrome
  - Hemizygous 22q11.2 chromosomal microdeletion
  - Mean IQ 70
  - Characteristic learning disability
  - Attention problems, anxiety, and mood lability
  - 30% develop Schizophrenia in adulthood.
- Williams Syndrome
  - Chromosome 7 microdeletion
  - ID
  - Excellent Verbal Skills
  - Hypersociability
Data indicate that persons with ID have more untreated caries and higher prevalence of periodontal problems than the general population.

Periodontal disease:
Medications, malocclusion, multiple disabilities and poor oral hygiene combine to increase risk.

- Encourage independence on oral hygiene
- Involve caregiver if independence is compromised
- Use of antimicrobials e.g. Chlorhexidine, and appropriate delivery. Rinsing may not be best. Alternative, spray or toothbrush.
- Awareness of other meds and importance of daily oral hygiene
Dental Caries Strategies for Care

- Diet and noncariogenic foods. Note: food is often used as a behavioral control by caregiver.
- If taking medicines cause xerostomia, stress drinking more water. Sugar free medications, and to be sure to rinse following dosing.
- Fluoride and sealants as preventive measures.

Malocclusion

- Prevalence similar to general population.
- Exception, CP and DS
- Not necessarily a barrier to treatment.
- Ability of patient or care giver to maintain oral hygiene.
- Don’t lower expectations.
- Quality of life can improve with self-image.

Damaging Oral Habits

- Bruxism
- Mouth breathing
- Tongue thrusting
- Picking at gingiva
- Biting lips
- Pica
Trauma and Injury
- Falls or accidents
- Physical abuse
- State laws protect
- Educate caregiver

Health Challenges
- Mental
- Behavior
- Physical
  - Cerebral Palsy
  - Cardiovascular
  - Visual
  - Seizure
  - Hearing loss

Physical Challenges
- Clear paths for movement in treatment setting
- Place in center of treatment chair. Pillows on either side to bolster and support
- Be familiar with wheel chair transfer techniques
- Be prepared to treat in their wheelchair.
  - Note, other physical modifications may be covered in other lectures (see wheelchair transfer technique "Practical Oral Care for People with Developmental Disabilities," www.nidcr.nih.gov)
**Behavioral Challenges**

- Talk with caregiver and/or physician about techniques found to be effective
- Schedule early in day
- Keep appointments short
- Allow extra time
- Use caregiver to support comfort and communication
- Consider sedation
- Immobilization techniques only as last resort.

**Mental Challenges**

- Team approach, Staff briefing
- Reduce distractions
- Talk with parent or caregiver to familiarize with patient's abilities
- Address patient directly. “Talk to me”
- Be consistent
- Keep instructions simple
- Listen actively. Show understanding, respect, and be sensitive to method of communication

**DEVELOPMENTAL DISABILITIES**

- Down Syndrome
- Autism
- Cerebral Palsy
- Intellectual Disability
Early professional treatment and daily oral care at home can allow people with Down Syndrome to enjoy benefits of a healthy mouth.

Definition
- First described by John Langdon Hayden Down in 1866
- A syndrome of phenotypic abnormalities that occur as a result of chromosomal translocation during gametogenesis yielding an individual with trisomy of the autosome #21.
- The normal human has twenty-three chromosome pairs: 22 pairs of autosomes and one pair of sex chromosomes.
- The Down syndrome patient has 21 normal autosome pairs, one autosomal triplet (21G), and a pair of sex chromosomes.

Incidence & Prevalence
- Incidence: 1:660-700 live births
- Prevalence: 2/1,000 U.S. Population
- Incidence is directly proportional to maternal age:
  - 15-29 yrs: 1:1500
  - 30-34 yrs: 1:800
  - 40-44 yrs: 1:100
  - Over 44 yrs: 1:50
**General Phenotypic Characteristics**

- Small stature
- Pelvic dysplasia, waddling gait
- Flat facies with low-set ears & sloping forehead
- Slanting palpebral fissures (mongoloid eyes)
- Medial epicanthal folds
- Brachycephaly with flat occiput
- Hypotonia
- Hyperflexibility
- Hand anomalies: clinodactyly, simian crease

---

**General Phenotypic Characteristics**

- Mental retardation: rarely with an I.Q. > 50
  - I.Q. range 20-80
  - But social development is usually greater than or equal to the mental age
- Impaired cell-mediated immunity
  - Impaired leukocyte antimicrobial activity
  - Predisposition to infections
    - Frequent and recurrent URIs
    - Increased susceptibility to periodontal disease

---

**General Phenotypic Characteristics**

- Increased incidence of Leukemia
- Increased incidence of Hepatitis B
- Cardiac anomalies (VSD & ASD)
  - Cardiovascular disease in older life
  - Shortened lifespan
### Oral Manifestations

- Angles Class III skeletal malocclusion
  - Aplasia/hypoplasia of the maxilla
  - Deficient vertical anterior growth of the maxillae
  - Palate of normal height, but narrow, and often with prominent rugae and lateral processes
  - Mandibles with oblique gonial angles, with sloping mentum leading to double chin
- Bilateral dental crossbites
- Relative macroglossia

### Oral Manifestations

- Dental anomalies
  - Partial anodontia
  - Delayed dental eruption
  - True microdontia, predominantly in the mesio-distal dimension
- Saliva changes which may be the cause of decreased prevalence of dental caries
- Severe periodontal disease secondary to decreased leukocyte chemotactic ability

### Intellectual Disability and Down Syndrome

- Wide variation in level of ID—many have mild to moderate which limits ability to learn, communicate and adapt
- Language development is delayed
- Can understand more than can verbalize
- Daily living can be challenging and frustrating
Strategy for Care
- Listen actively, be patient and take time
- Prepare by talking with parent or caregiver to determine intellectual and functional abilities
- Allow time for patient to process information
- Keep instructions simple and concrete

Behavior Management
- Can be warm and well behaved, and very trusting.
- Caregiver/Parent can provide insight in patients personality and motivation
- Schedule early in day
- Prepare dental team
- Do things in stepwise approach and be consistent at each visit
- Tell show do
- Immobilization is last resort.

Medical Conditions
- Life expectancy has risen and increase of conditions related age increased as well.
- Cardiac Disorders are common (Mitral valve prolapse)
- Compromised Immune Systems (Aphthous Ulcers, Candida, ANUG) (Chronic Respiratory infections)
- Hypotonia can affect mastication, swallowing, drooling, and speaking. Also AAI (Atlanto-Axial Instability)
- Seizures
- Hearing Loss
- Visual Impairment (strabismus, glaucoma and cataracts)
Oral Health Problems

- Periodontal Disease
- Dental Caries
- Orofacial features
- Malocclusion
- Dental Anomalies
- Trauma and Injury

Case Presentation
Hunter

DEVELOPMENTAL DISABILITIES

- Autism
- Intellectual Disability
- Cerebral Palsy
- Down Syndrome
Autism is a complex developmental disability that impairs communication and social, behavioral and intellectual functioning.

First described in 1943 by Leo Kanner, a child psychologist as a neurodevelopmental disorder, usually appearing in the first 3 years of a child’s life. In 1944 Asperger later describe a milder form.

A severe developmental brain disorder that appears in infancy, persists throughout life, and is characterized by impaired social interaction, abnormalities in communication (verbal and nonverbal), and restricted interests.

Prevalence 1:20 10,000 for Classic Autism
3:50: 10,000 for entire Autism Spectrum Disorder.
Autism is a Phenotype

- A clinical diagnosis not biologic, and is genetic but also may be environmental, such as prenatal condition.

Possible Etiology

Not entirely known and no single cause has been identified, however:
- Fragile X
- Tuberous Sclerosis
- Prenatal factors including intrauterine rubella, cytomegalic inclusion disease and FAS
- Postnatal factors including untreated phenylketonuria, and,
- Some concerns have arisen over rubella (MMR vaccine. Anecdotal evidence linking ASD to the preservative (Thymerasol) has not been confirmed in clinical studies (CDC IOM, and discredited)

ASD

Identical Twins have 75 percent concordance in developing autism
Fraternal twins however have only 3%
Subsequent children of parents with autistic child have between 2-8% likelihood of a second child with autism
Autism Spectrum Disorder

ASD

- In recent years variations in behavior patterns have become apparent creating the so-called Pervasive Development Disorder Umbrella
  - Classic Autism, most severe with likely ID
  - PDD-NOS - (Not otherwise specified)
  - Asperger’s Syndrome, relatively normal language skills, decreased ability to show empathy, and unusual interests that are pursued with great intensity. (Words are taken literally) “Raining cats and dogs” . Not associated with ID and may be very intelligent.
  - Rett Syndrome with higher incidence in girls.

ASD

Incidence

- Classic autism occurs in males 4:1 vs. females.
- Current findings dispute the original estimate of 5/10,000 because of wider definition and the entire spectrum of disorders the CDC has reported incidence as high as 1/250 births.

Diagnostic Signs in Early Childhood

- Lack of babbling or gesturing by age 1 yr.
- Lacks eye contact with mother by 1 yr
- Resistance to being held or cuddling by mother
- Non responsive to name
- Non verbal, single words by 16 mo.
- Two word together by age 2
- General screening is strongly indicated.
- Early intervention, while not a cure, can increase potential for development.
SENSORY PROCESS DISORDER
Modulation
Discrimination
Sensory -Based Movement

DYSPRAXIA
Inability to perform voluntary purposeful movement.
(this has very important role in dental management.)

SPD
Over responsive (resist or avoid)
Under responsive (seeks)
SPD
- Over responsive
  - Touch - aversion, wiping, kissing
  - Olfactory - perfume, cologne, smell of materials
  - Texture - avoid crunch, rough
  - Proprioceptive - open, close, tongue
  - Vestibular - head position,
    - Visual - light
    - Auditory - music, drills
    - Taste - "white diet, bland,
    - Proximity

SPD
- Under responsive
  Touch - mouthing, food pocketing
  Taste - spicy, strong, salty
  Proprioception -
  Vestibular - can’t sit still
  Proximity - crowds in, s
  Visual - lights
  Auditory
  Olfactory

Autism Summary
- Aloof, distant or detached from other people or their surroundings
- Inappropriate reaction to verbal or social cues
- Obsessive routines, repetitive behaviors, unpredictable body movements, and self-injurious behavior may be symptoms that complicate dental care.
- May be accompanied by co-existing disabilities such as ID or seizures.
Examples of Autistic Behavioral Characteristics

Summary
- Impaired social skills
- Echolalia (Repeating everything said to them)
- Sensorimotor deficiencies
- Limited interactive language skills
- Seizure disorders
- ID
- Stereotypic behavior
- Self-injurious
- Problems with symbolic thinking

ORAL HEALTH MANAGEMENT AND DENTAL CARE

Oral Sensory Profile
Dental Visit Preparation
Desensitizing Office Visits

Oral Sensory Profile
- Diet - This can be a clue to under or over responsiveness
- Oral Aversions- kiss, wipe, utensils,
- Oral Seeking- mouthing, biting, grinding, pocketing
- Other sensory- smells, lights, noise, crowds.
- Vocalization
Communication Problems and Intellectual Assessment

- Talk with parent or caregiver to assess intellectual and functional abilities. In some cases there is no mental retardation or disability. When ID is present, most are in the mild to moderate range. Adjust level of communication to patient.
- Use tell-show-do approach to care. Use demonstrations.

Behavior Management

- Can include hyper activity and frustration
- Desensitizing appointment
- Make future appointment short and positive
- Attention to treatment setting, distractions like light and instruments should be out of sight
- Praise and reinforce good behavior
- Least restrictive approach, encourage bringing comfort items e.g. stuffed animal
- Immobilization is last resort!

Behavior

- Pharmacological options when other techniques fail. Caution, unpredictable reactions may occur to medications.
  - (40% antidepressants, 30% antipsychotic, 40% stimulants, 30% anticonvulsant and other classes, i.e. mood stabilizers)
- General Anesthesia may be needed of behavior is not managed.
- Be aware of tendency of perseveration which is the continuous, meaningless repetition of words, phrases, sounds or movements. Tell-Show-Do may trigger.
**Responses to Stimuli**

- May be acutely sensitive to changes in environment.
- Sensitive to light, color, sound, or touch
- Be consistent with staff, operatory and appointment time
- Minimize distractions.
- Allow time for patient to adjust and desensitize
- Talk to caregiver to get impression of patient’s level of tolerance of physical contact.

**Unpredictable Body Movement**

- Path from waiting room to operatory and chair are clear and limited distractions
- Observe movements and assess patterns
- Anticipate movements and adjust to patient.

**Seizures**

- Consult with physician. Gain information on frequency and severity and medications that are being administered.
- Adjust appointment time and check if meds have been taken.
- Be aware of events or stimuli that trigger patients seizures.
Oral Care for People with Autism Challenges and Strategies for Care
- Communication Problems and intellectual assessment
- Behavior Problems
- Response to stimuli
- Unusual and unpredictable body movement
- Seizures

Dental Caries
- Risk increases due to preference for soft, sticky or sweet foods. Particularly used as behavior control. Recommend gradual alternatives.
- Fluoride and sealants.
- Caution about meds that reduce saliva or contain sugar.
- Encourage independence in daily oral hygiene
- If this repetitive activity can be encouraged, this is a good thing

Oral Care Problems to be Considered
- Periodontal disease
- Damaging Oral Habits
- Trauma and Injury
Recommended reading and supplement

- Addressing Dental Care for Individuals with Autistic Disorders. Hosted by Dr. David Tesini and Dr. Clive Friedman. (Proceedings of the International Symposium, Alberta, Canada, August 2004).
- Dr. Andrew Zimmerman from the KK1, Center on Autism and Related Disorders.

The Role of Special Olympics Healthy Athletes

- Special Olympics Healthy Athletes
  - 1996 formal relationship to the SO
  - Includes: Hearing, Vision, Health Promotion, Fitness, Podiatry, and Dental
  - 100 screening events worldwide in 2001
  - Over 600 events planned in 2006
  - 140 Special Smiles Events

Special Olympics Special Smiles Objectives

- Direct dental services to athletes:
  - Oral screening, assessment, mouth guards, counseling, and referral.
- Oral health education to athlete, parent, coach, care giver.
- Opportunity to see significant numbers of persons with ID for the dental professional volunteer
- Change lives.