Evaluation and Management of Special Populations
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Objectives
• Recall the major characteristics of the syndromes presented
• Identify prevalent ocular conditions
  – refractive error
  – accommodative status
  – binocular status
• Recall the clinical pearls
• Increase comfort level in evaluating individuals with special needs

Prevalence
• >3,000,000 individuals have one or more handicapping conditions
• Legal blindness is 200 x’s more frequent in this population

Developmental Disabilities
• Severe, chronic, mental, or physical impairments
• Occur at conception or soon after birth
• Characteristics
  – Continue indefinitely
  – Varied functional abilities
  – Need for early identification / intervention

Overview
• Three global conditions
  – Cerebral Palsy
  – Down Syndrome
  – Autism

Cerebral Palsy
• Multiply handicapping condition from brain maldevelopment or damage before, during, or shortly after birth
• Characterized by motor dysfunction and associated problems
### Cerebral Palsy

- **General signs of CP include**
  - Locomotor difficulty (100%)
  - Speech disorders (90%)
  - Visual disorders (70+%)  
  - Mental retardation (50%)
  - Seizures (35%)
  - Hearing impaired (20%)
  - Swallowing and drooling (10%)

- **Classifications**
  - Spasticity (~80%)
    - Hallmark = hypertonicity
    - Subclassifications:
      - Hemiplegia - one side of body, common in full term children
      - Quadriplegia - both arms, both legs, head and trunk
      - Diplegia - trunk and all 4 extremities, mostly legs, common in preterm infants
      - Paraplegia - involves the legs
  - Athetosis (~12%)
    - Etiology - Rh incompatibility
    - Hallmark = arrhythmic involuntary movement
  - Rigidity (~7%)
    - Hallmark = slow movements with limited range
  - Ataxia (~5%)
    - Hallmark = hypotonicity
  - Tremor (0.3%)
    - Hallmark = repetitive, rhythmic involuntary movement
  - Mixed (12%)
    - One type usually predominates

- **Incidence**
  - 2:1,000

- **Etiology**
  - It is only rarely inherited (10%)
  - Prenatal (50%)
  - Perinatal (33%)
  - Postnatal (17%)

- **Visual Characteristics**
  - Strabismus - 60%, mainly esotropia
  - Refractive error - 60%, mainly hyperopia
  - Amblyopia - 20%
  - Gaze restrictions ~18%

- **Ocular Disease Characteristics**
  - Optic Atrophy 7%
  - Nystagmus 6%
Cerebral Palsy

- Examining Pearls
  - Leave in their wheelchair
  - Improper positioning may lead to inappropriate responses to testing
  - Examining Technique
    - Preferential looking
      - Strabismus - mainly neurological in nature
        - Discuss before surgical referral

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Down Syndrome

- The most common chromosomal abnormality encountered
- An extra chromosome 21 or an extra part of it in each cell body (Trisomy 21)

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Down Syndrome

- Facts
  - Detected with ultrasound / amniocentesis
  - Associated with congenital heart defects
  - Acute leukemia more common
  - Mental retardation is universal
  - Good socialization skills

- Incidence
  - Occurring in 1:600 to 800 live births
  - 1:50 with women older than 44

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Down Syndrome

- Etiology - Genetic
- Three modes of genetic transmission
  - Nondisjunction Trisomy 21 (most common)
    - extra 21 chromosome
  - Translocation
    - normal number of chromosomes but a portion of a chromosome (21 or 22) attaches to another (13, 14, or 15)
  - Mosaicism (1%)
    - some affected cells some normal cells

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Down Syndrome

- Common Identifying Features
  - Short stature
  - Brachycephalic skull
  - Flat occipital lobe
  - Low set ears
  - Flat nasal bridge
  - Small oral cavity and protruding tongue

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Down Syndrome

- Common Ocular Features
  - Epicanthal folds
  - Upward slanted eyes/Oblique palpebral fissures
  - Iris anomalies
    - Brushfield's spots - pale grey irregular discoloration midperipheral irides
    - Iris hypoplasia - ~95% (population 2-9%)
  - Retinal changes
    - Early bifurcation of vessels
    - RPE disturbances
Down Syndrome

- **Visual Characteristics**
  - Strabismus ~40%, mainly high AC/A esotropia
  - Refractive error - 42 - 73%,
    - mainly hyperopes but myopia is also common
  - Accommodative dysfunction
    - 92% had amplitudes <10 D and 50% were <4 D
  - Nystagmus ~8%

- **Ocular Disease Characteristics**
  - Blepharitis/conjunctivitis ~40%
  - Keratoconnus - 15%
  - Cataracts - expect early – 9yo
    - Due to premature-aging complex

- **Examining Pearls**
  - Use objective tests
    - Test accommodation with MEM

Behavioral Disabilities

- Lie along a continuum or spectrum depending on the severity of the presenting symptoms

ADD  ADHD/ LD  PDD-NOS  Asperger’s Autism

- Least severe
- More severe
- Most severe

- Currently there is controversy about including ADD and ADHD on the PDD spectrum

Autism

- Behavioral syndrome characterized by abnormalities in:
  - understanding and using language
  - responses to sensory stimulation
  - responses to change and insistence on routines
  - social interaction

- Lifelong developmental disability

**Incidence**
- CDC estimates 1% of children
- More frequent in boys
- Recurrence risk of ~9%

**Etiology**

**Diagnosis**
- DSM-IV
- Usually diagnosed around age three or earlier

**Characteristics**
- Atypical, delayed, or unequal development
- Atypical responses to sensory stimuli
  - Tactile, auditory, visual, and/or gustatory dysf.
- Disorders of communication / social interactions
- Significant deficits in speech / language development
Autism

• Visual Characteristics
  – Strabismus - 21-84%, IXT (variable)
  – Poor fixations and pursuits
  – Refractive error - no specific trends

• Visual Behaviors
  – Limited or no eye contact
  – Visual avoidance
  – Extreme lateral gaze
  – Eye pressing
  – Hand flicking
  – Light gazing

• Treatment
  – Highly structured special education
  – Strict regulation in diet
  – Medication

  • Behavioral therapy
    – Applied Behavior Analysis (ABA)
      – Developed by Lovas
      – A: antecedent [directive] B: behavior C: consequence
    – Relationship Development Intervention (RDI)
      – Developed by Gutstein
      – Parent centered through lifestyle changes
      – Lays missing pathways
      – Improves flexible thinking
      – www.autismspeaks.org

Examination Pearls

• Limit distractions
  – Remove clutter

• Stimulate large muscle groups
  – Oral stimulation

Put it all together

• What we can learn from each condition:
  – Positioning of the patient should be addressed to achieve optimal testing readiness
  – Modify language/communication
  – Modify exam room
  – Constellation of signs/symptoms
  – Know what ocular findings are specific to condition
    • In general expect/look for:
      – Higher refractive errors
      – Accommodative problems
      – Ocular health concerns
### Evaluation of Special Populations

- **People First Language**
  - Example:
    - “child with autism” vs “autistic child”
- **Communication**
  - Keep it short and simple
  - Visually support what you say
  - Match facial expressions to words
  - Give choices
  - Recognize that behavior is communication
  - Show respect

### Case History

- **Goals of examination**
  - IEP goals vision related
- **School setting**
- **Other professionals**
- **Medications**
  - Many have ocular/visual side effects

### Visual Acuity Assessment

- **Assess at the highest level possible**
  - Modifications may be necessary
- **Observations**
- **Objective**
  - Visually Evoked Potential/Response (VEP/R)
  - Can this patient see?
  - What is their visual potential?

### Visual Acuity

- **Formal**
  - Use testing method appropriate for developmental level
    - Teller Acuity cards
    - Keeler Acuity cards
    - Cardiff Cards
    - Lea Symbols
    - Feinbloom
    - VEP

### Visual Acuity

- **Lea Visual Acuity**
Visual Acuity

- Teller Acuity

Keeler Cards

- Test distance 38cm
- Blank side of card has open circle that can distract patient

Visual Acuity

- Cardiff Visual Acuity

Clinical Pearls

- Modifications to testing may be necessary
- Measure VA in gaze other than primary
- Match, point, write instead of speak
- No VA test used in isolation can accurately and completely assess visual functioning
  - OD should combine
    - Data from history & outside reports
    - Data from observations
    - Data from formal and informal acuity measures
- Remember resolution test overestimate VA
  - Report should reflect this information

Refractive Error Assessment

- Cycloplegic
  - Views may be brief
    - Patient may not allow lenses near their face
    - Work on estimation skills
  - Heart arrhythmia, abdominal distress, or hepatic problems may have adverse reactions
    - (0.2% or 0.5% cyclopentolate)
  - Down Syndrome
    - Ok to use cyclopentolate
    - BrJOphthalmol2008;92
- Trial frame refraction

Refractive Status
Binocular Vision Assessment

- Eye movements
- Accommodation assessment
- Binocular alignment
- Sensory fusion
- Motor fusion
- Nystagmus - observe the characteristics

Motor Alignment

- Cover test
- Hirschberg/Kappa
- Bruckner

Cover test

- Most precise method
- Neutralize with prism
  - May need to estimate
- Use an interesting, detailed target
  - Ask the child to identify the target
- Change frequently
  - Use the one toy - rule

Motor Alignment

- Hirschberg/Kappa
- Bruckner

Ocular Health Assessment

- Anterior/Posterior health
  - Be aware of Tactile defensiveness
- Visual fields
- Color vision
Ocular Health
• 20D Transilluminator

What do I need to get started?
• Positive and patient attitude
• Alternative visual acuity testing method
• Multiple fixation targets
  – Spinning lighted toy
• Snacks
• Blanket/pad for floor

Management
• Assessment
  – Uncertainty lies the quality and quantity of data
  – Increase both quality and quantity by
    • Using alternate tests
    • Modifying traditional testing methods
    • Prioritizing testing based on expectations
    • Making the patient comfortable with altered communication and positioning
    • Using information from care-givers, parents and other professionals to learn how to best relate to the patient
• Helpful hints
  – Be confident in your objective test findings
  – Be familiar with the associated ocular dysfunctions

Management
• Plan - consider all treatment options
  – To Rx or not to Rx
    • VA?
    • Emmetropization?
    • Spectacles and Contact lenses?
  – Vision therapy
    • Condition correctable?
    • Good candidate?

Management
• Plan
  – Environmental modifications
    • Lighting, posture, seating, etc.
• Follow-up
  • Allows for fine tuning of the plan
• Interdisciplinary referrals
  • Get the entire team involved
• Your report

Vision Report
• Include Information
  – Visual Acuity
  – Refractive status
  – Sensory status
  – Ocular health
• Recommendations
  • Classroom accommodations
  • Spectacle wear schedule
  • Patching recommendations
  – Define everything in layman’s terms
Vision Report

JB was born full term at a birth weight of 6 lb 8 oz. JB is diagnosed with Dandy-Walker Syndrome (congenital brain malformation involving the cerebellum and surrounding fluid spaces), cardiomegaly (enlarged heart), hydrocephalus (build up of fluid inside the skull leading to brain swelling) s/p 14 ventriculoperitoneal shunt revisions (shunt surgically placed in the skull to relieve pressure secondary to hydrocephalus), and seizure disorder. JB is currently taking the following systemic medications: Prevacid, Nortriptyline, Enalapril, Lasix, Periactin, and Regulin.

Vision Report

VISUAL ACUITY
JB was able to respond to the 20/63 Cardiff acuity cards with both eyes open. However, it should be noted that Cardiff acuity overestimates the visual acuity by approximately three times. She showed equal objection to occlusion, which may indicate relatively similar acuities in both eyes.

REFRACTIVE STATUS
Through cycloplegic retinoscopy (objective measurement with drops administered to stabilize focusing system), JB was found to have equal and mild hyperopic (far-sighted) refractive errors in both eyes.

STRABISMUS AND BINOCULAR VISION
JB displayed an intermittent left hypertropia (eye turn upwards) using Hirshberg/Kappa testing. This test is an estimation of eye alignment according to corneal reflexes.

ASSESSMENT
JB was found to have mild hyperopic refractive error (far-sightedness) that is normal for her age. She was also seen to display a intermittent left hypertropia (left eye turns upwards) with a slow-moving, large amplitude nystagmus (dancing eyes). JB compensates for this eye turn and nystagmus with a preferred head turn to the right, head tilt to the left shoulder, and chin pointed downwards. Bilateral anterior (front) and posterior (back) segment health was within normal limits.

INDIVIDUAL VISION PLAN (IVP)
• JB was not prescribed spectacles at this visit as her hyperopic refractive error is minimal and normal for her age.
• JB adopts a head turn and tilt to help her align her eyes and slow her nystagmus. This allows her increase the time that her eyes are still and improves her ability to see details. She should be allowed to adopt this head position as needed. When in the classroom setting she should be seated at the front of the room and to the left of center. This will allow her to see the teacher while she adopts her preferred head position.
• JB should continue care with her Pediatrician.
• No treatment of the eye turn is needed at this time.
• JB should continue to receive occupational therapy and physical therapy, with heavy emphasis on speech/language therapy to improve her communication skills. A one-on-one speech/language therapist is recommended.
• JB should return for a full eye and vision assessment with Dr. Heyman in one year.

References
• Practice parameter for the assessment and treatment of children and adolescents with autism spectrum disorder.