Progressive Eye Conditions

Objectives

• To provide cost effective, professional development to Florida’s teachers of the visually impaired and others working with students with visual impairments.
• To enhance the knowledge base of teachers of the visually impaired in topics unique to students with visual impairments.

Review from Sessions 1 and 2

• Session 1 – Review of Eye Conditions
  – Nystagmus
  – Myopia (High Myopia)
  – Albinism (Ocular and Oculocutaneous)
  – Optic Nerve Hypoplasia
  – Optic Nerve Atrophy
  – Cataracts
  – Cortical Visual Impairment

• Session 2 – Impact of Eye Conditions on Reading and Learning
  – Which conditions have peripheral field loss
  – Which conditions have central field loss
  – How peripheral and central loss impacts reading
  – Legal and ethical issues related to selecting learning media
  – When to consider braille instruction

Retinal Conditions and Syndromes

• Retinitis Pigmentosa
• Infantile Glaucoma
• Stargardt Disease
• Leber Congenital Amaurosis
• Usher Disease
• Retinopathy of Prematurity
• Cone-Rod / Rod-Cone Retinal Dystrophies

• Leber Optic Neuropathy
• Batten
• Marfan
• Retinoblastoma
• Retinoschisis – Juvenile
• Bardet-Biedl Syndrome
Ocular Conditions

- Aniridia – group of disorders involving underdevelopment of the iris
  - Corneal problems and glaucoma may develop later in life
- Corneal Dystrophies - loss of corneal transparency progresses with age. Acuity is reduced, sensitive to glare, amblyopia can develop.

Ocular and Other Conditions

- Microphthalmos – absent or very small eyes may have glaucoma associated which can cause progressive loss
- Coloboma – a notch-like defect in any part of the eye that is non-progressive. Associated with CHARGE syndrome. If severe, can result in retinal detachment or complications can develop.

Retinitis Pigmentosa

- Retinitis Pigmentosa (RP) – genetic conditions involving progressive night blindness and peripheral visual fields loss. In some cases progresses to loss of useable vision.
- Inherited types are -- dominant, recessive or sex-linked
  - Sex-linked starts at an earlier age and affects vision more severely

RP Effects on Vision

- Night blindness (loss of rods)
- Peripheral field loss
  - Early in upper field, progressing to ring shaped, may lead to tunnel vision or loss of all vision
  - Central vision loss affected by involvement of cones

RP progression varies according to the type

- Sex-Linked have night blindness in early childhood, extensive field loss by early teens, central vision loss in twenties. By 40’s, down to count fingers
- Recessive is variable but usually early onset and severe
Retinitis Pigmentosa

- RP Progression (cont)
  - Dominant form has some night blindness and field loss in childhood, but most patients retain reasonable visual acuity until 40 or 50 years of age or throughout life.
  - Most children maintain reasonable acuity, but fields are severely restricted

Infantile or Juvenile Glaucoma

- Infantile Glaucoma is a range of conditions involving pressure inside the eyes being too high
  - Corneal stretching, damage to the optic nerve, peripheral field loss
  - Early treatment and surgery is often needed to control pressure
  - If pressure is controlled, should not progress
  - Uncontrolled pressure leads to damage to optic nerve and visual field

Infantile or Juvenile Glaucoma

- Symptoms:
  - Cloudy, enlarged corneas
  - One eye larger than the other
  - Light sensitivity
  - Excessive tearing without discharge
  - Elevated eye pressure – pressure can vary from one person to another. Typical pressure is in the 12 to 22 mm HG.

Glaucoma

Conditions with Associated Glaucoma

- Sturge-Weber Syndrome
- Neurofibromatosis
- Stickler Syndrome
- Marfan Syndrome
- Trisomy 13
- Trisomy 21 (Down)
- Warburg Syndrome

- Aniridia
- Congenital Ocular Melanosis
- Peters Syndrome
- Iris Hypoplasia
- Microphthalmos
- Retinoblastoma

Stargardt Disease

Photos from National Eye Institute / flickr
### Stargardt Disease

- Stargardt is a macular dystrophy and is sometimes called juvenile macular degeneration. It involves gradual deterioration of acuity of ranges from 20/200 to count fingers.
  - Color vision becomes abnormal
  - Peripheral field remains intact


### Leber Congenital Amaurosis

- Leber Congenital Amaurosis (LCA) – genetic retinal dystrophy involving both rods and cones.
- Roving eye movements and nystagmus are common.
- Present from birth or first few months
- Eye poking common


### Usher Syndrome

- Ushers Syndrome – RP type eye condition that has associated hearing loss.
- Type 1 – Profoundly deaf at birth, decreased night vision before age 10
- Type 2 – Moderate to severe hearing loss from birth, decreases night vision in late childhood or teens

Usher Syndrome

- Type 1 – Profoundly deaf at birth, decreased night vision before age 10, progresses rapidly until blind.
- Type 2 – Moderate to severe hearing loss from birth, decreases night vision in late childhood or teens. Does not result in blindness
- Type 3 – Normal hearing and vision at birth, progressive loss beginning in childhood or early teens. By mid-adulthood blind.

Retinopathy of Prematurity

- Retinopathy of Prematurity – scarring disease of the retina developing in premature and low birth-weight infants.
  - Immature blood vessels in the retina cause lack of oxygen or bleeding which leads to scarring

Retinopathy of Prematurity (cont)

- Stages:
  - Stage 1: Mild abnormal blood vessel growth. Many develop normal vision.
  - Stage 2: Moderate abnormal blood vessel growth. Many develop normal vision.
  - Stage 3: Severe abnormal blood vessel growth. Some develop normal vision. Stage 3 Plus disease means the retinas have become enlarged and twisted. Treatment can prevent retinal detachment.
  - Stage 4: Partially detached retina
  - Stage 5: Completely detached retina. Untreated will result in severe visual impairment or blindness.

Retinopathy of Prematurity

- Treatments – Laser, cryotherapy, and drugs to stop the growth of the blood vessels
  - Laser burns away periphery of the retina
  - Cryotherapy freezes edges of the retina
  - Both result in loss of some peripheral vision
  - Drugs showing promise with less peripheral loss.


National Eye Institute You Tube Video on ROP

Retinopathy of Prematurity

• Prognosis:
  – Most infants with severe vision loss have other problems related to early birth
  – About 1 in 10 develop severe retinal diseases
  – 2% of children with high-risk ROP developed glaucoma during the first 6 years of life
  – Complications (rare) include high myopia, cataracts, vitreous and retinal hemorrhage

Rod-Cone or Cone-Rod Dystrophies

Cone-Rod Dystrophies

• Wide range of eye conditions that affect the rods and cones of the eyes.
• Names of more common types:
  – Leber Amaurosis
  – Retinitis Pigmentosa
  – Usher Syndrome
  – Batten

Cone–Rod Dystrophy

• Cone-Rod Dystrophy (CRD) has early loss of color vision, central vision, and acuity followed by night blindness and loss of peripheral fields.
• Cone-Rod is generally more severe and rapid than that of Rod-Cone Dystrophies (RCDs) such as RP in leading to earlier legal blindness.
• At end stage there is not much difference between the vision of CRDs vs RCDs. Cone Rod Dystrophies are ten times less frequent than RP.

Leber Optic Neuropathy

• Leber Hereditary Optic Neuropathy
  – Usually males in teens or twenties
  – Vision problem begins in one eye and other eye is affected within several weeks or months
  – Vision loss in central field and color vision
  – Acuities are typically 20/200 or worse

Batten Disease

• Batten Disease – Inherited disease of the nervous system
  – Typically begins in childhood (5 to 10 years old)
  – Develops vision problems and/or seizures
  – Progressive mental decline, worsening seizures, loss of vision and motor skills

Cone-Rod Dystrophies: [http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1808442/](http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1808442/)


Cone –Rod Dystrophy: [http://www.viscotland.org.uk/eyeconds/rodconedystrophy.html](http://www.viscotland.org.uk/eyeconds/rodconedystrophy.html)


Batten Disease: [http://www.btsra.org/what-is-batten-disease/about-batten-disease](http://www.btsra.org/what-is-batten-disease/about-batten-disease)
Batten Disease

- Vision loss is often an early sign
- Four main types
  - Infantile (most severe)
  - Late Infantile
  - Juvenile
  - Adult
- Typically fatal by late teens or early 20s

Marfan Syndrome

- Marfan syndrome affects connective tissue. The severity of the effects of Marfan syndrome varies greatly even within the same family.
  - Heart and blood vessel problems
  - Skeletal abnormalities
  - Lung problems
  - Eye problems

Marfan Syndrome

- Eye Problems
  - Lens is often off-center
  - Increased risk for detachment of the retina
  - Cataracts
  - Glaucoma

Retinoblastoma

- Retinoblastoma – cancer of the eye that causes tumors of retinal cells and usually develops before the age of 4.
  - Can affect one eye or both
  - Location of the tumor and type of treatment determines affect on vision
  - New retinal tumors or recurrences are most likely to occur in first year after diagnosis

Retinoblastoma (cont.)

- Hereditary form of retinoblastoma is at risk for developing pineal tumors in the brain. This usually occurs more than 20 months after diagnosis.
- Increased risk of developing other types of cancer – bone or soft tissue sarcoma or melanoma in later years.
- Glaucoma is often associated


Disorders of vision in children: a guide for teachers and carers; Bowman, Bowman and Dutton, 2001, [www.ssc.education.ed.ac.uk/resources](http://www.ssc.education.ed.ac.uk/resources)

THE FOLLOWING LISTS ARE TO HELP GUIDE ELIGIBILITY DECISIONS AND SHOULD NOT BE CONSIDERED THE FINAL AUTHORITY. THE ELIGIBILITY TEAM IS RESPONSIBLE FOR ELIGIBILITY DECISIONS THEN THE IEP TEAM DECIDES SERVICES.
For Eligibility

These conditions should be considered “progressive” for eligibility purposes:

- Retinitis Pigmentosa
- Usher Syndrome
- Stargardt’s Disease
- Progressive Rod / Cone Dystrophies including Achromatopsia (not congenital)
- Leber’s Optic Neuropathy
- Marfan’s Syndrome
- Unstable Glaucoma
- Newly diagnosed Retinoblastoma

- Choroideremia (males)
- Juvenile Retinoschisis
- Best’s Disease (Macular Dystrophy)
- Batten
- Stickler Syndrome (due to likely retinal detachment)
- Cataract if not able to be removed
- Several Syndromes and other conditions

For Eligibility

These conditions have the possibility of being considered “progressive”

- Glaucoma
- Retinopathy of Prematurity (with unstable retina)
- Leber’s Congential Amaurosis
- Bardet-Biedl Syndrome
- Several other syndromes

For Eligibility

These conditions should be considered “progressive” for eligibility purposes:

- Choroideremia (males)
- Juvenile Retinoschisis
- Best’s Disease (Macular Dystrophy)
- Batten
- Stickler Syndrome (due to likely retinal detachment)
- Cataract if not able to be removed
- Several Syndromes and other conditions

Remember

- Each child is different and eye conditions can change in an instant
- Diagnosis of one condition does not preclude another condition from developing
- Do your own research from reputable sources.

Coping with Loss of Vision

- Common reactions
  - Shock and denial
  - Anger and questioning
  - Helplessness, fear, anxiety
  - Sadness and grief
  - Depression
  - Acceptance

Helping Students Cope

- Listen and be supportive
- Help make connections with others facing the same problems
- Get them the professional help they need – counseling, transition, low vision devices, O&M, etc.
- Involve parents, family members, and friends. They are also affected.

Royal National Institute of Blind
http://www.rnib.org.uk/livingwithblindness/copingwithblindness/emotionalupport/Pages/common_feelings.aspx#H2Heading4
Helping Students Cope

- Provide both practical and emotional support
  - Low vision devices
  - O&M
  - Braille and technology
  - Encouragement
- Find positive role models or mentors
- Promote independence and interaction

LEGAL AND ETHICAL CONSIDERATIONS

IDEA Rules

- In the case of a student who is blind or visually impaired, provide for instruction in braille and the use of braille unless the IEP team determines, after an evaluation of the student’s reading and writing skills, needs, including future needs, and appropriate reading and writing media, that instruction in braille or the use of braille is not appropriate for the student.

For a student staffed into a program for the visually impaired

- Braille is the default learning media. We must demonstrate and document that braille is not needed. This includes a reasonable expectation braille will not be needed in the future.

United States Department of Education
Office of Special Education and Rehabilitation (OSERs)

- Memo for ESE Directors on the importance of Braille Instruction, June 19, 2013
- Key Points:
  - Braille not being taught appropriately to students
  - Braille can be taught in combination with other methods
  - Must provide for current and future needs of students
  - Data-based decision making for learning media (FVLMA)
  - Systematic and regular instruction by trained personnel

Florida Department of Education
State Board Rules

- 6A-6.03014 Exceptional Student Education Eligibility Students Who Are Visually Impaired
- 6A-6.03022: Special Program for Students Who Are Dual-Sensory Impaired
State Board Rule

(b) If a medical criterion listed in SB 6A-6.0314 is met, then a comprehensive assessment of skills known to be impacted by a visual impairment, shall include, but is not limited to: functional vision evaluation, learning media assessment, and if appropriate, orientation and mobility.

Factors to Consider in Media Selection

- Efficiency
  - Time and effort
  - Print size
- Prognosis
  - Degenerative conditions
- Visual Fatigue

AER Position Paper

- Literacy Media Decisions for Students with Visual Impairments: September 2013
- http://aerbvi.org/downloads/5/0/AER_Fall_2013_5_FINAL.pdf

AER Position Paper Key Points

- Instruction in braille unless after an evaluation determines braille is not needed
- Instruction in print with or without optical devices after a clinical low vision evaluation
- Instruction in both braille and print (dual media) with or without prescribed optical devices

AER Position Paper

- Literacy media selection is based on a variety of assessments conducted by qualified professionals.
- IEP Team decides the amount of time to devote to instruction based on individual needs of the student
- Difficulties in literacy should be addressed as early as possible to narrow the achievement gap.

Perkins Path to Literacy: http://www.pathstoliteracy.org/dual-media
Using both print and braille????

MORE TOOLS IN THEIR “TOOLBOX”

Dual Media Students

- Braille for long reading assignments
- Enlarged print for math and science (usually with low vision devices)
- Braille (six-key entry) and technology for writing
- Introduction of braille for possible loss of vision later in life

Dual Media Instruction

- Research says that is best to teach braille reading early, even simultaneously with print.
- Print is everywhere and they learn print with exposure by parents and classroom teachers
- Braille must be specifically taught and included in the home and school environment
- Braille must be taught by qualified and knowledgeable TVIs

Summary

- Learning Media is based on assessed needs including future needs
- Every child is different
- The more “tools” in the “tool box” the better
- Braille instruction should be started as soon as possible … In Common Core Standards PreK students are expected to know the braille alphabet and punctuation

Resources

- AFB Family Connect
- National Association for Parents of Children with Visual Impairments (NAPVI)
- Disability specific support groups

Eye … Eye… Eye

POST-TEST
Name the Parts of the Eye

1. Clear covering on the front
2. White part of the eye
3. Works like a round muscle controlling the light that enters the eye
4. Black center of the eye
5. Directs the light to the retina

Name the Parts of the Eye (cont.)

6. Light sensitive layer of nerve cells
7. Area that allows for sharp, clear near vision
8. What takes the nerve impulses to the brain
9. Where “vision” happens
10. What part controls color vision?

Fill in the blank

11. A person with 20/400 vision can see at ______ feet what a person with normal or 20/20 vision can see at ______ feet.
12. A person is considered legally blind at ______ acuity.
13. Three progressive eye diseases that can result in blindness are ______, ______, and ______.

Name that Condition

14. Progressive, tunnel vision, night blindness, hereditary, usually results in blindness
15. Decreased acuity, severe myopia, possible retinal detachment, field loss, possible glaucoma, abnormal retinal blood vessel development leading to bleeding, scarring in premature infants.

Name that Condition

16. Poor visual acuity, near vision better than distance, nystagmus, photophobia, cones are absent, colors seen as shades of gray.
17. Dysfunction of optic nerve interrupting nerve impulses to brain, pale optic disc, caused by disease, pressure on optic nerve, trauma, glaucoma, toxicity, heredity, fluctuating vision.

Name that Condition

18. Refractive error where image is formed in front of retina, eyeball is elongated, can be severe and degenerative resulting in blindness.
19. Increased pressure in the eye because of blockage of fluid in the aqueous humor. Affects near and distant vision, photophobia, can be degenerative and result in blindness.
20. Decreased visual acuity, photophobia, high refractive error, nystagmus, lack of pigment causing abnormal retina and optic nerve development.

21. Decreased acuity, photophobia, nystagmus, birth defect which can cause a cleft in the pupil, iris, lens, retina, or optic nerve, hereditary.

22. Damage to the visual cortex or the visual pathways, fluctuating vision, eye intact, no nystagmus, color vision intact, often associated with neurological disorders.

23. __________ albinism affects the entire body, while __________ albinism only affects the eyes.

24. On an eye medical report OU means ________, OD means ________ and OS means ________.

25. ________ is the term typically used when referring to misalignment of the eyes.

**Fill in the Blank**

**Name that Condition**

**Follow-up Project**

- If you have a student with a progressive eye disease, research that disease. If you don't currently have a student that is diagnosed with a progressive eye disease, select one of the topics in this presentation.

**Follow-Up Project**

- Write a summary of the condition with the following:
  1. Hereditary factors
  2. Expected progression
  3. Current functional vision
  4. Plan for the student
     - Learning media selection
     - Psychological support
     - Post-high school support
Follow-up Project

• Written and sent electronically by email with subject line: LastName Webinar Homework (Ratzlaff Webinar Homework)
• Change the student’s name to your name to protect student confidentiality
• Submit by May 1, 2014

More Information

• Articles at TSBVI:
  – Syndromes Associated with Progressive or Degenerative Vision or Hearing Loss: http://www.tsbvi.edu/seehear/spring03/syndromes.htm

Resources

• Foundation Fighting Blindness: http://www.ffb.ca/eye_conditions/RD_diseases.html
• Retina International: http://www.retina-international.org/eye-conditions/retinal-degenerative-conditions/
• Lighthouse International: http://www.lighthouse.org/about-low-vision-blindness/childrens-vision/pediatric-eye-disorders/
• Batten Disease Support and Research Association: http://www.bdsra.org/what-is-batten-disease/about-batten-disease/

Resources

• Glaucoma Research Foundation: http://www.glaucoma.org/glaucoma/childhood-glaucoma-1.php

Resources

• Mayo Clinic - ROP treatment, what’s the latest approach? http://www.mayoclinic.com/health/rop-treatment/AN02150
• Journal of Medical Genetics: Inherited mitochondrial optic neuropathies: http://jmg.bmj.com/content/46/3/145.full

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