Hereditary Childhood Vision Impairment
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I. Epidemiology of Childhood Vision Impairment
   a. World-wide prevalence
   b. U.S. prevalence
   c. Causes of childhood vision impairment
      i. Hereditary
      ii. Traumatic
      iii. Perinatal
      iv. Unknown

II. Review of Inheritance Patterns
    a. Autosomal dominant
    b. Autosomal recessive
    c. X-linked recessive
    d. Mitochondrial
    e. Sporadic

III. Testing Useful for Differential Diagnosis
    a. ERG
    b. VEP
    c. Fundus photography

IV. Evaluation
    a. History
    b. External Observation
    c. Visual Acuity
       i. Grating
       ii. Optotype
       iii. Fixation and following
       iv. Observation of visual behavior
    d. Nystagmus
       i. Type (pendular, jerk)
       ii. Amplitude, frequency
       iii. Null point
    e. Refractive error
       i. With-the-rule astigmatism common in nystagmus
       ii. 80% of kids with VI have refractive error > ±1D
    f. Accommodation
    g. Ocular Health Assessment
V. Albinism
   a. Characterized by hypopigmentation, iris transillumination, foveal hypoplasia, abnormal decussation of fibers at chiasm
   b. Oculocutaneous (usually AR)
   c. Ocular (usually XLR)
   d. May be associated with systemic disease
      i. Hermansky-Pudlak
      ii. Chediak-Higashi
   e. Acuity variable (20/25 to 20/400) and dependent on amount of pigmentation, degree of foveal hypoplasia and amplitude/frequency of nystagmus
   f. Nystagmus – variable waveforms, but usually pendular
   g. Strabismus common
   h. Refractive error – runs the gamut, but often hyperopia and with-the-rule astigmatism

VI. Optic Nerve Hypoplasia
   a. Typically idiopathic (new research does NOT support previous reports of association with maternal drug or alcohol use
   b. Visual acuity highly variable (NLP to 20/20) and not linearly correlated to size of optic nerve head
   c. Unilateral or bilateral
   d. Nystagmus present when bilateral
   e. Refractive error variable
   f. Look for double ring sign and disc-macula/disc diameter ratio

VII. Hereditary Optic Atrophy
   a. AD (most common) or AR
   b. Acuity typically 20/80 to 20/200
   c. Temporal pallor
   d. May have centrocecal scotoma
   e. Mild color vision deficits, mild contrast deficits
   f. Not associated with nystagmus (onset typically before school entry, but after age 2)
   g. Always rule out other causes of optic atrophy
      i. Trauma
      ii. Hydrocephalus
      iii. Anoxia
      iv. Compression

VIII. Leber Hereditary Optic Neuropathy
   a. Inherited via mitochondrial DNA (common mutations: 11778, 14484)
   b. Most common onset 2nd to 4th decade
   c. Onset is acute, usually unilateral at first, becoming bilateral within weeks
   d. Color vision affected severely and early
IX. Leber Congenital Amaurosis
   a. AR (Gene therapy only for those with RPE65 mutation)
   b. Congenital retinal dystrophy
   c. Decreased vision in first year of life
   d. Nystagmus (often roving)
   e. Disc pallor, attenuation of vessels. RP like fundus pigmentation
   f. Refractive error commonly high hyperopia
   g. Extinguished scotopic and photopic ERG

X. Stargardt Macular Degeneration
   a. AR inheritance (ABCA4 gene)
   b. Less common AD inheritance (ELOVL4 gene)
   c. Onset between 1rst and 4th decade
   d. Children often suspected of malingering in early disease
   e. Lipofuscin deposited below retina
   f. Yellow flecks sometimes visible in posterior pole
   g. Dark choroid on fluorescein angiography
   h. Acuity at endstage is almost always 20/200 – 20/400

XI. Retinitis Pigmentosa
   a. Broad classification for a family of hereditary eye disease with the
      common feature of panretinal photoreceptor apoptosis
   b. AR, AD, XLR, Digenic, Mitochondrial
   c. Cone-Rod
   d. Rod-Cone
   e. May be syndromic (Bardet-Biedel, Refsum Disease)

XII. Progressive Cone Dystrophy
   a. Inherited most commonly as AD, but may be AR or XLR
   b. Progressive loss of cone function
   c. Bull’s eye maculopathy due to central area of uninvolved RPE
   d. Rods may be mildly involved in some pedigrees

XIII. Achromatopsia (aka Rod Monochromatism or stationary cone dystrophy)
   a. Complete – AR
   b. Incomplete (blue cone monochromat) – XLR
   c. Profound photophobia
   d. Nystagmus (often pendular)
   e. Benefit from spectacle or CL tints, especially red
   f. Visual acuity typically 20/200 if complete, and 20/60 -20/200 for
      incomplete forms
   g. Fundus appears normal
XIV. X-linked Juvenile Retinoschisis
   a. Near 100% have macular involvement
   b. Schisis of retina at NFL with vitreous liquefaction
   c. May have extensive vitreous opacities
   d. Macular changes may improve with topical dorzolamide
   e. VA variable

XV. Helpful websites
   b. Genetests.org
   c. www.blindness.org (Foundation Fighting Blindness)
The Low Vision Evaluation
10:20 - 12:00
November 11, 2014
Course objectives:

The Low Vision Evaluation

Course objectives

1. Be able to discuss the current low vision assessment tools in the evaluation of the low vision patient.
2. Be able to assess functional vision including visual acuity, visual field, contrast sensitivity, color vision, or other important parameters with the accepted low vision testing tools.
3. Be able to integrate the latest in health compliance, EMR, and time management into a sustainable low vision health care model.

Course outline

I. History (including Medicare compliance with meaningful use)
   A. The chief complaints
   B. The ocular history
      1. Diagnoses
      2. Medications
      3. Surgeries
      4. Glasses, Vision Assistive Equipment
      5. Filters, Lighting and Glare Control
      6. Other adaptive equipment
   C. The medical history
      1. Co-morbidities
      2. Medications
      3. Surgeries
      4. Meaningful use elements
      5. Patients affect (e.g. appearance, attitude, behavior).
II. Visual Acuities
   A. Understanding the essentials of low vision notations from the M to the N to LogMAR
      Measurement of visual acuity – the principles (Johnston, Riggs, Snellen, Bailey and
      Lovie)
   B. Standard distance and near low vision charts

III. Visual Skills: Pupils, motilities, saccades, binocularity

IV. Refraction

V. Visual Fields
   A. Confrontation
   B. The Amsler grid
   C. Automated perimetry in a low vision world
   D. Goldmann essentials
   E. Microperimetry and other useful tools (Fletcher Skills Chart - printed scotoma test)

VI. Contrast Sensitivity
   A. Concepts in measurement
   B. Current charts

VII. Glare Sensitivity

VIII. Reading assessment tools including the MN read and Pepper test

IX. Predicting the Add (Equivalent Power Considerations)

X. Prescribing
   A. Vision Assistive Equipment
   B. Lighting and Glare Control
   C. Visual Skills Training
   D. Referral for other Services
      1. ADL
      2. O&M
   E. Other Considerations in Prescribing
      1. The "Psychological" State of the Patient
      2. The "Physical" State of the Patient
      3. Cosmesis
      4. Cost

XI. Discussion with Patient – patient and doctor realistic expectations

XII. Examination strategies for complex situations
   A. For the unresponsive patient
   B. Language problems
   C. Profound hearing impairment
Optics Review for AAO LV Diplomate Exam

Course objectives

1. To review the categories and optical characteristics of the various low vision devices.
2. To understand the optical principles of these systems, with representative optics problems.
3. To have the background to be able to solve optics problems relating to these devices.
4. To review lighting principles used in conjunctions with low vision devices.

Course outline

I. Introduction – Categories and Characteristics of Low Vision Aids
   A. Categories of Optical Aids
      1. Spectacles
      2. Hand-held Magnifiers
      3. Stand Magnifiers
      4. Electro-optical Systems (including the Closed-Circuit Television)
      5. Telescopes
   B. Characteristics of Optical Aids
      1. Power
      2. Working Distance (Eye to Object Distance)
      3. Working Space (Lens to Object Distance)
      4. Field of View
      5. Light Gathering/Reduction
II. Optics of Near Low Vision Devices
   A. Object-Image Relationships
   B. Power Considerations –
      1. Equivalent Power of Near Optical System
         a. Case 1: Object at focal point of low vision device
            - Low vision device at spectacle plane
            - Low vision device away from spectacle plane
         b. Case 2: Object inside focal point of low vision device
            - Low vision device at spectacle plane
            - Low vision device away from spectacle plane
      2. Equivalent Power of a CCTV
      3. Equivalent Viewing Distance
   C. Field of View
   D. Magnification
      1. Traditional: Relative Size, Relative Distance, Angular
      2. Other notations: Effective (rated); Conventional; Angular (perceived/apparent)
   E. Verifying the Power
      1. Of a lens (spectacle or hand-held magnifier)
      2. Of a stand magnifier
      3. Of a telescope
   F. Labeling and ANSI Standards

III. Optics of Telescopes
   A. Distance Telescopes
      1. Galilean and Keplerian Telescopes
      2. Magnification \((F_e/F_o)\)
      3. Rx inclusion
   B. Near (and intermediate) Telescopes
      1. Accommodation/vergence amplification
      2. Compensation for vergence amplification
      3. Equivalent power of focal telescopes
      4. Equivalent viewing distance
   C. Verification of Telescopic Magnification
   D. Field of View through a Telescope
   E. Image Brightness
   F. Special Telescopic Systems
      1. Contact Lens Telescopes
      2. Telescopic Correction for Aphakia
      3. Implantable Miniature Telescope
      4. Reverse Telescopes and Field Expanders
Title
Low Vision Rehabilitation Grand Rounds

Description
Teach the advanced fundamentals of low vision rehabilitation practice by the examples of a grand rounds format.

Learning Objectives
Upon completion of this session, participants will be able to:
1. Cite the characteristics of the ocular conditions exhibited in the patients histories and relate the diagnosis to expected vision rehabilitation challenges.
2. Describe the principles of magnification that were used to assist the patients in the case histories presented.
3. List three important questions that can be asked during a case history that can give insight into the patient's challenges living independently with visual impairment.
4. Name three types of professionals other than optometrists who can assist the visually impaired and describe the role they played in the management of patients in the case histories presented.
5. Describe effective strategies for training a patient in the use of a low vision device.

Outline
1. Introduction
   a. Dr. Rosenthal's training, practice situation and general approach
   b. Dr. Squier's training, practice situation and general approach
   c. Format of the Grand Rounds course
2. Case 1 – Dr. Rosenthal
   a. Condition, age, gender of patient
   b. Challenges the patient was facing
   c. Clinical findings
   d. Rehabilitation strategies
   e. Outcomes
   f. Lessons learned, principles employed
   g. Dr. Squier's comments on the case
   h. Questions
3. Case 2 – Dr. Squier
   a. Condition, age, gender of patient
   b. Challenges the patient was facing
   c. Clinical findings
   d. Rehabilitation strategies
   e. Outcomes
   f. Lessons learned, principles employed
g. Dr. Rosenthal's comments on the case
h. Questions
4. Case 3 – Dr. Rosenthal
   a. Condition, age, gender of patient
   b. Challenges the patient was facing
   c. Clinical findings
   d. Rehabilitation strategies
   e. Outcomes
   f. Lessons learned, principles employed
   g. Dr. Squier's comments on the case
   h. Questions
5. Case 4 – Dr. Squier
   a. Condition, age, gender of patient
   b. Challenges the patient was facing
   c. Clinical findings
   d. Rehabilitation strategies
   e. Outcomes
   f. Lessons learned, principles employed
   g. Dr. Rosenthal's comments on the case
   h. Questions
6. Case 5 – Dr. Rosenthal
   a. Condition, age, gender of patient
   b. Challenges the patient was facing
   c. Clinical findings
   d. Rehabilitation strategies
   e. Outcomes
   f. Lessons learned, principles employed
   g. Dr. Squier's comments on the case
   h. Questions
7. Case 6 – Dr. Squier
   a. Condition, age, gender of patient
   b. Challenges the patient was facing
   c. Clinical findings
   d. Rehabilitation strategies
   e. Outcomes
   f. Lessons learned, principles employed
   g. Dr. Rosenthal's comments on the case
   h. Questions
8. Summary of cases