**2A Nurse Caring Family Concepts**

**Pediatric Chronic Illness, Sensory and Cognitive Impairment**

Week 17: May 12, 2003

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**Chronic Condition Definitions**

- Chronic condition:
  - Exists for a minimum of three months
  - Physical, psychological or cognitive condition that places limitations on day to day functioning or requires reliance on special treatments

- Disability: functional limitation that prevents or interferes with ability to perform age-expected activities

- Handicap: barrier imposed by society, environment, or one’s self in response to perceived differences

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**Dimensions of Childhood Chronic Conditions**

- Children with serious illnesses live longer
  - 18% in US have a chronic health condition
  - 6.5% have some disability

- Important Considerations:
  - Nature of onset
  - Progression of condition
  - Effects on appearance
  - Effects on daily function
  - Effects on behavior
  - Amount of care required
Impact of Chronic Condition on Family

- Child: condition does not define child; ask them what is impact of condition
- Siblings may have greater responsibility; worry, jealousy, negative emotions, restricted family events & increased family closeness
- Parents: emotional, physical & financial strain
  - Accept & manage condition
  - Meet developmental needs of child & family
  - Cope with stress & crises
  - Assist family to manage feelings
  - Educate others about condition
  - Establish support system

Family Members’ Feelings

- Shock & Denial
- Adjustment
  - Guilt
  - Anger
  - Overprotection
  - Rejection
  - Denial
  - Gradual acceptance
- Reintegration & Acknowledgement
  - Increased comfort
  - Social reintegration
  - Chronic sorrow

Parental Stress Points

- Diagnosis
- Developmental milestones
- Start of schooling
- Ultimate attainment
- Adolescence
- Future placement
- Death of child
Impact on the Child

- Infant: trust development dependent on attachment
- Toddler:
  - Encourage independence & maximum mobility
  - Resilient; but will manage stress better with strong parental bond & parental proximity
- Preschooler: may feel guilt that they caused illness
- School-age: peers influence self-esteem & socialization very important
- Adolescent: harder if illness occurs at this age; may interfere with development of identity

Normalization

- Preparation in advance when possible
- Participation in as many decisions as possible
- Sharing care regimen allow with family members & peers whenever possible
- Control: identify areas where child can be in control
- Expectations are same for child as well siblings
- Positive attitude: focus on areas of ability & competence to increase self-esteem

Supporting Family Coping

- Parent:
  - RN can model liking & acceptance of child
  - Affirm & build on strengths
  - Support groups: other parents with similar experiences are helpful
- Child:
  - Observe reaction to condition, functional ability & adaptive behaviors
  - Determine child’s understanding of illness; allow to ‘play’ out feelings
  - Provide support while child learns to cope
Support Family Coping

• Siblings:
  – Explain illness to siblings & prepare for physical changes
  – RN uses anticipatory guidance with parents to address negative feelings in siblings
  – Praise and show gratitude to siblings for taking on additional responsibilities

• Extended family/community:
  – Include grandparents in open discussion
  – Prepare to cope with comments from others

Supporting the Dying Child

• Discuss truth with children; they need honest, accurate information about their illness
• Help family determine whether to remain in the hospital or at home
  – Children’s hospice care is alternative to hospital care; holistic approach provides physical, respite & bereavement care
• Pain med routinely; rescue or bolus doses for breakthrough pain; opioids for severe pain
• Limit care to essentials

Hospital Deaths

• Prepare family by providing information
• Be available; consistent nursing staff if possible
• Ensure parents have full access to child at all times
• Provide quiet, respectful environment
• Make sure family’s needs are met
• Remove tubes/equipment from body after death
• Allow family unlimited time with child
• Contact organ/tissue donation organization
• Support grieving family; help siblings cope
• Acknowledge own grief reactions; attend funeral
Mental Retardation

- Defined as:
  - Significantly sub-average intellectual functioning: IQ < 70
  - Age at time of diagnosis: < 18 years
  - Functional strengths & weak limited adaptive skills in at least 2 or more areas of functioning:
    communication, self-care, home living, social skills, leisure, health/safety, self-direction,
    academics, community use, work

Mental Retardation Etiology

- Genetic
  - Down syndrome, Fragile X Syndrome
- Biochemical
  - Maternal alcohol/drug ingestion; overdose
  - Poisoning
  - Traumatic cranial injury
  - Drowning, aspiration
- Infectious
  - Maternal rubella or cytomegalovirus
  - Meningitis, encephalitis

Care of Cognitively Impaired Child

- Educate child/family & start early
- Self-care skills: many aids allow independence
- Promote optimum development:
  - Play/exercise: need
  - Communication
  - Discipline
  - Socialization
  - Sexuality
- Help family adjust to future care
- Care during hospitalization
Down Syndrome (DS)

• Etiology:
  – Extra chromosome 21 (Trisomy 21)
  – Greater risk in women >35
  – 4% have translocation
  – 1% have mosaicism
• Clinical manifestations include: epicanthic folds, square hands with a simian palmar crease, hyptonic muscles, protruding tongue, small nose, depressed nasal bridge, short neck

Down Syndrome Problems

• Congenital heart defects common
• Respiratory tract infections & chronic otitis media associated with decreased muscle tone, immune system dysfunction & anatomical differences in ears (narrow, short eustachian tube)
• Hypotonic; difficult to position with limp, flaccid extremities
• Feeding difficulties due to mucus & protruding tongue
• Constipation (Hirschsprung’s megacolon)
• Hypothyroidism, cataracts, leukemia & Alzheimer’s disease (after age 30)

Down Syndrome Nursing Interventions

• Life span of Down syndrome children has increased
• Screen for cardiac defects, hypothyroidism & anomalies
• Keep secretions liquefied; good pulmonary hygiene
• Teach why child less cuddly & how to swaddle
• Special eating utensils
• Fiber, fluids; stool softeners
Fragile X Syndrome

- Most common inherited cause of MR;
- Fragile site at tip of long arm of X chromosome
- X-linked; all affected males have sx disorder (no male carriers). Both affected sexes fertile
- Most males & 30% of females are mentally retarded (females affected to lesser degree)
- Prenatal diagnosis with direct DNA testing possible; but rare to identify during first year of life
- Care as for child with Down syndrome

Fragile X Clinical Manifestations in Males

- Intellectual disability (range from subtle learning disabilities to severe mental retardation)
- Physical features:
  - Long face, prominent jaw, large protruding ears, enlarged testicles, flat feet, double-jointed fingers
- Behavioral characteristics may include
  - Attention deficit disorders, speech disturbances, hand biting, hand flapping, autistic behaviors, poor eye contact, & aversion to touch and noise

Fragile X Clinical Manifestations in Females

- Same characteristics seen in males may also be present in some females
  - Smaller percentage of females are affected
  - Degree of impact usually diminished because females XX; males XY
  - 30% with significant intellectual disability; 70% have more moderate learning difficulties
- Physical & behavioral features
  - appear in smaller percentage of females & to a lesser degree.
### Hearing Impairment

- **Hard-of-hearing:** hearing sufficient to process linguistic information through hearing
- **Deaf:** unable to process linguistic information through hearing, with or without amplification
- **Incidence:** 3:1000 annually born with hearing impairment in US; 4000 are profoundly deaf
- **Etiology:** congenital or acquired factors: ototoxic drugs, malformation, low birth weight, perinatal infection or asphyxia, infections, environmental noise (especially neonates & adolescents)

### Hearing Impairment Pathology

Hearing disorders classified by location of defect:

- **Conductive:** temporary or permanent; from condition affecting sound transmission across middle ear; affects loudness of sound. Most common type; often result of chronic otitis media
- **Sensorineural:** usually permanent; from damage to inner ear &/or auditory nerve; causes distortion of sound & discrimination problems. Result of congenital defects or acquired conditions like kernicterus, infection, ototoxic drugs.

### Hearing Impairment Pathology

- **Mixed conductive-sensorineural:** from interference with middle ear sound transmission & along neural pathways; problems with sound distortion & hearing lower decibel sounds
- **Central auditory imperception:** all losses not conductive or sensorineural; organic or functional origin. Permanence of defect depends on origin
- **Organic:** damage interrupting sound transmission between brainstem & cerebral cortex
- **Functional:** no organic lesion exists to explain central auditory loss
### Classification of Hearing Impairment

- **Loudness described via decibels (dB).** 0 dB is softest sound heard; normal speech: 45-55 dB
- **Slight:** hard of hearing; no speech defects
- **Mild:** may miss 25-50% of conversations; may have speech difficulties
- **Moderate:** understands face-to-face at 3-5 ft; difficulty pronouncing words & limited vocabulary
- **Moderately severe:** hears loud conversational speech; needs special speech training
- **Severe:** deaf; may hear loud noises nearby, needs speech training
- **Profound:** deaf; more severe, may hear only loud sounds; requires extensive speech training

### Hearing Impairment Management

- **Prevention paramount (education, vaccination etc)**
- **Early detection to optimize speech development**
  - Neonatal screening programs & follow-up
  - Infants: no response to sound, absence of babble
  - Children: fail to develop speech by 24 mo, use gestures instead of verbalizations, monotone, yelling, request words repeated
- **Lipreading:** stand close, eye level, good lighting, no gum, speak clearly & slowly; sentences simple & short; eliminate background noise
- **Consider using cued speech, sign language, speech therapy, special aids (flashing lights, TDD)**

### Treatment

- **Conductive loss can be improved with:**
  - Surgery, antibiotics, tympanostomy tubes
  - Hearing aid to amplify sound
    - 4 types are commonly used for pediatric clients: behind ear; in ear, body aid & eyeglass aids.
    - Acoustic feedback from poor fit or high volume. Reinsert & turn down volume
- **Sensorineural hearing loss:** cochlear implants receive sound & transmit signal to electrodes so sound perceived. Long-term implications unknown
Visual Impairment

- Visual impairment is loss that cannot be corrected with regular prescription lenses
- School vision/partially sighted: acuity between 20/70 & 20/200; should be able to learn in school reading normal size print
- Legal blindness: 20/200 or less in better eye with correction. Qualifies for special schools & aid
- Etiology: perinatal infections, retinopathy of prematurity, trauma (penetrating or non-penetrating injuries, infections (meningitis)

Visual Impairment

- Myopia (nearsightedness): eyeball too long; light focuses in front of retina causing difficulty with far vision. Rubs eyes, holds book close to eyes. Treated with lenses to improve distance vision; laser surgery also corrective
- Hyperopia (farsightedness): eyeball too short; light focuses behind retina causing difficulty with near vision; most children farsighted until 7 years & usually outgrow. Lenses if needed, laser surgery also corrective
- Astigmatism: unequal curvatures of cornea or lens, prevents light from focusing on retina; special lenses, laser surgery also corrective

Visual Impairment

- Strabismus (crossed eyes) : mal-alignment of eyes that do not focus on same object in space due to lack of muscle coordination
- Amblyopia (lazy eye): reduced visual acuity or loss of vision in one eye unrelated to organic cause. Most common cause is strabismus
- Cataracts: lens become opaque, clouding light & obscuring vision
- Glaucoma: increased intraocular pressure causes increased pressure on optic nerve & eventual atrophy & blindness
Visual Impairment Management

• Prevention through education & safety precautions

• Assessment:
  – Identify at risk children by review of history
  – Observe for behaviors that indicate vision loss
  – Screen for visual acuity & ocular disorders

• Educate & refer as indicated to:
  – Promote child’s optimal growth & development
  – Prevent injury
  – Assist family to gain realistic concept of child’s abilities