CH 74 Child or Adolescent w/ Special Needs

COMMON DISORDERS

COLOR BLINDNESS

- ↓ or lack of ability to see certain colors
- inherited problem, but can become an acquired color vision problem due to aging, SE of some meds, injury to the eye, ↓ visual disorders such as glaucoma, cataracts, macular degeneration, or diabetic retinopathy
- common ex: inability to see reds ↓ greens; blue-yellow color blindness is less common
- monochromacy: black, white, ↓ gray vision
- reduced visual activity, myopic vision, ↓ nystagmus
- Dx: eye exams - Ishihara color blind test charts or the Cambridge color

HEMOCHROMATOSIS

- men 50-60; women after menopause
- a hereditary or an acquired condition in which the body absorbs iron from foods ↓ stores excess iron in body tissues ↓ organs
- common locations for excess stored iron: liver, heart, pancreas, reproductive organs, ↓ skin
- juvenile hemochromatosis becomes symptomatic during the teenage years up to the age of about 30 yrs
- Neonatal hemochromatosis occurs when iron accumulates rapidly in the developing fetus ↓ may be an autoimmune disease
- Secondary hemochromatosis is an acquired form of the disease due to iron deposit accumulation secondary to anemia, chronic liver disease, or an infection
- uncontrolled iron deposits can cause S/E: cirrhosis, liver cancer, diabetes, HF, erectile dysfunction, joint pain, ↓ skin color AS
- Early S/S are nonspecific ↓ can mimic other disorders
- Later, organ damage may be the 1st indicator of excess iron overload ↓ be the etiology of diabetes, loss of libido, cardiac dysrhythmias, or HF

TREATMENT

- reduce the amount of iron in the body; most common method is removal of blood from the body at regularly scheduled phlebotomy sessions · about 1 unit of blood (1 pt/ 470 mL) is removed
- slows the progression of the disease but will not reverse cirrhosis or improve joint pain
- chelation: process in which a chemical is injected into the bloodstream to remove heavy metals or minerals from the body
- heavy metals: iron, lead, mercury, copper, arsenic, ↓ aluminum; mineral calcium
- EDTA (ethylenediaminetetraacetic acid) is injected ↓ binds to the metal; causes the body to expel the substance through stool or urine

↑ absorption of iron

- NURSING CONSIDERATIONS: Avoid iron supplements, multivitamins containing iron, ↓ vitamin C
- Avoid eating raw shellfish because individuals w/ hereditary hemochromatosis are more susceptible to infections caused by bacteria in raw shellfish
- Avoid alcohol, which has the risk of ↑ liver damage
- Drink tannin-rich teas that may slow the storage of iron

NEUROFIBROMATOSIS

- genetically inherited conditions in which benign painful tumors develop as growths on nerve tissue
  - ↳ some cases, becomes malignant

3 types <

- NF2: grow on the 8th cranial nerve (vestibulocochlear nerves); NF1
- Schwannomas grow on cranial, spinal, ↓ peripheral nerves
- extensive café au lait spots on the skin of the facial area ↓ large neurofibromas on the legs; harmless ↓ can be a strong indicator of NF1 if there are >6 spots that are >0.5 cm in children prior to puberty
- present at birth or may appear during the first 5 years
- skeletal system effects: bones that form abnormally (scoliosis), bowing of the legs, osteoporosis, ↓ fractures that do not heal

- degree of hearing deficit or loss, vision deficit or loss, learning & cognitive impairement, cardiovascular complications such as HTN or blood vessel abnormalities, & severe pain
- Schwannomas - severe, debilitating pain; surgical intervention & pain management by a pain specialist
- meningiomas (multiple benign brain tumors) or spinal tumors may require surgeries to remove the growths
- Tx: palliative; pain relief, surgical removal of some problematic lesions, & PT

POLYCYSTIC KIDNEY DISEASE (PKD)

- an inherited disorder in which clusters of cysts develop, primarily w/in the kidneys, but the liver & other areas can also be affected
- cysts are noncancerous, round sacs filled w/ fluid
- HTN & headaches are common; precursors to kidney failure
- Other SIS: chronic back or side pain, abdominal distention, urinary frequency, hematuria, kidney stones, & urinary track or kidney infections
- Serious cardiovascular complications: development of an aneurysm in the brain & the development of mitral valve collapse
- Tx: 1st controlling HTN; antihypertensive drugs (ACE inhibitors)
- NURSING INTERVENTIONS: low-salt diet, fluid intake, & dietary intake of fruit, vegetables, & whole grains · maintain a healthy weight · monitor BP at home · avoid smoke & exercise about 30 min 5-6x-week

TAY-SACHS DISEASE (TSD)

- fatal inherited metabolic disorder
- certain lipids accumulate in the body due to the absence of a fundamental enzyme, resulting in progressive destruction of the neurons (nerve cells) w/in the nervous system
- As the fatty lipid proteins accumulate, damage occurs to the neurons of the brain, in turn causing damage to the eyes, ears, movement, & mental development

By age 2 →
- recurrent seizures & diminished capacity to function mentally & physically; motor abilities gradually regress until the child can no longer crawl, turn over, or sit

Characteristic →
- eye abnormality called a cherry-red spot, which develops at the back of the child's eyes

By age 3 & 4 →
- severely cognitively disabled, blind, paralyzed, & unresponsive

By age 5 →
- child dies because the nervous system can no longer function

- prenatal testing via chorionic villus sampling (CVS) b/n the 10th & 12th wks of pregnancy
- Amniocentesis b/n the 15th & 18th wks of pregnancy

PRADER-WILLI SYNDROME (PWS)

- complex genetic disorder that occurs equally among genders & all races · Ø family hx
- multiphase syndrome, SIS develop in stages
- Initially noted are hypotonia & feeding difficulties
- Later, hyperphagia & weight gain are noted; slow metabolism; calories are not used efficiently for energy
- excessive eating, weight gain, life-threatening obesity, & DMT2
- Intellectual impairement, learning disabilities, & behavioral problems range from mild to moderate in severity
- As the child ages, a short stature is noted & growth hormone may be prescribed; small hands & feets are common
- Incomplete sexual development
- child may have outbursts of temper, compulsiveness & stubbornness but may also be loving & friendly at times
- Distinctive facial features are common, such as narrow forehead, almond-shaped eyes, & triangular mouth
- Unusually fair skin & light-colored hair, underdeveloped genitals & delayed or incomplete puberty
- infertile

FETAL ALCOHOL SPECTRUM DISORDERS & FETAL ALCOHOL SYNDROME

SIGNS & SYMPTOMS

- facial abnormalities, slowed growth, developmental delays, below-normal mental functioning, & dysfunctions of the CNS
- Typical abnormal facial features (small, wide-set narrow eyes w/ drooping upper lids; short upturned nose; flattened cheeks; small jaw):
- SIDS · Failure to thrive · Microcephaly · Sleep & sucking abnormalities in infancy
- Extreme irritability · Epilepsy, seizures · Eye & ear defects · Heart defects
- Delayed gross motor skill development: difficulty rolling over, sitting, crawling
- Delayed fine motor skill development; does not learn new words additionally
- Impairment of fine motor abilities & poor coordination · Poor organ & bone development
- Learning disabilities; memory deficits, shortened attention span · Poor socialization skills
- Hyperactive behavior, distractibility, impulsiveness · Belligerance or stubbornness
- Problems adjusting to daily livings · Poor judgement & reasoning skills
- Inability to live independently · Difficulty understanding or following rules, regulations, & laws
- Worsening social, behavioral, & comprehension difficulties when approaching childhood

NEONATAL ABSTINENCE SYNDROME (NAS)

- group of medical problems that occur in a newborn who was exposed to addictive opiate or narcotic drugs as a fetus
- Withdrawal Sx: anxiety, insomnia, diaphoresis, vomiting, muscle pain, & goose bumps
- can last from a week to about 6 months
- A second type of NAS can occur if opiates (fentanyl/morphine) are used as long-term analgesics for an infant in severe pain
- SIS: tremors, hyperactive reflexes, high-pitched crying, irritability, seizures, & excessive crying; skin mottles (turns shades of reddish-blue)
- diarrhea, vomiting, fever, & diaphoresis
- despite excessive sucking, the infant does not feed well, has tachypnea, has difficulty sleeping, & shows a failure to thrive
- may start in the first 3 days or may take up to a week

DOWN SYNDROME (DS)

- a genetic disorder caused from abnormal cell division of chromosome 21, resulting in extra genetic material
- lifelong intellectual disabilities & developmental delays, that is, & mental acuity & & physical development
- 3 Types: trisomy 21, mosaic DS, & translocation DS
- Trisomy 21 (most common): occurring when either the sperm cell or the egg cell has abnormal cell division after fertilization
- mosaic DS is rare & caused by abnormal cell division after fertilization
- In translocation DS, the only form of the disorder is considered to be inherited. the mother or father has genetic material that is different or rearranged
- Advancing maternal age & the risk

SIGNS & SYMPTOMS

- abnormal crease straight across the palms called a simian line · Tendency toward overweight & obesity
- small flattened nose · small head & neck · Tongue that protrudes · Hypotonia · Extreme flexibility
- Eyes that slant upward · Brushfield spots (tiny white spots on the iris)
- Ears that are unusually shaped or small · Short stature
- Slow growth; remains smaller than peers · Delayed developmental milestones
- Small hands & feet w/ fingers that are shorter than normal
- Wide space b/n the big toe & the rest of the toes · Transverse crease across the soles of the feet

FRAGILE X SYNDROME (FXS) ·no cure

· an inherited intellectual disability carried on the X chromosome, which means that either parent may be a carrier
· specific protein is absent, or, in milder fragile X-associated disorders, is deficient ·autism
· ADD or ADHD; boys are more severely affected than girls
· SIS: intelligence & learning deficits, physical characteristics, social & emotional problems, speech & language difficulties, & sensory problems
· first developmental delays noticed typically are r/t the child's difficulty w/ balance, which makes sitting, moving, or walking difficult
· child may stutter & have disorganized speech

PHYSICAL CHARACTERISTICS
· elongated face, protruding ears, large testes, & a heart murmur

· stereotypic movements such as hand flapping or shyness & social anxiety

Table 74-1 Characteristics of Down Syndrome and Fragile X Syndrome

FEATURE	DOWN SYNDROME	FRAGILE X SYNDROME
Head	Round, small, short	Abnormally large
Face	Flattened profile	Long, large, protruding jaw
Ears	Small, low set	Large, protruding
Eyes	Upward, outward slant; epicanthal folds; Brushfield spots	Wide set; epicanthal folds
Nose	Small; depressed nasal bridge	Flattened nasal bridge
Hands	Short, square; simian creases	Simian creases
Mouth	High-arched palate; protruding tongue; mouth curved downward	High-arched palate
Behavioral	Low-normal intelligence to severe intellectual impairment; language delay	Mild to profound intellectual impairment; short attention span, hyperactivity; temper tantrums; autistic-like behaviors; speech delays

ATTENTION DEFICIT HYPERACTIVITY DISORDER ·ADHD/ADD

· condition associated w/ 3 categories of sx: inattentiveness, impulsivity, & hyperactivity
· 3 types: inattentive, hyperactive-impulsive, & combined
· to be diagnosed w/ ADHD, the sx must be severe, occur for at least 6 months, & significantly affect the child's developmental milestones

SIGNS + SYMPTOMS

· Inattentiveness
  · easily distracted & can't focus on one single task
  · seems forgetful; can't remember where items were placed
  · daydreams & becomes easily confused
  · becomes bored w/ an activity shortly after starting it
  · can't seem to follow directions or understand something new
  · has difficulty organizing a task or completing homework
· Hyperactivity
  · talks nonstop
  · wants to be constantly moving
  · can't sit still; fidgets or squirms during meals or at school
  · touches everything w/in sight
· Impulsivity
  · interrupts other's conversations
  · is emotionally inappropriate; shows no emotional restraint
  · acts out w/ inappropriate comments
  · shows impatience; can't wait to be given attention

TOURETTE SYNDROME (TS)

- an inherited, neurologic disorder of unknown cause characterized by repetitive, multiple involuntary movements + uncontrollable vocalizations called tics

SIGNS + SYMPTOMS

- 1st noticed in childhood, in the head + neck areas, generally b/n the ages of 3 + 9 years
- Motor tics: arm + head jerking, blinking, making a face, mouth twitching, + shoulder shrugging
- vocal tics: barking, yelping, grunting, coughing, + clearing of the throat
- Simple tics involve a limited # of muscle groups, such as blinking or making a face
- Complex tics: jumping + swearing
- Many situations can affect the intensity of tics, such as stress, illness, fatigue, or emotions
- coprolalia + echolalia - small *
  - ↳ socially inappropriate words
- Dx: presence of both motor + vocal tics for at least 1 year
- Tx: neuroleptic drugs - haloperidol (Haldol) - to suppress severe or significant tics that interfere w/ ADLs

PLUMBISM - lead poisoning

- occurs slowly over a period of weeks or months - at high levels it can be fatal

SIGNS + SYMPTOMS

- anemia, severe stomach aches, hearing loss, muscle weakness, + mental + physical damage
- permanent brain damage
- Adults: HTN, infertility, muscle + joint pains, nerve disorders
- pain, headache, irritability, memory loss, decline in mental functioning, mood swings
- Blue/blue-black line on the gums near the teeth - Hyperirritability - Anorexia, N/V
- Intermittent vomiting (lead colic) - Abdominal pain - Joint pain - Headache - Fatigue + ↓ play
- Anemia, pallor, + ↓ RBCs - Constipation - Behavior ds - Ataxia (unsteady gait), weakness, or clumsiness
- ↓ in intellectual + mental abilities - Impaired LOC - Seizures - Coma - Encephalopathy

- Dx: Blood lead level (BLL)
- Tx: individual first must be removed from the lead source
- mild: treated symptomatically - if lead levels are high, chelation is considered - EDTA IV + BAL in oil via deep IM (tree nut allergic)

MUSCULAR DYSTROPHY (MD)

- characterized by gradual, progressive weakness + loss of muscle mass w/ degeneration of the skeletal muscles - some affect cardiac muscle
- Duchenne muscular dystrophy (DMD) is the most common form of MD, characterized by progressive atrophy of symmetrical groups of skeletal muscles in boys b/n the ages of 2 + 12 years
- progresses rapidly; By age of 12 years, most boys are unable to walk
- At some point, the teen may not be able to breathe w/o mechanical assistance
- Becker muscular dystrophy (BMD) is similar to DMD, but the sx are less severe
- myotonic MD (MMD) is the most common adult form
- causes sustained muscle spasms, endocrine problems, cataracts, + cardiac abnormalities
- "swan-like" neck w/ a long, thin face, + drooping eyelids

SIGNS + SYMPTOMS

- beam to appear around the age of 3 years
- child may not be able to walk upstairs, run, or get up off the floor
- gait appears as a waddle + a (+) Gowers sign occurs, in which the child uses their upper extremity muscles to compensate for weak hip muscles
- may walk on the toes, fall frequently, + have difficulty hopping or running; lordosis, scoliosis, + contracture deformities, especially at the hips + knees
- delayed intellectual development + borderline IQ may be present

- gradual muscle atrophy occurs, & by the age of 11 or 12 years, the child is unable to walk & becomes wheelchair bound
- lose ability to feed themselves; muscles involved in swallowing are also affected, & tube feeding may be necessary
- difficulty breathing & begins to have cardiac failure
- in some cases, assisted respiratory ventilation can extend life
- Tx: support, corticosteroids, anticonvulsants, immunosuppressants, antibiotics
- wheelchair - pacemaker may be inserted