NR602 Final study guide notes

Primary Care Of The Childbearing (Chamberlain University)

NR602: Pediatric Study Topics

Pediatric Study Topics

Eye Disorders- 616- 646

Strabismus- a defect in In ocular alignment, or the position of the eyes in relation to each other; It is commonly called lazy eye. And strabismus, the visual axes are not parallel because the muscles of the eyes are not coordinated; When one eye is directed straight ahead, the other deviates.

Retinoblastoma is an intraocular tumor that develops in the retina. Although it is rare, this malignant retinal tumor is the most common tumor in childhood (some 4% of cancers in children younger than 15 years of age)

Clinical findings

- strabismus is the most common finding
- there is a decreased visual acuity uni- or bilateral white pupil (leukocoria), described often as an intermittent "glow, glint, gleam, or glare" by parents, is usually seen in low light settings or noted in photographs taken with a flash i.e. (cat's eye reflex)
- other symptoms include an abnormal red reflex, nystagmus, glaucoma, orbital Cellulitis and photophobia, hyphema, hypopyon (plus an anterior Chamber of eye); Signs of global rupture or also possible

Bulbar or palpebral conjunctival injection is a common presentation, which can be unilateral or bilateral. **differential diagnosis** should include allergy, conjunctivitis, infection, foreign body, chemical exposure, or systemic inflammatory disease, irritation of the conjunctiva or cornea, and congenital glaucoma.
Watery discharge can occur with allergies, nasolacrimal obstruction, foreign bodies, viral infection, and iritis.
Purulent or mucoid discharge can be noted with chronic dacrocystitis or nasolacrimal obstruction.
Advanced allergic conjunctivitis can have some mucoid production.

To differentiate, microscopic investigation of discharge may lead to other clues.

Photophobia is a symptom common of trauma and in infants with glaucoma or retinal disease. Other non-eye related causes of photophobia include migraines and meningitis.
A white pupil, *or* leukocoria is a serious finding and demands immediate referral to the pediatric ophthalmologist.
Causes of leukocoria include retinal detachment, cataract, retinal dysplasia, retinopathy of prematurity, and in newborns retinoblastoma.
All newborns should have a fundoscopic examination within 24 hours of birth and yearly on physical examinations.

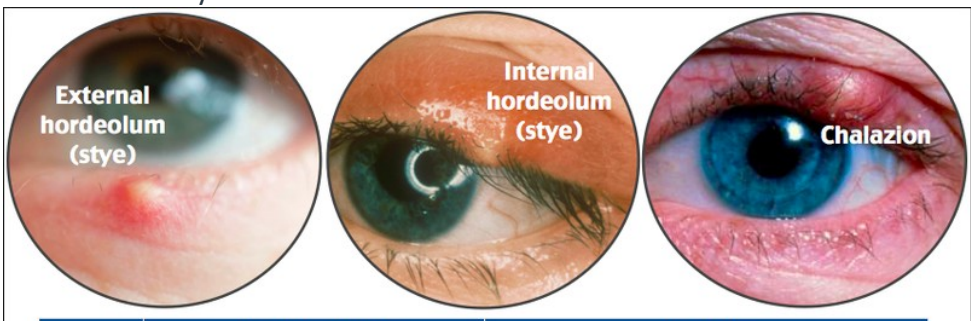
Conjunctivitis chart

TABLE 35.6 Types of Conjunctivitis

Type	Incidence/Etiology	Clinical Findings	Diagnosis	Management
Ophthalmia neonatorum	Neonates: *Chlamydia trachomatis, Staphylococcus aureus, Neisseria gonorrhoeae,* HSV (silver nitrate reaction occurs in 10% of neonates)	Erythema, chemosis, purulent exudate with *N. gonorrhoeae*; clear to mucoid exudate with *Chlamydia*	Culture (ELISA, PCR), Gram stain, R/O *N. gonorrhoeae, Chlamydia*	Saline irrigation to eyes until exudate gone; follow with erythromycin ointment For *N. gonorrhoeae:* ceftriaxone or IM or IV For chlamydia: erythromycin or possibly azithromycin PO For HSV: antivirals IV or PO
Bacterial conjunctivitis	In neonates 5-14 days of age, preschoolers, and sexually active teens: *Haemophilus influenzae* (nontypeable), *Streptococcus pneumoniae, S. aureus, N. gonorrhoeae*	Erythema, chemosis, itching, burning, mucopurulent exudate, matter in eyelashes; worse in winter	Cultures (required in neonate); Gram stain (optional); chocolate agar (for *N. gonorrhoeae*) R/O pharyngitis, *N. gonorrhoeae,* AOM, URI, seborrhea	Neonates: Erythromycin 0.5% ophthalmic ointment ≥1 year of age: Fourth-generation fluoroquinolone For concurrent AOM: Treat accordingly for AOM Warm soaks to eyes three times a day until clear No sharing of towels or pillows No school until treatment begins
Chronic bacterial conjunctivitis (unresponsive conjunctivitis previously treated as bacterial in etiology)	School-age children and teens: Bacteria, viruses, *C. trachomatis*	Same as above; foreign body sensation	Cultures, Gram stain; R/O dacryostenosis, blepharitis, corneal ulcers, trachoma	Depends on prior treatment, laboratory results, and differential diagnoses Review compliance and prior drug choices of conjunctivitis treatment Consult with ophthalmologist
Inclusion conjunctivitis	Neonates 5-14 days of age and sexually active teens: *C. trachomatis*	Erythema, chemosis, clear or mucoid exudate, palpebral follicles	Cultures (ELISA, PCR), R/O sexual activity	Neonates: Erythromycin or azithromycin PO Adolescents: doxycycline, azithromycin, EES, erythromycin base, levofloxacin PO
Viral conjunctivitis	Adenovirus 3, 4, 7; HSV, herpes zoster, varicella	Erythema, chemosis, tearing (bilateral); HSV and herpes zoster: unilateral with photophobia, fever; zoster: nose lesion; spring and fall	Cultures, R/O corneal infiltration	Refer to ophthalmologist if HSV or photophobia is present Cool compresses three or four times a day
Allergic and vernal conjunctivitis	Atopy sufferers, seasonal	Stringy, mucoid exudate, swollen eyelids and conjunctivae, itching (key finding), tearing, palpebral follicles, headache, rhinitis	Eosinophils in conjunctival scrapings	Naphazoline/pheniramine, naphazoline/antazoline ophthalmic solution (see text) Mast cell stabilizer (see text) Refer to allergist if needed

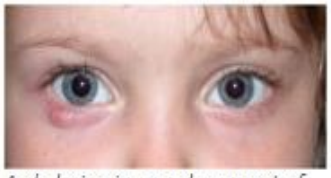
AOM, Acute otitis media; *EES,* erythromycin ethylsuccinate; *ELISA,* enzyme-linked immunosorbent assay; *HSV,* herpes simplex virus; *IM,* intramuscular; *IV,* intravenous; *PCR,* polymerase chain reaction; *PO,* (by mouth, orally); *R/O,* rule out; *URI,* upper respiratory infection.

Chalazion vs Stye



	Hordeolum (Stye)	Chalazion
Location	Most commonly found at or near an eyelash follicle	Most commonly found above the eyelashes on the upper lid
Cause	Bacterial infection either at the root of the eyelash follicle or in the oil gland of the lids	A blocked oil gland (Meibomian or Zeiss)
Symptoms	Tenderness, swelling	Firm, painless lump
Treatment	Spontaneous drainage, warm compresses	Warm compresses, antibiotic eyedrops, surgery

CHALAZIONS – Benign, chronic lipogranulomatous inflammation of the eyelid



A chalazion is an enlargement of an oil-producing gland in the eyelid.

Causes – blockage of the meibomian cyst

Risk – hordeolum or any condition which may impede flow through the meibomian gland. Also mite species that reside in lash follicles

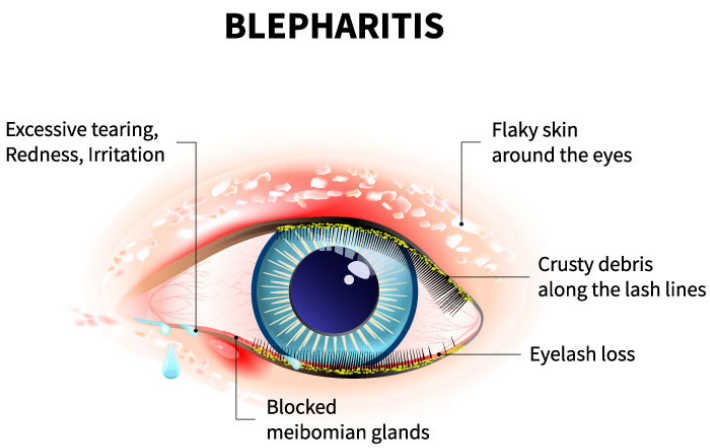
Assessment – PAINLESS, NOT INVOLVING LASHES
Lid edema, or palpable mass
Red or grey mass on the inner aspect of lid margin

Prevention – good eye hygiene

Treatment – warm, moist compresses 3x per day
Antibiotics not indicated because chalazion is granulomatous condition, if secondarily infected consider SULFACETAMIDE, ERYTHROMYCIN

Follow up – 2-4 weeks, if still present after 6 weeks follow up with ophthalmologist

Blepharitis-



BLEPHARITIS – Inflamation/infection of the lid margins (chronic problem)

2 Types:
seborrheic (non ulcerative) : irritants (smoke, make up, chemicals)

- s&s – chronic inflammation of the eyelid, erythema, greasy scaling of anterior eyelid, loss of eyelashes, seborrhea dermatitis of eyebrows and scalp

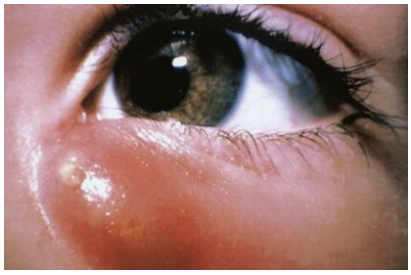
Ulcerative- infection with staphylococcus or streptococcus

- s&s – itching, tearing, recurrent styes, chalazia, photophobia, small ulceration at eyelid margin, broken or absent eyelashes
- the most frequent complaint is ongoing eye irritation and conjunctiva redness

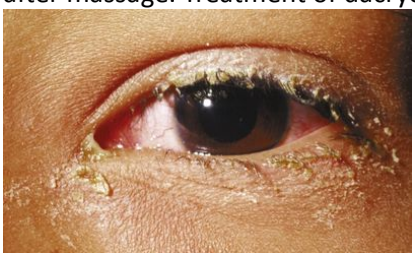
Treatment – clean with baby shampoo 2-4 times a day, warm compresses, lid massage (right after warm compress)

For infected eyelids – antistaphyloccocal antibiotics BACITRACIN, ERYTHROMYCIN 0.05% for 1 week AND QUIONOLONE OINTMENTS

For infection resistant to topical – TETRACYCLINE 250 MG PO X4
DOXYCYCLINE 100 MG PO X2



Dacryocystitis is an inflammation of the involved nasolacrimal duct; infection can result. Treatment: Gentle pressure applied in a downward and medial direction transmits hydrostatic force through the nasolacrimal duct to the obstruction. This technique should be performed two or three times a day. The eyelid should be cleaned with plain water after massage. Treatment of dacryocystitis is warm compresses AND oral or parenteral antibiotics



For uncomplicated bacterial conjunctivitis, treatment includes the following:
• Sodium sulfacetamide 10% ophthalmic solution or ointment; not effective against H. influenzae; stings; can cause allergic reactions (including Stevens-Johnson syndrome)
• Trimethoprim sulfate plus polymyxin B sulfate ophthalmic solution
• Erythromycin 0.5% ophthalmic ointment for patients with sulfa allergy and infants Azithromycin drops for children older than 12 months
• Fluoroquinolone ophthalmic drops including besifloxacin, ciprofloxacin, gatifloxacin, levofloxacin, moxifloxacin, or ofloxacin for children older than 12 months
The aminoglycosides (neomycin, tobramycin, and gentamicin) are to be avoided because of possible hypersensitization, severe allergic reactions, and increasing resistance.

CONJUCTIVITIS – inflammation or irritation of conjuctiva

Bacterial (PINK EYE) – in peds bacteria is the mosts common cause, contact lens, rubbing eyes, trauma,

S&S – purulent exudate, initially unilateral, then bilateral
Sensation of having foreign body in the eye is common
Key findings – redness, yellow green, purulent discharge, crust and matted eyelids in am
Self limiting 5-7 days. Eye drops – polytrim, erythromycin, tobramycin or cipro (fluoroquinolones, macrolides)
Improvement 2-4 days
Most common organism H. influenza <7

Viral – **adenovirus**, coxsackie virus, herpes, molluscum
S&S – profuse tearing, mucous discharge, burning, concurrent URI, enlarged or tender preauricular nose
Antihistamines/decongestant
Improvement, self limiting, 7-14 days

Chlamydial – chlamydia trachomatis
S&S – profuse exudate, associated with genitourinary symptoms, 1-2 weeks after birth
Gonococcal – 2-4 days after birth, most concern can cause blindess
PO azithromycin, doxycycline (tetracyclines increase photosensitivity, don't use in pregnancy)
Improvement 2-3 weeks

Allergic – IgE mast cell reaction, environmental, cosmetics
S&S – marked conjuctival edema, severe itching, tearing, sneezing
Topical antihistamine or topical steroids
Improvement 2-3 days

Chemical –thimerosal, erythromycin, silver nitrate
S&S conjuctival erythema, 30 minutes afer prophylactic antibiotics drops
Avoid contact
Can consider steroids
Conjunctivitis never accompany vision changes

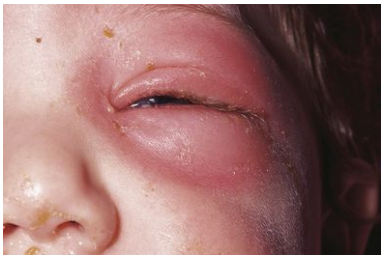
Diagnostic studies: swap and scraping must be done, gram and Giemsa staining, ELISA, PCR testing, newborn < 2 weeks needs to be tested for gonorrhea

Non –pharm – clean towels, change pillows, warm compress, no contacts, no eye make up – mascara
Gonococcal conjunctivitis: newborn – give Ceftriaxone IM once (don't give if hyperbilirubinemia,

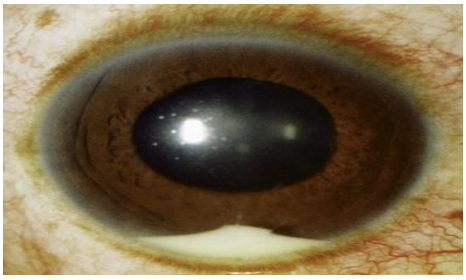
Non-gonococcal – erythromycin 0.5% ointment
Consider fluorescein staining if abrasion suspected

CDC recommends prophylactic administration of antibiotic eye ointment (ERYTHROMYCIN) 1 hour after delivery

Refer to ophthalmologist if herpes, hemorrhagic conjunctivitis or ulcerations present
May return to work/school 24 hours after topical



cellulitis. oral ABX are indicated. Per our text, a 7- to 14-day course. Amoxicillin with clavulanic acid, cefdinir, and cefpodoxime are first-line choices for treatment. Clindamycin is indicated if MRSA is suspected. clindamycin or a combination regimen of trimethoprim/sulfamethoxazole (TMP-SMX) PLUS amoxicillin or cefpodoxime or cefdinir



Uveitis - cycloplegics and systemic corticosteroids. Refer to ophthalmologist. treat underlying disease.

Common eye traumas that may present to primary care:
Corneal abrasion
Foreign Body
Retinal detachment

Musculoskeletal Injuries assessment and treatment

Osgood-Schaltter Disease – tibial tubercles and apophysis
Aseptic necrosis of the tibial tubercles and apophysis.
Signs and symptoms: painful swelling of tibial tubercle, limp, intermittent pain over months, hip pain may be aggravated by extension of knee against resistance, worsens with squatting, stair walking, forceful contraction of the quadriceps, usually due to overuse injury associated with athletic activity during rapid growth of tibial tuberosity, relived by rest. More common in males.
Diagnostics: X-ray, MRI if osteochondral lesion.
Treatment: Rest, modified activity, ice, NSAIDs, Quadriceps strengthening and stretch, tibial band during activity.

Talipes Equinovarus Congenita (Clubfoot)
Adduction of forefoot with plantar flexed at ankle (equines), or curves in (varus). Clinical **Diagnosis**: X-ray
Treatment Ortho, casting

Tibial torsion or femoral inversion
Foot turns in during walking or running. Pigeon-toed appearance. May be congenital or acquired from increased load on femur due to sitting in utero incorrectly, or FHx.
Diagnostics: usually clinical or leg and hip X-ray.
Treatment: Reassurance, braces, special shoes, and/or cast. Refer to ortho of severe deformity. Excellent prognosis.

Growth Pains
Idiopathic.
Signs and symptoms: pain at the lower limbs, bilateral, intermittent, localized to the muscles. Rule out neuro disorders.
Treatment Reassurance, massage, heat, NSAIDs if severe.

Overused injury "Little League" elbow
Physical stress produces forces in and around the elbow during throwing motion (baseball or softball), valgus stress is placed on elbow resulting in tension on the medial structures, repetition causes pathological changes.
Signs and symptoms: pain in elbow, decreased elbow ROM, mild flexion contracture, point tenderness, swelling, decreased performance.
Diagnostics: X-ray.

Treatment: Ice, resting arm, NSAIDs.

Genu Varum

Alignment of the knee with the tibia medially (varus) in relation to the femur. Bowlegged appearance as result of uterine position. Measure distance between knees with feet together, distance should be less than 5 inches. An angular deformity is physiologic and normal up to 3 y.o. Pathologic if more than 15 degrees. Associated with short stature, rapidly progressing

Treatment Brace/splint, NSAIDs for pain, and strengthening exercise. If rickets-prescribe Vitamin D. If overweight- weight management.

Genu Valgum

Alignment of the knee with the tibia laterally deviated (valgus) with relation to the femur. Commonly known as knock-kneed. Physiologic (over the first 3 years, 10-15 degrees, more common in girls) or pathologic (before age of 2 y.o, valgus angle >15 degrees, increasing in severity. Associated with short stature, obesity and asymmetry, due to metaphyseal dysplasia or injury).

Treatment Braces for angles more than 15 degrees. In some cases, resolves spontaneously by age 6. Refer to orthopedic for suspected pathology.

Toxic Transient Synovitis

Unilateral inflammatory arthritis of the hip with acute onset, decreased hip ROM-extension and internal rotation, painful limp, crying at night, more common in boys 3-6 y.o.

Clinical diagnosis: WBC w/ leukocytosis, ^ESR, hip X-ray normal.

Treatment Bedrest, no weight bearing on affected hip, NSAIDs, self-limiting

Legg-Calve Perthes Disease (LCPD) – femoral head

Avascular necrosis of the femoral head epiphyses associated w/ trauma, transient synovitis, coagulation abnormalities. Associated with low birth weight and socioeconomic status, and white race. Insidious onset of painful limp of thigh, knee, hip, worst with activity and not relived by rest. Restriction of voluntary motion limited passive motions, abduction and rotation of affected hip, atrophy of thigh or calf may be noticed.

Diagnostics: Hip X-ray, MRI

Treatment Abduction brace or long leg cast, surgery for bone reconstruction.

Muscular Dystrophy (MD)

Inherit muscle disorders-Congenital (6 types with CNS involvement), X-linked recessive (Duchenne's MD is most common and Becker's MD milder form manifests after exercise w/ muscle ache).

Signs and symptoms: hypotonia, muscle weakness, contractures, motor deficit. Diagnostics: CK is a good screening test-elevated, muscle biopsy, MRI, EMG.

Treatment IP involvement, no effective Tx as goal is maximal functioning. Corticosteroids slow progression 2-4 years, supportive therapies - PT, OT, nutrition, orthotics. Poor prognosis, once the chest muscle is affected, death will result.

Scoliosis

Lateral curve of spine. High incidents, greater in adolescent girls, onset in pubertal

Treatment

Functional Scoliosis: 5-10 degree curvatures are functional and are monitored.

Structural scoliosis: mild-curvature <20 degree-closely observe every 3-4 mo during growth spurts. Serious cases with 25-40 degrees, refer to ortho, Milwaukee brace for 23 hrs a day. Back exercise, spinal fusion - those may create psychological (negative body image, depression) and social issues (limited sport and group

participation). Use cognitive-behavior therapy. Functional scoliosis is self-limiting, structural scoliosis produces prominent scapula, ribs, uneven shoulder and asymmetric waistline or back pain. F/U annually during well child exam.

Slipped capital femoral epiphysis

Displacement of the femoral head from the femoral neck in a downward and backward position due to trauma, gradual slip from chronic abnormal forces, common in 9-15 y.o. boys before epiphyseal plate closes. More common in African American and Pacific Islanders. Associated with endocrine or systemic disorders.

Signs and symptoms: pain, at anterior thigh, knee, if hip is externally rotated or abducted, flexion is limited. In acute cases - injury proceeds pain, not weigh bearing on affected site, holds limb in external rotation. Painful passive ROM. Diagnostics: X-ray-AP and lateral pelvis, Frog-leg laterally.

Treatment: Refer to ortho STAT for surgical fixation.

Rashes and Dermatologic conditions 567-614

Pediculosis, or lice

three forms: head louse, pubic louse, or body louse.

Lice differ from scabies in that they live on the hair and only feed on the host for nutrients instead of burrowing in the skin.

Itching is also the prominent symptom in lice.

Children may exhibit no symptoms at all.

The presence of pubic lice in young children may be a red flag for sexual abuse and should be investigated further by the clinician. Lice on eyelashes can cause irritation and even blepharitis.

Primary treatment is removal of all nits, louse ova.

Brown nits on the hair shaft closer to the scalp indicate active infestation.

White nits that are more than 2 inches out on the hair shaft indicate a previous infection. These nits can remain stuck to the hair shaft for weeks after an active infestation and do not reflect current infestation.

may be noted more prominently in the hair directly behind the ears and at the base of the neck.

Resistance in treatment is growing and as many as 20% to 30% of nits may survive a preliminary treatment and may require a second treatment.

As with scabies, towels, clothing, bedding, pillows, stuffed animals, and so forth should be washed in hot water and dried if possible. Plastic bag enclosure should be for at least 2 weeks

Scabies

most common infestation found in humans and can have complications such as cellulitis.

The first clinical symptoms is 3 to 4 weeks after initial egg lay, creating an environment for easy transmission.

Signs and symptoms may vary, but itching is a classic symptom along with excoriation in folds of skin on extremities, between fingers or toes, or in antecubital spaces.

All household members should be treated simultaneously and the treatment of **permethrin 5% can be toxic for smaller children**, particularly with a larger body surface area or more immature body systems. To kill mites

and insects in the environment, sprays are not advised. Instead, heat and lack of oxygen are practical methods. Washing clothes and linen in hot water and drying in a drier on a maximum setting or dry-cleaning and keeping objects in a sealed plastic bag for at least 7 days is required

When documenting any skin lesions, it is important to

1) begin with the distribution

2) move to appearance of configuration

3) then color

4) then lesion type

5) and any primary or secondary changes.

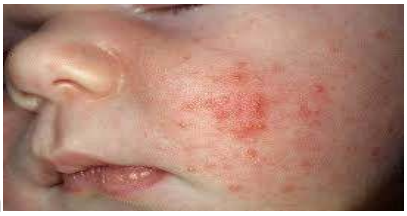
There are also some common terms in dermatology nomenclature.

• A primary lesion is the basic lesion or complaint characteristic of the disease and includes terms you are already familiar with, like macula, vesicle, petechial, and so forth.

• A secondary lesion is an effect of the primary lesion, such as a crusting from exudate, surrounding erythema, or excoriation marks from scratching

Neonatal Acne

tiny red or white bumps or pimples

no treatment, will resolve on its own 

Nevus

Hemangioma

bright red birthmark that shows up at birth or in the first or second week of life

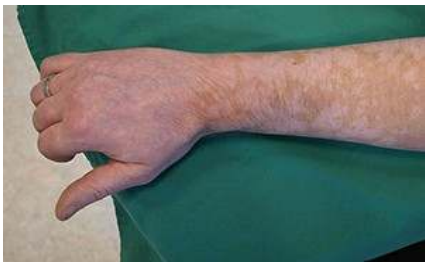
looks like a rubbery bump and is made up of extra blood vessels in the skin

Beta blocker, corticosteroids or laser treatment if does not resolve on its own

Cafe au lait spot

flat, pigmented spots on the skin

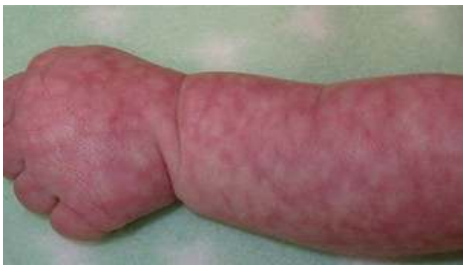
more than 6 can be a sign of neurofibromatosis



Mottling

skin looks blue or pale and blotchy.

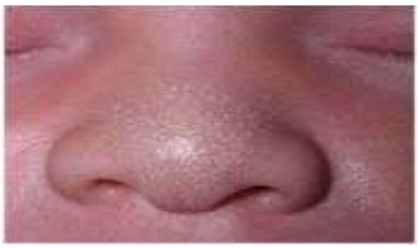
noticeable if the baby is uncovered or cold



Milia

tiny white bumps that appear across a baby's nose, chin or cheeks

You can't prevent milia. And no treatment is needed because they usually disappear on their own in a few weeks or month



Mongolian spots

Spots are flat, gray-bluish in color (almost looking like a bruise) and can be small or large. They are caused by some pigment that didn’t make it to the top layer when baby’s skin was bring formed. They are harmless and usually fade away by school age. Congenital birth mark seen most commonly over the lumbosacral area. Bluish green to black in color and oval to irregular shape. Most commonly found in individuals of African or Asian ethnic background.

Lead (Burn's Pediatric Primary Care – pp. 934-935)

Risk Factors

· Pica
· Child resides nearby a lead smelter, battery recycling plant, or any industry that is likely to release lead
· Family member or caretaker works with lead-based products/material
· Household members engage in hobbies such as ceramics, stained glass, or making their own fish tackle
· Painted or unusual materials burned on wood stoves or fireplaces
· Family uses folk or herbal remedies
· Food is prepared or stored on imported pottery, metal containers, or water from contaminated pipes

Physical Examination

· Clinical signs may not be present upon examination because lead is stored in the bones.
· It is NOT measured by blood lead levels (BLLs) (<5 mcg/dL to greater than or equal to 70 mcg/dL)

- A child can have high BLLs (45 mcg/dL) and have no obvious clinical signs of lead toxicity
- Another child may present with severe GI complaints and low BLLs (15 to 20 mcg/dL)

· Many children have subclinical symptoms or symptoms of other ailments such as anemia, constipation, abdominal pain, impaired hearing, learning disabilities, delayed growth, delayed growth, and/or hyperactivity

- GI infections, anemia (other causes/etiology), behavioral disorders, growth retardation, ADHD, and CNS infections are differential diagnoses.

· At higher levels, lead affects Vitamin D metabolism, velocity of nerve conduction, hemoglobin synthesis which can lead to myocardial excitability, increased ICP, coma, and death
· Many children do not exhibit signs of acute toxicity until they have high lead levels

Diagnostic Studies

· At risk children should be screened for lead by ages 1 to 2 years old.
  - Immigrant children ages 3 to 6 years old should be screened also if they have not been previously screened for lead toxicity
· Any child who appear to exhibit signs of lead toxicity should be screened
· If elevated lead levels are present, assessment of Free Erythrocyte Protoporphyrin (FEP), and Zinc Protoporphyrin (ZPP), and iron deficiency screening including serum ferritin are helpful in determining diagnosis and management.

Management

· Prevent the child's exposure to lead in the environment, monitoring BLLs, correcting any dietary deficiencies, investigating & removing lead (i.e. lead abatement) sources from the child's environment, and treating the child for lead toxicity.
  - Children in the same household should be tested and treated accordingly
· In all cases of lead toxicity (greater than or equal to 5 mcg/dL):
  - Inform caregiver of level of lead toxicity
  - Provide caregiver dietary and environmental education
  - Remove child from source of lead if known
  - Report to the public health department
  - Initiate environmental investigation. Some health departments may do this
  - Initiate lead hazard control/abatement
  - Follow up BLL every 3 months until BLL declines
  - Refer to social services if appropriate

· Chelation therapy is recommended for levels higher than 45 mcg/dL to treat acute, severe, and life- threatening poisoning

Patient & Family Education

· Prevention of lead poisoning is a public health responsibility, and public health officials use geographic information systems (GIS) to identify high risk areas
· PCPS play a critical role in preventing lead toxicity in children
  - Pediatric providers should take a lead exposure history on all children and families and identify all children who need to be screened, retested, and require appropriate intervention and treatment
· Parents need to be informed that it is critical to have professional assessment of the level of home contamination and professional abatement may be necessary
· Public health officials can work with parents to identify ways to control lead dust and paint chips in older homes and can work with landlords on lead abatement properties
· Other strategies parents can use:
  - Cover smaller peeling paint areas with sticky-backed paper
  - Damp-mop and damp-dust with household cleaners or lead-specific cleaning products (e.g., Ledizolv) twice weekly to decrease lead dust in the air; do not dry mop or sweep
  - Pick up and dispose of paint chips with a disposable rag or paper towel soaked in phosphate cleaner
  - Run water until temperature changes to flush pipes of lead sediment
  - Do not store or cook food in lead crystal or pottery that contains lead
  - Remove work clothes and wash hands before returning home if job is lead-related
· Inform parents that chelation therapy leads to a rapid fall in BLL, but most children have a rebound increase within days or weeks of treatment; repeated treatment may be necessary

Table 44.5 Management Recommendations for Lead Poisoning (p. 935)

Screening Sample: Blood Lead Levels	Action to Be Taken	Follow-Up Blood Levels Monitoring
< 5 mcg/dL	Not considered lead poisoning: · Provide caregiver/parent dietary and environmental education	· If high risk, retest in 6 months · If low risk, no further testing necessary; ongoing monitoring for changes in environment

	· Refer to social services if appropriate · Conduct environmental assessment to determine if child is exposed to a lead source (e.g., pre-1978 housing)	
Greater than or equal to 5-9 mcg/dL	· Reference value requiring intervention: ○ Confirmatory venous blood test within 1 week to 1 month	· Early follow-up (2 to 4 tests) by 3 months · Later follow up (after BLLs decline) at 6-9 months
10-44 mcg/dL	Confirmatory venous blood tests within 1 week to 1 month (the higher the BLL, the sooner confirmatory testing should be done)	
10-19 mcg/dL		· Early follow-up 1-3 months (may do at 1 month to ensure BLL are not rising rapidly) · Later follow-up at 3-6 months
20-24 mcg/dL		· Early follow-up 1-3 months (may do at 1 month to ensure BLL are not rising rapidly) · Later follow-up at 1-3 months
25-44 mcg/dL		· Early follow-up 2 weeks to 1 month · Later follow-up at 1 month · Retest every month until results <15 mcg/dL for at least 6 months, then retest every 3 months until child is 36 months old
45-59 mcg/dL	Confirmatory venous blood test within 48 h: · Complete history and physical	· Early follow-up as soon as possible · Retest every month until

	examination · Complete neurological examination · Lab work: Hbg, or Hct, and iron status (FEP or ZPP) · Abdominal x-ray with bowel decontamination if indicated · Chelation therapy, may be oral in ambulatory setting	results <15 mcg/dL for at least 6 months, then retest every 3 months until child is 36 months old
60-69 mcg/dL	Confirmatory venous blood test within 24 h: · Complete history and physical examination · Complete neurological examination · Lab work: Hbg, or Hct, and iron status (TIBC or SF) · Abdominal x-ray with bowel decontamination if indicated Chelation therapy, may be oral in ambulatory setting	· Early follow-up as soon as possible · Retest every month until results <15 mcg/dL for at least 6 months, then retest every 3 months until child is 36 months old
Greater than or equal to 70 mcg/dL	Medical emergency: · Retest immediately as an emergency lab test with venous blood sample · Hospitalize for IV chelation · Proceed according to action for 45-69 mcg/dL	· Early follow-up as soon as possible · Retest every month until results <15 mcg/dL for at least 6 months, then retest every 3 months until child is 36 years old

Headaches 978-983

Prevalence rates for migraine headaches are reported to be: age 3 (3% to 8%), age 5 (19.5%), age 7 (37% to 51%), and 7 to 15 years old (57% to 82%; Antonaci et al., 2014). Children as young as 2 years have been described as having migraine-like symptoms. Before 10 years old, the incidence is higher in males than females. During teenage years, females have a higher headache incidence, with hormonal influences likely playing a role.

Box 46.2 Most Common Types of Primary Headaches Seen in Primary Care Settings

Diagnostic Criteria Based on History

Pediatric Migraine Headache

- A. More than five attacks fulfilling features of B through D
- B. Duration: 2-72 h
- C. At least two of the following features:

- 1. Bilateral or unilateral (commonly bilateral in young children; unilateral pain usually emerges in late adolescence or early adult life)
  - a. Usually frontal/temporal
  - b. Occipital location is unusual and should be carefully evaluated (occipital headache in children whether unilateral or bilateral is rare and calls for diagnostic caution; many cases are attributable to structural lesions)
- 2. Pulsating quality
- 3. Moderate to severe intensity aggravated by routine physical activity
- 4. At least one of the following:
  - a. Nausea and/or vomiting
  - b. Photophobia and phonophobia (can infer from behavior)
- 5. Not attributed to another disorder

Infrequent Episodic Tension Type Headache

- A. At least 10 episodes occurring on <1 day per month on average (<12 days/year) and fulfilling criteria B through D
- B. Headache lasting from 30 min to 7 days
- C. Headache has at least two of the following characteristics:
  - 1. Bilateral location
  - 2. Pressing/tightening (non-pulsating) quality
  - 3. Mild or moderate intensity
  - 4. Not aggravated by routine physical activity, such as walking or climbing stairs
- D. Both of the following:
  - 1. No nausea or vomiting (anorexia may occur)
  - 2. No more than one of photophobia or phonophobia
- E. Not attributed to another ICHD-3 diagnosis

Chronic Tension Headache

- A. Headache occurring on 15 days per month on average for >3 months (180 days/year) and fulfilling criteria B through D
- B. Headache lasts hours to days or may be continuous
- C. Headache has at least two of the following characteristics:
  - 1. Bilateral location
  - 2. Pressing/tightening (nonpulsating) quality
  - 3. Mild or moderate intensity
  - 4. Not aggravated by routine physical activity such as walking or climbing stairs
- D. Both of the following:
  - 1. No more than one of photophobia, phonophobia, or mild nausea
  - 2. Neither moderate or severe nausea nor vomiting
- E. Not attributed to another ICHD-3 diagnosis

- *History*
- Duration:
- Frequency and triggers: Children with recurrent, low-intensity headaches, with no neurologic changes, and who recover completely between episodes are unlikely to have serious intracranial etiology. Triggers can include ovulation or menstruation, exercise, food or odors, and stress. Other triggers can include chocolate, processed meats, aged cheeses, nuts, altered amounts of caffeine intake, dairy products, shellfish, and some dried fruits. Consistent findings such as perimenstrual exacerbation, food triggers, and a stable pattern to the headache with intervals of wellness over a long time period are reassuring symptoms that suggest a primary headache. In most cases, a specific trigger or etiology is not ever identified.
- Location: Occipital or consistently localized headaches can indicate underlying pathology. Facial pain might be sinusitis. Oculomotor imbalance can produce a dull periorbital discomfort, whereas temporomandibular joint pain tends to localize around the periauricular or temporal areas. The child should be asked if the pain is unilateral or bifrontal.
- Quality and severity of pain: Sharp, throbbing, or pounding pain may indicate vascular migraine. Dull and constant pain may be tension or organic. Severity can be assessed by asking about limitations to activities and missed school days, although there are other factors

that contribute to missed school and limited activities. How many “different kinds of headaches” are experienced?

- Age of onset: Progression of the headaches over time and longest period of time without symptoms.
- Home management and medication dosages.
- Associated symptoms: nausea, vomiting, visual changes, dizziness, paresthesia, neck/shoulder pain, back pain, otalgia, abdominal pain, hypersomnia, food cravings, confusion, ataxia, pallor, photophobia, and phonophobia. Changes in gait, personality, vision, mentation, or behavior that do not occur at the same time as the headache are worrisome and merit further evaluation with referral. There are some precursor symptoms and conditions that can indicate a predisposition to migraines. These include cyclic vomiting and abdominal migraine (see Chapter 40) and benign paroxysmal vertigo (BPV; discussed later in the chapter). Alone, they do not warrant extensive or expensive workups unless the diagnosis is unclear.
- Head trauma: If associated with headache, a subdural hematoma or post-concussive syndrome must be considered.
- Psychologic symptoms: Evaluate for the presence of depression, school stressors, or concerns about family functioning. Additional factors to consider include bullying or peer issues at school, “over programming” and family expectations, and meal, hydration, and sleep status.

Distinguishing Features

- Migraine and migraine with aura: These can be differentiated by the presence or absence of aura symptoms (Table 46.3). Characteristics of migraines include nausea, abdominal pain, vomiting, unilateral pain, pulsating pain, relief with sleep, an aura, visual changes such as dark or blind spots, and a history of a family member (usually on the maternal side) with migraine without aura. Dizziness and motion sickness may be described. Infants and toddlers may present with irritability, sleepiness, and pallor. In preadolescents, common migraine symptoms are more likely. Nausea and vomiting might not occur, and the pain can be more frontal. Lethargy and sleep can follow. Visual changes are rare, and the pain quality is variable. Times between headaches are pain free. Pediatric migraine modifiers (variations in characteristics specific to pediatrics) include a duration of 1 to 72 hours and some evidence of pulsating (self-reported with heartbeat).
- Muscle contraction or tension headaches: The pain is dull and bifrontal or occipital, with nausea and vomiting occurring only rarely; there is no prodrome. Characteristic tension headaches feel like a band is squeezing the head. Tension headaches can last for days or weeks but generally do not interfere with activities. In children, it can be difficult to differentiate migraine and tension-type headaches. Psychosocial stress seems to be a major factor in tension and chronic daily headaches in both children and adolescents.
- Medication overuse headaches: Resulting from the overuse of agents such as acetaminophen, NSAIDs, and “migraine medications” (e.g., Excedrin migraine), these headaches are comorbid with primary headache disorders and are increasing in frequency in children. Medication overuse is generally defined as the use of medications more than 15 days per month, manifested by a gradual increase in headache frequency even in the face of increasing analgesic treatment.
- Abdominal migraine: Discussed in Chapter 40, symptoms include midline pain, nausea, and vomiting with minimal or no headache. This rare and somewhat controversial diagnosis can be suggestive of complex partial seizures and may merit further evaluation.
- Secondary headaches (or those headaches that have a pathologic process): Key historical markers are sudden onset of hyper acute or increasing pain severity or accompanying neurologic signs. Box 46.3 describes red flags indicative of a pathologic process requiring immediate referral.

Physical Examination

A complete physical and neurologic examination is needed:

1. • Blood pressure, height and weight, and head circumference (all ages)
2. • Vision screen
3. • Eyes: Palpate for tenderness; check for papilledema, movements
4. • Ears: Patency of canals, normal tympanic membranes
5. • Neck: Palpate muscles; check range of motion for nuchal rigidity

6. • Sinuses (frontal and maxillary)
7. • Teeth and temporomandibular joints (mouth and jaw): Palpate and check range of motion
8. • Thyroid gland
9. • Bones and muscles of skull: Palpate for tenderness; listen for cranial bruits; check range of motion of cervical spine
10. • Extremities: Tandem gait
11. • Nerves: Palpate supraorbital, trochlear, occipital nerves; assess CN IX to CN XII
12. • Reflexes: Pronator drift test (Romberg)

Box 46.3 Red Flags Suggestive of Secondary or Pathologic Headaches

1. • Headache upon awakening from sleep that then fades; increases in frequency and severity over a period of only a few weeks; is persistent and unilateral
2. • First or worst headache
3. • Pain that awakens the child from sleep
4. • Vomiting but not nauseated that may relieve the headache, or intractable vomiting
5. • Visual disturbances, diplopia, edema of the optic disc (papilledema)
6. • Increased pain with straining, sneezing, coughing, defecation, or changes in position
7. • Occipital region and neck pain
8. • Educational, mental, personality, or behavioral alterations; irritability
9. • New onset seizures or facial or extremity numbness
10. • Unsteadiness or dramatic changes in balance, gait abnormalities
11. • Fever with or without nuchal rigidity
12. • Family history of neurologic disorders (e.g., brain tumors, neurofibromatosis, vascular malformations)
13. • Child has a history of a ventriculoperitoneal shunt, meningitis, hydrocephalus, tumor, or prior history of a malignancy.

Box 46.4 Red Flags Suggestive of Intracranial Structural Pathology

Infants

- Full anterior fontanelle
- Open metopic and coronal sutures
- Poor growth
- Impaired upward gaze
- Abnormal head growth
- Shrill cry
- Lethargy
- Vomiting

Children

- Headache described as severe, excruciating of recent onset, unlike any previously experienced headache, no period of normal functioning between episodes, *or* persistent and unilateral
- Papilledema or abnormal eye movements (or one or both eyes suddenly turn in)
- Ataxia, hemiparesis, or abnormal deep tendon reflexes
- Cranial bruits
- Personality changes

Medication Treatment

Nonsteroidal anti-inflammatory drugs are the first-line pharmaceutical for acute treatment. These include acetaminophen, ibuprofen, and naproxen sodium. Zofran can be used for vomiting associated with headaches. Many of the newer medications for migraines (e.g., triptans) have not been adequately tested for safety and efficacy in children and adolescents, with the exception of sumatriptan and zolmitriptan (Table 46.5). Abortive medications should be taken at the onset of the headache and in the prescribed dosage, and should be available at home, school, or work. Importantly, the overuse of analgesics is to be avoided (more than three doses per week), as this can lead to medication overuse headaches (see “Complications”).

Prophylactic therapy is considered when migraines cause a child to miss school regularly and/or when the child suffers severe migraine headaches two to four times a month or tension or migraine three to four times per week with a clear sense of functional disability. Medication classifications to consider include specific β-blockers, antidepressants, anticonvulsants, or calcium channel blockers (Table 46.6). The recent CHAMP (Childhood and Adolescent Migraine Prevention) study (2017) by Powers, Coffey, and Chamberlin found no significant difference in headache frequency between the use of placebo versus preventive headache medications

(amitriptyline, topiramate, and placebo). The study postulated that pharmacotherapy may need to be reconsidered in this population.

Other treatments that are increasing in use include nutraceuticals. These are not regulated by the FDA and are used mainly for headache prevention, not acute treatment. Some nutraceuticals currently under study are magnesium (400 mg qd in adolescent, 200 mg in younger children), Coenzyme Q10, Riboflavin, butterbur, omega-3 fish oil, and Migraleif (riboflavin, puracol, magnesium). Melatonin is sometimes used to help establish good sleep patterns

Musculoskeletal Assessment of Infant:

· **Inspection and Palpation**: Inspection of the skin noting the skin color, presence of swelling or atrophy, erythema, ecchymosis, scars, or unusual pigmentation is essential. The provider should observe the child's posture while sitting, standing, and walking, as well as assess and evaluate the proportion of the upper extremities to the lower extremities. In addition, the provider should palpate skin for differences or inconsistencies in temperature and perfusion and palpate bone and joints to ascertain tenderness, prominence, indentations, and crepitus. Evaluation of symmetry as well as range of motion, muscle size, strength, and tone should be a part of a musculoskeletal examination. Furthermore, when there is a concern regarding sensory or motor deficits, the provider should assess and evaluate the child's spinal nerves and deep tendon reflexes.

· **Range of Motion Examination**: Range of motion is the normal range, flexion, extension, and rotation of a joint. Joint hypermobility is the ability of the joint to move beyond its normal range. Hypermobility of joints generally does not cause problems, although there is a slight increase in dislocation and sprain of the involved joint. The normal values of joint motion are age related and must be kept in mind (e.g., external hip rotation is greatest in early infancy). Passive range of motion, in which the examiner moves the joint, provides information about joint mobility and stability. It can also provide information about the limits of tendons and muscles that are contracted. Active range of motion, in which the child moves the joint, provides information about both muscle and bony structures working together for functional movement. Limited range of motion can be the result of mechanical problems, swelling, muscle spasticity, pain, infection, injury, or arthritis. Note pain, stiffness, limitations or deviations, and rigidity.

· **Gait Examination**: Ambulation typically begins between 8 and 16 months of age. The development of a normal gait is dependent on progressive neurologic maturation. Initially, a child's gait is characterized by a short stride length, a fast cadence, and slow velocity with a wide-based stance. The gait undergoes developmental changes. Walking velocity, step length, and duration of the single-limb stance increase with age, whereas the number of steps taken per minute decreases.

- **A mature gait pattern is well established by 3 years old**. **Normal neurologic maturation results in efficiency and smoothness of gait; and by 7 years old, the gait characteristics are similar to those of an adult**. A normal gait cycle consists of the stance phase, during which the foot is in contact with the ground, and the swing phase, during which the foot is in the air. The stance phase is further divided into three major periods: the initial double-limb support, followed by the single-limb stance, and then another period of double-limb support. Observe the child walking without shoes and minimal covering. The stance and swing phases should be compared in both legs, and the range of motion of each joint should be evaluated. Inspect from the front, side, and back as the child walks normally, on his or her toes, and then on the heels. The gait should be smooth, rhythmic, and efficient. Ankle, knee, and hip movements should be symmetric and full with little side-to-side movement of the trunk. Limping is a disturbance in the normal pattern of gait. Abnormal gait can be antalgic or non-antalgic. An antalgic gait is characterized by a shortening of the single-limb stance phase to prevent pain in the affected leg. Painful or antalgic gaits serve to reduce stress or pain at the affected area. The trunk shifts to the opposite side to keep balance and reduce stress; the stance phase and stride length are shortened as compensatory mechanisms. Causes of a painful gait include infection, trauma, or acquired disorders. A nonantalgic gait may be caused by general weakness, spasticity, muscular disorders, or leg length discrepancies.

For example, a Trendelenburg gait in which the trunk tips over the affected hip indicates hip disease and might or might not be painful because it also involves muscle weakness around the hip joint. Gait disturbances may become more apparent with fatigue.

· **Posture:** To assess posture adequately, the child should be examined undressed to his or her underwear. The examiner needs to look at the child from the front, side, and back.

  ○ Pelvis and hips should be level. Place hands on the iliac crest to test for a pelvic tilt caused by limb length discrepancy.

  ○ Legs should be symmetric in shape and size. The patellae should be straight ahead.

  ○ The feet should point straight ahead, with an imaginary line from the center of the heel through the second toe. There should be an arch (except in babies, in whom a fat pad obscures the arch) and straight heel cords.

  ○ The spine should be straight, and the back should look symmetric, with shoulder and scapula heights and waist angles equal. There should be slight lordotic curves at the cervical and lumbar areas.

· **Hip Examination:**

  ○ **Galeazzi Maneuver or he Galeazzi sign** can **signal conditions that cause leg length discrepancies**. The Galeazzi maneuver includes flexing the hips and knees while the infant or child lies supine, placing the soles of the feet on the table near the buttocks, and then looking at the knee heights for equality (Fig. 38-3, A). **The Galeazzi sign is positive if the knee heights are unequal.** However, it is not reliable in children with dislocatable but not dislocated hips or in children with bilateral dislocation.

  ○ **Barlow (dislocation) Maneuver dislocates an unstable or dislocatable hip posteriorly** (Fig. 38-4, A). The infant is placed in the supine position with knees flexed. The hip is flexed, and the thigh is brought into an adducted position applying downward pressure. With hip instability, the femoral head slips/drops out of the acetabulum or can be gently pushed out of the socket; this is termed a positive Barlow. The dislocation should be palpable as this maneuver is performed. The maneuver needs to be done gently in a non-crying neonate/infant to keep from damaging the femoral head. The hips should be examined one at a time. The hip generally spontaneously relocates after release of the posterior force.

  ○ **Ortolani Maneuver** can be done after the Barlow maneuver or separately (see Fig. 38-4, B). The Ortolani maneuver **reduces a posteriorly dislocated hip**. It is done to reduce a recently dislocated hip and is not done forcefully. The infant is in the supine position with both knees flexed. The thumb is placed near the lesser trochanter, and the pad of the second finger is positioned on the bony prominence of the greater trochanter. The leg is flexed at the hip and then abducted while pushing up with the fingers located over the trochanter posteriorly. The femoral head is lifted anteriorly into the acetabulum. **A palpable clunk as the femoral head is relocated is considered a positive Ortolani sign**. A mild click sound may be audible and is not a positive Ortolani sign. These clicks are common and normal sounds radiating from the knee or ankle. Of note, a positive Ortolani may only be achieved during the first few months of life. Dislocations can occur later in infancy; therefore, the provider must test the hips using other strategies and note limited abduction in older infants until they are walking independently (Fig. 38-5).

  ○ **Klisic Test** provides an **observational sign of hip placement**. The examiner places the tip of the third finger of one hand over the greater trochanter and the index finger of the same hand on the anterior superior iliac spine. An imaginary line is then drawn between the index and third fingers. Normally, the imaginary line points to the umbilicus. If the hip is dislocated, the imaginary line points halfway between the umbilicus and the pubis (i.e., the line points below the umbilicus). **The Klisic sign is another physical assessment marker of hip dislocation** (Fig. 38-6).

- **Trendelenburg Sign test** can be **used to identify conditions that cause weakness in the hip abductors**. The Trendelenburg sign is elicited by having the child stand and then raise one leg off the ground. If the pelvis (iliac crest) drops on the raised leg side, the sign is positive and indicates weak hip abductor muscles on the side that is bearing the weight. Normally the muscles around a stable hip are strong enough to maintain a level pelvis if one leg is raised (see Fig. 38-3, C). With bilateral dislocated hips, a wide-based Trendelenburg limp is noted.

- **Medial (Internal) and Lateral (External) Rotations** The child is placed prone, and the knees are flexed 90 degrees. Medial rotation is measured as the legs are allowed to fall apart as far as possible, using gravity alone or with light pressure. The angle between vertical (0 degree) and the leg position is the medial rotation. It is measured for each leg (Fig. 38-7, A). Asymmetric hip rotation is abnormal. Lateral rotation is measured by allowing the legs to cross while the child is still prone. The angle between vertical and the leg position is measured for each leg (see Fig. 38-7, B). Again, asymmetric hip rotation is abnormal. By 1-year-old, a normal child has approximately 45 degrees of internal and external hip rotation.

· **Back Examination:**

- **Adams Test or the Adams Forward Bend Test looks for asymmetry of the posterior chest wall on forward bending**. This **position allows for evaluation of structural scoliosis**. The child bends at the waist to a position of 90 degrees back flexion with straight legs, ankles together, and arms hanging freely or with palms together (in a diving position) but not touching the toes or floor (Fig. 38-8). The back is inspected for asymmetry of the height of the curves on the two sides or rib hump; the provider inspects the child's back by looking at it from the rear and side positions. The examiner should be seated in front of the child to best visually scan each level of the spine. If a rib hump is present, a scoliometer, if available, can be used to measure the angular tilt of the trunk. A spinal rotation greater than 5 to 7 degrees measured by placing scoliometer at the peak of the curvature indicates a need for further eval.

Cystic Fibrosis

Cystic Fibrosis: is a multi-system genetic disorder manifested by chronic obstructive pulmonary disease (COPD), GI disturbances, and exocrine dysfunction.

· It is an Autosomal recessive genetic disorder involving the mutation of **cystic fibrosis transmembrane conductance regulator (CFTR) protein**, which is expressed in epithelial cells and blood cells.

· CFTR functions in sodium transport through epithelial sodium channel, regulates the adenosine triphosphate (ATP) channels, and is involved in bicarbonate chloride exchange.

· CFTR gene defects cause deficits in ion transport, airway surface liquid depletion, and defective mucociliary clearance.
  - The most common defect (70%)= deletion of phenylalanine in position 508 (D508)
  - Table 32-4 CF: Transmembrane Conductance Regulator Mutation Classes (not sure if we need to know this?)

<u>Clinical manifestations of CF=</u> multisystem, progressive illness with varying levels of severity. (See Table 32-9 for the clinical manifestations seen in children at various ages).

· <u>Pulmonary:</u> CF is a major cause of severe chronic lung disease in children. The lungs of children with CF are normal at birth but become inflamed with chronic airway infection within a short time after birth.
  - Mucus is viscous, and dehydration of the airway secretions occurs, leading to dysfunctional mucociliary transport, airway obstruction, and chronic infections.
  - **Chronic, dry, frequent cough, sputum production and risk of respiratory failure.**
  - Bronchitis, bronchiolitis, bronchiectasis, and pneumonia occur frequently.
  - Bronchospasms resembling acute or chronic asthma may be present.
  - Airways become colonized with *S. aureus, H. influenza, and P. aeruginosa*. *Burkholderia cepacia* is a slower growing organism found in children with CF.
  - **Infection can begin in infancy, however pulmonary disease becomes more progressive and leads to cor pulmonale, respiratory failure, and death by adulthood (average age 41).**

- Other respiratory problems associated with CF= ARS, nasal polyps, allergic bronchopulmonary aspergillosis (ABPA), which starts by childhood and continues into adulthood.
- Digital clubbing is common.

· GI tract & nutrition: during infancy, meconium ileus, pancreatic insufficiency, and rectal prolapse can be seen in those with CF.
- **Meconium ileus** in 15% newborns, but can also develop in older adults causing GI obstruction
- **Failure to thrive** in 85% of affected children d/t pancreatic enzyme insufficiency.
- Edema with hypoproteinemia
- **Thick fat-laden stools (steatorrhea), poor muscle mass, and delayed maturation**.
- During childhood, intussusception, hepatic steatosis, biliary fibrosis, & rectal prolapse can occur.
- Other risk factors including 15% development of cirrhosis and portal hypertension and adenocarcinoma of the digestive tract can occur.
- Other GI problems: volvulus, duodenal inflammation, GERD, bile reflux, fibrosing colonopathy, and poor fat absorption leading to vitamin A, K, E, and D deficiencies with resulting anemia, neuropathy, night blindness, osteoporosis, and bleeding disorders.
- Distal intestinal obstructive syndrome (DIOS) occurs when viscous fecal matter causes blockage in the distal intestine and presents with abdominal pain and distention with pain. This can occur as a result of poor fat absorption, pancreatic insufficiency, and dehydration.

· Hepatobiliary tract: biliary cirrhosis occurs in 2-3% of children with CF and is characterized by jaundice, ascites, hematemesis from esophageal varices, and splenomegaly.
- Hepatic steatosis is also a known complication of CF.
- Adolescents may experience biliary colic and cholelithiasis (pain and inflammation of gallbladder)

· Endocrine: recurrent acute pancreatitis is not uncommon.
- CF related DM (CFRD) with relative insulin deficiency develops as the patient ages d/t autolysis of the pancreas as the pancreas body becomes fatty.
- CF patients need annual blood glucose screening, as up to 30% develop CFRD by adulthood.

· Musculoskeletal: Vitamin D deficiency may result in osteoporosis when bone reabsorption exceeds bone formation.

· Reproductive: Affected children have delayed sexual development.
- The vas deferens is nonfunctional and atrophied due to CFTR dysfunction, leading to azoospermia and male sterility.
- The incidence of inguinal hernia, hydrocele, and undescended testes is also high.
- Females experience secondary amenorrhea, cervicitis, and decreased fertility.
- A pregnancy is usually carried to term if pulmonary function is not severely compromised

· Sweat glands: Excessive salt loss can lead to hypochloremic alkalosis, especially in warm weather or after gastroenteritis.
- Children with CF often taste salty because of elevated amounts of sodium chloride (NaCl) lost in endogenous sweat.
- Dehydration and heat exhaustion are concerns.

Diagnosis= The diagnosis of CF is made on the basis of clinical features and laboratory findings.

· **The first and gold standard for diagnosis is the pilocarpine iontophoresis sweat test**.

· The child must have one of more of the clinical features of CF before ordering this test, which include chronic sinopulmonary disease, GI and nutritional abnormalities, salt loss syndrome, chronic metabolic alkalosis, or male urogenital abnormalities, resulting in obstructive azoospermia.

· The results of the sweat test are determined differently depending on age. Sweat tests should be done at a laboratory that regularly deals with children and routinely does these tests. Sodium chloride concentration goes up with age but a concentration greater than 60 mmol/L is still a diagnostic level. A result of greater than 60 mEq/L of chloride on two specimens is in the diagnostic range for CF. Children with hypoproteinemia may elicit false-negative sweat test results.

- The diagnosis of CF can be made in patients with clinical features of the disease under the following guidelines:
    - The concentration of sweat chloride is greater than 60 mmol/L.
    - The concentration of sweat chloride is in the intermediate range of 30 to 59 mmol/L for infants younger than 6 months old or in the range of 40 to 59 mmol/L for older individuals.
    - The child has two disease-causing CFTR mutations

- The second test is genetic analysis for the CFTR mutation. Commercial labs evaluate for the common mutations of the CFTR gene; however, complete sequencing of the gene is also available.

- A limited number of CF centers also test for nasal potential difference measurement.

- Newborn screening is now done in all 50 states by state labs that may measure immunoreactive trypsinogen (IRT) in the newborn's blood. The lab will either perform two IRT measurements (IRT/IRT) or test for CFTR mutation if the IRT is elevated (IRT/CFTR). If the IRT is elevated or the child has one CFTR mutation, the infant's primary provider is notified and the child is referred for sweat testing. There are false-negative screens, so if an infant has symptoms and signs of CF, a sweat test should be performed.

- Other diagnostic testing is indicated depending on secondary complications of CF. Glycosylated hemoglobin levels may be elevated in older children because of impaired pancreatic functioning. Pulmonary function tests are used to follow the clinical course.

Management= Children with CF have complicated treatment regimens and should be monitored by a multidisciplinary team at a CF-accredited center. The major aim of pulmonary disease treatment is to optimize lung functioning, prevent disease progression, and avoid complications. The treatment of CF-related lung disease requires control of airway infections, clearance of airway secretions, and decreasing inflammation in the lung. Pulmonary, nutritional, physical, and pharmacologic (antibiotic and anti-inflammatory) therapy and psychological counseling must be individualized for each child at each stage of the illness.

- Pulmonary
    - To promote airway clearance, inhaled dornase alfa (recombinant human deoxyribonuclease) selectively cleaves the DNA and reduces the mucus viscosity. Hypertonic saline works by drawing water into secretions and is used to thin secretions to allow removal of them. The use of postural drainage, active cycle of breathing, autogenic drainage, percussion, positive expiratory pressure, exercise, and high-frequency chest wall oscillation are done twice a day to facilitate secretion removal
    - The use of ivacaftor (Kalydeco; a medication that is a CFTR potentiator) is limited to patients who carry at least one G551D mutation
    - **To reduce chronic airway inflammation, high-dose ibuprofen and oral azithromycin dosed three times a week are used**. Patients must be screened for atypical mycobacterial infection before starting long-term azithromycin. Although high-dose ibuprofen decreases neutrophil migration in children from 6 to 17 years old, the therapy is not widely used due to risk of GI bleeding and frequent drug blood level measurements. Hemoptysis can be scant (less than 5 mL), moderate (5 to 240 mL), or massive (greater than 240 mL) and is associated with advancing lung disease as well as vitamin K deficiency. It results from the hypertrophy and proliferation of the bronchial arteries rupturing into the airways as a result of the disease process. The management includes antibiotic therapy, cessation of the anti-inflammatory drugs and limiting therapies for airway clearance

- Pneumothorax presents as acute onset of chest pain and dyspnea and is confirmed by chest x-ray. Smaller pneumothoraces are managed by observation and discontinuation of positive pressure. Surgical or chemical pleurodesis is used in recurrent large pneumothoraces.

- Lung transplantation is a viable therapy for selected patients who have terminal lung disease.

· Gastrointestinal

- Patients with CF have an 85% rate of pancreatic insufficiency. This leads to bulky, malodorous stools resulting in failure to thrive. Replacement with enzymes in the dosage ranges of 2000 to 2500 units/kg of lipase to a maximum of 10,000 units/kg/day. Higher dosing of lipase can lead to fibrosing colonopathy in a small number of patients.

- Fat malabsorption causes deficiency of vitamins A, D, E, and K; therefore, replacement must be started along with serum monitoring of the levels annually. Vitamin D deficiency can occur, resulting in osteopenia, osteoporosis, or rickets.

- Cystic fibrosis liver disease (CFLD) is diagnosed when there are at least two of the following: (1) hepatomegaly and/or splenomegaly, (2) liver function tests abnormalities on three tests in a 12-month period, (3) portal hypertension or abnormal liver echotexture on ultrasound, and (4) cirrhosis on liver tissue biopsy. Treatment with ursodeoxycholic acid is recommended. DIOS is managed using osmotic laxatives to promote lower bowel clearance. The use of sodium meglumine diatrizoate (Gastrografin) enemas can be used for a near complete obstruction by an experienced radiologist.

· Endocrine disorder

- CFRD is the result of the fatty infiltration and destruction off the islet cells due to the thick viscous secretions in the pancreas. Microvascular complication of diabetes including renal disease, retinopathy, and neuropathy are associated with hyperglycemia. Ketoacidosis is rare. At age 10, an oral glucose tolerance test is done annually to screen for CFRD. Hemoglobin A1C is not recommended because it underestimates overall glycemic control.

    - Insulin is used to treat CFRD.

Traumatic Brain Injury

- Involves tissue damage to the brain and its structures.
    - Open head trauma: produces more focal injuries
    - Closed head trauma: multifocal and diffuse damage
- Common causes
    - falls
    - sports-related injuries
    - motor vehicle accidents
    - violence and assaults
    - being struck by or against objects
- Boys are 2x more frequently than girls

Common Causes per age

- Infant - toddler: falls or non accidental trauma
- 0-4 yrs and 15 -25 yrs: HIGHEST RISK OF TBI
    - Young children: falls, pedestrian/bicycle accidents
    - Adolescents: motor vehicle accident, sport-related injuries and assaults.

Concussion (MILD TBI)

- History CDC Acute Concussion Evaluation (ACE) Tool.

- History of how injury occurred; if injury involved a fall, the height from which the child fell. Specifically, providers should ascertain injury cause, body part affected, forces, and circumstances.
- Loss of or alteration in consciousness or memory, confusion, irritability, inappropriate behavior, repetitive questioning
- Presence of vomiting and frequency
- Presence of headache, description of the headache pain
- Presence of blurred vision, diplopia, or other vision problem
- Numbness or loss of sensation, loss of balance, or difficulty walking
- Specific symptoms occurring at the time of injury and interval changes

TABLE 46.8

Pediatric Glasgow Coma Scale

	>1 Year	<1 Year		Score
Eye opening	Spontaneously	Spontaneously		4
	To verbal command	To shout		3
	To pain	To pain		2
	No response	No response		1
Motor response	Obeys	Spontaneous		6
	Localizes pain	Localizes pain		5
	Flexion-withdrawal	Flexion-withdrawal		4
	Flexion-abnormal (decorticate rigidity)	Flexion-abnormal (decorticate rigidity)		3
	Extension (decerebrate rigidity)	Extension (decerebrate rigidity)		2
	No response	No response		1

	>5 Years	2-5 Years	0-23 months	
Verbal response	Oriented	Appropriate words/phrases	Smiles/coos appropriately	5
	Disoriented/confused	Inappropriate words	Cries and is consolable	4
	Inappropriate words	Persistent cries and screams	Persistent inappropriate crying and/or screaming	3
	Incomprehensible sounds	Grunts	Grunts, agitated, and restless	2
	No response	No response	No response	1

TABLE 46.10

Classification of Head Injuries Based on Key Characteristics

Classification	Glasgow Coma Scale [a]	Neurologic Focal Deficit [b]	Loss of Consciousness	Other Neurologic Findings
Mild	13-15	No	No or brief loss (<30 min)	May have linear skull fractures
Moderate	9-12	Focal signs	Variable loss	May have depressed skull fracture or intracranial hematoma
Severe	≤8	Focal signs	Prolonged loss	Often have depressed skull fractures and intracranial hematoma

- Dx
  - Head trauma with a GCS score of 9-12 or 3-8 should have a Cranial CT scan

- perform cranial CT if the follow has occurred
  - • Penetrating trauma or depressed skull fracture or signs of basilar injury
  - • Altered level of consciousness (excessive irritability or lethargy)
  - • Loss of consciousness (exceeding 1 minute)
  - • Amnesia about the injury
  - • Focal neurologic signs or deficit
  - • Persistent vomiting or seizures
  - • History of coagulopathy
- CT preferred in emergency
- Skull fractures are better seen on skull radiography
- CT is order 3-8 days past injury order with and without contrast to pick up extravasated blood

Management

- Child With Minor Closed Head Injury and NO LOC
  - Observation in clinic/ED under the care of a competent caregiver to recognize abnormalities and seek assistance
- Child With Minor Closed Head Injury and Brief LOC
  - Observation in clinic/ED under the care of a competent caregiver
  - CT scan is accepted
  - if caregiver is unable to monitor at home hospita;ization is indicated.
- Child With Moderate Head Injury or Worrisome Symptoms
  - Admitted for overnight observations in hospital if LOC is normal
  - Moderate injury GCS 9 - 12 may require prolonged observation in ED
  - Severe injury GCS less than 8 or coma: immediate hospitalization and consult with neurologist/ ciritical care team
- Cognitive and physical brain rest are the essential components

Box 46.5 Head Injury Education Key Points for Parents

Parents or caregivers should be given specific instructions regarding:

- Waking the child every 2-4 h for the first 24 h after injury, ensuring the child wakes easily and is able to stay awake for a few minutes.
- Making sure child is moving his or her arms and legs normally.
- Giving only acetaminophen, if needed, for headache or relief of soft tissue pain.

Contact the health care provider or take the child to an ED if the following symptoms are observed:

- Increased drowsiness, sleepiness, inability to wake up, unconsciousness
- Vomiting more than twice
- Neck pain
- Watery or bloody drainage from ear or nose
- Seizures or fainting
- Unusual irritability, personality change, confusion, or any unusual behavior
- Headache that gets worse or lasts more than a day
- Unequal pupils, blurred vision, abnormal or changing hearing, or speech
- Gait abnormality (e.g., clumsiness or stumbling), weakness of any muscle of arms, legs, or face

HOSPITALIZATION if the following occur:

- Changing vital signs
- Seizures, altered mental status or slurred speech
- Prolonged unconsciousness (>30 seconds)
- Persisting memory deficit or focal neurologic signs
- Depressed or basilar skull fractures
- Persistent headache (particularly with stiff neck)
- Recurrent vomiting or unexplained fever
- Unexplained injury (suspected child abuse)
- CT scan or MRI findings that are worrisome

Complications

- - Posttrauma Sequelae (can happen up to months after trauma)
  - • Headache, vertigo or dizziness

- • Difficulty concentrating or loss of memory
- • Poor school performance and neurobehavioral problems
- • Depression, fatigue

MILD INJURY

- no resulting physical deficit
- weeks to months after may result in cognitive deficits

MORE SEVERE INJURY

- cognitive function changes generally will not improve after 12 months
- speech and motor difficulties may continue to improve for up to several years

- Post-Concussion Syndrome
  - Adolescents S/Sx: headache, dizziness, irritability, and impaired ability to concentrate
  - Younger Children S/Sx: aggression, disobedience, behavioral regression, inattention, and anxiety sleep related issues

Prevention

- Use seat belts in motor vehicles.
- Protect children from falls. Discourage the purchase of residential trampolines.
- Wear helmets for sports participation. The proper fitting of helmets is important.

Sickle cell anemia 753-755

Sickle Cell Anemia & Trait: describes a group of complex, chronic disorders characterized by hemolysis, unpredictable acute complications that may become life threatening, and the possible development of chronic organ damage. Children who have homozygous inheritance have SCA or disease (Hgb SS). Their bodies do not form the normal Hgb A molecule, but rather synthesize hemoglobin S (Hgb S), which carries the amino acid valine instead of glutamic acid. Because of this change, Hgb S tends to polymerize or come out of solution at low PaO2, low pH, low temperature, and low osmolality. This process collapses the RBC, giving it a "sickled" shape, and produces a chronic hemolytic anemia. **The new shape is rigid and clogs small blood vessels, producing ischemia, pain, and other vaso-occlusive problems. Sickle cell disease has an autosomal recessive inheritance pattern**. It is found **most often in people of African descent**. SCA exceeds most other hematologic disorders only alpha-thalassemia is more common. Routine neonatal screening identifies most infants with sickle cell disease born in the United States, because it is mandated in all states and the District of Columbia. It is still important to do a careful family medical history because many adults do not realize they are carriers.

Clinical Findings= The symptoms of sickle cell disease are multisystem, necessitating vigilant care to minimize occurrence of crises and complications. Common symptoms include:

· Fatigue and anemia
· Pain crises
· Dactylitis (swelling and inflammation of the hands and/or feet) and arthritis
· Bacterial infections
· Lung and heart injury
· Leg ulcers
· Priapism
· Splenic sequestration (sudden pooling of blood in the spleen) and liver congestion
· Aseptic necrosis and bone infarcts (death of portions of bone)
· Eye damage
· Abdominal pain

· Children with sickle cell trait who are heterozygous (Hgb A + Hgb S) for the gene essentially have a benign clinical course. Their RBCs contain only 30% to 40% Hgb S, and sickling does not occur under most conditions. Extreme exercise, typically to exhaustion, dehydration, and relative hypoxia (altitude) are major confounding factors.

Physical Examination= **SCA symptoms typically begin to emerge in the second 6 months of life** as the amount of Hgb S increases and Hgb F declines. Subsequently, **painful, vaso-occlusive crises** occur.

- D/t the multisystem nature of complications these children need prompt, detailed evaluation and intervention.
- After 5 years old, splenomegaly usually disappears because of auto infarction of the organ.
- Rates of height and weight gain usually slow after 7 years old, and puberty may be delayed 3 to 4 years.

Diagnostic Studies=The following laboratory results are seen in sickle cell disease:

- Hct of 20% to 29%
- Hgb 6 to 10 g/dL (severe)
- Reticulocyte count elevated: 5% to 15%
- Normal to increased WBC and platelet count
- MCV greater than 80 fL; mean corpuscular hemoglobin concentration (MCHC) greater than 37 mg/dL
- Hgb electrophoresis (after infancy), isoelectric focusing or high performance of liquid chromatography showing a predominance of Hgb S and no Hgb A.
- Morphology: Irreversibly sickled cells or chronic elliptocytes, Howell-Jolly bodies, nucleated RBCs

Hgb electrophoresis results in a newborn with sickle cell trait will be Hgb FAS, and Hgb FS for a child with either SCA or sickle beta-zero thalassemia (SBO). Normal results of Hgb electrophoresis are Hgb FA.

Management= of the child with SCA is complicated and should be done in consultation with a pediatric hematologist. Children with sickle cell disease still need regular primary care services and coordination of consultative services and information. Growth is closely monitored, immunizations need to be done on time, parents require support, and communication with specialty services should be coordinated, such as an annual ophthalmologic examination by a retinal specialist. Care is comprehensive, spanning normal well-child issues through acute crises and hospitalization.

Some of the key aspects of care for the child with SCA are as follows:

- **Hydration, illness prevention, and pain management are fundamental aspects of disease management**. NSAIDs or acetaminophen may be adequate for mild to moderate pain, but narcotics should be used when these are not adequate for management.

- **CBC and reticulocyte count are monitored every few months.**

- **All the usual immunizations of childhood are to be administered on time** including 13-valent pneumococcal conjugate (four doses at appropriate intervals) and 23-valent pneumococcal polysaccharide vaccines (first does at or after 24 months of age) with a second dose of PPSV23 given 3 years after the first dose. The conjugate Hib and meningococcus vaccine (HibMenCY) is recommended for infants with sickle cell disease at 2, 4, 6, and 12 to 15 months of age and booster doses of MCV4 every 5 years thereafter. An annual flu vaccination is essential.

- **Invasive bacterial infection is the leading cause of death in young children with SCA. Penicillin V prophylaxis (125 mg orally, twice daily) is initiated by 2 months old.** At 3 years old, increase the dose to 250 mg orally twice a day, and continue at least until the fifth birthday or until the child has received two doses of PPSV23.

- **Folic acid supplementation at 1 mg/day is typically given to adults to prevent folate deficiency due to hemolysis.** It is not standard therapy for children unless a folic acid deficiency is suspected and should be individualized for each patient.

- **Aggressive treatment of infections and maintenance of hydration and body temperature are used to prevent hypoxia and acidosis**; volume replacement may be necessary to prevent circulatory collapse.

- **Treatment of coexisting medical problems** associated with lower oxygen saturations, such as asthma and obstructive sleep apnea.

- **In children with severe SCA, hydroxyurea is used to reduce the number of painful crises and incidences of acute chest syndrome (a leading cause of death in adolescents with SCA).** It is a preventive medication and not effective during the acute crisis. Hydroxyurea use is associated with a lower need of blood transfusions and fewer hospital visits by

reducing the frequency and severity of painful events and acute chest syndrome episodes. It increases Hgb F levels within cells, which decreases Hgb S levels, increases RBC water content, and alters adhesion of RBCs to endothelium. There is some early evidence suggesting it helps improve growth and preserves organ function.

· Annual stroke prevention screening of major intracranial vessels with transcranial Doppler ultrasound evaluation is planned for 2-to 16-year-old children or as long as their bone windows allow meaningful evaluation. A reading of greater than 200 cm/sec time-averaged mean maximal velocity indicates high risk for stroke and an indication to start transfusion programs to maintain Hgb S levels less than 30%

Emergency admission or referral is necessary in the presence of the following:

• Fever (to rule out sepsis) greater than 101° F (38.3° C)
• Pneumonia, chest pain, or other pulmonary symptoms (acute chest syndrome)
• Sequestration crisis (splenomegaly with decreased Hgb or Hct)
• Aplastic crisis (decreased Hct and reticulocyte count)
• Severe painful crisis
• Unusual headache, visual disturbances
• Priapism

Complications= Because of functional asplenia (absence of spleen), the greatest concern is febrile illness indicating infection and possible sepsis.

· Increased risk of pneumococcal sepsis in children younger than 5 yrs. old therefore all complaints of fever, poor feeding, lethargy, & irritability should be clinically evaluated.
· The consequences of hemolysis may include chronic anemia, jaundice, cholelithiasis, & delayed growth and sexual maturation.
· Vaso-occlusion & tissue ischemia may result in acute & chronic injury to virtually every organ system, with stroke being the major concern.

Patient and Family Education= The parents of children with SCA need a great deal of support in raising a child with a genetically transmitted chronic disease. Initial education includes the genetics and pathophysiology of the disease and the importance of regular health maintenance visits. The need for early evaluation and treatment of febrile illness, acute splenic sequestration, aplastic crisis, and acute chest syndrome. Parents can be taught to palpate their child's spleen. Any downward displacement or enlargement of the spleen below the left costal margin should be evaluated by a health care professional and blood counts monitored for increasing anemia. As the child grows, the family should be educated about other potential clinical complications, such as stroke, enuresis, priapism, cholelithiasis, delayed puberty, retinopathy, avascular necrosis of the hip and shoulder, and leg ulcers.

Preventive care measures also include the following:
• Timely admin. of routine immunizations, including pneumococcal, meningococcal vaccines & yearly influenza vaccine
• Prophylactic antibiotics
• Genetic counseling for those with sickle cell trait
• Support groups
• Educating adolescents with the trait about their status and the risk of disease transmission
• Hematopoietic stem cell transplant (the only intervention that can cure sickle cell disease with strict inclusion criteria identified for transplant eligibility)
• Gene therapy (under investigation)

Microcytic Anemia: Iron Deficiency Anemia

Iron deficiency anemia (IDA) is the most common nutritional disorder and hematologic condition in the world. Some 9% of adolescent girls develop iron deficiency; 2% to 3% of such cases were due to rapid growth, heavy menses, and nutritionally inadequate diets (Abrams, 2017). The incidence of IDA among children in the United States has been declining slightly during the past four decades, although the prevalence remains high among children living at or below poverty level. Other risk factors include childhood obesity and a history of prematurity or low birth weight (Powers & Mahoney, 2019). IDA correlates with diets low in iron, as occurs with an overuse of goat's milk, cow's milk, or other milk substitutes. Deficient iron intake is also associated with prolonged bottle feeding.

Dietary iron is primarily absorbed in the duodenum. Malabsorption of iron occurs in diseases that affect this segment of the intestine, such as celiac disease, Crohn disease, giardiasis, or resection of the proximal small intestine. Disorders causing rapid transit, GI blood loss due to inflammatory bowel disease, cow's milk–induced colitis, or chronic use of aspirin or nonsteroidal anti-inflammatory drugs (NSAIDs) can also deplete iron stores and contribute to iron deficiency (Mahoney, 2017).

The minimal laboratory screening for iron deficiency is the Hgb level. Often the simplest and most cost-effective measurement is a CBC, which includes the Hgb, Hct, MCV, and RDW. A serum ferritin level is helpful because it reflects current stores of iron, but it must be interpreted carefully because ferritin is an acute-phase reactant and may be increased with inflammatory conditions (Powers & Mahoney, 2019). The American Academy of Pediatrics (AAP) Committee on Nutrition recommends universal Hgb screening for anemia at 12 months of age. This screening should include an assessment of risk factors for iron deficiency and IDA (Box 39.3).

Screening Hgb can be performed on children younger than 1 year of age when risk factors warrant it. When IDA screening or any other routine health screening recommendation is being implemented, it is important to keep in mind that screening is not just a one-time test; the effectiveness of treatment must be determined through follow-up testing. Thus, after the routine 12-month Hgb testing, risk assessment for anemia should be performed at all preventive pediatric health care visits, with follow-up blood testing if positive. If children are at risk for IDA, a repeat Hgb should be performed as often as indicated.

Effects of Iron Deficiency

Many studies demonstrate that iron-deficient states in the first few years of life are associated with cognitive deficits that extend well into adulthood, although direct causality is difficult to prove. Children's brains reach 95% of adult size by 2 years of age; therefore any nutritional deficit, including iron, can cause lasting damage. Lead poisoning (plumbism) is often a comorbid condition to IDA. A child at risk for lead exposure should be screened at 9 to 12 months of age and again at 24 months. An estimated 99% of lead-poisoned children are identified through screening procedures rather than clinical recognition. Additional screening should be done at 15 and 30 months of age based on at-risk status. Local health departments determine the prevalence of lead poisoning in their area and issue guidelines related to blood lead screenings for targeted children in their catchment areas (Simon et al, 2014). If the initial blood lead level is 5 mcg/dL or greater on a single visit, it is a concern for public health purposes. The United States has made great strides in reducing lead toxicity through the elimination of tetraethyl leaded gasoline, banning lead-containing solder to seal food and beverage cans, and a federal rule to limit the amount of lead allowed in paint intended for household use.

Clinical Findings

Conduct a detailed history and complete physical examination, keeping in mind children with moderate to severe anemia can be asymptomatic. Some key elements to remember are as follows:

1. • Infants and toddlers may be irritable and restless, but this only occurs with Hgb less than 8 g/dL and is often noticed in retrospect—after treatment.
2. • Pica (appetite for nonfood items such as paper, dirt, and clay) and pagophagia (the desire to ingest ice) may be present.
3. • Anorexia has been reported with Hgb levels less than 8 g/dL.
4. • Developmental delays (mental and motor areas) and social-emotional behavioral disturbances have been reported in infants and young children; adolescents may experience cognitive impairment

Diagnostic studies

IDA is likely if there is a low Hgb level for age (in the range of 8 to 11 g/dL), a history of low iron intake, and no concern about other possible causes for the anemia or the possibility of another hemoglobinopathy. If the age of the child and the dietary patterns are consistent with IDA and there is microcytic anemia (Hgb >9 g/dL), many clinicians begin a trial of iron supplementation for 4 to 6 weeks without further diagnostic testing and then follow the child's Hgb and reticulocyte counts. The RDW is the earliest marker of iron deficiency. Serum ferritin is low with iron deficiency.
Mild to moderate IDA is characterized by Hgb levels of 7 to 10 g/dL. Levels less than 4 g/dL necessitate consultation with a hematologist; and levels of 7 or less should be carefully evaluated as to whether the child needs referral to hematology. If treatment with oral iron supplements is effective, follow-up Hgb in 1 month should reveal a minimal improvement of 1 to 2 g/dL; the reticulocyte count will increase to greater than 3% in 48 to 96 hours

Differentiating Features of Microcytic Anemias

Test	Iron Deficiency Anemia	Thalassemia Minor [a]	Anemia of Inflammation [b]
Serum iron	Low	Normal	Low
Serum iron-binding capacity	High	Normal	Low or normal
Serum ferritin	Low	Normal or high	Normal or high
Marrow iron stores	Low or absent	Normal or high	Normal or high
Marrow sideroblasts	Decreased or absent	Normal or increased	Normal or increased
Free erythrocyte protoporphyrin	High	Normal or slightly increased	High
Hemoglobin A_2 or F	Normal	High β-thalassemia; normal α-thalassemia	Normal
Red blood cell distribution width [c]	High	Normal	Normal/↑

BOX 39.3 Risk Factors for Iron Deficiency Anemia

1. • Premature and/or low-birth-weight infants
2. • Exclusive breastfeeding beyond 6 months without iron supplementation
3. • Early weaning to whole cow's milk (before 1 year)
4. • Excessive (>25 oz) intake of cow's, goat's, or soy milk in children 1-5 years of age
5. • Infants fed formula not fortified with iron
6. • Children with feeding problems
7. • Children with special health care needs/restricted diets
8. • Alternative diets (e.g., vegan)
9. • Low socioeconomic status, food insecurity
10. • Exposure to lead
11. • Eating disorders, including obesity
12. • Hookworm
13. • Adolescent female, excessive menstrual bleeding

The typical profile for IDA is

1. • Microcytic, hypochromic RBCs on CBC
2. • Low or normal MCV; low to normal RBC number
3. • High RDW (>14%), low ferritin, high TIBC
4. • Mentzer index greater than 13 (IDA more likely)

There is a high comorbidity between IDA and lead poisoning (Pb >5 mcg/dL) because lead molecules block iron from binding to protoporphyrin by inhibiting essential mitochondrial membrane function and interfering with enzymes.

Management of IDA

For infants and children, the treatment for IDA consists of iron supplementation, typically as ferrous sulfate (3 to 6 mg/kg/day of elemental iron in two to three divided doses or 3 mg/kg/day in one or two divided doses for mild or moderate IDA) (Sills, 2016). In adolescents the daily maximum of elemental iron is 200 mg. The child's reticulocyte count should be reassessed within 1 week, as reticulocytosis may be seen within a few days of treatment. Hgb should return to a normal level within 4 to 6 weeks. If a therapeutic response is observed (Hgb increase of >1 g/dL or >3% increase in Hct), iron supplementation should continue to normalize Hgb, then continue for 2 to 3 months to replete iron stores. Serum ferritin levels should be rechecked 6 months after iron supplements are stopped to determine resolution of the anemia and adequacy of iron stores

Iron Requirements/Anemia of prematurity

Full-term infants accumulate almost 80% of their iron stores during the last trimester of pregnancy. Delayed clamping of the umbilical cord also ensures that newborns receive the maximal transfusion to begin life. Along with prematurity, maternal anemia, maternal hypertension with intrauterine growth retardation, and/or gestational diabetes can result in less iron transferred to the fetus. In the case of preterm births, the decreased iron stores are depleted rapidly and the normal physiologic nadir occurs earlier, aggravated by a smaller total blood volume at birth and poor GI absorption. The use of erythropoietin to prevent and treat anemia of prematurity also increases the risk of iron deficiency (Powers & Mahoney, 2019). In general, preterm (<37 weeks' gestation) infants require an oral supplement of elemental iron at 2 mg/kg per day from 2 weeks through 12 months of age (Powers & Mahoney, 2019).

Breast milk provides an average iron content of 1.0 to 0.3 mg/L with a high bioavailability. Because there is large variation in the iron content in human milk, the content of maternal milk may not always provide for the needs of the growing infant. It is recommended that the exclusively breastfed term infant receive elemental iron supplementation of 1 mg/kg/day (15 mg maximum) beginning at 4 months of age and continuing until iron-containing complementary foods are introduced and taken in adequate quantities (Powers & Mahoney, 2019). This same iron supplementation recommendation holds for the partially breastfed infants who receive more than half of their daily feeding as human milk. Infants fed with standard infant formulas receive a sufficient iron intake of 12 mg/dL.

The intake of iron should increase dramatically to 11 mg/day between 7 and 12 months of age based on cells sloughing and the demands of increasing body mass (Hernell et al., 2015). As the rate of growth decreases in early childhood, so does the nutritional requirement of iron, down to 7 mg/day between 1 and 3 years of age. Iron deficiency becomes more prevalent during these ages as well, reaching 6.6% to 15.2% depending on ethnicity and socioeconomic status, although the occurrence of IDA is 0.9% to 4.4%. Despite these seemingly low levels of incidence, IDA accounts for almost half of the anemias of early childhood. Liquid supplementation for this age group is appropriate until 36 months of age. Chewable multivitamins can be used for children older than 3 years of age, but the supplement formulation must be evaluated for adequate replacement.

Thalassemias

Thalassemias are categorized into two types: alpha (α) and beta (β), based on the affected chain. In the carrier of *α thalassemia* (minor/trait), only one α chain is present, which enables the production of adequate amounts of Hgb with few or no symptoms; however in α-thalassemia disease (major), there are no α chains, so the β-globulin subunits cluster into groups of four. These β tetramers are incapable of carrying oxygen, and the affected fetuses die in utero (hydrops fetalis). In the carrier of *β thalassemia* (minor/trait), there are sufficient β chains to bind with the abundant α chains and create functional Hgb molecules; however, a resultant asymptomatic mild microcytic anemia is present (see Box 39.3).

In β thalassemia major, the α chains do not bind with each other but rather degrade in the absence of β chains; ongoing degradation results in severe anemia and the need for ongoing transfusions. The possibility of thalassemia increases if the onset of anemia and symptoms occurs prior to 3 to 6 months of age and there is a prior family history of thalassemia or a family history of anemia, miscarriage, or fetal demise or jaundice, gallstones, anemia, or splenomegaly.

Categorizing the thalassemias is less straightforward than with many anemias because although the heterozygous disease is hypochromic and microcytic, the homozygous diseases are also hemolytic. There is anemia and increased erythropoiesis. The erythropoiesis results in bone marrow expansion, but the pathogenesis of this is not fully understood. Focal osteomalacia and delayed bone maturation are at least partially explained by suboptimal blood transfusions and iron overload. Furthermore, the marrow expansion results in frontal bossing and hyperplasia of the maxillary bones, leading to typical facies (DeBaun et al., 2016).

α Thalassemias

The α thalassemias are composed of several variant Hgbs that are responsible for the various presentations. Current nomenclature often refers to the subtypes by including an indication of the number of gene deletions of α globin; the severity of symptoms increases with more deletions. Three gene deletions result in severe, even fatal, manifestations of disease. Two gene deletions present with hypochromia; the absence of gene deletions causes mild anemia and often erythrocytosis. A single globin gene deletion is clinically insignificant (DeBaun et al., 2016).

There are two manifestations of the disease expression of α thalassemia. The homozygous Hgb type, Hgb Bart (or Hgb H) with four γ-chains, results in *hydrops fetalis* and is incompatible with life because of severe anemia. Homozygous Hgb H disease can also present as a microcytic, hypochromic anemia that most often manifests as a hemolytic anemia, hepatosplenomegaly, and mild jaundice and sometimes includes thalassemia-like bone changes. During times of physiologic stress, the child may require RBC transfusion.

There are two different carrier states of α thalassemia. In α-thalassemia trait, the child exhibits microcytosis and hypochromia but has normal percentages of Hgb $A_2$and Hgb F. The other trait state is referred to as a *silent carrier state* but can have either a silent hematologic phenotype or present with microcytic hypochromia and some erythropoiesis.

Management

Hgb H disease exacerbations may necessitate occasional transfusion during hemolytic or aplastic crises. No treatment is indicated for the carrier trait expressions of disease, and the microcytosis seen in these expressions require that serum iron studies should be done before starting any iron supplements. Those carrying the α-thalassemia trait alleles require careful genetic counseling, because there are complex patterns of inheritance that could affect the phenotype of such individuals' offspring (Benz, 2017).

β Thalassemia Minor/Minima

β thalassemia minor and minima disease, also known as β thalassemia *trait,* is associated with a mild hypochromic, microcytic anemia in which Hgb levels are 2 to 3 g/dL below normal and the MCV averages 65 fL. These children must be monitored for iron accumulation but are otherwise asymptomatic. The disease may be confused with iron deficiency or lead poisoning and can be differentiated by measuring serum iron or lead levels, transferrin saturation, or serum ferritin levels. It is particularly important to diagnose this condition correctly in order to avoid unnecessary administration of iron supplements, which do not improve the Hgb level and could result in iron overload. The primary diagnostic feature is increased Hgb A_2 (>3.5%) on electrophoresis (Benz, 2017).

BOX 39.5 Mentzer Index

Mentzer index = MCV/RBC. If the ratio is less than 13, the anemia is more likely to be thalassemia trait; if the ratio is greater than 13, the anemia is more likely to be due to iron deficiency.

Clinical Findings

Clinically most individuals with thalassemia trait are asymptomatic, although mild pallor and splenomegaly may be found. A Hgb of 9.5 to 11 g/dL, Hct less than 30%, and a MCV of less than 75 fL are commonly seen in thalassemia minor. The MCV/RBC count per milliliter is less than 13 (Box 39.5). In contrast, the Mentzer index of iron deficiency is usually greater than 13; however, some sources use 13.5 as the indicator for IDA (Benz, 2017). The degree of anemia may be exacerbated in concurrent illness or pregnancy.

Management

No specific treatment is known for β thalassemia minor. Primary emphasis should be on the education of all family members and genetic testing, and counseling should be offered.

β Thalassemia Intermedia

This variant of thalassemia is the result of various mutations that cause a disorder with a clinical severity that spans from the mild symptoms of the β-thalassemia trait to the severe manifestations of β-thalassemia major. Classification is typically based on the severity of the symptoms and the type of treatment necessary rather than by the specific genotype. Diagnosis and management are clinically based with the goal of maintaining a satisfactory Hgb of at least 6 to 7 g/dL without the regular need for RBC transfusions. Transfusions alleviate thalassemic features, but there is controversy as to whether these children should receive transfusions. This decision must be balanced against the future need for chelation if there is iron overload from repeated transfusions (DeBaun et al., 2016).

β Thalassemia Major

Homozygous forms are thalassemia intermedia and thalassemia major. Homozygous β thalassemia major (or Cooley anemia) is associated with severe anemia resulting from the decreased or absent production of Hgb A and hemolysis caused by the precipitation of excess α chains in the RBCs.

Clinical Findings

Affected infants usually become symptomatic in the first year of life and have pallor, failure to thrive, hepatosplenomegaly, and a severe anemia with an average Hgb of 6 g/dL and low MCV (60 to 70 fL). RBC morphology reveals significant microcytosis, poikilocytosis, hypochromia, target cells, and nucleated RBCs. Hgb A_2 and Hgb F levels are elevated.

Management

Proper management of the child requires collaboration with a pediatric hematologist. Standards of care for thalassemia patients should be followed (Vichinsky and Levine, 2012). RBC transfusions are usually necessary every 2 to 4 weeks with the goal of maintaining a pretransfusion Hgb level between 9.5 and 10.5 g/dL. To help with future cross matching, the provider should obtain a complete typing of the patient's erythrocyte profile before the first transfusion (phenotyping). This helps to decrease difficulties with subsequent transfusions. Splenectomy may also be indicated. Hematopoietic stem cell transplantation is the only curative modality for β thalassemia major. This has been most successful in children younger than 15 years of age without excessive iron overload and hepatosplenomegaly who have sibling-matched human leukocyte antigen (HLA) allogeneic hematopoietic transplantation (Benz, 2017; DeBaun et al., 2016). Gene therapy is being investigated and holds promise for those with this major disorder.

Iron chelation is necessary to treat the hyperferric state produced by repeated transfusions and prevent complications primarily of the heart, liver, and endocrine system. Iron overload can develop even without the use of blood transfusions because of the increased iron absorption associated with high rates of erythropoiesis and red cell destruction. Monitoring for iron stores should be done on a regular basis (Benz, 2017). Chronic iron chelation therapy is necessary in order to remove the excess iron resulting from frequent transfusions. Indications for chelation occur when the serum ferritin concentrations are excessive (variably stated as >300, >400, or >1000 mcg/L) and/or magnetic resonance imaging (MRI) T_2 suggest the presence of iron loading in critical organs such as the liver (>3 mg of iron per gram of dry weight) and the heart (Benz, 2017). Deferoxamine is administered parenterally or subcutaneously, usually via a pump overnight. It is time-consuming and associated with pain. Subcutaneously infused medications have been replaced by oral chelators. Deferasirox is an oral agent taken once daily at 20 to 30 mg/kg/day; it stabilizes the ferritin levels, thus achieving a negative iron balance (DeBaun et al., 2016). Iron excretion through chelation is further aided by the ingestion of vitamin C. Because the iron is excreted through the kidneys, hydration and monitoring of renal status are vital.

Otitis Media With & Without Effusion- p. 656

AOM is an acute infection of the middle ear

The AAP Clinical Practice Guideline requires the presence of the following three components to diagnose AOM:

- **§ Recent, abrupt onset of middle ear inflammation and effusion (ear pain, irritability, otorrhea, and/or fever)**
- **§ MEE confirmed by bulging TM, limited or absent mobility by pneumatic otoscopy, air-fluid level behind TM, and/or otorrhea**
- **§ Signs and symptoms of middle ear inflammation confirmed by distinct TM erythema or ear pain (holding, tugging, rubbing the ear)**

Etiology

- **AOM often follows eustachian tube dysfunction (ETD).**
- **Common causes of ETD include upper respiratory infections, craniofacial anomalies (cleft palate), allergies, adenoid hypertrophy, and tobacco smoke**

exposure. ETD leads to functional eustachian tube obstruction and inflammation that decreases the protective ciliary action in the eustachian tube.

- **Respiratory syncytial virus and influenza are two of the viruses most responsible for the increase in the incidence of AOM seen from January to April. Although a virus is usually the initial causative factor in AOM, strict diagnostic criteria, careful specimen handling, and sensitive microbiologic techniques have shown that the majority of AOM is caused by bacteria or bacteria and virus together.**

Most common infecting organisms in AOM:

- **§ *S. pneumoniae-most common bacteria responsible for AOM***
- **§ Nontypeable *Haemophilus influenza***
- **§ *Moraxella catarrhalis***
- **§ *S. pyogenes* (group A streptococci)**

Pathophysiology

Eustachian tube obstruction causes negative pressure as air absorbs in the middle ear. The negative pressure pulls fluid from the mucosal lining and causes a sterile fluid accumulation that may be colonized by bacteria and result in purulent fluid. Young children have shorter, more horizontal, and more flaccid eustachian tubes that are easily disrupted by viruses, which predisposes them to AOM.

Types of OM

Type	***Characteristics***
AOM	**Suppurative effusion of the middle ear**
Bullous myringitis	**AOM in which bullae form between the inner and middle layers of the TM and bulge outward**
Persistent AOM	**AOM not resolved after antibiotic therapy or AOM recurs within days of Tx**
Recurrent AOM	**3 separate bouts of AOM within 6-months or 4 within a 12-month period; often a positive family history of otitis media and other ENT disease**

Risk Factors for Otitis Media, Chronic Otitis Media, or Otitis Media With Effusion

Genetic susceptibility/sibling with history of otitis media
Native Americans and Native Alaskans
Non-Hispanic Caucasian
Prematurity
Younger than 2 years of age
Unimmunized
Day care attendance
Sharing a bedroom
Breastfeeding for less than 6 months
Parental smoking and other ETS exposure
Environmental pollution exposure
Overweight or obese
Feeding in supine position
Autumn season
Male gender

Early onset otitis media
Bilateral OME
Lower socioeconomic status
ETS, Environmental tobacco smoke; OME, otitis media with effusion.

Clinical Findings

Rapid onset of:

- **Ear pain that may interfere with activity and/or sleep, especially when lying flat**
- **Irritability and ear pulling in an infant or toddler**
- **Otorrhea**
- **Fever**

Other key risk factors or symptoms include prematurity, craniofacial anomalies or congenital syndromes associated with craniofacial anomalies, daycare attendance, disrupted sleep or inability to sleep, lethargy, dizziness, tinnitus, unsteady gait, diarrhea and vomiting, sudden hearing loss, stuffy nose, rhinorrhea, and sneezing.

- **Presence of MEE, confirmed by pneumatic otoscopy, tympanometry, or acoustic reflectometry, as evidenced by:**
    - **Bulging TM**
    - **Decreased TM translucency**
    - **Absent or decreased TM mobility**
    - **Air-fluid level behind the TM**
    - **Otorrhea**

Signs and symptoms of middle ear inflammation indicated by TM (amber color is usually seen in otitis media with effusion [OME]; white or yellow may be seen in either AOM or OME).

In addition, the following TM findings may be present:

- **Increased vascularity with obscured or absent landmarks (see Fig 36.4).**
- **Red, yellow, or purple TM. (Redness alone should not be used to diagnose AOM, especially in a crying child.)**
- **Thin-walled, sagging bullae filled with straw-colored fluid seen with bullous myringitis**

Diagnostics for AOM

- **Pneumatic otoscopy is the simplest and most efficient way to diagnose AOM.**
- **Tympanometry reflects effusion (type B pattern).**
- **Tympanocentesis identifies the infecting organism and is helpful in the treatment of infants younger than 2 months old. In older infants and children, tympanocentesis is rarely done and is useful only if the patient is toxic or immunocompromised, or in the presence of resistant infection or acute pain from bullous myringitis. If a tympanocentesis is warranted, refer the patient to an otolaryngologist for this procedure.**

Management of OAM

Treatment is based on the child's age, illness severity, and the certainty of diagnosis.

Treatment Guidelines

1. Pain management is the first principle of treatment with weight-appropriate doses of children's ibuprofen or acetaminophen to decrease discomfort and fever. Distraction, oil application, or external use of heat or cold may be of some use.

2. Antibiotics are also effective for AOM.

- **Amoxicillin remains the first-line antibiotic for AOM if there has not been a previous treated AOM in the previous 30 days, there is no conjunctivitis, and no penicillin. β-lactam coverage (amoxicillin/clavulanate, third-generation cephalosporin) is recommended when the child has been treated with amoxicillin in the previous 30 days, there is an allergy to penicillin, or the child has concurrent conjunctivitis or has recurrent otitis that has not responded to amoxicillin.**
- **If the child is younger than 2 years of age, treatment with amoxicillin or amoxicillin/clavulanate for 10 days, or for children older than 2 years of age, treatment for 5 to 7 days.**
- **Ceftriaxone may be effective for the vomiting child, the child unable to tolerate oral medications, or the child who has failed amoxicillin/clavulanate.**
    - **Clindamycin may be considered for ceftriaxone failure but should only be used if susceptibilities are known.**
    - **Prophylactic antibiotics for chronic or recurrent AOM are not recommended.**

3. Observation or "watchful waiting" for 48 to 72 hours allows the patient to improve without antibiotic treatment. Pain relief should be provided, and a means of follow-up must be in place. Recommendations for follow-up include: parent-initiated visit or phone call for worsening or no improvement; scheduled follow-up appointment; routine follow-up phone call; or give a prescription to be started if the child's symptoms do not improve or if they worsen in 48 to 72 hours.

4. Routine follow-up is not needed if the child improves within 48 hours. If the child has not shown improvement in ear symptomatology after 48 to 72 hours, the child should be seen to confirm or exclude the presence of AOM. If the initial management option was an antibacterial agent, the agent should be changed.

Management of Persistent and Recurrent Acute Otitis Media p. 658

- **Persistent AOM occurs when antibiotic therapy is completed and AOM is still present or AOM recurs within days of treatment. Retreatment with a broader-spectrum antibiotic is suggested.**
- **Persistent MEE is common after resolution of acute symptoms and should not be seen as a need for continuing antibiotics (see Otitis Media with Effusion section).**
- **Recurrent AOM is defined as more than three distinct and well-documented bouts of AOM in 6 months or four or more episodes in 12 months.**

An otolaryngology referral is indicated when appropriate therapy for otitis media fails. Placement of tympanostomy or PE tubes can help relieve discomfort, reduce time with OME, improve hearing, and decrease the likelihood of further infection. Indications for tympanostomy and the insertion of pressure-equalizing tubes is discussed below.

Other Treatment Issues for AOM

- **The PCP is encouraged to maintain understanding of current recommendations for AOM management because of rapid changes in resistance patterns and newly developed treatments.**
- **Decongestants and antihistamines are not indicated.**
- **Antimicrobial ototopical drops (ofloxacin or ciprofloxacin) or ophthalmic drops (tobramycin or gentamicin) are indicated if the TM is perforated (Fig 36.5), the child has otorrhea, or the child has patent, draining PE tubes.**

• Xylitol, a sugar found in fruits and birch bark, has bacteriostatic effects against S. pneumoniae and interferes with bacterial adhesion to mucous membranes. It appears to have some suppressive effects in preventing ear infections. Xylitol is available in an oral solution, lozenges, and chewing gum. The lozenges and chewing gum are more effective than the oral solution. Children younger than 2 years old cannot have chewing gum or lozenges. Xylitol must be given three to five times a day on a regular basis to be effective.
• There is no safe or effective herbal treatment for AOM or OME.

Complications of AOM

Persistent AOM, persistent OME, TM perforation, OE, mastoiditis, cholesteatoma, tympanosclerosis (Fig 36.6), hearing loss of 25 to 30 dB for several months, ossicle necrosis, pseudotumor cerebri, cerebral thrombophlebitis, and facial paralysis are possible complications.

Medications Used to Treat Acute Otitis Media p. 658

Prevention and Education for AOM

The following interventions, shown to be helpful in preventing AOM, should be encouraged:

- **Exclusive breastfeeding until at least 6 months of age protects against AOM (Bowatte et al., 2015)**
- **Avoid bottle propping, feeding infants lying down, and passive smoke exposure**
- **Pneumococcal vaccine; specifically, PCV13, which contains subtype 19A**
- **Annual influenza vaccine helps prevent otitis media**
- **Xylitol liquid or chewing gum as tolerated**
- **Choose licensed day care facilities with fewer children**
- **Educate regarding the problem of drug-resistant bacteria and the need to avoid antibiotic use unless absolutely necessary. If antibiotics are used, the child needs to complete the entire course of the prescription and follow up if symptoms do not resolve.**

Otitis Media With Effusion p 659

The diagnosis of OME is made in the presence of MEE without signs or symptoms of acute ear infection. MEE decreases the mobility of the TM and interferes with sound conduction.

Etiology

OME can occur spontaneously with ETD caused by an inflammatory process after AOM, viral illness, anatomic abnormalities, barotrauma, allergies, or a combination of these conditions. ETD changes the middle ear mucosa in the following sequence: (1) the mucosa becomes secretory with increased mucus production, (2) the mucus absorbs water as the mucosa becomes viscous, and (3) fluid becomes stuck behind the TM. Bacterial biofilms may explain the persistence of OME. Biofilms are mixed microorganisms enclosed in a polymeric matrix that adhere to surfaces, such as the middle ear mucosa.

Risk Factors for Hearing Loss Caused by Otitis Media with Effusion p. 660

Bilateral Otitis media with effusion (OME) for 4 months or longer
If two or more present:

- **OME present for longer than 8 weeks**
- **Speech development slower than peers**
- **Speech less clear than previously**
- **Child decreases amount of talking**

Child less responsive to name and other familiar sounds
Child says “Huh?” or “What?” frequently
Child sits close to TV or wants volume louder
Child has difficulty learning (reading, spelling)
Child is hyperactive or overly inattentive
OME, Otitis media with effusion.

Clinical Findings of OME

- **Children with OME are often afebrile and asymptomatic**
- **May present with intermittent complaints of mild ear pain, fullness in the ear (“popping” or feeling of “talking in a barrel”), dizziness or impaired balance.**
- **The older child may complain of hearing loss.**
- **The young child may request that you speak louder or require a higher volume than usual for the radio or television.**
- **Pneumatic otoscopy reveals decreased TM mobility**
- **An abnormal-appearing TM, often described as dull, varying from bulging and opaque with no visible landmarks to retracted and translucent with visible landmarks and an air-fluid level or bubble may be seen**

Diagnostics

The tympanogram is flat-type B. The audiogram can show hearing loss ranging from mild to moderate (25 to 60 dB).

Management

Recommendations for management of OME in children 2 months to 12 years of age include:

1. Pneumatic otoscopy should be performed to document OME, particularly in children with ear pain and/or hearing loss. Tympanometry may be indicated in children to confirm OME diagnosis.
2. Manage the child with OME with watchful waiting for 3 months from date of diagnosis.
3. Intranasal steroids or systemic steroids, system antibiotics, antihistamines, and decongestants are not recommended for treatment of OME.
4. Children with OME who are at risk for speech, language, and learning problems should be identified. An age-appropriate hearing test should be performed if OME persists for 3 months or longer.

- **At-risk children are defined as having developmental delays because of sensory, physical, cognitive, or behavioral factors (e.g., hearing loss independent of OME, speech or language delays, pervasive or other developmental disorders, syndromes or craniofacial disorders, blindness, and/or cleft palate). These children should be promptly referred for hearing, speech, and language evaluation.**

5. Reevaluate a child with OME every 3 months until the effusion resolves, or every 3 to 6 months for children with chronic OME. When managing a child with OME, clinicians should document resolution of OME in the medical record.
6. Communication is key to families understanding the duration and course of OME, the need for follow-up, and the associated sequelae including potential hearing impairment and impact on speech and language development.
7. Referral to an otolaryngologist is recommended when otoscopy suggests possible or impending structural damage of the TM, significant hearing loss is identified, and/or for chronic or persistent OME for more than 6 months. The need for referral should be clearly communicated to the family.

8. In a child younger than 4 years old, insertion of tympanostomy tubes is performed for persistent or chronic OME with associated sequelae; adenoidectomy is not recommended unless an indication exists other than OME. In a child 4 years old or older, insertion of tympanostomy tubes and/or adenoidectomy is recommended for OME.

Tourette syndrome—427

Most severe chronic tic disorder; symptoms include multiple motor and vocal tics, varying in nature and severity over time

1. Frequently have additional problems such as aggressiveness, social withdrawal, self-harming acts, and sleep disorders
2. Symptoms become more unpredictable during adolescence and may result in school refusal

· Etiology/Incidence

1. Uncertain etiology; possible genetic central nervous system disturbance
2. Believed that abnormal neurotransmitters in the brain contribute
3. Transient tics
    1. Occur in 25% of normal children
    2. Often begin in school-age children and can be intensified by anxiety, fatigue, or excitement
4. Tourette syndrome
    1. Onset between 2 and 15 years of age with mean age 6 to 7 years
    2. Incidence—1:10,000 persons
    3. Males 3 times more frequently affected

· Signs and Symptoms

1. Characteristics of tics
    1. Variable expression—frequency, intensity, and severity
    2. Exacerbated by stress
    3. Some degree of voluntary control may be present
    4. Typically subside during sleep
2. Simple tics
    1. Movements present that resemble nervous habits
    2. Facial "twitches," head shaking, eye blinking, shoulder shrugging, or throat clearing

Tourette syndrome

1. Simple tics
2. Complex sequences of coordinated movements (e.g., bizarre gait, kicking, jumping, body gyrations, scratching, and seductive or obscene gestures)
3. Involuntary vocalizations occur, ranging from simple to complex noises
4. Expression is gender influenced
    1. (1) Motor and vocal manifestations more prevalent in boys
    2. (2) Behavioral problems, such as obsessive-compulsive disorder, more common in girls

Management/Treatment

1. Transient tics—support and education for child and family
2. Chronic tics/Tourette syndrome
    1. Referral to mental health specialist
    2. May involve psychotherapy, behavior management, stress management
    3. Pharmacologic management
        1. Consultation and/or referral— pediatric neurologist
        2. Medications useful to suppress behavioral symptoms but interfere with daily functioning

UTI- p.824

Types:

- Asymptomatic bacteriuria
- Cystitis
- Pyelonephritis

  - Asymptomatic bacteriuria is bacteria in the urine without other symptoms, is benign, and does not cause renal injury.
  - Cystitis is an infection of the bladder that produces lower tract symptoms but does not cause fever or renal injury.
  - Pyelonephritis is the most severe type of UTI involving the renal parenchyma or kidneys and must be readily identified and treated because of the potential irreversible renal damage.

Clinical Signs of Pyelonephritis

· Fever
· Irritability, and vomiting in an infant
· Urinary symptoms associated with fever, bacteriuria, vomiting, and renal tenderness in older children.

UTIs are the most common cause of serious bacterial infection in infants younger than 24 months old with fever without a focus.
A complicated UTI is defined as a UTI with fever, toxicity, and dehydration, or a UTI occurring in a child younger than 3 to 6 months old.

UTIs may be classified based on their association with other structural or functional abnormality such as VUR, obstruction, dysfunctional voiding, or pregnancy.

A UTI must be identified as a first occurrence, recurrent (within 2 weeks with the same organism or any reinfection with a different organism), or chronic (ongoing, unresolved, often caused by a structural abnormality or resistant organism).

Age and gender of the pediatric patient are important factors in determining the evaluation method and the course of treatment.

Prevalence
The risk of UTI in infants 2 to 24 months old is about 5%. The incidence in females is more than twice that of males (2.27%); uncircumcised boys have a rate 4 to 20 times greater than circumcised boys (AAP Subcommittee on Urinary Tract Infection, 2016). There is a greater frequency in premature and low-birth-weight infants. Females older than 12 months old have 2.1% prevalence; after the first year of life, it is also more common to find a UTI in females than in males with an overall incidence of 1% to 3% in girls and 1% in boys (Elder, 2016d). The incidence of UTI is often increased in sexually active adolescent girls. The risk of recurrence within the first year after an initial infection is common.

Etiology
The organism most associated with UTI is Escherichia coli (70%), although other organisms (such as Enterobacter, Klebsiella, Pseudomonas, and Proteus) can cause infection.

UTI secondary to group B streptococcus is more common in neonates.

Contributing Factors:

- Most UTIs are thought to be ascending (i.e., the infection begins with colonization of the urethral area and ascends the urinary tract). If the infection progresses to the kidney, intrarenal reflux deep into the kidneys can lead to scarring.
- The most important risk factor for the development of pyelonephritis in children is VUR, which can be detected in 10% to 45% of young children who have symptomatic UTIs.
- Reflux of infected urine from the bladder increases the risk of pyelonephritis. Kidney damage occurs in the compound papillae, which have wide and gaping openings allowing intrarenal reflux. The compound papillae are located in the upper and lower poles of the kidney, the usual site of scarring.

Host resistance factors and bacterial virulence factors are also important in the etiology of UTIs:

- Presence of a structural abnormality or dysplasia (such as VUR, obstruction, or other anatomic defect), or the presence of functional abnormalities (such as dysfunctional voiding or constipation).
- Female gender (having a short urethra), poor hygiene, irritation, sexual activity or sexual abuse, and pinworms.
- Several bacterial factors are known, but the two most important ones are adherence and bacterial virulence. Bacteria that have fimbriae or pili are able to adhere to the surface of the bladder mucosa; this allows the bacteria to resist the bladder's defensive cleansing flow of urine and causes tissue inflammation and cell damage.
- Adherence may also play a role in bacteria ascending the urinary tract. Virulence refers to the toxicity of substances released by bacteria. The greater the virulence, the greater the damage to the urinary tract. Both of these factors enhance colonization of the urinary tract and aid in the persistence and effect of the bacteria.

Clinical Findings of UTI (p. 825)

Neonates	Infants	Toddlers and Preschoolers	School-Age Children and Adolescents
Jaundice	Malaise, irritability	Altered voiding pattern	"Classic dysuria" with frequency, urgency, and discomfort
Hypothermia	Difficulty feeding	Malodor	
Failure to thrive (FTT)	Poor weight gain	Abdominal/flank pain [a]	Malodor
Sepsis	Fever [a]	Enuresis	Enuresis
Vomiting or diarrhea	Vomiting or diarrhea	Vomiting or diarrhea [a]	Abdominal/flank pain [a]
Cyanosis	Malodor	Malaise	Fever/chills [a]
Abdominal distention	Dribbling	Fever [a]	Vomiting or diarrhea [a]
Lethargy	Abdominal pain/colic	Diaper rash	Malaise

[a] Findings increase likelihood of pyelonephritis.

Labs/Diagnostics

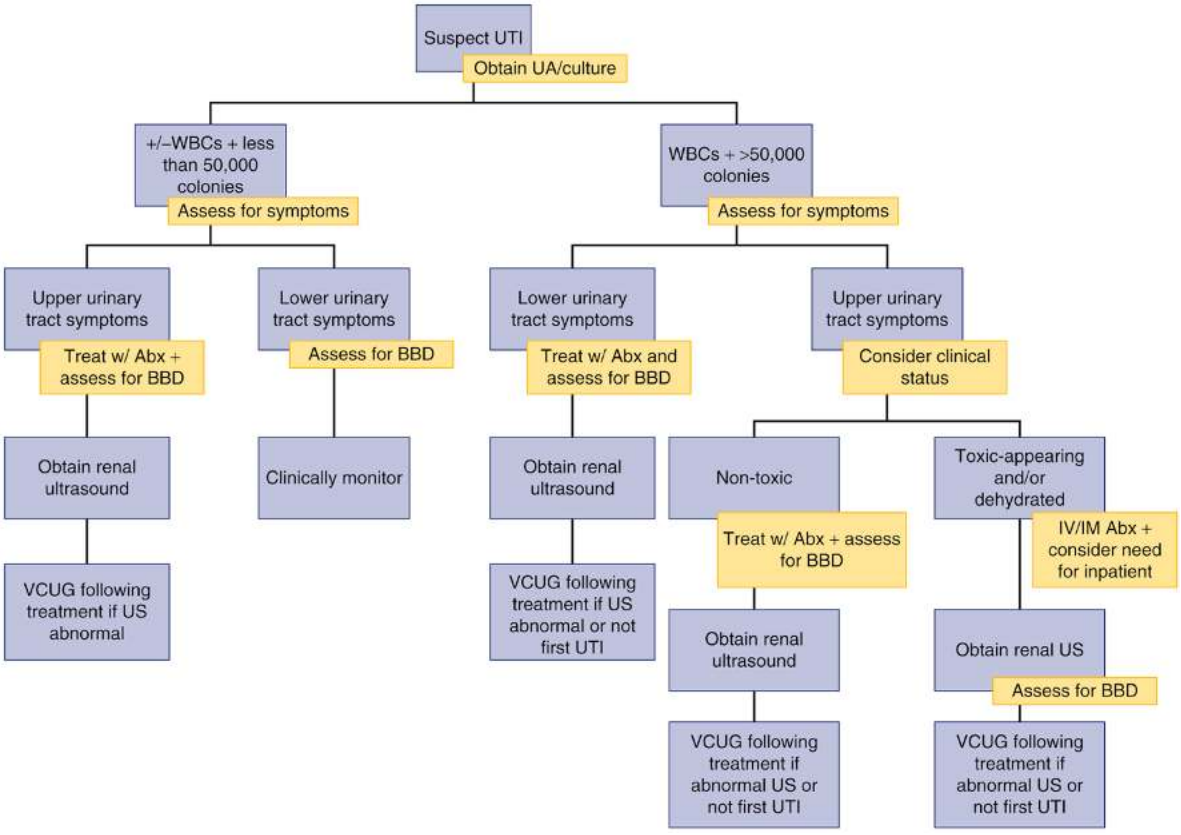
The method used to collect urine has an effect on the interpretation of results. Bagged urine specimens, even after cleaning the external genitalia prior to placement, produce a high incidence of false positive UA results due to contamination and thus must not be used to determine UTI. If antimicrobial therapy must be initiated due to an ill-appearing child, catheterization and urine culture must be obtained prior to administering antibiotics. Older children who can void on command should be able to obtain a clean-catch void. Having the female child sit with knees apart, feet supported, and torso leaned forward while on the toilet separates the labia and decreases contamination.

Urine culture is essential to confirm the diagnosis. UTIs cause cultures with greater than 100,000 colonies of a single pathogen in a clean catch urine specimen, greater than 50,000 in a catheterized or suprapubic specimen, or 10,000 colonies of a single pathogen in the symptomatic.

• UA should be used only to raise or lower suspicion. Suspicious findings include foul odor, cloudiness, nitrites, leukocytes, alkaline pH, proteinuria, hematuria, pyuria, and bacteriuria.
• Leukocyte esterase chemical tests detect pyuria, but pyuria may arise from causes other than UTI.
• Consider obtaining a lab UA with reflexive gram stain and microscopy if dipstick findings are positive.
• Bacterial identification and determination of sensitivities are necessary in patients who appear toxic or could have pyelonephritis, have relapses or recurrent UTI, or are nonresponsive to medication.
• Complete blood count (CBC) (elevated WBC count), erythrocyte sedimentation rate (ESR), C- reactive protein (CRP), BUN, and creatinine should be done if the child is younger than 1 year old, appears ill, or if pyelonephritis is suspected.
• Blood culture should be done if sepsis is suspected (see Chapter 28).

FIG 41.1 Suspect Urinary Tract Infection (UTI) Algorithm.

Imaging and Prophylaxis (p. 828)



Why Do a Radiologic Workup?
• To identify any structural or functional abnormality of the urinary tract
• To identify any renal scarring or damage
Who Requires a Workup?
• Order a renal and bladder ultrasound on children with the first positive urine culture and with fever and systemic illness. In children with one or more infections of the lower urinary tract (dysuria, urgency, frequency, suprapubic pain), renal and bladder ultrasound may be considered; however, assessment and treatment of bladder and bowel dysfunction is most important.
• If the renal ultrasound is abnormal, voiding cystourethrogram is indicated.
What about Prophylaxis?

• There is controversy about if and when prophylaxis should be used (AAP Subcommittee on Urinary Tract Infection, 2016). If a decision to use prophylaxis is made and depending on the source, between one-quarter to one-half of the treatment dose of antibiotic may be given at bedtime.
• Nitrofurantoin: Older than 2 months old: 1-2 mg/kg as a single daily dose; expensive; liquid form poorly tolerated; consider sprinkling capsules over applesauce, yogurt, pudding
• TMP-SMX, trimethoprim-sulfamethoxazole:
TMP 2 mg/kg as a single daily dose or 5 mg/kg twice per week (based on TMP component) if older than 1 month
• Cephalexin: 10 mg/kg as a single daily dose
• Amoxicillin: 10 mg/kg as a single daily dose; can be used for a newborn or premature infant; not used past the first 2 postnatal months; shelf life for liquid is 14 days

Study	Cost	Advantages	Disadvantages	Use
Ultrasound	Least expensive	Shows structure, shape, and growthDetects structural abnormality, obstruction, pyelonephritis, large scarsPainless, low risk, no radiation, noninvasive, available	Does not detect small scars of VUR Poor visualization of uretersDoes not measure renal function or transient injury to kidney	Initial evaluation with first UTI and follow-up
Voiding cystourethrogram (VCUG) (radiographic)	Least expensive	Detects and grades VUR if high or low pressure, high or low bladder volumes, during voiding, during early or late bladder fillingVisualizes bladder and urethra (especially in males) and diverticula	Does not detect obstruction, pyelonephritis, scarsGreater radiation than with scanRequires intravesical administration of contrast	Indicated in infants and children with abnormal ultrasound
Dimercaptosuccinic acid (DMSA) renal scan (nuclear)	Most expensive	Detects acute inflammation, scars, and obstructionEarlier detection of parenchymal damage—large or small scars, permanent or focal—than with IVP (1-3 years old)	Does not detect VUR or measure renal functionDoes not evaluate calyces, ureters, bladder, or urethra	Follow-up for fever of unknown origin and negative ultrasound in neonatesTo diagnose acute pyelonephritisTo detect renal scars
Computed tomography (CT) (contrast)	Expensive	Detects obstruction, pyelonephritis, large scars	Does not detect small scars or VUR Risk of allergic reaction, acute renal failure	Trauma

IVP, Intravenous pyelogram; *UTI*, urinary tract infection; *VUR*, vesicoureteral reflux.

Management

Goals of treatment are to quickly identify the extent and level of infection; to eradicate infection; to provide symptomatic relief; to find and correct anatomic or functional abnormalities; and to prevent recurrence and renal damage. When deciding on a treatment plan, the child's age, gender, symptoms, the suspected location of the UTI, and antibiotic resistance patterns in the community must be considered.

Asymptomatic Bacteriuria

If there are no leukocytes on UA, no treatment is indicated.

Uncomplicated Cystitis

Use regional antibiotic resistance patterns and culture and sensitivity results when choosing antibiotics. Short-term (3 to 5 days) antibiotics may be as effective in treating non-febrile bladder infections as standard 7- to 10-day dosing with no increased risk of recurrence. Children 2 to 24 months old and febrile children should have 7 to 14 days of antibiotics. Additional specific recommendations for the febrile child less than 24 months can be found in the AAP Guidelines.

Recommended oral medications:

• Trimethoprim-sulfamethoxazole (TMP-SMX): More than 2 months old—8 to 12 mg/kg TMP component in two divided doses; adolescents, 160 mg TMP component every 12 hours.
• Amoxicillin: Younger than 3 months old—20 to 30 mg/kg/day in two divided doses every 12 hours; older than 3 months old—25 to 50 mg/kg/day in two divided doses; adolescents, 250 to 500 mg every 8 hours or 875 mg every 12 hours.
• Amoxicillin clavulanate (doses for amoxicillin component): Younger than 3 months old—30 mg/kg/day in two divided doses; older than 3 months old—20 to 45 mg/kg/day in two or three divided doses; adolescents—250 to 500 mg every 8 hours or 875 mg every 12 hours.
• Cephalexin: 50 to 100 mg/kg/day divided in four doses and given every 6 hours (maximum dose of 4 g/day).
• Cefixime: Older than 6 months old—16 mg/kg/day divided every 12 hours for first day, then 8 mg/kg/day divided every 12 hours to complete 10-day treatment; adolescents—400 mg every 12 to 24 hours.

• Nitrofurantoin: Older than 1 month old—5 to 7 mg/kg/day divided every 6 hours (maximum 400 mg/24 hours); adolescents—50 to 100 mg/dose every 6 hours (macrocrystals) or 100 mg twice a day (dual release).

• Recurrent UTI: Further evaluation required. Prophylactic antibiotic use (Box 41.2) is controversial and not routinely recommended.
• Acute pyelonephritis: Oral therapy is equally as effective as parenteral therapy in treating pyelonephritis and preventing kidney damage.
• Hospitalization is required if severity of symptoms warrants—dehydrated, vomiting, or not drinking. Children 1 month old and younger should be admitted and provided a parenteral regimen.
• Infants over 1 month and children with uncomplicated pyelonephritis (well hydrated, no vomiting, no abdominal pain) can be effectively treated with cefixime, cephalexin, or amoxicillin clavulanate.
• Adolescents with uncomplicated pyelonephritis can be treated with either amoxicillin clavulanate (875/125 mg twice a day) or ciprofloxacin (500 mg twice a day or extended release 1000 mg once a day).
• Follow-up cultures are not routinely needed. However, follow-up urine culture should be done 48 to 72 hours after initiating treatment if symptoms persist or organism resistance is found in the community.
• If the culture is not sterile or if no clinical improvement is seen, antibiotic change should be based on sensitivity report. Urine should be sent for bacterial identification and sensitivity studies if not performed initially, and an alternative broad-spectrum antibiotic should be used pending those results. Culture should again be repeated after 48 to 72 hours if response to therapy limited.
• Phenazopyridine may be given at 12 mg/kg/day for 6- to 12-year-olds and 200 mg for those older than 12 years old, three times a day for dysuria.

• Radiologic workup (Table 41.4) is recommended to identify any structural or functional abnormality of the urinary tract and any renal scarring or damage.
• Children younger than 2 years old with the first UTI should have a renal and bladder ultrasound as soon as the urine is sterile or when the prescribed antibiotic has been completed. Additionally, all children with fever, diagnosed with pyelonephritis, or with recurrent UTIs should have a renal and bladder ultrasound. VCUG does not need to be done routinely with first febrile UTI. However, if ultrasound reveals hydronephrosis, scarring, or other atypical or concerning findings, VCUG should be utilized (American Urological Association [AUA], 2010/2017).
• DMSA scan ordered by the urological specialist may be obtained when renal scarring is suspected or when diagnosis of pyelonephritis is uncertain (AUA, 2010/2017).

Education
• Clear explanation of the cause, potential complications, and overall treatment plan, including short- and long-term plans.
• Frequent and complete voiding and increased fluid intake, especially water. Scheduled voiding times, voiding with knees spread apart, or double voiding (voiding and then immediately attempting to void again) is helpful.
• Proper hygiene and avoiding irritants, such as bubble baths, sitting in soapy water, and perfumed soaps. Avoid wearing tight pants, especially spandex pants. Wear cotton underwear. Treat perineal inflammation to help prevent UTI.
• Treat constipation, pinworms.
• Encourage sexually active females to drink water before intercourse and void immediately afterward.
• Decrease intake of bladder irritants, such as the “four Cs” (caffeine, carbonated beverages, chocolate, citrus), aspartame (NutraSweet), alcohol, and spicy foods.
• Seek prompt medical attention with recurrence of fever and/or duration of fever for more than 48 hours, especially if younger than 24 months old.

Cerebral Palsy: 995-999

- chronic nonprogressive motor disorder that is the result of damage in the brain that control motor function

- Earlier Dx and treatment - impact long term outcomes
- prevalence 1.5 to more than 4 per 1000 live births
- Risk factors:
    - Prenatal/birth history risk factors
    - Hearing and vision or ocular problems, such as strabismus, nystagmus, and optic atrophy
    - Change in growth parameters, especially decreased head circumference
    - Early head injury, meningitis, or seizures
    - Muscle tone: hypotonic/hypertonic; tone can be hypotonic before 6 months old, then become hypertonic in the affected extremities
    - Developmental milestones: They may be delayed but should still be attained depending on the extent of CP; persistent primitive reflexes are common (e.g., Moro and tonic neck). Hand preference before 1 year old is highly suspect.
    - Feeding history of regurgitating through the nose, inability to coordinate suck and swallow, inability to advance the diet to textured foods—oral-motor coordination problems
    - Irritability or depressed affect (including unusual sleepiness) as a neonate
    - Difficulty with movement, grasp and release, self-feeding, and head control to look around; inability to change position per developmental level
    - Communication problems, either in language or speech proficiency
- Dx
    - MRI : structural abnormalities
    - Chromosomal/metabolic studies
    - Lumbar Puncture if sepsis
- Management
    - Multidisciplinary team referral
    - family education/support about Dx
    - nutrition: oral motor coordination problems
    - elimination: constipation is common
    - Dentistry: problems with oral mobility lead to gum disease
    - Drooling: inability to manage oral secretions

Chronic Genitourinary Conditions in Males

Hypospadias

Hypospadias is a common congenital abnormality in which the urethral meatus is located anywhere from the proximal glans to the perineum on the ventral surface (underside) of the penis.
The etiology of hypospadias is unknown.
Hypospadias occurs in 1 in 250 male infants with an increased risk if family members have hypospadias

History and Clinical Findings

1. • A family history of a male relative with GU problems may be reported.
2. • There is report of an unusual direction, particularly downward, to the urine stream.
3. • Other findings include inguinal hernia or undescended testicles (10%), and/or chordee

Physical Examination

It is essential to visualize the urethral meatus, which is facilitated by pulling the ventral shaft skin in a downward and outward direction. The deformity is described by location: distal (60%), mid-penile (25%), or proximal (15%).

Management

The goal of surgical repair is to have a functional penis that appears normal. Historically, circumcision was avoided because the foreskin may be used in the surgical repair. However, newer surgical techniques that do not require the use of skin flaps change this standard of care. Physiologic phimosis may prevent visualization of a urethral anomaly during a well-child exam, especially with a mild form of hypospadias. Surgical success is not compromised in these cases. Referral should be made to a pediatric urologist at birth or at detection of the

anomaly. Surgery to correct hypospadias is best done around 6 to 12 months old. Repair is usually accomplished in a one-stage outpatient procedure unless it is a complex defect.

Cryptorchidism (Undescended Testes)

Cryptorchidism is a testis that does not reside in and cannot be manipulated into the scrotum. A Undescended testes is a common disorder that often causes great anxiety for parents.

This condition is the most common GU disorder in boys, occurring in 3.4% of term newborns. Testicular descent occurs at 7 to 8 months gestation, so it is therefore more common in preterm (30%), low-birth-weight, and twin infants. A great majority of undescended testes descend spontaneously during the first 3 months of life but after 6 months old it is rare (0.8%) for them to descend. Cryptorchidism is bilateral in 10% of cases. Retractile testes are bilateral and most common in boys 5 to 6 years old (Elder, 2016b).

History and Clinical Findings

1. • Family history of undescended testes or testicular malignancy
2. • Testes not consistently descended during the infant's bath/warm environments
3. • Risk factors include prematurity, hypospadias, congenital hip subluxation, low birth weight, Down syndrome, Klinefelter syndrome
4. • Other congenital, endocrine, chromosomal, or intersex disorders

Physical Examination

Having the child sit cross-legged, frog-legged, squat, or stand can facilitate testicle descent and palpation.

1. • Scrotal rugae less fully developed
2. • Bilateral or unilateral absence of a testicle
3. • Retractile testes, which move between the scrotum and external ring, but can be manipulated to the lower part of the scrotum and remain there; in children 3 months to 7 years old, retraction is especially common with tactile stimulation of the area or cold
4. • Gliding testes that lie between the scrotum and external ring and can be manipulated to the lower part of the scrotum, but return to the high position
5. • Location of the testis is described as prescrotal (at the external inguinal ring); canalicular, high or low (between the external and internal rings), the most common type; ectopic (superficial inguinal, femoral, or perineal); or intraabdominal (above the internal inguinal ring), not palpable, occurring in less than 15% of males with undescended testes.

Diagnostic Studies

None are indicated except in newborns with potential sex abnormalities, hypopituitarism, Down syndrome, or congenital adrenal hyperplasia.

Management

The goals of treating undescended testes are to improve fertility outcome, decrease malignancy risk, and minimize the psychological stress associated with an empty scrotum. Management is surgical intervention between 9 and 15 months old. Hormonal therapy is not effective in stimulating testicular descent. Surgery at 6 months old is appropriate if orchiopexy is performed by a skilled pediatric urologist or surgeon with an attendant and skilled pediatric anesthesiologist. In a child younger than 1 year old, regular examination to assess the position of the testes should be performed at every well-child care visit. If the testes remain undescended, referral to a pediatric urologist or surgeon should occur by 6 months old. If undescended testes are found after 1 year old, the child should be immediately referred to a pediatric urologist or surgeon for treatment. Teach self-palpation of testes.

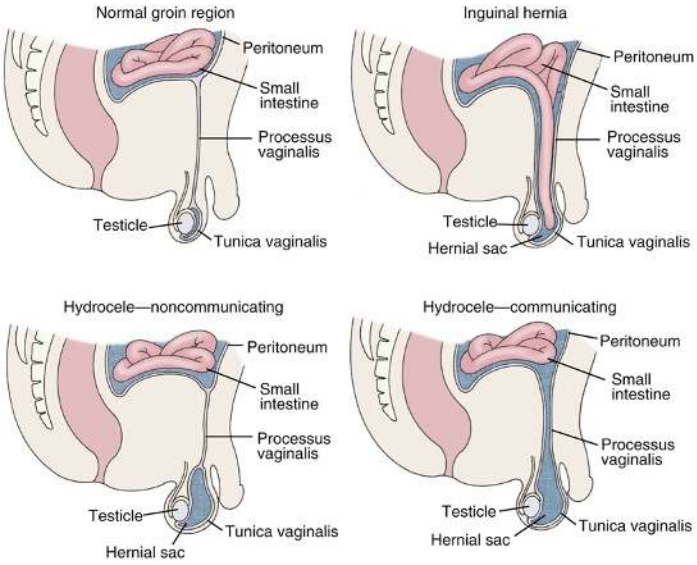


FIG 41.6 Hydroceles and Hernias.

Hydrocele

A common cause of painless scrotal swelling is a hydrocele, a collection of serous fluid in the scrotal sac.

History and Clinical Findings

Hydroceles that persist beyond 1 year old are assumed to be in conjunction with a hernia. In older children, they also occur after trauma, with an inflammatory illness, or neoplasm.

1. • Intermittent or constant bulge or lump in the scrotum, often more distally placed. Scrotal size increases with activity and decreases with rest.
2. • Overlying skin may be tense.
3. • Bluish discoloration in the area of bulge.
4. • No distress or vomiting.

Physical Examination

1. • Asymmetry or a scrotal mass present; if swelling is present in the inguinal area, a hernia is probable; swelling is usually unilateral
2. • Testes descended
3. • Cremasteric reflex present
4. • Noncommunicating hydrocele—scrotal sac tense, slightly blue tinged, fluctuant, and does not reduce; no swelling in the inguinal region
5. • Communicating hydrocele—fluid in the scrotal sac comes and goes (probably flat in the morning, swollen later in the day)

Management

1. • Noncommunicating hydrocele: Fluid is generally absorbed spontaneously; no treatment is indicated unless the hydrocele is so large that it is uncomfortable or persists longer than 1 year.
2. • Communicating hydrocele: Many communicating hydroceles will resolve without surgery and deserve observation (Guerra and Leonard, 2017). If the hydrocele persists for more than 1 year, referral for surgical intervention is recommended.

For children over 12 months old, a diagnostic evaluation by the urologist is warranted if the congenital hydrocele has not resolved or with the initial onset of clinical findings. Surgery is usually done on an outpatient basis.

Spermatocele

A benign, painless scrotal mass or cyst on the head of the epididymis or testicular adnexa containing sperm is called a *spermatocele.* A spermatocele is an uncommon, generally benign finding and occurs in the mature male or older adolescents.

History and Clinical Findings

1. • Scrotal swelling but otherwise asymptomatic
2. • Painless, mobile cystic nodule usually less than 1 cm in size, superior and posterior to the testicle
3. • No change in size with the Valsalva maneuver

4. • An ultrasound may be ordered if large and bothersome or painful

Management

No treatment is required unless the cyst is large and bothersome or painful in which case referral to a urologist is recommended.

Varicocele

A varicocele is a benign enlargement or dilation of testicular veins causing a painless scrotal mass of varying size that may feel like a "bag of worms." It is usually found on the left side.

History and Clinical Findings

1. • Usually, a painless swelling is noted in the left side of the scrotum, occasionally a "dull ache" or "heavy" feeling if large.
2. • Scrotal swelling with prolonged standing causes pain; swelling and pain resolve upon reclining. Pain can occur with strenuous physical activity.

Physical Examination

1. • In the standing position, a "bag of worms" can be felt posterior and superior to the testis that collapses on lying and enlarges with the Valsalva maneuver.
2. • Measure and compare the size of both testes (length, width, and depth) using a standard orchidometer.
3. • Grade 3 varicocele, the classic "bag of worms," is larger than 2 cm and easily visualized; grade 2 varicocele is 1 to 2 cm in diameter and is easily palpable when the adolescent is standing but not visualized; grade 1 varicocele is the most common, very small, and difficult to palpate (the Valsalva maneuver may help).
4. • Cremasteric reflex is present.

Diagnostic Studies

1. • Ultrasonography to rule out malignancy in children younger than 10 years old
2. • Serial ultrasonography to measure testicular size every 6 to 12 months of age

Management

Asymptomatic grade 1 varicocele with normal testicular volumes usually does not require intervention in adolescence but involves ultrasonographic monitoring of testicular size every 12 months (AUA, 2010/2017). Any change in comfort level should be reported. Referral to a surgeon or urologist should be made if the varicocele is grade 2 or 3, if the varicocele is painful, if the difference in testicular volume is marked (greater than 2 mm by ultrasound), if the varicocele is right sided or bilateral, or if testicular growth becomes retarded over a 6- to 12-month period (Elder, 2016b). Ligation is the usual procedure, completed on an outpatient basis with few complications.

Inguinal Hernia

A scrotal or inguinal swelling (or both) that results in bulging of abdominal contents through a weakness in the abdominal wall is an inguinal hernia (see Fig 41.6). In females, inguinal hernias cause swelling in the inguinal area and labia majora.

Inguinal hernias are much more common in males than in females (8 to 10:1), occurring in 1% to 5% of boys. Premature infants are at increased risk (7% to 30% of males, 2% of females). Indirect hernias are a congenital condition and are the most common type in children. Direct hernias are rare in childhood and normally are the result of straining and weakened abdominal muscles.

History and Clinical Findings

1. • Family or personal history of undescended testes
2. • Swelling in the inguinal area, scrotum, or both that comes and goes and increases with crying or straining
3. • Prematurity, weight lifting, or obesity

Physical Examination

1. • Swelling is found in the inguinal area, scrotal area (labia majora in females), or both.
2. • The hernia is reducible with pressure on the distal end.
3. • Direct hernias push outward through the weakest point in the abdominal wall.
4. • Indirect hernias push downward at an angle into the inguinal canal.
5. • The child is fussy and has a distended abdomen if the hernia is incarcerated.

6. • Silk glove sign: A sensation of two surfaces rubbing against each other while one palpates the spermatic cord as it crosses the pubic tubercle.

Diagnostic Studies

An abdominal radiograph is helpful if air is present below the inguinal ligament. Ultrasonography differentiates a hernia from a hydrocele and is especially helpful if an incarcerated hernia is suspected.

Management

If a child is seen with a hernia, an attempt should be made to reduce it, and the child should be referred to a surgeon or urologist for repair within 1 to 2 weeks. Even if no swelling is seen at the visit but is elicited by the history, the child should be referred to a surgeon or urologist. Inguinal hernias do not resolve spontaneously. Premature infants should have the hernia repaired prior to discharge. If the hernia is not easily reduced; if it is painful; or if a hard, tender, or red mass is present, refer immediately. If reduction is difficult and ischemia is ongoing, hospitalization and surgical repair within 24 to 48 hours are indicated.

Testicular Masses

A mass located on the testicle is most often a malignancy. Testicular tumors can occur at any age; 35% of prepubertal testicular tumors are malignant. Most of the tumors are yolk sac tumors; however, rhabdomyosarcoma and leukemia can appear in this age group; 98% of painless testicular tumors in adolescents are malignant (Elder, 2016b).

History and Clinical Findings

1. • Family history of testicular cancer
2.
3. • Sensation of fullness or heaviness
4. • Possibly no complaints because testicular masses cause little or no pain and are often small
5. • Cryptorchidism, trauma, and atrophy

Physical Examination

1. • A hard, painless testicular mass.
2. • There may be an associated hydrocele.
3. • The abdomen and supraclavicular areas should be assessed for any palpable nodes.

Diagnostic Studies

1. • Serum levels of alpha-fetoprotein, β-human chorionic gonadotropin (β-hCG), and lactate dehydrogenase if tumor is suspected.
2. • Scrotal sonography establishes the location of the mass and differentiates a cystic from a solid mass.
3. • CT scan is indicated to evaluate for metastasis and ordered by specialists.

Management

Any child or adolescent with a testicular mass must be referred immediately for further evaluation. Treatment is dependent on the stage and type of tumor and includes orchiectomy, irradiation, and/or chemotherapy.

Acute Male Genitourinary Conditions

Scrotal Trauma

Trauma to the scrotum most often occurs as a result of sports participation or play, usually from direct blows to the scrotum and straddle injuries. In a prepubertal child, the testicle is often spared damage because of the small size and mobility of the testes. Damage occurs when the testicle is forcibly compressed against the pubic bones. Significant symptoms (swelling, discoloration, and tenderness) from minor trauma suggest an underlying tumor.

Clinical Findings

1. • Pain after injury; older children and adolescents usually report a specific mechanism of injury, time, and place.
2. • Swelling, discoloration, ecchymosis, and tenderness of the scrotum are common.
3. • Clear transillumination is compromised if a hematoma is present.
4. • Ultrasound differentiates the degree and type of injury and assesses for testicular rupture.

Management

NSAIDs, cool compresses, scrotal support or elevation, and bed rest are modalities used to help relieve pain. An enlarging scrotum merits immediate surgical exploration, as does hematocele.

Epididymitis

Epididymitis is an inflammation of the epididymis that is painful, acute, and commonly caused by *Neisseria gonorrhoeae* or *Chlamydia trachomatis* in the sexually active adolescent, often with infection in the urethra or bladder. It is rare before puberty, but it occurs in younger boys from *E. coli* infection. It may occur in children younger than 2 years old with GU tract abnormalities

History and Clinical Findings

1. • Trauma or sexual encounters within past 45 days
2. • Painful scrotal swelling, usually gradual but may be acute
3. • Dysuria and frequency, or obstructive voiding
4. • Fever, nausea, vomiting

Physical Examination

1. • Scrotal edema and erythema are noted.
2. • The epididymis is hard, indurated, enlarged, and tender; the spermatic cord is tender.
3. • The testis has normal position and consistency.
4. • The cremasteric reflex is normal (not present in older adolescents).
5. • Elevation of testis may relieve pain (in torsion it increases pain).
6. • Hydrocele may be present due to inflammation.
7. • Urethral discharge may be present: purulent in gonorrhea, and scant and watery in chlamydial infection.
8. • Rectal examination reveals prostate tenderness and can produce a urethral discharge.

Diagnostic Studies

1. • UA: Pyuria and occasional bacteria may be present
2. • CBC: Elevated WBC count
3.
4. • Urethral culture and Gram stain: Urine nucleic acid amplification tests for gonococci and *Chlamydia*
5. • Testing for other STIs and HIV if there is a history of sexual activity
6. • Doppler ultrasonography to differentiate torsion of the testis
7. • If the above tests are not diagnostic, refer to urology to identify urogenital problems with evaluation by VCUG, ultrasonography, or both in prepubertal children and in those who deny sexual activity.

Management

Management involves symptom relief and treatment of a causative organism, if found. Bed rest, scrotal support, and elevation are indicated. Apply ice packs as tolerated. Sitz baths and analgesics or NSAIDs relieve pain. Antibiotic treatment includes the following (Centers for Disease Control and Prevention [CDC], 2015):

1. • First line: Ceftriaxone (250 mg intramuscularly one time) plus doxycycline (100 mg twice a day for 10 days).
2. • Alternative treatments: Ofloxacin (300 mg twice a day for 10 days) or levofloxacin (500 mg once a day for 10 days).
3. • Referral to a urologist is indicated if a solitary testicle is involved, if a prompt response to treatment does not occur, or if a question about the diagnosis remains. Treatment of sexual partner(s) from the past 60 days is indicated if caused by an STI. Intercourse should be avoided until cured. Follow-up is needed within 3 days if no improvement or if symptoms recur after treatment. Follow-up after antibiotics is recommended to ensure that no palpable mass remains.

Phimosis and Paraphimosis

Phimosis refers to a foreskin that is too tight to be retracted over the glans penis. Physiologic or primary phimosis occurs over the first 6 years of life when the glans does not completely separate from the epithelium. Pathologic or secondary phimosis occurs after puberty or when the foreskin cannot be retracted after previously being retracted. Paraphimosis is a retracted foreskin that cannot be reduced to the normal position.

Phimosis can be congenital or acquired from infection and foreskin inflammation. Paraphimosis causes penis constriction and results in pain, glans edema, and possible necrosis. Paraphimosis is most common in adolescents and can follow masturbation, sexual activity, or forceful retraction.

History and Clinical Findings

1. • May be a history of infection or inflammation of the penis
2. • Retraction of the foreskin with an inability to reduce it (paraphimosis)

3. • Pain and dysuria
4. • Signs of urinary obstruction—ballooning of the foreskin with urination and/or abnormal intermittent urinary stream

Physical Examination

1. • Phimosis—a tight, pinpoint opening of the foreskin with minimal ability to retract the foreskin; foreskin flat and effaced
2. • Pathologic phimosis—thickened rolled foreskin
3. • Paraphimosis—edema and bluish discoloration of the glans and foreskin

Management

1. • Phimosis: Normal cleansing with gentle stretching of the foreskin until resistance is felt. Most foreskins are retractable by 5 or 6 years old. Never forcefully retract the foreskin. Circumcision is indicated if urinary obstruction or infection is present. Persistent phimosis can be treated with 0.05% betamethasone cream twice daily for 2 to 4 weeks. This frequently allows successful retraction of the foreskin, promotes awareness of improved hygiene, and offers an alternative to circumcision. (Elder, 2016a).
2. • Paraphimosis: Reduction may be accomplished by lubricating the foreskin and glans, simultaneously compressing the glans, and placing distal foreskin traction. If this technique is not successful, surgical release of the constricting band must be done to prevent necrosis of the glans. Paraphimosis is a surgical emergency (Elder, 2016a). Investigation of events leading to the paraphimosis is needed to rule out sexual abuse.

Balanitis and Balanoposthitis

Balanitis is an inflammation of the glans; *balanoposthitis* is an inflammation of the foreskin and glans penis occurring in uncircumcised males or those with phimosis. Debris accumulation under the foreskin, probably resulting from poor hygiene, irritates the foreskin and glans and leads to infection. If purulent discharge with fiery-red erythema and moist translucent exudates are present, consider streptococcal etiology. Normal skin flora is the usual cause of infection, but gram-negative bacteria are possible. If a urethral discharge is present, a sexually transmitted infection (STI) must be considered. Occasionally trauma or allergy can be the cause.

History and Clinical Findings

1. • A fussy infant or pain and dysuria in an older child. Edema and inflammation are noted on the foreskin and glans.
2. • Cultures may help determine infectious causes

Management

Prescribe oral and topical antibiotics as directed by the cultures, along with warm bathtub soaks. Depending on the swelling, topical steroids might also be prescribed.

Diabetes 952-960

Comparison of Type 1 and Type 2 Diabetes in Youth

	Type 1 Diabetes	Type 2 Diabetes
Age at onset	All ages	≥10 years old
Gender	Equal distribution by gender	More frequent in females
Race/ethnicity	Most frequent in non-Hispanic whites May occur in all racial and ethnic groups	More frequent in African Americans, Asians, Native Americans, Hispanics
Obesity	Similar to the general population; not related to type 1 diabetes	>90%
Family history of diabetes	5%-10% have first-degree relative affected	Approximately 80% have first-degree relative affected

Comparison of Type 1 and Type 2 Diabetes in Youth

	Type 1 Diabetes	Type 2 Diabetes
Insulin secretion	Very low	Low, normal, or high
Insulin sensitivity	Normal	Decreased
Onset	Acute, severe	Subtle to severe
Ketosis, DKA	Approximately one-third of new cases	Uncommon
Hypertension	Uncommon	Common
Acanthosis nigricans	Rare	Common
Polycystic ovary syndrome	Rare	Common
Islet autoimmunity	Present	Uncommon

Type 1 Diabetes

Type 1 diabetes is caused by autoimmune destruction of pancreatic beta cells in the islets of Langerhans thought to be triggered by a preceding environmental event in genetically susceptible individuals. This destruction of beta cells results in an absolute deficiency in insulin secretion, reduced biologic effectiveness, or both. Normal metabolic function depends upon sufficient amounts of circulating insulin. Insulin deficiency results in uninhibited gluconeogenesis and a blockage in the use and storage of circulating glucose. Therefore high blood glucose levels are a result of the defective metabolism of carbohydrate, protein, and fats.

The following early symptoms are often reported:

1. • Polydipsia, polyphagia, polyuria
2. • Nocturia, blurred vision
3. • Weight loss or poor weight gain
4. • Fatigue and lethargy
5. • Vaginal moniliasis

As ketoacids accumulate, the following history is reported:

1. • Abdominal pain, nausea and/or vomiting
2. • Fruity-smelling breath
3. • Weakness (caused by dehydration)
4. • Mental confusion
5. • Coma

Physical Exam

1. • Dehydration (child may not look clinically dehydrated unless actively vomiting)
2. • Weight loss or slow weight gain
3. • Muscle wasting
4. • Tachycardia
5. • Vaginal yeast, thrush, or other infection

If ketosis develops:

1. • Slow, labored breathing (Kussmaul breathing)
2. • Flushed cheeks and face
3. • Fruity-smelling breath

Diagnosis Studies:

1. Urine for glucose and ketones
2. • Metabolic screen for acid-base status to exclude DKA
3. • Hemoglobin A_{1c} (HbA_{1c})
4. • Blood glucose
5. • Screen for the presence of pancreatic autoantibodies: This should be considered to confirm the diagnosis of type 1 diabetes, particularly in those cases where there may be uncertainty regarding type

Diagnostic Criteria for Diabetes

The presence of one or more of the following criteria indicates a diagnosis of diabetes:

1. • $HbA_{1c} \geq 6.5\%$
2. • Fasting plasma glucose 126 mg/dL (7 mmol/L) or greater
3. • Random plasma glucose 200 mg/dL or greater plus presence of classic symptoms (polyuria, polydipsia, polyphagia)
4. • Postprandial (2 hours after eating) plasma glucose 200 mg/dL (11.1 mmol/L) or greater

Initial Management

All children with type 1 diabetes should be started on insulin at diagnosis. Children with ketoacidosis should be admitted to the hospital for IV insulin treatment, fluid replacement, and careful monitoring to prevent cerebral edema that, although rare, can cause significant morbidity or mortality. In general, the goal of insulin therapy is to achieve optimal glycemic control typically defined as HbA_{1c} less than 7.5% and/or a fasting blood glucose level between 90 and 130mg/dL.

All children require medical nutrition therapy (MNT) that must match the insulin schedule.

Intensive basal bolus insulin therapy can be delivered using either multiple daily injections (MDIs), or continuous subcutaneous insulin infusion (CSII) using an insulin pump based on patient and family preferences. MDI regimens can be implemented using a long-acting insulin analogue administered typically once a day to provide a steady basal insulin replacement with boluses of rapid-acting insulin at meal and snack times. This regimen requires a minimum of four injections per day.

In CSII therapy, the pump infuses rapid-acting insulin into the subcutaneous tissue through a small, flexible, soft cannula. The cannula is replaced in a new site by the wearer or the family every 2 or 3 days. . Bolus insulin is delivered at meals and snacks. Bolus doses can be calculated by the pump if the user enters the amount of carbohydrate to be eaten and/or his/her blood glucose.

CSII via an insulin pump is particularly useful for delivering small bolus doses of insulin and varying the dose of basal insulin delivered over the 24-hour period.

Insulin Dosages (units/kg/day)

Age	Total Daily Insulin (units/kg/day)
Partial Remission (Honeymoon) any age	<0.5
Prepubertal	0.7-1.0
Pubertal	1.0-2.0

BOX 45.8 Types of Insulin Analogues

Rapid acting
Duration: 3-5 h

1. • Aspart (NovoLog)
2. • Lispro (Humalog)
3. • Glulisine (Apidra)

Short acting
Duration: 5-8 h

1. • Regular insulin

Intermediate acting
Duration: 12-24 h

1. • NPH

Long acting
Duration: Up to 24 h

1. • Detemir (Levemir)
2. • Glargine (Lantus)
3. • Degludec (Tresiba) (duration up to 42 h per manufacturer)

Adjusting Insulin Dosages

Parents and teens can be educated to make insulin adjustments based on blood glucose patterns. They analyze what time of day the blood glucose is consistently outside of the target range (either too low or too high) and adjust the insulin dose that most directly is related to the problematic blood glucose pattern. Usually, parents can safely make up to a 10% adjustment to the basal or bolus insulin dose.

Medical Nutrition Therapy

Nutrition is an essential component of diabetes management. Diets should be healthy and daily calories spread over three meals and snacks. Caloric requirements are based on the child's age, body weight, and activity level. Calories are distributed between protein (15%), carbohydrates (55%), and fat (less than 30% of caloric intake with less than 7% in the form of saturated fats) and account for food preferences, including those pertinent to culture. The meal plan for a child with diabetes should include the same healthy foods recommended for all pediatric patients. The goal is to balance food intake with insulin dose and activity to maintain blood glucose levels within the target range and to prevent both hyperglycemic and hypoglycemic episodes. A pediatric dietician nutritionist is essential to provide ongoing guidance to the child and family.

Exercise

Glucose levels can vary by the type, duration, and intensity of exercise performed. Typically, aerobic exercise, such as long distance running or cycling, results in a decrease in blood glucose levels, whereas anaerobic exercise, such as weight training, sprinting (running or cycling), or jumping, may temporarily increase glucose (Riddell et al., 2017). Therefore, to compensate for the effect of exercise on blood glucose levels, children and families are taught to either decrease the insulin dose or take extra carbohydrates prior to exercise. Anaerobic exercise requires conservative blood glucose correction for higher glucose post exercise as hypoglycemia can still occur later on in the day.

Ongoing Management

Children with type 1 diabetes should be seen every 3 to 4 months, with the visit tailored by age and developmental stage and careful attention paid to diabetes management including:

1. • Self-monitoring blood glucose results
2. • Frequency of hypoglycemia
3. • HbA_{1c}
4. • Physical activities
5. • Emotional adjustment to the disease
6. • Social issues, such as peer pressure
7. • Eating issues: Young women and men with diabetes have an increased incidence of eating disorders, such as "diabulemia" in which insulin dosage is decreased to lose weight (Doyle et al., 2017). Providers should have a high index of suspicion for disordered eating in youth with elevated BMIs and HbA_{1c} level
8. • A physical examination that focuses on:
    1. • Growth and weight gain
    2. • Blood pressure
    3. • Stage of puberty
    4. • Injection site assessment for lipodystrophy
    5. • Clues for other autoimmune disease (thyroiditis and celiac disease)

Recommended Screening Parameters for Youth With Diabetes

		Frequency	
	Measure	**Type 1**	**Type 2**
Glycemic control	HbA_{1c}	• Every 3 months	• Every 3 months (every 6 months if stable)
Thyroid disease	Antithyroid peroxidase and antithyroglobulin antibodies	At diagnosis	• Not required
	TSH	• Every 1-2 years	• Not required

Recommended Screening Parameters for Youth With Diabetes

	Frequency		
	Measure	**Type 1**	**Type 2**
Celiac disease	tTG-IgA, IgA	• At diagnosis • 2 and 5 years later, and as clinically indicated	• Not required
Dyslipidemia	Fasting lipids	• If positive family history, shortly after diagnosis. • Otherwise start screening if ≥10 years of age. • If lipids are normal, repeat every 3-5 years. • If lipids are elevated, screen yearly	• At diagnosis, if ≥10 years of age, repeat every 3-5 years
Nephropathy	Random spot urine albumin to creatinine ratio	• 5 years after diagnosis • Repeat yearly	• At diagnosis, repeat yearly
Hypertension	Blood pressure	• At each visit	
Retinopathy	Dilated eye exam	• Yearly once ≥10 years old OR reached puberty AND diabetes duration of 3-5 years	• At diagnosis, repeat yearly
Neuropathy	Foot exam	• Yearly once ≥10 years old OR reached puberty AND diabetes duration of 5 years	• Yearly
Psychosocial		Screen for emotional well-being at each visit	

Type 2 Diabetes

Clearly, type 2 diabetes in youth is a serious and growing public health problem. Type 2 diabetes begins with increased tissue resistance to insulin, resulting in hyperinsulinemia and hyperglycemia. Although pancreatic beta cells initially produce insulin, hyperglycemia creates an increased insulin demand; with increasing demand for insulin over time, the pancreas loses its ability to effectively secrete insulin. Autoimmune destruction of pancreatic beta cells does not typically occur. During puberty, GH secretion as part of the pubertal growth spurt further increases resistance to insulin action for those predisposed to type 2 diabetes. Adolescents with normally functioning pancreatic beta cells secrete additional insulin to compensate for this puberty-related effect. However, when beta cells do not function properly, metabolic decompensation begins and leads to a state of prediabetes (impaired fasting glucose and/or impaired glucose tolerance) with eventual progression to type 2 diabetes

Screening Guidelines

The symptoms of type 2 diabetes may be absent or subtle, so children at risk should be screened. The American Diabetes Association (2017) provides guidelines for screening children at risk:

1. • Screen if overweight (BMI is greater than 85th percentile for age and gender, or weight is greater than 120% of ideal weight), plus any two of following risk factors:
    1. • Family history of type 2 diabetes in first- or second-degree relative
    2. • Race/ethnicity (Native American, African American, Latino, Asian American, Pacific Islander)
    3. • Signs of insulin resistance or conditions associated with insulin resistance (e.g., acanthosis nigricans, polycystic ovary syndrome, hypertension, dyslipidemia)
    4. • Maternal history of diabetes or gestational diabetes during pregnancy with this child
2. • Screen every 3 years.
3. • Use fasting plasma glucose test following diagnostic criteria for diabetes discussed earlier.
4. • Use clinical judgment to screen for type 2 diabetes in high-risk patients who do not meet these guidelines.

History

The history of patients with type 2 diabetes may include:

1. • Polydipsia, polyphagia, polyuria
2. • Nocturia or bedwetting
3. • Blurred vision
4. • Obesity, especially central
5. • Report of a hyperpigmented, velvetlike rash in skin folds
6. • Frequent or slow-healing infections
7. • Fatigue, symptoms of sleep apnea
8. • History of premature adrenarche
9. • Family history of type 2 diabetes

Physical Examination

The physical examination should include assessment of height, weight, stage of pubertal development, and blood pressure. The following findings may be present:

1. • Dehydration
2. • Overweight (BMI greater than 85th percentile for age and gender) or obesity
3. • Weight loss (less common)
4. • Acanthosis nigricans noted in the axilla, base of the neck, groin, knuckles, and other skin folds
5. • Vaginal yeast, thrush, other infection
6. • Polycystic ovary syndrome symptoms (e.g., acne, hirsutism)
7. • Hypertension

Diagnostic Studies

Screening should be conducted in high-risk children without symptoms (see earlier "Screening Guidelines") and should include (American Diabetes Association, 2017):

1. • Urine for glucose and albumin (can be performed in the office)
2. • Children can have ketoacidosis if they have gone undiagnosed for a long time
3. • Fasting blood sample for blood glucose, HbA_{1c}, lipid panel, TSH and free T_4, and insulin level

Management

Treatment of type 2 diabetes in youth remains in its infancy and currently lacks the strong evidence base of type 1 diabetes. The primary treatment for children and adolescents with type 2 diabetes is education and lifestyle modification, particularly improvement of nutritional practices and physical activity behaviors leading to weight loss. However, in the United States, fewer than 10% of children with type 2 diabetes are successful in achieving glycemic control with diet and exercise alone. If lifestyle changes are not successful in normalizing blood glucose levels, pharmacologic agents should be added to the treatment regimen. Two pharmacologic agents are approved for the use of type 2 diabetes in the pediatric population: metformin and insulin.

As with type 1 diabetes, the treatment plan for children with type 2 diabetes must be individualized. The treatment goal of both type 1 and type 2 diabetes is the same—the normalization of blood glucose values through the achievement of optimal glycemic control ($HbA_{1c} \leq 7.5\%$). This typically requires:

1. • Daily self-monitoring by the child: If the child is treated with multiple insulin injections or continuous pump therapy, check blood glucose levels three or more times each day; for those on less-frequent insulin injections, oral medication, or MNT alone, a daily check may be adequate.
2. • HbA_{1c} and plasma glucose levels monitored every 3 to 4 months for those whose therapy has changed or who are not meeting glycemic goals. Children who are meeting goals and have stable glycemic control can be monitored every 6 months.
3. • Follow-up every 3 to 4 months on lifestyle, nutrition, and other complications of obesity, discussed in more detail later.

4. • Screening and successful control of the associated complications, such as hypertension and hyperlipidemia, are important. See Table 45.5 for screening recommendations.

Lifestyle Changes: Nutrition and Exercise

When discovered early, type 2 diabetes may respond to lifestyle changes, such as alterations in diet and exercise. These changes must be comprehensive and family based. MNT is an important part of the treatment plan. Referral to a registered pediatric dietician nutritionist is essential, with the goals of weight loss and regulating nutritional intake. A low-fat diet, self-monitoring of weight, and being physically active are important components of MNT. Successful weight management may consist of weight maintenance rather than weight loss depending on the child's age and BMI. Changes in family eating patterns can contribute to weight maintenance or loss that may normalize insulin levels. Nutrition counseling should be provided both at the time of diagnosis of type 2 diabetes and as part of ongoing clinical management and should be consistent with guidelines of the Academy of Nutrition and Dietetics.

Inactivity and the increasing obesity epidemic are directly related to the escalating incidence of type 2 diabetes in young people. Physical activity is not only one of the major type 2 diabetes prevention messages, but it is also a critical component in treatment. Youth with type 2 diabetes should be strongly encouraged to be physically active by participating in sports and regular exercise. Daily vigorous exercise (30 to 60 minutes a day) helps to control weight, and even modest weight loss has been shown to reduce insulin resistance

Medication

Metformin is the only oral agent approved by the FDA for use in children with type 2 diabetes, and, although rosiglitazone is used in combination with metformin in adults, the FDA has found insufficient evidence to approve it for pediatric use. Many other medications prescribed for adults with type 2 diabetes are used off label in pediatrics. Metformin decreases the amount of glucose produced by the liver and increases insulin sensitivity of the liver and muscles.

Most children are started on metformin in doses up to 1000 mg twice a day. Metformin rarely causes hypoglycemia, so blood glucose needs to be checked only before breakfast and 2 hours after dinner. Mild gastrointestinal side effects may occur with metformin use but are usually self-limiting. Metformin users can experience vitamin B_{12} deficiency probably secondary to malabsorption, especially with higher doses and longer use. Providers should counsel youth taking metformin to increase foods high in vitamin B_{12}; they should also have a high suspicion of vitamin B_{12} deficiency if clinical signs appear (see Chapter 39). Although there is no consensus on requiring laboratory testing, youth taking metformin, especially long-term users, should probably be assessed regularly for vitamin B_{12} deficits and high homocysteine levels (Thomas and Gregg, 2017). Liver function tests should also be monitored. Should the youth fail to respond to metformin, a combination of two oral agents may be used.

***Epilepsy 990-993

Epilepsy

Epilepsy is a neurologic disorder characterized by recurrent unprovoked seizures. A seizure is an event caused by an abnormal electrical signal arising from within the cerebral cortex. Seizures can present in many different ways. The two primary types of seizures are generalized (arising across the cortex) and focal (arising from one specific area of the cortex). The International League Against Epilepsy (ILAE) has created a system for classification of seizure types and presentation (Fig 46.5). The primary care provider (PCP) should be familiar with the presentation for various types of seizures. The diagnostic criteria for epilepsy is a single unprovoked seizure with a known increased risk for future provoked seizures OR two unprovoked seizures that occur greater than 24 hours apart OR diagnosis of an epilepsy syndrome

History

Historical questioning should include the following:

1. • Description of the seizure: Focal or generalized (if known), semiology (presentation characteristics of the seizure), loss of consciousness, aura, length of postictal sleep or confusion, duration of the episode, history of prior seizures and associated illness or injury
2. • Any underlying medical diagnosis (e.g., diabetes, renal disease, cardiovascular disorder)
3. • Previous CNS infection or birth trauma
4. • Intrauterine infection, trauma, bleeding
5. • Toxic exposure or drug use
6. • AED noncompliance (stopped abruptly or doses missed) or changes in drug manufacturer or change to generic from name brand
7. • Family history of seizures
8. • Any history of developmental or learning delay or regression

Physical Examination

The following should be determined on physical examination:

1. • Weakness or focal abnormalities on the neurologic exam
2. • Presence of seizure activity during the examination
3. • Hypertension (for renal disease)
4. • Signs of systemic disease or cardiovascular disorder
5. • Skin findings suggestive of neurocutaneous syndromes (i.e., café au lait spots, ash leaf spots, hypopigmented macules, or facial hemangiomas)
6. • Signs of head trauma
7. • Transillumination of the skull in infants

For children presenting with staring spells that are questionable for childhood absence epilepsy (CAE), the provider can perform 2 to 3 minutes of hyperventilation using a pinwheel or similar object. Often times, this will provoke a seizure, which is diagnostic for CAE. Appropriate safety precautions should be taken prior to performing this, and the child and family should also be informed of the intent prior to proceeding.

Diagnostic Studies

These are the typical diagnostic test recommendations:

1. • Complete blood count (CBC), including platelets and liver function tests (LFTs)—useful for diagnostic purposes or as a baseline before AED therapy is started.
2. • Metabolic screen may be considered later in the workup, not initially.
3. • Blood glucose—standard in all patients.
4. • Urine and serum toxicology—only if illicit drug exposure is suspected.
5. • Genetic testing—expanding use in epilepsy assessment but not routinely ordered. Need typically determined by specialist.
6. • LP—only if child is younger than 6 months; child of any age with persistent changes in mental status or failure to return to baseline functioning; patients with meningeal signs.
7. • EEG—standard in all children after first unprovoked seizure. An abnormal EEG supports the diagnosis and can be useful in differentiating the type of epilepsy. However, a normal EEG when the child is not seizing does not rule out epilepsy. Video electroencephalogram (VEEG) over 1 to 6 days is another option to help characterize events concerning for seizures.
8. • MRI—imaging studies are not routinely indicated for generalized seizures provided the seizure is followed by a normal neurologic examination and return to baseline mental status. Imaging is recommended: (1) following a focal seizure or if the EEG reveals focal electrographic abnormality; (2) if the patient demonstrates cognitive changes after several hours and postictal focal dysfunction (signs of increased intracranial pressure, such as found with tumors, abscesses, strokes, or vascular malformations); (3) if the seizure lasted more than 15 minutes; (4) in infants younger than 6 months old; and (5) if any new onset of focal neurologic deficit has occurred.
9. • CT scan—used only in emergent cases of marked cognitive, motor, or neurologic dysfunction of unknown etiology
10. • Polysomnography (simultaneous EEG, electromyogram, electrocardiogram [ECG], and electrooculogram) can be useful to assess symptoms occurring during sleep.

Management

Referral

If epilepsy is suspected, refer to a provider with expertise in child neurology for diagnosis and initiation of treatment. The PCP can monitor stable children with epilepsy, including continuing the prescription for their AED, monitoring laboratory studies, and performing case management. However, most neurology providers will perform these tasks at routine follow up visits.

Initial Management of Epilepsy

Medication is generally the first-line treatment for epilepsy. The goal for treating epilepsy should always be two-fold: no seizures and no side effects. AED selection should be individualized based on type of epilepsy but should also account for other factors, such as potential side effects, medical history, gender, and age. Providers should be aware that much of AED use in children is done off-label, but there is a large body of evidence supporting this practice. Medication information including

potential side effects should be reviewed with the patient and family prior to initiating therapy. Antiepileptic Drug Withdrawal- For many children with epilepsy, it may be possible to taper medication if the child is able to go 2 years without a seizure.

Ketogenic Diet

The ketogenic diet is a valuable nonpharmacologic treatment option that should be considered for children with intractable epilepsy (particularly nonsurgical candidates) or as a first-line treatment for specific types of epilepsy (such as GLUT1 transporter deficiency). The ketogenic diet is a very rigid diet that allows for high ratios of fats to protein plus carbohydrates in order to transform the body's primary source of energy from glucose to ketones (ketosis).

Surgical Interventions

Surgical intervention for intractable epilepsy can be another effective nonpharmacologic treatment option. Surgery is now considered earlier in the treatment process than it was in the past. All children with intractable focal epilepsy should be evaluated at a comprehensive epilepsy center to determine if they may be a candidate for surgical intervention. There are a variety of surgical options that can be considered and are individualized based on the patient. These can range from resection of a seizure focus or hemisphere (sometimes curative) to procedures that disrupt pathways in the brain (generally palliative), such as with a corpus callosotomy or multiple subpial transection.

Complications

Status epilepticus (SE) are seizures that may be continuous, or frequent, without recovery between episodes. SE can either be classified as Convulsive SE or Nonconvulsive SE based on clinical presentation. In general, SE refers to a seizure that lasts for greater than 30 minutes or multiple seizures without recovery between events. A child who has Convulsive SE may be at increased risk for morbidity and mortality due to lack of oxygenation, decreased cerebral perfusion, metabolic acidosis, hypoglycemia, hyperkalemia, lactic acidosis, increased temperature, and increased intracranial pressure. Such an occurrence needs to be handled as a medical emergency. SE can be triggered by an acute brain infection, progressive neurologic disease, medication failure or noncompliance, electrolyte imbalance, or rarely, a febrile seizure in an otherwise healthy child without other risk factors. Adverse outcomes can include behavioral problems, acquired intellectual disability, and focal deficits. Administration of a rescue medication in the prehospital setting by parents or caregivers (including school personnel) is recommended. Medications that are commonly used for this purpose include rectal diazepam, intranasal midazolam, or buccal clonazepam.

Leukemia 760-761

The leukemias are a group of malignant hematologic diseases in which normal bone marrow elements are replaced by abnormal, poorly differentiated lymphocytes known as *blast cells.* Genetic abnormalities in the hematopoietic cells take over, resulting in the unregulated clonal proliferation of malignant cells. Leukemias are classified according to cell type involvement (i.e., lymphocytic or nonlymphocytic) and by cellular differentiation. ALL is characterized by preponderantly undifferentiated WBCs.

The leukemias are the most common form of childhood cancer, accounting for up to 30% of all pediatric cancers. Although there have been dramatic improvements in survival for ALL over the past four decades, with outcomes approaching 90% in the latest studies, progress has been slower for myeloid leukemia and certain subgroups such as infant ALL, adolescent/young adult ALL, and relapsed ALL]

ALL accounts for about 80% of childhood leukemia cases, with a peak incidence between 2 and 6 years of age, and 56% of leukemia cases in adolescents (Siegel, 2018). Acute myeloid leukemia (AML) is less common in children than ALL and accounts for about 15% of leukemia cases in children and 31% of those in adolescents.

Clinical Findings

Most of the clinical signs and symptoms of leukemia are related to leukemic replacement of the bone marrow and the absence of blood cell precursors. The child may be anemic, pale, listless, irritable, or chronically tired and may have the following:

1. • A history of repeated infections, fever, weight loss
2. • Bleeding episodes characterized by epistaxis, petechiae, and hematomas
3. • Lymphadenopathy and hepatosplenomegaly
4. • Bone and joint pain

Central nervous system (CNS) symptoms—such as headache, vomiting, or lethargy—are rare at the time of diagnosis but can present due to an intracranial or spinal mass. All these symptoms may be vague or nonspecific; providers must maintain a high index of suspicion for cancer.

Diagnostic Studies

The following are used to diagnose leukemia:

1. • CBC with differential, WBCs, platelet and reticulocyte counts. Thrombocytopenia and anemia are present in most cases. The WBC count may be elevated, normal, or low with varying levels of neutropenia.
2. • A peripheral smear may demonstrate malignant cells.
3. • Bone marrow examination showing an infiltration of blast cells replacing normal elements of the marrow
4. • Chromosomal and genetic abnormalities are found in most children with ALL. The genetic alterations in their leukemic blast cells may include changes in the number of chromosomes and a structure with recurrent translocations and deletions, which provide important prognostic information

Management

Approximately 90% of children diagnosed with ALL can now be cured; they are considered cured after 10 years in remission. Key genetic features are critical factors in the management plan. The treatment program for most types of acute leukemia involves 4 to 6 weeks of induction phase (usually with vincristine, prednisone, and L-asparaginase), with the goal of inducing a complete remission and restoring normal hematopoiesis. This is followed by a consolidation phase of therapy lasting several months and then a maintenance phase for 2 to 3 years. Chemotherapy, CNS therapy (cranial irradiation, which is now reserved only for a high-risk child—those with CNS disease or high WBC counts at diagnosis), or intrathecal administration of chemotherapy and systemic administration of corticosteroids are the key interventions. The use of cranial irradiation as part of therapy is decreasing. For children with ALL who relapse, the need for allogeneic stem cell transplantation is not considered until the second complete remission. However, children with certain chromosomal rearrangements or those who are not in remission by the end of the first induction phase are considered at high risk for relapse; thus transplantation must be considered sooner.

For those with AML, treatment consists of induction chemotherapy, CNS prophylaxis, and postremission therapy. Allogeneic stem cell transplantation from an HLA-matched sibling or parent is considered in the first complete remission for children with high-risk disease.

Long-term sequelae of cancer therapy for ALL have been identified in research studies and include effects on cognition, neuropsychologic functioning, and growth deficiencies and an increased risk for second malignancies, such as AML or lymphoma. CNS irradiation has been linked to learning disabilities and impaired IQ, especially in children younger than 5 years of age who also received intrathecal therapy. As a result, cranial radiation dosages have been reduced, and earlier neuropsychologic testing is recommended. Other documented potential late effects of ALL treatment include congestive heart failure, avascular necrosis, and osteoporosis.

***ADHD, 287/ 433-441

ADHD is one of the most commonly diagnosed disorders in childhood. It is considered a neurodevelopmental disorder because it has a clear neurologic base with symptoms that can profoundly affect the behavior of individuals across many settings in their lives. It is a chronic condition that persists for many into adolescence and adulthood. The symptoms of ADHD affect cognitive, educational, behavioral, emotional, and social functioning and core symptoms include inattention, hyperactivity, and impulsivity occurring at a developmentally inappropriate level observed in at least two settings (home, school, or work) with clear evidence of clinical impairment in social, academic, or occupational functioning. There is a range of severity of symptoms from one individual to the next (mild, moderate, and severe), and the scope and severity of behaviors may change within an individual as maturation occurs. ADHD criteria are listed in Table 30.5 with information about how to differentiate between types in Box 30.2.

Current recommendations for the initiation of pharmacotherapy in children with ADHD include assessment of sleep conditions before initiating the medications. Methylphenidate, which is a first-line FDA-approved medication for ADHD, has been linked to insomnia in short- and long-term clinical trials.

DSM-5 Criteria for Attention-Deficit/Hyperactivity Disorder

A. A persistent pattern of inattention and/or hyperactivity-impulsivity that interferes with functioning or development, as characterized by (1) and/or (2)	
Symptom	Criteria
Inattention (1)	Six (or more) of the following symptoms have persisted at least 6 months to a degree that is inconsistent with developmental level and that negatively impacts directly on social and academic/occupational activities: a. Often fails to give close attention to details or makes careless mistakes in schoolwork, at work, or during other activities (e.g., overlooks or misses details, work is inaccurate) b. Often has difficulty sustaining attention in tasks or play activities (e.g., has difficulty remaining focused during lectures, conversations, or lengthy reading. c. Often does not seem to listen when spoken to directly (e.g., mind seems elsewhere, even in the absence of any obvious distraction. d. Often does not follow through on instructions and fails to finish schoolwork, chores, or duties in the workplace (e.g., starts tasks but quickly loses focus and is easily sidetracked). e. Often has difficulty organizing tasks and activities (e.g., difficulty managing sequential tasks; difficulty keeping materials and belongings in order; messy, disorganized work; has poor time management; fails to meet deadlines). f. Often avoids, dislikes, or is reluctant to engage in tasks that require sustained mental effort (e.g., schoolwork or homework; for older adolescents and adults, preparing reports, completing forms, reviewing lengthy papers). g. Often loses things necessary for tasks or activities (e.g., school materials, pencils, books, tools, wallets, keys, paperwork, eyeglasses, mobile telephones). h. Is often easily distracted by extraneous stimuli (for older adolescents and adults, may include unrelated thoughts). i. Is forgetful in daily activities (e.g., doing chores, running errands; for older adolescents and adults, may include unrelated thoughts).
Hyperactivity/impulsivity (2)	Six or more of the following symptoms have persisted for at least 6 months to a degree that is inconsistent with developmental level that negatively impacts directly on social and academic/occupational activities: a. Often fidgets with or taps hands or feet or squirms in seat. b. Often leaves seat in situations when remaining seated is expected (e.g., leaves his or her place in the classroom, in the office or other workplace, or in other situations that require remaining in place). c. Often runs about or climbs in situations where it is inappropriate. (**Note:** In adolescents or adults, may be limited to feeling restless. d. Often unable to play or engage in leisure activities quietly.

	e. Is often “on the go” acting as If “driven by a motor” (e.g., is unable to be or uncomfortable with being still for extended time, as in restaurants, meetings; may be experienced by others as being restless or difficult to keep up with). f. Often talks excessively. g. Often blurts out an answer before a question has been completed (e.g., completes people’s sentences; cannot wait turn in conversation). h. Often has difficulty waiting his or her turn (e.g., while waiting in line). i. Often interrupts or intrudes on others (e.g., butts into conversations, games, or activities; may start using other people’s things without asking or receiving permission; for adolescents and adults, may intrude into or take over what others are doing).
B. Several inattentive or hyperactive-impulsive symptoms were present prior to age 12 years. C. Several inattentive or hyperactive-impulsive symptoms are present in two or more settings (e.g., at home, school, or work; with friends or relatives: in other activities). D. There is clear evidence that the symptoms interfere with, or reduce the quality of, scial, academic, or occupational functioning. E. The symptoms do not occur exclusively during the course of schizophrenia or another psychotic disorder and are not better explained by another mental disorder (e.g., mood disorder, anxiety disorder, personality disorder, substance intoxication or withdrawl).	

Dyslexia,

Dyslexia refers to difficulties with word recognition, decoding, and spelling. Difficulties in writing include spelling, punctuation, grammar, organization, and clarity of written expression.

ODD (oppositional defiant disorder)

ODD is a pattern of negative, hostile, and defiant behavior that is excessive compared with other children of the same age (APA, 2013). Symptoms often occur in early childhood, from 3 to 7 years old with the disorder typically beginning by 8 years old.

Etiologic factors include many of the parenting and family dysfunctions identified for social aggression. Precursors to the disorder are common in early childhood, especially defiance and negativism. More common in boys before puberty, the gender distribution is approximately equal thereafter.

Clinical Findings

The essential feature of ODD is a recurrent pattern of behavior that is negative, defiant, disobedient, and hostile toward authority figures. Behavior is typically directed at family members, teachers, or peers that the child knows well. The child manifests the following behaviors to an extent that leads to impairment

1. • Actively defies or refuses adult requests or rules
2. • Is argumentative, angry, resentful, touchy, or easily annoyed
3. • Easily loses temper; is vindictive
4. • Blames others for own mistakes or difficulties
5. • Deliberately does things to annoy others
6. • Children often see their own behavior as justifiable, not oppositional or defiant

Management

Attend to the early signs of defiant and oppositional behavior or aggression, or both, and educate parents about positive parenting strategies and exercising consistent, healthy discipline, which is similar to the management of CDs. Because these children typically do not perceive themselves as having a problem and cause distress within the family system, referral for intervention is indicated. As described for CD, parent training programs are more successful if they include information about child behavior in multiple environments (e.g., school and home) and target dysfunctional family processes. Child training groups provide added benefit if combined with parent training groups. Again, collaboration with the school is important. These multiple approaches, conducted simultaneously, are most effective.

Bipolar 432-433

Bipolar disorder, formerly known as *manic depression,* is characterized by unusual shifts in mood, energy, and functioning and may begin with manic, depressive, or a mixed set of manic and depressive symptoms. Approximately 1% to 2% of children younger than age 18 meet the diagnostic criteria for bipolar disorder. The risk of suicide in bipolar depression is the highest of all the psychiatric disorders.

A characteristic pattern usually evolves for a particular person, with manic episodes preceding or following major depressive episodes. Most individuals with bipolar disorder return to a full level of functioning between episodes; 20% to 30% experience persistent mood lability and interpersonal difficulties. Sometimes psychotic symptoms develop after several days or weeks of manic symptoms. Such features tend to predict that the individual with subsequent manic episodes will again experience psychotic symptoms.

Parents who are bipolar are at greater risk for having bipolar children. The most common onset of symptoms occurs between 15 and 19 years old. There is no differential incidence based on race, ethnicity, or gender. Children with ADHD seem to be vulnerable to bipolar illness, or it may be that ADHD is a misdiagnosed early sign of the mania to come. If children are also bipolar, treatment of ADHD with psychostimulants or antidepressants may precipitate a manic episode. Antidepressants in depressed children may also precipitate mania and the onset of bipolar illness.

Clinical Findings

Bipolar disorder in childhood or early adolescence appears to be a different, more severe form of the illness than occurs with late adolescent or adult onset. The early-onset form is characterized by irritability and continuous, rapid-cycling, and mixed-symptom state that may also co-occur with disruptive behavior disorders (e.g., ADHD or CD); features of ADHD or behavior disorder are often early symptoms. Prepubertal and early adolescent bipolar disorder is fairly consistent with no differences according to gender, puberty, or comorbid ADHD. In the late adolescent or adult form, the hallmark features include classic manic episodes, episodic patterns of mania and depression, and relative stability between episodes. Symptoms include the following:

1. • Severe mood changes—extreme irritability or overly elated and silly
2. • Inflated self-esteem or grandiosity—“I am the best in the world at X”
3. • Increased energy and physical agitation
4. • Decreased need for sleep (sleeps few hours or no sleep for days without tiring)
5. • Talkativeness or compulsion to talk; frequent topic changes or cannot be interrupted
6. • Racing thoughts
7. • Distractibility, with attention moving constantly from one thing to another
8. • Increase in goal-directed activity (socially or at school)—get “stuck” on activities and can’t stop doing them
9. • Risk-taking behaviors or activities; taking “more dares”
10. • Hypersexuality in talk, thoughts, feelings, or behaviors
11. • Psychosis—visual or auditory hallucinations
12. • Suicidal thoughts and behaviors in 76% of cases and suicidal attempts in 31% of cases

Most adolescents experience depression as their initial symptom. The most common symptoms of mania include irritable mood and grandiosity, elevated mood, decreased sleep, racing thoughts, poor judgment, flight of ideas, and hypersexuality. With mania, children appear to be the happiest of people, but the happiness and laughter do not match the situation or context. Grandiosity may manifest in efforts to correct teachers or critique their efforts, seeing themselves as above rules and laws, or devoting time to an activity for which they have no

talent. Children's sleep difficulties (a hallmark sign) are reflected in high activity levels before bed (e.g., rearranging the furniture), whereas adolescents need little sleep at all. Risk-taking behavior ranges from children climbing excessively high trees or hopping between rooftops to adolescents driving recklessly and speeding. In adolescents, manic episodes are more likely to include psychotic features and may be associated with school truancy, school failure, substance use, or antisocial behavior. The child or adolescent who has depression but also manifests symptoms of ADHD that seem severe (e.g., extreme temper outbursts and mood changes) should be evaluated by a child behavioral health specialist with experience in bipolar disorder. Symptoms are manifested in relatively age-specific ways.

Assessment for comorbid conditions is important. Anxiety disorders, including panic disorder, affect about half (54%) of patients with bipolar disorder. Other common comorbidities include ADHD (48%), disruptive behavior disorders (31%), and substance abuse (31%).

Management

Referral to a child behavioral health provider is critical. Current recommendations for pharmacologic treatment include the use of mood stabilizers, such as lithium, alone or in combination with antiseizure medications (e.g., valproate, divalproex) and atypical antipsychotics (e.g., risperidone). Neither antidepressants nor stimulants have proven effective. Antidepressant use may potentiate manic responses. The use of lithium must be carefully monitored. The best clinical responses occur when pharmacotherapy is combined with individual and family psychotherapy. Therapy should focus on minimizing comorbidities, enhancing problem-solving and communication skills, and reducing negative self-thoughts. Other nonpharmacologic interventions with proven effectiveness include stress reduction, healthy diet, routine exercise, and developing good sleep hygiene.

Depression

There are three categories of depression that occur during childhood and adolescence: (1) MDD, (2) dysthymic disorder, and (3) adjustment disorder with depressed mood. MDD is defined as either a depressed or irritable mood or a markedly diminished interest and pleasure in almost all of the usual activities, or both, for a period of at least 2 weeks. A dysthymic disorder is characterized by depressed or irritable mood for the majority of days in the past 2 years that is less intense but more chronic than major depressive episodes. Adjustment disorder with depressed mood typically occurs within 3 months after a major life stressor, involves less-severe symptoms, and is relatively mild and brief.

There are multiple subtypes of MDD. Children and adolescents with psychotic depression (e.g., affected individuals hallucinate or have delusions) have a greater incidence of adverse long-term outcomes, resistance to psychopharmacotherapy, and a much higher risk of developing bipolar depression. Atypical depression affects approximately 15% of children with depression and is characterized by hypersomnia, increased appetite, psychomotor retardation, and weight gain. Seasonal affective disorder is most common during the fall and winter months when there is less daylight. Premenstrual dysphoric disorder occurs within a week of menstruation and lasts until a few days after menstruation.

Clinical Findings

Older children and adolescents with depression usually present with symptoms similar to those of adults. School-aged children rarely spontaneously admit to depression symptoms. Parents and teachers may report decreased mood, impaired concentration, inattention, irritability, fluctuating mood, temper tantrums, social withdrawal, somatic complaints, agitation, separation anxiety, or behavioral problems. Males are more likely to have externalizing symptoms (e.g., aggression, acting-out, anger), and females are more likely to have internalizing symptoms (e.g., somatic complaints, feelings of sadness). Females have two times higher depression rates than males after puberty, but prepubertal gender rates are roughly equal. Major depression symptoms represent a persistent change that occurs across settings, activities, and relationships and causes the child distress, impaired functioning, or developmental alteration. Infants and young children may present with failure to thrive, speech and motor delays, repetitive self-soothing behaviors, withdrawal from social interaction, poor attachment, and loss of developmental skills. Infants may not respond to extra efforts to soothe or engage them.

Toddlers and preschoolers may lack energy, be too eager to please others, be excessively or unusually clingy or whiney, and have developmentally inappropriate problems with separation. Preschoolers with MDD may

present with sad or grouchy mood, lack of pleasure in play or activity, poor appetite and weight loss, sleep problems, low energy and activity levels, low self-esteem, or increased death or suicide play or talk. School-age children may be irritable, angry, or hostile or have externalizing behavior, such as hyperactivity, difficulty handling aggression, or reckless behavior. Frequent absences from school, perhaps because of school phobia, or poor performance and other school problems are common. On the other hand, school-age children may have internalizing symptoms, such as boredom, lack of interest in playing with friends, social withdrawal, somatic complaints (e.g., stomachaches, headaches, muscle aches, or tiredness), eating or sleeping disturbances, enuresis, or encopresis. Some children with depression describe themselves in negative terms, whereas others, in an effort to compensate for feelings of poor self-worth, become preoccupied with attempting to please others. Talking directly with the child or adolescent is essential because it is thought that half of depression cases are missed when only parents are interviewed. The following symptoms are common:

1. • Depressed mood: Sad, "blue," down, angry, bored
2. • Loss of interest and pleasure in usual activities
3. • Change in appetite or weight (loss or increase)
4. • Insomnia or hypersomnia
5. • Low energy and fatigue
6. • Difficulty concentrating; indecision
7. • Feelings of worthlessness or inappropriate or excessive guilt
8. • Recurrent thoughts of death or suicidal ideation

A diagnosis of MDD is made if there have been at least 2 weeks of depressed mood or loss of interest and at least four additional symptoms of depression. The symptoms cause considerable distress and impairment in social and academic functioning and cannot be caused by bereavement. Therefore, it is important to assess the following:

1. • Recent life events and losses
2. • Family history of depression or other psychiatric disorders
3. • Family dysfunction
4. • Changes in school performance
5. • Risk-taking behavior, including sexual activity and substance use
6. • Deteriorating relationships with family
7. • Changes in peer relations, especially social withdrawal

Management

The first goals of management are to determine suicidal risk and intervene to prevent suicide. Suicidal risk is greatest during the first 4 weeks of a depressive episode. Patients with acute suicidal intent that includes a plan, psychosis, risk of abuse, and unstable behavior require immediate psychiatric evaluation. Cumulative suicidal risks—prior suicidal behavior or attempts, depression, and alcohol, tobacco, or drug abuse/dependence—require behavioral health intervention as well, and immediate referral must be made. Attention must also be paid to establish a safe environment (e.g., removal of firearms, knives, and lethal medications, including tricyclic antidepressants [TCAs]). Families of adolescents with depression may be noncompliant with recommendations to remove guns from the home in spite of compliance with other aspects of treatment. Vigilant follow-up in this regard is crucial. Other management strategies by the PCP include referral to community resources, such as hotlines, and to identify an emergency plan for the family should the patient become actively suicidal, psychotic, or a danger to others. It is important to note that suicidal ideation often increases during the treatment phase known as *emergence.* Emergence occurs in the first week to month of treatment when the patient's energy levels increase, but feelings of hopelessness and helplessness have not yet receded. A major depressive episode requires intervention by a behavioral health specialist. Unfortunately only about half of all individuals with depression achieve full remission of their symptoms. Therapies typically include CBT in a group or individual psychotherapy format. Group CBT may help adolescents. Often, family therapy or psychoeducation is indicated

Substance Abuse Disorder

Substance use is a precursor to abuse or dependence, and regular use clearly increases the risk for developing a SUD. Substance abuse is a maladaptive pattern of the use of alcohol or drugs manifested in significant impairment or distress. The criteria for substance dependence in the DSM-5 is divided into specific categories (alcohol, cannabis, inhalants, etc.), and in adults it includes tolerance, withdrawal, and compulsive substance use. Instead, substance-related blackouts, craving, and impulsive sexual or risk-taking behavior tend to be more important criteria.

The cause of SUD is multifaceted. Many contributing factors exist, including:

1. • Genetic vulnerability (family history)
2. • Parental substance use
3. • Dysfunctional family relationships (i.e., rigidity, distant relationships, neglect) and negative life events
4. • Psychiatric conditions (e.g., CD, ADHD, depression), low self-esteem, poor body image, ineffective coping (poor emotional regulation, poor problem-solving skills)
5. • Poor sleep hygiene
6. • School failure
7. • Low religiosity
8. • Competitive athleticism

Precipitating life events tend to center around loss of relationships (e.g., parental separation, divorce, or death; death of a close friend) and chronic negative circumstances (e.g., parental substance abuse, maltreatment).

Clinical Findings

Identifying an adolescent's problem with substance abuse requires a careful assessment, conducted with an accepting, nonjudgmental, nonthreatening, matter-of-fact attitude. The covert nature of substance abuse and the dynamic of denial make it crucial to avoid a critical tone

Interviewing the adolescent with the parents is a key strategy for obtaining information about etiologic factors and behavioral, cognitive, emotional, and physical changes that they have observed in the adolescent. However, it is essential that the adolescent also be interviewed alone at every visit to assess mental health and family issues. When talking about substance use with an adolescent, it is important to begin with general questions that are not overly personal. Begin by asking the adolescent about acquaintances or friends who smoke, drink, or use drugs; whether anyone in the family has had problems with these; and what the adolescent does with friends when they get together. It is helpful to ask about experimentation, under what circumstances it occurs, and the adolescent's feelings about it. To obtain a chronologic history of tobacco, alcohol, or drug use, it may be helpful to approach the subject by inquiring about prescription drugs and moving to illicit substances. The key is to remain nonjudgmental to elicit information that will indicate whether the adolescent is experimenting, a regular user, or dependent on substances. Ask about the adolescent's source of drugs or alcohol; the adolescent who uses substances provided by a friend or acquaintance is less advanced than one who purchases them directly. The practitioner should ask, "What? How much? How often? When? How? Where? With whom? Does the patient use substances at parties, home, school, alone, or with friends?"

Teens who screen positive should receive Screening, Brief Intervention, and Referral to Treatment (SBIRT). The CRAFFT questionnaire is an appropriate screening instrument for substance abuse in the primary care setting. Positive responses to two or more items indicate a high likelihood for substance abuse and merits further evaluation and treatment.

Significant behavioral changes that may reflect drug use include the following:

1. • Infants and young children: Excessive crying; poor feeding or failure to thrive; irritability, jitteriness, or excessive lethargy; poor eye contact; sleep disorders
2. • Older children and adolescents: Decreased school performance; lethargy, hyperactivity or agitation, hypervigilance, decreased attention; disinhibition; deviant or risk-taking behavior; repeated absences or suspensions from school; loss of interest in previously enjoyed activities; withdrawal from family and usual friends, or change in friends to those involved in drugs and alcohol; irritability, fighting, or acting out; hypersexuality; exaggerated mood swings; sleep pattern changes or nightmares; altered menstruation; and change in appetite (from anorexia to unusual hunger)

Mood changes include swings from depression to euphoria, nervousness, unreasonable anger, and frequent expressions of hopelessness or failure. Low self-esteem typically characterizes those who abuse substances.

Physical signs that indicate a substance use problem include the following:

1. • Weight loss
2. • Red eyes with inhaled or smoked substances
3. • Hoarseness, chronic cough, wheezing, frequent “colds” or “allergy” symptoms, epistaxis, and perforations of nasal septum with cocaine and inhalant use
4. • Accidents, trauma, injuries
5. • Intoxication
6. • Complete or partial amnesia for events during intoxication with alcohol and date rape drug use
7. • Dilated or constricted pupils
8. • Gynecomastia, irregular periods, small testes with marijuana
9. • Needle tracks occur with intramuscular (IM) steroids or intravenous (IV) use
10. • Generalized pruritus with opiate use
11. • Reflux, diarrhea, gastritis, and constipation with opiate and alcohol use
12. • Perioral sores or pyodermas from huffing and bagging

Diagnostic Studies

Urine toxicology can be helpful to verify adolescent truthfulness, although a positive drug screen result does not indicate substance abuse or dependence; it only indicates substance use. A negative drug screen result does not rule out an SUD. The approximate duration that drugs can be detected in the urine is as follows:

1. • Alcohol—10 to 12 hours
2. • Amphetamines—2 to 4 days
3. • Cocaine and its major metabolite—1 to 3 days
4. • Benzodiazepines—1 day to 1 week
5. • Barbiturates—1 day to 1 week
6. • Opiates—2 to 5 days
7. • Marijuana—up to 30 days

Management

Exposure to tobacco and alcohol and illicit substances begins in early childhood. The pediatric PCP should discuss parental modeling for the use of alcohol, tobacco, and other substances in early childhood during routine well-child visits. It is important to educate school-age children and their parents about substance use and its consequences. For adolescents, a direct assessment and an interview about substance use are essential. Parents should be advised not to involve their child in their own substance use. Something as seemingly innocuous as “getting dad a beer from the refrigerator” gives the child practice in alcohol use.

Substance abuse must be treated, and referral to a substance abuse program is crucial. The initial goal is to help adolescents take positive steps toward changing their substance use and abuse behavior. If the adolescent denies any problem, efforts should focus on helping the adolescent acknowledge problems. Clarifying reported negative consequences, creating doubts about substance use, and raising awareness of the risks related to current use are motivational interviewing strategies that may be helpful. It is important to remain empathic and yet emphasize the adolescent’s responsibility to make healthy choices. If the adolescent does not have chronic use, harm prevention is the goal of the intervention. Guide the adolescent to examine his or her substance use responsibly and identify ways to prevent harmful consequences.

If the adolescent progresses to chronic substance use, a number of options exist. Outpatient or day treatment programs are effective for those who can live and be managed at home. For adolescents with more serious addiction, comorbid psychiatric conditions, or suicidal ideation, residential treatment or hospitalization may be necessary. Given the prominence of family dysfunction and family life events in the cause of the problem, family-based treatment programs are essential. Family treatment, rather than family psychoeducation or family support groups, has been shown to be superior to other modalities. Follow-up assessments should include substance use issues and other predictors of use: stress or negative life events, depression or negative affect regulation, and the presence of positive support within or outside of the family. Self-help or 12-step groups are thought to be an essential element in the recovery process.

Pinworms

Nematode parasite with infestation of intestines and rectum

1. Human pinworm is ubiquitous; adult worm lives in rectum, comes out at night to lay eggs on perianal skin and dies causing pruritus; scratching and finger to mouth contact transfers eggs to intestine; these develop into mature worms and repeat cycle (approximately 2 weeks)

-Found in children of all socioeconomic classes

-Eggs float easily in air and can be swallowed by others

• **Signs and Symptoms**

1. Nocturnal anal itching
2. Vaginal itching (pinworm crawls into vagina)
3. Insomnia (itching)
4. Worm-like "threads"—seen in toilet or on underwear

• **Physical Findings**

1. Excoriation of perianal and perineal area
2. Thread-like worms will be seen on visualization of anus (early morning using flashlight)

• Diagnostic Test/Findings**: Adhesive cellophane tape "paddle" with kits available for parental use;** or can be made with clear Scotch tape and glass slide; prior to arising and bathing, paddle is pressed against anus and then examined for eggs

• **Management/Treatment**

1. Medication (over age 2 and non-pregnant)
    1. Pyrantel pamoate 11 mg/kg one dose (maximum dose 1 g); repeat in 2 weeks
    2. Mebendazole 100 mg single dose (same dose for all ages and weights); repeat in 2 weeks
2. Reassure parents ubiquitous nature of organism; reinfection likely
3. Test other family members and treat at same time if infected
4. Prevention
    1. Keep nails clean and short
    2. Bathing will remove eggs from skin and decrease pruritus
    3. Excellent hand washing

Congenital heart defects in children

Acyanotic- Left-to-right shunting (increase Pulmonary blood flow)

> Atrial Septal Defect- middle of the wall positioning called "Ostium secundum"

- **A hole between the left and right atria is there for fetal circulation but is supposed to close soon after birth. If it fails to close, you get the atrial septal defect.**
- **Asymptomatic**
- **Split S2 heart sound "lub durub, lub durub"**
- **Highest risk (uncommon)-paradoxical embolus. potentially leading to stroke.**
  https://www.khanacademy.org/science/health-and-medicine/circulatory-system-diseases/acyanotic-heart-diseases/v/atrial-septal-defect

> Ventricular Septal Defect

- **A hole in the center of the ventricular wall between the ventricles that did not close after birth.**
- **Usually located in the top of the wall in the membranous septum. Can be located in the muscular septum down lower.**
- **Asymptomatic**
- **Holosystolic murmur**
- **Treatment: usually it is left alone. If large, surgery can be done to repair.**
  https://www.khanacademy.org/science/health-and-medicine/circulatory-system-diseases/acyanotic-heart-diseases/v/rn-ventricular-septal-defect

> Patent Ductus Arteriosus

- **Treatment: Indomethacin or ibuprofen may be given to preterm infants to effect closure when there is significant left-to-right shunt. It is contraindicated and ineffective in term or older infants.**

https://www.khanacademy.org/science/health-and-medicine/circulatory-system-diseases/acyanotic-heart-diseases/v/patent-ductus-arteriosus

Obstruction to ventricular outflow
> Coarctation of the Aorta
> Pulmonary Stenosis
> Aortic Stenosis

Cyanotic - right to left shunting

Decreased Pulmonary blood flow
> Tetralogy of Fallot

https://www.khanacademy.org/science/health-and-medicine/circulatory-system-diseases/cyanotic-heart-diseases/v/tetralogy-of-fallot

> Tricuspid Atresia
> Hypoplastic left heart syndrome

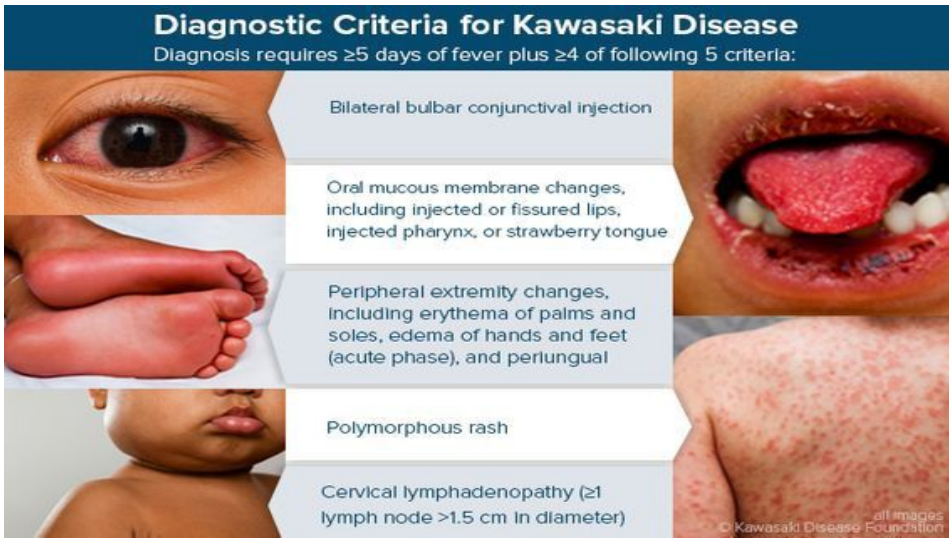
Mixed blood flow
> Transposition of the great vessels

https://www.khanacademy.org/science/health-and-medicine/circulatory-system-diseases/cyanotic-heart-diseases/v/transposition-of-great-arteries

> Truncus arteriosus
> Critical pulmonic stenosis

Kawasaki Disease 560-561

KAWASAKI DISEASE (also known as *mucocutaneous lymph nodesyndrome or infantile polyarteritis*– an acute, febrile, immune-medicated, self-limited disease characterized by vasculitis. Leading cause of acquired heart disease in children



85% <5 years old
most prevalent in Japan

S&S
Stage 1 -Acute (1-2 weeks) – high fever 103-105 for at least 5 days unresponsive to antibiotics, oral mucosa lesions may last 1-2 weeks, perineal rash, non-tender cervical adenopathy, painful rash and edema on feet
Diagnosis requires **fever for 5 days** and 4 of these criteria:

Edema or erythema of hands and feet, conjuctival injection (bilat), cervical adenopathy, rash (non-vesicular and polymorphous), exudative pharyngitis, diffuse oral arythema, STRAWBERRY TONGUE, crusting of lips and mouth

Stage 2 - Subacute (2-8 weeks after onset) – without treatment: desquamation of palms, feet, periungual area, coronary artery aneurysm, joint aches and pains, acute MI may be seen, Pancardidis, diarrhea, jaundice, hepatosplenomagaly, platelet couns >10, 000 000 per mm

Stage 3 – Convalescent – clinical signs have resolved, completed when all lab values are normal, however nail changes include Beau lines (deep transverse grooves across the nails)

It is a fatal disease in small % of children who develop coronary artery problems despite treatment

Differentials:
Group A strep: scarlet fever
Measles
Epstein barr
Toxic shock
Rocky mountain spotted fever
Steven-johnson syndrome
Juvenile RA

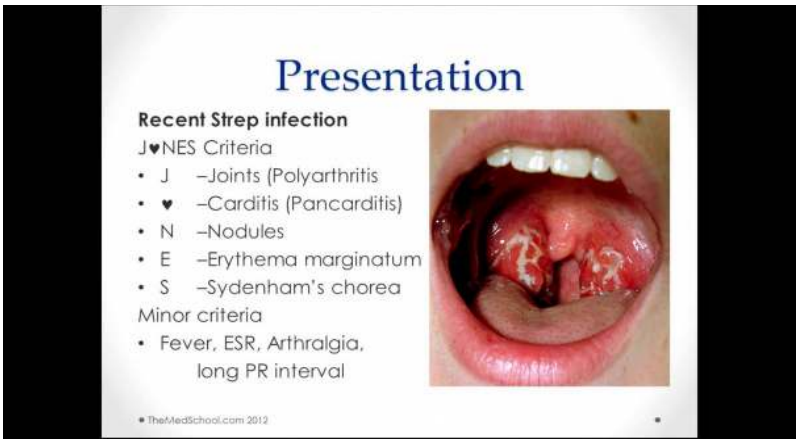
Tests: based on S&S and diff
CBC, anemia, platelets 50% > 450 000
ESR >100
C-reactive protein
EKG – prolonged PR intervals, decrease QRS
Chest Xray – dilated heart, pleural effusion
Pyuria/mild proteinuria

Pharmacology
IVIG (IV immunoglobulin) single dose of 2g/kg for over 12 hours in the first 10 days
Aspirin 80-100 mg/kg/d in 4 doses (Reye's syndrome)

Complications
MI
Development and rupture of coronary artery aneurysm may lead to emboli, HF, heart valve problems, dysrhythmias, myocarditis

Rheumatic Fever 557-559

RHEUMATIC FEVER – An inflammatory disease that develops in 1-3% of children who have **untreated infection with group A strep (GAS).** This can affect the heart, blood vessels, joints , skin, CNS, connective tissues



S&S – hx of pharyngitis 2-4 weeks prior onset of symptoms.

Modified Jones criteria used to diagnose patient:
2 major, or 1 major and 2 minor criteria must be presented as evidence

Major – carditis: 65% have with murmurs
Polyarthritis:75%
Chorea: 15%
Erythema marginatum (macular rash with erythematous border
Subcutaneous nodules

Minor
Fever 101-104F
Artharlgias
Elevated ESR, C-reactive protein
Prolonged PR intervals on EKG

Tests: throat cultures, negative antigen test
ESR, C-reactive protein
ASO tites
EKG
Chest xray
CBC

Treatment: first line Penicillin, if allergic Azithromycin
Prednisone
Aspirin
AHA 2010 no longer recommends prophylaxis treatment for endocarditis in those with rheumatic fever

RSV Bronchiolitis rhinosinusitis virus

Bronchiolitis is a viral disease most often caused by respiratory syncytial virus (RSV), characterized by wheezing. Bronchiolitis often occurs in children less than 2 years of age. Other causes of bronchiolitis include influenza, parainfluenza, adenoviruses, and rhinoviruses. Some rare bacteria can on occasion cause bronchiolitis, such as mycoplasma pneumonia. These diseases are highly contagious and spread through droplet transmission. Children with lung disease, prematurity, immunocompromise, or those in day care are at higher risk of contracting the illness (Marcdante & Kleigman, 2019).
Symptoms follow several days of incubation and begin with URI like symptoms, progressing to worsening cough, rhinorrhea, and wheezing. In infants and young children, irritability may also be present. Upon exam, these children may also show increased work of breathing, prolonged expiration, grunting, intercostal retractions, and nasal flaring. Lung sounds may include wheezing and crackles. Bronchiolitis can be present with or lead to pneumonia. In patients who present with symptoms of respiratory distress, pulse-oximetry measurement, and anterior-posterior and lateral chest radiographs, a full cardiopulmonary assessment is advised.
Treatment is purely supportive and includes monitoring of pulse oximetry, oxygenation, hydration, nutrition, frequent observation.

CROUP- p. 482

Human parainfluenza virus (hPIV), a paramyxovirus, is similar to the influenza and mumps viruses and is an important cause of laryngotracheobronchitis (croup), bronchitis, bronchiolitis, and pneumonia.

There are four antigenic hPIV types. Types 1 and 2 usually strike children 1 to 5 years old and are usually associated with croup; outbreaks are seen more in summer and fall and in odd-numbered years; reinfections occur at any age. Type 3 is endemic, associated more with bronchitis, bronchiolitis, and pneumonia in those younger than 12 months old, results in shorter immunity (a particular problem for immunocompromised patients), and outbreaks peak in the spring and summer (sometimes into fall months). Type 4 infections are less well pathologically and clinically

understood, may be more pervasive than once thought, and can cause mild to severe respiratory illness. By the time most children are 5 years old, they have been exposed to all of the types.

Pathophysiology

This virus spreads by direct person-to-person contact through infected nasopharyngeal secretions or from fomite contamination. It replicates in the superficial ciliated epithelial lining the airways of the upper and lower respiratory tract and spreads readily. The incubation period is 2 to 6 days. Healthy children shed virus for 4 to 7 days before symptom onset and up to 7 to 21 days after resolution of symptoms. The virus lives on nonporous surfaces for up to 10 hours.

Clinical Findings

- **Acute onset of mild fever**
- **Sore throat**
- **Rhinitis**
- **Hoarseness**
- **Barky cough**
- **Lower respiratory involvement symptoms include dyspnea, crackles, wheezing, and hyperaeration. In older children and adolescents, recurrent infection may manifest as a mild URI.**

Diagnostic Studies

Routine testing is not needed. Specific RT-PCR assays are the standard diagnostic test when needed. WBC count may be normal or slightly elevated with a mild lymphocyte elevation.

Treatment

The treatment is supportive. Symptoms management of croup: steroids, nebs, racemic epinephrine

- **Reliable studies using ribavirin are lacking; therefore, aerosolized ribavirin should only be considered for high-risk patients with severe lower respiratory involvement.**
- **Antibiotics are reasonable in cases of severe infection when secondary bacterial invasion is suspected (e.g., otitis media, bronchitis, tracheitis, pneumonia).**
- **No vaccine is available.**
- **Good hand hygiene is important.**
- **Immunocompromised individuals are more prone to developing secondary bacterial infections.**

Asthma-all levels of severity

Asthma is a disease of airway inflammation and hyper-responsiveness that occurs not only due to environmental allergens, but also due to illness or exercise (Marcdante & Kleigman, 2019). It is one of the most common chronic childhood diseases and it poses a significant risk to immediate and long-term health.
The Asthma Action Plan is recommended for all ages. The written document is the primary care providers instructions for self-management of asthma. The Asthma Action Plan should be given to anyone in care of a pediatric child with asthma.

The following charts below provide a reference in how to manage asthma in the pediatric population. For example, a patient comes into the clinic is experiencing asthma symptoms more than 2 days per week, wakes up at night 1-2 times per month, and uses their short acting beta agonist medication more than 2 days a week. As the primary care provider, the recommended step for initiating therapy is Step 2. The Step 2 approach means to recommend a low dose inhaled corticosteroid.

Stepwise approach to managing asthma

Steps	Preferred treatment
Step 1	SABA prn
Step 2	Low dose ICS
Step 3	0-4 years: Medium dose ICS + subspecialist referral ≥5 years: Low dose ICS + LABA or medium dose ICS
Step 4	Medium dose ICS + LABA or montelukast + subspecialist referral
Step 5	High dose ICS + LABA or montelukast + subspecialist referral
Step 6	High dose ICS + LABA or montelukast + OCS + subspecialist referral

Notes

- The stepwise approach is meant to assist—not replace—clinical decision making.
- Before step up, review adherence, inhaler technique, environmental control and comorbid conditions.
- If clear benefit is not observed within 4-6 weeks and/or technique and adherence is not satisfactory, consider adjusting therapy and/or alternative diagnoses.

Acronyms

SABA = Short acting beta agonist

LABA = Long acting beta agonist

ICS = Inhaled corticosteroid

OCS = Oral corticosteroid

ED = emergency department

Classifying asthma severity and initiating therapy

Components of severity		Intermittent	Persistent		
			Mild	Moderate	Severe
Impairment	Symptoms	≤2 days/week	>2 days/week	Daily	Throughout the day
	Nighttime awakenings	0 (≤4 years) ≤2x/month (≥5 years)	1-2x/month (≤4 years) 3-4x/month (≥5 years)	3-4x/month (≤4 years) >1x/week (≥5 years)	>1x/week (≤4 years) Often 7x/week (≥5 years)
	SABA use for symptoms	≤2 days/week	>2 days/week	Daily	Several times per day
	Limitation of normal activity	None	Minor	Some	Extreme
	Lung function*	FEV1>80% FEV1/FVC>85% (5-11 years) FEV1/FVC normal (≥12 years)	FEV1>80% FEV1/FVC>85% (5-11 years) FEV1/FVC normal (≥12 years)	FEV1>60% FEV1/FVC>75% (5-11 years) FEV1/FVC reduced by 5% (≥12 years)	FEV1<60% FEV1/FVC<75% (5-11 years) FEV1/FVC reduced >5% (≥12 years)
Risk	Exacerbations requiring OCS	0-1/year	≥2/6 months (0-4 years) ** ≥2/year (≥5 years)		
Recommended step for initiating therapy***		Step 1	Step 2	Step 3	Step 3 (≤4 years) Step 3 or 4 (5-11 years) Step 4 or 5 (≥12 years)

* Some individuals with smaller lungs in relation to their height (such as a thin individual with narrow A-P diameter to their chest) may normally have FEV1<80% and/or FEV1/FVC<85%. Lung function measures should be correlated with clinical assessment of asthma severity.

** For 0-4 years, ≥4 wheezing episodes per year each lasting >1 day and risk factors for persistent asthma meets risk criteria for persistent asthma.

*** For initial therapy of moderate or severe persistent asthma that is poorly controlled, consider a short course of OCS.

**** Recommended guidelines

Gastroenteritis

The term *acute gastroenteritis* was formerly used to describe acute diarrhea, but this term is technically a misnomer because the etiology of diarrhea does not technically involve the stomach With acute diarrhea, there is a disruption of the normal intestinal net absorptive versus secretory mechanisms of fluids and electrolytes, resulting in excessive loss of fluid into the intestinal lumen. This can lead to dehydration, electrolyte imbalance,

and in severe cases, death in those also malnourished. In children younger than 2 years old, this translates to a daily stool volume of more than 10 mL/kg (this definition excludes the normal breastfeeding stooling of five or six stools per day). In children older than 2 years old, diarrheal stooling is described as occurring four or more times in 24 hours. The duration can last up to 14 days.

Viruses can injure the absorptive surface of mature villous cells, which reduces the amount of fluid absorbed. Some can release a viral enterotoxin (e.g., rotavirus). A loss of water and electrolytes ensues, and there can be volumes of watery diarrhea, even if the child is not being fed. Bacterial and parasitic agents can adhere and/or translocate, causing noninflammatory diarrhea. Bacteria can also damage the anatomy and functional ability of the intestinal mucosa by direct invasion. Some bacteria release endotoxins, whereas others release cytotoxins that result in the excretion of fluid, protein, and cells into the intestinal lumen and an inflammatory response in some cases. Abnormal peristalsis for any reason can result in acute diarrhea. The enteric pathogens are spread through the fecal-oral route and by ingestion of contaminated food or water.

History

1. • Pattern of diarrhea: Onset, number of stools, volume, frequency
2. • Appearance of stool: Odor, mucoid, and/or bloody
3. • Associated symptoms: Abdominal pain, nausea, vomiting, or fever
4. • Number of wet diapers in the past 24 hours and approximate time of last void
5. • Dietary consumption: Changes in diet that might correlate with increased stooling; ingestion (and when) of raw or poorly cooked foods (e.g., raw or undercooked eggs, meat, shellfish, fish, poultry), unpasteurized or under-pasteurized milk or juices, home-canned foods, fresh produce (fruits/vegetables), soft cheeses, deli meats
6. • If given, response to oral rehydration therapy
7. • Food allergies
8. • Family members or close friends with similar illness or other GI diseases
9. • Day care, school attendance, recreational swimming exposure (even if chlorinated): Illness patterns and contacts at these locations; walking in soil without shoes
10. • Travel history: Foreign or coastal areas; camping or travel where untreated water might have been consumed
11. • Attendance at picnics or other outings where food was consumed
12. • Most recent weight and previous growth pattern
13. • Medications: Antibiotics, laxatives, antacids, opiates (withdrawal), vitamins (toxicity)
14. • Pica (metals, plants)
15. • Chemotherapy
16. • Recent surgeries (abdominal)

Physical Examination

1. • Complete physical examination including vital signs and assessment of behavior/mental status changes
2. • Assess for dehydration **Diagnostic Studies**

Diagnostic studies are ordered if the symptoms (discussed earlier) of more serious infection are present. Molecular diagnostic tests (e.g., polymerase chain reaction [PCR]) have greatly improved the ability to diagnose diarrheal illness due to bacteria. The following tests may be ordered:

1. • Stool examination (color, consistency, blood, mucus, pus, odor, volume): In endemic areas, microscopy examination for parasites (e.g., *G. lamblia* and *E. histolytica*).
2. • Stool: pH (<5.5 suggests carbohydrate intolerance typically seen in viral infections), leukocytes (suggest bacterial invasion), reducing substances (viral infections), and occult blood. Normal stool: pH greater than 5.5, carbohydrate negative.
3. • Stool cultures should be considered early in the course of illness for bloody or prolonged diarrhea; in the presence of leukocytes; if clinical signs of colitis are present; for suspected food-borne illness outbreaks (especially with *E. coli* O157:H7); in the immunocompromised; or after recent travel abroad.
4. • Electrolytes, if indicated, to evaluate degree of dehydration and for more serious signs and symptoms of infectious disease.
5. • CBC, as indicated for serious infectious disease.

Management

The foundation of all treatment of acute diarrhea is fourfold:

1. • Restore and maintain hydration and correct/maintain electrolyte and acid-base balance.
2. • Maintain nutrition.
3. • Prescribe antibiotics prudently.
4. • Treat any related conditions, such as sepsis and cardiovascular collapse.

Some adjunct medications and treatments have received wider use in countries outside of the United States and show efficacy in some studies. Some of these include:

1. • Antidiarrheals (antimotility agents or adsorbents) are not generally recommended. However, a review of literature demonstrated that loperamide in children older than 3 years old is safe and decreases the duration and frequency of diarrhea compared with placebo. Children younger than 3 years old and those who are malnourished, those with moderate or severe dehydration, those who are systemically ill, or those who have bloody diarrhea should not be treated with this drug. Some over-the-counter products intended for diarrhea contain salicylates (e.g., Pepto-Bismol), and there is concern for Reye syndrome.

GERD

Reflux of gastric contents into esophagus, due to immaturity of lower esophageal sphincter

1. Occurs in 50% of infants in first 3 months of life, 67% of 4-month-old infants and 5% of 10- to 12-month-old infants.
2. Resolves as child becomes more upright and starts solids
3. Often caused by overfeeding or incomplete burping
4. Childhood diagnoses with higher incidence of GERD include: neurological impairment and delay, esophageal atresia, hiatal hernia, bronchopulmonary dysplasia, asthma, and cystic fibrosis

• **Signs and Symptoms**

1. Effortless, painless spitting of varying amounts, often within 40 minutes of eating
2. No choking or color changes
3. Normal growth—growth chart is key
4. Feeding history may indicate excessive intake; burp heard during vomiting may indicate incomplete burping

1. Reflux may cause other physical complications, such as:
    1. (1) Failure to thrive (FTT)—caused by long-term, forceful regurgitation
    2. (2) Esophagitis—causing irritability, anemia, and guaiac positive stools or hematemesis; dysphagia
    3. (3) Aspiration—pneumonia, wheezing, apnea
    4. (4) Sandifer syndrome—abnormal posturing of head and neck
2. May be "silent GER"—no overt vomiting, but complications may be presenting symptom
3. 60% show improvement by 16 months; 30% may remain symptomatic up to 4 years

• Physical Findings

1. May have wheezing or respiratory symptoms with aspiration
2. Abdominal examination normal—no masses, olive, or peristaltic waves
3. Neurological examination normal—no signs of increased ICP

Diagnostic Tests/Findings

1. Diagnosis often made by observation and history; testing only to determine if reflux is causing problems since vomiting indicates reflux
2. UGI to ligament of Treitz
3. pH probe—indicates amount of reflux occurring;
4. Upper endoscopy.

5. Scintigraphy—"Milk scan;" evaluates for slow gastric emptying and aspiration;
6. Guaiac stool/emesis—positive for occult blood if abnormal
7. Empiric medical therapy—trial of medication to relieve specific symptoms. If symptom is relieved with medication then GER can be determined to be the cause

Management/Treatment

Conservative therapy

1. Positioning—(most important) post- prandial, prone position for 1 to 2 hours if infant can be observed; infant seats/ swings worsen by increasing intra- abdominal pressure; caution with diapering/playing postfeeding
2. Breastfeeding or predominately whey formula
3. A 1 to 2 week trial of hypoallergenic formula may be warranted if vomiting or other symptoms severe enough to con- sider drug treatment
4. Thickening agents such as rice cereal have not been proven to decrease reflux but may decrease vomiting
5. Avoid over-feeding—age in months plus 3 equals number of ounces every 3 to 4 hours for most infants to age 5 months
6. Small feedings with frequent burping
7. Reassure parents with growth charts

Medications: (if conservative therapy has failed)

1. H2 blockers (1st line), proton pump inhibitors (2nd line), and antacids (short term) if irritable from esophagitis (aluminum-containing antacids increase plasma aluminum levels in infants; there are no studies on safety of magnesium or calcium carbonate preparations, antacids should not be used long term)

Genu Varum

Alignment of the knee with the tibia medially (varus) in relation to the femur. Bowlegged appearance as result of uterine position. Measure distance between knees with feet together, distance should be less than 5 inches. An angular deformity is physiologic and normal up to 3 y.o. Pathologic if more than 15 degrees. Associated with short stature, rapidly progressing

Treatment Brace/splint, NSAIDs for pain, and strengthening exercise. If rickets-prescribe Vitamin D. If overweight- weight management.

Genu Valgum

Alignment of the knee with the tibia laterally deviated (valgus) with relation to the femur. Commonly known as knock-kneed. Physiologic (over the first 3 years, 10-15 degrees, more common in girls) or pathologic (before age of 2 y.o, valgus angle >15 degrees, increasing in severity. Associated with short stature, obesity and asymmetry, due to metaphyseal dysplasia or injury).

Treatment Braces for angles more than 15 degrees. In some cases, resolves spontaneously by age 6. Refer to orthopedic for suspected pathology.

Cholesteatoma

Complications of AOM

Persistent AOM, persistent OME, TM perforation, OE, mastoiditis, cholesteatoma, tympanosclerosis (Fig 36.6), hearing loss of 25 to 30 dB for several months, ossicle necrosis, pseudotumor cerebri, cerebral thrombophlebitis, and facial paralysis are possible complications.

Legg-Calve Perthes Disease (LCPD)

Avascular necrosis of the femoral head epiphyses associated w/ trauma, transient synovitis, coagulation abnormalities. Associated with low birth weight and socioeconomic status, and white race. Insidious onset of painful limp of thigh, knee, hip, worst with activity and not relived by rest. Restriction of voluntary motion limited passive motions, abduction and rotation of affected hip, atrophy of thigh or calf may be noticed.

Diagnostics: Hip X-ray, MRI

Treatment Abduction brace or long leg cast, surgery for bone reconstruction

Idiopathic Scoliosis 897-899

1. Idiopathic: Etiology is unknown and is likely multifactorial. It is the most common type of scoliosis. There are three types classified by age at onset:
    1. • Infantile (0 to 3 years of age)
    2. • Juvenile (3 to 10 years of age)
    3. • Adolescent (11 years of age and older)

Idiopathic scoliosis is the most common type. Its etiology is unknown, but it often has a familial or genetic pattern. The overall incidence of idiopathic scoliosis is approximately 2% to 3% with between 0.3% and 0.5% of children with scoliosis having curves greater than 20 degrees on radiography and less than 0.1% demonstrating curves greater than a 40-degree Cobb angle.

Treatment decisions are based on the natural history of each curvature. Infantile scoliosis can resolve spontaneously; however, progressive curves require bracing and surgery in an attempt to slow the curve progression and prevent complications (e.g., thoracic insufficiency syndrome). The treatment goal is to delay spinal fusion, allowing time for the pulmonary system and thoracic cage to have matured and maximal trunk height to be achieved. (See the various surgical procedures described in the following section.)

Observation is always indicated for curves less than 20 degrees. Bracing or surgery may be indicated for larger curves. Brace treatment may reduce the need for surgery, restore the sagittal profile, and change vertebral rotation. Indications for bracing are a curve of more than 30 degrees. Additional indications for brace therapy include skeletally immature patients with curves of 20 to 25 degrees that have shown more than 5 degrees of progression. The efficacy of bracing for adolescent idiopathic scoliosis remains controversial. Some studies show brace treatment to be effective in preventing progression; however, it has been found that the success of treatment is proportional to the amount of time that the patient wears the brace. Various brace treatment protocols suggest wearing a brace as much as 23 hours per day; therefore compliance is a significant factor for this treatment modality.

Surgical treatment is indicated for children and adolescents who do not respond to bracing and for those with curvature exceeding 45 to 50 degrees. There are various surgical procedures; all aim to control progressive curvatures. In the past, surgery was limited to arthrodesis (surgical fusion) of the spine. Referral to an orthopedist or a center that specializes in working with infants and children with scoliosis is essential.

Dysplasia of the hip

DDH-or congenital hip dysplasia

Due to abnormal dislocation of the hip resulting in limited abduction of the hip. Signs and symptoms: thigh fold asymmetry, and leg length inequality. In walking children-painless limp. Diagnostics: Positive Barlow maneuver, Ortolani or Allis signs, US for <4 mo, X-ray AP of pelvis.

Treatment

Pavlik harness, surgical reduction. Excellent prognosis.

Scoliosis

Lateral curve of spine. High incidents, greater in adolescent girls, onset in pubertal

Treatment

Functional Scoliosis: 5-10 degree curvatures are functional and are monitored.

Structural scoliosis: mild-curvature <20 degree-closely observe every 3-4 mo during growth spurts. Serious cases with 25-40 degrees, refer to ortho, Milwaukee brace for 23 hrs a day. Back exercise, spinal fusion - those may create psychological (negative body image, depression) and social issues (limited sport and group participation). Use cognitive-behavior therapy. Functional scoliosis is self-limiting, structural scoliosis produces prominent scapula, ribs, uneven shoulder and asymmetric waistline or back pain. F/U annually during well child exam.

Osgood-Schlatter

Aseptic necrosis of the tibial tubercles and apophysis. May be caused by recent physical activity like running track or playing soccer/football (overuse). Pain is increased during and after activity. Pain goes away once activity has stopped. Often seen in adolescent years, after rapid growth spurt. More common in males than females. 10-15 years of age in boys, and 8-12 years in girls.

Signs and symptoms:
Painful swelling of tibial tubercle, limp, intermittent pain over months, hip pain may be aggravated by extension of knee against resistance, worsens with squatting, stair walking, forceful contraction of the quadriceps, usually due to overuse injury associated with athletic activity during rapid growth of tibial tuberosity, relieved by rest.

Physical Exam Findings:
- Patients may complain of pain by extending the knee against resistance, stressing the quadriceps, or squatting with the knee in full extension.
- Focal swelling, heat, and point tenderness found at the tibial tuberosity
- May palpate a bony prominence over the tibial tuberosity.
- Full range of motion of knee

Diagnostics:
Usually based on history and physical. X-ray or MRI may be done if there is an osteochondral lesion.

Treatment:
Osgood-Schlatter disease is a self-limiting condition, with symptom management the key consideration. Rest, modified activity, ice, NSAIDs, quadriceps strengthening and stretching, tibial band during activity. Neoprene sleeve can be used for stabilization

Febrile Seizures

- Most common seizure in children and can occur in up to 5% of children.
- The fever is typically 38 degrees Celsius and can occur before or after the seizure occurs.
- Febrile seizures can occur between 6-60 months.

Simple febrile seizure: generalized and lasts less than 15 minutes.
Complex febrile seizures: can present as generalized or focal seizures, and last longer than 15 minutes, and/or with clustering (multiple seizures without recovery in between).

Etiology:
- Unclear

- Excludes seizures that are caused by intracranial illness or related to other underlying CNS problems.
- Risk is high for children who have family history of febrile seizures or with predisposing factors, such as NICU for more than 30 days, developmental delay, or attending day care.
- Recurrence of febrile seizure: if 1st febrile seizure was before 18 months of age, low temp at the time of seizure, fever after seizure, or family history.

History:
- Description such as duration, type (generalized or focal), frequency in 24 hours.
- Temperature level, abnormal neurological findings, family hx, maternal smoking in perinatal period, prematurity, NICU > 28 days, parent's perception of child's development.

Physical Exam Findings:
- weakness or focal abnormalities on neuro exam
- Presence of a seizure activity upon exam
- May see HTN (renal disease), signs of systemic or cardiovascular disease
- Head trauma
- Transillumination of the skull in infants

Diagnostic studies:
- LP to r/o bacterial meningitis or CNS infection
- EEG and MRI for complex febrile seizures. MRI for focal exam findings.

Management:
- A(airway), B(breathing), C's(circulation) if seizure is still occurring
- Place child inside lying position to prevent aspiration
- Put NOTHING in patient's mouth
- Time the duration of the seizure. If seizure lasts longer than 5 min, call 911.
- Acetaminophen or ibuprofen after seizure has stopped (PO or rectally)
- Rescue medication (rectal diazepam, intranasal midazolam, or buccal clonazepam) can be used in prehospital settings for prolonged febrile seizures or cluster seizures.

Education:
- Education should be provided on risks, first aid, and management. Education should also include recurrence and reassurance that nothing can be done to prevent the seizures. No long-term consequences occur with simple and most complex febrile seizures.

Testicular torsion

The result of twisting of the spermatic cord, which subsequently compromises the blood supply to the testicle.

· There is a 6-hour window before significant ischemic damage and alteration in spermatic morphology and formation occurs.
· Testes rotate and blocks blood and lymphatic flow
· Can occur after physical exertion, trauma, or on arising
· It can also occur at any age
· Most common in adolescence
· Uncommon before 10 years of age
· Left side is twice as likely to be involved because of longer spermatic cord

Signs and symptoms:
· Sudden onset of unilateral scrotal pain, often associated with nausea/vomiting
· There may be a history of testicular pain.
· Minor trauma, physical exertion, or onset of pain on arising
· Patient may complain of abdominal or inguinal pain for a child who is embarrassed
· There could be a fever or none at all

Physical Exam Findings:
· Patient may appear ill, anxious, and resisting movement

· Gradual but progressive swelling of the involved scrotum
· Involved scrotum will be red, warm, and tender
· "BLUE DOT" sign – subtle blue mass that is visible through the scrotal skin
· One testis is larger than the other, lying transversely, exquisitely painful
· Spermatic cord may be thickened, twisted, and tender
· Elevation of testes increases the pain
· Cremasteric reflex is absent on side of torsion
· Neonate: hard, painless, mass with edema or discolored scrotal skin

Diagnosis:

· Doppler ultrasound or testicular flow scan if doppler WNL
· UA is typically WNL

Management:

· Surgical emergency (within 6-12 hours to prevent retorsion, preserve fertility, and prevent abscess, and atrophy).
· Occasional manual reduction can be performed
· Contralateral orchiopexy may also be done

Education, Prevention, and Prognosis:

· Can result in necrosis if torsion persists for more than 24 hours.

Wilm's Tumor

- Most common malignancy of the GU tract. Ages 2-5 years old. Higher in African Americans, lower in Asian
- Firm, smooth mass in the abdomen or flank that **DOES NOT cross the midline**
    - Stage 1- limited to kidney and can be completely excised
    - Stage 2 – extends beyond the kidney but can still be completely excised
    - Stage 3- post-surgical residual non-hematogenous extension confined to the abdomen
    - Stage 4- hematogenous metastasis, frequently to the lung
    - Stage 5- bilateral kidney involvement
- An important feature of Wilms tumor is that it is associated with congenital abnormalities including renal, cryptorchidism, hypospadias, duplication of the collecting system, ambiguous genitalia, hemihypertrophy, aniridia, cardiac abnormalities, and Berkwith- Wiedemann, Denys-Drash, and Perlman syndromes
- **S/S**- increased abdominal size or palpable mass
- **Diagnostic Studies-** Chest and abdominal can be performed to differentiate neuroblastoma, abdominal ultrasonography to differentiate a solid from cystic mass or hydronephrosis, UA shows hematuria in 25-33%, CT to stage disease
- **Differential-** Neuroblastoma but this will cross the midline.
- **Management**- based on stage and patient condition. Surgery, chemo and radiotherapy are options
- **Complications-** lungs and liver are most common sites of metastasis, HTN b/c of renal ischemia
- **Prognosis-** 80-90% for infants with stage 4, re-occurrence- less than 50% response to chemo

Turner Syndrome – (Monosomy X) – (sex chromosome disorder)

Common clinical findings-

- Short stature, short neck with webbing, low posterior hairline, posteriorly rotated ears, ptosis, short $4^{th}/5^{th}$ metacarpals, short legs, hyperconvex nails, bicuspid aortic valve, COA, hip dysplasia, scoliosis, horseshoe kidney, chronic OM with conductive hearing loss, delayed puberty/infertility

Primary Care Concerns

- Monitor growth- short stature is expected, GH tx is typically begun early (4-5 yrs)
- Nonverbal (math) learning disabilities are common

- Annual hearing exam, recurrent otitis media, progressive mid frequency sensorineural hearing loss
- Ongoing vision assessment- strabismus
- Early onset osteo-penia/ -porosis, vitamin D supp, appropriate estrogen therapy, exercise
- Monitor BP (HTN)
- Annual thyroid screen (Hyper/ hypo)
- Ongoing assessment for celiac disease (tissue transglutaminase immunoglobulin A)
- Careful early monitoring for kyphosis, scoliosis, lordosis
- Increased risk for hyperlipidemia, cardiac defects (aortic root dilation, bicuspid aortic valve, coarctation of aorta (35%), renal anomalies, pulmonic stenosis
- Supplemental estrogen therapy for sexual development and preservation of bone mineral density
- Tendency to form keloids

Down Syndrome- Trisomy 21

Common Clinical Findings

- Short stature, brachycephaly, midface hypoplasia with flat nasal bridge, brushfield spots, epicanthal folds with up-slanting palpebral fissures, small mouth with protruding tongue, Myopia/cataracts, small ears/narrow canals, extra skin at nape of neck, lax joints, short broad hands/feet/digits, single palmar crease, clinodactyly, exaggerated space/plantar groove between great and second toes, congenital heart disease, at risk for leukemia, hypothyroidism, and Alzheimer disease

- Intellectual/cognitive disability/ developmental delays, hearing loss, hypotonia (infant)

Primary Care Issues

- Careful review of newborn screen or hypothyroid
- Careful review of newborn critical congenital heart disease
- Ongoing ophthalmologic exam for cataracts
- Increased risk for duodenal atresia
- High risk for atlantoaxial instability
- Monitor for neurological conditions (infantile spasms, seizures, moyamoya malformation)
- Systemic screening for celiac disease
- Increased risk for leukemia