# Pediatrics Summary

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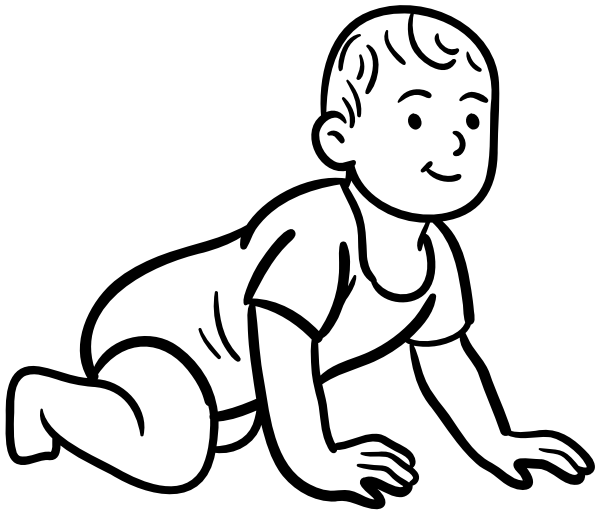

## Resources:

- Slides
- Final PP

# Cardiology

## 1. Fetal and Transitional Physiology

- **Fetal Circulation:** In utero, the **umbilical vein** carries the highest oxygenated blood from the placenta to the fetus. Blood shunts from the right atrium to the left atrium through the **foramen ovale**. The **ductus arteriosus** directs blood from the pulmonary artery to the aorta, bypassing the high-resistance fetal lungs.
- **Transition at Birth:** After birth, pulmonary vascular resistance drops, the foramen ovale closes, and the ductus arteriosus typically closes within days. Abnormal persistence of these structures leads to congenital heart disease (CHD).
- **Normal Vital Signs:** Heart rate decreases with age, while blood pressure is lowest at birth and increases over time. A heart rate of 165 in a crying 6-month-old is normal, but a rate of 250 in a newborn is a sign of pathology (likely SVT).

## 2. Acyanotic CHD (Volume Overload / Left-to-Right Shunts)

These lesions involve blood shunting from the left (high pressure) to the right (low pressure) side of the heart, causing pulmonary overcirculation.

- **Ventricular Septal Defect (VSD):** The most common CHD. It typically causes a **holosystolic murmur** at the left sternal border. Symptoms like tachypnea, poor feeding, and failure to thrive (FTT) usually appear between **4-8 weeks** of life as pulmonary resistance drops. A **louder** murmur often indicates a smaller (and potentially closing) defect.
- **Patent Ductus Arteriosus (PDA):** Characterised by a **continuous "machinery" murmur** at the left subclavicular area. It causes **bounding pulses** and a **wide pulse pressure** (e.g., 100/40 mmHg). On X-ray, a large PDA shows cardiomegaly and left-sided chamber enlargement.
- **Atrial Septal Defect (ASD):** Often asymptomatic in childhood. The classic sign is a **fixed, wide splitting of the second heart sound (S2)** and a mid-systolic murmur at the left upper sternal border. ASD causes dilatation of the **right atrium and right ventricle**, *not* the left ventricle. Complications like Eisenmenger syndrome are rare in childhood and usually occur much later.

## 3. Acyanotic CHD (Pressure Overload / Obstruction)

- **Coarctation of the Aorta (CoA):** A narrowing of the aorta. The hallmark physical finding is a **radio-femoral pulse delay** and lower blood pressure/weak pulses in the legs compared to the arms. It causes left ventricular hypertrophy (LVH) on ECG. In older children, X-rays may show **rib notching** due to collateral vessel development.
- **Aortic Stenosis (AS):** Presents with a systolic murmur at the right upper sternal border radiating to the **suprasternal notch or neck**. An ejection **click** is often heard.

## 4. Cyanotic CHD (The "5 T's")

- **Transposition of the Great Arteries (TGA):** The most common cause of cyanosis **at birth**. It often presents with **no murmur**. The X-ray classically shows an **"egg on a string"** cardiac shadow with a narrow mediastinum.
- **Tetralogy of Fallot (TOF):** The most common cyanotic CHD overall. It consists of four features: VSD, overriding aorta, RVH, and pulmonary stenosis (PS). The murmur is due to the **PS**. X-ray shows a **boot-shaped heart** with dark (oligemic) lungs. "Tet spells" (increased cyanosis) can be triggered by crying or exercise; **squatting** helps by increasing systemic resistance.
- **Tricuspid Atresia:** Often suggested by the presence of **Left Axis Deviation (LAD)** on an ECG in a cyanotic infant.
- **Total Anomalous Pulmonary Venous Return (TAPVR):** Classically shows a **"snowman"** appearance on X-ray.
- **Truncus Arteriosus:** Presents with mild cyanosis, increased pulmonary blood flow, and a wide pulse pressure.

## 5. Duct-Dependent Lesions and Catastrophic Shock

Certain severe defects (Critical AS, Critical CoA, Hypoplastic Left Heart Syndrome) require a patent ductus arteriosus (PDA) to maintain systemic blood flow.

- **Presentation:** When the PDA begins to close, the infant develops **catastrophic heart failure or shock**, mimicking sepsis. Signs include mottled skin, weak pulses, metabolic acidosis, and oliguria.
- **Emergency Management:** For any unstable infant with suspected cyanotic or duct-dependent CHD, the first step is to administer **Prostaglandin E1 (PGE1/Alprostadil)** to keep the ductus open.

## 6. Heart Failure (HF) in Children

HF occurs when the heart cannot meet the body's metabolic demands.

- **Clinical Picture:** In infants, HF presents as tachypnea, **diaphoresis (sweating) during feeds**, irritability, and poor weight gain (FTT). Physical signs include **hepatomegaly**, a **gallop rhythm (S3)**, and displaced apical impulse.
- **Management:**
    - **Nutritional support:** High-calorie feeds.
    - **Diuretics (e.g., Lasix):** Reduce congestion. *Note: MCQ 62 states that Lasix use can lead to hyponatremia and hypokalemia, but metabolic acidosis is NOT a typical complication (it usually causes alkalosis)*.
    - **Inotropes (e.g., Dopamine, Digoxin, Milrinone):** Increase heart contractility. **Propranolol** (a beta-blocker) is *not* used to increase contractility.
    - **Afterload reducers:** ACE inhibitors.

Summary Table for X-ray Findings

| **Disease** | **Classic X-ray Finding** |
|---|---|
| **TOF** | Boot-shaped heart, decreased pulmonary markings |
| **TGA** | Egg-on-side shape, narrow mediastinum |
| **TAPVR** | Snowman appearance |
| **CoA** | Rib notching (in older children), cardiomegaly |
| **VSD/PDA** | Cardiomegaly, increased pulmonary markings |

# Rispiratory

## I. Upper Respiratory Tract Infections (URTI) & Sinusitis

- **The Common Cold:** Primarily a viral illness (most common: **Rhinoviruses**) characterized by rhinorrhea and nasal obstruction without severe systemic symptoms. Children average **6–7 colds per year**. Physical findings are limited to the upper respiratory system, such as swollen erythematous turbinates.
- **Acute Bacterial Sinusitis:** Usually follows a viral URTI where inflammation blocks drainage.
    - **Clinical Diagnosis:** It is diagnosed based on a persistent history of RTI (cough or nasal discharge) for **>10–15 days** without improvement, or **severe symptoms** (fever >39°C and purulent discharge for 3–4 days). **Thick mucus secretions** are a hallmark.
    - **Pathogens:** *S. pneumoniae* (30%), nontypable *H. influenzae* (20%), and *M. catarrhalis* (20%).
    - **Management: Amoxicillin** is first-line. CT is **not required** for simple diagnosis but is used if orbital or intracranial complications (like brain abscesses) are suspected.
- **Nasal Physiology:** In infants, the **nasal airway** is the major contributor to total respiratory resistance during spontaneous breathing. A **unilateral purulent discharge** with a foul smell in a child who does not respond to antibiotics strongly suggests a **nasal foreign body** rather than sinusitis.

## II. Pharyngitis and Neck Abscesses

- **Pharyngitis:** Caused by viruses or **Group A Beta-Hemolytic Streptococcus (GABHS)**.
    - **Treatment: Penicillin** is used primarily to **prevent Acute Rheumatic Fever**. Note that while antibiotic treatment prevents rheumatic fever, **acute glomerulonephritis** (AGN) can still follow streptococcal infections of both the pharynx and the skin.
- **Retropharyngeal Abscess:** Occurs mainly in children **under 3–4 years**. Symptoms include fever, drooling, neck stiffness (**torticollis**), and a bulge in the posterior pharyngeal wall.
- **Peritonsillar Abscess:** More common in adolescents; presents with a **"hot potato" voice**, trismus (difficulty opening the mouth), and displacement of the uvula.

## III. Stridor and Upper Airway Obstruction

- **Acute Stridor:**
    1. **Croup (Laryngotracheobronchitis):** The most common cause of acute stridor, usually caused by **Parainfluenza virus**. Characterized by a **barking cough**, hoarseness, and inspiratory stridor that is **worse at night** and with agitation. The X-ray shows the **"Steeple sign"** (subglottic narrowing). Treatment involves **oral/IM dexamethasone** and **nebulized epinephrine**.
    2. **Epiglottitis:** A life-threatening emergency caused by ***H. influenzae* type b** (incidence decreased by vaccines). Features: sudden onset of high fever, **drooling**, and a **"tripod position"**. Hallmark: **"Thumb sign"** on lateral X-ray. **Crucial:** Never examine the throat or provoke anxiety in these patients, as it can cause total airway obstruction; they must be **intubated in a controlled environment** (theatre/ICU).
    3. **Bacterial Tracheitis:** A serious superinfection (usually ***S. aureus***) that follows a viral croup-like illness, characterized by high fever and a **toxic appearance**.
- **Chronic Stridor:**
    - **Laryngomalacia:** The **most common cause of chronic stridor**. The inspiratory stridor **worsens in the supine position**, when crying, or during URTIs, and improves in the prone position.
    - **Tracheomalacia:** Causes **expiratory stridor** due to tracheal collapse.
    - **Vocal Cord Paralysis:** Often associated with **biphasic stridor**.

## IV. Bronchiolitis and RSV

- **Etiology:** Primarily caused by **Respiratory Syncytial Virus (RSV)** in infants.
- **Clinical Features:** Tachypnea, wheezing, and **bilateral hyperinflation** on CXR. In infants, RSV can also cause **apnea** and otitis media, but it is **not a cause of asthma** (though it may increase future risk of reactive airway disease).
- **Management:** Treatment is **supportive** (oxygen, fluids). **Bronchodilators and steroids are not recommended**. **Secondary bacterial infection** is an uncommon complication.

## V. Bronchial Asthma Management

- **Diagnosis and Monitoring: Spirometry** shows **decreased FEV1** and increased Forced Residual Capacity (FRC) and Residual Volume (RV) due to air trapping. A **Skin Prick test** for inhaled allergens is the best investigation to evaluate the **predictive risk of future asthma** in a wheezing child.
- **Triggers: Upper respiratory infections** are the most common trigger for asthma in children aged 0–4 years.
- **Acute Exacerbation:**
    - **Management:** First-line is **inhaled SABA** (e.g., Albuterol) plus **systemic corticosteroids** (Oral/IV).
    - **Severity:** A **silent chest** (no air movement) and **PaCO2 retention** (hypercarbia) are the most concerning findings, indicating impending **respiratory failure**.
- **Long-term Control:** Controller therapy (like ICS) should be considered if the patient uses rescue bronchodilators **more than two days per week**. If low-dose ICS fails, the first step is to **check compliance** before increasing the dose.

## VI. Pneumonia in Children

- **Etiology by Age:**
    - **4 months to 4 years: Viral** pathogens are most common, specifically **RSV**.
    - **5 years and older:** Atypical bacteria, specifically ***Mycoplasma pneumoniae***, are most common.
    - **Specific cases: *S. pneumoniae*** is the leading cause in children with **agammaglobulinemia**. ***S. aureus*** is the most common cause of **empyema** and **pneumatoceles** (cavitary lesions).
- **Diagnosis:**
    - **CXR:** Viral shows **bilateral hyperinflation**; Bacterial (*S. pneumo*) shows **lobar consolidation**. **Round pneumonia** is typically caused by *S. pneumoniae* or *Klebsiella*.
    - **Physical Exam:** Signs of consolidation include **bronchial breathing**, **dullness to percussion**, and **increased tactile vocal fremitus**.

- **Hospitalization Indications:** Required for infants **<3–6 months**, patients with **respiratory distress** (SpO2 <90%), **immunodeficiency**, or those **failing oral antibiotics**. **Age less than 6 years** is NOT a reason for admission.
- **Treatment: Amoxicillin** is first-line for outpatient bacterial pneumonia. **Erythromycin** (or other macrolides) is the drug of choice for ***Mycoplasma***. If a patient fails amoxicillin and develops a pleural effusion, the next step is **IV Ceftriaxone and Vancomycin**.

## VII. Cystic Fibrosis (CF)

- **Genetics:** An **autosomal recessive** disease involving a defect in the **chloride channel** on **Chromosome 7**.
- **Diagnosis:**
    - **Meconium ileus** in a newborn is a diagnostic clinical feature.
    - **Sweat Chloride Test:** Value **≥60 mmol/L** is positive. **False negatives** occur with **hypoproteinemia** or edema. The test **does not normalize** with treatment and is not used to monitor management.
    - **Newborn Screening:** Measures **IRT**; a low IRT is not diagnostic, but an elevated one suggests CF.
- **Manifestations:** Chronic cough, **digital clubbing**, malabsorption (**foul-smelling stools**, failure to thrive), and **nasal polyps**. CF causes **delayed puberty**, NOT precocious puberty. It is NOT associated with perianal abscesses or immune deficiency.
- **Management:** Aim is to clear secretions and treat infection. **Pseudomonas aeruginosa** indicates progressive disease and is treated with an **aminoglycoside plus Ticarcillin or Ceftazidime**. Vitamin supplementation (A, D, E, **K**) is essential

# GI

## I. Breastfeeding and Infant Nutrition

- **Composition and Benefits:** Breast milk is considered the gold standard of nutrition, with its composition varying according to the infant's age. It is easily digested due to a high whey:casein ratio. While highly beneficial, breast milk is naturally deficient in Vitamin D, requiring supplementation. It also has a lower sodium load compared to formula. Benefits include promoting mother-infant bonding, reducing the frequency of diarrheal disease, and lowering the risk of recurrent infections and sudden infant death syndrome.
- **Antigenicity and Allergies:** The most antigenic component of breast milk is typically the protein component. In cases of Cow's Milk Protein Allergy (CMPA), which affects up to 5% of infants, symptoms include atopic dermatitis, chronic cough, and bloody stools. Management requires switching to extensively hydrolyzed or amino acid-based formulas; regular or partially hydrolyzed formulas are not appropriate. CMPA typically resolves before the age of three.
- **Dietary Milestones:** Solids should be introduced to a healthy infant's diet at 6 months. Fresh cow's milk is not allowed until 12 months of age. Low-fat milk should be delayed until the child is 24 months old.
- **Contraindications:** In developed countries, maternal HIV infection is a absolute contraindication to breastfeeding. However, conditions like breast abscess or hepatitis B (if the infant receives proper prophylaxis) are not absolute contraindications. For an infant born to a Hepatitis B carrier, the most appropriate management is to administer both Hep B immunoglobulin and the Hep B vaccine within the first hours of life.

## II. Acute Gastroenteritis (AGE) and Diarrhea

- **Definitions:** Acute diarrhea is generally defined as a decrease in stool consistency and/or an increase in frequency (typically ≥ 3 evacuations in 24 hours). It typically lasts less than 7 days and is considered chronic if it persists beyond 14 days.
- **Etiology and Mechanism:** Rotavirus is the most common cause of acute watery diarrhea in children and induces diarrhea through an osmotic mechanism. Most cases of acute diarrhea are self-limited and do not require extensive diagnostic workups unless the patient is very young, has underlying chronic conditions, or shows signs of invasive disease. Invasive or bloody diarrhea is often caused by organisms like *Shigella*, *Salmonella*, *Campylobacter jejuni*, and *Yersinia*. Notably, *Vibrio cholerae* and *Giardia* typically cause watery, non-invasive diarrhea.

- **Management:** Oral Rehydration Solution (ORS) is the first-line therapy for mild to moderate dehydration, with a recommended dose of 75ml/kg over 3–4 hours. Clinicians should encourage the early resumption of regular feeding and the continuation of breastfeeding throughout the illness. Routine use of lactose-free or diluted milk is not recommended.
- **Complications:** Dehydration is the most significant complication of AGE and is a leading cause of acute kidney injury (AKI) and shock in children. Indications for hospital admission include shock, intractable vomiting, failure of oral rehydration, or neurological abnormalities like lethargy or seizures.

## III. Chronic Diarrhea and Malabsorption

- **Pathogenesis:** Chronic diarrhea results from osmotic, secretory, inflammatory, or dysmotility processes. In secretory diarrhea, stool volume is not dependent on diet and persists during fasting. In osmotic diarrhea, such as that caused by lactose intolerance, the stool pH is typically acidic (< 5.5) due to carbohydrate malabsorption.
- **Celiac Disease:** This is an immune-mediated sensitivity to gluten found in wheat, barley, and rye (rice is safe). It is associated with other autoimmune conditions like Type 1 Diabetes and Trisomy 21. Presentation can include diarrhea, constipation, short stature, and Vitamin D deficiency. The best serological screening test is the anti-tissue transglutaminase (TTG) antibody, while the gold standard for diagnosis is a small bowel biopsy (histology).
- **Cystic Fibrosis (CF):** Failure to thrive in CF is primarily caused by fat malabsorption due to exocrine pancreatic insufficiency. Chronic diarrhea in these patients often presents as steatorrhea (fatty stools). Diagnosis is confirmed via a sweat chloride test; a reading above 60 mmol/L is diagnostic.
- **Toddler's Diarrhea:** Also known as chronic non-specific diarrhea, this is the most common cause of chronic diarrhea in children aged 2–4. It is characterized by the presence of undigested food particles in stools and is often linked to the excessive consumption of fruit juices. Crucially, it does not result in malnutrition or poor weight gain.

## IV. Failure to Thrive (FTT) and Abdominal Pain

- **Growth Evaluation:** Adequate weight gain is the best way to evaluate the adequacy of an infant's nutrition. FTT is categorized into types; Type 1 FTT is the most common, where weight crosses percentiles while head circumference and length are initially preserved. Causes of organic FTT include persistent diarrhea, malabsorption (Celiac, CF), and excessive vomiting.
- **Warning Signs:** "Red flags" for chronic diarrhea that suggest serious systemic disease include night stools, blood or mucus in the stool, and poor weight gain.
- **Abdominal Pain:** Pain that awakens a child at night is a classic indicator of organic (rather than functional) abdominal pain.

## V. Neonatal Jaundice

- **Physiologic Jaundice:** This is caused by increased bilirubin production (shortened RBC lifespan) and immature hepatic uptake or conjugation. It typically peaks at 3–4 days in term infants and disappears by 7–10 days.
- **Pathologic Jaundice:** This is considered a medical emergency and is recognized if jaundice appears within the first 24 hours of life, if bilirubin rises rapidly (>0.3 mg/dL/h), or if it persists beyond 14 days in a term infant. Direct (conjugated) bilirubin levels above 1 mg/dL are always pathologic.
- **Breastfeeding-Associated Jaundice:** This occurs in the first few days of life and is related to mild dehydration and low caloric intake. Management focuses on increasing the frequency of breastfeeding (every 2–3 hours) to improve hydration.
- **Management:** Management decisions are guided by hour-specific nomograms (like the Bhutani nomogram) to assess the risk of severe hyperbilirubinemia. Phototherapy is used to isomerize toxic bilirubin into a non-toxic product, preventing bilirubin-induced neurologic dysfunction (BIND) and kernicterus.

# Endocrine

## Growth Parameters and Milestones

Average term birth weight ranges from 2.5 to 4.2 kg, with a mean of approximately 3.5 kg. An infant typically loses up to 10% of their birth weight in the first week of life but should regain it shortly after. A child's weight generally doubles by 4 to 5 months of age and triples by their first birthday. During the first year, an infant should gain approximately 6 to 7 kg.

The average birth length is 50 cm. Height milestones include doubling birth length by 4 years of age and tripling it by 13 years. Growth in length during the first year is roughly 25 cm, followed by 12 cm in the second year and 6 cm in the third. Head circumference at birth is approximately 35 cm and increases by 12 cm during the first year of life. Macrocephaly is defined as a head circumference greater than the 97th percentile. The anterior fontanelle typically begins to close around 9 months of age. Teeth usually begin to erupt between 6 and 7 months. Upper-to-lower segment proportions change significantly throughout childhood and are not adult-like in infancy.

## Short Stature and Bone Age

Evaluation of short stature requires assessing growth velocity and bone age.

- **Constitutional Delay of Growth and Puberty:** Characterised by short stature, delayed puberty, and a bone age that is less than chronological age, though growth velocity is usually normal or at the lower limit of normal. Management is typically reassurance.
- **Growth Hormone (GH) Deficiency:** Associated with low growth velocity and delayed bone age. In neonates, signs include microphallus, hypoglycemia, prolonged jaundice, and midline defects like cleft palate. Birth weight is typically normal, as intrauterine growth is not primarily GH-dependent.
- **Familial (Genetic) Short Stature:** Bone age is equal to chronological age, linear growth velocity is normal, and height is consistent with the mid-parental height.
- **Mid-Parental Height Calculation:** For boys, (Father's height + Mother's height + 13) / 2; for girls, (Father's height + Mother's height - 13) / 2.

Screening tests for short stature include CBC, ESR, tissue transglutaminase (for celiac disease), and renal/thyroid function tests. A random growth hormone level is not a useful screening tool due to its pulsatile secretion.

## Pubertal Development

The normal onset of puberty is between 8 and 13 years in females and 9 and 14 years in males.

- **Females:** The first sign is typically breast budding (thelarche). Menarche usually occurs at Tanner stage 4, following peak height velocity. Obesity may lead to early menarche.
- **Males:** The first sign is testicular enlargement. Linear growth acceleration begins in the early stages (Tanner 1–2), while peak height velocity occurs later, often at Tanner stage 4.
- **Sexual Maturation Rate (SMR):** Staging is based on breast development in females, testicular and penile size in males, and pubic hair in both. Axillary hair is not a primary Tanner parameter. Precocious puberty is defined as onset before age 8 in girls.

## Diabetes Mellitus (DM)

Diabetes is a metabolic disorder of hyperglycemia caused by defective insulin secretion or action.

- **Classification:** Type 1 DM (autoimmune/antibody-mediated), Type 2 DM (insulin resistance), and rare forms like MODY (maturity-onset diabetes of the young, involving beta-cell dysfunction).
- **Diagnosis:** Fasting plasma glucose  126 mg/dL, random glucose  200 mg/dL with symptoms, or a 2-hour glucose  200 mg/dL during a 75-g OGTT.
- **Pathogenesis (T1DM):** Autoimmune destruction of beta cells; overt symptoms appear when less than 10% of beta-cell mass remains.
- **Insulin Action:** Insulin inhibits hepatic gluconeogenesis and glycogenolysis while stimulating protein synthesis and lipogenesis. In its absence, lipolysis and ketogenesis increase. Counter-regulatory hormones (epinephrine, cortisol, GH, glucagon) oppose insulin and raise blood glucose.
- **Management:** Multiple daily injections or insulin pumps are used. Education must cover home monitoring, nutrition, and identifying hypoglycemia; children should not be restricted from sports.
- **Monitoring:** HbA1c provides a 2- to 3-month average of glycemic control; the target is often below 7.5%. Screening for retinopathy and nephropathy should occur annually starting at age 11 if the disease duration is at least 2 years.

## Diabetic Ketoacidosis (DKA)

DKA results from severe insulin deficiency and is entirely preventable with proper management.

- **Diagnosis:** Glucose > 200 mg/dL, arterial pH < 7.3, and serum bicarbonate < 18 mmol/L, along with ketonuria or ketonemia.
- **Clinical Picture:** Signs include polyuria, polydipsia, abdominal pain (due to peritoneal irritation from acidosis), Kussmaul breathing (deep, rapid respiration), and confusion.
- **Cerebral Edema (CE):** A severe complication. Risk factors include age < 5 years, new-onset DM, high initial urea, low initial CO2, rapid administration of hypotonic fluids, and the use of bicarbonate.

## Hypoglycemia

Defined as a blood glucose level less than 72 mg/dL.

- **Symptoms:** Hunger, sweating, trembling, pallor, blurred vision, and headache. These typically precede neuroglycopenia.
- **Action:** Confirm the level and treat with simple carbohydrates (e.g., juice). Severe cases require glucagon via intramuscular/subcutaneous injection or nasal powder (Baqsimi, approved in 2019). The Gvoke HypoPen was also FDA-approved in September 2019 for severe rescue.
- **Sick Day Rules:** Never omit insulin during illness, as counter-regulatory hormones raise glucose levels; monitor glucose and ketones (urine or blood) frequently.

## Congenital Hypothyroidism

- **Causes:** The most common cause is thyroid dysgenesis (agenesis or ectopy). Thyroid dyshormonogenesis is suspected when a goiter is present. Maternal antithyroid medications can cause transient hypothyroidism.
- **Manifestations:** Early signs (first 4 weeks) are non-specific: prolonged jaundice, constipation, and poor feeding. By 3 to 6 months, the full picture includes a large tongue, umbilical hernia, hypotonia (not hypertonia), coarse hair, and broad hands with short fingers.
- **Management:** Routine screening (common in Jordan) and early treatment with levothyroxine are essential to prevent mental retardation.

## Adrenal Disorders

- **Congenital Adrenal Hyperplasia (CAH):** Most commonly caused by 21-hydroxylase deficiency (CYP21A2 gene mutation), leading to an accumulation of 17-hydroxyprogesterone. Features include ambiguous genitalia in females, hyponatremia, hyperkalemia, and metabolic acidosis. 11-beta hydroxylase deficiency is distinct because it is associated with hypertension.
- **Primary Adrenal Insufficiency (Addison's):** Characterised by skin hyperpigmentation, salt craving, low cortisol, and high ACTH. The high ACTH levels distinguish it from secondary insufficiency.
- **Adrenal Crisis Treatment:** Immediate management includes an intravenous bolus of normal saline (10–20 mL/kg), 10% dextrose for hypoglycemia, and stress-dose intravenous hydrocortisone. Mineralocorticoid replacement is generally not required during high-dose hydrocortisone therapy.

## Obesity

Pediatric obesity is defined as a BMI  95th percentile, while  85th is overweight. Complications include insulin resistance, hepatic steatosis, sleep apnea, pseudotumor cerebri, cholelithiasis, and early menarche. Genetic causes include Prader-Willi syndrome, which can be diagnosed using fluorescence in situ hybridization (FISH).

# Nephrology

## I. Fluid and Electrolyte Management

Fluid management in children involves calculating maintenance requirements and replacing deficits from dehydration.

- **Maintenance Requirements (Holliday-Segar Formula):**
    - **First 10 kg:** 100 mL/kg/day (or 4 mL/kg/hr).
    - **Second 10 kg:** 50 mL/kg/day (or 2 mL/kg/hr).
    - **Each kg > 20 kg:** 20–25 mL/kg/day (or 1 mL/kg/hr).
    - *Example:* A 12 kg child requires 1100 mL/day (1000 + 2 × 50). A 20 kg child requires 1500 mL/day (1000 + 500).
- **Dehydration Assessment and Management:**
    - **Degrees of Dehydration:** Categorised as mild, moderate, or severe based on clinical signs.
        - **Mild:** Normal pulse/BP, <5% weight loss (infant).
        - **Moderate:** Tachycardia, normal to slightly low BP, 6–10% weight loss, delayed capillary refill (>3s).
        - **Severe:** Weak pulse, hypotension (shock), >9% weight loss, capillary refill >5s, anuria.
    - **Earliest Sign:** Increased heart rate (tachycardia) is the earliest sign.
    - **Fluid Deficit Calculation:** Deficit (mL) = % Dehydration × Weight (kg) × 10.
        - *Example:* For a 15 kg child with 5% dehydration, deficit = 5 × 15 × 10 = 750 mL. Total 24-hr fluid = Maintenance (1250) + Deficit (750) = 2000 mL.
    - **Shock Management:** Administer 20 mL/kg Normal Saline (NS) bolus over 20 minutes; repeat up to three times (max 60 mL/kg) until stable.
- **Electrolyte Imbalances:**
    - **Hyponatremia (Na < 135):** Caused by gastroenteritis (extrarenal loss), SIADH (gain of water), or adrenal insufficiency. Correcting hyponatremia too rapidly (>10–12 mmol/day) risks **Central Pontine Myelinolysis**.

- **Hypernatremia (Na > 145/150):** Features include "doughy" skin, irritability, and brain hemorrhage. Correction should be slow (over 48 hours) to prevent **cerebral edema**.
- **Hypokalemia:** Causes include alkalosis and Renal Tubular Acidosis (RTA). Addison's disease is a cause of *hyperkalemia*, not hypokalemia.
- **Metabolic Alkalosis:** Classically seen in recurrent vomiting (e.g., pyloric stenosis) presenting with high pH, high HCO3, and low Cl.

## II. Urinary Tract Infections (UTI)

UTIs are common in children and require specific diagnostic and management approaches.

- **Epidemiology and Risk Factors:** *E. coli* is the most common causative organism. Neonatal boys are more prone to UTI than girls, but girls are more common later. Risk factors for recurrence include constipation, high-grade vesicoureteral reflux (VUR), and neurogenic bladder.
- **Diagnosis:**
  - **Specificity:** Positive Nitrite test is more specific than leukocyte esterase.
  - **Sample Collection:** Urethral catheterization or suprapubic aspiration is preferred in infants/newborns to avoid contamination. A suprapubic tap is definitive even at low counts (e.g., 2000 CFU).
  - **Clinical Presentation:** Pyelonephritis (upper UTI) is suggested by high-grade fever and loin pain, whereas cystitis involves lower tract symptoms.
- **Evaluation and Imaging:**
  - **DMSA Scan:** The gold standard for detecting **renal scarring**.
  - **MCUG:** The test of choice to detect Posterior Urethral Valves (PUV) and grade VUR.
  - **US:** Often indicated after the first UTI to detect structural anomalies.
- **Management:** Treatment duration is typically 7–14 days. Asymptomatic bacteriuria is generally *not* treated with antibiotics unless in specific contexts (e.g., pregnancy), but it can be a normal finding in neurogenic bladders.

## III. Nephrotic Syndrome (NS)

Nephrotic syndrome is characterized by the failure of the glomerular filtration barrier.

- **Diagnosis Criteria:**
    1. **Edema** (typically periorbital/scrotal).
    2. **Heavy Proteinuria** (>40 mg/m²/hr or spot ratio >2).
    3. **Hypoalbuminemia** (<2.5 g/dL).
    4. **Hyperlipidemia**.
- **Most Common Type:** Minimal Change Disease (MCD) accounts for >85% of pediatric cases.
- **Complications:**
    - **Infections:** Increased risk of spontaneous bacterial peritonitis.
    - **Hypovolemia:** Patients may present with abdominal pain and signs of shock despite edema.
    - **Hypercoagulability:** Low levels of Antithrombin 3 and increased bleeding/clotting tendency.
- **Management:**
    - **First-line:** Steroids (Prednisone). Most children (>90%) respond to steroids.
    - **Acute Phase:** Includes no-added-salt diet and albumin/furosemide for severe edema. ACE inhibitors are used for long-term proteinuria control but not typically in initial acute stabilization.
- **Biopsy Indications:** Indicated if the child is <12 months or >12 years, has steroid resistance, low C3, or persistent hematuria.

## IV. Acute Kidney Injury (AKI) and CKD

- **AKI Definitions:** Defined by abrupt loss of function and elevated serum creatinine.
    - **Prerenal:** Most common form; caused by reduced perfusion (e.g., dehydration). Renal tubular function remains intact (FENa < 1%).
    - **Intrinsic:** Involves parenchymal damage (e.g., Acute Tubular Necrosis [ATN]); FENa is usually >2%.

- **AKI Management:**
    - **Hyperkalemia Treatment:** Immediate stabilization with calcium gluconate if ECG changes (peaked T waves) are present. Shift potassium with insulin/glucose or sodium bicarbonate. Remove with diuretics or dialysis.
    - **Renal Replacement Therapy (RRT):** Indicated for fluid overload >20% unresponsive to diuretics, severe acidosis, or hyperkalemia.
- **Chronic Kidney Disease (CKD):**
    - **Definition:** Functional or structural abnormalities persisting for at least 3 months.
    - **Complications:** Anemia (EPO deficiency), CKD-MBD (bone disease due to hyperphosphatemia and low Vitamin D), growth failure, and hypertension.
    - **Staging:** Stage 1 (GFR >90) to Stage 5 (Kidney failure, GFR <15).

## V. Quick Reference for MCQ Answers

- **UTI Gold Standard Scarring:** DMSA scan.
- **NS Edema Mechanism:** Underfill (low albumin/renin activation) or Overfill (primary Na retention).
- **Earliest Dehydration Sign:** Tachycardia.
- **PUV Test of Choice:** MCUG.
- **Shock Bolus:** 20 mL/kg normal saline.
- **AKI Compensatory Mechanism:** Afferent arteriole dilation (Prostaglandins) and Efferent arteriole constriction (Angiotensin II).

# Anemia

## 1. Definition and Broad Classification

**Anemia** is clinically defined as a reduction in hemoglobin concentration or red cell volume that falls below the standard range for a person's specific age. Pediatric hematologists generally classify anemias into four functional categories based on their underlying cause:

- **Inadequate Intake:** This involves a deficiency of specific nutritional factors required for red cell production, namely **Iron, Vitamin B12, and Folic Acid**.
- **Production Defects (Bone Marrow Failure):** This includes congenital conditions like pure red cell anemia, ineffective erythropoiesis, or anemias secondary to **chronic infection, renal failure, and cancer**.
- **Hemolytic Anemias:** Conditions where red blood cells are destroyed prematurely due to membrane defects, enzyme deficiencies, or hemoglobin synthesis issues.
- **Blood Loss:** Direct loss of blood from the circulatory system.

## 2. Pediatric Physiology and Early Infancy

The concentration of hemoglobin changes significantly in early life. While newborns typically have a higher hemoglobin level, they experience **"physiologic anemia"** at approximately 2 to 3 months of age. This process is often **exaggerated in premature babies**, who may develop more severe anemia during this period.

## 3. Nutritional Anemias (Inadequate Intake)

**Megaloblastic Anemia**

This condition is characterized by the presence of **megaloblasts in the bone marrow and macrocytes (abnormally large red cells) in the blood**.

- **Folic Acid Deficiency:** This often presents between **4 to 7 months of age**. Causes include a diet lacking folate, poverty, or the consumption of **goat's milk**. It can also result from increased requirements due to hemolysis or metabolic disorders like MTHFR deficiency.
- **Vitamin B12 Deficiency:** This typically manifests between **9 months and 10 years of age**. It can be caused by the mother's diet (in breastfed infants), defective absorption due to a lack of **intrinsic factor**, or transport defects like Transcobalamin II deficiency.

- **Clinical Presentation:** Patients often exhibit failure to thrive, generalized weakness, glossitis (tongue inflammation), anorexia, and pallor. B12 deficiency is distinct for causing **neurologic manifestations**, including ataxia, paresthesia, and hyporeflexia.
- **Treatment:** Folic acid is treated with a **5 mg daily tablet**. Vitamin B12 deficiency requires **intramuscular (I.M.) injections** (1 mg daily for 2 weeks if neurologic defects are present, followed by monthly doses).

**Iron Deficiency Anemia (IDA)**

IDA is a common pediatric diagnosis, often caught during routine check-ups when a physician notices **pallor** in a child (such as a 20-month-old).

- **Diagnosis:** Important history elements include a **detailed nutritional history**, checking for gastrointestinal symptoms like **chronic diarrhea**, and investigating a family history of anemia. Lab results typically show **low hemoglobin (e.g., 5 gm), low MCV (e.g., 50), and high RDW (e.g., 20)**.
- **Associated Conditions:** IDA can sometimes be associated with **Peutz-Jeghers Syndrome**, which presents as brown pigmentation on the lips and gums.
- **Treatment and Response:** Therapy involves an iron-fortified diet and **oral ferrous supplements (6 mg/kg/day)** for 2–3 months. The **first response to therapy is an increase in appetite and activity** because iron is essential for the function of certain body enzymes. This is followed by an **increase in reticulocyte count after 3–5 days**.
- **Failure to Respond:** If a patient does not improve, doctors consider **poor compliance, ongoing blood loss, inadequate dosing**, or coexistent diseases like malabsorption or malignancy.

## 4. Hemolytic Anemias

Hemolysis is the premature destruction of red cells. Clinical evidence includes a **low Packed Cell Volume (PCV), increased indirect bilirubin, high reticulocyte count, and low haptoglobin**.

- **Hereditary Spherocytosis (HS):** This is usually an **autosomal dominant** defect (75% of cases) in cell membrane proteins like **spectrin and ankyrin**. Symptoms include moderate anemia, jaundice, **splenomegaly (enlarged spleen)**, and the formation of **gallstones**.
- **G6PD Deficiency:** This is the most common red cell enzyme defect and is **X-linked**, affecting males more frequently. Hemolysis is triggered by **oxidant stress**, such as infections, fava beans, or drugs like **aspirin and sulfa**. This leads to the formation of **Heinz bodies** (denatured globin) and premature cell destruction.

- **Alpha Thalassemia:** This is classified by the number of gene deletions: **Silent Carrier** (1 gene), **Trait** (2 genes), **HbH disease** (3 genes), and **Hydrops Fetalis** (4 genes).

## 5. Management of Chronic Anemias (Thalassemia focus)

- **Prevention:** Strategies include identifying carriers (long-term) and providing **genetic counseling** for couples at risk before marriage.
- **Hypertransfusion Protocol:** To ensure normal growth and development, patients are maintained at a **pre-transfusion hemoglobin level of 10.5 gm**. This helps minimize skeletal abnormalities and reduces the enlargement of the spleen.
- **Iron Overload and Chelation:** Ongoing transfusions lead to toxic iron levels. **Chelation therapy** is used to bind and remove this iron. Common agents include:
    - **Desferal:** Given via SC or IV infusion; highly selective but suffers from poor patient compliance.
    - **Ferriprox:** An oral tablet taken three times daily; carries a risk of **neutropenia**.
    - **Deferasirox (Ex jade):** A once-daily oral tablet; carries a risk of **kidney toxicity and skin rashes**.

## 6. Bone Marrow Failure and Aplastic Anemia

Bone marrow failure involves disorders of the hematopoietic stem cells that can affect **one cell line or all lines (pancytopenia)**.

- **Single Line Defects:** Examples include **Diamond-Blackfan** (red cells only) or **Kostman Syndrome** (white cells only).
- **Aplastic Anemia:** Defined by **pancytopenia** and a hypocellular bone marrow. A well-known familial form is **Fanconi Anemia**, which is associated with physical anomalies like **renal and skeletal defects, strabismus, microphthalmia, and mental retardation**.
- **Management:** Treatments include steroids, androgens, cyclosporine, or **Bone Marrow Transplantation (BMT)**.
- **Transfusion Risks:** Side effects of blood transfusions include allergies, fever (non-hemolytic febrile reactions), viral transmission, and **Graft-versus-host disease (GVHD)**.

# Neurology

## I. Developmental Milestones and Evaluation

Correctly identifying developmental ages is essential for pediatric neurology assessments.

- **Vision and Social Interaction:** Visual fixation and tracking are present at birth. Social smile typically emerges at 2 months but may be delayed until 3 months; delay beyond this suggests cognitive or neurological issues. Infants track objects 180 degrees by 2 months.
- **Motor Milestones:**
    - **Head Support:** Full head support is usually established by 4 months.
    - **Rolling and Sitting:** Rolling (prone to supine) begins around 4–6 months. Infants sit with pelvic support by 6 months.
    - **Walking and Running:** The normal range for walking is 9–15 months. Walking with one hand held is typical at 12 months, and running stiffly is expected by 18 months.
    - **Advanced Mobility:** Riding a tricycle and walking upstairs alternating feet occur around 3 years (30–36 months).
- **Fine Motor and Drawing:**
    - **Grasp:** The immature pincer grasp is expected by 12 months. Putting objects in the mouth begins at 6 months.
    - **Copying Shapes:** A child draws a vertical line at 18–20 months. A horizontal line is typical by 2 years. A circle is copied at 3 years. A cross is copied at 4 years. A square is copied at 4 years. A triangle is typically copied at 5 years.
- **Language and Cognitive:**
    - **Words:** At 12 months, a child says 1-3 words besides "mama" and "baba" specifically. By 18 months, they should speak at least 10 words.
    - **Sentences:** Two-word phrases/sentences appear by age 2. Pronouns (like "I") are used by 2.5 to 3 years.
    - **Self-Knowledge:** A child knows their name and sex by 3 years. Knowing self-age and sex is NOT typically expected as early as 18 months.

- **Correction for Prematurity:** For developmental assessment of premature infants, their age should be corrected for their gestational age (GA) at birth. For example, a 7-month-old born at 28 weeks (12 weeks/3 months premature) has a corrected developmental age of 4 months.

## II. Approach to Hypotonia and Neuromuscular Disorders

Hypotonia is a clinical sign defined as decreased resistance to passive movement.

- **Central vs. Peripheral:**
    - **Central (Upper Motor Neuron):** Features include lethargy, seizures, dysmorphism, hand fisting, scissoring on vertical suspension, and normal or increased reflexes.
    - **Peripheral (Lower Motor Neuron):** Features include alertness/responsiveness, hyporeflexia or areflexia, and muscle atrophy. **Fasciculations** (specifically of the tongue) are a classic sign of peripheral involvement.
- **Spinal Muscular Atrophy (SMA):**
    - **SMA Type I (Werdnig-Hoffmann):** Onset <6 months, severe, majority die before age 1.
    - **SMA Type II:** Onset 6–18 months; child can sit but cannot walk.
    - **SMA Type III:** Onset >18 months, milder progression.
    - **Diagnosis:** Creatine kinase (CPK) is typically normal in SMA.

## III. Seizures and Epilepsy Syndromes

- **Definitions:** A seizure is a transient occurrence of abnormal neuronal activity; epilepsy requires at least two unprovoked seizures >24 hours apart.
- **Febrile Seizures:** Occur between 6 months and 5 years.
    - **Simple:** Generalized, <15 minutes, does not recur in 24 hours.
    - **Complex:** Focal, >15 minutes, or recurs within 24 hours.
    - **Prognosis:** The risk of recurrence is highest if the first seizure occurs in infancy. Risk of future epilepsy is generally low (~2%) unless there is a family history or complex features.
    -

- **Common Syndromes:**
    - **Infantile Spasms (West Syndrome):** Spasms in series, often associated with Tuberous Sclerosis. EEG shows **hypsarrhythmia**. Treatment includes ACTH, steroids, or Vigabatrin.
    - **Childhood Absence Epilepsy:** 4–8 years, staring spells, EEG shows generalized **3 Hz spike-wave discharges**. Valproic acid is often the drug of choice.
    - **Benign Rolandic Epilepsy (BECTS):** 4–11 years, focal seizures often on awakening, EEG shows centrotemporal spikes.
    - **Lennox-Gastaut Syndrome:** Multiple seizure types, abnormal EEG, intellectual dysfunction; least likely to respond well to medical treatment.
    - **Juvenile Myoclonic Epilepsy (JME):** Adolescents, myoclonic jerks upon awakening, often requires life-long treatment with Valproate.
- **Neonatal Seizures:** Most common types are **subtle seizures** (lip-smacking, eye deviation, apnea). **Phenobarbital** is the first-line treatment. Hypokalemia is NOT a common cause (think hyponatremia or hypoglycemia).
- **Status Epilepticus:** Seizure >5 minutes. Emergency treatment starts at 5 minutes (T1). Initial rescue medications include Midazolam, Lorazepam, or Diazepam.

## IV. Meningitis in Children

- **Microbiology:**
    - **Neonates (<3 months):** Group B Streptococcus (GBS), *E. coli*, and *Listeria monocytogenes*. *E. coli* after 6 days of life should prompt exclusion of galactosemia.
    - **Older children:** *S. pneumoniae* and *N. meningitidis*.
- **CSF Analysis (Bacterial):** Characterised by WBC >1000 with neutrophil predominance, elevated protein (>100 mg/dL), and decreased glucose (<40 mg/dL or <60% of serum glucose).

- **Management:**
    - **Empiric Antibiotics:** Vancomycin plus Ceftriaxone (or Cefotaxime) for most older children. Ampicillin plus Gentamicin for early-onset neonates.
    - **Dexamethasone:** Indicated for infants ≥6 weeks with suspected *Hib* or *S. pneumoniae* meningitis. It should be given **before or within 20 minutes** of the first antibiotic dose to reduce hearing loss and mortality.
- **Prophylaxis:** Rifampin is used for close contacts of *Hib* or meningococcal cases. Prophylaxis for *Hib* is indicated for all household contacts if there is an unimmunized child <4 years or an immunocompromised child.

## V. Cerebral Palsy (CP) and Other Conditions

- **Cerebral Palsy:** A non-progressive motor disorder.
    - **Risk Factors:** Prematurity (major risk), low birth weight, birth asphyxia, and multiple gestations. Consanguinity is NOT a standard risk factor for CP.
    - **Types:** Diplegic CP (common in preterm infants with periventricular leukomalacia); Choreoathetotic CP (often related to high bilirubin/kernicterus).
- **Specific Clinical Signs:**
    - **Tuberous Sclerosis:** Seizures, ash leaf spots (hypopigmented lesions), and subependymal calcification on MRI.
    - **Sturge-Weber Syndrome:** Epilepsy, port-wine stain, and hemangiomas.
    - **Tay-Sachs Disease:** Developmental delay and cherry red spots on eye exam.
    - **Hurler Syndrome:** Coarse features, hepatosplenomegaly, and joint limitations.

# Neonatology

## I. Neonatal Classification and Mortality

- **Gestational Age (GA) Categories**:
    - **Term**:  37 completed weeks.
    - **Preterm**: < 37 weeks. Further divided into:
        - **Extremely preterm**: < 28 weeks.
        - **Very preterm**: 28 to < 32 weeks.
        - **Moderate to late preterm**: 32 to < 37 weeks.
    - **Post-term**: > 42 completed weeks.
- **Birth Weight (BW) Categories**:
    - **Low Birth Weight (LBW)**: < 2500g.
    - **Very Low Birth Weight (VLBW)**: < 1500g.
    - **Extremely Low Birth Weight (ELBW)**: < 1000g.
- **Growth Status**: Infants are plotted on charts to determine if they are **Small for Gestational Age (SGA)** (< 10th percentile), **Appropriate for Gestational Age (AGA)** (10th–90th percentile), or **Large for Gestational Age (LGA)** (> 90th percentile).
- **Jordan Statistics**: The leading causes of neonatal mortality in Jordan are **congenital malformations** followed by **prematurity**.

## II. Immediate Care and the "Golden Hour"

The "Golden Hour" refers to the evidence-based steps taken in the first 60 minutes for a term baby with good tone and breathing.

- **Step 1: Thermal Care**: Immediate drying, using warm preheated sheets, and hats are essential to maintain a temperature between . An unclothed newborn in a  room feels like an adult in .
- **Step 2: Delayed Cord Clamping (DCC)**: Delaying for **1–3 minutes in term** infants (or until pulsation stops) and **30–60 seconds in preterms**.

    - **Benefits**: Increased red cell volume, higher iron stores up to 6 months, and significantly reduced risk of **anemia, intraventricular hemorrhage (IVH), and necrotizing enterocolitis (NEC)**.

- **Step 3: Skin-to-Skin Contact**: Should be continuous for  60 minutes to maintain stability, reduce crying, and promote bonding.
- **Step 5: Early Breastfeeding**: Initiation should occur within the **first hour**.
- **Step 6: Postponing Procedures**: Bathing (delay 6–24 hours), weighing, and non-urgent injections should wait until after the first hour.
- **Step 7: Vitamin K**: Administered intramuscularly (IM) in the first hour to prevent **Vitamin K-deficiency bleeding (VKDB)**.
    - **Why neonates are deficient**: Immature gut flora (slow acquisition of K-producing bacteria), low transplacental transfer, poor fetal liver storage, and low content in human breast milk.
    - **VKDB Signs**: Classic (day 1–7) or Late (1–12 weeks) presenting as skin bruising, umbilical/circumcision bleeding, or **fatal intracranial hemorrhage**.

## III. Newborn Assessment and Apgar Scoring

- **Clinical Examination**: The nurse performs an initial assessment immediately at birth. The full, complementary paediatric clinician's exam is typically completed between **24–48 hours** (ideally with parents present).
- **Apgar Score Components (Evaluated at 1 and 5 minutes)**:
    - **Appearance (Color)**: Pink (2); Acrocyanosis/Body pink with blue extremities (1); Blue/Pale (0).
    - **Pulse (Heart Rate)**: > 100 bpm (2); < 100 bpm (1); Absent (0).
    - **Grimace (Reflex Irritability)**: Sneezing/Coughing/Vigorous cry (2); Grimace only (1); No response (0).
    - **Activity (Muscle Tone)**: Active motion/flexed limbs (2); Some flexion (1); Limp/Flaccid (0).
    - **Respiration**: Regular breathing/Strong cry (2); Irregular/slow (1); Absent (0).
    - *Note*: The Apgar score is **not** used to guide immediate resuscitation (which starts before the 1-minute score is calculated) and does not predict future IQ.

- **Common Normal Findings**: Brick dust-coloured urine (urates), breast engorgement, vaginal spotting (pseudomenses), and Mongolian spots.
- **Alarming/Abnormal Signs**: White pupillary reflex (**leukocoria**), diastolic murmurs, **bifid uvula**, bulging fontanelle, or a liver edge palpable > 1cm below the costal margin.

## IV. Neonatal Jaundice

- **Physiologic Jaundice**: Typically appears **after the first 24 hours** (usually day 2–3), increases at a rate < 5 mg/dL/day, and has direct bilirubin < 2 mg/dL.
- **Pathologic Jaundice**: Suspected if it appears in the **first 24 hours of life**. **Hemolysis** (ABO/Rh incompatibility) is the most common cause.
- **Breastfeeding Jaundice**: Early onset (first week) due to low caloric intake/dehydration (e.g., feeding only every 5–6 hours).
- **Breast Milk Jaundice**: Late onset (after day 5–7); a common cause of prolonged unconjugated hyperbilirubinemia.
- **Direct (Conjugated) Hyperbilirubinemia**: Always pathologic; includes biliary atresia (presents with white stools, dark urine, and palpable liver) or metabolic disorders.

## V. Respiratory Distress and Resuscitation

- **Resuscitation**: If a baby is not breathing, the first step is **Positive Pressure Ventilation (PPV)**. Chest compressions start only if HR remains < 60 despite effective ventilation. The drug for persistent bradycardia is **Adrenaline (Epinephrine)**; Atropine is not used.
- **Respiratory Distress Syndrome (RDS)**: Caused by **surfactant deficiency** in preterms. CXR shows a "**ground glass**" appearance and **air bronchograms**. Treatment includes surfactant (LISA method), oxygen, and early nasal CPAP.
- **Transient Tachypnea of the Newborn (TTN)**: Delayed clearance of lung fluid, often after **elective C-section**. Onset is usually within 2 hours of birth.
- **Meconium Aspiration Syndrome**: Seen in term/post-term babies; complications include pneumothorax and persistent pulmonary hypertension.

## VI. Complications of Prematurity

- **Apnea of Prematurity**: Cessation of breathing > 20 seconds; treated with **caffeine**.
- **Necrotizing Enterocolitis (NEC)**: A serious GI emergency; risk is significantly lower (6–10 fold) in infants receiving **human breast milk**.

- **Intraventricular Hemorrhage (IVH)**: Bleeding in the brain; grading depends on severity; can lead to post-hemorrhagic hydrocephalus.
- **Retinopathy of Prematurity (ROP)**: Due to incomplete retinal vascularization; linked to low GA and BW.
- **Metabolic Bone Disease (Osteopenia)**: Diagnosed by low serum phosphorus, **high Alkaline Phosphatase** (> 600), and high PTH.
- **Anemia of Prematurity**: Due to blood loss and a shortened RBC lifespan (40–60 days).

## VII. Infant Nutrition

- **Human Milk vs. Cow's Milk**: Human milk contains more **lactoalbumin, vitamins A, C, E, and lactoferrin**, and secretes **IgA**. However, it has **less calcium and phosphate** than cow's milk.
- **Human Milk Oligosaccharides (HMOs)**: Major solids (after lactose and lipids) that protect against infections like NEC and diarrhea.
- **Vitamin D**: Exclusively breastfed babies require **400 IU daily supplementation**, even if the mother has adequate stores.
- **Contraindications to Breastfeeding**: HIV, active TB (until treated), herpes lesions on the breast, or the infant having **galactosemia**. Maternal Hepatitis B is **not** a contraindication if the infant is vaccinated and receives HBIG.

## VIII. Growth, FTT, and Short Stature

- **Normal Growth**: Infants lose up to 10% of birth weight but regain it by **10–14 days**. They double BW by 4 months and triple it by 1 year.
- **Failure to Thrive (FTT)**: Inability to maintain expected growth rates.
    - **Type I**: Most common; due to inadequate intake (e.g., incorrect formula preparation) or excessive loss (vomiting/diarrhea).
    - **Wasting**: Weight-for-length < 3rd percentile (indicates acute malnutrition).
    - **Stunting**: Height < 3rd percentile (indicates chronic malnutrition).
- **Refeeding Syndrome**: Risks in severely malnourished children; characterised by **hypokalemia, hypomagnesemia, and hypophosphatemia**.

- **Short Stature**: Height < 3rd percentile or < -2 SD. The **ICP model** phases are: Infancy (nutrition-dependent), Childhood (GH/Thyroxine-dependent), and Puberty (sex steroid-dependent).

## IX. Development and Screening

- **Developmental Milestones**:
    - **4 months**: Full head control, reaches for and mouths objects.
    - **7 months**: Transfers objects hand-to-hand, rolls over.
    - **10 months**: Pincer grasp, cruises/walks holding furniture, "mama/dada".
    - **15 months**: Walks alone, uses 3 cubes.
    - **2 years**: Runs well, puts 3 words together.
    - **Red Flags**: No single words by 16 months or **loss of any skill at any age**.
- **Mandatory Screenings in Jordan**:
    - **Metabolic**: TSH, G6PD, and **PKU** (tested at 2 weeks).
    - **Hearing**: Officially launched in 2021 (prevalence of loss 1.4/1000).
    - **CCHD (Heart)**: Pulse oximetry on the **right hand and either foot** after 24 hours.
- **Safe Sleep (SIDS Prevention)**: The baby should sleep **alone, on their back, in a bare crib**, and share a room (but not a bed) with parents for at least 6 months.

# Rheumatology

Pediatric rheumatology is a complex medical specialty dedicated to the diagnosis and management of **autoinflammatory and autoimmune conditions** in children. These diseases are often multisystemic, meaning they can affect many different parts of the body simultaneously, and because most lack a single definitive diagnostic test, clinicians must rely heavily on a detailed **clinical history and physical examination**.

While musculoskeletal (MSK) problems are common in children—affecting roughly 1 in 8—the causes range from benign issues like hypermobility to life-threatening conditions such as malignancy or infection. To help differentiate these, doctors look for **inflammatory features**, such as "gelling" (stiffness after rest), morning stiffness, and pain that improves with movement, versus **mechanical features**, where pain is typically worse during or after physical activity.

## Juvenile Idiopathic Arthritis (JIA)

**JIA** is the most common chronic rheumatic illness in children. It is defined as arthritis in one or more joints that lasts for at least **six weeks** in a child under 16, with no other identifiable cause. It is classified into several distinct subtypes:

- **Oligoarthritis:** This is the most common form of JIA. It typically affects **four or fewer joints**, usually medium-to-large ones like the knee or ankle. It most frequently occurs in young girls between 1 and 3 years old. A critical risk for these children is **chronic uveitis** (asymptomatic eye inflammation), especially if they test positive for **Antinuclear Antibodies (ANA)**. Because uveitis does not cause a red or painful eye initially, regular "slit lamp" eye screenings are essential to prevent permanent vision loss.
- **Polyarthritis:** This subtype involves **five or more joints** and can affect both large and small joints, including the fingers, shoulders, and cervical spine.
    - **RF-Negative:** This form is more common in girls and often has a "biphasic" age of onset.
    - **RF-Positive:** Typically appearing in older children (ages 9–11), this form is often more aggressive and is associated with the presence of **Rheumatoid Factor (RF)** and **anti-CCP antibodies**.
- **Systemic JIA (Still's Disease):** This is a unique and serious form characterized by **quotidian (daily) spiking fevers** and a classic **salmon-pink, evanescent rash** that comes and goes with the fever. Unlike other forms, it often involves internal organs, leading to an enlarged liver or spleen (**hepatosplenomegaly**) and swollen lymph nodes.

- **Enthesitis-Related Arthritis (ERA):** This involves both arthritis and **enthesitis** (inflammation where tendons or ligaments attach to the bone). It is most common in boys over the age of 6 and is strongly linked to the **HLA-B27 gene**. Patients often experience inflammatory pain in the lower back or sacroiliac joints, which can lead to limited flexibility in the spine.
- **Psoriatic JIA:** This diagnosis is made when a child has arthritis combined with either psoriasis or signs like **dactylitis** (swelling of an entire finger or toe, known as "sausage digit") and **nail pitting**. It often affects the distal interphalangeal (DIP) joints of the hands.

## Life-Threatening Complications and Emergencies

- **Macrophage Activation Syndrome (MAS):** This is a life-threatening complication most often seen in systemic JIA. It occurs when the immune system overreacts, leading to a massive inflammatory response. Key signs include persistent high fever, falling blood cell counts (**cytopenia**), liver dysfunction, and **extremely high ferritin levels** (often exceeding 10,000 ng/mL).
- **Septic Arthritis:** This is a **medical emergency** caused by an infection in the joint. It typically presents with a very high fever and a single joint that is intensely painful, hot, and red. It requires urgent joint aspiration and treatment with intravenous (IV) antibiotics.

## Other Common Musculoskeletal Conditions

- **Reactive Arthritis:** This is common following a viral illness, such as a sore throat. It causes joint swelling and pain that usually resolves on its own within 2 to 3 weeks.
- **Hypermobility:** This is a non-inflammatory, benign condition where joints have an unusually large range of motion. Children often experience aches **after exercise** or after long periods of handwriting. Management involves reassurance, appropriate footwear, and physiotherapy to strengthen the muscles around the joints.

## The Diagnostic Process

To reach a diagnosis, pediatric rheumatologists use a combination of tools:

- **pGALS:** A quick, validated screening tool (Gait, Arms, Legs, Spine) used to identify joint problems in school-aged children.
- **Laboratory Tests:** These include markers of inflammation (**ESR and CRP**), full blood counts to check for anemia or low platelets, and tests for specific autoantibodies like **ANA, RF, and anti-dsDNA**.
- **Imaging:** X-rays are often used to look for joint damage or bone changes, while **Ultrasound and MRI** are highly effective at detecting early joint inflammation and soft tissue changes.

# Infectious

## 1. Clinical Approach to Fever

- **Pathogenesis:** Fever is a controlled rise in the hypothalamic temperature 'set-point'. It is triggered by **exogenous pyrogens** (microorganisms, toxins, drugs) that induce **leukocytes** to release **endogenous pyrogens** (IL-1, IL-6, TNF-, IFN). These stimulate **PGE2 synthesis**, affecting the Thermoregulatory Center (TRC) to increase heat production and reduce loss.
- **Fever vs. Hyperthermia:** Fever is a controlled hypothalamic response. **Hyperthermia** is a rise in core temperature *beyond* the set-point and regulation of the hypothalamus (e.g., heat stroke).
- **Benefits:** Fever inhibits bacterial growth and viral replication while enhancing immunological processes like T-cell activity and antibody synthesis. Phagocytosis is significantly greater at temperatures >40°C.
- **Categorisation:**
    - **Low-grade:** 37.3°C to 38.0°C.
    - **Moderate-grade:** 38.1°C to 39.0°C.
    - **High-grade:** 39.1°C to 41.0°C.
    - **Hyperthermia:** >41.0°C.
- **Serious Infection Indicators:** Age <1 month, poor arousability, petechial/non-blanching rash, delayed capillary refill, and bulging fontanelle.
- **Traffic Light System (Red/High Risk):** Includes age <3 months with temp 38°C, grunting, respiratory rate >60, reduced skin turgor, and status epilepticus.

## 2. Acute Bacterial Meningitis

- **Pathogens by Age:**
    - **Neonates (<3 months):** Group B Streptococcus (GBS), *Escherichia coli* (major causes of early-onset), and *Listeria monocytogenes*. If *E. coli* occurs after 6 days of age, exclude galactosemia.
    - **Older Infants/Children:** *S. pneumoniae*, *N. meningitidis*, and *H. influenzae* type b (Hib).

    - **Adolescents:** Predominantly *N. meningitidis*.

- **Clinical Presentation:**
    - **Neonates:** Non-specific signs like poor feeding, vomiting, respiratory distress (33-50%), and temperature instability. Classic signs like **nuchal rigidity (15%) and bulging fontanelle (25%) are uncommon** in this age group.
    - **Older Children:** Fever, headache, photophobia, and positive meningeal signs (**Kernig sign**: cannot extend knee >135° with flexed hip; **Brudzinski sign**: neck flexion causes involuntary hip/knee flexion).
- **CSF Analysis (Bacterial):** Characterised by high WBC (1000-5000 with >80% neutrophils), elevated protein (100-500 mg/dL), and **low glucose** (<40 mg/dL or ratio <0.4).
- **Management:**
    - **Empiric Therapy:** Post-neonatal cases require **Vancomycin** (to cover resistant *S. pneumoniae*) plus a 3rd-generation cephalosporin (**Ceftriaxone** or Cefotaxime).
    - **Neonatal Therapy:** Ampicillin plus an aminoglycoside (Gentamicin).
    - **Dexamethasone:** Indicated for infants **6 weeks** and children. It must be given **before or within 10-20 minutes of the first antibiotic dose** to reduce hearing loss and mortality, particularly for Hib and pneumococcal meningitis. It is **not indicated for neonates**.
- **Chemoprophylaxis:**
    - **Meningococcal:** Early administration (ideally <24 hours) for close contacts using **Rifampin (2 days)**, Ceftriaxone (IM), or Ciprofloxacin.
    - **Hib:** Rifampin prophylaxis for all household contacts if there is an unimmunized child <4 years or an immunocompromised child present.

## 3. Urinary Tract Infections (UTI)

- **Epidemiology:** Females have a higher overall prevalence (8% vs 2% in males). However, in **neonates, males have a higher incidence**, especially if uncircumcised.
- **Pathogens:** *E. coli* causes 80% of cases. Others include Klebsiella, Proteus, and Pseudomonas.
- **Clinical Features:** Neonates may present with sepsis, failure to thrive, or prolonged jaundice. Older children show classic symptoms like **dysuria, urgency, and flank pain**.

- **Risk Factors for Testing:** White race, age <12 months, temperature 39°C, and fever duration 24-48 hours without another source.
- **Imaging:**
    - **RBUS (Ultrasound):** Should be performed after the first febrile UTI in infants.
    - **VCUG:** Not routine after the first UTI; indicated only if RBUS shows hydronephrosis/scarring or for recurrent febrile UTIs.
- **Treatment:** Pyelonephritis (febrile UTI) requires 7-14 days of antibiotics (e.g., Ceftriaxone). Non-febrile cystitis can be treated for 3-5 days with oral TMS or 2nd/3rd generation cephalosporins.

## 4. Immunisation Facts (MCQ Focus)

- **Live Attenuated Vaccines:** MMR, Oral Polio (OPV), BCG, Varicella, and Rotavirus.
- **Inactivated/Killed Vaccines:** Inactivated Polio (IPV), Hepatitis A, DTaP (acellular pertussis/toxoids).
- **Contraindications:**
    - **General:** Severe anaphylactic reaction to a previous dose.
    - **Pertussis (DTaP):** Progressive CNS disease or **encephalopathy within 7 days** of a previous dose. Precautions (not absolute contraindications) include seizures, high fever (>40.5°C), or persistent crying >3 hours.
    - **OPV:** Contraindicated for household contacts of immunocompromised patients (use IPV instead).
    - **Pregnancy:** Pneumococcal, Varicella, and MMR vaccines are generally contraindicated.
- **Schedules & Specifics:**
    - **Rotavirus:** Given at 2, 3, and 4 months in Jordan. Contraindicated after 8 months.
    - **Hib:** Not indicated for healthy children **older than 5 years**.
    - **Hepatitis B:** Newborns of HBsAg-positive mothers must receive **both the HepB vaccine and IVIG** immediately.

## 5. Febrile Exanthems & Other Infections

- **Measles:** essential prodrome includes cough, coryza, and conjunctivitis. **Koplik spots** are pathognomonic. Vitamin A is used for treatment.
- **Rubella:** maximal fetal risk occurs in the first two months of pregnancy. Rash typically starts on the face.
- **Roseola Infantum:** characterized by high fever for 1-5 days followed by an **abrupt drop in temperature** and the appearance of a rash.
- **Chickenpox (Varicella):** rash appears first on the **trunk**. It is more severe in adults and neonates. Vaccine is effective post-exposure if given within 3-4 days.
- **Diphtheria:** suspected cases require immediate **antitoxin**. The vaccine contains a smaller antigen amount for adults.
- **Tetanus:** a toxin-mediated disease that is **not contagious**. Maternal immunization prevents neonatal tetanus.
- **Kawasaki Disease:** Requires fever 5 days plus criteria like strawberry tongue, polymorphous rash, and cervical lymphadenopathy.
- **Rotavirus:** Highly contagious cause of osmotic diarrhea; dehydration is the primary complication. Not associated with HUS (which is usually *E. coli* O157:H7).