

NGN NCLEX 2026 • VISUAL REFERENCE • PEDIATRIC CARDIOLOGY

Tetralogy of *Fallot*

The most common cyanotic congenital heart defect after infancy • A four-part right-to-left shunt

CYANOTIC DEFECT

PEDIATRICS

CARDIAC

RIGHT → LEFT SHUNT

PRIORITY: AIRWAY/O₂

SOURCE DISCLOSURE

This topic was not present in your uploaded personal notes for this session. Content compiled from standard NCLEX references: *Saunders Comprehensive Review for the NCLEX-RN*, *ATI RN Nursing Care of Children*, *Lippincott Pediatric Nursing*, and the 2024 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease.

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Definition & Overview

WHAT IS IT • WHY IT MATTERS

- ▶ **Tetralogy of Fallot (TOF)** is a congenital heart defect made up of **four** structural abnormalities occurring together.
- ▶ It is the **most common cyanotic** congenital heart defect **after age 1**. (Transposition of the Great Arteries is more common in the newborn period.)
- ▶ The combination produces a **right-to-left shunt** — deoxygenated blood bypasses the lungs and enters systemic circulation, causing **hypoxemia** and **cyanosis**.

The Four Defects

P

Pulmonary Stenosis

Narrowing of the pulmonary valve / RV outflow tract — blocks blood flow to the lungs.

R

RV Hypertrophy

Right ventricle thickens from pumping against the stenosis (high afterload).

O

Overriding Aorta

Aorta sits directly over the VSD and receives blood from *both* ventricles.

V

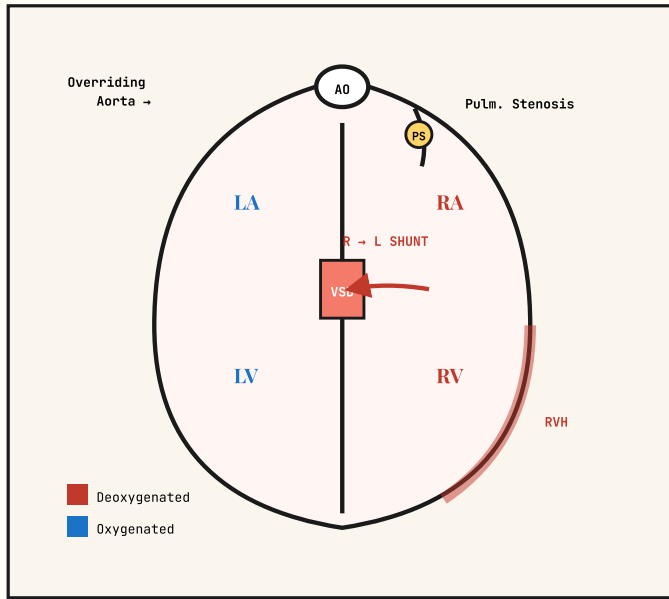
VSD

Ventricular septal defect allows deoxygenated blood to cross to the left side.

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Pathophysiology (Simplified)

STEP-BY-STEP MECHANISM WITH ANATOMIC DIAGRAM



1 **Pulmonary stenosis** obstructs blood flow leaving the right ventricle toward the lungs.



2 Pressure builds in the RV → **right ventricular hypertrophy** develops as the wall thickens.



3 RV pressure exceeds LV pressure, forcing **deoxygenated blood right-to-left** through the VSD.



4 The **overriding aorta** receives *mixed* blood from both ventricles and sends it to the body.



5 **Hypoxemia + cyanosis** — systemic O_2 saturation falls. Chronic hypoxia drives **polycythemia** and **clubbing**.

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Signs & Symptoms

EARLY INFANCY VS. CHRONIC / LATE • RED FLAG HIGHLIGHTED

Early (Infant)

- ▶ **Cyanosis** worse with *crying, feeding, defecating*
- ▶ Loud **systolic ejection murmur** at left sternal border
- ▶ Poor feeding • tachypnea with feeds
- ▶ Failure to thrive (FTT)
- ▶ Easily tired • prolonged sleep
- ▶ SpO₂ typically 65–85%

Chronic / Late

- ▶ **Clubbing** of fingers and toes
- ▶ **Polycythemia** (compensatory ↑ RBCs) → ↑ stroke / thrombus risk
- ▶ **Squatting** in toddlers after exertion (compensatory)
- ▶ Delayed growth and development
- ▶ **Boot-shaped heart** on chest X-ray
- ▶ Risk of brain abscess, stroke, bacterial endocarditis

RED FLAG: TET SPELL (Hypercyanotic / “Blue” Spell)

Acute episode of **profound cyanosis, hypoxia, and irritability** — often triggered by crying, feeding, defecation, or waking. Infant becomes deeply blue, may have **syncope** or **seizures**. Murmur may *disappear* (no flow across stenosis). **This is a medical emergency.**

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Priority Nursing Interventions

ABC FRAMEWORK · SAFETY · NCLEX PRIORITIZATION

Routine / Maintenance Care

- ✓ **Monitor** SpO₂, HR, RR, and color continuously
- ✓ **Cluster nursing care** to minimize crying & agitation
- ✓ **Small, frequent feedings** with rest periods · use a soft preemie nipple
- ✓ Maintain **neutral thermal environment** (cold = ↑ O₂ demand)
- ✓ Strict **I&O**, daily weights, monitor for CHF
- ✓ **Iron-rich diet** — despite polycythemia, RBCs are iron-poor
- ✓ Adequate **hydration** — dehydration thickens blood, ↑ thrombus risk
- ✓ **Endocarditis prophylaxis** before dental and invasive procedures
- ✓ Up-to-date immunizations including **RSV (palivizumab)**, flu, pneumococcal

Surgical / Long-Term

- ✓ **Palliative:** Modified Blalock-Taussig (BT) shunt — used in neonates too small for full repair
- ✓ **Definitive repair** typically at **4–12 months** of age — close the VSD, relieve RV outflow obstruction
- ✓ Postop: monitor for **arrhythmias, low cardiac output, residual VSD, RV failure**
- ✓ Pre-op teaching for family · prepare for ICU stay



TET Spell Emergency Protocol — Memorize This Order

1

Knee-to-Chest

↑ SVR, ↓ R→L shunt —
push blood toward lungs

2

Calm & Comfort

Quiet, dim room — ↓ O₂
demand

3

100% O₂

Blow-by initially; mask if
tolerated

4

Morphine

↓ infundibular spasm &
agitation

5

IV Fluids · Propranolol

Volume + β-block;
correct acidosis with
bicarb

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Pharmacology

ACUTE SPELL + SUPPORTIVE MEDICATIONS

DRUG · CLASS	MECHANISM	KEY SIDE EFFECTS	NURSING CONSIDERATIONS
Morphine Sulfate Opioid	↓ Infundibular spasm · ↓ preload · sedation during TET spell	Respiratory depression · hypotension · constipation	Have naloxone at bedside · monitor RR & SpO ₂ · IV/SubQ during spell
Propranolol Beta-blocker	↓ HR & contractility · relaxes RV outflow tract spasm · prevents recurrent spells	Bradycardia · hypotension · bronchospasm · hypoglycemia	Hold for HR < 60 (or age-appropriate threshold) · take apical pulse · do not stop abruptly
Prostaglandin E₁ (Alprostadil) Prostaglandin	Keeps ductus arteriosus open in neonates → maintains pulmonary blood flow until surgery	Apnea · fever · flushing · hypotension	NICU only · continuous IV · have intubation equipment ready (apnea risk)
Furosemide (Lasix) Loop Diuretic	↓ Preload · treats CHF symptoms	Hypokalemia · dehydration · ototoxicity	Monitor K ⁺ · daily weight · I&O · caution — dehydration ↑ thrombus risk in polycythemia
Digoxin Cardiac Glycoside	↑ Contractility · ↓ HR for heart failure support	Bradycardia · N/V · yellow-green halos · dysrhythmias	Apical HR × 1 min · hold if < 90 (infant) / < 70 (child) · therapeutic 0.8–2.0 ng/mL
Sodium Bicarbonate Alkalizer	Corrects metabolic acidosis from severe hypoxia during spell	Hypernatremia · alkalosis · extravasation injury	Central line preferred · do not mix with calcium · ABG monitoring

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NCLEX Test Traps

WRONG VS. RIGHT · WHAT STUDENTS CONFUSE

❌ COMMON WRONG ANSWER “Discourage the toddler from squatting — teach the parents it’s a bad habit.”	✅ CORRECT THINKING Squatting is compensatory — it ↑ SVR and pushes blood through the pulmonary circuit. Allow and support it.	❌ COMMON WRONG ANSWER “During a TET spell, call the rapid response team first.”	✅ CORRECT THINKING Knee-to-chest position FIRST, then O₂ — these are independent nursing actions you do immediately. You can call for help <i>while</i> positioning.
❌ COMMON WRONG ANSWER “Lay the cyanotic infant flat to improve perfusion.”	✅ CORRECT THINKING Flat positioning ↓ SVR and worsens the right-to-left shunt. Use knee-to-chest (infant) or squatting (older child).	❌ COMMON WRONG ANSWER “TOF is the most common cyanotic defect in newborns.”	✅ CORRECT THINKING TOF is the most common cyanotic defect after age 1. In the immediate newborn period, Transposition of the Great Arteries (TGA) is most common.
❌ COMMON WRONG ANSWER “Polycythemia means the child has plenty of iron — restrict iron-rich foods.”	✅ CORRECT THINKING Polycythemia is a compensatory response to chronic hypoxia. RBCs are often iron-deficient. Give iron-rich foods and adequate hydration.	❌ COMMON WRONG ANSWER “A baseline SpO₂ of 80% on a TOF infant means immediate intubation.”	✅ CORRECT THINKING Chronic baseline SpO₂ of 65–85% can be expected. Compare to <i>this child’s</i> baseline. A drop from baseline matters more than the absolute number.

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Patient & Family Education

HOME MANAGEMENT • SAFETY • WHEN TO CALL

Recognize a TET Spell

Sudden deep cyanosis, irritability, hyperventilation, possible loss of consciousness. **Place baby in knee-to-chest position immediately** and call 911 if the spell doesn't resolve quickly.

Feeding Strategy

Small, frequent feeds. Use a **soft preemie nipple** with a larger hole. Burp often. Stop and rest if cyanosis or fatigue appears. Higher-calorie formula may be ordered.

Avoid Dehydration

Dehydration **thickens the blood** (polycythemia) and increases stroke/thrombus risk. Watch for vomiting, diarrhea, fever — push fluids and call provider.

Infection Prevention

Keep immunizations current. **RSV prophylaxis (palivizumab)**, annual flu vaccine. Avoid sick contacts. **Antibiotic prophylaxis before dental work** to prevent endocarditis.

Promote Calm

Avoid prolonged crying when possible. Respond promptly. Cluster routines. Maintain a comfortable temperature — cold & overheating both ↑ O₂ demand.

Call the Provider For

Worsening cyanosis · poor feeding · ↑ work of breathing · lethargy · seizure · fever · signs of dehydration. **911 for unresolved TET spell or unresponsiveness.**

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Memory Boosters & Mnemonics

PATTERN RECOGNITION • PICTURES THAT STICK

PROVe

THE FOUR DEFECTS

1. Pulmonary stenosis
2. Right ventricular hypertrophy
3. Overriding aorta
4. VSD (ventricular septal defect)

TET • K-O-M-P

TET SPELL SEQUENCE

1. Knee-to-chest position
2. Oxygen 100%
3. Morphine
4. Propranolol & fluids

BOOT

CLASSIC CXR FINDING

The heart looks like a **boot** on chest X-ray — small pulmonary artery + upturned cardiac apex from RV hypertrophy. *Coeur en sabot.*

4 = 4

PATTERN LOCK

4 letters in "Fall" (Fallot) = **4 defects**. Cyanotic = right-to-left shunt (blood goes the *wrong* way). Compare with the **3 acyanotic L→R defects: VSD, ASD, PDA.**



NGN CLINICAL JUDGMENT MEASUREMENT MODEL (NCJMM)

Full 6-Step Scenario

Scenario: A 7-month-old infant with known Tetralogy of Fallot is brought to the pediatric clinic by their parent for a routine immunization visit. While in the waiting room, the infant becomes inconsolable, develops deep cyanosis around the lips and nail beds, and is breathing rapidly. SpO₂ on the pulse oximeter reads **62%** (baseline 80%). HR 184, RR 64. The infant's previously loud murmur is now difficult to auscultate.

Step 1

Recognize Cues

Acute cyanosis, irritability, SpO₂ drop from 80% → 62%, tachypnea (64), tachycardia (184), **vanishing murmur**. Triggers: clinic visit, possible crying/agitation.

Step 2

Analyze Cues

Findings are consistent with a **TET (hypercyanotic) spell**. Vanishing murmur = **no flow across pulmonary stenosis** — severe R→L shunt. Hypoxia → acidosis if untreated.

Step 3

Prioritize Hypotheses

Primary: TET spell (highest priority — airway/breathing/circulation compromise). Rule out: pneumonia, sepsis, dehydration — but symptoms + history point to spell first.

Step 4

Generate Solutions

- ① Knee-to-chest position immediately ·
- ② Calm, quiet environment ·
- ③ 100% O₂ blow-by ·
- ④ Call provider/RRT ·
- ⑤ Prepare morphine, IV access, propranolol ·
- ⑥ Anticipate bicarb for acidosis.

Step 5

Take Action

Nurse places infant in knee-to-chest on parent's lap, applies blow-by O₂, calls for help, draws up morphine per order, secures IV, monitors continuous SpO₂ & cardiac rhythm.

Step 6

Evaluate Outcomes

Effective: SpO₂ rises to baseline (≥ 80%), color improves, infant calms, murmur returns, RR & HR trend down.
Ineffective: persistent cyanosis, ↓ LOC → escalate, transfer to ED/ICU.