Tetralogy of Fallot

What the Nurse Caring for a Patient with CHD Needs to Know

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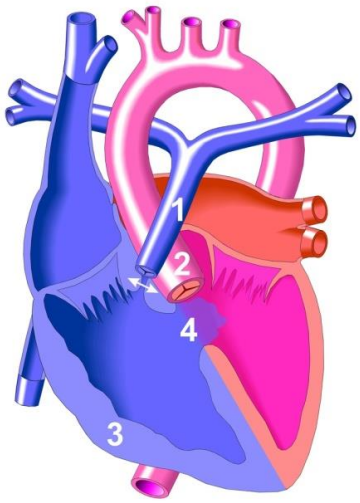
Embryology

- Most common cyanotic congenital heart disease (CHD)
  - 7 – 10% of all CHD
  - Spectrum of severity
- Normal development
  - The heart starts as a tube
  - Two sections of the tube, grow towards each other
    - During 5-6th weeks of gestation
    - Truncus arteriosus (TA) and the bulbus cordis (BC)
    - TA twists 180° as it grows down towards the BC
    - Twisting separates the aorta and the pulmonary artery
    - Transition point also point of formation of semilunar valves
  - Bulbus cordis
    - Contains genetic material from cardiac neural crest cells
      - Contributes to significant proportion of TOF patients with syndromic or genetic conditions
    - Contributes to formation of
      - Truncal septum
      - Perimembranous ventricular septum
- Abnormal development
  - Occurs in cono-truncal area of heart
    - Anterior deviation of the twisting results in abnormal truncal septation and ventricular septal formation found in tetralogy of Fallot (TOF)
  - Four distinct anatomic features include:
    - Smaller right ventricular outflow tract (RVOT) and pulmonary valve (PV) (Number 1 in illustration below)
    - Impeded flow from right ventricle (RV)
    - Anterior malaligned ventricular septal defect (VSD) (Number 4 in illustration below)
    - Enlarged aortic root which overrides the VSD (Number 2 in illustration below)

        - Right ventricular hypertrophy (RVH) develops as a result of the RV pumping against the small RVOT and PV (Number 3 in illustration below)

Anatomy (See illustration below)

- Right sided obstruction may occur at three levels. (Number 1 in illustration below)
    - Obstruction along the RVOT
    - Hypoplasia/stenosis of the PV
    - Stenosis of the pulmonary arteries (PAs)
- Aorta (Number 2 in illustration below)
    - Sits over the VSD, called an 'overriding' aorta
    - Aortic root dilation
        - Occurs with significant sub-pulmonary stenosis and right to left shunting across the VSD
        - Results from increased blood flow across the aortic valve (AV)
    - Right-sided aortic arch in ¼ to 1/3 of patients with TOF
- Right ventricular hypertrophy (RVH) (Number 3 in illustration below)
    - Results from RVOT obstruction
    - Worsens over time
- VSD (Number 4 in illustration below)
    - Large perimembranous defect
    - Has extension into subpulmonary area
- Coronary arteries
    - Abnormal in 5% of patients
    - Anterior descending branch of right coronary artery (RCA) most common
        - Crosses RVOT where incision usually made
        - Must be identified prior to surgical repair



Tetralogy of Fallot

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- Variants of TOF (See Defect Documents in these guidelines for each variant)
    - TOF with pulmonary atresia (PA)
    - TOF with PA and with multiple aorta-pulmonary collaterals (MAPCA's)
    - TOF with absent pulmonary valve (APV)
    - TOF with double outlet RV
    - TOF with atrial septal defect (ASD)
        - Called pentalogy of Fallot
        - May have a small LV

Physiology/clinical findings

- Clinical findings widespread
    - Depend on severity of RVOT obstruction and presence of a patent ductus arteriosus (PDA)
    - Range from significant to mild or no cyanosis
        - Mild cyanosis may become significant with spontaneous closure of PDA
        - Mild cyanosis from mild pulmonary outflow obstruction
            - Called "Pink tet"
            - May develop symptoms of pulmonary overcirculation with normal decrease in pulmonary vascular resistance
            - May protect pulmonary vascular from pulmonary hypertension
        - Even mild cyanosis will become more significant over time
            - Increase in infundibular stenosis
            - Development of polycythemia
- Neonate and children
    - Cyanosis
        - Cardinal symptom
        - Results from decreased flow through RVOT
            - Loud, harsh systolic ejection murmur (SEM)
                - Indicates obstructed flow across RVOT
                - Decrease in murmur may indicate decreased flow
                    - From decreased volume through RVOT
                    - From right-to-left shunting across VSD
            - Stenosis may occur at any and/or all levels
        - Secondary to right to left shunting across the ASD/VSD
        - Neonate with significant cyanosis
            - Pulmonary blood flow dependent on PDA
            - Maintain PDA with prostaglandin
        - Will increase over time due to increased RVOT obstruction
    - Increased RVOT obstructions
        - Will see increasing cyanosis
        - Leads to RVH
        - Increased infundibular stenosis from RVOT hypertrophy
        - May progress to severe sub-PS with very little forward flow through the RVOT

        - May hear decreased systolic ejection murmur (SEM) from decreased flow
    - Clubbing
        - Not evident at birth
        - Develops over time
- Hypercyanotic episodes ("Tet" spells)
    - Result from significant increase in right-to-left shunting across VSD with decreased ratio of pulmonary blood flow to systemic blood flow
    - Can occur at any time
        - Seems to be most frequent between 2-4 months of age
        - Not dependent upon severity of RVOT obstruction
    - Occur most frequently
        - With activities that increase oxygen consumption - crying, eating, vagal maneuvers (defecating, passing a nasogastric tube, suctioning, venous puncture to start IV or draw blood)
        - With changes in contractility due to endogenous catecholamines or hypovolemia - acidosis, anemia, fever, dehydration
        - Suspected pathology
            - Resistance of pulmonary vascular bed > resistance of systemic vascular bed
            - Dynamic changes in degree of subpulmonic obstruction
    - Symptoms
        - Increasing irritability
        - Diaphoresis
        - Severe cyanosis
        - Hyperpnea
        - Murmur significantly diminished due to increase in outflow tract obstruction
        - Rapidly become hypoxic with metabolic acidosis
        - Progresses to severe lethargy and possible death
    - Management
        - Aimed at decreasing pulmonary vascular resistance (PVR) and increasing systemic vascular resistance (SVR)
        - Quite baby/child
        - Place in knee-chest position
        - Administer 100% oxygen
        - Follow hospital or home protocol
            - Call for help
            - Administer morphine/sedative
            - Administer fluids
            - Administer beta blocker
            - Administer phenylephrine
            - Treat acidosis – sodium bicarbonate
    - Not seen as frequently due to earlier surgical intervention
    - Absolute indication for surgical intervention
    - "Squatting" seen in toddlers and older children

        - Due to posture of children who controlled own spells
        - Squatting increases systemic vascular resistance with self-imposed knee-chest position
- Repaired Adult with TOF
    - Pathology related to RVOT
        - RV dilation and dysfunction from increased volume load due to pulmonary valve regurgitation (PR)
        - Murmurs
            - To and fro murmur heard best at the left upper sternal border (LUSB) from PS and PR
            - Systolic ejection murmur on upper back from PA
            - Systolic murmurs on lateral lung fields from branch PA stenosis
    - Pathology related to tricuspid regurgitation (TR)
        - Chordal tissue of the septal leaflet of the tricuspid valve (TV) attach to the intraventricular septum
        - Leaflets separate as RV dilates resulting in TR
    - Pathology related to AV
        - Regurgitation may be present
        - Dilation of aortic root
        - Diastolic murmur from aortic valve regurgitation (AR)
    - Pathology of conduction system
        - Right bundle branch block (RBBB)
            - Almost always present and results from:
                - Ventriculotomy
                - Infundibular resection
                - RV dilation
            - S2 may be widely split
            - Risk of ventricular tachycardia and sudden death from RBBB > 180 milliseconds

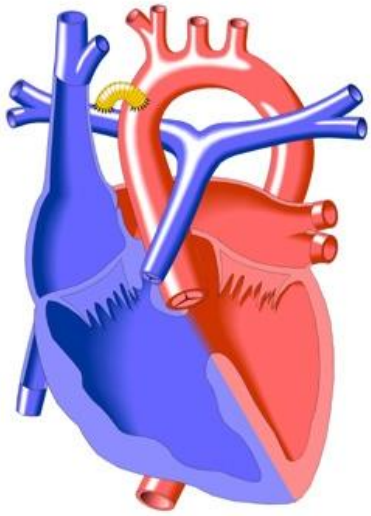
Interventions

- Cardiac catheterization
    - Diagnostic
        - Necessary if echocardiogram not definitive
        - Location of coronary arteries
        - Associated lesions
        - Post-surgical evaluation
            - Residual stenosis of RV outflow
            - Small pulmonary arteries
            - Hemodynamic assessment of residual VSD
    - Interventional
        - Balloon pulmonary valvuloplasty
        - RVOT stent
        - Coil embolization of collateral vessels
        - PV replacement (Melody valve)

- FDA approved for replacement of a stenosed and/or regurgitant PV in a valved conduit used in repair of TOF
- Specific criteria for conduit size, RV function
- Constructed with a venous bovine valve sutured inside a stent
- Placed inside the original conduit valve and stent with valve dilated to open stent
    - Dilation compresses the original conduit valve
- Functions as new PV
- Risk endocarditis

- Surgical intervention
    - Type and timing of intervention dependent on patient symptoms, associated cardiac and noncardiac lesions and institutional preference
    - Cyanosis
        - Neonates with severe cyanosis and ductal dependent pulmonary blood flow
            - Complete repair, often with maintenance of PFO
            - Aorto-pulmonary shunt and complete repair at 4-6 months of age
    - Infants with increasing cyanosis
        - Medical management
            - Generally related only to hypercyanotic spells
            - Relax infundibular muscle and potentially slow development of further infundibular obstruction with beta blocker (Propranolol)
            - Avoid diuretic and dehydration
            - Maintain adequate hematocrit
            - Spells or increasing cyanosis require surgical intervention and medical therapy only temporizing
        - Repair depending on coronary anatomy, patient size
            - With RCA across RVOT
                - Palliation with aorto-pulmonary artery shunt such as Modified BT shunt or central shunt
                - Complete repair with RV to PA conduit in infancy or if palliated at 6-12 months of age
            - Normal coronary anatomy
                - Palliation with aorto-pulmonary artery shunt
                - Primary repair in asymptomatic patients per institutional preference at 3-6 months of age
        - Repair depending on PV anatomy, patient size
            - Depends on institutional preference
            - Early repair vs cardiac catheterization with intervention
                - Balloon valvuloplasty of pulmonary valve
                - RVOT stent
    - Hypercyanotic spells
        - Repair should be scheduled after first spell
        - Goal to repair BEFORE spell

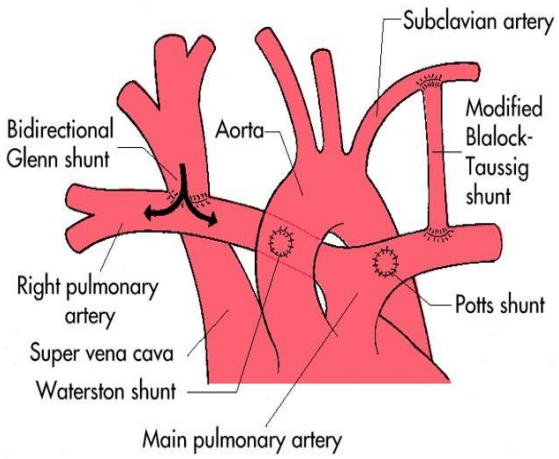
- Palliative Shunts (See illustrations below for types and locations of shunts)
  - Classic Blalock-Taussig Shunt (BT shunt)
    - Subclavian artery anastomosed to pulmonary artery
    - First performed in 1944
    - Palliative procedure done in infancy with complete 'repair' later in life
    - Thoracotomy scar on the side where the shunt placed
    - Diminished pulse on affected arm due to use of subclavian artery to supply blood to the pulmonary arteries
    - Arm on operated side should not be used for blood pressure assessment or to draw blood
    - Rarely used
  - Modified BT shunt
    - Connection with Gore-Tex tube graft between subclavian artery and pulmonary arteries
    - Pulse and BP should be obtainable in both arms
    - Most common of palliative aorto=pulmonary artery shunt
    - Modification includes Gore-Tex graft placed closer to bifurcation of LPA and RPA (Central shunt)



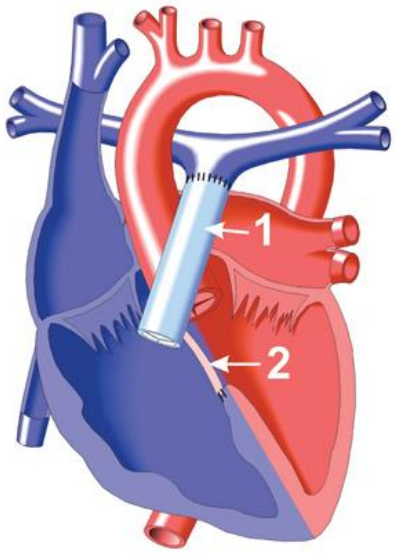
Tetralogy of Fallot with Modified Blalock-Taussig Shunt

  - Waterston shunt
    - Ascending aorta connected to main or right pulmonary artery (RPA)
    - May lead to pulmonary hypertension

- Rarely used
    - Difficult to control pulmonary blood flow (PBF)
    - Distorted RPA
- Potts Shunt
    - Descending Aorta connected to left pulmonary artery (LPA)
    - Rarely used as noted with Waterston



- Primary TOF Repair
    - Closure of VSD through tricuspid valve
    - Resection of RVOT obstruction
        - Limited muscle resection, division muscle bundles
    - Transannular patch and/or valvuloplasty or annuloplasty
        - Depends on PV z-score
        - Valvuloplasty/annuloplasty without transannular patch best long-term outcomes
    - Augmentation of PAs
        - Generally in tetralogy of Fallot, PA size is normal, confluent without collaterals
    - RV to PA conduit
        - Per institutional preference, utilized by some routinely instead of transannular patch to avoid pulmonary regurgitation especially in neonatal repairs
        - Utilized in patients with anomalous RCA to avoid TAP into coronary artery



Repair of Tetralogy of Fallot with RV to PA Conduit

- Repeat intervention in the pediatric patient may include:
  - PV dilation
  - RVOT dilation/reoperation
  - RV-PA conduit replacement
  - Closure of residual VSDs
  - Repair of TV
  - Pulmonary valve replacement
- Repeated intervention in the adult patient may include:
  - PV replacement (surgical or catheter)
  - Relief of RVOTO
  - RV-PA conduit change
  - PA augmentation
  - Cavo-tricupsid isthmus ablation (MAZE procedure for intractable arrhythmias) (See Adult Guidelines on Arrhythmia Management)
  - TV replacement or annuloplasty
  - Aortic root replacement
  - Aortic valve replacement
  - Closure of residual VSD's or ASD's

Specific considerations and routine care

- Preoperative considerations
  - Neonates with tetralogy of Fallot
    - Management based on the degree of outflow tract obstruction
    - Mild to moderate outflow tract obstruction

        - Anticipate discharge until surgery scheduled between 4-6 months
        - Discharge teaching for parents/caregivers - specific information for hypercyanotic ("Tet") spells
            - Triggers
                - Mornings
                - Crying
                - Post bathing
                - Vagal stimulation – defecation, vomiting
                - Dehydration
            - Signs/symptoms of "Tet" spell
                - Irritability
                - Inconsolable
                - Sudden increase in cyanosis
                - Hyperpnea
            - Interventions for "Tet" spells
                - Hold and quiet baby
                - Place in knee-chest position
                - Administer oxygen (if available at home)
                - Call for help (Plan for local support)
            - When to call cardiologist
                - Diarrhea, vomiting, fever
                - Hypercyanotic spell
- Preoperative Management
    - Initial care
        - Maintaining adequate oxygenation/hydration
        - Anticipate/preventing hypoxic spells
            - Hold and quiet baby
            - Call for help
            - Place in knee-chest position
            - Administer oxygen
            - Administer morphine sulfate
            - Administer volume
            - Treat anemia if present
            - Sedation
        - Identify comorbidities
            - Associated cardiac defects – PA, Atrioventricular Septal Defects, ASD
            - Syndrome/genetic defects
                - 10% with chromosomal abnormalities
                - Complete chromosomal analysis and/or fluorescent in situ hybridization (FISH) analysis prior to by-pass surgery
                - Most common 22q11 microdeletion (DiGeorge)
                    - Provide leuko-reduced, irradiated, red blood cells for surgery

            - Parathyroid dysfunction leads to poor regulation of calcium with hypocalcemia
            - Immunodeficiency from abnormal T-cell response predisposes to infections
            - Physical defects including abnormal facies and oral/pharyngeal defects, kidney and GI abnormalities
        - Prevent hospital acquired complications (See Peds/Neo Guidelines for Infection Prevention)
            - Air emboli from intravenous lines
                - Monitoring all venous lines
                - Use on 0.2 micron air filters on all venous lines
            - Infections- use all Infection Prevention Bundles (central line, endotracheal, urinary catheter)
            - Cerebral thromboembolic events from dehydration, anemia
            - Hypercyanotic event
                - Manage painful events – starting IVs, phlebotomies
                    - Consider topical analgesics, Sweeties, pacifiers, swaddling
                - Ensure adequate hydration
                    - Oral hydration with clear liquids up to 2 hours preoperative/preprocedure
                    - Start intravenous fluids for longer periods of NPO
    - Initial care for tetralogy of Fallot with severe PS or Pulmonary Atresia (TOF/PA) (See Peds/Neo Guidelines for tetralogy of Fallot with Pulmonary Atresia)
        - Immediate stabilization with PGE_1 infusion
        - With closure of ductus arteriosus may see profound cardiogenic shock
            - Ensure administration of intravenous prostaglandins (PGE_1)
            - Control/provide ventilation – intubation with mechanical ventilation
            - Support ventricular function and cardiac output – continuous infusion of inotropes
            - Assess/manage end organ complications
- Postoperative Management (See Peds/Neo Guidelines for Postoperative Care, Infection Prevention, Hemodynamic Monitoring, Nutrition, Developmental Care, Arrhythmia Management)
    - Residual valvar stenosis or regurgitation
        - Significant residual stenosis may prolong RV dysfunction, worsen TR and increase right to left atrial shunt and hypoxemia
        - PR well tolerated early postop
        - PR progression over time results in:
            - RV dilation
            - Deceased function
            - May necessitate re-intervention on TV and/or RVOT/**PV**
    - Hypoxemia

        - Potential cause: Inadequate relief of PS, RV dysfunction with shunting through residual atrial defect
        - Management if profound hypoxemia: Re-intervention in catheterization lab or operating room
    - Low cardiac output (CO)
        - Potential cause: Injury to TV apparatus, residual VSD, RV dysfunction, arrhythmia
        - Management, if necessary: May require TV repair, VDS repair, vasoactive support
    - RV dysfunction with decreased compliance
        - Monitor volume status closely
            - Maintain adequate preload for non-compliant RV
            - Avoid hypovolemia
        - Avoid tachycardia
            - Improve preload to RV
            - Allow time for ventricles to empty
        - May require support with inotropic agent and/or Milrinone
        - Prevent increased RV afterload
            - Good pulmonary mechanics and ventilation
            - Avoid effusions, pneumothorax, hemothorax, atelectasis/collapse
    - Arrhythmias (See Adult and Peds/Neo Guidelines for Arrhythmia Management)
        - Associated with:
            - VSD closure
            - RV muscle resection
            - RVOT Patch angioplasty or placement of RV to PA conduit
            - RV dysfunction
        - Potential arrhythmias
            - Junctional Ectopic Tachycardia (JET)
                - Potential for significant hemodynamic compromise
                - Reduce degree of hemodynamic impairment
                    - Early recognition
                    - Prompt treatment
                        - Cooling to core temperature less than 36 degrees
                        - Antiarrhythmic mediations
                        - Sedation
                        - Overdrive pacing
            - Atrial tachycardia
            - Ventricular tachycardia
                - Rare in younger child
                - More common in adult
                - Monitor electrolytes, provide electrolyte management
            - Heart block uncommon but possible due to VSD closure

Long Term Complications/Follow-up Care (See both Peds/Neo and Adults Guidelines specific to these complications – Cyanosis, Arrhythmia Management, Ventricular Dysfunction)

- Follow-up Care
  - Neonatal, pediatric, and adult patients require periodic, routine follow-up
  - Should be done by age appropriate pediatric and/or adult cardiologist trained in CHD at a center that provides comprehensive CHD care
  - Frequency depends on complications, time/type of repair and symptoms
    - Minimum - annually
  - Assessment includes monitoring of rhythm, valvar and ventricular function
- Right ventricular dysfunction
  - Tricuspid regurgitation
  - Coronary artery injury
  - Pulmonary valve regurgitation
    - Leads to RV dilation/dysfunction
    - Symptoms
  - Residual/recurrent RVOTO: branch PA, PV, infundibular
    - Branch PA stenosis (make bullet under RVOTO)
    - May develop at the site of the original BT shunt
    - May have been present since birth but never adequately repaired
- Aortic regurgitation +/- aortic root dilation
  - May result from damage to valve from VSD closure
  - May be intrinsic aortic root abnormality
- LV dysfunction
  - Systolic heart failure
  - Diastolic heart failure
- Atrial arrhythmias
  - Common arrhythmias - AV block, atrial flutter, and/or atrial fibrillation
  - May be result of atrial dilation from TR or PR with ventricular dilation
- Ventricular arrhythmias
  - Most common - sustained ventricular tachycardia
  - Potential causes
    - Ventriculotomy for conduit or transannular patch (TAP)
    - Long standing ventricular dilation from PR
    - Conduction delay from VSD closure
- Sudden death
  - Generally arrhythmia related
  - Consideration for ICD implantation
- Endocarditis (See 2015 American Heart Association for SBE Prophylaxis Guidelines for Adults and Children)

Management during Pregnancy

- Challenges in cardiovascular and maternal-fetal management for pregnant patient with cardiac disease
  - Physiologic changes during and after pregnancy in patients without cardiac disease
    - Changes upon the cardiovascular system

            - Increase in plasma volume by 50%
            - Increase in resting pulse by 17%
            - Increase in cardiac output by 50%
        - After delivery
            - Normalization of heart rate within 10 days
            - Return to pre-pregnancy state by 3 months
                - Stroke volume
                - Cardiac output
                - Systemic vascular resistance
    - Impact of cardiac pathology on pregnancy
        - Regurgitant lesions tolerated fairly well
        - Increased risk with
            - Obstructive lesions, especially left-sided lesions
            - Arrhythmias
- Patients with TOF need a plan for pregnancy (See Adult Guidelines on Pregnancy and Congenital Heart Disease)
    - Pre-pregnancy visit with ACHD trained cardiologist
        - Identify risk factors
        - Modify risk factors as needed before pregnancy
    - Plan should include
        - Partnership with ACHD cardiologist and high-risk obstetrical/fetal medicine
        - Birthing site plan

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