Truncus Arteriosus

What the Nurse Caring for the Patient with Congenital Heart Disease Needs to Know

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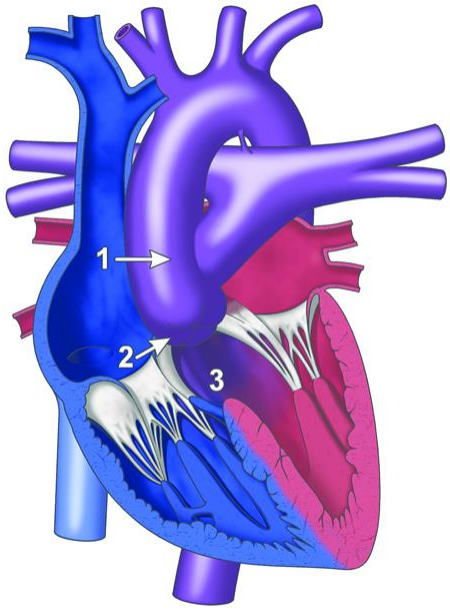
Embryology

- Rare congenital heart disease
- Before the 4th week of embryonic life
  - Endocardial tube expands, elongates and develops areas of dilation
    - Includes bulbus cordis
    - Ventricular outflow tracks
    - Truncus arteriosus
  - Results in cardiac looping – completed by day 28 of gestation
    - Stretching creates torsion in truncus arteriosus
    - Contributes to formation of spiral septum
- Septation of the truncus arteriosus – day 26 to 42
  - Mesenchymal cells
    - Include neural crest cells
    - Actively proliferate
    - Form ridges in bulbus cordis and truncus arteriosus
    - Ridges create 180 degree spiraling
    - Create aorticopulmonary septum
    - Contribute to closure of conal ventricular septum
  - Spiraling enhanced by forward blood flow
  - Failure of septation results in:
    - Truncus arteriosus (TA)
    - Large ventricular septal defect (VSD)
- Development of semilunar valves
  - At base of the truncus arteriosus
    - Result from:
      - Swelling of endocardial tissue
      - Endocardial cushions
  - Failure of septation
    - Results in one valve
    - Valve abnormal
      - Usually have more than 3 cusps
      - Cusps usually thickened and deformed
      - May be regurgitant
      - Sometimes stenotic
- Neural crest cells
  - Arise from genetic material
  - Influence the development of thymus and parathyroid glands from the pharyngeal pouches

- Result in increased prevalence of DiGeorge Syndrome (micro deletion of chromosome 22 q 11) in truncus arteriosus
  - Abnormal calcium regulation
  - Abnormal glucose regulation
  - Immunodeficiency

Anatomy

- Large, single, arterial trunk
  - Arises from the base of the heart (As indicated by #1 in Illustration below)
  - Gives rise to pulmonary, systemic, and coronary circulations
- Ventricular septal defect (VSD) (As indicated by #3 in Illustration below)
- Single semilunar valve (As indicated by #2 in Illustration below)



Truncus Arteriosus Type I

- Four types of truncus arteriosus (TA) as described by Van Praagh
  - Type determined by origin of pulmonary arteries
  - Type I: Short main pulmonary trunk arises from truncus (As illustrated above)
  - Type II: No main pulmonary artery trunk. Both branch pulmonary arteries arise separately from trunk
    - May arise close to one another from the left posterolateral aspect of the truncus
    - May arise from distinct and separate locations

    - Type III: Aorticopulmonary septum absent
        - Only one of branch pulmonary arteries arises from trunk
        - Blood flow to ipsilateral lung from collateral vessel(s)
    - Type IV: Abnormality includes aorta and aortic arch
        - Blood flow to descending aorta via a patent ductus arteriosus
        - Main, right, and/or left pulmonary arteries comprise trunk
- Associated cardiac conditions
    - Pulmonary artery stenosis
    - Absent pulmonary artery (s)
    - Underdeveloped aortic arch
    - Interrupted aortic arch
    - Abnormal coronary arteries
- Associated non-cardiac conditions
    - Most relate to DiGeorge Syndrome
    - DiGeorge Syndrome (Chromosome 22q11.2 deletion)
        - Also called Velo-Cardio-Facial Syndrome (VCFS)
        - Immunodeficiency disorder
            - Thymic gland dysfunction
            - Parathyroid glands not fully developed or complete absent
                - Various degree of poor calcium regulation
                - Depending on extent of gland tissue missing
        - May be asymptomatic
        - May have many different symptoms, both major and minor
            - **CATCH 22 – Syndrome**
                - **C**ardiac abnormality (especially tetralogy of Fallot)
                - **A**bnormal facies
                - **T**hymic aplasia – decrease immunity
                - **C**left palate
                - **H**ypocalcemia/**H**ypoparathyroidism
            - Facial features
                - Eyes – wide-set (hypertelorism), down-slanting
                - Low-set ears
                - Prominent nose with squared nasal root
                - Micrognathia (Small size of lower jaw)
            - Behavioral concerns
                - Learning disorders
                - Impulsive
                - Uninhibited
                - Affectionate
                - Can function socially
            - Kidney abnormalities (Hydroureter)
            - Significant feeding difficulties as babies
            - Gut malrotation, decreased motility
    - Skeletal deformities

Physiology

- Clinical Features
    - Cyanosis
        - Usually evident in the neonatal period
        - Volume of pulmonary blood flow
            - Decreased flow may result from:
                - Stenosis of pulmonary arteries
                - Pulmonary vascular resistance
                - May protect lungs from pulmonary overcirculation
        - Cardiac output from both ventricles
            - Complete intracardiac mixing
            - Combined ventricular output through single trunk
    - Heart failure
        - Overwhelming abnormal finding
        - Presence of significant truncal valve insufficiency – 50% of patients
            - Increased systemic ventricle volume load
            - With severe insufficiency, heart failure may be present in neonatal period
        - Large left-to-right shunt
            - Increases as neonatal pulmonary vascular resistance (PVR) falls
                - Output nearly equal at birth
                - Pulmonary flow increases to around 3 times systemic flow
                - Results in pulmonary overcirculation
            - With increased shunt see decrease in cyanosis
- Pulmonary vascular obstructive disease
    - See with later repair (Beyond infancy)
    - Results from increased pulmonary blood flow
        - Congestive heart failure
        - Leads to pulmonary hypertension
    - May develop into Eisenmenger syndrome (See Adult Guidelines on Eisenmenger Syndrome)

Medical Management/Procedures/Interventions

- Neonate
    - Most have some symptoms of congestive heart failure
    - Medical care depends on clinical presentation
        - Hypoxemia
            - Due to complete mixing of pulmonary and systemic circulations
            - Decreased pulmonary blood flow due to normally elevated pulmonary pressures
            - Do NOT administer supplemental oxygen
                - Will lead to pulmonary vasodilation
                - Will increase risk of pulmonary overcirculation

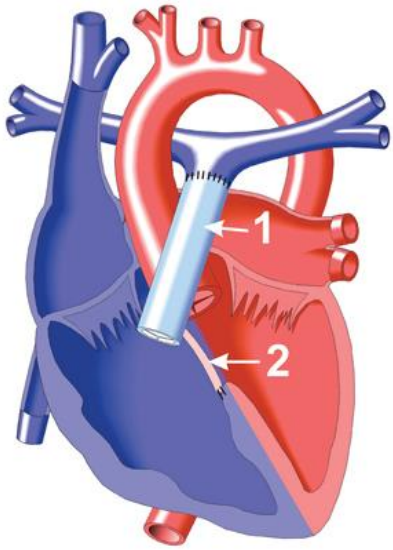
            - May administer if oxygen saturation less than 75%
        - Treatment of heart failure
            - Digitalis
            - Diuretics
            - Vasodilators
        - Medical management usually necessary prior to surgical intervention
        - Prevent secondary injury
            - Neurological injury
                - Mixing of venous and arterial blood
                - May result from air emboli
                - Use air filters on ALL intravenous lines
                - Avoid hemoconcentration (from aggressive diuresis)
            - Pulmonary dysfunction (See Peds/Neo and Adult Guidelines on Pulmonary Hypertension)
                - Avoid use of supplemental oxygen
                - Avoid hypocarbia
            - Poor nutrition (See Peds/Neo Guidelines on Nutrition)

    - Non cardiac differential diagnosis
        - Sepsis
            - Present with signs and symptoms of shock
            - Symptoms from high output heart failure with significant pulmonary overcirculation
                - Resemble the presentation of neonatal sepsis
                - Occur with higher ratio of pulmonary-to-systemic blood flow
                - Do not include cyanosis
        - Necrotizing enterocolitis (NEC)
            - Either preoperatively or postoperatively
            - Requires appropriate evaluation to rule out NEC

- Cardiac catheterization
    - Infrequently done preoperatively
    - May be necessary to define:
        - Truncal anatomy
        - Anatomy and location of pulmonary arteries
        - Location of coronary arteries
    - Post operatively
        - Evaluate status of pulmonary vascular disease
        - Evaluate surgical repair
        - Intervene in stenosis at any site
    - Manage stenosis
        - Conduit
        - Suture lines in pulmonary arteries

        - Percutaneous placement of pulmonary valve with Melody valve in older teens and adults

- Surgical intervention
    - Invariably requires operative repair
        - Symptoms usually develop in the early neonatal period
        - Complete repair indicated when symptoms not controlled with medical management
    - Palliative intervention
        - Pulmonary artery banding
            - Initial procedure done in 1967
            - Usually before 6 weeks of age
            - High risk operation
        - Complete repair in later infancy
        - Rare in 2015 - done only if increased risk with complete repair
    - Complete repair accepted standard
        - Started in 1980s
        - Procedure (Modified Rastelli Procedure as Illustrated below)
            - Closure of VSD
            - Commits outflow from LV to common arterial trunk
            - Reconstructs right ventricular outflow tract (RVOT)



Rastelli Procedure

        - RVOT reconstruction determined by location of pulmonary arteries

- Both branch pulmonary arteries arising from separately, but in close proximity, on posterior aspect of common trunk
    - Standard method of RVOT/PA reconstruction:
        - Remove central pulmonary arteries from the common trunk en bloc and attach to valved conduit
        - Place a valved conduit from RV proximally to the central PAs distally
- One PA arising from the common trunk and one from the underside of the aortic arch
    - PAs disconnected separately
    - PAs then anastomosed together, then anastomosed to the conduit
    - Anastomosed independently to conduit
- Reoperation to replace conduits required throughout life span
    - Due to somatic growth of infant
    - Due to calcification and/or degeneration of conduit and/or valve

- Repair of coexisting anomalies
    - As appropriate
        - With cardiopulmonary bypass, cardioplegia, and sometimes deep hypothermic arrest
        - Depends on
            - Anatomic features
            - Preference of surgeon
    - Increases risk of procedure
- Repair/replace truncal valve
    - Outcomes significantly worse with regurgitant truncal valve
    - If abnormal, repair at time of initial TA repair
    - May require replacement in infancy (See Defect Document on Aortic Valve and Peds/Neo Guidelines on Anticoagulation)

Specific Considerations

- **Neonatal/Infant** (See Peds/Neo Guidelines for Neonatal Care, Developmental Care, Nutrition)
    - Monitor for signs/symptoms of congestive heart failure
        - Degree and severity of symptoms depend upon degree of pulmonary blood flow
        - As pulmonary pressures fall with normal pulmonary vascular development
            - Pulmonary blood flow increases
            - Cyanosis decreases
            - Oxygen saturation increases
        - Development of heart failure accelerated with truncal valve regurgitation

    - Monitor/treat abnormal glucose and calcium regulation
    - Monitor for sepsis
    - Monitor for low cardiac output
    - Maintain adequate nutrition
- **Post-operative** (See Peds/Neo Guidelines for Postoperative Care, Nutrition, and Developmental Care)
    - Low Cardiac Output Syndrome
        - RV dysfunction from right ventriculotomy for conduit placement
        - Increased with pulmonary hypertension
        - Without atrial shunt (creation of small ASD), decreased RV output may lead to decreased LV function and poor systemic perfusion
    - Congestive heart failure
    - Pulmonary hypertensive crisis (See Peds/Neo Guidelines on Pulmonary Hypertension)
        - Anticipate if pulmonary pressures elevated prior to surgery
        - Meticulous attention to:
            - Nursing procedures, especially endotracheal tube suctioning
            - Alveolar oxygenation by:
                - Preventing/treating atelectasis
                - Treat pneumothorax
                - Avoid hypoventilation
                - Monitor for pulmonary edema
        - Decrease myocardial demand
            - Maintain normothermia
            - Maintain slight alkalosis
            - Manage agitation, pain, and sedation
    - Bleeding
        - Multiple suture lines, including right ventriculotomy and aortic repair
        - Prevent hypertension
        - Monitor/maintain coagulation factors within normal limits
        - Monitor/maintain normal hematocrit
        - Monitor for tamponade
    - Arrhythmias (See Peds/Neo Guidelines for Arrhythmia Management)
    - Prevent infection (See Peds/Neo Guidelines for Infection Prevention)
        - Know results of pre-operative genetic studies for DiGeorge Syndrome
        - Frequently have open chest (See Peds/Neo guidelines for Open Chest)
        - Follow infection prevention bundles for central lines, urinary catheters, endotracheal intubation/ventilation

Long-term Complications

- Requires life-long care by a cardiologist who specializes in congenital heart disease

- Conduit stenosis/replacement
- Truncal valve insufficiency /Aortic regurgitation
    - Valve repair
    - Valve replacement
    - Pulmonary vascular disease/Eisenmenger's syndrome (Refer to Peds/Neo and Adult Guidelines on Pulmonary Hypertension and Eisenmenger's syndrome for further discussion and management.)
    - Anticoagulation (See Peds/Neo and Adult Guidelines on anticoagulation)
- Progressive myocardial failure (Refer to Guidelines on Ventricular Failure for further discussion and management.)
- Provide endocarditis prophylaxis (See 2015 AHA Endocarditis Prophylaxis for both children and adults)
- Pregnancy (See Adult Guideline on Pregnancy in ACHD for further discussion and management)
    - Requires cardiology evaluation prior to pregnancy
    - Requires careful multidisciplinary coordination
    - Successful pregnancy and delivery has been achieved after complete repair
    - Increased risk to both woman and fetus (See Problem Sections for the complications listed below for further discussion and management)
        - Pulmonary vascular disease
        - Pulmonary hypertension
        - Aortic valve surgical interventions including valve replacement

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