Truncus Arteriosus

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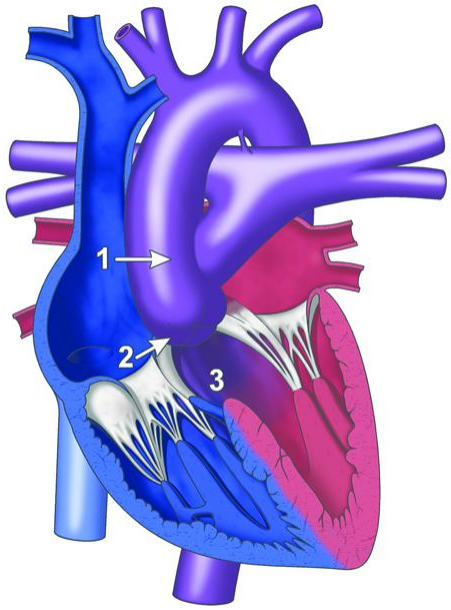
I. Embryology
- A. During embryonic life, the truncus arteriosus normally begins to separate and spiral into a pulmonary artery and aorta
- B. Neural crest cells develops into the pharyngeal pouches (thymus and parathyroid glands) this association could explain the prevalence of DiGeorge syndrome (micro deletion of chromosome 22 q 11)

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



Truncus Arteriosus Type I

D. Three types of truncus arteriosus
    1. Type I: Pulmonary trunk arises from truncus (As illustrated above)
    2. Type II: Right pulmonary artery (RPA) and left pulmonary artery (LPA) arise separately but close to one another from the left posterolateral aspect of the truncus
    3. Type III: Pulmonary arteries each arise from respective lateral aspects of the truncus

C. Associated conditions
    1. Skeletal deformities
    2. Hydroureter
    3. Bowel malrotation
    4. DiGeorge Syndrome
        a. Hypoglycemia
        b. Thymic dysfunction
        c. Mental retardation

III. Physiology

A. Large left-to-right shunt
    1. Overwhelming abnormal finding
    2. Increases as neonatal pulmonary vascular resistance (PVR) falls
    3. Results in pulmonary overcirculation

B. Truncal valve regurgitation
    1. Present in ~ 50% of patients
    2. May result in alterations in ventricular function from volume overload

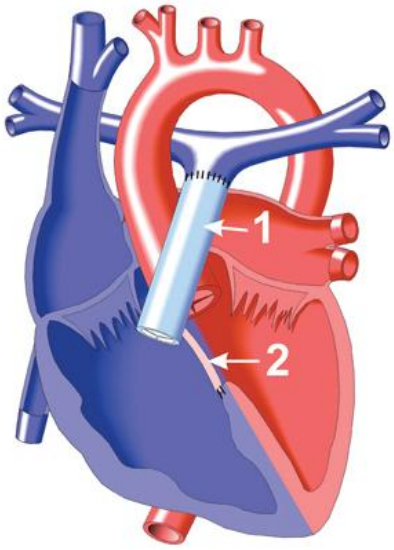
C. Pulmonary vascular obstructive disease with later repair (Beyond infancy)

IV. Types of Repairs

A. PA Band in neonatal period if risk of repair is prohibitive

B. Correction during infancy

C. Total correction (Rastelli Procedure)
    1. Patch closure of VSD
    2. Place a conduit between the right ventricle (RV) and pulmonary arteries (PA)



Rastelli Procedure

D. Truncal valve repair/replacement (See Problem Section on Valve Repair/Replacement for further discussion and management)

V. Long-term Complications
- A. Conduit stenosis/replacement
- B. Truncal valve insufficiency /Aortic regurgitation
  1. Valve repair
  2. Valve replacement
- C. Pulmonary vascular disease (Refer to Problem Section on Pulmonary Hypertension for further discussion and management.)
- D. Progressive myocardial failure (Refer to Problem Sections on Systemic and Pulmonary Ventricular Failure for further discussion and management.)

VI. Pregnancy (See Problem Section on Pregnancy in ACHD for further discussion and management)
- A. Requires cardiology evaluation prior to pregnancy
- B. Requires careful multidisciplinary coordination
- C. Successful pregnancy and delivery has been achieved after complete repair
- D. Increased risk to both woman and fetus (See Problem Sections for the complications listed below for further discussion and management)
  1. Pulmonary vascular disease
  2. Pulmonary hypertension
  3. Prosthetic valves

References:

Emmanouilides, et al: *Clinical Synopsis of Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including fetus and young adults*, Philadelphia, 1998, Lippincott, Williams & Wilkins.

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