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TRUNCUS ARTERIOSUS REPAIR

Surgical Specialty:	Cardiac Surgery
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Background:

- General Considerations
    - Truncus Arteriosus (TA) occurs due to failure of embryonic truncal septation, resulting in a single arterial trunk and semilunar valve for the pulmonary, systemic, and coronary circulations. It comprises 1-4% of congenital heart defects.
    - This lesion is classified based on the origin of the pulmonary arteries in relation to the common trunk. The two systems for describing the anatomy are called the Collett and Edwards (described in 1949) and Van Praagh (described in 1965) classification.[1]
        - The type of truncus does not necessarily affect anesthetic management but does give an understanding to the surgical approach.
    - This lesion is typically associated with a large non-restrictive ventricular septal defect (VSD), which results in systemic and pulmonary venous blood mixing in the ventricles before being ejected via the common truncal root.
        - Below are two links demonstrating the 3-dimensional anatomy of this lesion (links obtained with permission from the Paul W. Weinberg Cardiac Registry at CHOP).
            - https://sketchfab.com/3d-models/91-truncus-arteriosus-type-1-fbf367bde00848c7af341e52c4b4b57f
            - https://sketchfab.com/3d-models/93-truncus-arteriosus-type-1-1a178d2eb4c0499c9e2b22727fd4de62
    - The resulting arterial oxygen saturation is dependent on the delicate balance between the pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR). This ratio of blood flow through the pulmonary and systemic circulation is referred to as Qp:Qs.
        - $\frac{Qp}{Qs} = \frac{\text{Aortic Oxygen Saturation-Mixed Venous Oxygen Saturation}}{\text{Pulmonary Venous Oxygen Saturation-Pulmonary Arterial Oxygen Saturation}}$
        - In a mixing lesion when Qp:Qs is balanced 1:1, as in the newborn period, oxygen saturation is typically around 80%.
        - If Qp:Qs is >1, oxygen saturation may be high (>90%), at the expense of systemic perfusion.
        - If Qp:Qs is <1, systemic blood flow will increase at the expense of pulmonary blood flow, resulting in hypoxemia.

        - Following birth, PVR decreases, resulting in an increase in Qp:Qs. This increased pulmonary circulation results in decreased systemic and coronary perfusion, and congestive heart failure (CHF). Additionally, over time this overcirculation causes vascular endothelial and smooth muscle changes, resulting in pulmonary arterial hypertension if left untreated.
        - Ventricular end-diastolic and subendocardial pressure is elevated, compromising coronary perfusion and increasing the risk for ischemia and ventricular arrhythmias.
    - The majority of cases are diagnosed prenatally; post-natal diagnosis is usually detected once pulmonary vascular resistance (PVR) has dropped, resulting in neonatal CHF symptoms, including tachypnea, increased work of breathing, poor feeding, failure to thrive, and cool extremities with delayed capillary refill.[2]
- Patient Considerations
    - Disease Specific Considerations
        - Without surgery, the mortality rate is 74-100% within the first year of life due to CHF or multisystem organ failure from poor systemic perfusion.
        - Definitive repair is typically performed in the neonatal period, prior to the development of pulmonary vascular disease.
            - Patients too small for full repair may undergo pulmonary artery banding as a way to limit pulmonary overcirculation while awaiting definitive repair.[1]
        - Truncal valve pathology
            - The truncal valve is tricuspid in 60-70% of cases, though may have multiple cusps, and the valve may be insufficient or stenotic.
                - Truncal valve insufficiency can result in a more rapid onset of CHF due to increased ventricular end-diastolic pressure and decreased aortic diastolic pressure, resulting in decreased coronary perfusion.
                - Truncal valve pathology carries the highest risk for increased mortality.
        - Coronary Anatomy
            - Coronaries in close proximity to the origins of pulmonary arteries may be in jeopardy during reconstruction of the aorta
            - Aberrant coronaries on the right ventricle may be at risk for compression by the newly constructed right ventricular outflow tract. [3]
    - Associated Comorbidities/Syndromes
        - Chromosome 22q11 deletion (present in 30-40% of cases)
        - Other cardiac anomalies include atrial septal defect, coronary artery variations, right aortic arch, and interrupted aortic arch.[2]

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Evaluate for signs of CHF, as described above
        - Pulmonary over-circulation is typically managed with diuretics, afterload reduction, and inotropes pre-operatively
    - Transthoracic Echocardiography (TTE) with particular attention to the origin and size of the pulmonary arteries (PA), size and location of the VSD, anatomy (expound on

which anatomy) , and function of the truncal valve, coronary artery anatomy, and ventricular function.
  - Chest X-ray may demonstrate cardiomegaly and signs of pulmonary congestion.
  - Evaluate for associated comorbidities
    - Chromosome 22q11 deletion may have variable presentation and is associated with hypocalcemia, craniofacial abnormalities, and absent thymus (thus requiring blood product irradiation).
  - Discussions to have with surgeon/family
    - The early mortality rate is 3-20%, with increased risk in cases requiring truncal valve repair. However, those who survive 1 year following repair have 93% survival at 20 years.[4]
    - Code status
    - ECMO candidacy
- Specific or Unique Room Set-up Requirements
  - Airway
    - Consider nasal or oral endotracheal tube per institutional protocol
  - Drugs/Infusions
    - Anti-fibrinolytic (tranexamic acid or aminocaproic acid) bolus and infusion
    - Heparin, protamine
    - Calcium chloride or calcium gluconate
    - Inotropic support and afterload reduction medications (dopamine, epinephrine, milrinone) should be readily available following cardiopulmonary bypass
    - Sedative and narcotic infusions for transport to the ICU
  - Monitors
    - Standard ASA monitors prior to induction. Neonates may have an umbilical arterial catheter (UAC) in place. Peripheral arterial access location may depend on arch orientation and anatomy, but most commonly is on the ipsilateral side of the innominate artery.
    - Central venous access (location may be institution-dependent including femoral, internal jugular, or atrial)
      - If there is pre-existing umbilical access, verify correct positioning of the line with X-ray.
    - Consider multisite near-infrared spectroscopy (NIRS) to detect low system oxygen delivery (DO_2).
    - Transesophageal Echocardiography (TEE)
  - Blood Availability
    - Blood prime will be required for cardiopulmonary bypass (CPB) given the patient's age and weight. These blood products should be irradiated given the high incidence of 22q11 in this population.
    - Neonates can be prone to coagulopathy following CPB, therefore cryoprecipitate, platelets, and fresh frozen plasma (FFP) should be readily available following CPB.
  - Other Indicated Resources: perfusionist, cardiology for intra-operative TEE

Intraoperative Considerations:

- General
  - Surgical Considerations

        - Pulmonary vessel loop pre-bypass can help reduce pulmonary flow and increase systemic flow if signs of hypotension and/or coronary ischemia are present
        - Cardiopulmonary bypass cannulation strategy
            - The aortic cannula is placed near the base of the innominate artery to allow sufficient room for the aortic cross-clamp above the origin of the branch PAs from the common arterial trunk.
            - Bicaval or single atrial venous cannulation
            - For cases requiring arch reconstruction, deep hypothermic circulatory arrest (DHCA) may be performed (institution-dependent). If antegrade cerebral perfusion (ACP) is performed, a right arm arterial line is preferred. Goal pressures should be established with the surgeon and perfusionist.
            - If truncal valve insufficiency is present, the cardioplegia can cause left ventricular distention if not applied properly. It may require direct infusion into coronaries.
        - Following initiation of cardiopulmonary bypass, surgical aims consist of:
            - Separation of the great vessels
            - VSD closure
            - Establishing a new source of pulmonary blood flow, often accomplished with a conduit from the RV to the PA. This requires a right ventriculotomy.
            - Repair of truncal valve insufficiency (if applicable)[2]
- Induction
    - TA carries the highest risk of intraoperative cardiac arrest of any congenital heart surgery[2]. Venous access is preferred prior to induction.
        - A variety of pharmacologic agents are acceptable including ketamine, midazolam, fentanyl, and/or volatile agent with careful titration to achieve a balanced technique prior to muscle relaxation and tracheal intubation.
    - Utilizing 100% FiO_2 can significantly lower PVR, compromising systemic perfusion. Pre-oxygenation can be considered, but given potential detrimental effects, the primary objective should be to minimize apnea time with the most skilled person performing the intubation. Video laryngoscopy should be considered to optimize first-attempt success[5]. FiO_2 should be reduced as soon as feasible following the establishment of airway protection with the goal to maintain a SpO2 of 70-80%. Mechanical ventilation should be adjusted to avoid hypocarbia.
- Positioning
    - Supine, shoulder roll
- Maintenance
    - Volatile (with caution, as the neonatal population may be sensitive to the hemodynamic effects of volatile anesthetics).
    - Muscle relaxant
- Hemodynamic/Physiologic goals
    - Pre-Cardiopulmonary Bypass
        - As common with neonatal mixing lesions, management focuses on balancing pulmonary and systemic flow (Qp:Qs) and maintaining coronary perfusion. Signs of inadequate systemic perfusion may include lactic acidosis, delayed capillary refill, ischemic changes on EKG, or higher than expected oxygen saturation.

        - For patients with low PVR, aim is to avoid further decreases in PVR. Ventilator settings should be adjusted to avoid hypocapnia and hyperoxia.
        - Increases in SVR may further exacerbate pulmonary overcirculation, but an excessive drop in SVR may result in hypoxemia.
        - If hypotension and ST changes are present before a pulmonary vessel loop is feasible, use inotropic support rather than fluid to avoid further volume overload and maintain diastolic pressure
        - Avoid extreme tachycardia, as this increases myocardial oxygen demand
        - For patients with pulmonary arterial hypertension (PAH), ventilatory maneuvers to reduce PVR are necessary. This scenario is less common given that most surgical interventions occur in the early neonatal period.
    - Post-Cardiopulmonary Bypass
        - Inotropic and vasopressor support should be readily available (such as epinephrine 0.02-0.04 mcg/kg/min, milrinone 0.25-0.5mcg/kg/min, or dopamine 3-5mcg/kg/min)
            - Right ventricular systolic dysfunction may result from the ventriculotomy required for conduit placement, or from high PVR.
                - Inhaled nitric oxide may be beneficial in this scenario[1]
            - Left ventricular systolic dysfunction may result from volume overload if the truncal valve is incompetent or from myocardial ischemia.
        - Post-operative heart block is usually uncommon because a septal band separates the VSD from the conduction bundle, but is possible.
            - An epicardial pacemaker should be readily available.
        - Bleeding
            - The presence of a thymus is noted on sternal entry and will determine whether irradiated blood products will continue to be required.
            - Expect coagulopathy given the patient's age and size. There may be significant bleeding from aortic suture lines. Cryoprecipitate, platelets, and FFP should be readily available. Consider ROTEM or TEG to guide transfusion strategy. Also, consider recombinant hemostatic agents such as prothrombin complex concentrate (PCC), Factor VII (not to be given together), or fibrinogen concentrate.
        - Target normocapnia, ensure adequate sedation to minimize excessive elevations in PVR.
- Emergence/Disposition
    - These patients are often kept sedated in the early postoperative period due to the extent of the surgical procedure and are transferred to the ICU intubated.
        - Options for sedation include dexmedetomidine or midazolam and fentanyl or morphine for analgesia.
- Post-op Care
    - Analgesia, ventilator support, and inotropic support are weaned per the ICU and surgical team.
    - Following hospital discharge
        - Reoperation rates are high due to the need for re-intervention of the right ventricular outflow tract (RVOT). The RV to PA conduit does not grow with the child, and therefore is likely to necessitate further intervention. Occasionally, interventions for conduit stenosis can be performed in the catheterization lab, but many children eventually require repeat operations to address the RVOT or truncal valve pathology at some point during their lifetime.

- Studies have shown that despite a higher re-operation rate, adult survivors have a similar quality of life to age-matched controls, though physical function may be reduced.[4]

Case-Specific Complications/Pitfalls

- Maintaining Qp:Qs and optimizing ventilatory and hemodynamic strategies are key to maintaining systemic perfusion in the pre-CPB period. Failure to recognize an increase in the ratio will compromise end-organ perfusion.
  - Coronary ischemia is of significant concern, as described previously.
- Anesthesiologists should be ready to quickly address myocardial dysfunction and bleeding that results following separation from CPB.

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