SACROCOCCYGEAL TERATOMA

Surgical Specialty:	Pediatric General Surgery
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Background[1-4]:

- General Considerations
    - Sacrococcygeal teratoma (SCT) is a common extragonadal germ tumor in neonates and infants but a rare disease overall.
    - SCT originates from the tip of the sacrum and has both solid and cystic components
    - Staging of the SCT is based on the amount of external and internal pelvic component of the tumor.
    - Timing of surgical resection:
        - In-utero
        - Neonatal period
        - Post-neonatal period
    - The focus of this SPA Case Guide will be on the immediate neonatal repair.
- Patient Considerations
    - SCT is the most common extragonadal germ cell tumor in neonates/infants
    - It occurs in 1:27,000 – 40,000 births
    - Females are affected more than males
    - Antenatal diagnosis: routine prenatal ultrasound, magnetic resonance imaging (MRI)
    - High mortality during the neonatal period
    - Survival rate (assuming appropriate prenatal care/management):
        - 85% of postnatally diagnosed SCTs
        - 50% of prenatally diagnosed SCTs
    - Staging of the SCT is based on the amount of external and internal pelvic component of the tumor (Altman's classification). There are 4 types (Figure 1):
        - Type I: most of the tumor is outside the pelvis
        - Type II: tumor inside and outside the pelvic cavity, but it is larger outside the pelvis
        - Type III: tumor inside and outside the pelvic cavity, but larger inside pelvis/abdomen
        - Type IV: no tumor outside the pelvis, all the components are inside the pelvic cavity and abdomen

- Predictors of poor prognosis
  - Solid tumor morphology, significant spinal canal invasion, fetal hydrops, placentomegaly, highly vascular tumor, rapid growth of SCT, high output cardiac failure, and tumor volume to fetal weight ratio (TFR) greater than 0.12
  - TFR is calculated by dividing the total tumor volume by the estimated fetal weight
  - Total tumor volume is calculated using the greatest length, width, and height measurements of the tumor by US or MRI
  - TFR >0.12 is associated with an 80% incidence of hydrops and a 60% mortality rate while; TFR < 0.12 is associated with 100% survival.[4]
- Common complications:
  - Prenatally diagnosed: polyhydramnios, fetal cardiac failure, fetal hydrops, placentomegaly, tumor hemorrhage, and prematurity
  - Tumor recurrence, malignant transformation, sexual dysfunction, bladder and bowel dysfunction, and lower leg palsy
- Associated Comorbidities/Syndromes
  - Approximately 15-30% have associated congenital defects including nervous, cardiac, gastrointestinal, genitourinary, and musculoskeletal.

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - Review fetal and neonatal imaging studies (ultrasound, MRI, echocardiography)
  - Additional labs/tests indicated during work-up
    - Complete blood cell count, type & cross, serum chemistry, coagulation panel
  - Discussions to have with surgeon/family
    - Code Status
    - ECMO Candidacy
- Specific or Unique Room Set-up Requirements
  - Airway: General endotracheal intubation; consider using neonatal intensive care unit (NICU) ventilator
  - Drugs/Infusions
    - Fentanyl, non-depolarizing muscle relaxant
    - Resuscitation drugs: Atropine, epinephrine, calcium
    - Dextrose administration to maintain glucose homeostasis
  - Monitors: Standard American Society of Anesthesiologists monitors and invasive arterial blood pressure monitoring
  - Access: need reliable venous access (peripheral intravenous or umbilical venous catheter), arterial access (umbilical arterial catheter or radial arterial line)
  - Blood Availability:
    - Type & Cross
      - Packed red blood cells
        - If surgery is occurring immediately after birth, use irradiated, leukocyte-reduced, O-negative blood
      - Fresh-frozen plasma and platelets may be required in the setting of massive blood loss
  - Blood products should be readily available, as well as other pharmacologic methods, such as recombinant Factor VIIa, to attenuate bleeding while the surgical team does its part to correct the anatomic causes of bleeding. It has been reported that transfusion volumes are greater in patients with non-cystic tumors.[5]

- PICU Bed Availability (if indicated): NICU disposition

Intraoperative Considerations:

- General
  - General endotracheal anesthesia
  - Positioning for laryngoscopy may be challenging. Consider lateral position or supine with appropriate padding with blankets/foam to protect the teratoma
  - Video laryngoscopy may be helpful when securing the airway
- Induction: Opioid-based for better hemodynamic stability and muscle relaxation
- Positioning for surgery: Lateral or prone position
- Maintenance: Intravenous and inhaled anesthesia as tolerated
- Hemodynamic/Physiologic goals
  - Pulmonary status: Risk of respiratory distress syndrome
    - Caused by surfactant deficiency. Surfactant is produced by type II pneumocytes
    - Relatively stiff lungs with high peak inspiratory pressures
    - Increased oxygen requirement
    - Providing artificial surfactant at delivery helps to decrease the surface tension in alveoli
  - Massive Transfusion[6]
    - There is no consensus on the definition of massive transfusion in children.
    - The table below may be used for guidance; however, ongoing losses with transfusion of more than 10% of the total blood volume per hour may also be considered as a massive transfusion intra-operatively.

Blood volumes and massive transfusion definitions by age group.

Age group	Estimated blood volume	Massive transfusion
Premature neonate	100 mL/kg	50 mL/kg over 12 h 100 mL/kg over 24 h
Term neonate	90 mL/kg	45 mL/kg over 12 h 90 mL/kg over 24 h
Infants	80 mL/kg	40 mL/kg over 12 h 80 mL/kg over 24 h
Older children	70–80 mL/kg	35–40 mL/kg over 12 h 70–80 mL/kg over 24 h

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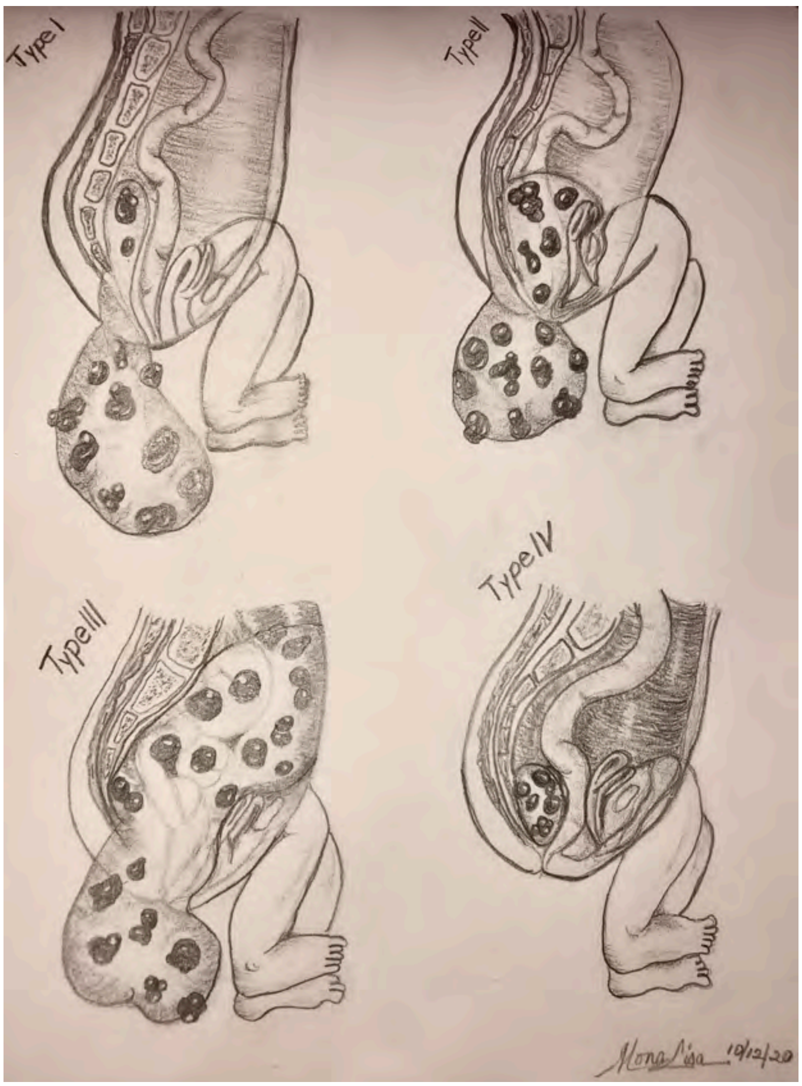
  - Hypothermia
- Surgical Considerations (such as neuromonitoring, muscle relaxation, anticipated blood loss)
  - Massive blood loss
  - Venous air embolism
    - Rare reports during giant tumor resection
  - Possible cardiac arrest from pulseless electrical activity (PEA)
    - Reversible causes of PEA arrest are hypovolemia, hypoxia, hypoglycemia, hypokalemia, hyperkalemia, hypothermia, tension pneumothorax, cardiac tamponade, toxins, coronary thrombosis, pulmonary thrombosis
    - Prolonged surgery duration
- Emergence/Disposition

    - Remain intubated and transfer to the NICU
- Postoperative care
    - Wean to extubate in the NICU
    - Patients often remain hospitalized for many months

Complications/Pitfalls

- High-output cardiac failure
- Massive transfusion
- Disseminated intravascular coagulation

Figure 1: Altman Classification



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Created: August 2022; Last revised: Date 4/22/2024