Sacrococcygeal Teratomas



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Sacrococcygeal Teratomas

- Tumor derived from germ cells (3 layers) that are foreign to the anatomic site in which they arise
- Typically midline or paraxial
- May be solid, cystic or mixed
- Mature vs Immature vs Malignant
 - neuroepithilium
- Most common neonatal tumor
 - **1:35,000 1:40,000**
 - Females more commonly affected

Types

- Yolk Sac Tumors (67%)
 - Most commonly Sacrococcygeal > Ovary
 - Other: Mediastinum, Retroperitoneum, Vagina, Testicle, Intracranial
- Embryonal
- Choriocarcinoma
- Mixed (10% worse prognosis)

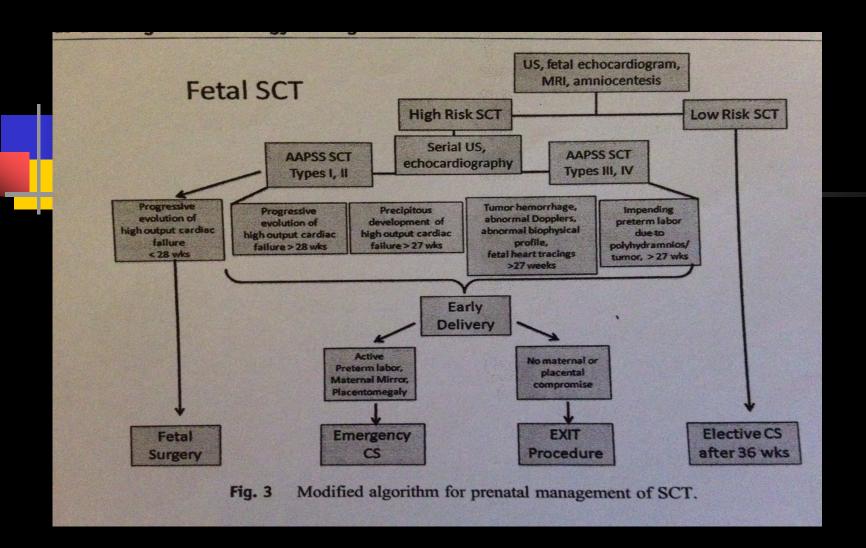
Clinical Presentation

- Present as a protruding mass arising from the coccyx
- Vascular Steal
- High Output Cardiac Failure (Hydrops)
- Tumor rupture/hemorrhage (> 5cm)
- Maternal Mirror Syndrome
- Lower Extremity Weakness
- Bladder, Rectal Obstruction

Early delivery as an Alternative Management Strategy for Selected High Risk Fetal Sacrococcygeal Teratomas

Risk Serial US Delivery

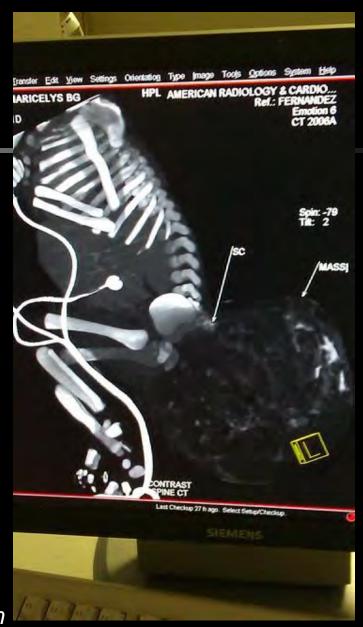




- 28-32 weeks maternal/fetal decompensation → Labor
- <27 weeks Type III/IV, maternal health overrides</p>
- Ex Utero Intrapartum Treatment

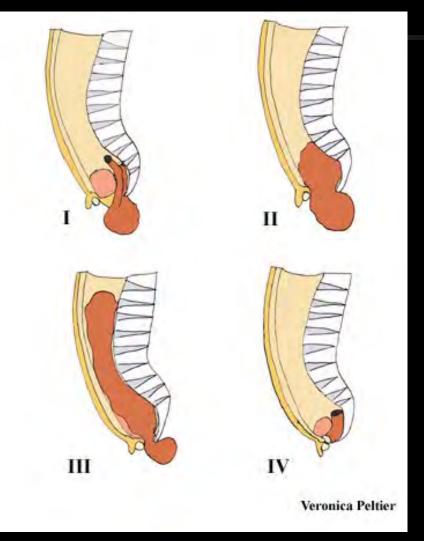


- Type I tumors (47%, most common)
 - Completely external
 - Identifiable prenatally
 - Least morbid
 - Usually Benign





- Type II (35%)
 - IntrapelvicComponent
- Type III (8%)
 - Intrabdominal Component

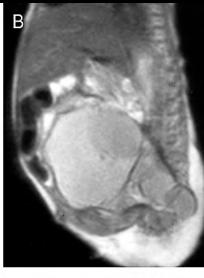


Classification

- Type IV (9%)
 - Completely Internal
 - Recognized Late
 - Malignant Transformation
 - Poor prognosis









Tumor Markers

- Alpha Feto Protein, B-HCG
- Useful in monitoring treatment response and tumor recurrence

- Our Patient's: B-HCG <2, AFP 12,472 (7/11/2011)
 - 1360 Aug 2011, 372 Oct 2011
 - AFP return to adult levels by 8 mo

Treatment

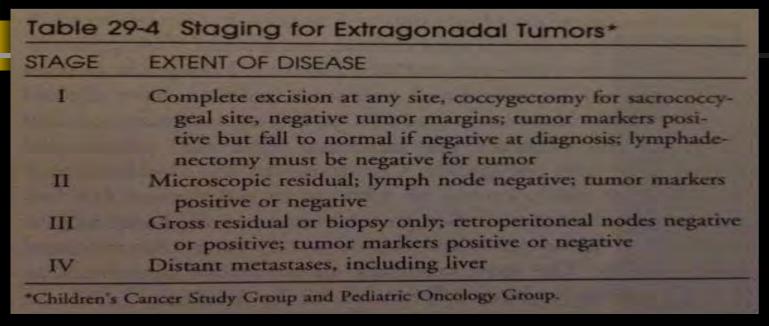
- Posterior anorectal approach vs Combined abdominal approach
- Blood Supply: Midsacral Artery and Hypogastric Artery Branches
- Coccygectomy: recurrence >35%







Staging



- Platinum based Chemotherapy
 - Cisplatin, Carblopatin + Bleomycin, Etoposide
- Neoadjuvant if advanced
 - 83% 5 yrs neoadjuvant + Surgery vs 49% surgery alone
- Radiation for presence of metastasis
 - www.uptodate.com, Pediatric Surgery, 5th Ed, O'Neill Jr, et al.

Prognosis

- Benign: disease free survival > 90%
- Time is of the essence:
 - <2 months, 7-10% malignant</p>
 - > 1 year, 75% malignant
- Follow up: 3-6months for 3 yrs
 - Most recurrences occur within 1st 3 yrs.

Ashcraft's Pediatric Surgery 5th ed. Holdcomb et al.

Characteristics

- Malignancy characteristics increases with:
 - Age at diagnosis
 - Surgical type (type IV is worst)
 - Male
 - Presence of necrosis and hemorrhage
 - Degree of immaturity doesn't correlate with malignancy except in ovary

References

- Pediatric Surgery, 5th Ed, O'Neill Jr, et al.
- www.uptodate.com
- www.emedicine.com
- Early delivery as an Alternative Management Strategy for Selected High Risk Fetal Sacrococcygeal Teratomas. Flake, et al. <u>Journal of Pediatric</u> <u>Surgery</u>, Aug 2010.
- Ashcraft's Pediatric Surgery 5th ed. Holdcomb et al.