#### 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

### **Fypes**

#### Yolk Sac Tumors (67%)

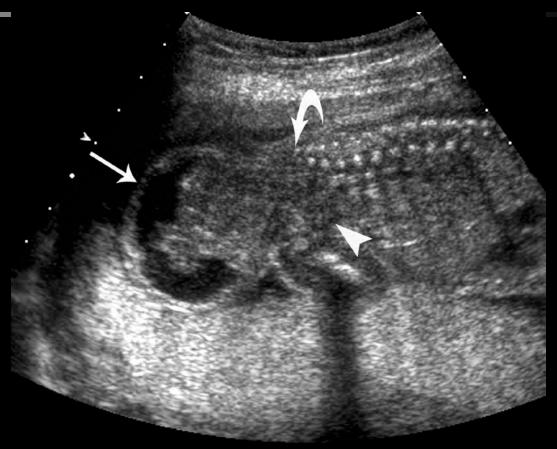
- Most commonly Sacrococcygeal > Ovary
  - Other: Mediastinum, Retroperitoneum, Vagina, Testicle, Intracranial
- Embryonal
- Choriocarcinoma
- Mixed (10% worse prognosis)

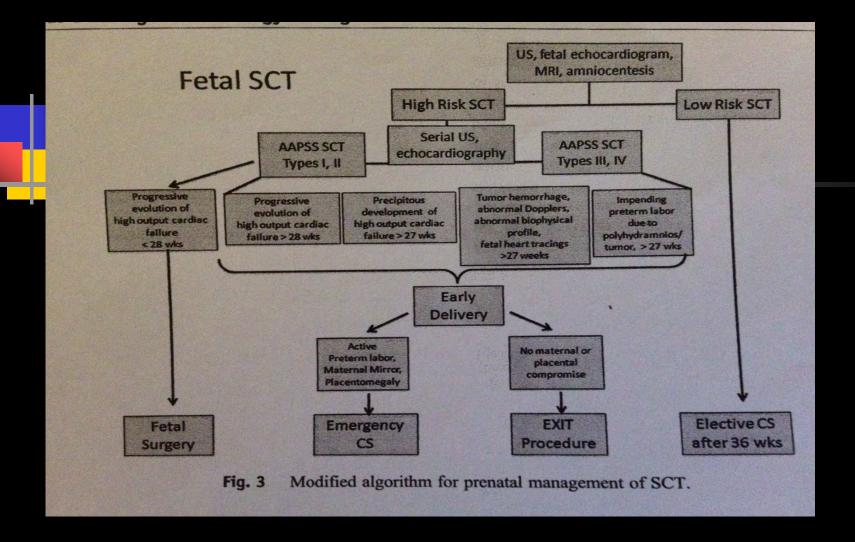
#### **Clinical Presentation**

- 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  $\rightarrow$  Labor
- <27 weeks Type III/IV, maternal health overrides</p>
- Ex Utero Intrapartum Treatment

## Classification

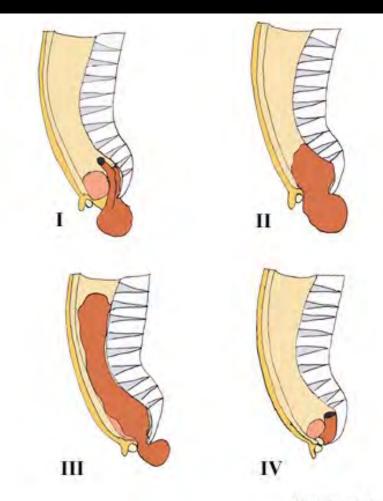
- Type I tumors (47%, most common)
  - Completely external
  - Identifiable prenatally
  - Least morbid
  - Usually Benign



American Academy of Pediatrics Surgical Section

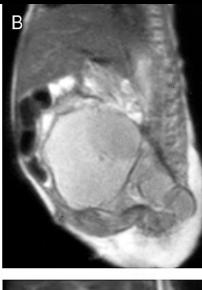
### Classification

- Type II (35%)
   Intrapelvic Component
- 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%

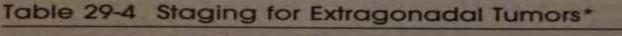






# Staging

II



#### STAGE EXTENT OF DISEASE

	Complete excision at any site, coccygectomy for sacrococcy-
	geal site, negative tumor margins; tumor markers posi-
	tive but fall to normal if negative at diagnosis; lymphade-
	nectomy must be negative for tumor

- Microscopic residual; lymph node negative; tumor markers positive or negative
- III Gross residual or biopsy only; retroperitoneal nodes negative or positive; tumor markers positive or negative
   IV Distant metastases, including liver

"Children's Cancer Study Group and Pediatric Oncology Group.

#### 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</li>
> 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
- <u>www.emedicine.com</u>
- 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.