American Pediatric Surgical Association

Prenatal Counseling Series

Sacrococcygeal Tumors

APSA
American Pediatric Surgical Association
Saving Lifetimes

from the
Fetal Diagnosis and Treatment Committee

of the
American Pediatric Surgical Association

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Sacrococcygeal Tumors

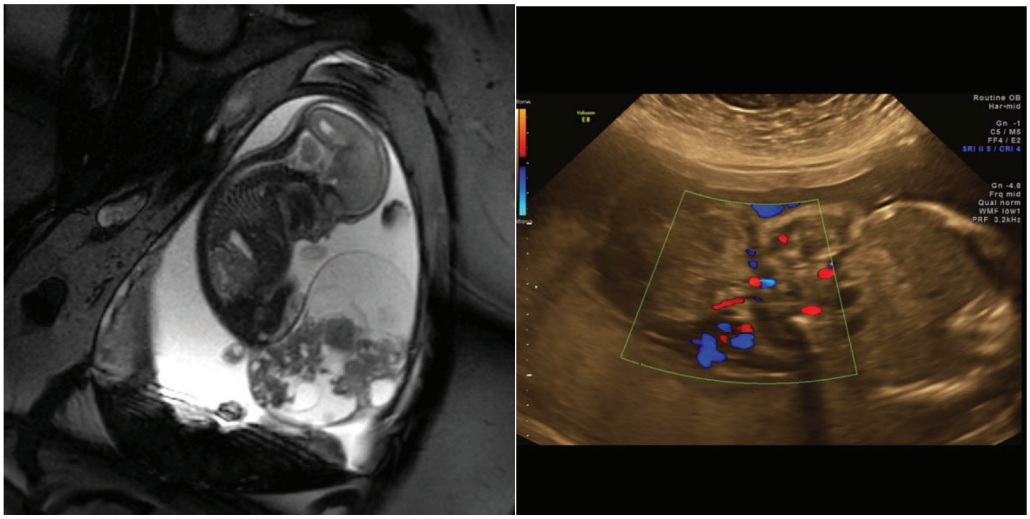
- Sacrococcygeal tumors (SCT) are one of the most common congenital neoplasms of the newborn period with a prevalence of 1:27-40,000 live births.
- They arise from a totipotent stem cell in the coccyx (Henson's node) and are generally benign in fetal and early neonatal life.
- Incidence is 4 times more common in females.
- Sacrococcygeal tumors are classified into four categories :

AAPSS Staging Classification of Sacrococcygeal Teratoma

Type	Description
Type I	Completely external, no presacral component
Type II	External component and internal pelvic component
Type III	External component and internal component extending into abdomen
Type IV	Completely internal and no external component

Source: Fetology: *Diagnosis and Management of the Fetal Patient*
By Diana W. Bianchi, Timothy M. Crombleholme, and Mary E. D'Alton

- Complications related to prenatally diagnosed SCTs may include polyhydramnios, fetal cardiac failure, fetal hydrops, placentomegaly, maternal mirror syndrome, tumor hemorrhage and prematurity.
- Prenatally diagnosed SCTs have 3 times the mortality rate compared to postnatally diagnosed neonates with a mortality rate ranging from 15-35%.
- Approximately 15-30% have associated congenital defects including nervous, cardiac, gastrointestinal, genitourinary and musculoskeletal.
- Local abnormalities such as rectovaginal fistula, urethro-vaginal fistula, urethral atresia and imperforate anus are directly related to tumor growth.



Sagittal MRI images of a fetus with a large pre sacral mass composed of mixed cystic and solid components. The majority of the mass is exophytic with a small component located within the pelvis (type 1). Color Doppler ultrasound image shows internal blood flow within the solid components of the mass. Images courtesy of Jill Stein, MD - Colorado Fetal Care Center

Initial Evaluation

- Referral to a fetal center should be considered for a multidisciplinary consultation with surgery, neonatology, genetics and maternal fetal medicine
- Detailed obstetrical ultrasound with amniotic fluid index
- Consider fetal MRI
- Fetal echocardiogram
- Aneuploidy has not been reported with SCT. Amniocentesis is not recommended for karyotype analysis unless there are multiple anomalies, advanced maternal age or if fetal surgery is indicated
- Close follow up and monitoring for the development of polyhydramnios and/or high output cardiac failure

Prenatal Diagnosis

Typically diagnosed by finding a mass on routine ultrasound.

Ultrasound and Color Doppler

Sacrococcygeal tumors are sonographically seen as large, heterogenous masses due to their different tissue components. Important features to consider include size, location, solid and cystic areas and septation, vascular supply and calcifications.

- Detailed anatomy
- Amniotic fluid index
- Cardiac/thoracic ratio
- Increased descending aortic blood flow (>120cm/s)
- Increased diameter of inferior vena cava (>1cm)
- Umbilical artery Doppler systolic to diastolic (S/D) ratio.
- Placental thickness
- Important markers of hydrops fetalis include the presence of ascites, pleural or pericardial effusions and skin or scalp edema
- Tumor Volume to Fetal Weight Ratio

Differential Diagnosis of an SCT

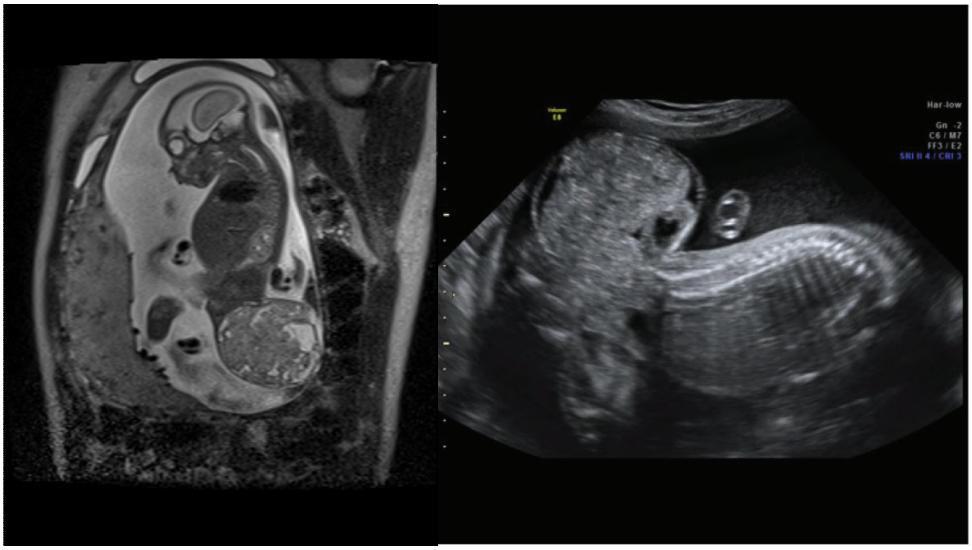
- Lumbosacral myelomeningocele
- Dermoid
- Lipoma
- Neuroblastoma
- Other malformations of the sacrococcygeal region

Fetal Echocardiography

- Detailed cardiac anatomy
- Combined cardiac output measurement (normal combined cardiac output is 550 ml/min/kg)
- Cardiac/thoracic ratio
- Descending aortic blood flow
- Inferior vena cava diameter
- Umbilical artery Doppler systolic to diastolic (S/D) ratio.

Fetal Magnetic Resonance Imaging

- MRI can further determine size of mass, anatomic relations, impact on other pelvic structures, hemorrhagic changes and intrapelvic or intraspinal extent of mass.
- MRI may also be helpful in operative fetal surgery planning.
- Useful in cystic SCT cases to differentiate them from myelomeningocele



Sagittal MR and ultrasound images show a large mixed cystic and solid presacral mass that is primarily external (type 1). Dark foci scattered within the mass on the MRI image suggests calcification and/or hemorrhage. Images courtesy of Jill Stein, MD - Colorado Fetal Care Center

Tumor Volume to Fetal Weight Ratio

- Tumor volume to fetal weight ratio (TFR)= total tumor volume / estimated fetal weight
- Is an important prognostic indicator for SCT and is calculated using greatest length, width and height measured of the tumor by US or MRI and fetal weight calculated by US using Hadlock formula

Predictors of Poor Prognosis

- Solid tumor morphology
- Significant spinal canal invasion as risk for paraplegia
- Presence of fetal hydrops – placentomegaly – maternal mirror syndrome
- Tumor with high vascularity
- A phase of unpredictable rapid growth of SCT
- High output cardiac failure
- TFR >0.12 associated with 80% incidence of hydrops and 60% mortality rate while TFR <0.12 associated with 100% survival

Prenatal Counseling

- Standard prenatal care should be continued with frequent serial ultrasound scans and echocardiographic surveillance.
- Depending on complete clinical picture, families are counseled on the options of:
  1. termination of pregnancy if GA is <24 weeks or per local state regulation
  2. continued standard prenatal care with US scans and echocardiographic surveillance
  3. possible fetal intervention if fetus and mother fit criteria
- Mode of delivery: determined by the size of the tumor
  1. Small tumors can be delivered vaginally.
  2. Cesarean delivery is recommended in larger tumors (>5cm) to avoid tumor-induced hemorrhage or dystocia.

Fetal Intervention

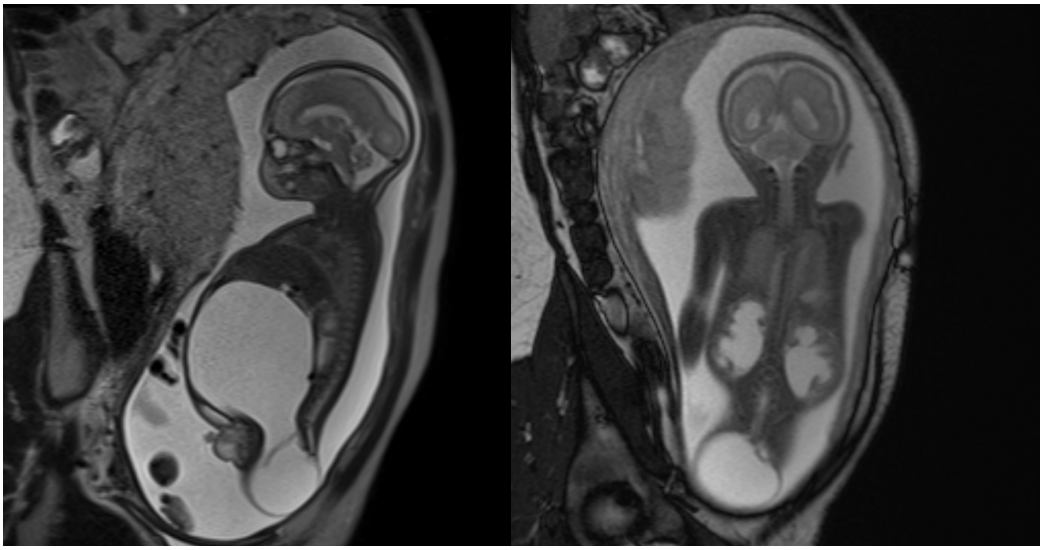
Fetal intervention for SCTs should be performed at highly specialized fetal centers to guarantee best maternal and fetal outcomes. Fetal interventions have been reserved for those fetuses <26 weeks gestational age. For fetuses presenting >27 weeks, the morbidity of fetal intervention must be weighed against the risk of early delivery and postnatal resection.

The goal of fetal surgery for SCT is to reduce vascular supply to the tumor through vascular occlusion, to remove the low-resistance tumor vascular bed from fetal circulation to reduce morbidity and mortality related to the SCT and to allow for continued fetal development prior to delivery.

Fetal interventions for SCT have included cyst decompression, open fetal surgery for debulking of SCT, EXIT-to-resection and vascular flow interruption via fetoscopic laser ablation, radiofrequency ablation or interstitial ablation +/- vascular coiling.

- Trans-abdominal and transvaginal aspirations of large cysts may be considered to facilitate delivery.

Cyst decompression may assist with maternal discomfort, and cyst-amniotic shunts may relieve bladder outlet obstruction.



Sagittal and coronal MR images demonstrate a large presacral mass that is primarily cystic with internal septation. The internal portion of the mass is larger than the external portion (type 3). There is resultant mass effect with obstruction of the renal collecting systems as demonstrated by dilated renal calyces, pelves and ureters. The urinary bladder is compressed. Images courtesy of Jill Stein, MD - Colorado Fetal Care Center

- Survival following SCT interventions have ranged from 38-75%. However, survival in hydropic SCT patients not undergoing fetal intervention is likely < 10%.

- **Open fetal surgery for debulking of SCT:** High-risk SCT with evidence of impending high-output cardiac failure – absence of maternal risk factors for anesthesia and surgery – and singleton pregnancy with normal karyotype analysis
  1. Gestational age ideally less than 26 weeks, and favorable anatomy (classification type I or II).
- **EXIT-to-Resection:** is an approach utilized to manage high risk SCTs that are at risk of rupture with exsanguination presenting after 32 weeks' gestation.
- **Post Fetal Surgery Considerations:** Continued surveillance with US and echo**.** Betamethasone in anticipation of preterm delivery. Delivery by Cesarean section is done for impending preterm labor.

Postnatal Considerations

- Postoperative surveillance: physical examination including digital rectal exam and AFP levels every 3 months until at least 3 years of age with imaging if indicated
- Type III and IV tumors have a higher risk of urinary and fecal incontinence and should be followed in specialized clinics

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