

Sacrococcygeal Teratoma

Brock W. Hanisch
Creighton University,
Department of Clinical Anatomy



Sacrococcygeal Teratoma (SCT)

- The most common tumor found in newborns.
- "Sacro" refers to sacrum and "coccy" refers to coccyx.
- Teratoma refers to the type of tissues that make up these growths. They are comprised of chaotically arranged tissues of all types (fat, bone, nerves, muscle, etc...) that are found in an area that they are not normally found.
- Sacrococcygeal teratoma (SCT) is rare, occurring in 1 in 35,000 to 40,000 live births.
- It is four times more common in females than males. The cause is not known. One theory is it is a failed twinning attempt. Another is it is a growth from an abnormally placed set of germ or stem cells.

Sacrococcygeal Teratoma (SCT)

- **Those diagnosed in utero carry 50% risk of premature delivery.**
- **Sacrococcygeal teratomas can be quite large. Many are approximately the size of the unborn baby. Tumors greater than 10cm in diameter require cesarean.**
- **Some of the SCTs are cyst-type tumors, meaning they are filled with fluid. Others are solid tumors that may have a significant amount of blood flow through them. The most common type of SCT is a combination of both solid and cystic.**
- **Tumor develops from pluripotential embryonic cells (remnants of primitive streak).**

Origin of SCT

Embryology:

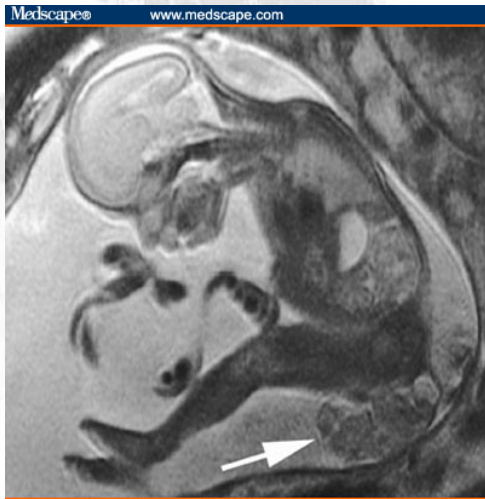
- Original germ cells migrating from the yolk sac to the gonad pathway are thought to persist, deviate, differentiate, and mature, typically resting anterior to the future coccyx at Hensen's node.
- Growth of these primitive pluripotential cells escapes the control of embryonic inductors and organizers, resulting in a teratoma.
- Rearrangement within the proto-oncogene or in a regulatory sequence result in a molecular transformation of cells foreign to that anatomical site.

Origin of SCT

- Often occurs near the coccyx, where the greatest concentration of primitive cells exists for the longest period of time.
- SCT is formed from multiple neoplastic tissues that:
 1. lack organ specificity
 2. Foreign to the sacrococcygeal region
 3. Are derived from all three germ layers

Diagnosis of SCT

- Neonate with large soft buttock mass; identified as a cystic SCT
- Sacrum and coccyx are intact but small tongue of tissue attached to coccyx



Diagnosis of SCT

- Usually made by ultrasound (US) as a screening procedure or to assess uterine size.
- Characteristic findings are a caudal or intrapelvic mass.
- Full evaluation is best performed by MRI. If there is invasion of soft tissues or bone it implies malignancy, but majority of teratomas do not show this and proper diagnosis must be made histologically.
- Following diagnosis, Echocardiographic and Doppler US measurements are essential to gauge the severity of the teratoma.
- Once a diagnosis of SCT is made US and EKG are made weekly to ensure proper fetal heart development.

SCT Classification

Sacrococcygeal teratomas have been classified into four groups depending upon the amount of the tumor outside the body:

Type I is completely external, evident at birth, and more easily resected or surgically removed. Type I does not typically spread.

Type II has external and internal components. The internal portion is confined to the pelvic region. This type will spread in about 6% of cases.

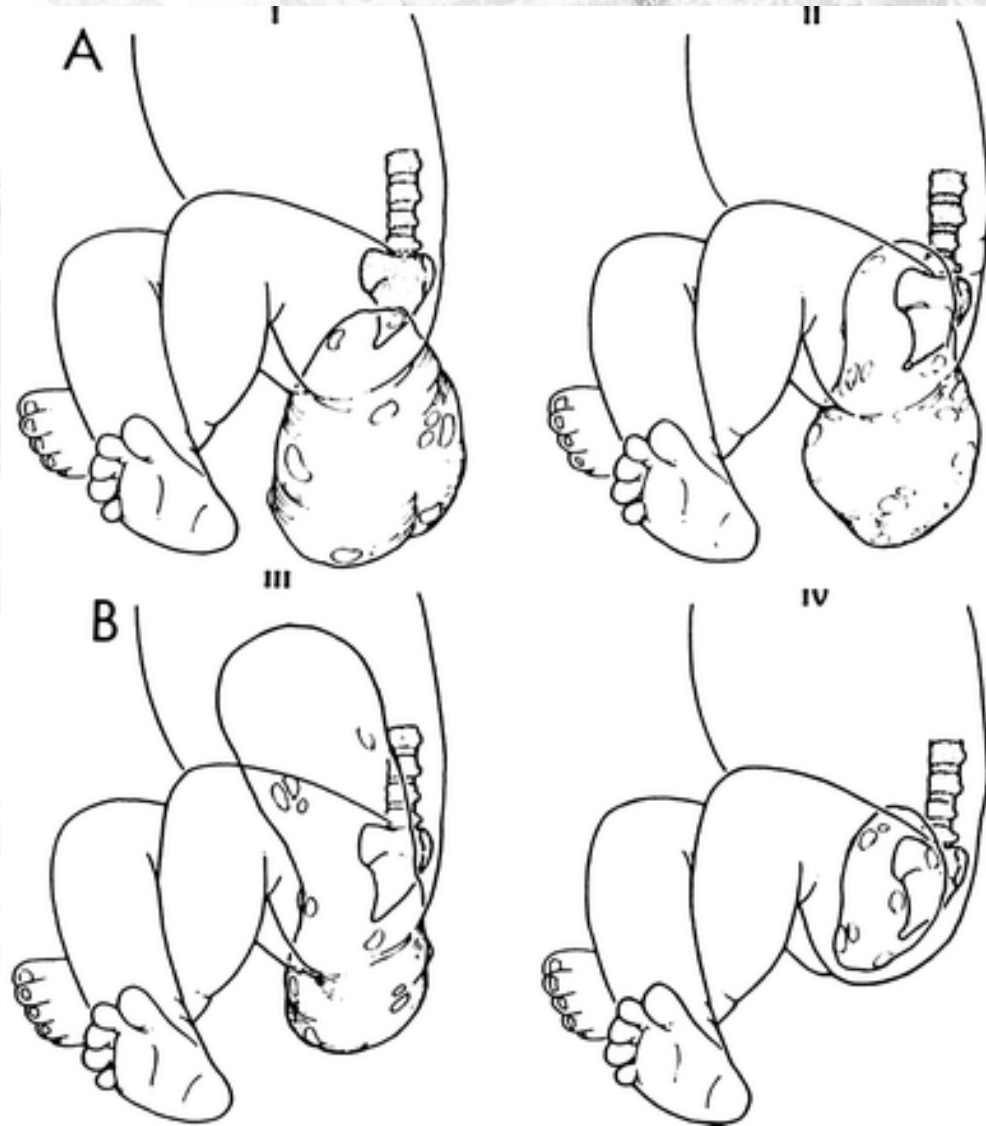
Type III also has external and internal components but the internal portion extends into the abdominal area. These types, II and III, are also evident at birth but the resection may be more difficult requiring access both from the back of the baby and from the front. Type III will spread in about 20% of cases.

Type IV is completely internal. For this reason it may go undiagnosed for some time. Later, symptoms may develop that warrant an investigation and at this time the diagnosis is made. This type will spread in about 8 % of cases.

SCT Classification

Altman classification system:

- Type I (A, left) and type II (A, right) teratomas have predominant extrapelvic components.
- Type III (B, left) and type IV (B, right) teratomas have a primarily intrapelvic location.



SCT Complications

**•Most serious problem with prenatal SCT is high output cardiac failure resulting in fetal hydrops (build-up of serous fluid in fetal tissues) and respiratory insufficiency
Hydramnios (excess amniotic fluid) and Placentomegaly (enlargement and thickening of the placenta) may also occur.**

**•These conditions may result in premature labor.
Without treatment, hydrops is universally fatal for fetus (Exception: fetuses who develop hydrops near term)
Thus, fetal surgery is the only real hope for pre-viable fetuses with SCT accompanied by hydrops**

SCT Complications

- **Approximately 11-38% of fetuses with an SCT will have other anomalies or birth defects. These associated defects increase the mortality rate as well.**
- **Malignancy is not a primary cause of death for these infants. Other complications for SCT include hemorrhage within the tumor, development of hydrops and risk of preterm labor due to the size of the tumor and/or polyhydramnios (high volume of amniotic fluid).**
- **The babies with large solid tumors are more prone to develop hydrops. This is, however, a small percentage of babies with SCT (less than 20%).**

SCT Complications

- Hydrops is an abnormal accumulation of fluid in two or more areas of the body. Some of the more common areas can include fluid in the abdomen (ascites), fluid around the lungs (pleural effusion), fluid around the heart (pericardial effusion), or extra fluid under the skin (anasarca) or scalp.
- If hydrops develops after 30 weeks' gestation, the mortality rate is approximately 25%. If, however, the hydrops develops before 30 weeks' gestation, the mortality rate is more than 90%. These babies may become candidates for fetal intervention if hydrops does develop early. This means some form of treatment may be available for the baby before it is born.

SCT Treatment and Recovery

- When tumors are excised properly, prognosis is usually good with survival rates greater than 95%.
- Most tumors are benign, and only 11% recur after resection.
- Incidence of malignancy increases from 10% at birth to 50-70% at two months.
- Even with recurrence, modern chemotherapy treatment carries a 98.4% survival rate.



SCT Treatment and Recovery

- **Physician may in some circumstances remove growth prior to birth.**
- **High risk! Some physicians have attempted to use laser or radiofrequency ablation (RFA) or tumor embolization rather than risk open fetal surgery. However, this too has significant risks due to close proximity of tumor to anorectal complex, vagina, urethra, sciatic nerves and hip joints.**

Case Study

- **A 29-year-old woman who had previously undergone four normal deliveries was admitted to the emergency room in labor.**
- **The infant's head, upper limbs, and trunk were delivered normally, whereupon delivery was halted due to obstruction by the infant's buttocks and lower limbs.**
- **Delivery of the newborn was accomplished by means of cesarean.**

Case Study (Continued)

- **A 3.6kg (7lb14.98oz) female infant was delivered, appearing active and well upon initial examination.**
- **Protruding from the coccygeal region, slightly displaced to the left of the midline, was a skin-covered mass devoid of hair and 20 cm in diameter.**
- **X-ray studies of the pelvis revealed a soft tissue mass with no calcification of the coccygeal vertebra.**
- **Pelvic extension of the mass could not be determined and surgery was performed at age 48hr.**

Case Study (Continued)

- **Provisional diagnosis: benign sacrococcygeal teratoma**
- **Treatment: the teratoma was excised through an inverted V incision over the lower sacrum and buttocks. Prior to the removal of the tumor, the middle sacral artery and vein as well as the lateral sacral vessels supplying it were ligated. Upon removal, the tumor was separated from the rectum while the coccyx was excised with the tumor.**

Case Study (Continued)

- **Provisional diagnosis was confirmed by histological analysis: Histological sections of the tumor revealed cysts lined by glandular epithelium; fat, muscle, and connective tissue; acini resembling the pancreas; cartilaginous material, sebaceous glands; hair follicles and sweat glands.**
- **The infant was discharged at eight days of age, and at 15 months of age remained a healthy and well developed child with no recurrence of tumor.**

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Suggestions?
Comments?

