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# Sacrococcygeal Teratoma (SCT)

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***Abstract:*** *Sacrococcygeal teratoma (SCT) is the most common congenital germ cell tumor, with an incidence of 1 in 35,000-40,000 live births, This birth defect is more common in female than in male babies.female predominance (3:1-4:1 ratio). The tumor arises from embryologically multipotent cells from the Hensen node, which is located in the coccyx. Most SCTs are now diagnosed prenatally, because of the widespread use of routine obstetric ultrasonography. Which is the most common germ cell neoplasia in the newborn, that develops before birth and which are seen on the frontal surface of coccyx (tailbone) and sacrum, and is thought derive from the primitive streak. SCT is usually discovered either because a blood test performed on the mother at 16 weeks shows a high alpha fetoprotein (AFP) amount, or because a sonogram is performed because the uterus is larger than it should be. The increased size of the uterus is often caused by extra amniotic fluid, called polyhydramnios.*

***Keywords:*** *Alpha fetoprotein (AFP),bowel obstruction, hydronephrosis, hydropsfetalis, hip dysplasia, urinary obstruction.*

## I. Introduction

Sacrococcygeal teratoma (SCT) is the most common congenital germ cell tumor, with an incidence of 1 in 35,000-40,000 live births, This birth defect is more common in female than in male babies. Female predominance (3:1-4:1 ratio). The tumor arises from embryologically multipotent cells from the Hensen node, which is located at the anterior aspect of coccyx, by about the 2nd to 3rd weeks of gestation. The tumour is composed of the all three germ cells (i.e. ectoderm, mesoderm and endoderm)

Most SCTs are now diagnosed prenatally, because of the widespread use of routine obstetric ultrasonography. Which is the most common germ cell neoplasia in the newborn that develops before birth and which are seen on the frontal surface of coccyx (tailbone) and sacrum, and is thought derive from the primitive streak.

As a result of ultrasonographic developments in recent years, sacrococcygeal teratoma can be easily diagnosed in the prenatal period.Sacrococcygeal teratomas are benign 75% of the time, malignant and life-threatening 12% of the time, and the remainder are considered "immature teratomas" that share benign and malignant features. Benign sacrococcygeal teratomas are more likely to develop in younger children who are less than 5 months old, and older children are more likely to develop malignant sacrococcygeal teratomas.

Although the tumors can grow very large, they are usually not malignant (that is, cancerous). They can usually be cured by surgery after birth, but occasionally cause trouble before birth. SCT is usually discovered either because a blood test performed on the mother at 16 weeks shows a high alpha fetoprotein (AFP) amount, or because a sonogram is performed because the uterus is larger than it should be. The increased size of the uterus is often caused by extra amniotic fluid, called polyhydramnios. The diagnosis of SCT can be made by an ultrasound examination.

## II. Materials & Methods

In the present study 530 Infants and fetuses obtained from Rajiv Gandhi Institute of medical sciences general Hospital, Srikakulam from department of Obstetrics and Gynaecology, and private hospitals during the year April 2013 to March 2015.

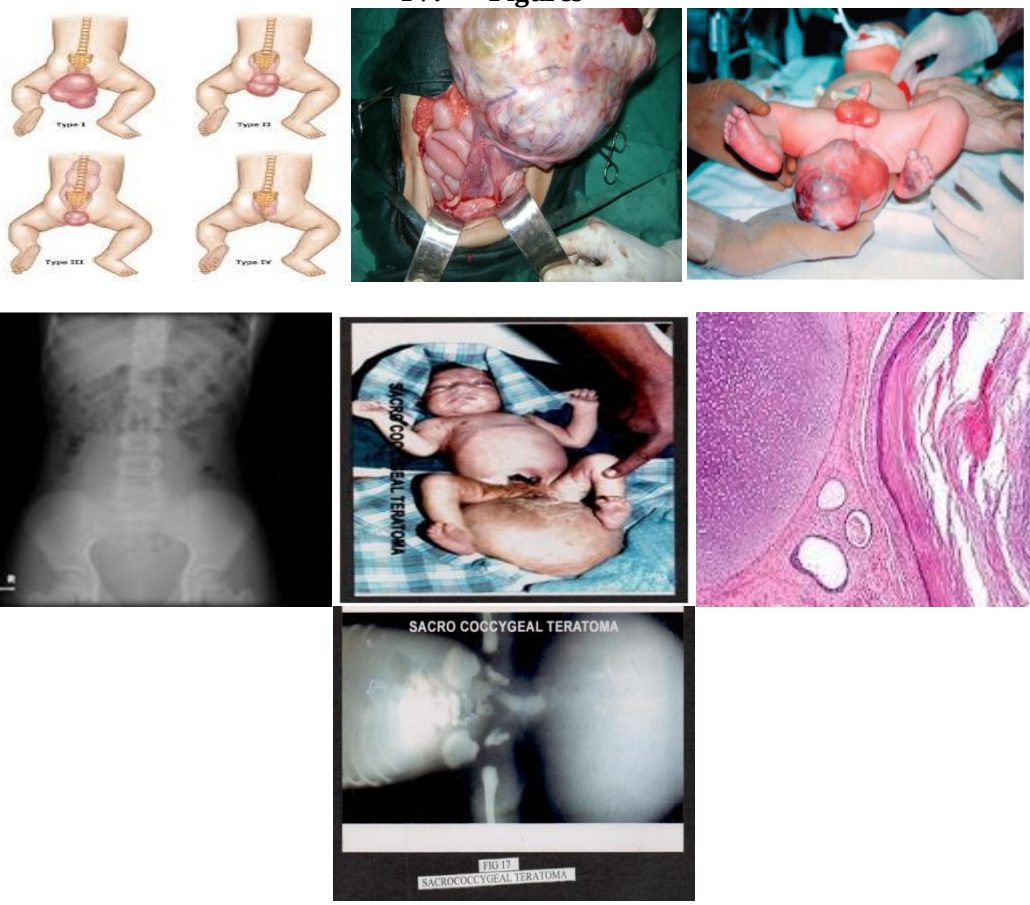
Weight of the foetus, Head circumference, crown - rump length, both right and left arm and forearm length, groin to tip of the first hallux and right and left foot lengths were taken with help of Thread, vernier calipers & Scale for the present study.

## III. Observations

Among the 530 new born 17 were having congenital abnormalities out of which, only one new born is with, Sacrococcygeal teratoma (SCT) and the measurements of foetus are as follows:

Weight of the foetus: 2500 grams
Head circumference: 40cms
Crown rump length: 42 cms
Arm length:
Right: 10cms
Left: 10cms
Forearm length:
Right: 9cm
Left: 9cm
Femur length: 12cms
Foot length
Right: 8.45cms
Left: 8.45cms.

## IV. Figures




## V. Discussion

The clinical significance of SCT results from its concurrence with other pathologies, thereby increasing prenatal and perinatal mortality and morbidity.. Mortality and morbidity rates are relatively high in patients suffering from SCT as a result of high output heart failure, polyhydramnios, hydrops, preterm delivery, anemia, and tumor rupture.. STC is diagnosed in the early period in the form of a cystic, solid or mixed mass extending from the sacral zone to the perineum or hip .Ultrasonography is valuable in monitoring the size of the tumor, early diagnosis of complications, and in the determination of the most favorable time and method of delivery as well as prenatal diagnosis.

**Classification of sacrococcygeal teratoma(sct)**

SCTs are categorized according to the classification developed by the American Academy of Pediatrics, Surgical Section (AAPSS), as follows:

- Type I - Primarily external or has a minimal presacral component
- Type II - Predominantly external but has a significant intrapelvic component
- Type III - Predominantly intrapelvic with abdominal extension, with a small external component
- Type IV - Entirely within the pelvis and abdomen

Whereas type I tumors, being primarily external to the fetus, are easily diagnosed prenatally and are amenable to fetal resection, type IV tumors can be difficult to diagnose and are not amenable to fetal resection. The AAPSS classification describes surgical anatomy and identifies tumors that are amenable to fetal resection, but it does not provide prognostic information, nor does it identify fetuses who would benefit from fetal intervention.

Historically, sacrococcygeal teratomas present in 2 clinical patterns related to the child's age, tumor location, and likelihood of tumor malignancy. With the advent of routine prenatal ultrasound examinations, a third clinical pattern is emerging.

- Fetal tumors present during prenatal ultrasound exams, with or without maternal symptoms. SCTs found during routine exams tend to be small and partly or entirely external. The internal SCTs are not easily seen via ultrasound, unless they are large enough to reveal their presence by the abnormal position of the fetal urinary bladder and other organs, but large fetal SCTs frequently produce maternal complications which necessitate non-routine, investigative ultrasounds.
- Neonatal tumors present at birth protruding from the sacral site and are usually mature or immature teratomas.
- Among **infants and young children**, the tumor presents as a palpable mass in the sacropelvic region compressing the bladder or rectum. These pelvic tumors have a greater likelihood of being malignant. An early survey found that the rate of tumor malignancy was 48% for girls and 67% for boys older than 2 months at the time of sacrococcygeal tumor diagnosis, compared with a malignant tumor incidence of 7% for girls and 10% for boys younger than 2 months at the time of diagnosis. The pelvic site of the primary tumor has been reported to be an adverse prognostic factor, most likely caused by a higher rate of incomplete resection

In older children and adults, the tumor may be mistaken for a pilonidal sinus, or it may be found during a rectal exam or other evaluation. The preferred first treatment for SCT is complete surgical removal (i.e., complete resection). The preferred approach to a small SCT is through the perineum; a large SCT may require an additional approach through the abdomen. Resection should include the coccyx and may also include portions of the sacrum. The surgery should include reattachment of the small muscles and ligaments formerly attached to the coccyx, in effect reconstructing the posterior perineum. If not, there is an increased risk of perineal hernia later in life.

SCTs are classified morphologically according to their relative extent outside and inside the body:

- Altman type I — entirely outside, sometimes attached to the body only by a narrow stalk
- Altman type II — mostly outside
- Altman type III — mostly inside
- Altman type IV — entirely inside; this is also known as a presacral teratoma or retrorectal teratoma

The Altman type is significant in the contexts of management of labor and delivery, surgical approach, and complications of SCT. Serial ultrasound and MRI monitoring of SCTs in fetuses in utero has demonstrated that the Altman type can change over time. As the tumor grows, it can push between other organs and through the perineum to the body surface where the tumor appears as a bulge covered only by skin. Sometimes, the tumor bulge later slips back inside the perineum.Like all teratomas, a sacrococcygeal teratoma has the potential to be malignant, and the standard of care requires long-term followup by an oncologist.

During prenatal ultrasound, an SCT having an external component may appear as a fluid-filled cyst or a solid mass sticking out from the fetus' body. Fetal SCTs that are entirely internal may be undetected if they are small; detection (or at least suspicion) is possible when the fetal bladder is seen in an abnormal position, due to the SCT pushing other organs out of place.

At birth, the usual presentation is a visible lump or mass under the skin at the top of the buttocks crease. If not visible, it can sometimes be felt; gently prodded, it feels somewhat like a hardboiled egg. A small SCT, if it is entirely inside the body, may not present for years, until it grows large enough to cause pain, constipation and other symptoms of a large mass inside the pelvis, or until it begins to extend out of the pelvis. Even a

relatively large SCT may be miss if it is internal, because the bony pelvis conceals and protects it. Mediastinal tumors, including teratomas, are similarly concealed and protected by the rib cage.

Maternal complications of pregnancy may include mirror syndrome Maternal complications of delivery may include a Cesarean section or, alternatively, a vaginal delivery with mechanical dystocia.

Complications of the mass effect of a large SCT may include hip dysplasia, bowel obstruction, urinary obstruction, hydronephrosis and hydrops fetalis. Even a small SCT can produce complications of mass effect, if it is presacral (Altman Type IV In the fetus, severe hydronephrosis may contribute to inadequate lung development. Also in the fetus and newborn, the anus may be imperforate.

Later complications of the mass effect and/or surgery may include neurogenic bladder, other forms of urinary incontinence, fecal incontinence, and other chronic problems resulting from accidental damage to or sacrifice of nerves and muscles within the pelviRemoval of the coccyx may include additional complications. In one review of 25 patients however, the most frequent complication was an unsatisfactory appearance of the surgical scar. The day an expectant parent receives a diagnosis of SCT (sacrococcygeal teratoma) for their unborn or newly born child is a dark and painful moment in time. Frequently, as in our case, the occasion of the diagnosis is a much anticipated regularly scheduled ultrasound test to determine, among other things, the gender of the baby in utero. In other cases, the mother or baby in utero is showing some sign of distress or abnormality and an ultrasound is ordered to identify the cause of the distress or abnormality. In still other cases, the diagnosis is not made until the baby is actually delivered. In any event, parents are ill prepared to learn that the baby they already love and cherish is suffering from a rare (1 in 40,000) fetal tumor called a sacrococcygeal teratoma.

Fetuses with larger tumors or tumors that go up inside the baby's abdomen will require more complex surgery after birth, but in general do well. Again, they will have to be followed by an oncology service with blood tests for several years. Fetuses with very large tumors, which can reach the size of the fetus itself, can pose a difficult problem both before and after birth.

We have found that those SCTs that are largely cystic (fluid-filled) generally do not cause a problem for the fetus before birth. However, when the SCT is made up of mostly solid tissue, and has a lot of blood flow in it, the fetus can suffer adverse effects. This is because the fetus's heart has to pump not only to circulate blood to its body, but also to all the blood vessels of the tumor, which can be as big as the fetus. In essence, the heart is performing twice its normal amount of work. The amount of work the heart is doing can be measured by fetal echocardiography. This sensitive test can determine how hard the heart is working when the fetus is approaching hydrops, or heart failure. If hydrops does develop, usually in rapidly growing solid tumors, the fetus usually will not survive without immediate intervention before birth.

Fetuses with large tumors and a great deal of blood flow to the tumor have to be followed closely for the development of hydrops, or fetal heart failure which can lead to fetal death. If hydrops does not develop, these babies may require Cesarean-section delivery and an extensive operation after birth. Most babies will do well once the tumor is completely removed. Blood testing for alpha-fetoprotein (AFP) levels should be performed routinely. There can also be long-term consequences which include the reccurence of the tumor or difficulty with bowel and/or bladder control as a consequence of the surgical procedure

The vascular supply to an SCT commonly arises from the middle sacral artery, which can enlarge to the size of the common iliac artery and cause a vascular steal syndrome. These large vascular tumors can lead to high-output cardiac failure as a consequence of arteriovenous shunting through the tumor, resulting in placentomegaly, hydrops.

**Radiographic features:**

**Plain film:** may show a large mass projecting from the lower pelvic region or within the abdominopelvic cavity, may show calcification.

**CT:** Not part of routine investigation. Identifies bone, fat and cystic components. Calcification may again be seen.

**Ultrasound:** Mature types tend to be more cystic which show as anechoic components. Solid types (which are much rarer) often show an echogenic mass within the pelvis. The correlation between sonographic appearances and malignant components are thought to be poor. Colour Doppler interrogation in some tumours may show marked hypervascularity with arterio-venous (AV) shunting.

Signal characteristics can significantly vary depending on the constituent of the teratoma .

- T1: fat components appear high signal, calcific/bony components low signal
- T2: fluid (cystic) components appear high signal, calcific bony components low signal
- T2* GRE: magnetic susceptibility artifact because of calcifications
- T1 C+ (Gd): enhancing solid components.

## VI. Conclusion

The cause of Sacrococcygeal Teratoma / SCT is not known. Though a rare condition (1 in 35,000-40,000 births), Sacrococcygeal Teratoma / SCT is one Sacrococcygeal Teratoma / SCT is detected by ultrasound as a mass on the fetal buttocks associated with a uterus larger than it should be at a specific gestational age. The reason for this is often polyhydramnios While the mortality rate for Sacrococcygeal Teratoma / SCT diagnosed in a newborn is less than five percent, the mortality rate for Sacrococcygeal Teratoma / SCT in utero is 50 percent.

Thus, there should be close observation during pregnancy. Weekly ultrasounds should be performed to confirm or rule out associated abnormalities, monitor amniotic fluid, tumor growth, fetal well being and early signs of hydrops Weekly fetal echocardiograms will detect early changes in cardiac function with increases in combined ventricular output, increased aortic flow and dilatation of the SVC. After 30 weeks of gestation, weekly amniocentesis may be recommended to determine pulmonary maturity, which may allow for an early delivery. The mother should be observed for signs of preterm labor, preeclampsia (toxemia), or the "mirror syndrome," in which the mother mirrors the symptoms of the fetus when placentomegaly has developed. Delivery should occur in a tertiary care hospital where neonatologists and pediatric surgeons are available to provide care.

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