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## CASE REPORT

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# Sacrococcygeal Teratoma — Role of Ultrasound in Antenatal Diagnosis and Management

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### ABSTRACT

*Sacrococcygeal teratomas are common congenital tumours that develop early in foetal life. Foetuses with this malformation are at risk for significant perinatal morbidity and mortality. This report demonstrates the role of foetal sonography in the diagnosis of sacrococcygeal teratoma and reviews the role of imaging in guiding antenatal and postnatal management.*

*Key Words: Antenatal diagnosis, Sacrococcygeal region, Teratoma, Ultrasound*

### INTRODUCTION

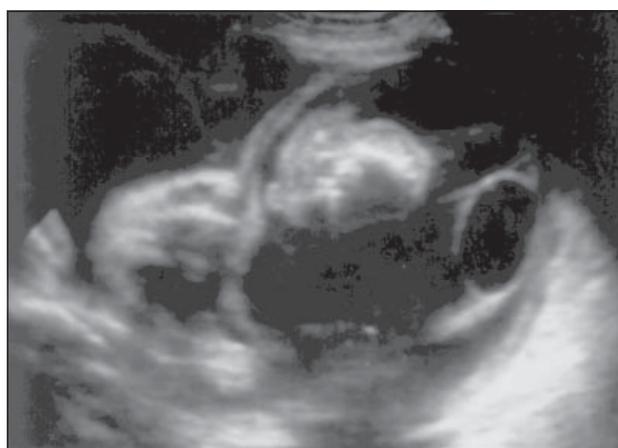
Sacrococcygeal teratomas (SCT) are relatively common congenital tumours that develop early in foetal life.<sup>1</sup> SCT may become large in utero and can be confidently diagnosed by prenatal sonography in the second or early third trimester of pregnancy.<sup>2</sup> Although the majority of these tumours are histologically benign, they are nevertheless associated with significant morbidity and mortality due to such concomitant factors as prematurity of the infant, dystocia, traumatic delivery, and intratumoural haemorrhage. This report demonstrates the role of foetal sonography in the diagnosis of SCT and reviews the role of imaging in guiding antenatal and postnatal management.

### CASE REPORT

A 28-year-old woman, gravida 2 para 1, was referred for level II ultrasound scan at 22 weeks gestation. There was no family history of birth defects. The sonographic examination revealed a single intrauterine pregnancy with an estimated gestational age of 23 weeks. The study revealed a large 5 x 6 x 5 cm mixed echogenic mass arising from the sacrococcygeal region (Figure 1). There were cystic areas as well as calcific foci within the mass (Figure 2). The spine appeared intact (Figure 3), and



**Figure 1.** Longitudinal ultrasonography scan shows a complex septated cystic solid mass protruding from the caudal part of spine.



**Figure 2.** Parasagittal scan showing normal lumbosacral spine.

the lower limbs appeared normal. The foetal kidneys and bladder appeared normal. There was no evidence

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**Figure 3.** Ultrasonography scan showing calcific foci within the mass with posterior acoustic shadowing.

of possible invasion of foetal pelvis and abdomen. The surrounding structures in the foetal pelvis and abdomen were normal. Liquor was adequate.

Based on the above findings, a diagnosis of purely external (type I) variety of SCT was made. The patient decided to continue the pregnancy and was scheduled for follow up ultrasound 4 weeks later. However, she did not keep the appointment for follow up ultrasound and presented at 30 weeks gestation with rapid increase in the size of the uterus, premature rupture of membranes, and spontaneous labour pains. She delivered a dead foetus with a large sacrococcygeal mass measuring 10 x 10 x 8 cm, which had external haemorrhagic areas. At histopathological examination, the mass was confirmed to be a mature variety of SCT. Autopsy was refused. At local examination, however, there was no evidence of foetal hydrops. Placental size was within normal limits.

## DISCUSSION

Teratomas are neoplasms that contain derivatives of more than one of the 3 primary germ cell layers of the embryo.<sup>3</sup> Although these tumours are benign, one or more of the germ cell layer derivatives may develop malignant characteristics. Teratomas usually arise as a mass in the sacrococcygeal region. Although the exact aetiology of most SCT is unknown, the majority are thought to be sporadic.<sup>3</sup> SCT is a common neoplasm with a reported incidence of 1 in 30,000 to 40,000 births.

SCT are usually large (average size, 8 to 10 cm), well encapsulated, and grossly lobulated. These masses possess both cystic and solid components.<sup>4</sup> They are classified according to the degree of exterior component

or intrapelvic extension. Type I tumours (46.7%) predominantly lie external to the foetus and type II tumours (34.1%) present externally but have significant intrapelvic extension. Type III tumours (8.8%) are apparent externally but predominantly lie within the pelvis and abdomen. Type IV tumours (9.8%) are entirely presacral with no external presentation.<sup>3,4</sup> Only 2% of SCT grow into the spinal canal.

Embryologically, SCT are thought to derive from a pluripotential cell line that escapes from the control of embryonic inductors and differentiates into various tissues not normally found in the sacrococcygeal region.<sup>1,5</sup> Their predilection for this area is most likely to be related to the large number of pluripotential cells usually found in the caudal region of the embryo, closely associated with the distal sacrum and coccyx.<sup>1,5,6</sup> Most SCT are histologically benign and may have mature adult type components or immature elements (20%).<sup>1</sup> Malignant SCT are extremely rare in the foetus and uncommon in newborn infants. The likelihood of malignancy greatly increases in tumours diagnosed after the infant is 2 to 4 months old.<sup>1</sup>

In recent years, the practice of routine obstetrical ultrasound scan has led to a significant increase in the number of SCT diagnosed in utero. The teratoma in the patient reported here appeared as a mixture of cystic and solid components (Figure 1). The majority of prenatally diagnosed teratomas are solid or mixed in echogenicity with interspersed or peripheral cystic components. A purely anechoic form of SCT has also been described.<sup>7-10</sup> The imaging of tumours that contain calcification varies in terms of increased echogenicity or echogenicity with shadowing. Dystrophic calcification in areas of haemorrhage and necrosis can also be found. Fragments of bone or rudimentary limbs may be present and appear as bright foci with distal acoustic shadowing.<sup>7-10</sup> Serial sonograms may show a change in appearance of the mass, since areas of haemorrhage or degeneration develop in large SCT. Congenital anomalies may be present in association with SCT, including genitourinary, anorectal, and lower vertebral malformations, and need to be ruled out during prenatal sonography.<sup>2,3,5</sup>

Recently, prenatal magnetic resonance imaging (MRI) has been added to the armamentarium of radiologists as a tool for imaging antenatal foetal anomalies. MRI has several advantages over obstetric ultrasound, including a large field of view, superior soft tissue contrast, more precise volumetric measurements, and greater accuracy

in demonstration of associated abnormalities and intrapelvic extension.<sup>11</sup>

Several factors can be predicted on the basis of prenatal sonographic appearance, which influence the prognosis for the foetus. Foetal SCT are rarely malignant but large benign tumours are associated with significant morbidity and mortality. In such foetuses, complications result from massive intratumoural haemorrhage and dystocia.<sup>10,12</sup> Normal development of vital organs, particularly kidneys and lungs, may be hampered by compression from a large retroperitoneal component. The presence of hydronephrosis appears to be a very unfavourable prognostic sign.

Several mechanisms may be responsible for renal obstruction and secondary dysplasia, including urethral obstruction by the perineal component of the tumour and direct ureteral compression or invasion by a large retroperitoneal component of the mass.<sup>7</sup> Jouannic et al reported a patient with SCT diagnosed at 22 weeks gestation with a significant intrapelvic component leading to bladder outlet obstruction at 27 weeks.<sup>13</sup> Ultrasonography-guided prenatal percutaneous shunting of a cystic teratoma was performed at 28 weeks gestation to avoid prolonged foetal compression by the tumour that could have adverse effects by stretching pelvic and sacral nerves. Urinary dilatation resolved completely after shunting.<sup>13</sup>

Foetal hydrops associated with ascites, pleural effusions, skin thickening, and placentomegaly is another unfavourable prognostic sign.<sup>8-10</sup> The hydrops may be secondary to direct compression of major vascular structures. If the hydrops develops in a foetus with SCT and the foetus is sufficiently advanced in gestation to have developed lung maturity, emergency delivery and standard postnatal resection is appropriate. However, for foetuses that become hydropic before the development of lung maturity, foetal surgical intervention may be the only hope for survival.<sup>12</sup> Prenatal SCT resection is aimed solely at tumour debulking and devascularisation to eliminate large arteriovenous shunts through the tumour, but this should be considered only by experienced surgeons.<sup>12</sup> Close maternal foetal observation and administration of corticosteroids in an attempt to accelerate foetal pulmonary maturation is advisable.

The patient presented here had preterm labour with sudden increase in size of the uterus, probably due to rapid increase in amniotic fluid volume and size of the

mass. Large pelvic or abdominal extension of SCT is rare and necessitates a more extensive surgical approach. Prenatal detection can alert the surgeon and ultrasound, computed tomography, or MRI can be performed in the postnatal period allowing for better surgical planning.<sup>10</sup> Three-dimensional ultrasound may better define the degree of involvement of the sacrum and pelvic structures of prognostic importance.<sup>14</sup> Widening of the lumbosacral canal, which may also be better appreciated on 3-dimensional scanning, indicates spread of teratoma within the vertebral canal. Recently, prenatal MRI has also been shown to positively and incrementally influence management for a substantial portion of patients being considered for foetal intervention. However, precise indicators for prenatal MRI are not yet established. Avni et al evaluated the usefulness of MRI in the diagnosis and assessment of foetal SCT to determine whether MRI provides information not seen on obstetric sonography.<sup>15</sup> These authors concluded that SCT had characteristic MRI appearances that allowed a complete assessment of most foetuses. Due to MRI, the prenatal evaluation was changed for 3 patients, including 1 foetus with an extension of tumour within the spinal canal. This recognition at MRI altered the treatment and counselling for the patient.<sup>15</sup>

The amniotic fluid volume is usually markedly increased and onset of polyhydramnios is often acute, as happened in this patient. The presence of polyhydramnios may be a factor in precipitating preterm labour secondary to uterine distension. The cause of polyhydramnios is usually unclear. Volume reduction amniocentesis and tocolysis may be required to treat symptomatic polyhydramnios and prevent preterm delivery.<sup>10,11</sup>

SCT can usually be confidently diagnosed on prenatal sonography when a complex or solid mass attached to the foetal rump is depicted. In certain patients, however, other entities have to be excluded. A predominantly cystic SCT must be distinguished from a myelomeningocele. The latter is always associated with spinal dysraphism and careful examination of the posterior element of the lumbosacral spine is critical.<sup>8</sup> In an unusual case reported in the literature, colour Doppler imaging assisted prenatal sonographic diagnosis of a large type I cystic SCT closely simulating a myelomeningocele.<sup>16</sup> Other tumours such as lymphangiomas, chordomas, and ependymomas need be considered only rarely since they are usually much smaller, seldom have a large external component, or occur in a different age group after birth.

The gestational age of the foetus closely affects the perinatal outcome. Most foetuses with large SCT are born prematurely, with resultant increased morbidity due to lung immaturity and other complications associated with prematurity.<sup>10,12</sup> Foetal well-being should be monitored because SCT are associated with an increased incidence of stillbirths and foetal deaths in utero. The causes of foetal death include massive haemorrhage within the tumour and profound anaemia, high output cardiac failure, arteriovenous shunting, and steal phenomenon within the mass.<sup>17</sup> Hase et al felt that several considerations, including control of haemorrhage, were necessary to facilitate surgical management of massive SCT.<sup>18</sup> Foetal Doppler could be used to identify feeding vessels and vascularity of the tumour and aid appropriate devascularisation techniques during postnatal resection of the tumour. Lam et al performed ultrasound-guided thermocoagulation of feeding vessels of SCT at 18 weeks.<sup>19</sup> The blood supply was successfully reduced, although foetal death resulted 2 days after the procedure. Paek et al used an innovative percutaneous technique of ultrasound-guided radiofrequency ablation to interrupt the blood flow to SCT in 4 foetuses and concluded that targeted ablation of the feeding vessels diminishes blood flow sufficiently to reverse high output cardiac failure.<sup>20</sup> The selection of foetuses that may benefit from in utero surgery is being developed and prenatal diagnostic techniques are utilised to gain as much knowledge about the foetus so that appropriate counselling of the parents can be undertaken.

Thus, prenatal sonography of SCT allows for optimal antenatal, perinatal, obstetric, and surgical management. It allows characterisation of the nature of the mass (solid versus cystic, vascularity), follow up of tumour size, and rapid identification of any complications. MRI is a valuable adjunct to obstetric sonography for the prenatal evaluation of SCT, allowing for a more timely and appropriate delivery or the possibility of in utero surgery. By recognising the problem of dystocia at birth from a large tumour mass, which could lead to tumour rupture, haemorrhage, or birth asphyxia, elective caesarian section has become the rule for lesions greater than 5 cm in size.<sup>21,22</sup> The optimal uterine incision is one that will permit atraumatic delivery of the infant and minimise the risk of tumour rupture. In 2 patients described in the literature, ultrasound-guided needle aspiration of cystic SCT was performed in utero to facilitate delivery.<sup>23</sup> However, since the majority of SCT have solid components, early delivery as soon as foetal lung maturity is achieved is recommended.

After delivery, perinatal management of foetal SCT requires an interdisciplinary team of radiologists, obstetricians, neonatologists, and paediatric surgeons (Figure 4). Surgical removal of SCT should be performed without delay to avoid complications such as haemorrhage and ulceration and to minimise the risk of malignant degeneration. The coccyx should be removed with the tumour to avoid local recurrence. Some authors suggest that most infants can safely undergo a short period of stabilisation before surgery, including

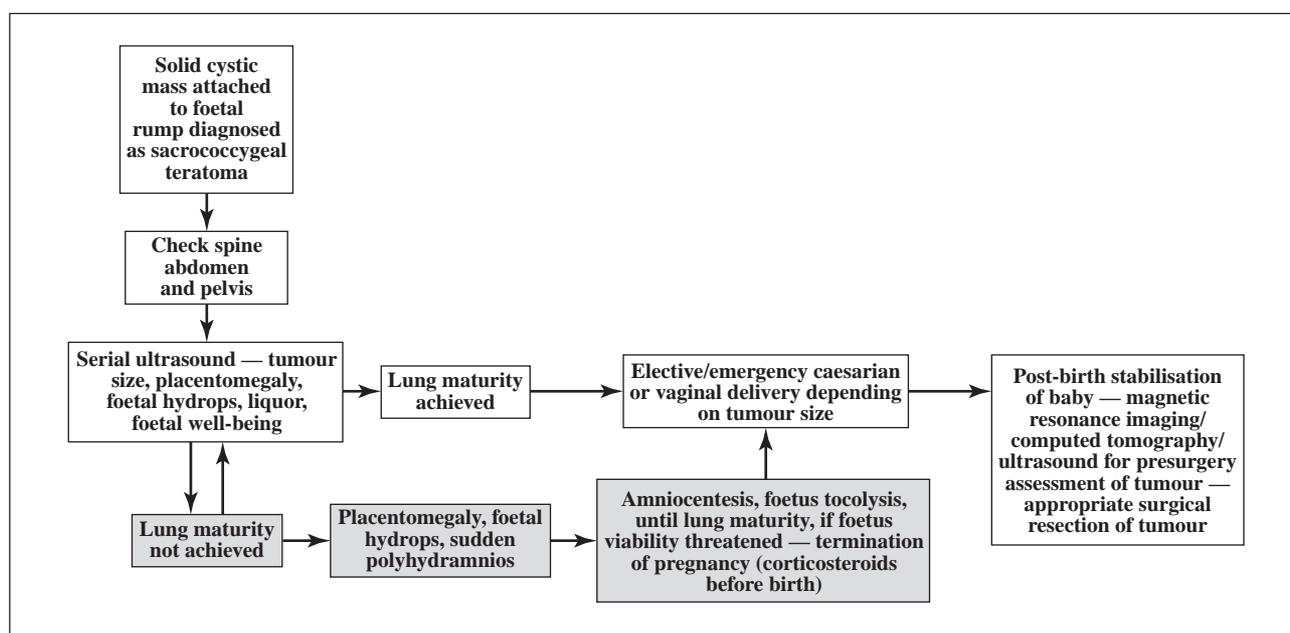


Figure 4. Flow chart showing role of ultrasonography in prenatal management of sacrococcygeal teratomas.

assessment of cardiovascular status, MRI or ultrasound to determine the extent of intra-abdominal tumour extension, and assist in surgical planning.<sup>10,12,24,25</sup> The prognosis for cure is generally extremely good after successful complete removal of benign SCT.

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