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## Cystic Type IV Sacrococcygeal Teratoma

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### CASE HISTORY

A 3050-g female infant was born at 30 weeks' gestation to a 28-year-old mother after a pregnancy complicated by a sonogram at 20 weeks' gestation demonstrating a large cystic mass in the abdomen and oligohydramnios. The infant was delivered by Cesarean section and was cyanotic without spontaneous respiration. The patient was resuscitated in the delivery room but had APGAR scores of 1 at 1 minute and 7 at 5 minutes. Plain radiographs, a sonogram (Figure 1), and a magnetic resonance imaging (MRI) (Figure 2) of the abdomen were obtained. The plain radiograph demonstrated a distended abdomen with multiple calcifications.

### DENOUEMENT AND DISCUSSION

This large abdominopelvic mass with calcifications, suggested by sonogram and MRI (Figure 3), represented a sacrococcygeal teratoma with a small presacral component and a massive abdominopelvic portion. This was confirmed at biopsy. Resection was undertaken at the age of several weeks because of the large size of the mass and bleeding. Definitive resection followed embolization of internal iliac arterial feeding vessel bilaterally to decrease the risk of bleeding during definitive resection.

Sacrococcygeal teratoma is the most common neonatal neoplasm. It is found predominantly in females and discovered in the neonatal period.<sup>1</sup> These tumors are believed to arise from embryologically multipotent cells of Hensen's node, which lies within the coccyx.<sup>1</sup>

The tumors are classified, according to Altman Classification of the Surgical Section of the American Academy of Pediatrics, into four types: Type I, those tumors that are predominantly external projecting from the sacrococcygeal region and presenting with

distortion of the buttocks; Type II, those tumors that are predominantly external, but have a large intrapelvic component; Type III, those that are predominantly intrapelvic with a small external, buttock mass; Type IV, those that are entirely internal with no external or buttock component.<sup>1</sup> Type IV sacrococcygeal teratomas may occur as a familial form inherited as an autosomal dominant condition. In this entity, a presacral mass (teratoma, anterior meningocele, dermoid cyst, lipoma, neurofibroma, enteric cysts, or hamartoma) is associated with anal stenosis and typical Scimitar defect of the sacrum. This is often referred to as Currarino triad.<sup>2,4</sup> Approximately 45% of teratomas is Type I, 35% is Type II, 10% Type III, and 10% Type IV.<sup>4</sup>

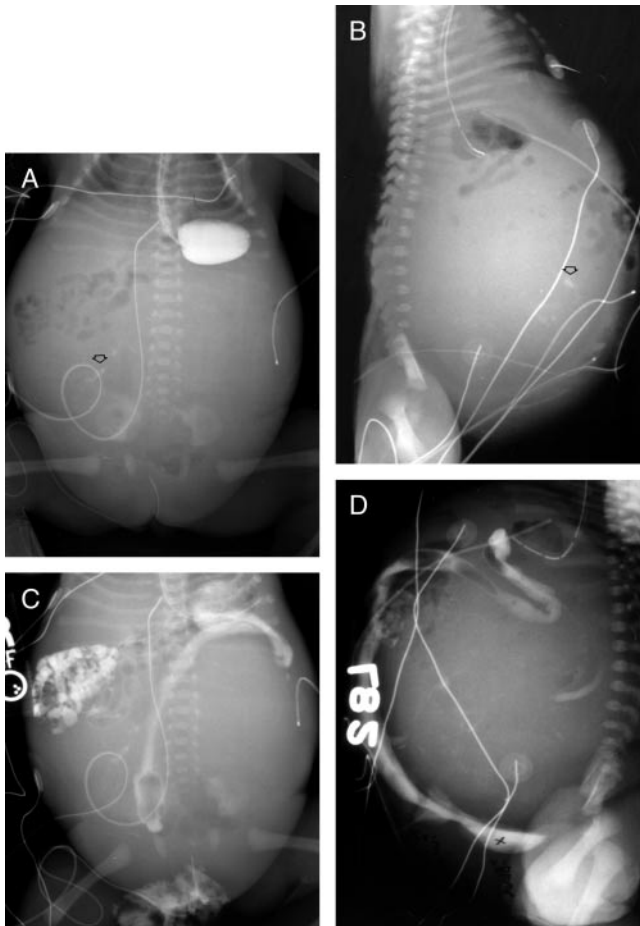
All of the four types may have an intraspinal component. Spinal involvement may be associated with neurologic deficit. This did not occur in the patient described here.

Malignant transformation appears to increase in frequency with the time to diagnosis from birth. Only 7% to 10% of tumors diagnosed before age 2 months are malignant. However, after 2 months, the incidence of malignancy rises to 66% in boys and 50% in girls.<sup>4</sup> Thus, the sex incidence of malignant lesions is equal or even slightly predominantly male in distinction to the female predominance in all patients with sacrococcygeal teratomas.<sup>3</sup> Almost all reported cases of malignant degeneration have been malignant teratomas with endodermal sinus tumor.<sup>3</sup> The teratomas may be cystic, solid, or mixed. Predominantly cystic lesions are usually benign. Type I lesions are almost always benign.<sup>4</sup> Calcifications occur in one third of sacrococcygeal teratomas,<sup>1</sup> and more frequently seen in benign than malignant lesions.

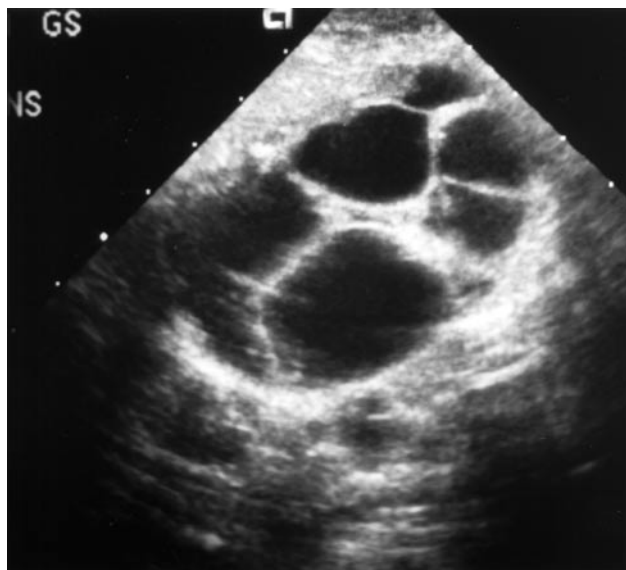
The recommended treatment is resection of the tumor en bloc with the coccyx.<sup>5</sup> Surgical difficulties include hemorrhagic complications and coagulopathy.<sup>5</sup> Hemorrhage is the most common cause of death among neonates with sacrococcygeal teratomas. The mortality in the neonatal period is approximately 16%.<sup>5</sup> Features that correlate with an increased risk of hemorrhage include polyhydramnios, large-size lesions, and fetal distress.<sup>5</sup> Other poor prognostic features include congestive heart failure, placentomegaly, and hydrops fetalis. These latter three findings have been associated with 100% mortality.<sup>5</sup> Sacrococcygeal teratomas are often very vascular tumors, sometimes having hemodynamically significant arteriovenous shunting. In addition, some patients with sacrococcygeal teratomas have coagulopathy possibly due to tumor trauma at time of delivery. The trauma is thought to cause release of thromboplastins into the bloodstream, resulting in a coagulopathy and even DIC.

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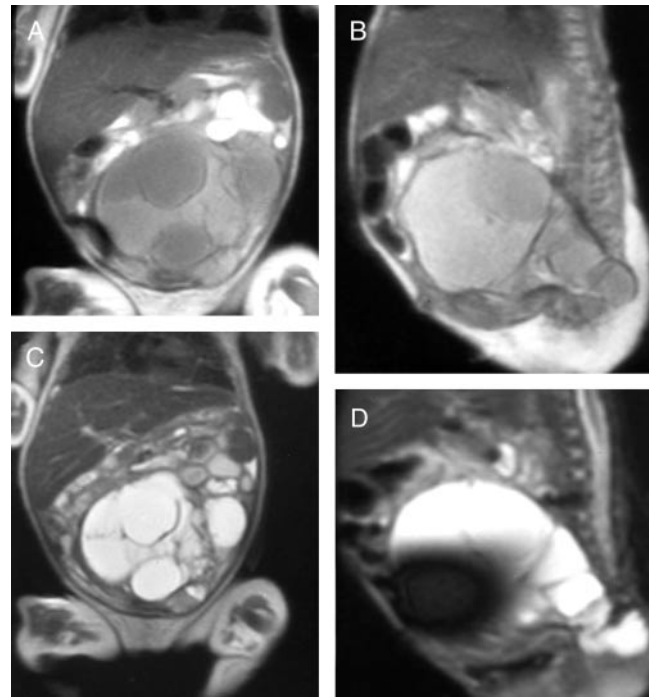
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**Figure 1.** Plain radiographs of the abdomen during small bowel follow through study anteroposterior (A) and lateral (B) and with contrast in the colon; anteroposterior (C) and lateral (D). A large mass is present with calcifications (arrows). This mass displaces the sigmoid and left colon far anteriorly.



**Figure 2.** Sonograms of the lower abdomen demonstrate the mass to be composed of multiple cysts.



**Figure 3.** MRI scans of T1-weighted coronal (A) and midline sagittal (B) and T2-weighted coronal (C) and midline sagittal; (D) the large mass of cysts is seen extending from the infracoccygeal and presacral regions into the abdomen.

### References

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