Society of Radiologists in Ultrasound
2011 Toshiba Resident Teaching Case

Logan McLean, MD
University of Utah
Salt Lake City, UT

CLINICAL HISTORY

Size greater than dates at 28 weeks gestation. Normal obstetrical ultrasound for fetal anatomy at 19 weeks.

FIGURES: See attached

FIGURE LEGENDS:

Figure 1. Longitudinal (A, B)) ultrasound images of a 28-week fetus shows an exophytic, heterogeneous mass (arrows) with solid and cystic components. The mass appears to arise from the distal sacrum and, invades into the pelvis (B, arrowheads) resulting in obstruction of the vagina (*) and bladder (+). Transverse (C) ultrasound image with color Doppler shows the vascularity of this tumor.

Figure 2. Transverse ultrasound image of the fetal pelvis shows a mildly dilated urinary bladder (arrowhead) and bilateral hydronephrosis (arrows). The intrapelvic component of this mass is causing compression of the bladder and subsequent dilatation of the ureters.

Figure 3. Transverse ultrasound image of the fetal chest was obtained to calculate the cardiothoracic ratio. The cardiothoracic ratio was mildly increased, 0.54, suggesting developing high-output cardiac failure. No pericardial effusion or additional features of hydrops were demonstrated.

Figure 4. Axial (A), sagittal (B) T2 HASTE images of the fetus show the mass is originating from the sacrum (arrow). On the sagittal image (B), the intrapelvic extension (arrowhead) of the mass is well delinated with MRI, along with the resultant hydrocolpos (*) and anterior compression of the bladder (+).

Figure 5. Clinical photograph of the baby in a right lateral decubitus position shows the solid mass protruding from the sacrococcygeal region (arrowheads). Note the large vessels on the tumor surface. This baby was delivered at 30 weeks after the mother developed mirror syndrome. The mass ruptured during the cesarean section. An attempt to ligate the tumor was performed however, the baby did not survive.

DIAGNOSIS

Sacrococcygeal teratoma, Type II

DISCUSSION

A teratoma is a neoplasm derived from all three germ cell layers: ectoderm, mesoderm and endoderm [1]. Approximately 70-80% of teratomas in the fetus occur in the sacrococcygeal region [2]. SCGT is the most common neonatal tumor with an incidence of 1:40,000 live births [1].

On ultrasound, sacrococcygeal teratomas (SCGT) are most likely to be diagnosed in the second trimester. The classic sonographic appearance of a SCGT is a heterogeneous mass with mixed solid and cystic components, which may contain calcifications. SCGT's are purely cystic in only 15%. The more solid the SCGT is the more likely it is to have a poor outcome compared to the more cystic tumors. The solid tumor to head volume of (STV/HV > 1) identifies a fetus with risk for poor outcome [3]. Intratumoral hemorrhage may be present and also portends a poor prognosis. Color Doppler is useful to evaluate vascularity and arteriovenous shunting, which increases the risk for hydrops [2].

To stage this tumor, it is important to evaluate for extension into the pelvis or abdomen [4]. MR imaging is superior in evaluation of the extent of intrapelvic, abdominal and spine involvement, which is very useful for surgical planning. MR is also helpful in identifying intratumoral hemorrhage.

The American Academy of Pediatric Surgery Section classifies sacrococcgyeal teratoma as follows [5]:

> Type 1: Completely external or minimal presacral component.
>
> Type 2: External and internal component extending into the presacral space.
>
> Type 3: External and internal component extending into abdomen.
>
> Type 4: Completely internal, no external component. These tumors are most likely to undergo postnatal malignant degeneration.

Although chromosomal abnormalities are associated with sacrococcygeal teratoma, there are several associated findings to evaluate for on prenatal imaging. Genitourinary abnormalities including hydronephrosis, renal dysplasia, ureteral atresia, urinary ascites and hydrocolpos are often associated with this diagnosis [4,6]. Other anomalies include rectal atresia or stenosis, hip dislocations and clubbed feet.

The primary differential diagnosis of SCGT is a myelomeningocele (MMC). A MMC is a complex cystic mass that contains both meninges and neural elements. MMC causes splaying of the dorsal ossification centers, unlike SCGT, and most often extends posteriorly from the spine. Other fetal solid tumors, including rhabdomyosarcoma and fibrosarcoma, are in the differential diagnosis for a SCGT, however are incredibly rare.

In the clinical setting, the mother often presents measuring size greater than dates secondary to the SGCT itself or associated polyhydramnios. Polyhydramnios may cause premature labor and delivery. Delivery complications include dystocia, tumor avulsion and fetal exsanguination [7]. Specifically, there is a 50% mortality for the fetus due to hydrops from a high-output state and intratumoral hemorrhage. Poor prognostic factors include hydrops, diagnosis before 30 weeks and having a large solid component [2]. The prognosis is significantly worse for the fetus than neonate. Maternal complications include hyperemesis, preeclampsia and preterm labor [4]. The mother can also develop a potentially life-threatening condition, mirror syndrome, where her symptoms "mirror" that of the fetus. This syndrome presents with maternal fluid retention and hemodilution, progressive edema develops and ultimately requires immediate delivery.

Treatment options are limited but include therapeutic amnioreduction for symptomatic polyhydramnios [7]. Fetal surgery has been performed in rare cases where fetal decompensation and hydrops occurs at < 30 weeks but the success of surgery has been quite limited [4,7]. Ultimately, the outcome is universally fatal once the fetus develops placentomegaly and hydrops. Delivery in a tertiary care center is recommended when the fetal lungs reach maturity. Cesarean section delivery is recommended for tumors > 5cm.

REFERENCES

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2. Woodward PJ, Sohaey R, Kennedy A, Koeller KK. From the archives of the AFIP: a comprehensive review of fetal tumors with pathologic correlation. *Radiographics.* 2005 25(1): 215-42.

3. Sy ED, Filly RA, Cheong ML, Clifton MS, et al. Prognostic role of tumor-head volume ratio in fetal sacrococcygeal teratoma. *Fetal Diagn Ther.* 2009 26(2): 75-80.

4. Hedrick HL, Flake AW, Crombleholme TM, Howell LJ, et al. Sacrococcygeal teratoma: prenatal assessment, fetal intervention and outcome. *J Pediatr Surg*. 2004 39 (3): 430-8.

5. Altman, RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American academy of pediatrics surgical section survey –1973. *J Pediatr Surg.* 1974 9: 389-398.

6. Milam DF, Cartwright PC, Snow BW. Urological manifestations of sacrococcygeal teratoma. *J Urol.* 1993 149:574-576.

7. Coleman BG, Adzick NS, Crombleholme TM, Johnson MP, et al. Fetal therapy: state of the art. *J Ultrasound Med*. 2002 21(11): 1257-88.

AUTHOR: Logan A. McLean, MD

INSTITUTION

University of Utah Department of Radiology

30 North 1900 East, #1A071

Salt Lake City, UT 84132-2140

The University of Utah

Department of Radiology

UNKNOWN CASE, TOSHIBA RESIDENT

SRU Meeting Chicago, IL October 21-23, 2011

Logan A. McLean, MD

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FIGURE 1

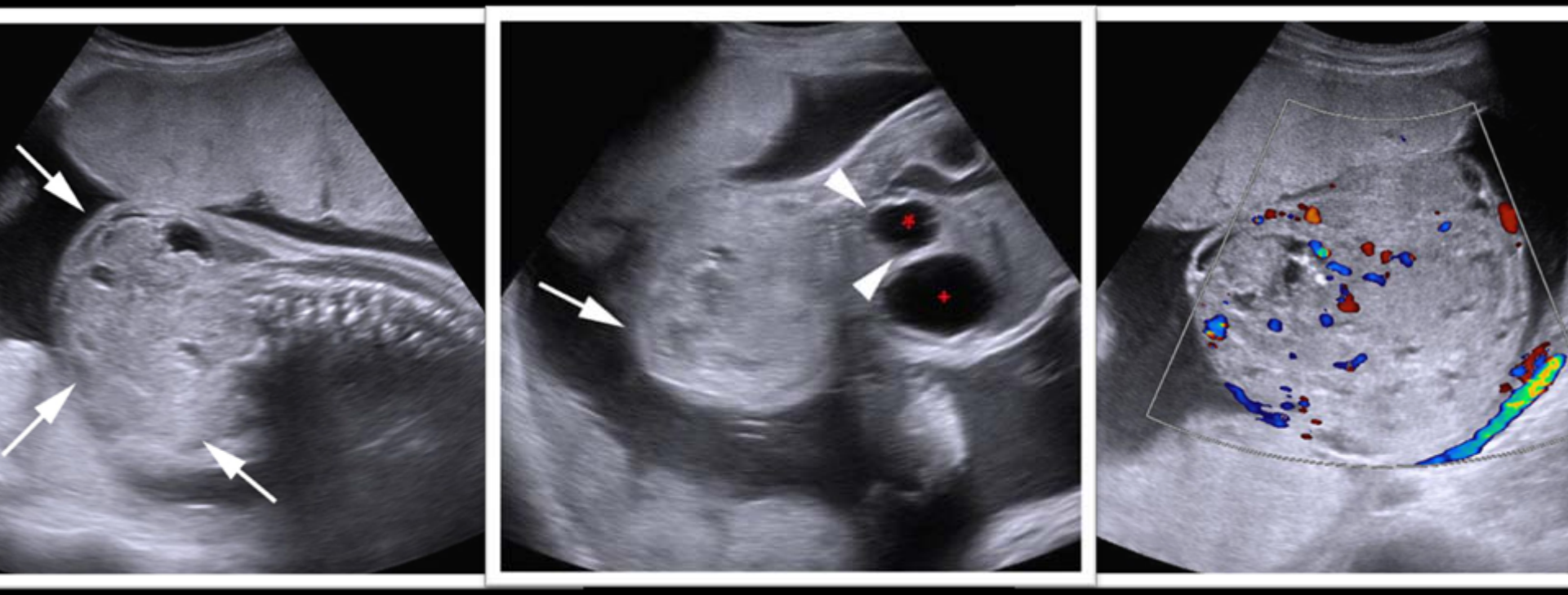
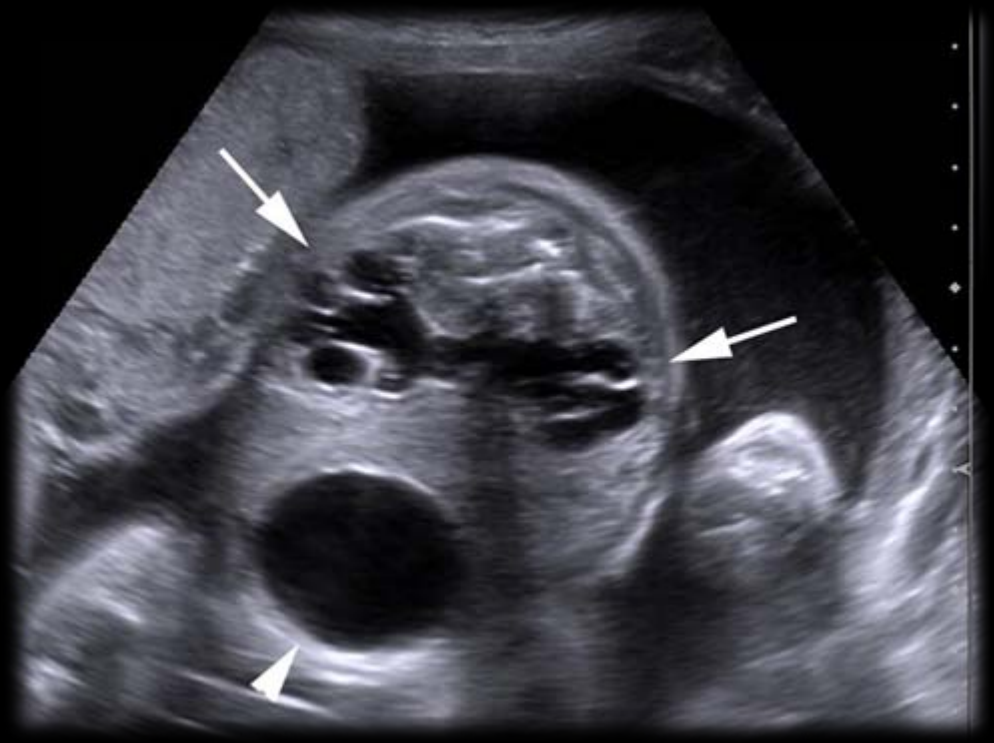


FIGURE 2



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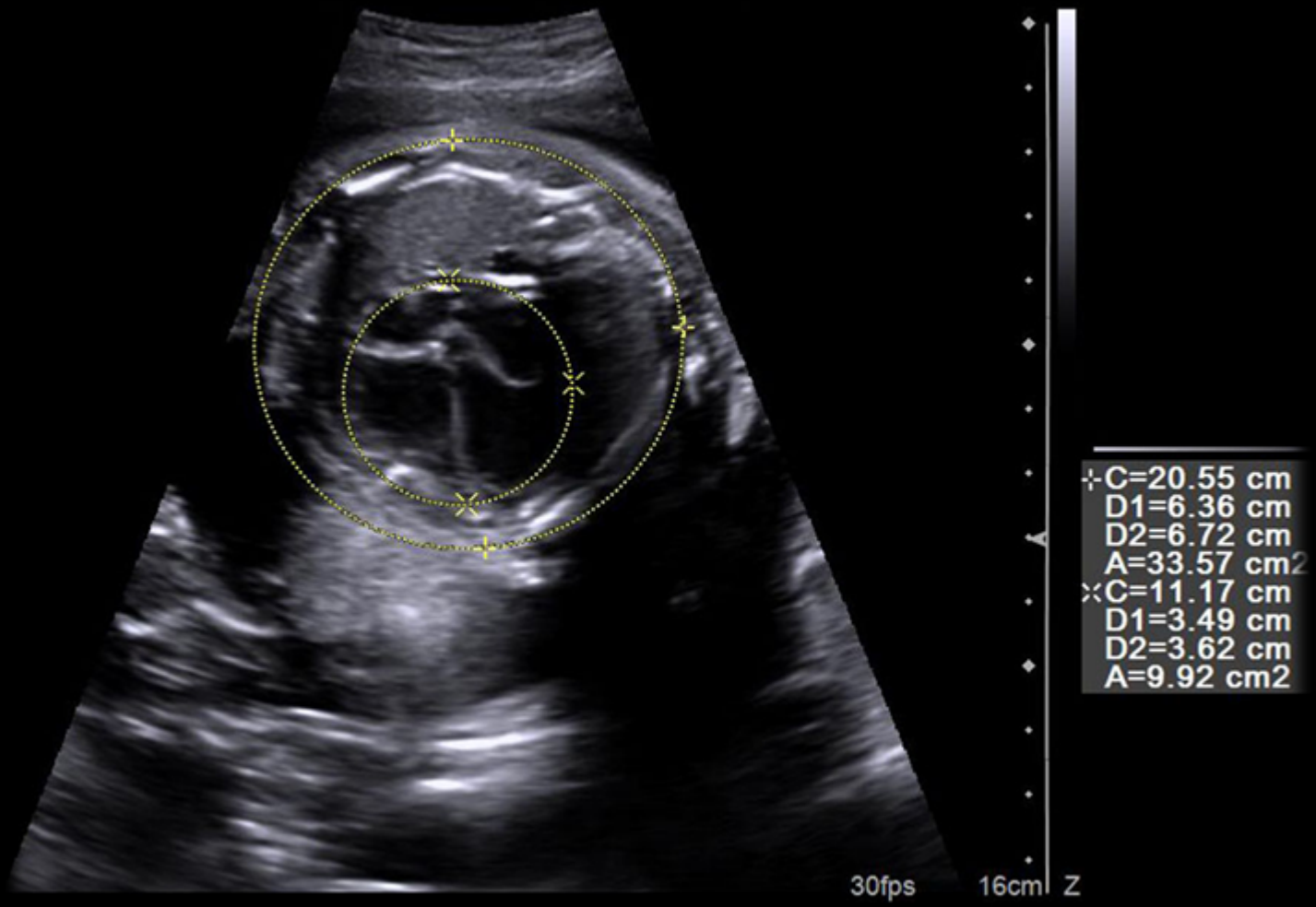
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D1=6.36 cm
D2=6.72 cm
A=33.57 cm2
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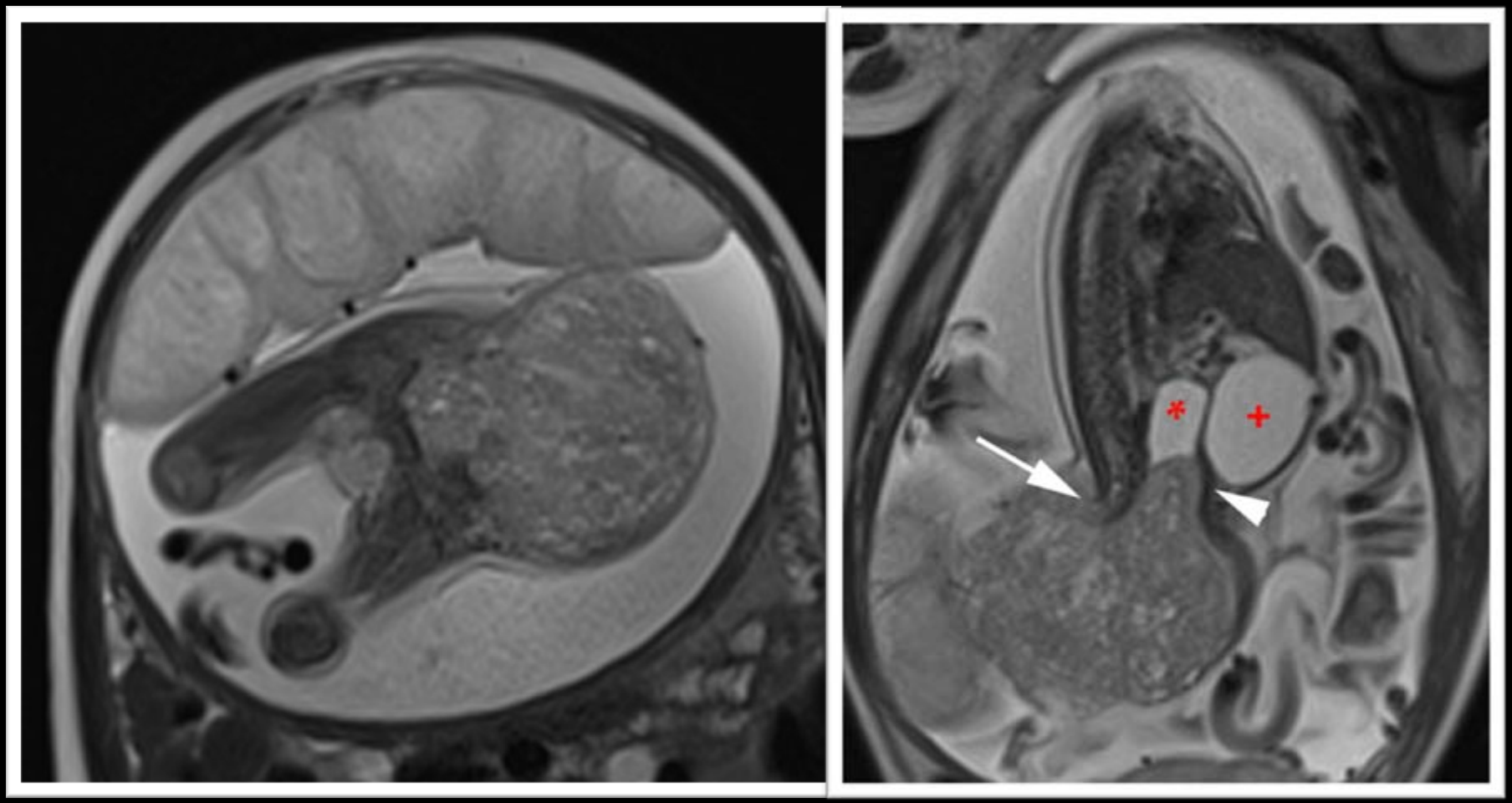
D1=3.49 cm
D2=3.62 cm
A=9.92 cm2
30fps
16cm
Z

FIGURE 4



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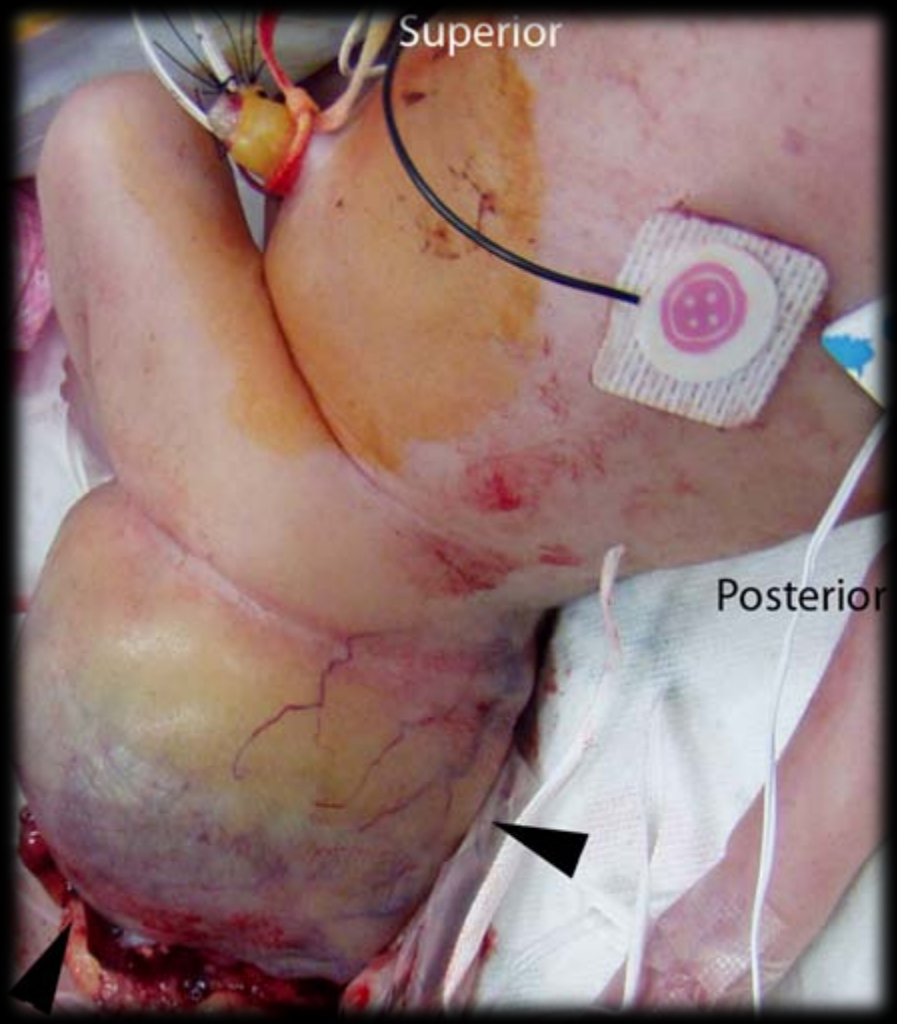

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