KARYA ILHAM PUBLISHING
CONNECTING MINDS, BRIDGING IDEAS, INSPIRING READERS

Journal of Health and Quality of Life

Journal homepage:
https://karyailham.com.my/index.php/jhqol/index
ISSN: 3030-5101

Sacrococcygeal Teratoma: Tales of the "Tailbone" Tumour in a Newborn

Tharveen Nair Chandrasekaran[1,2], Pwi Jia Yee[3], Bhuwaneswaran Vijayam[2,4,5*]

[1] Surgical Department, Hospital Sultan Ismail, 81100, Johor Bahru, Johor, Malaysia
[2] Johor Bahru Parenteral and Enteral Nutrition Society (JPENS), 80100, Johor Bahru, Johor, Malaysia
[3] NUMed Surgical Society, Newcastle University Medicine Malaysia (NUMed), 79200 Iskandar Puteri, Johor, Malaysia.
[4] Regenerative Medicine Working Group, Newcastle University Medicine Malaysia (NUMed), 79200 Iskandar Puteri, Johor, Malaysia.
[5] Department of Clinical Medicine, Newcastle University Medicine Malaysia (NUMed), 79200 Iskandar Puteri, Johor, Malaysia.

ARTICLE INFO

Article history:
Received 13 May 2024
Received in revised form 11 Jun 2024
Accepted 10 July 2024
Available online 16 August 2024

Keywords:
Sacrococcygeal teratoma; tailbone tumour; tailbone teratoma

ABSTRACT

Sacrococcygeal teratoma (SCT) is a rare tumour with a low incidence, but it is the most common extragonadal tumour in the newborn. It occurs at the base of the coccyx (tailbone) and is derived from embryonic germ cell layers. Although the majority are benign in nature, they are associated with significant morbidity and mortality for the mother and the foetus. A multidisciplinary approach is needed where an accurate early diagnosis should be made antenatally, leading to anticipation of complications during intrapartum for the mother and foetus, and ultimately early, complete surgical excision of the tumour. The prognosis for survival is good after resection. We report a case of a newborn delivered at 29 weeks who was diagnosed with SCT at post-partum but unfortunately passed away due to the complications of prematurity before surgery could be performed. This case report highlights the literature review of this pathology, especially the management of SCT, which begins in the antenatal period soon after the diagnosis is made.

1. Introduction

Sacrococcygeal teratoma (SCT) is a rare condition that affects one in every 35,000 to 40,000 live births. An epidemiological study at a tertiary centre in Malaysia showed that SCT accounted for almost half of all the neonatal tumours treated in that setting [1]. Typically, an infant with SCT would also co-manifest other sensory-motor, gastrointestinal and excretion abnormalities. Despite being easily detectable during pregnancy via an ultrasound, the SCT lacks clear, unified recommendations. This case report will highlight our encounter with SCT in an infant, as well as the scientific discussion behind the clinical experience.

* *Corresponding author.*
E-mail address: bhuwaneswaran.vijayam@newcastle.edu.my

https://doi.org/10.37934/jhqol.3.1.5562

2. Case Report

A 20-year-old primigravida was sent to our obstetrics and gynaecology colleagues for spontaneous preterm labour at 29 weeks of gestation. At 28 weeks of pregnancy, she was advised to get an ultrasound abdomen to check for foetal abnormality. The ultrasound confirmed the suspicion of polyhydramnios with hydrocephalus. She went into early labour and gave birth to a male infant via spontaneous vaginal delivery, weighing 1.81kg, and having an APGAR score of 2 at one minute. After that, the infant was intubated and taken to the neonatal intensive care unit (NICU). Upon delivery, there was presence of a large SCT arising from the sacral region about 15cm x 20cm as depicted in Figure 1 and 2. The tumour was fluctuant indicating cystic features. The anus was displaced cephalad and anteriorly. There was also bruising at the base of the tumour without any apparent bleed.

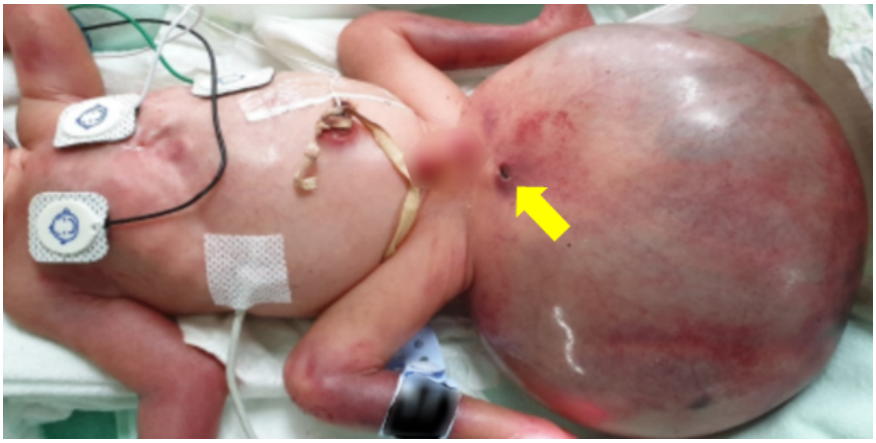

Fig. 1. Superior view of the SCT where a large tumour is seen with the anus displaced cephalad and anteriorly (indicated with yellow arrow)

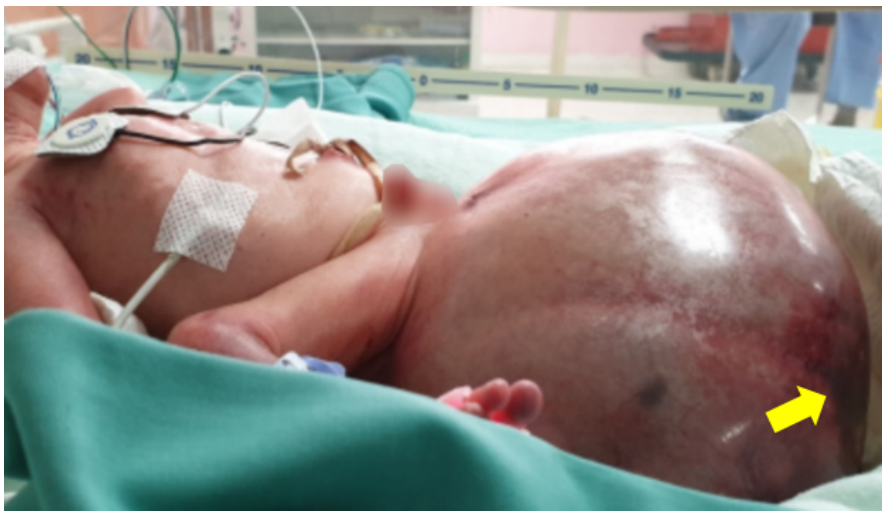

Fig. 2. Lateral view of the SCT where bruises were noted at the base of the tumour (indicated with yellow arrow)

2. Discussion

SCTs are a form of Germ Cell Tumour (GCT). The sacrococcygeal region is the most prevalent location for GCT development in newborns [2]. SCT is the most frequent extragonadal tumour of

infancy and is usually benign at this age. Despite their benign nature, SCTs are associated with significant morbidity and death. The gonads (ovaries and testes), the anterior mediastinum, the retroperitoneal space, the presacral and coccygeal regions, the pineal and other intracranial sites, the neck, and abdominal viscera are all common locations of teratomas [3]. Primordial Germ Cells (PGC) are needed for the physiologic differentiation and production of gametes. However, the PGCs are subject to form Type 1 GCT or Type 2 GCT along the process of germline differentiation via PGC migration and proliferation, sex specific differentiation, and embryonic differentiation [5,7]. Fig. 3 shows the germline fate under physiologic and pathologic condition. GCTs are formed during the pathologic pathway. Abbreviation: GCT: Germ Cell Tumour, PGC: Primordial Germ Cells. Image modified from[7].

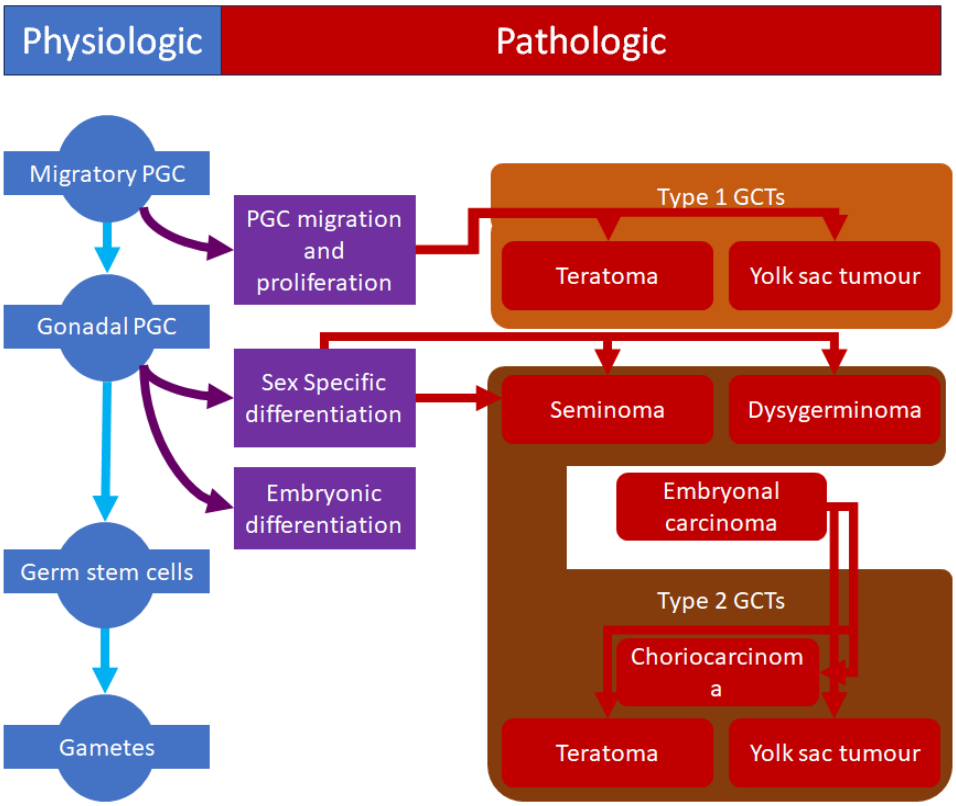

Fig. 3. Germline fate under physiologic and pathologic conditions. GCTs are formed during the pathologic pathway. *Abbreviation: GCT: Germ Cell Tumour, PGC: Primordial Germ Cells.* Image modified from [7]

Embryologically, SCTs are derived from the pluripotent cells in Hensen's node of the primitive streak, where it contains two or three germ cell layers [4]. In addition to the gonads, abnormally sequestered midline embryonic rests can also contain pluripotent cells[6]. Figure 4 shows the primordial germ migration from the yolk sac to the other locations of embryo. These areas are subject to GCT growth.

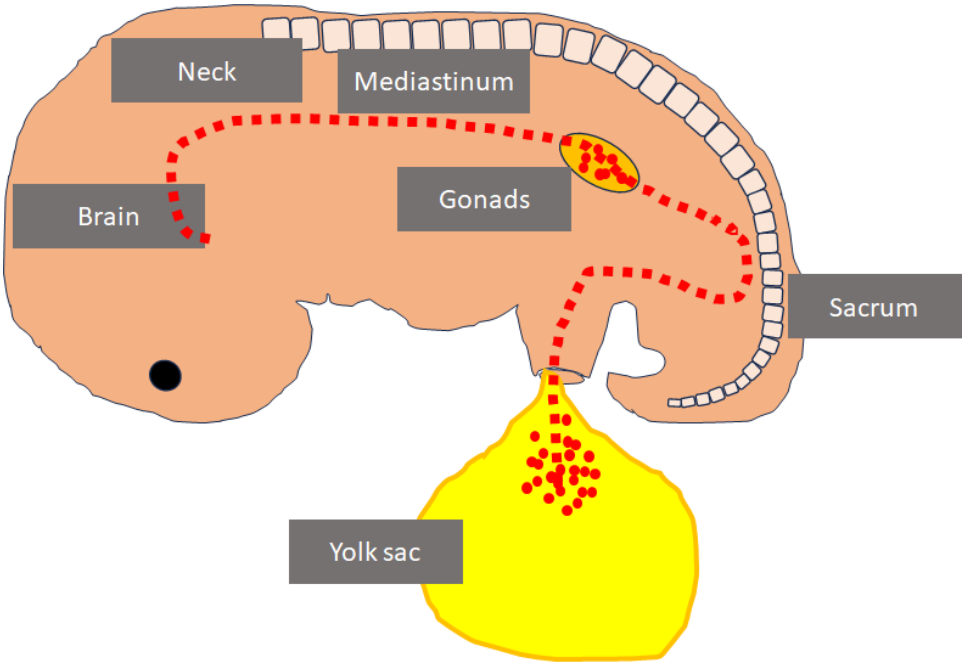

Fig. 4. The primordial germ cells migration

SCTs usually have a solid and a cystic component, with the solid component having a very substantial blood supply, which can be a major issue during pregnancy or after surgery [4]. The four types of SCTs are Type I, II, III, and IV [2]. Type I SCT represents around 45% of the cases. The rest of the percentages and its components are summarized in Table 1 [2]. Figure 5 shows the sacral component (light orange) and intrapelvic (reddish) components in relation to each SCT classification.

Table 1
Sacrococcygeal Teratoma Type, Incidence and Components

Sacrococcygeal Teratoma Type	Incidence Percentage (%)	Components
Sacrococcygeal Teratoma Type I	45	predominantly external with a minimal pre-sacral component.
Sacrococcygeal Teratoma Type II	35	intra-pelvic component
Sacrococcygeal Teratoma Type III	9	intra-pelvic and abdominal extension with a minimal external component,
Sacrococcygeal Teratoma Type IV	10	intra-pelvic component and abdominal extension

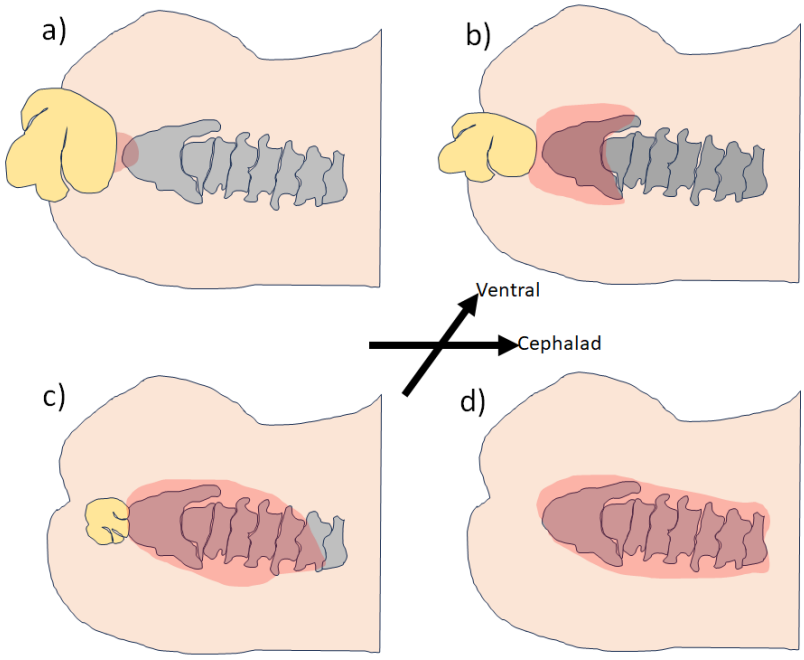

Fig. 5. SCT Type 1 – IV (a-d). Arrows indicate position and placement of the infant

Recognising the significance of a sacral mass in the differential diagnosis should be a top priority. Myelomeningocele, meningocele, lipoma, haemangioma, perineal cyst, lymphangioma, or rectal prolapse must be clearly identified, recognised, and distinguished. For in-utero cases, an MRI could be done to distinguish these instances [1]. In contrast, in ex-utero cases, clinical examinations including per rectal examination, ultrasound, and computed tomography (CT) scans may further aid and strengthen the diagnosis [8]. Figure 6 shows the differential diagnosis for sacrococcygeal malformations. These could be divided into benign and malignant in general or to more specific subgroups which are congenital, inflammatory, neurogenic, and others.

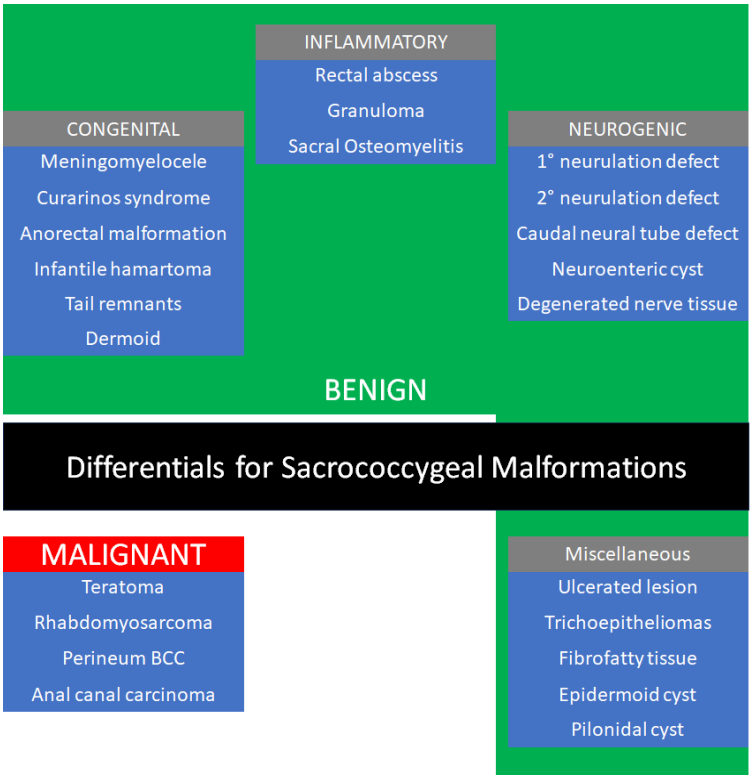

Fig. 6. Differential for Sacrococcygeal Malformations.
Abbreviations: 1°: Primary, 2°: Secondary, BCC: Basal Cell Carcinoma

Typically, infants with SCT have an excellent prognosis, depending on the timing of diagnosis, the malignant potential of the tumour, and the challenges expected in surgical excision [9]. The overall survival rate exceeds 95% [5]. This was similar to the data reported at a Malaysian tertiary centre [1]. In contrast, factors such as tumour size of more than 10 cm, Type III and IV SCTs, presence of solid tumours, and presentation after the second month of life enhances the chance of malignancy [9].

Prenatal diagnosis can be done early in this era with the advancement of technology. This is significant since early prenatal presentation is associated with high foetal morbidity and mortality [4]. Moreover, presentation after 30 weeks yields a better prognosis index in terms of foetal survival [4]. Another important reason for diagnosing SCT antenatally is for foetal monitoring during pregnancy. Large tumours can cause a shift in the blood away from the foetus due to their high blood flow demand [4]. As the tumour grows, this can result in high-output heart failure or hydropic foetus. Polyhydramnios, haemorrhage into the tumour, and preterm labour are all possible consequences [4]. Additionally, this could also impact the gravid physiology resulting in conditions such as water retention, placentomegally, pre-eclampsia and even cardiac failure [4]. Elsewhere, Zhang *et al.,* [12] reported elevation of alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 and carbohydrate antigen 72-4 (CA72-4) in a rare case of mucinous tumour arising from SCT.

The usage of ultrasonography for SCT detection could be carried out as early as the first trimester [4]. Another reason for detecting SCT early in the pregnancy is to arrange the mode of birth. Caesarean section is advised in SCTs larger than 10cm in diameter or highly vascular tumours after foetal lung maturity is achieved [9]. This is done to reduce the chance of tumour rupture, which may be fatal. The second alternative is post-natal surgical excision, which is the most used technique for SCT patients. The most significant principle in preventing recurrence is complete tumour removal

with coccygectomy [9]. However, detailed attention needs to be given to the possible intra-operative complications, such as massive blood loss and cardiac arrest secondary to electrolyte imbalance, as these are the most common causes leading to on-table deaths [9]. Although, the sacrum is the most utilised surgical approach, the sacro-abdominal surgery is indicated if the tumour is those from Type III or IV [9].

In this case that we reported earlier, the SCT pathology was missed during the prenatal period as it was misdiagnosed as hydrocephalus. We presume that this may be one the many factors that had led to poor anticipation of the condition and poor foetal outcome upon delivery. The treatment modalities include both in-utero and ex-utero options. These are ultrasound-guided percutaneous radiofrequency, laser ablation, percutaneous drainage, thermocoagulation, foetal resection, and ex-utero intrapartum treatment (EXIT) [10,11]. In addition, there is also a role of shunt placement namely vesicoamniotic shunt, percutaneous amniotic shunt and tumour-amniotic shunt [11]. Of note, the mode of surgery could also be determined after establishing the presence of cardiac insufficiency in the SCT patient [10]. In this case, the EXIT option had a better outcome [10].

Despite complete excision and regardless of whether the tumour is benign or malignant, the recurrence rate is about 1-22% during the first three years of life [9]. Close follow-up with digital rectal examination, tumour marker (serum alpha-fetoprotein), and imaging should be done for at least 3 years after surgery to look for recurrence [9]. Recurrence can be diagnosed in adulthood as late as 40 years following neonatal SCT resection, accounting for 6.7% of cases [9]. Malignant degeneration of a mature teratoma, insufficient resection or intra-operative tumour spillage or lack of coccygectomy and unapparent tiny foci of malignant cells in a reported histology of a mature teratoma were proposed reasons for this late recurrence [9]. As a result, monitoring should continue into adulthood.

Because the SCT lacks clear, unified recommendations, the combination of good physical examination, laboratory analysis, and imaging may aid in distinguishing different lesions to enable prompt and precise management [2]. Figure 7 is the proposed algorithm to manage SCT. These were clinically evaluated and modified from previous algorithms by two different authors [13,14].

Fig. 7. Proposed management algorithm for SCT. *Abbreviations: CS: Caesarean Section, EXIT: ex-utero intrapartum treatment, SCT: sacrococcygeal teratoma, US: ultrasound*

4. Conclusions

The prompt diagnosis and management of SCT begins antenatally, with a careful approach during intrapartum and a timely manner of surgical excision in the post-partum period. As a result, the ultimate hallmark in increasing the survival of SCT patients is a multidisciplinary approach of early diagnosis with anticipation of maternal and foetal problems and early complete surgical resection.

References

[1] Yeap, Badrul Hisham, and Zakaria Zahari. "Neonatal tumours in Malaysia: a call for heightened awareness." *Pediatric surgery international* 26 (2010): 207-212. https://doi.org/10.1007/s00383-009-2523-7

[2] Phi, Ji Hoon. "Sacrococcygeal teratoma: a tumor at the center of embryogenesis." *Journal of Korean Neurosurgical Society* 64, no. 3 (2021): 406-413. https://doi.org/10.3340/jkns.2021.0015

[3] Engel, Rainer M., Ronald C. Elkins, and Barry D. Fletcher. "Retroperitoneal teratoma. Review of the literature and presentation of an unusual case." *Cancer* 22, no. 5 (1968): 1068-1073. https://doi.org/10.1002/1097-0142(196811)22:5<1068::AID-CNCR2820220525>3.0.CO;2-3

[4] Girwalkar-Bagle, A., Thatte, W.S., Gulia, P., *Sacrococcygeal teratoma: A case report and review of literature*, vol. 18, n.d

[5] Lew, Chong-Zhi, Hsi-Che Liu, Jen-Yin Hou, Ting-Huan Huang, and Ting-Chi Yeh. "Pediatric Extracranial Germ Cell Tumors: Review of Clinics and Perspectives in Application of Autologous Stem Cell Transplantation." *Cancers* 15, no. 7 (2023): 1998. https://doi.org/10.3390/cancers15071998

[6] Monteiro, M., Teresa Margarida Cunha, A. Catarino, and V. Tomé. "Sacrococcygeal teratoma with malignant transformation in an adult female: CT and MRI findings." *The British journal of radiology* 75, no. 895 (2002): 620-623. https://doi.org/10.1259/bjr.75.895.750620

[7] Pierce, Joshua L., A. Lindsay Frazier, and James F. Amatruda. "Pediatric germ cell tumors: a developmental perspective." *Advances in urology* 2018, no. 1 (2018): 9059382. https://doi.org/10.1155/2018/9059382

[8] Agarwal, Akhilesh, Sukanta Das, Dipak Ghosh, and Anshu Agarwal. "Sacrococcygeal masses other than meningomyelocele." *Indian Journal of Surgery* 73 (2011): 206-209. https://doi.org/10.1007/s12262-010-0014-7

[9] Sinha, Shalini, Yogesh Kumar Sarin, and Vidyanand P. Deshpande. "Neonatal sacrococcygeal teratoma: our experience with 10 cases." *Journal of Neonatal Surgery* 2, no. 1 (2013). https://doi.org/10.47338/jns.v2.16

[10] Ding, Yunping, Mengmeng Yang, Min Lv, Ying Jiang, Tian Dong, Baihui Zhao, and Qiong Luo. "The ex-utero intrapartum treatment (EXIT) strategy for fetal giant sacrococcygeal teratoma with cardiac insufficiency: a case report and review of the literature." *Frontiers in Oncology* 12 (2022): 1035058. https://doi.org/10.3389/fonc.2022.1035058

[11] Hirose, Shinjiro, and Diana L. Farmer. "Fetal surgery for sacrococcygeal teratoma." *Clinics in perinatology* 30, no. 3 (2003): 493-506. https://doi.org/10.1016/S0095-5108(03)00059-9

[12] Zhang, Fengtian, Xiaolong Yu, Jin Zeng, and Min Dai. "Mucinous tumor arising in a giant sacrococcygeal teratoma: a rare case report." *Medicine* 96, no. 47 (2017): e8759. https://doi.org/10.1097/MD.0000000000008759

[13] Baumgarten, Heron D., Juliana S. Gebb, Nahla Khalek, Julie S. Moldenhauer, Mark P. Johnson, William H. Peranteau, Holly L. Hedrick, N. Scott Adzick, and Alan W. Flake. "Preemptive delivery and immediate resection for fetuses with high-risk sacrococcygeal teratomas." *Fetal diagnosis and therapy* 45, no. 3 (2019): 137-144. https://doi.org/10.1159/000487542

[14] Varlas, Valentin Nicolae, Eliza Maria Cloțea, Roxana Georgiana Varlas, Anca Pop, Ovidiu Peneș, Dragoș Crețoiu, Vlad Dima, and Laura Bălănescu. "Immature sacrococcygeal teratoma: a case report and extensive review of the literature." *Diagnostics* 14, no. 3 (2024): 246. https://doi.org/10.3390/diagnostics14030246