From Sacrum to Spine: A Complex Case of Sacrococcygeal Teratoma

Jennifer Young, MN, RN(EC) NP-Pediatrics, NNP-BC
Hazel Pleasants-Terashita, MN, RN(EC) NP-Pediatrics, NNP-BC

Continuing Nursing Education (CNE) Credit

The test questions are provided in this issue. The posttest and evaluation must be completed online. Details to complete the course are provided online at academyonline.org. Sign into your ANN account or register for a non-member account if you are not a member. A total of 1.5 contact hour(s) can be earned as CNE credit for reading this article, studying the content, and completing the online posttest and evaluation. To be successful, the learner must obtain a grade of at least 80 percent on the test. The test expires three (3) years from the publication date of this journal.

Disclosures: The authors, members of the activity planning committee, and the test panel have no relevant financial interests or affiliations with any commercial interests related to the subjects discussed within this article. No commercial support or sponsorship was provided for this educational activity. Neither ANN nor ANCC endorses any commercial products discussed/ displayed in conjunction with this educational activity.

The Academy of Neonatal Nursing is accredited with distinction as a provider of continuing nursing education by the American Nurses Credentialing Center's Commission on Accreditation. Provider, Academy of Neonatal Nursing, approved by the California Board of Registered Nursing, Provider #CEP 6261; and Florida Board of Nursing, Provider #FBN 3218, CE Broker # 50–3066.

The purpose of this article is to present a case study of a premature infant with a large sacrococcygeal teratoma. Background pathophysiology, etiology, and medical and nursing care are addressed.

Abstract

Neonatal tumors occur infrequently; sacrococcygeal teratoma (SCT) is a rare and abnormal mass often diagnosed on antenatal ultrasound. An SCT may cause serious antenatal complications, requires surgery in the neonatal period, and can lead to various long-term sequelae including fecal incontinence or constipation, urinary incontinence, and lower extremity mobility impairment. Even rarer are SCTs that include intraspinal extension necessitating complex neurosurgical intervention to relieve possible spinal cord compression or tumor tissue resection. A comprehensive understanding of the natural history of SCT provides frontline neonatal nurses and nurse practitioners with the expertise and language to support families during an infant's NICU admission. A glossary of key terms accompanied by a case review of a premature infant born with a large external SCT with intrapelvic and intraspinal components aids in enhancing knowledge related to the potential impact of an SCT on the central nervous system.

Keywords: neonate; sacrococcygeal teratoma; tumor; intraspinal; neurological complications; neurology; spinal innervation

Sacrococcygeal teratoma (SCT) is one of the most common tumors seen in neonates, growing antenatally from the coccyx of a developing fetus.[1–5] The tumor may grow in a posterior direction forming an exteriorized protrusion, or develop anteriorly distorting surrounding internal tissues or organs. SCTs are different than other tumors found in children because of their composition of tissue elements that are unrelated to the area from which they arise.[6] Teratomas vary in size, vascularity, and degree of mass effect upon nearby structures. An SCT creating mass effect displaces or compresses nearby structures which may include bladder, uterus, vagina, or rectum. All SCTs require resection early in life to reduce the risk of malignancy formation.[7,8]

PATHOGENESIS OF SCT

Embryology

The complexities of human development are fascinating as seen through the embryologic origin of SCT formation. While not fully understood, theory suggests SCTs are a rare form of germ cell tumor arising from abnormal migration of germ cells from the primitive yolk sac.[9,10] Rapid cell division occurs after egg fertilization and implantation. At the beginning of the third week after conception, the ongoing production of cells generates a structure known as the primitive streak which forms a thickened band of embryonic tissue, giving rise to the three germ cell layers: the ectoderm, mesoderm, and endoderm.[11] Further

Accepted for publication
January 28, 2020.

© 2020 Springer Publishing Company

http://dx.doi.org/10.1891/0730-0832.39.3.147

differentiation of germ cell layers occurs which establishes certain types of embryonic tissue[11]:

- Ectoderm: outer skin layer, hair, nails, teeth, certain glands, and the nervous system
- Mesoderm: connective tissue and muscle of viscera, bones, and the urogenital system
- Endoderm: epithelial lining of the gastrointestinal tract, respiratory system, and glandular cells of other organs including liver and pancreas

These primitive germ cell layers may become misplaced during cell migration, and lead to the development of a teratoma.[6] Other theories suggest that cells originating from Henson's node, also known as the primitive node and a part of the evolving primitive streak, give rise to teratoma formation.[1] Because Henson's node resides within the coccyx, this may account for the high number of teratomas that form in the sacrococcygeal region.[1,12] Additional processes occurring during the third postconceptional week include the evolution of the primitive streak in forming the neural tube and neural crest cells, which may be the source of SCT development and account for the recurrent link with spinal abnormalities.[9,11]

INCIDENCE

SCT tumors occur once in approximately 35,000 to 40,000 live births.[1,3,4,7,9,13,14] The incidence of SCT may be underrepresented when fetal demise and pregnancy interruption are included.[7] Approximately 75 to 80 percent of affected infants are female.[1,5,7,13,15] The majority of SCTs are benign tumors when diagnosed, however malignancy may ensue if not resected in the neonatal period.[7,16] While SCTs do not commonly have associations with other congenital malformations, there is potential for abnormal formation of the genitourinary system, anorectal structures, or bony spinal structures arising from the coccyx because of the mass effect from the growing tumor.[1,3] Intraspinal extension remains a rare finding, occurring in <5 percent of infants born with an SCT.[2]

DIAGNOSIS

SCTs are usually diagnosed antenatally, particularly if there is a large external component. Because the majority of SCTs are exophytic structures, many develop into a tumor that is easily detected with ultrasound imaging.[7,9] Fetal magnetic resonance imaging (MRI) allows caregivers to better understand both the characteristics of the tumor as well as its impact on surrounding structures, most commonly the rectum, bladder, or uterus.[1,17] Imaging findings identify heterogeneous lesions comprised of cystic components including fat, solid tumor that demonstrates evidence of tissue features from germ cell layers, or a combination of both cystic and solid components.[1] Other antenatal investigations that may be suggestive of an SCT include maternal biochemical screening results. Serum alpha-fetoprotein (AFP) is a protein made by the yolk sac and then the liver of the developing fetus.[18] Because AFP passes through the placenta and into maternal circulation, elevated maternal AFP levels may signal genetic disorders or fetal congenital malformations including SCT.[10,18]

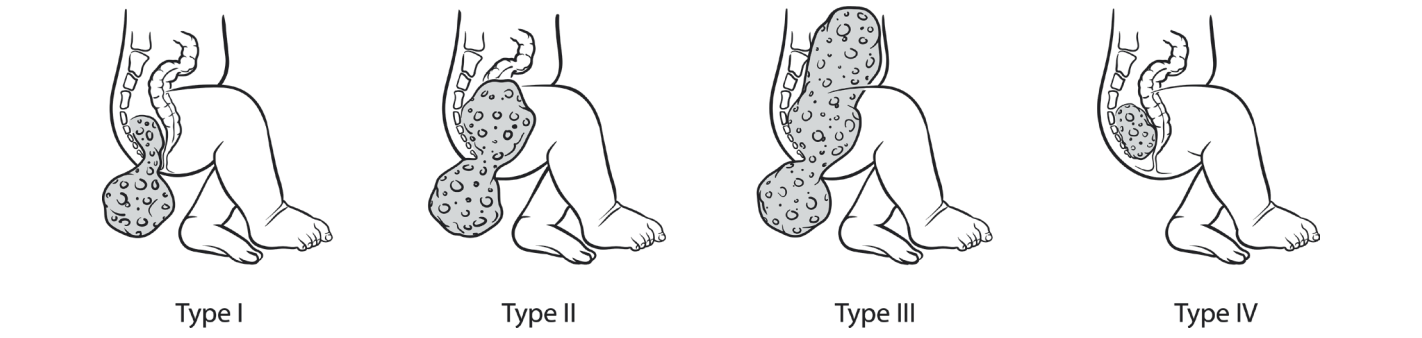
CLASSIFICATION

Altman[13] describes a grading classification of SCT compiled from data obtained through survey of the membership of the American Academy of Pediatrics Surgical Section (see Figure 1, Table 1). Based on the results of 405 cases of SCT occurring over a ten-year period, Altman's classification system reflects the degree of internal and external components of a teratoma and remains the gold standard for describing SCT.[13]

TABLE 1 ■ Altman Classifications of Sacroccocygeal Teratoma[13]

Type	Description
I	Predominantly external tumor with minimal presacral component
II	External tumor with significant intrapelvic extension
III	Some external component but predominant pelvic mass and extending into abdomen
IV	Presacral tumor only; no external component visible

FIGURE 1 ■ Altman classifications of sacrococcygeal teratoma.



Source: Courtesy of The Hospital for Sick Children, Toronto, Ontario.

TABLE 2 ■ Pathology Characteristics of Sacrococcygeal Teratoma[19]

Tumor Tissue Grade	Description
0	Mature well-differentiated tissue; may include skin, muscle, bone
1	Rare foci of immature or embryonal tissue
2	Moderate amounts of immature elements
3	Large amounts of immature tissue elements; may contain malignant cells (e.g., yolk sac components)

After surgical resection, tissue pathology analysis provides further information through histological grading of the teratoma. Valdiserri and Yunis[19] characterize tumors according to the maturity of the tissue elements evident in the resected teratoma sample (Table 2).

Approximately 75 to 90 percent of SCTs reveal benign pathology, including mature and immature tissue histology.[1,13,19] However, both immature and mature teratomas may undergo malignant transformation if not removed.[1] Neither tumor size or pathology findings independently determine a neonate's prognosis as multiple factors may influence pregnancy and neonatal courses.[8,10]

ANTENATAL COMPLICATIONS

Close monitoring throughout pregnancy is essential; the pregnant woman carrying a fetus diagnosed with SCT requires the care of a high-risk multidisciplinary perinatal team. As pregnancy progresses, the developing fetus may demonstrate high cardiac output failure resulting from the metabolic demands created by arteriovenous shunting within the highly vascularized tumor.[1,20] A teratoma creates a "vascular steal" from the fetus, reducing systemic resistance. The fetal heart compensates by increasing cardiac output to maintain essential blood flow to all tissues. Fetal anemia resulting from hemorrhage within the tumor may exacerbate the potential for high cardiac output failure.[1] Anemia and cardiac failure can lead to the development of hydrops fetalis. Regular surveillance through serial fetal echocardiography to evaluate cardiac function combined with frequent ultrasonography to assess tumor growth, amniotic fluid index, and fetal well-being provides important data in monitoring the development of hydrops fetalis.[20] If a fetus develops hydrops, the risk of intrauterine demise increases.[10] Severe hydrops may require urgent premature delivery of the fetus or consideration of alternative fetal therapies.

Fetal Interventions

Fetal interventions are not indicated for small or cystic teratomas, but instead are reserved for complications of fetal high cardiac output failure and hydrops.[21] While antenatal strategies to reduce blood flow to the tumor have been attempted, success rates remain limited and the complication rate high.[20,21] Open fetal surgery to debulk the teratoma may allow for fetal cardiovascular recovery prior to birth although premature labor may result. Minimally invasive approaches include radiofrequency or laser ablation, which are techniques that occlude blood vessels supplying the tumor. Hemorrhagic complications may occur as the tumor mass itself may also be "burned" in the process.[20] Amnioreduction may be helpful in preventing preterm delivery by decreasing uterine muscle stress caused by polyhydramnios. Perhaps the most important role of the antenatal multidisciplinary team, aside from close monitoring of both the pregnant woman and fetus, is to prepare for a coordinated delivery to avoid the risk of hemorrhagic complications at the time of birth.[7,20]

NEONATAL MANAGEMENT

Initial Stabilization

Vaginal delivery carries a risk of dystocia, tumor rupture, and ensuing hemorrhagic shock; controlled cesarean section delivery is recommended for external tumor size exceeding 5 cm in diameter to avoid dystocia.[8,12] Immediate neonatal resuscitation focuses on hemodynamic stability and ensuring the airway is protected and the cardiovascular status is satisfactory. Emergency treatment measures including volume replacement, cardiotropic support, and urgent debulking of the SCT may be required to control bleeding if severe hemorrhagic shock occurs. After initial stabilization, the tumor requires protection until a pediatric surgeon performs an assessment and collaboratively establishes a surgical plan with the neonatal healthcare team. Most SCTs are covered in skin; however, if ablation techniques were performed in utero, skin necrosis may have ensued. Nonadherent dressings may be applied as required to ensure no breach in the tumor skin occurs that may precipitate bleeding before surgical repair.[8] Transition of the newborn to extrauterine life and completion of diagnostic imaging prior to the surgical repair contributes to greater intraoperative stability. Assessment of neonatal serum AFP levels provides baseline data as a tumor marker, although it may be difficult to interpret as the AFP level is naturally elevated at birth.[18] Surgical excision of the teratoma usually occurs in the first week of life. Infants with a malignant SCT may undergo chemotherapy and radiation treatment in addition to surgical intervention.[3]

Surgical Repair

The goal of operative repair of an SCT is to remove the entire tumor with clear margins and no spillage of tumor contents during the procedure. The intricate dissection may cause the procedure to last several hours as the surgical team carefully removes the tumor in effort to minimize injury to any surrounding organs or structures. Most Type I and Type IV SCTs undergo resection through a posterior surgical approach as the tumor can usually be completely excised through one surgical site at the infant's lower back. A Type II or Type III SCT, with significant intrapelvic or abdominal extension, may require both a laparotomy incision to mobilize the superior part of the tumor along with a posterior approach to complete removal.[3] The coccyx must be removed; coccygectomy reduces the risk for tumor

recurrence as nests of cancerous cells are commonly found in or adjacent to the coccyx.[1,3,5,7,8,22] Multiple subspecialty surgical teams work together to ensure an intricate and complete excision occurs, often including pediatric general surgeons, neurosurgeons, and plastic surgeons.

Postoperative Nursing Care

Because of the operative approach required to fully excise an SCT, the incision may be significant in size and suture count. To avoid pressure on healing tissues, neonates are commonly nursed prone in the early postoperative period, often for up to a week postprocedure. Pediatric surgeons may collaborate with plastic surgeons for the final stage of the surgical repair in an attempt to preserve normal gluteal folds and skin coverage as much as possible. In some cases, a drain may be left in situ to collect any fluid that may pool in potential space under the skin closure. Prevention of infection is vital. Antimicrobial therapy and wound care, which may include local bacteriocide dressings, are individualized according to the complexity of the surgical resection. Insertion and maintenance of an indwelling urinary catheter reduces soiling of the incision with urine that can be harmful to healing tissue. Strategic positioning of a diaper and meticulous removal of stool is required so as not to disrupt healing tissue or stitches. The use of a plastic drape secured above the anus but below the lesion to isolate fecal material from the surgical site may be necessary. Neonatal nurses and occupational therapists play important roles in helping parents care for their baby in the postoperative period when conventional holding, feeding, and diapering might not occur. As healing progresses, discharge planning includes assessment for positioning in a car seat.

CASE STUDY

The following case study illustrates antenatal complications, premature birth, and complex surgical interventions for an infant born with an SCT which contributed to a protracted NICU admission. Highly specialized perinatal and postnatal multidisciplinary care, in collaboration with family members, led to successful discharge home.

Antenatal History

Malaki is a male infant born at 29 1/7 weeks gestation, birth weight 2,500 g, to a 35-year-old mother. Her antenatal ultrasound completed at 18 weeks' gestation revealed a sacral mass; fetal MRI at 24 weeks' gestation confirmed a large SCT with intraspinal extension and associated spinal scoliosis. Features of fetal high output cardiac failure were also noted, including hepatomegaly and placental edema, necessitating referral to a high-risk obstetrical unit with fetal medicine subspecialty. Fetal echocardiogram demonstrated mild to moderately reduced right ventricular function. In order to reduce the strain on Malaki's heart, fetal laser ablation of the arteries feeding the teratoma occurred at 25 and 27 weeks' gestation. Fetal blood transfusion was provided; amnioreduction for ongoing evidence of polyhydramnios also occurred.

Course at Delivery

The birth of an infant with a large SCT requires elaborate preparation to ensure vital personnel and equipment are present. Malaki's mother received care in a high-risk perinatal center and anticipatory birth plans included a cesarean section delivery by a fetal-medicine specialist and full perinatal team including neonatology service to be present. Antenatal referral to pediatric general surgery allows for further preparation planning to incorporate their presence at time of delivery in case the tumor requires immediate attention. At 29 1/7 weeks, Malaki required urgent delivery by emergency cesarean section because of absent fetal movement and poor biophysical profile scoring. Apgar scores were 1, 3, and 7 at one, five, and ten minutes respectively; cord blood gases revealed significant acidosis (pH 6.8/CO_2 95 mmHg/HCO_3 10 mEq/L/base deficit -18). Malaki's grave condition at birth commanded extensive resuscitation efforts including intubation and ventilation by high-frequency oscillation, inotropic support, fluid resuscitation including blood product, and sodium bicarbonate administration. Emergency tumor debulking occurred following the arrival of a pediatric surgeon who was urgently called to the resuscitation room. Malaki subsequently transferred to a tertiary/quaternary NICU for ongoing management.

Postnatal Course

Aside from his large SCT, Malaki was observed to be nondysmorphic and without evidence of other congenital anomalies. Malaki's extensive SCT, comprised of external, intrapelvic, and intraspinal components, ruptured shortly after birth causing him to experience critical hypovolemic shock and hemodynamic instability with ensuing consequences of pulmonary hypertension and renal failure secondary to acute tubular necrosis. Malaki's significant ascites at birth was attributed to his rapidly evolving hydropic state prior to delivery. Ongoing complex rescue care during Malaki's initial days included peritoneal drain insertion to relieve the intra-abdominal pressure created by the ascites, inhaled nitric oxide and ongoing high-frequency oscillation ventilation, multiple cardiotropic agents, and frequent blood product administration. After several days, Malaki's condition stabilized and he was able to go for surgery. Malaki required four complex operative interventions over his NICU admission to resect his teratoma. Retrospectively, Malaki's teratoma was estimated to weigh 1.4 kg, representing approximately 55% of his birth weight (Figure 2).

Resection #1. On day of life (DOL) 4, Malaki had a partial excision of the external component of his SCT. He experienced profound blood loss intraoperatively, required cardiac compressions and pressor administration while surgical intervention to stem further blood loss ensued. Tumor pathology of his excised tissue identified an immature teratoma without elements indicative of malignancy.

Despite the complex course during his first weeks of life, Malaki was able to successfully extubate to noninvasive

FIGURE 2 ■ Large highly vascular exophytic sacrococcygeal teratoma (SCT).

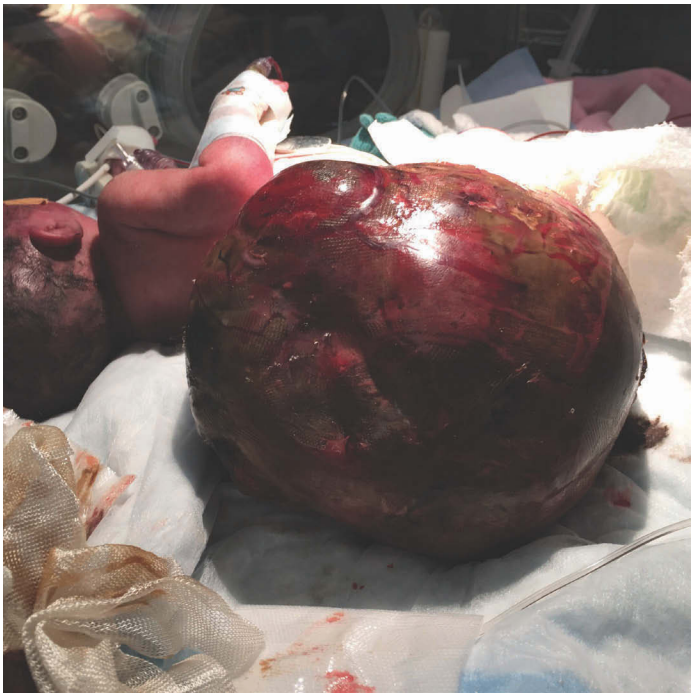
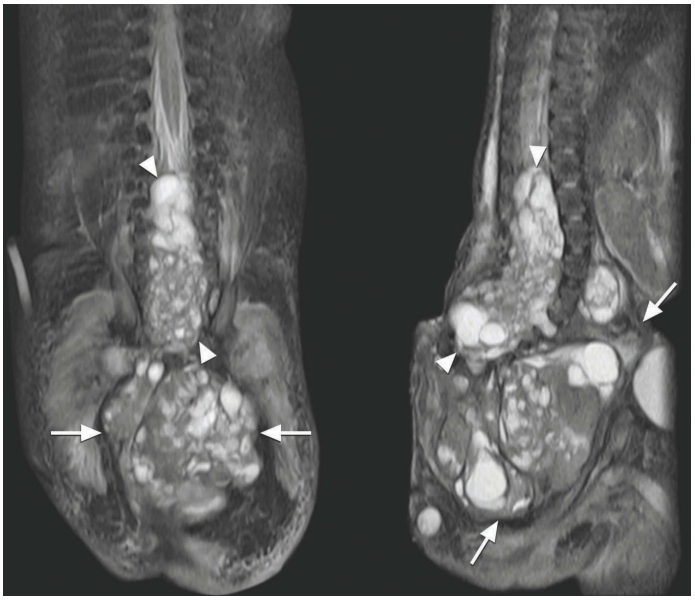


FIGURE 3 ■ Mixed cystic-solid sacrococcygeal teratoma (SCT). Arrows point to exophytic intrapelvic component. Arrowheads note intraspinal component.



respiratory support on DOL 21 (postmenstrual age [PMA] 32 1/7 weeks) and required only low flow oxygen shortly after. Progression in enteral feeding gradually occurred. Brain imaging completed in the first two weeks of life demonstrated bilateral intraventricular hemorrhages and increased periventricular white matter echogenicity. Several weeks later, in preparation for further teratoma resection, an MRI revealed the large pelvic mass extending into the spinal canal through multiple lumbosacral neural foramina with expansion of the spinal canal and splaying of the cauda equina noted, representing possible spinal cord compression (Figure 3). Displacement of the rectum and urethra was also demonstrated; Malaki required cautious urinary catheterization as his penis was tortuous from birth, attributed to the in utero mass effect of his SCT. Throughout this time period, Malaki was noted to have oozing passage of stool without evidence of an anal wink. He appeared to have some spontaneous movement at his hip and knee but no ankle movements were noted.

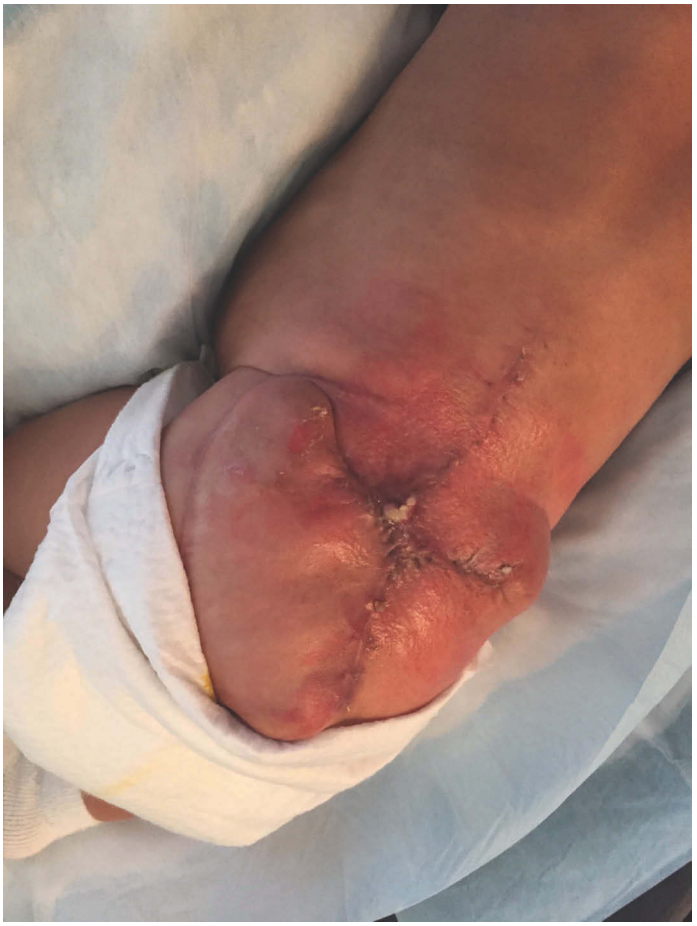
Resection #2. On DOL 34 (PMA 34 weeks), Malaki had further partial resection of the pelvic mass; however, residual intraspinal tumor tissue remained. Tumor pathology revealed an immature teratoma with tumor cells present at the outer margins of the tissue resected. The presence of tumor cells necessitate ongoing monitoring of AFP levels to gauge potential tumor transformation.

Over the following weeks, Malaki's SCT fungated, becoming a sizeable external mass. An MRI completed on DOL 68 (PMA 38 6/7 weeks) revealed an interval increase in the intraspinal element, including a large component extruding from the distal spinal canal. During Malaki's NICU admission, AFP levels were followed approximately every 2 weeks and remained elevated throughout. Although it is not uncommon for AFP to be elevated in newborn infants, Malaki also had residual tumor present. The focus of evaluation of AFP is to note changes in trend; a significant rise in Malaki's AFP coincided with tumor growth (peak value 2,743 mcg/mL).

Resection #3. On DOL 105 (PMA 44 1/7 weeks), Malaki underwent complex and collaborative surgical intervention including excision of the external aspect of his SCT, multilevel laminectomies starting at L1 with partial resection of intraspinal tumor components, as well as coccyx resection (Figures 4 and 5). Malaki's tumor pathology analysis revealed mixed histology with persistence of an immature teratoma and evidence of various tissue types including viscera, bone, muscle, and nerves. No malignancy was identifiable. Future implications of laminectomy surgery include possible progressive spinal abnormalities such as scoliosis or kyphosis.[23]

Resection #4. Magnetic resonance imaging continued to reveal the remaining intraspinal component of Malaki's SCT within the presacral space from below the L1 level and extending through multiple lumbosacral neural foramina, with the largest element extending through the L5/S1 neural foramen (Figure 6). Alpha-fetoprotein levels continued to increase. On DOL 140 (PMA 49 1/7 weeks), Malaki was taken to the operating theater for partial resection of his remaining intra-abdominal tumor and intraspinal tumor below S1. Residual intraspinal tumor was left in situ because

FIGURE 4 ■ Postoperative wound after partial excision of complex sacrococcygeal teratoma.



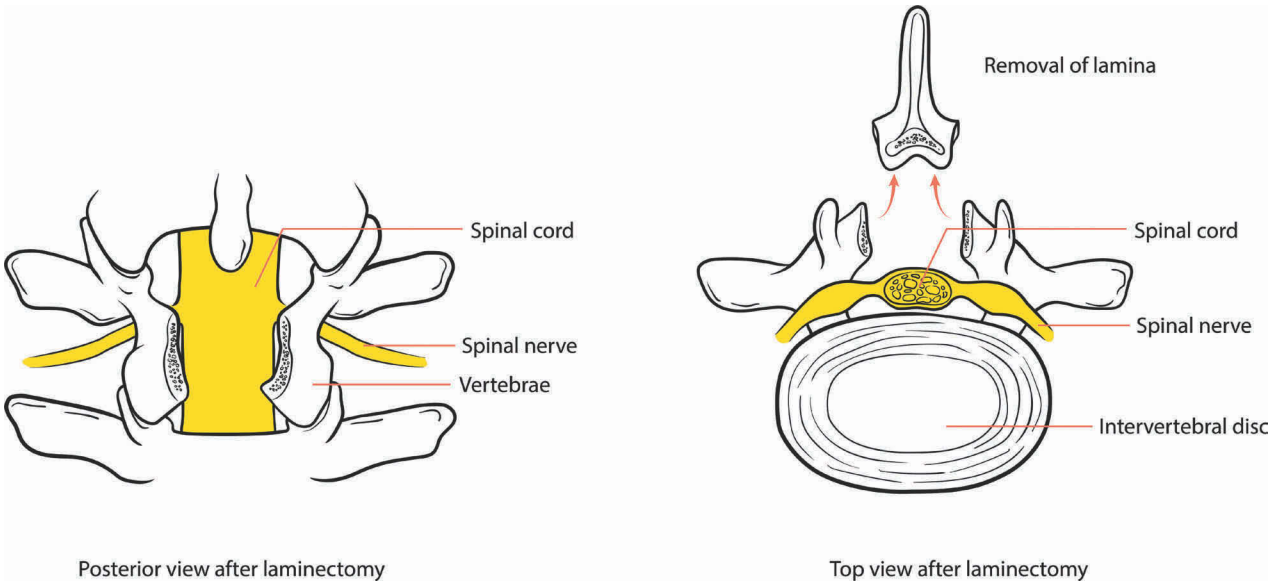
of the difficulty in access given its position within the neural foramina. Mature teratoma pathology was reported without evidence of malignancy.

Malaki required meticulous wound care management following each operation which was optimized by collaboration between plastic surgeons, wound care specialists, neonatal nurse practitioners, and registered nurses. Specialized antimicrobial dressings containing either chlorhexidine acetate or nanocystalline silver were used, depending on the progress of each surgical wound healing, which also assisted in preventing common wound pathogen proliferation. Prophylactic postoperative antibiotics were provided but not vigilantly discontinued as would be encouraged in the current era of antibiotic stewardship.

Preparation for Discharge

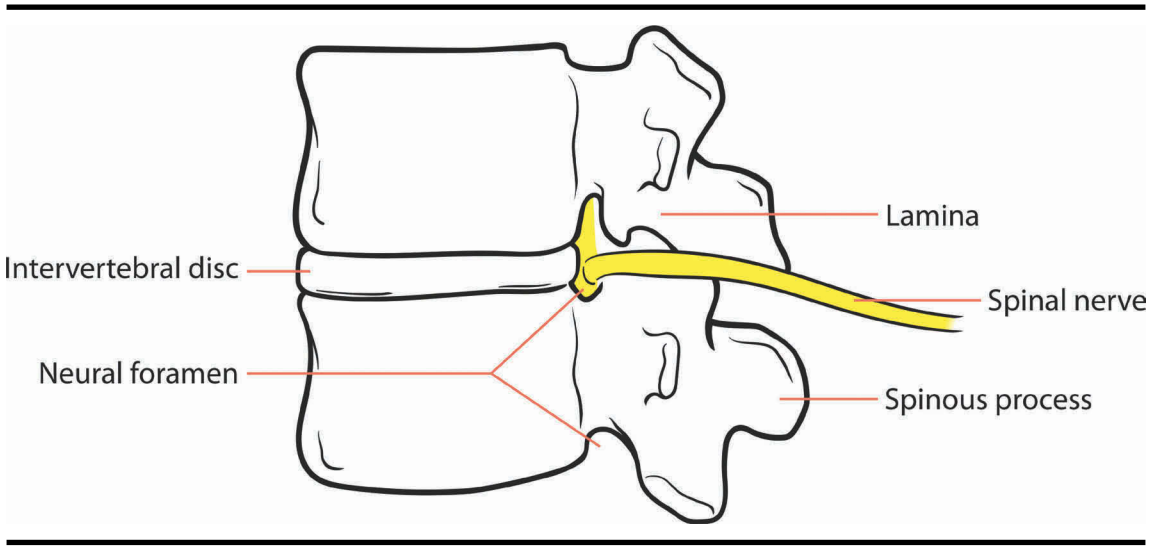
At almost six months chronological age, or approximately three months corrected age, Malaki graduated home from the NICU on ad lib oral feeding and receiving daily dressing changes for his surgical wound. Remarkably, he was alert, interactive, and responsive to his environment. Brain imaging completed at term revealed grossly normal brain parenchyma with mild white matter volume loss; head circumference growth had steadily progressed but plateaued in the weeks preceding discharge. Malaki demonstrated normal tone and smooth spontaneous movements of his upper extremities. Spontaneous movements were reduced in his lower extremities; some hip movements and knee extension were present while hip flexion and ankle movement were absent. Abnormal plantar reflexes were demonstrated. Reduced anal tone and absence of anal wink persisted. A weak urine stream was noted. A voiding cystourethrogram did not show evidence of

FIGURE 5 ■ Laminectomy.



Source: Courtesy of The Hospital for Sick Children, Toronto, Ontario.

FIGURE 6 ■ Vertebrae and neural foramen.



Source: Courtesy of The Hospital for Sick Children, Toronto, Ontario.

urinary reflux; renal ultrasonography demonstrated normal parenchyma without evidence of hydronephrosis.

Anticipatory education for Malaki's parents included a discussion about the likelihood of Malaki experiencing a neurogenic bladder and bowel dysfunction. Concerns for Malaki's future ambulation were also identified. Supportive, community-based, outpatient services included occupational therapy and physiotherapy to aid in optimizing Malaki's development. Subspecialty follow-up included ongoing assessment by pediatric surgery and neurosurgery for evaluation of Malaki's remaining tumor; hepatology follow-up services were also included as Malaki had developed hepatic fibrosis with findings suggestive of portal hypertension. Antenatal red blood cell transfusions introduced through the hepatic circulation, coupled with protracted administration of total parenteral nutrition and the initial liver insult resulting from hypovolemia injury at birth had contributed to progressive hepatic cirrhosis. His residual intraspinal tumor initially required AFP analysis every two weeks with interval increases as it declined. Additional surveillance included an annual MRI to monitor for potential tumor growth.

NEUROLOGICAL IMPLICATIONS OF SCT

Understanding the structural and functional organization of the central and peripheral nervous system, including nerve innervation, provides foundational knowledge for healthcare professionals in understanding the impact of an SCT and, in particular, the nature of Malaki's complex teratoma. The central nervous system, comprised of the brain and spinal cord, is the overall system that controls and coordinates various functions of the body. The brain is surrounded by the skull; the spinal cord is surrounded by rings of bony vertebrae and protective membranes known as the meninges.[24] Together the vertebrae and the meninges form the spinal column. The spinal column begins at the base of the skull and ends at the coccyx, elongating more rapidly than the spinal cord during the later stages of fetal life.[25]

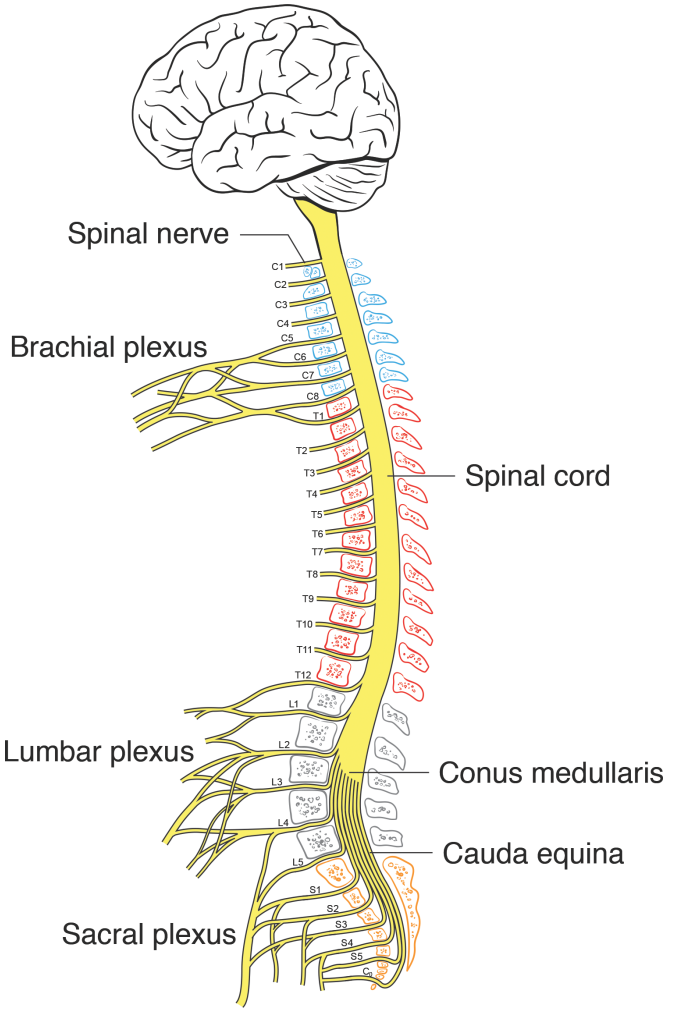
The spinal cord is a cylinder of nervous tissue that extends from the base of the brain to between the second and third lumbar vertebrae at term gestation.[25] The final resting position of the caudal end of the spinal cord in adulthood may range from the twelfth thoracic vertebrae to the third lumbar vertebrae.[25] The spinal cord functions as the communication system between the brain and various parts of the body by carrying messages that influence movement and provide sensation. Spinal nerves, part of the peripheral nervous system, leave the spinal cord through openings in the vertebrae called neural foramina (Figure 6).[25] As noted in the case study, Malaki's SCT extended through multiple foramina, creating considerable surgical dilemma. Spinal nerve roots branch off the spinal cord in pairs, one going to each side of the body. The spinal neurons carry messages to and from the spinal cord. Each nerve has a specific job for movement and sensation in various parts of the body. When the spinal cord develops abnormally or experiences compression or injury, sensation and/or movement can be interrupted resulting in a temporary or permanent loss of function and paralysis.[26] The higher on the spinal cord a congenital anomaly or compression occurs, the greater the loss of function the infant may experience.[27]

Vertebrae are grouped into sections based on their location: cervical, thoracic, lumbar, and sacral. The spinal nerves are designated according to where the nerve root emerges from the vertebral column. There are 31 pairs of spinal nerves: eight pairs of cervical nerves (C1–C8) emerge from the vertebral column in the neck region, 12 pairs of thoracic nerves (T1–T12) exit in the chest region, five pairs of lumbar nerves (L1–L5) arise from the lower back area, five pairs of sacral nerves (S1–S5) exit from the tailbone region, and a single coccygeal nerve that emerges from the tip of

the coccyx.[24] Spinal nerves appear along the entire vertebral column, however the spinal cord only extends to the lumbar region in a term neonate; the caudal end of the spinal cord is referred to as the conus medullaris.[11] The nerve bundle existing in the lower part of the spinal column resembles a horse's tail, known as the cauda equine.[11] Nerve fibers from different spinal nerves recombine, creating networks called nerve plexuses.[24] Four nerve plexuses innervate distinct parts of the body: cervical plexus (C1–5), brachial plexus (C5–T1), lumbar plexus (T12–L4), and sacral plexus (L4–S4).[24] Nerve plexuses contain both sensory and motor fibers (Figure 7).[24]

Each pair of spinal nerves monitors a specific region of the body surface known as a dermatome; damage to a spinal nerve produces a characteristic loss of sensation in the corresponding region of the skin.[24] Reflexes are automatic, rapid, involuntary responses to stimuli and test the integrity of the CNS.[28] Absent, weak, or abnormal responses may demonstrate the location of a potential insult to the spinal cord.[26]

FIGURE 7 ■ Spinal cord and nerve plexuses.



Source: Courtesy of The Hospital for Sick Children, Toronto, Ontario.

Examples of reflexes in neonates include stretch reflexes (e.g., patellar reflex) and withdrawal reflexes, as noted by touching the sole of the foot with a sharp point which provokes flexion of the stimulated extremity and extension of the opposite extremity.[24,29]

Tumor mass effect, direct pressure of the SCT, and/or surgical trauma to nerves and nerve plexuses can contribute to long-term functional sequelae that correlate with the level of spinal cord involved (Table 3).[12,14,30] Evaluation of the neonate with an SCT requires particular attention be paid to the motor, sensory, and sphincter responses to stimulation in the areas of spinal nerve innervation associated with the location of the SCT. Conducting a thorough examination includes observation for spontaneous movement of extremities at rest and when stimulated. Assessment of tone and reflexes including the hip, upper and lower leg, and ankle assists in determining possible corresponding spinal nerve insufficiency and subsequent functional outcome deficit. The anocutaneous reflex, also known as "anal wink," is evaluated by providing cutaneous stimulation to the perianal skin.[29] A normal response is contraction of the external sphincter.[28] Assessment for bladder sphincter function is more challenging in neonates; bladder distention or constant dribbling may signify a neurogenic bladder[29] (Table 4).

COMPLICATIONS ARISING FROM SCT

Morbidity and mortality associated with SCT are impacted by premature birth and prenatal complications including high cardiac output failure, tumor hemorrhage, and hydrops fetalis.[3,20] Survival rates for infants born with benign SCT remain high, ranging from 80 to 95 percent.[5,30] Predictions regarding long-term sequelae vary internationally.[3,5,15,20,30] Bladder and bowel dysfunction may be caused by tumor mass effect or surgical trauma.[3,5,14,30] Urological complications from SCT include uncontrolled voiding and difficulties in bladder emptying as well as evidence of renal injury involving hydronephrosis and vesicoureteral reflux.[14,15] Formal urodynamic studies assist in diagnosing neurogenic bladder but are not feasible in the neonatal period. Routine assessment for clinical symptoms of a distended bladder or urinary dribbling is necessary. Strategies to reduce the potential development of renal injury may include clean intermittent catheterization and anticholinergic medication for infants and children diagnosed with neurogenic bladder.[15,31] Bowel dysfunction problems reported in children born with SCT include constipation and soiling.[14] Protective aids may be required beyond infancy.

The social stigma of bladder and bowel accidents may contribute to self-esteem issues, isolation, and interfere with school performance.[31] Timed voiding, adequate hydration, pelvic floor training, and structured bowel regimens may provide assistance in overcoming milder forms of bladder and bowel dysfunction for older children.[31] Ongoing supportive counseling may assist in providing education and additional coping strategies. There is a paucity of literature exploring

TABLE 3 ■ Nerve Plexuses and Neonatal Functional Sequelae if Injured[23,25,26,28]

Nerve Plexus	Major Nerve(s)	Distribution	Assessment (Primitive, Deep Tendon or Sphincter Reflex; Range of Motion)	Functional Sequelae
Cervical (C1–5) Innervates muscles of neck and diaphragm	Phrenic	Diaphragm		Respiratory insufficiency
Brachial (C5–T1) Innervates pectoris and upper limbs	Axillary Median Radial Ulnar	Shoulder, arm, forearm, wrist, hand, digits	Moro Palmar grasp Antecubital fossa tap	Asymmetrical shoulder, arm, and/or hand movement
Lumbosacral Innervates pelvis and lower limbs				
Lumbar (T12–L4)	Femoral Obturator Saphenous	Hip, knee	Patellar tap Hip flexion, adduction, external rotation	Deficient/assisted ambulation likely
Sacral (L4–S4)	Gluteal Sciatic	Hip, knee, ankle	Plantar reflex Ankle tap Anocutaneous/anal wink	Independent ambulation preserved Constipation, soiling, bladder distention, urine leaks

TABLE 4 ■ Glossary of Terms

Term	Explanation
Amnioreduction	Intervention for polyhydramnios; needle aspiration of amniotic fluid under ultrasound guidance
Caudal	Latin term for tail; along the axis of the central nervous system at the inferior end
Debulk	Surgically reduce external tumor mass
Dermatome	A region of skin supplied by a specific spinal nerve that perceives sensations (e.g., pain, temperature, pressure)
Exophytic	Growing outward from point of origin
Fungate	Rapidly growing lesion with irregular borders and/or shape
Innervate	Supply an organ or body part with nerves
Laminectomy	Surgical technique; removal of the bony arch or the "roof" of the posterior portion of a vertebral bone
Laser ablation	Surgical intervention completed by inserting a small scope into the amniotic sac via a small abdominal incision in the pregnant woman and "sealing" blood vessels using laser energy
Mass effect	Effect of a growing mass pushing on or displacing surrounding tissue or organs
Neural foramen	Opening between vertebral bodies through which spinal nerves exit on each side
Presacral	Presacral space is inside the pelvis, behind the rectum, and in front of the coccyx and sacrum; normally it is empty

cosmetic outcomes after surgery for SCT. Unsatisfactory cosmesis may include dimples, prominences, keloids, or disfiguring scars that can lead to reduced mobility of the buttock region.[32] Corrective cosmetic surgery may be sought by survivors, however previous follow-up studies focused on functional outcomes have not reported on this aspect of postoperative experience.

SURVEILLANCE FOR TUMOR RECURRENCE

While positive functional outcomes are critical to the overall success of an SCT excision, vigilant monitoring for recurrence of a teratoma remains a priority for the first five years following surgery. Dependable indicators predicting later malignancy when initial teratomas are benign do not exist.[30] As previously discussed, removal of the coccyx is vital in the effort to remove residual tumor cells and reduce the risk of recurrence. Other factors that may influence the risk of relapse include spillage of tumor cells during excision, inability to completely remove the entire tumor, and the presence of immature histology.[1,14] Follow-up care for children born with SCT is recommended every three to six months with the surgeon for the first one to two years and then annually until school age. Assessments will include a digital rectal exam and serum AFP measurement. The rectal exam allows the surgeon to evaluate for any changes near the sacrum through assessment of the posterior wall of the rectum as this is where a tumor recurs most frequently. Serial monitoring of AFP provides a trend over time; an elevation from baseline may trigger diagnostic imaging to aid in determining tumor recurrence.

CONCLUSION

Malaki is four years old and attends public school and is supported by educational assistants in the classroom. He easily integrates with his peers through their shared enjoyment

FIGURE 8 ■ Malaki, age four on customized bicycle.

of sensory applications offered at the water table and sand box. Malaki demonstrates early proficiency at letter and word recognition. Despite having multiple laminectomy surgeries in the newborn period, Malaki does not currently show evidence of spinal curvature abnormality. He crawls and shuffles, while holding on to objects when upright. Malaki benefits from ankle foot orthoses to stabilize his ankles and protect his feet. He takes great pleasure in pedaling his customized bicycle and uses a wheelchair to assist in his mobility, both of which positively enhance his independence. Malaki also uses protective aids to manage bladder and bowel needs. Malaki continues to have residual tumor anterior to the sacrum that extends through the first and second sacral neural foramina bilaterally and posteriorly into the left-sided soft tissues. Annual MRI surveillance confirms it has not grown significantly and AFP levels do not signal potential malignant transformation. Surgical intervention would likely require resection of the superior sacrum including nerve roots, resulting in permanent sequelae and therefore expectant management is currently employed.

Neonates born with an SCT survive complex surgeries, some experiencing neurological insufficiencies similar to Malaki. Regular surveillance for tumor recurrence creates a burden of uncertainty for families. Lifestyle adaptations may be necessary to accommodate activities of daily living. Integration into educational systems may require additional supports to facilitate elimination and other needs for young students. Malaki serves as an inspiring example of a baby born with a giant SCT with external, intrapelvic, and intraspinal components who experienced numerous prenatal and postnatal complications. His complex health care needs continue into early childhood, requiring assistive devices to facilitate mobility and protective aids to manage elimination, as well as ongoing monitoring for tumor transformation. Malaki's initial comprehensive multidisciplinary neonatal care, coupled with his resilience, early community-based interventions, and ongoing strong family support allows him to thrive in his classroom—making new friends, positively and fully engaged in learning and, in his mother's words, "living his best life" (Figure 8).

ACKNOWLEDGMENTS

The authors would like to thank their colleagues Drs A. Daneman, S. Himidan, and J. Langer for their assistance and support whilst preparing this manuscript. They would also like to thank their nurse practitioner colleagues N. de Silva and S. Bernardo for their expertise. Most importantly they wish to acknowledge Baby Malaki and his family for inspiring them while they cared for him in the NICU, and for allowing them to share his story in an effort to advance the knowledge of health care professionals.

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate consent forms. The substitute decision maker has provided consent for images and other clinical information, including first name disclosure, to be reported in this journal.

REFERENCES

1. Yoon HM, Byeon S, Hwang JY, et al. Sacrococcygeal teratomas in newborns: A comprehensive review for the radiologists. *Acta Radiol.* 2018;59(2):236–246. https://doi.org/10.1177/0284185117710680
2. Perrone E, Jarboe M, Maher C, et al. Early delivery of sacrococcygeal teratoma with intraspinal extension. *Fetal Diagn Ther.* 2018;48:72–76. https://doi.org/10.1159/000472714
3. Draper H, Chitayat D, Ein SH, Langer JC. Long-term functional results following resection of neonatal sacrococcygeal teratoma. *Pediatr Surg Int.* 2009;25:243–246. https://doi.org/10.1007/s00383-009-2322-1
4. Swamy R, Embleton N, Hale J. Sacrococcygeal teratoma over two decades: Birth prevalence, prenatal diagnosis and clinical outcomes. *Prenat Diagn.* 2008;28:1048–1051. https://doi.org/10.1002/pd.2122
5. Schropp KP, Lobe TE, Rao B, et al. Sacrococcygeal teratoma: The experience of four decades. *J Pediatr Surg.* 1992;27(8):1075–1079. https://doi.org/10.1016/0022-3468(92)90563-M
6. Guvenc BH, Etus V, Muezzinoglu B. Lumbar teratoma presenting intradural and extramedullary extension in a neonate. *Spine J.* 2006;6:90–93. https://doi.org/10.1016/j.spinee.2005.08.013
7. Graf, JL, Albanese, CT. Fetal sacrococcygeal teratoma. *World J Surg.* 2003;27:84–86. https://doi.org/10.1007/s00268-002-6741-6

8. Tuladhar R, Patole SK, Whitehall JS. Sacrococcygeal teratoma in the perinatal period. *Postgrad Med J*. 2000;76:754–759. https://doi.org/10.1136/pgmj.76.902.754
9. Wessell A, Hersch DS, Ho CY, Lumpkins KM, Groves MLA. Surgical treatment of a type IV cystic sacrococcygeal teratoma with intraspinal extension utilizing a posterior-anterior-posterior approach: a case report. *Childs Nerv Syst*. 2018;34:977–982. https://doi.org/10.1007/s00381-018-3718-9
10. Flake AW. Fetal sacrococcygeal teratoma. *Semin Pediatr Surg*. 1993;2(2):113–120.
11. Moore KM, Persaud TVN, Torchia M. *The Developing Human: Clinically Oriented Embryology*. 11th ed. Philadelphia, PA: W. B. Saunders Company; 2020: 47–51.
12. Kunisaki SM, Maher CO, Powelson I, Gemmete JJ, Hirschl RB, Mychaliska GB. Benign sacrococcygeal teratoma with spinal canal invasion and paraplegia. *J Pediatr Surg*. 2011;46:E1-E4. https://doi.org/10.1016/j.jpedsurg.2011.05.013
13. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American academy of pediatrics surgical section survey 1973. *J Pediatric Surg*. 1974;9(3):389–398. https://doi.org/10.1016/S0022-3468(74)80297-6
14. Hambreus M, Hagander L, Stenstrom P, Arnbjornsson E, Borjesson A. Long-term outcome of sacrococcygeal teratoma: A controlled cohort study of urinary tract and bowel dysfunction and predictors of poor outcome. *J Pediatr*. 2018;198:131–136. https://doi.org/10.1016/j.jpeds.2018.02.031
15. Le LD, Shumyle A, Lim FY, Keswani SG, Crombleholme TM. Prenatal and postnatal urologic complications of sacrococcygeal teratomas. *J Pediatr Surg*. 2011;46:1186–1190. https://doi.org/10.1016/j.jpedsurg.2011.03.045
16. Bechtel, AS, Gauger, CA. Sacrococcygeal teratoma with multiple recurrences by intradural extension in the neonate: A common neonatal mass with an unusual course. *Glob Pediatr Health*. 2014;1. https://doi.org/10.1177/2333794X14560820
17. Duczkowska A, Bekiesinksa-Figatowska M, Herman-Sucharska I, et al. Magnetic resonance imaging in the evaluation of fetal spinal canal contents. *Brain Dev*. 2011;33:10–20. https://doi.org/10.1016/j.braindev.2010.01.003
18. Davidson MR, London ML, Ladewig, PA. *Olds' Maternal Newborn Nursing & Women's Health Across the Lifespan*. 9th ed. Upper Saddle River, NJ: Pearson Education Inc.; 2012.
19. Valdiserri RO, Yunis EJ. Sacrococcygeal teratomas: A review of 68 cases. *Cancer*. 1981;48(1): 217–221. https://doi.org/10.1002/1097-0142(19810701)48:1<217::AID-CNCR2820480135>3.0.CO;2-R
20. Hedrick HL, Flake AW, Crombleholme TM, et al. Sacrococcygeal teratoma: Prenatal assessment, fetal intervention and outcome. *J Pediatr Surg*. 2004;39(3):430–438. https://doi.org/10.1016/j.jpedsurg.2003.11.005
21. Van Mieghem T, Al-Abrahim A, Deprest J, et al. Minimally invasive therapy for fetal sacrococcygeal teratoma: Case series and systematic review of the literature. *Ultrasound Obstet Gynecol*. 2014;43(6):611–619. https://doi.org/10.1002/uog.13315
22. Raveenthiran, V. Sacrococcygeal teratoma. *J Neonatal Surg*. 2013;2(2):18.
23. Papagelopoulos PJ, Peterson HA, Ebersold MJ, Emmanuel RP, Choudhury SN, Quast LM. Spinal column deformity and instability after lumbar or thoracolumbar laminectomy for intraspinal tumors in children and young adults. *Spine*. 1997;22(4):442–451. https://doi.org/10.1097/00007632-199702150-00019
24. Martini F, Bartholomew EF. *Essentials of Anatomy & Physiology*. 7th ed. Hoboken, NJ: Pearson Education, Inc.; 2017:280–285.
25. Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 8th ed. Baltimore, MD: Lippincott Williams & Wilkins; 2005:64–84.
26. Volpe JJ, ed. *Volpe's Neurology of the Newborn*. 6th ed. Philadelphia, PA: Elsevier; 2018:1109–1120.
27. Noetzel MJ. Myelomeningocele: Current concepts of management. *Clin Perinatol*. 1989;16(2):311–329. https://doi.org/10.1016/S0095-5108(18)30634-1
28. Hubert RJ, VanMeter KC. *Gould's Pathophysiology for the Health Professions*. 6th ed. St. Louis, MO: Elsevier; 2018:331–339.
29. Heaberlin PD. Neurologic assessment. In: Tappero EP, Honeyfield ME, eds. *Physical Assessment of the Newborn: A Comprehensive Approach to the Art of Physical Examination*. 6th ed. New York, NY: Springer Publishing Company; 2018:144–186.
30. Schmidt B, Haberlik A, Uray E, Ratschek M, Lackner H, Hollwarth ME. Sacrococcygeal teratoma: Clinical course and prognosis with a special view to long-term functional results. *Pediatr Surg Int*. 1999;15:573–576. https://doi.org/10.1007/s003830050675
31. Dos Santos J, Lopes RI, Koyle MA. Bladder and bowel dysfunction in children: An update on the diagnosis and treatment of a common but underdiagnosed pediatric problem. *Can Urol Assoc J*. 2017;11(1–2 suppl 1):S64–S72. https://doi.org/10.5489/cuaj.4411
32. Bittmann S, Bittmann V. Surgical experience and cosmetic outcomes in children with sacrococcygeal teratoma. *Curr Surg*. 2006;63(1):51–54. https://doi.org/10.1016/j.cursur.2005.04.011

About the Authors

Jennifer Young, MN, RN (EC) NP-Pediatrics, NNP-BC, and Hazel Pleasants-Terashita MN, RN (EC) NP-Pediatrics, NNP-BC, are neonatal nurse practitioners at The Hospital for Sick Children in Toronto, Canada.

For further information, please contact:
Jennifer Young, MN, RN (EC) NP-Pediatrics, NNP-BC
The Hospital for Sick Children
555 University Avenue
Toronto ON Canada
M5G 1X8
E-mail: jennifer.young@sickkids.ca