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ABSTRACT

SUMMARY: Dysraphic malformations of the spine and spinal cord (DMSSC) represent a spectrum of common congenital anomalies typically (though not exclusively) affecting the lower spinal segments. These may be responsible for varying degrees of neurologic, orthopedic, and urologic morbidity. With advances in neuroimaging, it is now possible to better diagnose and evaluate these disorders both prenatally and postnatally. Neuroimaging, performed at the right time and with technique optimization, is integral in guiding clinical management. However, the terminology used to describe these lesions has become increasingly confusing, and there is a lack of consensus regarding the essential radiologic features and their clinical weighting. This variability in radiologic practice risks unstructured decision making and increases the likelihood of suboptimal, less informed clinical management. In this manuscript, the first of a series of consensus statements, we outline a standardized international consensus statement for the radiologic evaluation of children with suspected DMSSC derived from a critical review of the literature, and the collective clinical experience of a multinational group of experts. We provide recommendations for plain radiography, sonography, CT, and MR imaging in the evaluation of DMSSC with an emphasis on technique of imaging and imaging protocols.

ABBREVIATIONS: DMSSC = dysraphic malformations of the spine and spinal cord; FS = fat suppression; GRE = gradient-echo; SSFSE = single-shot fast spin-echo; US = ultrasound

Dysraphic malformations of the spine and spinal cord (DMSSC) represent a spectrum of congenital malformations presumed to have their origins in defects of early embryogenesis. The clinical consequences of these disorders affect the development of children worldwide and result in significant personal and socioeconomic costs.[1,2] Despite being relatively common (estimated incidence 1–3/1000 live births), their etiology is largely unknown.[3,4] The terms "spinal dysraphism" or "tethered cord" are often used as umbrella terms for these disparate malformations; however, the term "dysraphism" implies a known etiology (anomaly of midline fusion) and "tethered cord" implies a known mechanism of clinical deterioration. Because both of these assertions are incorrect, we use the term DMSSC to encompass these disorders.[1]

Neuroimaging plays a vital role in the diagnosis, classification, and management of DMSSC. With competent image acquisition and interpretation, diagnostic accuracy can be potentially excellent, providing correct anatomic delineation and aid in appropriate management. Despite this, there is currently no consensus as to how children with suspected DMSSC should be radiologically evaluated. In the absence of this guidance, clear differences arise between centers in terms of diagnostic approach, classification schema, clinical management, and, by virtue, prognosis in terms of neurologic and urologic outcomes.[1] This worrying clinical heterogeneity risks unstructured decision making, missed diagnoses, and potentially suboptimal management of the child.

These challenges highlight the need for an expert-driven multidisciplinary effort to better understand the radiologic and clinical classification of these disorders. To this aim, we established an international multidisciplinary DMSSC group with the aims of disseminating knowledge to the broad medical community, improving the diagnosis and management of DMSSC, and accelerating research in the field.

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In this manuscript, the first of a series of consensus statements, we outline a standardized international consensus statement for the radiologic evaluation of children with suspected DMSSC derived from a critical review of the literature, and the collective clinical experience of a multinational group of experts.

MATERIALS AND METHODS

Literature Review

PubMed was systematically queried for papers reporting 1) radiologic protocols for the investigation of congenital DMSSC and 2) radiologic findings in congenital DMSSC. The keywords used in the search were as follows: "spinal dysraphism," "spine malformation," "spinal cord malformation," "spina bifida," "myelomeningocele," "lipomyelocele," "lipomyelomeningocele," "terminal myelocystocele," "nonterminal myelocystocele," "abortive myelocystocele," "split cord malformation," "neurenteric cyst," "diastematomyelia," "spinal lipoma," "dorsal lipoma," "transitional lipoma," "caudal lipoma," "filar lipoma," "tethered cord," "thickened filum terminale," "caudal regression syndrome," "dermal sinus tract," and "limited dorsal myeloschisis." The most recent search was performed on April 1, 2022. Following this literature search, the final reference list was generated on the basis of 1) relevance to the scope of our recommendations and 2) relative importance and originality within the field.

The Consensus Process

This article represents an international consensus statement based on 5 meetings of the International DMSSC Consensus Group: a panel of 17 recognized experts invited to participate in this modified Delphi consensus process on the basis of prior scholarship in the field and the need for global representation. Participating experts are pediatric neuroradiologists ($n = 9$), pediatric neurosurgeons ($n = 3$), pediatric urologists ($n = 2$), and developmental neurobiologists ($n = 3$). Delphi rounds were mediated by an independent, nonparticipating author.

Meetings were held on June 26, 2020; July 24, 2020; September 25, 2020; November 17, 2020; and January 8, 2021. Of these meetings, the first 2 contained specific focus discussions on the radiologic evaluation of congenital DMSSC. Before each meeting, consensus statements were prepared by a core team (Asthik Biswas, J.S., S.S., and K.M.) based on evidence from the literature and expert opinion. During meetings, the panel discussed consensus statements and agreed on new or modified recommendations for the radiologic evaluation of congenital DMSSC. Consensus statements were subsequently revised in view of these discussions, and the process iterated until consensus was achieved. Consensus was defined as ≥80% agreement (≥14/17 experts). Unless otherwise stated, all recommendations are reported at this level of consensus. The final manuscript was revised and endorsed by all panel members before submission.

In the first Delphi round, all authors voted on 24 recommendation statements. Agreement was reached for 9 statements, and the remaining 15 revised as per the reasons each author provided for disagreement. In the second round, all authors voted on 16 revised recommendation statements and consensus was achieved in all remaining areas.

Consensus Recommendations

This consensus statement should be applied to the radiologic evaluation of all fetuses, children, and adults with suspected DMSSC.

Plain Film Radiography

Conventional radiography with anteroposterior and lateral views is often the first-line screening investigation to assess abnormalities of the vertebral column in children. Findings seen on plain radiographs may include, but are not limited to, spina bifida, widened spinal canal, lumbosacral soft tissue swelling, segmentation anomalies, and the bony spur of diastematomyelia. Plain film radiographs may also aid the evaluation of associated kyphotic and/or scoliotic deformities in patients with certain malformations, such as segmental spinal dysgenesis.[5] However, plain radiography exposes the child to ionizing radiation and images have poor soft tissue resolution, resulting in low diagnostic sensitivity. In addition, overlying gas and stool shadows can limit the evaluation of the spine. Therefore, in the current era, we recommend that plain radiography should only be used as 1) a preliminary screening investigation when other imaging modalities are not available, 2) an adjunct (with the aid of a marker) to aid vertebral counting if there is uncertainty in determining the lumbosacral junction on ultrasonography, or 3) where there is a need to evaluate/monitor associated spinal deformity.[6,7]

Sonography (Ultrasound)

Ultrasound (US) is the first-line technique for the antenatal diagnosis of DMSSC. Despite this, it has a limited role in the postnatal evaluation of suspected DMSSC.[8] The partially ossified, predominantly cartilaginous, not yet fused posterior vertebral elements in this age group provide a good acoustic window for detailed visualization of the spinal cord and caudal structures. Studies have confirmed good concordance between US and MR imaging. Beyond 3–4 months of age, this acoustic window of opportunity is lost due to ossification and closure/fusion of the posterior arches of the vertebral column. After this time, MR imaging becomes the first-line technique for older children.[2,8] Though individual operator expertise is its main limitation, when performed by an experienced operator, US may be used exclusively for the evaluation of DMSSC in low-risk infants less than 3–4 months of age.[9] The advantages of US are its cost-effectiveness, wide accessibility, bedside acquisition, and rapid image acquisition time, which negates the need for sedation. This said, US has lower resolution than MR imaging, and so we recommend that MR imaging is performed in all children in whom DMSSC is suspected on US. Cranial US performed in the same setting can also expedite the diagnosis of associated intracranial anomalies, such as hydrocephalus and Chiari deformity.[10] US may be of further relevance to exclude other/associated non-neurologic findings, such as urogenital abnormalities.

US Technique. We recommend feeding the infant before the examination as a soothing technique. US should then be performed primarily in the prone position, with the child's head slightly elevated above the feet to permit better filling of the lower CSF spaces.[11,12] The child's neck must be slightly flexed when

evaluating the craniocervical junction; a rolled towel or blanket, placed under the child's abdomen or pelvis, may also help to accentuate the lumbar lordosis and widen the posterior interspinous spaces. Real-time scanning in the lateral decubitus position results in free movement and clustering of the cauda equina nerve roots toward the dependent side, thereby permitting the assessment of cord movement.[13] Positioning the child in a semi-erect fashion, with the head held by the sonographer, may enhance the detection of meningocele.[9] US should not be used to image open DMSSC as this provides limited additional information and increases susceptibility to infection.[14] In open DMSSC, US should be used to image more rostral parts of the vertebral column for the assessment of associated anomalies, such as hydrosyringomyelia and hydrocephalus.

High-frequency linear-array (7–12 MHz) and curved-array (8–10 MHz) transducers should be used to evaluate the spine and spinal cord in the longitudinal and transverse planes with the study limited to the area of interest, usually lumbosacral and lower thoracic region with evaluation and characterization of the filum terminale, cauda equina nerve roots and distal thecal sac, ossified parts of the bony vertebrae (including its posterior elements), and any skin lesions or masses. Sonography aids in assessment of overlying soft tissues for the presence of hemangioma, lipoma, skin covered masses (meningocele), and tracts extending from the skin surface toward the spinal canal. A thick layer of coupling gel or a standoff pad may help in better assessment of superficial soft tissues. Color or power Doppler sonography may also be used as an adjunct to better characterize soft-tissue masses (eg, cutaneous hemangiomas) found on the skin or within the spinal canal.[6,7] The study may be extended to include the entire spinal canal from the craniovertebral junction to the coccyx. If available, a small footprint sector probe may be used for detailed evaluation of the craniocervical junction. Panoramic or extended FOVs can visualize the neonatal spine from T12 to the coccyx in a single image, potentially permitting full visualization of any abnormalities. 3D US is not essential but may be of use in complex cases for visualization in the additional coronal plane.[15]

The position of the conus medullaris should be assessed by identifying the lumbosacral junction and thus the location of the L5 vertebra at the lordotic angle between the lumbar and sacral vertebrae and should be confirmed by counting the vertebral level down from rib 12 or counting cephalad from S5 (rounded or triangular shape of first coccygeal segment when ossified).[6,7,9,12,15] In neonates, wherein the acute angle may not be seen clearly, flexion and extension movements of the pelvis may help to identify the point of motion of the sacrum. Alternatively, comparison with a marked lateral plain radiograph may be used.

Antenatal US. Imaging plays a crucial role in the prenatal diagnosis and classification of DMSSC, as emphasized by recent advances in intrauterine repair.[16] 2D and 3D US is invariably the first-line technique for the morphologic study of the fetus.[17] Second trimester US, in particular, has a high sensitivity for the detection of DMSSC and is employed in routine screening programs across the world, making it possible to suspect and detect neural tube defects early in gestation.[18] Maternal serum alfa-fetoprotein screening can also help to identify high-risk children and define the need for more detailed fetal imaging (US or MR imaging) and/or invasive tests, namely, amniocentesis.[19] As such, the radiologic investigation of suspected DMSSC should always be interpreted in tandem with maternal serum alfa fetoprotein levels.[20]

To screen for suspected DMSSC, the fetal head and entire length of the fetal spine should be studied in the coronal, parasagittal, and transverse planes.[21] US is particularly sensitive in the evaluation of the skin, soft tissues, vertebral body ossification centers, brain for features of Chiari II deformity, in addition to any mass lesions, sacral anomalies, and sac(s), if present. Antenatal US is more sensitive for the diagnosis of open DMSSC than for closed DMSSC.[18]

In cases of myelomeningocele, the anatomic level of the lesion is important both for prognostication and, these days, as an eligibility criterion for possible fetal surgery. Studies have confirmed the comparable accuracy of fetal US and MR imaging in ascertaining level of myelomeningocele defect.[22] Additionally, antenatal sonography also has the advantage of detecting associated anomalies, including cardiac, renal, and bowel anomalies, which are important in determining eligibility for fetal surgery. Antennal US also aids in diagnosis of lower limb abnormalities (eg, equinovarus feet, vertical talus) and assessment of lower limb movements of the fetus, adding a functional perspective to this imaging technique.

MR Imaging

MR imaging is the technique of choice for the evaluation of suspected DMSSC because of its excellent spatial and contrast resolution with multiplanar and multicontrast capabilities in the absence of ionizing radiation.

Sedation. One of the main challenges in pediatric MR imaging acquisition is the varying abilities of children to tolerate the environment of the scanner and the requirements of imaging, namely, the need to remain stationary. In neonates and young infants, imaging during spontaneous sleep following feed with the baby wrapped up in a blanket (feed-and-swaddle or feed-and-wrap) is a viable option and obviates the need for sedation in this age group.[23] Attempts to keep the baby awake, hungry, and due for feed before the scheduled examination help to increase the likelihood of a spontaneous sleep following feeding. Similarly, the availability of dedicated quiet rooms for patient preparation and subsequent awakening greatly improves the chances of success for imaging small infants without sedation. Children aged 4 years and older may be sufficiently cooperative, especially with the support of a child life specialist, including mock-MR training, though this may vary because of acute illness and the developmental stage of the child.[2,24] Younger or severely ill children will typically require sedation, administered according to local guidelines. Cardiorespiratory monitoring with MR imaging–compatible equipment is required in all sedated children. Further techniques to minimize sedation during MR imaging, including fast sequences, motion correction, noise reduction, and reducing scan time, are beyond the scope of this manuscript and are discussed in detail in previously published literature.[25]

Table 1: Recommended MR imaging sequences and parameters for the assessment of children with suspected DMSSC

Sequence	Plane	Imaging Parameters	Notes
Essential sequences			
3 plane scout/localizer	Axial, sagittal, coronal		For subsequent planning
T1-weighted TSE whole spine	Sagittal	3.0 mm thickness (TR, 600 ms, TE, 30 ms)	—
T2-weighted TSE whole spine	Sagittal	3.0 mm thickness (TR, 3000 ms, TE, 120 ms)	—
T2-weighted FS, Dixon, or STIR	Coronal	3.0 mm thickness (TR, 3000 ms, TE, 40 ms)	FS preferred over STIR; whole spine
T1-weighted TSE	Axial	≤3.0 mm thickness	Lumbosacral region (conus and filum terminale) and the suspected area of abnormality (group of axial images through the disc level not applied)
T2-weighted DRIVE, CISS, or FIESTA	Sagittal	0.6 mm thickness	Sagittal acquisition centered on the area of suspected abnormality with 3D reconstructions
Optional sequences			
T2-weighted TSE	Axial	3.0 mm thickness, non-fat-suppressed	Suspected area of abnormality (group of axial images through the disc level not applied)
T1-weighted TSE	Coronal	3.0 mm thickness	Centered onto and along the major axis of the sacrum (for suspected sacral abnormalities)
T1-weighted FS	Sagittal	3.0 mm thickness	Confirmation of lipoma
T1-weighted FS C+	Axial, sagittal, coronal	3.0 mm thickness	Suspected infections/tumors
DWI	Axial or sagittal	3.0–4.0 mm thickness	Suspected dysontogenic abnormalities, epidermoids, dermoids, abscesses
T2-weighted GRE or EPI-GRE	Axial	3.0 mm thickness	Evaluation of bony septum in diastematomyelia
T1-weighted TSE C+	Axial, sagittal, coronal	3.0 mm thickness	Suspected mass lesions, dysontogenic abnormalities, or infections

Note:—DRIVE indicates driven equilibrium; C+ = postcontrast.

Scanner Magnetic Field Strength. Both 1.5T and 3T scanners are suitable for imaging suspected DMSSC. As such, the choice of magnetic field strength depends on local availability and radiologist preference. 1.5T scanners remain the most widely available.[26] Advantages of 3T MR imaging include higher spatial and contrast resolution; the potential for reduced scan times without compromising image quality; and reduced motion artifacts with higher temporal resolution.[27] 3T scanners are, however, more costly, and artifacts caused by field inhomogeneity, magnetic susceptibility, vascular pulsation, and chemical shift are exaggerated. Spinal imaging remains particularly challenging at 3T despite technical advances, such as thin section imaging, parallel imaging, and increasing the receiver bandwidth.[27]

Standardized Spinal MR Imaging Protocol. In cases of myelomeningocele or syndromes associated with dysraphism (eg, VACTERL, cloacal exstrophy), whole spine imaging is required. In isolated, closed dysraphic states, there is limited clinical utility in imaging beyond the lumbosacral region.[28] Optimized MR imaging protocolling is crucial to maximize diagnostic yield and reduce scanning time, thereby limiting the necessity or duration of sedation. We recommend imaging of the whole spine at baseline, including dedicated, high-resolution imaging of the area of the suspected abnormality. Given the inherent challenges of MR imaging in children, essential sequences should be acquired first, with optional sequences acquired subsequently as required.

The standardized spinal MR imaging protocol for DMSSC evaluation is presented in Table 1. Following localizer or scout imaging, high-resolution T1- and T2-weighted TSE images of the whole spine are acquired in the sagittal plane without fat suppression (FS). Advances in MR imaging, the use of multichannel phased array coils, and the combination of multiple images into a single full FOV have enabled visualization of the entire spine, from the craniocervical junction to the coccyx, in a single image, thereby permitting panoramic appraisal and the counting of vertebral levels to identify the exact level of abnormality. In addition, 1 panoramic coronal sequence (T2-weighted TSE [T2-TSE]) with FS (T2-TSE FS, Dixon, or short tau inversion recovery [STIR]) is acquired of the whole spine. T2-weighted FS is preferred because of its inherently high signal-to-noise ratio, good visualization of small anatomic detail, and shorter acquisition time.[29] Axial acquisition on T1-weighted imaging without FS is then used to study specific regions as indicated by clinical findings or by findings on the previously acquired sagittal images (block acquisition and not at the level of intervertebral discs). The section thickness for these sequences should be ≤3.0 mm with submillimeter in-plane resolution and intersection gaps of 0.30–0.50 mm.[2,30] A volumetric acquisition of high-resolution heavily T2-weighted images in the sagittal plane with retrospective multiplanar reconstructions should also be performed, either driven equilibrium (DRIVE), CISS, or FIESTA. These provide exquisite delineation of the cord/root/CSF interfaces and are particularly useful for evaluating subtle structural abnormalities, such as those found in DMSSC.

Routine DWI is not required in children with DMSSC, but it should be performed for the identification and assessment of dysontogenetic mass lesions. The high lesion conspicuity of postoperative inclusion epidermoids/dermoids after repair of a spinal dysraphism on DWI may be advantageous. Similarly, gradient-echo (GRE) or EPI-GRE are useful for the evaluation of the bony septum in children with diastematomyelia.

The intravenous injection of gadolinium-based contrast agents is not routinely indicated and should only be used to evaluate suspected infections and mass lesions inadequately characterized on noncontrast MR imaging. MR angiography may also be used for preoperative identification of the artery of Adamkiewicz (great anterior radiculomedullary artery).

Additional screening of the cranial vault should be considered to exclude associated cerebral and/or cerebellar abnormalities. Other optional sequences may be added to the protocol depending on clinical indication, findings on initial imaging, and national guidance.

With the advances in imaging, it is now possible to decrease examination times while maintaining diagnostic performance which is of paramount importance in radiologic evaluation of DMSSC. These advances include faster sequences, powerful computers for faster image reconstruction, 3D sequences, acceleration techniques, such as parallel imaging, simultaneous multislice imaging, compressed sensing, and deep learning reconstructions. Parallel imaging is the most used technique, available in most modern scanners without the need for specialized software or hardware. In simultaneous multislice imaging, excitation of more than 1 section is done at a time and uses the same coil technology and reconstruction methods as parallel imaging. Both these techniques allow acceleration to a factor up to 2 times without degrading the image quality and when used in combination, can provide an acceleration factor of 4 with similar signal-to-noise ratio and contrast-to-noise ratio. Further, deep learning models can reconstruct the undersampled data to simulate the fully sampled reconstructions.[31]

Follow-up MR Imaging. At follow-up, imaging can be limited to the area of interest with screening T2-weighted images of the whole spine without FS.[2]

Fetal MR Imaging

Fetal MR imaging is the preferred second-line technique (after prenatal sonography) for imaging of the fetus.[32,33] Though not indicated in all children because of availability and technical limitations, it is a powerful adjunct to prenatal US, providing additional information crucial for prenatal counseling, assessing eligibility for prenatal surgery, predicting neurologic outcomes, and guiding perinatal management.[34]

DMSSC become more evident on fetal MR imaging in the second trimester. This fortunately coincides with the optimal age for MR imaging. We recommend waiting until 17–18 gestational weeks (15–16 weeks postfertilization) before performing fetal MR imaging because of the potential risks posed to the developing fetus and the current technical limitations of fetal MR imaging in younger fetuses due to their smaller size and even greater fetal motion. Pregnant women should only undergo MR imaging earlier in their pregnancy if the risk-benefit ratio to the child is favorable and if other nonionizing imaging modalities are inadequate. In all instances, it is important to counsel parents on the likelihood of diagnosing a DMSSC in their child and of the potential effects this may have on their child's development.

Fetal MR imaging aims to identify pertinent anatomic features of DMSSC, such as the level of the spinal defect, by 1) establishing the most caudal hyperintense spinal disc space as L5–S1 and the lowest horizontal vertebral body as L5, and 2) counting the vertebral bodies superior to the highest level of the absence of the posterior elements at the bone/skin defect. Fetal MR imaging can also define and characterize the presence or absence of a spinal cord syrinx; diastematomyelia; and sac; and the continuity of cutaneous soft tissues with the neural tube sac.[35,36] Associated anomalies of the fetal extremities and intracranial anomalies may also be detected on fetal MR imaging, including the severity of the Chiari II deformity according to the degree of posterior fossa hindbrain herniation, lateral ventricular size, and third ventricular size.[36]

Standardized Fetal MR Imaging Protocol. Before undergoing fetal MR imaging, child-bearing women must empty the urinary bladder. A phased array body surface coil is then wrapped around the mother's pelvis and centered over the fetal ROI. Maternal comfort is the priority; both supine and left lateral decubitus positions are acceptable and should be adopted as per maternal preference.[37]

Prenatal imaging of the fetus is a dynamic process that starts with an initial scout or localizer followed by a series of sequences with each sequence acting as a localizer for the next. Images are acquired in all 3 anatomic planes with respect to the fetus. The main challenge of fetal MR imaging is fetal motion artifact. If persistent and severe, it may be necessary to prioritize image acquisition on planes that best visualize the anatomy in maximizing the yield of the study. Due to this, monitoring by the radiologist is essential.

The standardized fetal MR imaging protocol for DMSSC evaluation is presented in Table 2. Fetal MR imaging should be performed at 1.5T or 3T depending on local availability and radiologist preference. 3T is superior to 1.5T for the visualization of cartilage and spine because of the use of single-shot turbo spin-echo and steady-state free precession sequences. The fetal head and entire length of the fetal spine should be studied on all 3 planes (axial, sagittal, and coronal) by using T2-single-shot fast spin-echo (T2-SSFSE) or HASTE and balanced fast-field echo or FIESTA at 3–4 mm section thickness with no intersection gaps and the smallest FOV possible.[38] A minimum of 2 stacks of images in each plane should be obtained (which may be omitted in case of excessive fetal motion). Gradient-echo sequences (ie, EPI and true FISP) have greater ferromagnetic susceptibility and provide greater resolution of bony and vascular structures, especially in fetuses aged less than 27 gestational weeks.

Optional sagittal and coronal T1-weighted spoiled gradient-echo acquisition of the fetus with section thickness 5 mm and with no intersection gaps may also be performed and should have the smallest possible FOV.[35] Prenatal imaging (including T1-weighted images) does not adequately demonstrate fat within the

Table 2: Recommended MR imaging sequences and parameters for the assessment of fetuses with suspected DMSSC

Sequence	Plane	Imaging Parameters	Notes
Essential sequences			
3 plane scout/localizer	Axial, sagittal, coronal	—	For subsequent planning
T2-weighted TSE maternal pelvis	Sagittal	—	To assess the position of the fetus; reposition the coil if the fetal ROI is not in the center of the coil
T2-weighted SSFSE or HASTE	Axial, sagittal, coronal[a]	3-4 mm thickness, no intersection gaps (TR, 2000–3000 ms, TE, 150 ms), FOV 340 mm, flip angle 160°	Provides excellent anatomic detail
T2-weighted EPI-GRE or true FISP	Axial, sagittal, coronal[a]	4 mm thickness, no intersection gap (TR, 4.22 ms, TE, 1.75 ms), FOV 380 mm, flip angle 65°	Evaluation of bony and vascular structures
Optional sequences			
T1-weighted SPGR	Sagittal, coronal	5 mm thickness, no intersection gaps (TR, 600 ms, TE, 30 ms), FOV 340 mm	Improves spatial resolution with increasing gestational age
Cine imaging	Volumetric acquisition	—	Assesses fetal extremity mobility

Note:—SPGR indicates spoiled gradient recalled-echo.
[a] Acquisition of all 3 planes in T2-weighted SSFSE (HASTE) and T2-weighted true FISP may not be feasible if the fetus is moving excessively; and in such a scenario, the protocol can be curtailed with T2-weighted SSFSE in axial and coronal planes (providing anatomic detail) and T2-weighted true FISP in sagittal plane (providing assessment of osseous structures).

defect of closed dysraphism. This is attributable to several factors, including low spatial resolution of T1-weighted images in the fetus, underdevelopment of fetal fat in early gestational age, and relatively increased proportion of brown fat in the fetus and neonate that has slightly different signal characteristics than white fat on MR imaging. Furthermore, optional cine imaging may convey an idea of the motility of fetal extremities, offering an insight into the child's postnatal prognosis.

CT

CT is of limited value for the evaluation of DMSSC because of its poor soft tissue resolution and correspondingly poor sensitivity, exposure of the child to ionizing radiation, and invasiveness in the case of CT myelography.[2,39] This said, we support the use of CT for several specific indications:

1) Vertebral anomalies where there is a need to define bony anatomy, eg, as part of preoperative planning prior to instrumented fixation.
2) Identification of the bony septum in diastematomyelia.
3) Preoperative identification of the artery of Adamkiewicz (great anterior radiculomedullary artery) via CT angiography when MR angiography either fails to identify the vessel or is not feasible.[40]
4) Patients with absolute contraindications to MRI.

Standardized CT Protocol. We recommend that children undergo a low-dose noncontrast CT of the spinal area of interest (section thickness ≤2 mm), acquired continuously in the axial plane with no intersection gaps. Multiplanar 2D- and 3D-reconstructions in bone and soft kernel should also be performed as they have been shown to increase the sensitivity and specificity of the study.[41] As CT is reserved for the elucidation of specific features, it should, therefore, always be performed with the minimum possible FOV and not extended beyond the region of the abnormality to minimize radiation exposure, as per the as low as reasonably achievable principle.[2,42]

CT has limited soft tissue contrast; thus, evaluation of the thecal sac and its contents is limited. Intrathecal injection of iodinated contrast media in CT myelography may facilitate visualization of the thecal sac and its contents. However, the use of CT myelography is not recommended when MR imaging is available as CT myelography is invasive, less sensitive, and exposes the child to ionizing radiation.[2,39]

Imaging Guideline Adaptations and Further Considerations

The challenges of imaging children vary across institutions and countries depending on 1) clinical management and 2) the availability of resources given the expense of additional imaging and the cost and risk of sedation if required. Therefore, the principal adaptation to this consensus statement is for clinical settings without routine access to MR imaging, in which we recommend that children are referred to institutions with MR imaging; however, we do agree that it may not be possible in certain resource-limited care environments. CT should not be performed in lieu of MR imaging given its markedly reduced diagnostic accuracy. US is the first-line technique for the antenatal diagnosis of DMSSC and has a significant but limited role in the evaluation of neonates and infants with suspected DMSSC. US is the first method of screening for infants up to 3–4 months of age before ossification of the vertebral bodies. MR imaging is the technique of choice for the evaluation of suspected DMSSC because of its excellent spatial and contrast resolution.

We recommend imaging patients with combined cutaneous stigmata (combination of 2 or more midline cutaneous lesions) or an atypical skin dimple with MR imaging; however, US may be used in some cases. Atypical dimples are larger than 5 mm and located within 25 mm of the anus. Other criteria include deep dimples, dimples located cranially to the gluteal crease or outside the midline, and multiple dimples. On the other hand, a simple sacral dimple is smaller in size (<5 mm in diameter) with

a midline placement within 25 mm of the gluteal crease from the anus and has no other cutaneous abnormalities (such as asymmetry of the gluteal crease, capillary hemangioma, hypertrichosis, dermal sinus tract, lipoma, subcutaneous dermoid cyst, pseudotail, or true tail).[43,44] In patients with combination of less than 2 cutaneous stigmata, atypical dimple, and deviation of gluteal cleft, we recommend performing an US during the first month of life; if anomalies are detected, MR imaging should be performed. In patients with sacral dimple alone, pigmentary nevus, and little hemangioma, we recommend regular clinical follow-up and to perform MR imaging only in the presence of neurologic or orthopedic alterations.[43]

CONCLUSIONS

Neuroimaging is central to the multidisciplinary evaluation of children with suspected DMSSC. It is our hope that this international consensus statement will provoke the standardization of image acquisition and evaluation, thereby increasing the diagnostic yield of studies and improving care for children worldwide.

Disclosure forms provided by the authors are available with the full text and PDF of this article at www.ajnr.org.

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