SPINAL DURAL ARTERIOVENOUS FISTULAS

THE UTILITY AND ACCURACY OF CONTRAST-ENHANCED MR ANGIOGRAPHY FOR LOCALIZATION OF SPINAL DURAL ARTERIOVENOUS FISTULAS

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ENGLISH ABSTRACT

Background and purpose: Spinal Dural Arteriovenous Fistulas (SDAVFs) are challenging to diagnose. Often the diagnosis is made when advanced neurological symptoms are present. Radiological examination plays a key role in making the diagnosis. Conventional MR imaging may give rise to the suspicion of a SDAVF and contrast-enhanced MR angiography (CE-MRA) can serve as a useful non-invasive examination to detect SDAVFs and predict their location prior to digital subtraction angiography (DSA). Few experiences are published with CE-MRA but only in preliminary fashion or comprised small number of cases. By using CE-MRA as guidance for selective DSA, burdensome DSA can be avoided. The purpose of this study was to determine the utility and accuracy of contrast-enhanced MR angiography in Spinal Dural Arteriovenous Fistulas in a large number of cases.

Methods: A retrospective analysis from 1999 – 2012 in the Toronto Western Hospital/University Health Network (TWH/UHN) identified 70 patients clinically suspected of harboring a SDAVF. Each patient underwent conventional MR imaging, CE-MRA and DSA. We evaluated for the presence or absence of serpentine flow voids, T2-weighted hyperintensity and patchy cord enhancement on conventional MR imaging as well as the level and side of the fistula as predicted by CE-MRA. DSA was used as the reference standard for the true location of the fistula. Institutional Research Ethic Board approval was obtained.

Results: Of the 70 cases, 53 were determined to be a SDAVF, 10 cases were shown to be other forms of spinal vascular malformation and 7 were negative on DSA. On conventional MR imaging all reported cases of SDAVF showed serpentine flow voids (100%). T2-weighted hyperintensity was seen in 96% extending to the conus in 85% of cases. Patchy cord enhancement was seen in 93%. CE-MRA correctly localized the level and side of the SDAVF in 43 of the 53 cases (81%).

Conclusion: CE-MRA is a useful non-invasive examination in the detection of the level and side of a SDAVF. A negative CE-MRA combined with negative conventional MR imaging can exclude a SDAVF and obviate the need for DSA. CE-MRA facilitates but does not replace diagnostic DSA in cases of SDAVF as confirmation of specific location, type of fistula and arterial detail are required prior to contemplated treatment of these lesions.

Abbreviation key:
SDAVF = spinal dural arteriovenous fistula
CE-MRA = contrast enhanced magnetic resonance angiography
DSA = digital subtraction angiography
FOV = field of view
CMM = case of SDAVF missed or mislocalized
PC = phase contrast
TOF = time of flight
Appx = appendix
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1. INTRODUCTION

Spinal Dural Arteriovenous Fistulas (SDAVFs) is a rare entity of the spinal vascular disease that can be challenging to diagnose. Making the diagnosis of a SDAVF in an early state is important, because without accurate treatment SDAVFs may lead within a short time to considerable morbidity with progressive spinal cord symptoms. In 6 to 24 months the clinical state can deteriorate to a state of paraplegia and incontinence. However, in a majority of patients the diagnosis is made within 1 to 3 years, when most already suffer from significant neurological impairment1,2. Adequate and timely imaging is inevitable in making the diagnosis at the time that symptoms are still reversible after treatment.

Epidemiology and classification

SDAVFs are the most common of all spinal vascular lesions (70 to 80 percent). Annually it affects approximately 5-10 patients per million people, albeit this is probably underestimated3. The vascular lesion is most frequently diagnosed between the 4th and 6th decade with a male-female ratio of 5:1. Almost all SDAVFs are single lesions and are most are found in the lower thoracic or thoracolumbar region. Over 80 percent of all lesions are found between T6 and L2 and there seems to be predominance for the left side2,4. Various classifications are developed for spinal vascular lesions, among other based on embryologic development5, angio architecture and pathophysiology6 and shunt origin with relation to underlying diseases7. Not all classifications are practical for clinical purposes and evolving knowledge in spinal vascular diseases has led to the reappraisal of existing classifications and proposal for other classifications.

A spinal vascular fistula can be located epidural (or extradural), intradural or perimedullar (or pial), whereby the intradural fistula (SDAVF) is called the ‘classical’ type and represent more than 90% of all spinal fistula types. The fistulous connection is typically located on the dura adjacent to the nerve root sleeve and directly connects a radiculomeningeal artery (appendix I, further appx I) with a bridging vein that pierces the dura mater. When a vascular nidus exists between the arterial feeder and the draining vein, the lesion is called a spinal arteriovenous malformation (AVM). Spinal AVMs represent 20-30% of all spinal vascular lesions, also classifiable in various subtypes.8

Pathophysiology and Clinical features

The pathological fistulous connection between the radiculomeningeal artery and perimedullary vein causes an abnormal retrograde flow in the perimedullary venous plexus (appendix II, further: appx II) which led to venous congestion and venous hypertension9. As a result, the perimedullary veins may become thickened and tortuous, the blood-cord barrier may disrupt and a decreased pressure gradient develops with consequently cord edema and a decreased spinal cord perfusion. Without treatment this may end in irreversible ischemia10. Symptoms of the myelopathy are related to the level of the progression of the venous congestion and consecutive edema in caudocranial direction11,12. Often the lower thoracic spinal cord and conus medullaris region are involved, resulting in associated neurological symptoms. Clinically, patients suffer from aspecific sensory symptoms such as paresthesias or hypo-esthesia, gait disturbances and lower back pain with or without irradiation to the legs. Later on, also motor symptoms and bowel, bladder and sexual dysfunction may appear which often persists to a certain extent after treatment1,2. Because of the non-specificity of the symptoms, the disease can easily be attributed to more common diseases, such as a peripheral nerve disorder10. This reason, but also the rarity and the difficulty to diagnose SDAVFs, has resulted in a great discrepancy in the severity of symptoms between the time of presentation and the time of diagnosis. Van Dijk et al. reported in 2002 a motor, sensory and bladder impairment at onset in 55%, 47% and 10% respectively, which was increased at the time of diagnosis to 96%, 90% and 82% respectively. Comparable results were published by Narvid et al. in 20081,2.
Radiological Features

Radiological examination plays a key role in establishing the diagnosis and includes: conventional spinal MR imaging, MR angiography and intra-arterial digital subtraction angiography (DSA)\(^{6,13}\). Findings of SDAVF on conventional MR imaging have been well described and include: T2-weighted hyperintensity central in the spinal cord that span over several levels, T1-weighted patchy Gadolinium-enhancement within the spinal cord and (predominantly posterior) located serpentine flow voids along the spinal cord. These findings lead one to suspect a SDAVF, but cannot predict the level or side of a SDAVF\(^{13-17}\). Pathophysiologically, the usually homogeneous, not well-delineated T2-weighted hyperintensity reflects spinal cord edema and extends often to the conus medullaris. Although not frequently reported, a hypo intense rim can be seen surrounding the T2-weighted hyperintensity and it represent deoxygenized blood within the dilated capillary vessels\(^{13}\). The serpentine flow voids reflect the thickened and tortuous vessels along the spinal cord. The patchy Gadolinium-enhancement is caused by a disruption of the blood-cord-barrier due to the venous congestion\(^{18}\).

In MR angiography significant advances have been made the last decades in depicting the spinal vessels and the spinal vascular lesions\(^{19,20}\). Several MR angiographic techniques are used with various levels of success. The early efforts in using MR angiography starts in the 90's and include Phase-contrast (PC)\(^{21,22}\) and Time-Of-Flight (TOF)\(^{23}\) techniques in 2D and 3D mode. However they could depict in most cases abnormal dilated spinal arteries and veins, both techniques failed in many cases to depict the location where the arterial feeder connects to the draining vein i.e. the fistulous connection. Reason for this was the limited spatial and temporal (since the techniques strongly rest on high blood-flow) resolution and the long acquisition time that caused for overlapping enhancement of veins and background tissue.

In 2002 a successful small initial experience was published with patients that undergone first-pass contrast-enhanced MR angiography (CE-MRA)\(^{24}\). In 89% of the cases the location was correctly predicted using CE-MRA. This CE-MRA technique is independent from the blood-flow dependent PC and TOF techniques, has a shorter acquisition time and uses a contrast agent for a high Contrast-to-Noise ratio to depict the vessel lumen and the potential fistula\(^{19}\). Small vessels with a slow or turbulent flow, like in SDAVFs, can therefore still be depicted. Other reports show that CE-MRA is successful in supporting the diagnosis and direct the DSA\(^{17,25-28}\). Most of these techniques to date have been reported in a preliminary fashion and comprised small patient numbers. Further investigation with larger case series is needed to determine the accuracy and utility of CE-MRA in the depiction of spinal fistulas.

Treatment

The treatment has to be tailored to each patient’s peculiar disease and requires a multidisciplinary approach that eventually will lead to an approach that will comprise endovascular embolization with N-butyl-2-cyanoacrylate (NBCA) and/or, in case of failing or not feasible, surgical ligation of the fistulous connection. However surgical treatment seems to be superior to endovascular management, confirmatory DSA and embolization can nowadays be combined in one procedure because of the decreased DSA procedural time by using CE-MRA as guidance. This justifies an endovascular attempt when feasible\(^{4,29-32}\).

Spinal vascular anatomy\(^{4,30,33-38}\)

For a correct interpretation of the radiological examination and contemplated endovascular treatment it is essential to have a thorough knowledge of the vasculature of the spinal cord. In the adult, the spinal cord is supplied by the segmental arteries which originate from the deep and ascending cervical arteries or the vertebral arteries at cervical level, from the aortic arch and descending aorta at the thoracolumbar level (the intercostal and lumbar arteries, \(\text{appx I B}\)) and from the internal iliac arteries and median sacral artery at lower lumbar level. Incidentally, they can also
origin from other arteries, especially in the cervical and upper thoracic level. Connections with the pharyngeal, occipital, esophageal, pleural, pericardial and right bronchial arteries are described. Between the segmental arteries the arterial intersegmental anastomoses (appx I Ba,C,G,I) which preserve an adequate blood supply to the spinal cord. The segmental arteries split up in three different trunks; (I) the lateral (or ventral) trunk (appx I F) that supplies the ribs, adjacent intercostal muscles and surrounding tissues of blood, (II) the middle (or dorsal) trunk (appx I H,I,J) which provides the accompanying muscles from blood and (III) the medial (or spinal) trunk (appx I K) that enters the intervertebral foramen and supplies the local epidermal and dural elements and at certain levels also the spinal cord. The local epidermal elements are fed ventrally by retrocorporeal (appx I La) and dorsally by radiculoradial (appx I Lb) arteries that split off from the spinal trunk before it pierces the dura. By entering the dura, the spinal trunk can divide in three types of radicular arteries; (a) the radicular arteries (appx I Ka) that supplies the nerve roots and sometimes the nerves itself from blood; (b) the radiculomeningeal arteries (appx I M,N) that supplies the nerve root and the dura mater; (c) and the radiculomedullary arteries (appx I P) which are supplying the spinal cord, but are not present at all vertebral levels. Once present, they follow the anterior (or ventral) nerve root and drain into the anterior spinal artery (ASA, appx I Q). Occasionally they give off branches to contribute to the anterior pial network. When it follows the dorsal nerve root or do not connect directly to the ASA, it is called a radiculopial artery (appx I O) and these connect subsequently to one of the two posterior spinal arteries (PSAs, appx I S,T) or direct to the dorsal pial network. The number of radiculomedullary arteries anterior varies from 2 – 14 (mean of 6); the radiculopial arteries from 11 – 16 (mean 12). The largest anterior radiculomedullary artery is called the ‘artery of Adamkiewicz’, and is clinically important because it contributes substantially to the blood supply of the spinal cord. Losing this artery may result in colonic ischemia, sensory deficits, paraplegia and loss of bowel and bladder function”.91

The ASA arise from the one or two vertebral arteries and confluence at the C2-C4 level; the two posterior spinal arteries (PSAs) arise from the pre-atlantal portion of the vertebral arteries or from the posterior inferior cerebellar arteries (PICA). The ASA and PSAs pursue their way down to the conus were they forming together the ‘rami cruciantes’ (or arcade, appx I Z). Along their course they have extensive anastomoses that surround the spinal cord and are called the pial plexus (or ‘vasacorona’, appx I R,U,V). This network forms the base of the intrinsic medullar blood supply. Small perforors (appx I X) branch off the pial plexus into the spinal cord. From the ASA sulco-commissural (or sulcal) arteries (appx I W) travel into the anterior median fissure were they dispersing centrifugally in small branches (appx I Y). The ASA supplies two third of the spinal cord, which includes the gray matter; the PSAs the dorsal one third which is mostly white matter.

The venous system shows an enormous variation with even more anastomoses as seen in the arterial system and is divided in an intradural (intrinsic and extrinsic part) and an extradural system. Intradural in the intrinsic part, sulcal (central, appx II A,B) and radial (peripheral, appx II C) veins collects the deoxygenated blood within the spinal cord and drains equally the blood through an extrinsic venous ring (appx II H) around the spinal cord to the also extrinsic situated longitudinal anterior and posterior spinal veins (appx II E,F). These anterior vein and (up to three) posterior veins are cranially connected to the brainstem veins and basal sinuses of the foramen magnum and caudally it continues in 80% as a terminal vein (appx II I) to the end of the dural sac. In between, numerous radiculomedullary (or radicular, appx II K,O) veins branches off these spinal veins. Disregarding the smaller radiculomedullary veins (<0.25mm), there are 8 – 14 anterior and 5 – 10 posterior radiculomedullary veins. They pierce the dura in 60% adjacent with the nerve roots and in 40% there is a separate foramen in the dura. The transdural part of these radiculomedullary veins is substantially narrowed and has a zigzag course, which results in a high resistance and acts as a ‘valvelike’ anti-reflux system. Furthermore the spinal venous system, including the extradural system is valveless. Once outside the dura mater there is a continuation into the now called extradural (or epidural)
system which is divided in three intercommunicating parts: (a) the anterior and posterior internal vertebral venous plexus \((appx\ II\ P,Q)\) that is located inside the vertebral canal but outside the dura mater; (b) the anterior and posterior external vertebral venous plexus \((appx\ II\ U,V)\) that surrounds the entire vertebral bony column and (c) the basivertebral veins \((appx\ II\ S,T)\) that travels transversally through the vertebral corpus and connects the anterior internal and external vertebral venous plexus. The plexuses drains cervical via the vertebral veins in the internal jugular, vertebral and deep cervical veins and is cranially connected with the suboccipital venous system. At thoracic and thoracolumbar level the plexuses drains via the intercostal veins in the azygos, hemiazygos or accessory hemiazygos veins, at lumbar level via lumbar veins \((appx\ II\ N)\) in the ascending lumbar vein \((appx\ II\ R)\) or renal vein and sacral via the lateral sacral vein in the iliac vein. Finally they all drain into the superior and inferior vena cava towards the right atrium.
2. Research question

The University of Toronto Brain Vascular Malformation Study Group has collected data from patients who presented at the Toronto Western Hospital/University Health Network with vascular malformations from the foundation of the group. After updating this database, patients assessed for SDAVFfs are selected for further analysis. The purpose of this study was to evaluate the accuracy and utility of contrast-enhanced MR angiography in localizing and visualizing Spinal Dural Arteriovenous Fistulas prior to Digital Subtraction Angiograph. Following this, the research question of this project is:

*What is the utility and accuracy of contrast-enhanced MR angiography in localizing and visualizing Spinal Dural Arteriovenous Fistulas?*
3. MATERIALS AND METHODS

Patients

From September 1986 to August 2012, the multidisciplinary AV malformation clinic at Toronto Western Hospital, University Health Network (TWH, UHN) has seen 127 patients that were assessed for a SDAVF. The records and imaging of the 127 patients were retrospectively reviewed to identify a subgroup of patients initially considered for a diagnosis of SDAVF who underwent consecutive CE-MRA (available since 1999) and catheter based spinal intra-arterial digital subtracted angiography at TWH. Patients with a history of treated SDAVF were excluded. All patients in our study group underwent CE-MRA and subsequently DSA as described below. The radiologist’s original report of the CE-MRA and DSA were used as the tabulated results for this study. Angiographic images, CE-MRA or DSA, were not routinely re-reviewed as part of this study. The presence or absence of SDAVF and its location as well as data regarding findings at conventional MR imaging were also tabulated. Approval for this retrospective project was obtained from the local hospital institutional Research Ethics Board.

MR imaging

All patients underwent conventional MR imaging of the cervical, thoracic and/or lumbar spine prior to the CE-MRA. This examination was clinical indicated and requested by the referring physician. The images were obtained with a superconducting 1.5 T system (Signa cv/I, version 8.2.5 – 12.0 software: GE Medical Systems, Milwaukee, Wis) with the patient in a supine position. A posterior standard phased-array spinal coil was used and the location of FOV was chosen thoracolumbar or based on clinical and/or radiological suspicion of the location of the fistula. Standard sagittal T1 (with and without gadolinium) and T2 weighted MR images were performed. Sequence parameters for T1-weighted images include: FOV 32-40 x 32-40 cm; Repetition Time 400-560 msec; Echo Time 9-24 msec; fractional echo acquisition; Echo Train Length 0-1; matrix 512 x 192-224; bandwidth 32.0kHz; slice thickness 3.0 mm; section spacing 0.5 mm. For T2-weighted imaging the following parameters were used: FOV 32-40 x 32-40 cm; Repetition Time 3550-6450 msec; Echo Time 103-119 msec; fractional echo acquisition; Echo Train length 10-27, matrix 512 x 192-224; bandwidth 32.0kHz; slice thickness 3.0 mm; section spacing 0.5 mm. When there was a clinical or radiological suspicion for the existence of a SDAVF, the patients underwent subsequently also a MR angiography. MR angiography was performed as a contrast enhanced first-pass technique employing real-time visualization of gadolinium inflow and manual or automated triggering of the elliptic centric ordered (former known as ATECO) 3D gadolinium enhanced angiographic sequence similar to that previously reported. Similar CE-MRA protocols and sequences are routinely commercially available from all MR-system vendors. Sequence parameters for the 3D gadolinium enhanced sequence include: flip angle 35°; FOV 36 x 27cm; Repetition Time 4.2msec; Echo Time 1.2msec, fractional echo acquisition; number of excitations 1; matrix 352 x 352; bandwidth 62.0 kHz; slice thickness 1.2mm, zero skip, no interpolation, sections acquired 124 and scan time of 147 seconds.

As described by Farb et al., the CE-MRA technique consisted of 4 integrated parts: the first is an initiation of a two dimensional single-section bolus-detection sequence oriented in the transverse plane and located at approximately the level of T10. Secondly the patient receives, via a 22-gauge intravenous catheter in the antecubital vein, an intra-venous power injection using a two-cylinder MR-compatible injector (Spectris; Medrad, Indianaol, Pa) of 10mL gadobutrol (Gadovist; Bayer Schering Pharma, Berlin, Germany) with an injection rate of 1.5mL/s followed by a flush of 30mL of saline (1.5ml/sec), followed by manual or automated detection of the arrival intra-arterial gadolinium contrast agent (aortic) at the level T10. This results in termination of the detection sequence, insertion of an approximate 4 second delay and subsequent initiation of the three-dimensional fast-gradient-echo MR angiographic sequence with elliptic centric ordered phase
encoding. The source images obtained with this CE-MR angiographic technique were post processed on a workstation (Advantage for Windows, version 4.2; GE Medical Systems) creating multiplanar volume reformatted (MPVR) images, contiguous 7 mm thick sections displayed as Maximum Intensity Projection (MIP), in axial and coronal planes through the entire imaged volume. The formal report for each CE-MRA was rendered based on these MPVRs as well as the source images were reviewed by one of 5 neuroradiologists all experienced (>5 years) in spinal vascular imaging.

**DSA Examination**

DSA was, in accordance with the literature, considered as the gold standard in the depiction of SDAVF. The DSA examinations were performed using a biplane dedicated neuroangiographic system (Advantx LCN+, GE Medical Systems, Buc, France; INFX-8000 Vfi-BP, Toshiba, Nasu, Japan) following the protocol as described by Willinsky et al. The patients were under general anesthesia and multiple selective arterial injection with iodinated contrast agent (Omnipaque 300; GE Healthcare, Chalfont St. Giles, UK) were performed to identify the anterior spinal artery and the level, side and angio architecture of the fistula. In all cases the angiographers were informed about the CE-MRA results and were encouraged to direct the injections to the presumed fistula location early in the procedure.

**Data collection**

Data was extracted from the original neuroradiology reports available in the hospital in the Radiology Information System (RIS) and PACS viewing workstation (eFilm 2.1.2; Merge Healthcare, Milwaukee, Wis). When the original reports were not available, the data was collected from medical letters in the Electronic Patient Record (EPR) Online Documentation (SIMS Education). In two instances where documentation regarding the interpretation of conventional imaging was not available the images were reviewed by an experienced neuroradiologist (Dr. R.I. Farb). The data collected from the conventional MR imaging included;

- presence or absence of T2-weighted hyperintensity within the spinal cord
- extension of T2-weighted hyperintensity to the conus medullaris
- intradural serpentine flow voids along the spinal cord
- intramedullary spinal cord enhancement on T1-weighted images after gadolinium injection.

The location, level and side of the SDAVF as predicted from the MR angiographic report was also recorded for each patient. In cases where a discrepancy of only one vertebral level occurred between that predicted by CE-MRA and the true site of fistula at DSA, the images were reviewed together with a neuroradiologist with more than 20 years of experience (Dr. R.I. Farb). This occurred in 3 cases, in all three the level was incorrectly reported at DSA due to miscounting of vertebral segments. Images showed complete anatomic congruence once this correction was made. For the purposes of this study these cases were each tabulated as a correct CE-MRA localization of SDAVF.

Information extracted from each patients DSA report included: confirmation of the diagnosis of SDAVF versus other form of vascular lesion; the location of the fistula; complications during DSA procedure; total DSA procedural time and subsequent disposition of the patient regarding attempted embolization versus surgical referral.
4. Results

Of the original 127 patients referred for evaluation of possible spinal dural arteriovenous fistula, 108 were confirmed SDAVFs on DSA. Seventy cases were identified where the patient underwent consecutive conventional MR imaging, CE-MRA and DSA. In 53 of these 70 cases (76%) the diagnosis of SDAVF was confirmed at DSA after having been suspected at CE-MRA in each case; the ‘SDAVF-group’. The remaining 17 (24%) are described as the ‘non-SDAVF-group’ discussed below and include the variant forms as well as the DSA negative cases.

SDAVF-group

In the 53 confirmed cases of SDAVF the mean age was 60.6 (range, 10 – 86 years) with a strong male predominance (49:4, male/female).

In 43 of the 53 cases (81%, 95% confidence interval (CI) 0.69- 0.89) the CE-MRA correctly predicted the level and side of the fistulous connection. In one case, where the level was correctly predicted, both sides were reported as possible locations (this instance was counted as a correct prediction by CE-MRA). In 10 cases the CE-MRA was either incorrect regarding the perceived location of the fistula (6 cases, 11%) or the neuroradiologist felt that a location of the fistula could not be discerned or suggested (4 cases, 8%) (Table 1).

Table 1.

Missed or mislocalized SDAVF-cases (‘CMMs’)

<table>
<thead>
<tr>
<th>Patient-number</th>
<th>Age, Sex</th>
<th>Year of study</th>
<th>MRI findings</th>
<th>HC on CE-MRA</th>
<th>MRA level fistula</th>
<th>DSA findings</th>
<th>Reason of missing/misinterpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. 1</td>
<td>38Y, M</td>
<td>2001</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>T11 Right</td>
<td>L1 Left</td>
<td>● Vascularity seen at T11 Right</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>● Fistulous connection too small (&lt;1mm)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>● Gap fistulous site &gt; 1.5 cm</td>
</tr>
<tr>
<td>No. 15</td>
<td>48Y, M</td>
<td>2009</td>
<td>Y / Y / N / Y</td>
<td>Y</td>
<td>Indeterminate</td>
<td>T1 Left</td>
<td>● Gap fistulous site &gt; 1 cm</td>
</tr>
<tr>
<td>No. 38</td>
<td>62Y, M</td>
<td>2012</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>Indeterminate</td>
<td>T6 Left</td>
<td>● Fistulous connection too small</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>● Gap fistulous connection &gt; 1 cm</td>
</tr>
<tr>
<td>No. 50</td>
<td>80Y, M</td>
<td>2001</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>T10 or T12 Right</td>
<td>Int. iliart. Left</td>
<td>● Very complex and small fistula (5 DSA’s needed to identify the fistula)</td>
</tr>
<tr>
<td>No. 74</td>
<td>56Y, M</td>
<td>2010</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>Indeterminate</td>
<td>T7 Left</td>
<td>● Degraded by motion artifacts</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>● Very small fistulous connection</td>
</tr>
<tr>
<td>No. 86</td>
<td>64Y, M</td>
<td>2009</td>
<td>Y / Y / Y / N</td>
<td>Y</td>
<td>T9-T10 Left</td>
<td>T6 Right</td>
<td>● Gap fistulous connection 2- 3 cm</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>● Vertical A-V connection</td>
</tr>
<tr>
<td>No. 89</td>
<td>63Y, M</td>
<td>2009</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>T11 Right</td>
<td>L2 Right</td>
<td>● Very small fistulous connection</td>
</tr>
<tr>
<td></td>
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<td></td>
<td></td>
<td></td>
<td>● Gap fistulous connection &gt; 1 cm</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>● Fistula on the edge of FOV</td>
</tr>
<tr>
<td>No. 100</td>
<td>61Y, M</td>
<td>2001</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>Indeterminate</td>
<td>T6 Left</td>
<td>● Data not available</td>
</tr>
<tr>
<td>No. 105</td>
<td>70Y, M</td>
<td>2006</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>T8 or T10 Left</td>
<td>T6 Right</td>
<td>● Very small fistulous connection</td>
</tr>
<tr>
<td>No. 112</td>
<td>76Y, M</td>
<td>2002</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>T11-T12 Left</td>
<td>L2 Right</td>
<td>● Data not available</td>
</tr>
</tbody>
</table>

T2-HI; T2-weighted hyperintensity,
CO; conus involvement of T2-weighted hyperintensity
FV; Serpentine Flow Voids
GE; (patchy) gadolinium enhancement
HC; high confidence on MRA for SDAVF
Gap; distance between arterial feeder and draining vein i.e. fistulous connection below spatial resolution on MR angiography.
Location of SDAVF

When incorrect regarding level of the fistula, the CE-MRA showed a maximal deviation of 3 vertebral levels. The highest level involved was C1/C2 (nerve root C2); the most caudal vertebral level was L4. In 28 cases (53%) the fistula was located on the right side, in 24 cases (45%) left sided. Multiplicity was seen in one case (2%), which was a bilateral fistula at level C1/C2 (congruent with the perceived level at CE-MRA however separate fistulas were not discerned at MRA). In 45 cases (85%) the fistula was located between T6 – L2 with a remarkable peak at T6 (18 fistulas, 34%) (Table 2).

In one case 3 CE-MR angiographies over different regions were performed because of a strong suspicion of the presence of a SDAVF but neither CE-MRA nor DSA could confirm the location of a SDAVF which was subsequently (on the 5th DSA) perceived to arise from the branches of the left internal iliac artery.

Conventional MRI findings

Complete data regarding the conventional MR findings of SDAVF at MR examination was available in 40 cases (75%). These findings include the presence or absence of:

1) T2-weighted hyperintensity
2) Extension of T2-weighted hyperintensity within the conus medullaris
3) Serpentine flow voids
4) Mild patchy cord enhancement.

Of the 40 SDAVF-cases where all conventional MR data was available, 35 cases (88%) were reported as positive for all four findings. In 4 cases (10%) there were 3 positive findings; 3 cases without extension of T2-weighted hyperintensity to the conus and one case without patchy gadolinium enhancement. These two findings were reported as negative in 1 out of 40 cases (2%), resulting in total 2 positive findings. In all 10 SDAVF-cases missed or mislocalized on CE-MRA the conventional MR findings could be fully retrieved (Table 1). All findings regarding conventional MR imaging, including the non-reported/non-retrievable cases, are depicted in Figure 1.

Treatment

In the 53 patients with SDAVF confirmed at DSA, 39 underwent attempted (74%) endovascular embolization. Embolization was initially angiographical successful in 20 cases when attempted (51%). In the remaining 19 cases (49%) embolization failed due to technical difficulties, procedural complications or there appeared to be an incomplete occlusion on post embolization DSA. These patients received subsequently secondary surgery shortly after the procedure. Fourteen of

![Figure 1: Conventional MRI findings of all 53 SDAVF-cases.](image)
the 53 patients (26%) were initially not amendable for endovascular treatment during the diagnostic procedure and underwent conventional surgical ligation.

**DSA procedural time**

The mean total time of the DSA procedure was available in the majority of cases. In the endovascular treated group (20 procedure times reported) the mean time was 198 minutes, in the combined treated group (15 reported times) 172 minutes and in the surgery group (11 reported times) 143 minutes. It should be noted that retrospective delineation could not be made confidently between the diagnostic and interventional portions of each procedure.

**Non-SDAVF-group**

Seventeen of the 70 cases (evaluated for SDAVF with CE-MRA and DSA) were not found to be a SDAVF at DSA. These 17 consisted of 7 negative DSA’s, as predicted by a (very) low confidence in 5 on MRA, and the remaining 10 cases were shown to be other forms of vascular malformations i.e. 4 cases of a perimedullary (or pial) fistula, 4 cases of a perimedullary nidal arteriovenous malformation (AVM) and 2 cases of an epidural fistula. The levels and side of the malformations are shown in table 3.

CE-MRA predicted the correct level and side in 6 of these 10 variant cases (60%, 95% confidence interval (CI) 0.31- 0.83). In 7 of 10 of these variant cases (70%) the CE-MRA report suggested a non-SDAVF malformation. The missed or mislocalized cases are shown in table 4.

<table>
<thead>
<tr>
<th>Table 3. Location of the variant forms of vascular malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Epidural type</strong></td>
</tr>
<tr>
<td><strong>Perimedullary type</strong></td>
</tr>
<tr>
<td>Costocervical</td>
</tr>
<tr>
<td>S2</td>
</tr>
<tr>
<td>Filum Terminale</td>
</tr>
<tr>
<td><strong>Perimedullary AVM</strong></td>
</tr>
<tr>
<td>T9</td>
</tr>
<tr>
<td>T12</td>
</tr>
<tr>
<td>L1</td>
</tr>
</tbody>
</table>

**Table 4. The non-SDAVF group, the missed and mislocalized cases.**

<table>
<thead>
<tr>
<th>Patient-number</th>
<th>Age, Sex</th>
<th>Year of study</th>
<th>MRI findings</th>
<th>HC on CE-MRA</th>
<th>MRA level fistula</th>
<th>Reason of misses/misinterpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. 52 52Y, F</td>
<td>2010</td>
<td>N / N / Y / N</td>
<td>Y</td>
<td>Indeterminate</td>
<td>Pial AVF</td>
<td>Gap fistulous connection &gt; 1 cm</td>
</tr>
<tr>
<td>No. 56 62Y, M</td>
<td>2009</td>
<td>Y / Y / Y / Y</td>
<td>Y</td>
<td>SDAVF T10-T11 Left</td>
<td>Pial AVF S2 Right</td>
<td>Increased vascularity seen at T10-T11</td>
</tr>
<tr>
<td>No. 104 57Y, F</td>
<td>2003</td>
<td>Y / Y / Y / N</td>
<td>Y</td>
<td>Pial AVF or SDAVF, level indeterminate</td>
<td>Pial AVF Filum terminale</td>
<td>Region was correct predicted by detecting a prominent vein (dural sac)</td>
</tr>
<tr>
<td>No. 106 21Y, M</td>
<td>2007</td>
<td>Y / N / Y / ?</td>
<td>Y</td>
<td>Pial or Dural AVF or Pial AVM T11-T12</td>
<td>Pial AVM L1 Left</td>
<td>Low lying AVM (out FOV)</td>
</tr>
</tbody>
</table>

T2-HI; T2-weighted hyperintensity, CO; conus involvement of T2-weighted hyperintensity 
FV; Serpentine Flow Voids 
GE; (patchy) gadolinium enhancement 
HC; high confidence on MRA for SDAVF 
Gap; distance between arterial feeder and draining vein i.e. fistulous connection below spatial resolution on MR angiography
Conventional MRI findings in the non-SDAVF group

Conventional MR imaging was also evaluated in this variant group. In the perimedullary fistula group as in the perimedullary AVM group 3 out 4 cases showed T2-weighted hyperintensity extending to the conus (75%). All 8 of these cases showed (100%) serpentine intradural flow voids. In 2 cases of each of these groups (50%) gadolinium enhancement of the cord was reported. All four findings were positive in the 2 epidural cases.

In the 7 DSA-negative cases was in 3 (43%) T2-weighted hyperintensity, in 2 (29%) conus involvement, in 4 (57%) serpentine flow voids and in 2 cases (29%) gadolinium enhancement seen on conventional MR imaging.

Treatment in the non-SDAVF group

Of the 17 patients in the non-SDAVF-group, 5 (29%) received endovascular embolization and one of those 5 needed secondary surgery because the endovascular treatment was not successful (20%). In 4 cases (24%) direct surgical treatment was performed. Eight cases (47%) received no treatment because there was no fistula (7 cases) or the patient was asymptomatic (1 case).

DSA procedural time in the non-SDAVF group

The mean total time of the procedure was in the endovascular treated cases (2 epidural fistula and 2 perimedullary fistula cases) 196 minutes, in the one combined (endovascular and surgery) treated case 141 minutes and in the surgery treated group (2 AVMs and 2 perimedullary fistulas) 208 minutes. The mean time in the 8 cases without treatment was 133 minutes.

Complications DSA

Of 70 procedures there were in 5 cases (7%) complications reported which resulted in early abortion of the DSA. All of them occurred during the therapeutic portion of the DSA. In 3 cases (4%) a dissection was seen and in 2 cases (3%) evident vasospasms. Except one, all cases were without post procedural consequences. One patient that showed vasospasms on DSA had significant neurological deterioration after the procedure. Spinal-CT showed embolic material in the anterior spinal artery as a consequence of the endovascular treatment. The patient was referred to a revalidation center and after months of revalidation he gained significant neurological recovery.

Time-interval CE-MRA and DSA

In 64% of the 70 cases the patient underwent consecutive MR, CE-MRA and DSA, within a 2 week period. In the remaining 36% the time interval was quite variable due to perceived medical urgency and ranged from 16 to 333 days, of these in 66% the interval was less than 60 days. Clinical considerations which resulted in a prolonged interval included: an asymptomatic or minimally symptomatic patient or a patient with stable neurologic condition.
5. Discussion

This study gives further insight in the accuracy of CE-MRA in detecting and localizing SDAVFs, with DSA as a reference standard. The sensitivity in predicting the correct level of the SDAVF after CE-MRA in our case series was 81%, 95% confidence interval (CI) 0.69 - 0.89. CE-MRA was also correct in the determination of other types of fistulas in 60%, 95% confidence interval (CI) 0.31 - 0.83.

Conventional MR imaging

Conventional MR imaging is obviously useful in the diagnosis of a SDAVF. However, conventional MR imaging is not the conclusive examination, as shown in our results it yield a strong suspicion on a SDAVF when the typical signs are present. Also, it has the additional advantage of excluding other, more common causes of myelopathy. In our series we could retrieve data regarding stated presence or absence of serpentine flow voids on conventional MR imaging in 50 of 53 cases, which were all positive (100%). This was the most consistently identified finding of SDAVF in our patient population. The next most consistent finding was cord T2 hyperintensity (96%) with extension to the conus in 85%. This is in agreement with previously reported experience15-17. Muralidharan et al. reported that of 141 cases 131 (93%) had T2-weighted hyperintensity that extended to the conus medullaris in 95%. In 93 cases they could retrieve data about vessel abnormality, which was seen in 78 cases (84%). Toossi et al. published similar results and found from a series of 36 cases in 89-92% T2-weighted hyperintensity and flow voids in 81%. When those findings combined, at least one typical finding was found in 100% of the cases. Based on their results, they stated that if both findings are absent then a symptomatic spinal fistula can be effectively ruled out. It should be noted that these authors also report cases of SDAVF where tortuous intradural flow voids are not present. In those papers images were reviewed from 198515, 17 and 199516. The latter did neither separate dural, perimedullary and epidural fistulas into subgroups. It is possible for these reasons that serpentine flow voids were not seen in 100% of cases. On the other hand, normal findings should not be confused for serpentine flow voids. It has been described that enhancing (normal) vessels may be prominent, however can be differentiated from abnormal dilated vessels. Normal perimedullary veins are usually more vertical oriented and less tortuous15. A second pitfall is that pulsation artifacts can be misinterpreted as flow voids4, 23. Lack of other MR findings further argues against the presence of a SDAVF.

Two cases of SDAVF in our series did not show the usual T2 weighted hyperintensity within the cord and these patients may be thought of as having less likelihood of harboring a SDAVF15 nonetheless in both of these cases ‘SDAVF-like’ serpentine flow voids were present which lead to the suspicion of a SDAVF and subsequent CE-MRA. In both of these cases the CE-MRA indicated the diagnosis and depicted the correct location of the fistula (Figure 2). When T2-weighted hyperintensity is absent, neurological symptoms are usually mild17. In our series both patients did not suffer from neurologic symptoms.

Mild patchy spinal cord enhancement was the least consistent of the MR findings, reported in 41 cases from which 38 were positive. The three cases without spinal cord enhancement had recently revealed symptoms (< 4 months). Its presence does indicate a disrupted blood-cord barrier and serve to reinforce the suspicion of SDAVF. It is usually seen 40 to 45 minutes after administration of gadolinium16, 18. The short time of symptoms might indicate that disruption of the blood-cord barrier occur later on in the progression of the disease.
Use of CE-MRA

In our series CE-MRA strongly supported the diagnosis in all cases of spinal vascular malformation and demonstrated an accuracy of 81% for identifying the level and side of the SDAVF. Aiding the diagnosis of SDAVF in this manner does not obviate the need for confirmatory DSA but facilitates the DSA and has been shown to decrease the time and volume of contrast material required to complete the diagnostic procedure\(^{25}\). The exact decrease in diagnostic DSA procedural time could not be measured in our series because distinction between the diagnostic and therapeutic portion was not possible. Furthermore, comparison could not have been made, because cases without CE-MRA were excluded in this study. However, in our center and others, CE-MRA enables an attempted endovascular embolization during the same procedure what implies a substantial decrease in time of the confirmatory diagnostic portion of DSA\(^{19,20,24}\).

With CE-MRA the fistulous connection can be traced by following the feeding intercostal/radiculomeningeal artery to the fistula, which is most often located in the dura near the root sleeve where the dilated medullary vein connects upstream to the intradural engorged venous plexus. The small size of these structures and the typically slow flow through these fistulas render other forms of MRA (Time-Of-Flight and Phase-Contrast) impractical\(^{19-21}\). Temporal and spatial resolution will continue to improve as MR systems continue to evolve, this enables more accurate tracing of these structures. Nowadays fast-gradient-echo CE-MRA performed at the most recent MR systems may detect vessels with a minimal diameter about 0.5mm\(^{20,26,42}\). Nonetheless, the fistulous site itself is still commonly difficult to visualize with the current CE-MRA technique. McCutcheon et al. found that the true fistulous site in the dura is micro vascular with multiple tangled and looped

Figure 2a, b, c and d: a. Sagittal T2-weighted image shows no T2-weighted hyperintensity within the cord however small foci of hypointensities along the dorsal aspect of the cord are present representing serpentine flow voids (white arrows) b. Sagittal CE-MRA image shows enhancement of the engorged medullary veins (white arrows) c. Coronal Multi Planar Volume Reformatting (MPVR) MIP image of the CE-MRA shows on the right T4 level ‘hazy enhancement’ of the arterial radiculomeningeal feeder (white arrowhead) and fistulous site (black arrowhead) draining into the medullary vein (white arrow) d. Anteriorposterior view of DSA following contrast injection of the right T4 intercostal artery (asterix). There is opacification of the radiculomeningeal artery (white arrowhead) and a fistulous connection (black arrowhead) with early venous filling into the medullary vein (white arrow).
vessels that individually are beyond the spatial resolution capabilities of those current MR systems.

Comparison of the CE-MRA and DSA images in cases of correctly localized SDAVs showed that when the fistula was ‘visible’ at CE-MRA, it actually appeared as a short hazy segment of enhancement (‘smudge’) that connected the medullary vein to the feeding artery. The smudge likely represents the multiple tangled, tortuous microscopic vessels that provide enough enhancement to overcome background noise and provide the continuity between the larger vessels even though the individual small vessels are not seen discretely (figure 3). In many cases where the fistula (‘smudge’) itself was not seen, the prediction of the location of the fistula could be made by inference i.e. the small short ‘signal gap’ between the arterial feeder and medullary vein was the actual site of fistula. This gap was, in these cases, quite small (several mm at most) this method of fistula localization by inference was further facilitated if the course of the feeding artery and the medullary draining vein were in line.

Retrospective analysis of the ‘cases of SDAVF missed or mislocalized’ (CMMs) together with a neuroradiologist experienced in spinal MRA for 20 years (Dr. R.I. Farb) revealed that in the majority of CMMs the reason for the ‘miss’ could be attributed to the ‘gap’ being too large (1>cm) moreover, in these missed cases, the feeding artery and medullary vein were commonly not aligned (figure 4). Spatial resolution remains the limiting factor in these cases (table 1 and 4).

Although in 10 cases the CE-MRA was inconclusive or incorrect in localizing the SDAVF, the exam was still useful in the diagnostic process. In one of these CMM the CE-MRA was severely degraded by motion artifacts, in this case and all other CMMs the CE-MRA showed a high confidence for the existence of a SDAVF because of demonstrated early venous reflux into abnormal dilated vessels around the spinal cord.
Other potential causes for mislocalization of SDAVF at CE-MRA include; miscounting of the vertebral levels (which we attempted to correct for in this study) leading to incorrect reporting of the level of the fistula and the occurrence of the SDAVF located outside the chosen FOV of the CE-MRA. We tried to minimize this by using a large FOV to include at least T2 to L2. Also, when the conventional MR suggested the exciting lesion lies within the cervical or lumbar region we appropriately altered the FOV to include that area understanding that (not withstanding) the location of the fistula and the cord edema are not necessarily congruent. Authors have acknowledged this FOV issue. In an initial experience of this CE-MRA technique published in 2002 by Farb et al. 5 of 9 patients had to be reexamined because the location of FOV (36 cm) was – based on study of Aminoff et al. caudally aligned and included approximately T8 to S1 what explained the high number of initially missed cases. In a study of Saraf-Lavi et al. the location of FOV (26 cm) was chosen based on clinical findings and encompassed between 7 and 11 vertebral levels. In their series, 5 of 20 cases were missed because the level of the SDAVF was not included in the FOV. Luetmer et al. used a FOV of 28 cm and mislocalized 6 of 31 SDAVFs. After five missed cases they overcame the FOV limitation by using two overlapping acquisitions which allowed coverage from C2 to S1. Mull et al. used a novel technique of CE-MRA combining various advancements in MR angiography, including a phased array spine coil with a FOV of 50 cm. In this series, only one tentorial SDAVF was missed out of 19 true SDAVF cases.
Location of SDAVFs
In our study 84% of all SDAVFs the location was found between T6 and L2. This is in agreement with earlier reports\textsuperscript{2, 4}. Van Dijk et al. found a predominance of 70% on the left side, in contrary to a slight predominance for the right side (53%) in our study. The reason for the peak of 34% of SDAVFs at T6 in our study remains unexplained.

Differentiation between types of vascular malformations
In this series there were 10 cases of spinal vascular malformations which at DSA were shown not to be SDAVFs. Seven of these 10 cases (70%) were correctly questioned as variant (non-SDAVF) malformations at CE-MRA. In the remaining 3 cases (30%) a SDAVF was suspected but appeared on DSA to be a perimedullary fistula in 2 cases and an epidural fistula in one case. Differentiation between SDAVF and other malformations can thus be estimated but remains challenging. At best distinguishing between different types of spinal vascular lesions requires a thorough knowledge of the anatomy and the pathophysiology of the various types of spinal vascular malformations\textsuperscript{13, 30, 40}. Nonetheless differentiation limitations at CE-MRA is possible in a minority of cases and the reader must remain cognizant of this possibility\textsuperscript{26}.

Epidural (or extradural) fistulas are located outside the dura mater and are associated with an engorged epidural venous plexus (apx II P,Q) what may cause spinal cord and/or nerve root compression. On conventional MR imaging these lesions typically appear as extradural enhancing masses (flow voids) representing the engorged epidural venous plexus. The manifestation of this type of fistula depends on the transdural ‘valvelike’ anti-reflux system. When dysfunctional, the high pressure in the epidural veins may cause reflux or stasis in the intradural venous system and radiological findings may be similar from those seen in SDAVFs\textsuperscript{8, 13}. Perimedullary (or pial) arteriovenous fistulas are usually located on the ventral pial surface of the spinal cord. There is a direct connection via one or more arterial feeders arising from the ASA (or in some cases from the PSA’s) (apx I Q.T) to the vein, resulting in an engorged venous network. Gradation in type is been made based on the number of arterial feeders and the size and flow of the fistula. Non-invasive imaging findings are similar to those in SDAVFs, but in perimedullary fistulas are serpentine flow voids rather seen ventrally along the spinal cord. Subarachnoidal hemorrhage (SAH) may occur due to venous rupture, especially in high-flow fistulas.\textsuperscript{8, 13} Distinction between a perimedullary fistula and a SDAVF can be made by determination of the arterial feeder(s) at DSA. Perimedullary fistulas are fed by radiculomedullary arteries (connected to the ASA) which have a typical ‘hairpin’ loop after piercing the dura\textsuperscript{33, 35, 36, 39}. SDAVs are supplied by radiculomeningeal arteries which have commonly a tortuous, horizontal course. However SDAVs can have anastomoses with perimedullary arteries, which then opacify in these cases also at DSA. Spinal AVMs can be located intramedullary, on the surface of the spinal cord or in the epidural space (or at a combination of these locations). Patients harboring a spinal AVM usually present with a SAH and acute clinical deterioration. However symptoms can be similar to a SDAVF, the acute clinical presentation of a SAH is rarely confused with a SDAVF. On conventional MR imaging a nidus is seen and there may be serpentine flow voids, especially around the nidus\textsuperscript{8, 13}. In our series all four AVMs were correctly diagnosed at non-invasive examination.

DSA-negative cases
In 7 cases DSA did not demonstrate a spinal vascular malformation, in 5 of these (71%) CE-MRA was reported as having a very low suspicion for a SDAVF i.e. none or very subtle enhancement of the spinal veins. Also, conventional MR images were not convincing for a SDAVF in these 5 cases and other diagnoses were considered. In these cases, DSA was performed to further exclude a vascular lesion. In the remaining two cases however the conventional MR images as well as the CE-MRA supported the presence of a SDAVF. In one case the suspicion was only moderate as there was no T2-weighted hyperintensity within the cord and there were mildly enlarged intradural serpentine
vascular structures on CE-MRA (Figure 5a and b). In the remaining case the MR showed cord T2-weighted hyperintensity and CE-MRA showed serpentine flow voids thought to be typical of SDAVF however DSA performed on two separate occasions failed to identify a vascular abnormality. This patient’s lower extremity weakness improved and was subsequently lost to follow-up (Figure 5c and d).

**Figure 5a and b (patient 1), c and d (patient 2):** a. On sagittal T2-weighted image no T2-weighted hyperintensity is seen within the spinal cord; at T9-T11 subtle hypointensity is seen i.e. suspected flow voids posterior to the cord (white arrow). b. Sagittal contrast enhanced T1-weighted image (magnified) shows local enhancement of the medullary veins from T9-T11 (white arrows). c. Sagittal T2-weighted image shows central T2-weighted hyperintensity within the lower spinal cord (black arrows). d. Sagittal (upper image) and axial (lower image) Maximal Intensity Projection (MIP) images of CE-MRA show enhancement of the dorsal medullary veins (white arrow on upper image) and the suspected fistulous connection, not confirmed at DSA (not shown), at T11 (black arrowhead on lower image).

**Limitations**

To our knowledge this is the largest series examining the utility of CE-MRA in the diagnosis of SDAVFs with reference DSA. Limitations of the current study include shortcomings of the MRA technique itself, specifically: the FOV limitations as described above; spatial resolution limitations, also described above, which contribute to the CMMs; and the use of 1.5T imaging system rather than a 3T imager. Earlier reports suggest that 3T systems may be advantageous compared to 1.5T systems in the depiction of the angio architecture of fistulas owing to the available increase in signal-to-noise ratio (SNR) which could be reinvested in improved spatial resolution. Vessels measuring 0.2 – 0.3 mm in diameter could be depicted in 3T systems. In reality however, spinal imaging at 3T has been hindered by field inhomogeneity, the scan time requirements as well as specific absorption rate (SAR) limitation issues thus negating some of the perceived advantages at 3T. Few successful experiences are published with a 3T system, but none of them showed a comparison or direct benefit from images obtained at 3T system.

The technique of CE-MRA evaluated here employs a two phase technique i.e. an ‘arterial phase’ and a ‘blood-pool phase’ acquisition. The ‘phase’ properties of the images are conferred from the first pass – centrically encoded collection of data where the center of K-space (data field of the MR
images) is collected following a 4 seconds inserted delay and continues from low frequency data to high frequency data over the ensuing 147 seconds yielding an arterial phase image. The data set is reacquired at 147 seconds to collect the ‘blood-pool’ data. Other authors have emphasized use of a multiphase technique of CE-MRA to discern multiple sequential arterial through venous phases which they believe may help in localizing the SDAVF. The TWH/UHN intentionally does not pursue a multiphase technique for the evaluation of SDAVF and chose rather to maximize the spatial resolution ability of the CE-MRA over the temporal resolution.

A further limitation of our study was the lack of information provided by CE-MRA regarding additional arterial feeders of the SDAVFs and the location of the Adamkiewicz artery (AKA). These structures are essential in the decision for treatment but are invariably not discernable using the technique of CE-MRA described here. Difficulty visualizing these structures has been experienced by other authors.

The series of patients discussed here suffers from selection bias in that the prevalence of SDAVF in the study population was obviously very high. DSA was not routinely performed in patients thought to be frankly negative for SDAVF at both conventional MR and CE-MRA. This is justified given the additional lack of compelling clinical findings commonly encountered in these ‘negative’ patients. Angiography in this negative population would be inappropriate. Thus the true sensitivity of CE-MRA for SDAVF has not been rigorously addressed.

Clinical implications
When there is a clinical and/or radiological suspicion of a SDAVF or other type fistula, CE-MRA should be performed. Conventional MR imaging combined with the results of the CE-MRA gives reliable information about the diagnosis and correctly determines in a high number of cases the location of the fistula. Referrals from peripheral centers can be from higher quality when CE-MRA is performed and repeated non-invasive examination may be avoided. This may also have a positive effect on the time-to-diagnose and the time-to-treat.

Based on our data our current recommendation is to strategically place the FOV of the CE-MRA such that it includes at a minimum T4 to L2 unless there is compelling clinical and/or radiological evidence to shift the FOV cephalad or caudad.

In cases where both conventional MR imaging and CE-MRA are negative, a symptomatic SDAVF is excluded and DSA can be avoided. When at non-invasive imaging a fistula is suspected and the location is predicted, CE-MRA can be used as guidance for selective DSA to avoid investigating the whole spinal cord. DSA is in this manner more time-efficient and an attempt to treat with endovascular embolization during the same procedure is now part of our standard procedure at our center.

CE-MRA facilitates but does not obviate confirmative diagnostic DSA. DSA is also still inevitable for identifying the exact angio architecture of the fistula and for the determination of possible additional feeders or anastomoses with the normal spinal vasculature. How convincing CE-MRA can be, the location and type of fistula can still be misinterpreted and DSA can alter the definitive diagnosis or choice of treatment.

Avoiding diagnostic DSA in both negative conventional MR imaging and CE-MRA and the use of CE-MRA as guidance during diagnostic DSA in suspicious cases implies a reduction of procedural complications. However conventional diagnostic DSA might be a relative safe examination, there is always the risk of vasospasms, dissections and plaque embolization, especially in the elderly people with severe atherosclerosis and more fragile vessels.
6. CONCLUSION

Serpentine flow voids are the most consistent finding at conventional MR imaging of SDAVFs, accompanied by T2-weighted imaging in almost all cases. CE-MRA can be expected to correctly localize the site of the SDAVF in over 80% of cases. Also in the determination of other types of fistulas CE-MRA can be helpful. This study provides further insight into the accuracy and role of CE-MRA in the work-up of patients with suspected SDAVF and why localization of the fistula may be incorrect. When there are no typical findings on conventional MRI and CE-MRA, the existence of a symptomatic fistula is effectively excluded and DSA can be avoided. CE-MRA facilitates but does not replace diagnostic DSA in cases of SDAVF as the definitive test specifically confirming location, type of fistula and arterial detail which are required for treatment decisions.
7. References


Achtergrond en doel: Een Spinale Durale Arterioveneuze Fistula (SDAVF) is een spinale vasculaire aandoening die lastig te diagnosticeren is. Vaak wordt de diagnose pas gesteld als er al vergevorderde neurologische problemen bestaan. Radiologisch onderzoek speelt een sleutelrol in het stellen van de diagnose. Conventioneel MR onderzoek kan het vermoeden doen rijzen op het bestaan van een SDAVF. Contrast-enhanced (contrast-geïntensiveerd) MR angiografie (CE-MRA) kan bijdragen in het opsporen van een SDAVF en kan tevens de meest waarschijnlijke locatie van de fistula bepalen voordat de patiënt een uitgebreide digitale subtractie angiografie (DSA) ondergaat. De afgelopen jaren zijn een aantal artikelen gepubliceerd over het gebruik van CE-MRA in het opsporen van SDAVFs. Echter, al deze onderzoeken zijn initiële ervaringen of bevatten kleine patiëntenaantallen. Wel blijkt uit deze onderzoeken al dat het gebruik van CE-MRA nut kan hebben voor de DSA. Uitgebreid onderzoek van alle spinale arterien middels DSA kan worden voorkomen en alleen selectieve DSA hoeft te worden uitgevoerd door CE-MRA als leidraad te gebruiken. Het doel van deze studie is het vaststellen van het nut en het bepalen van de accuraatheid van contrast-enhanced MR angiografie in het vaststellen van Spinale Durale Arterioveneuze Fistulas.

Materiaal en Methode: In het Toronto Western Hospital/University Health Network (TWH/UHN) zijn middels een retrospectieve analyse vanaf 1999 tot en met 2012 in totaal zeventig patiënten geïdentificeerd die klinisch verdacht werden op het hebben van een SDAVF en hiervoor conventioneel MR onderzoek, CE-MRA en DSA hebben ondergaan. In dit onderzoek hebben we gekeken op de conventionele MR beelden naar de aan- of afwezigheid van serpentine flow voids (tortueuze en gedilateerde vaten), T2-gewogen hyperintensiteiten in het ruggenmerg en diffuse enhancement van het ruggenmerg na gadolinium toediening. Van de CE-MRA werd het vastgestelde niveau en de zijde van de fistula verzameld. DSA werd in deze studie gebruikt als referentie voor de werkelijke locatie van de fistula.

Resultaten: Van de zeventig geselecteerde casussen bleken er 53 een SDAVF te zijn. Van de overige 17 casussen waren er 10 die een ander type spinale vasculaire malformatie hadden en in 7 casussen was de DSA negatief. Op de conventionele MR beelden van de SDAVFs bleek in 100% serpentine flow voids aanwezig te zijn. T2-gewogen hyperintensiteit in het ruggenmerg werd gezien in 96% van de gevallen wat in 85% tot in de conus medullaris uitstrekte. Diffuse enhancement van het ruggenmerg werd gezien in 93%. CE-MRA localiseerde het niveau en de zijde van de SDAVF in 43 van de 53 casussen (81%).

Conclusie: CE-MRA is een bruikbaar niet-invasief onderzoek in het aantonen van het niveau en de zijde van een SDAVF. Als zowel conventioneel MR onderzoek en CE-MRA geen tekenen laten zien van een fistula kan een DSA worden vermeden. CE-MRA faciliteert het diagnostische deel van een DSA maar vervangt het niet. DSA blijft in deze gevallen nog steeds noodzakelijk om de specifieke locatie te bevestigen, het type fistula vast te stellen en de precise angioarchitectuur te identificeren om tot een juiste behandeling te komen bij deze leasies.
APPENDIX I

SPINAL ARTERIAL VASCULATURE

A: aorta
B: intercostal or lumbar artery (segmental artery)
Ba: intersegmental anastomosis
C: prevertebral anastomotic network
D: vertebral body feeding arteries
E: dorsal spinal artery
F: intercostal/muscular artery
G: pretransverse anastomotic network
H: dorsal division of dorsal spinal artery
I: post transverse anastomotic network
J: muscular branches of the post transverse anastomotic network
K: ventral division of the dorsal spinal artery
Ka: radicular artery
La: ventral epidural arcade
Lb: dorsal epidural arcade
M: radiculomeningeal artery (in nerve root sleeve)
N: radiculomeningeal artery (dural branch)
O: radiculopial artery
P: radiculomedullary artery
Q: anterior spinal artery (ASA)
R: mesh like pial vessels arterial network
S/T: posterior spinal artery (PSA)
U/V: pial arterial network (vasacorona)
W: sulco commissural artery
X: rami perforantes of the peripheral (centripetal) system
Y: central (centrifugal) system of sulco commissural arteries
Z: rami cruciantes

Schematic drawing of the spinal arterial vasculature (derived from http://neuroangio.org)
APPENDIX II

SPINAL VENOUS VASCULATURE

A: centripetal network of veins
B: central (sulcal) veins
C: peripheral (radial) centrifugal veins
D: venous anastomosis between the centripetal and centrifugal systems
E: anterior (ventral) median vein
F: posterior (dorsal) median veins
G: transmedullary anastomosis between dorsal and ventral median venous systems
H: extrinsic surface anastomosis between dorsal and ventral median veins
I: vein of filum terminale
J: dominant radicular vein of the cauda equina
K: radiculomedullary (radicular) vein
L: nerve root sleeve
M: angular point of radiculomedullary vein piercing the dura of the nerve root sleeve
N: lumbar vein (segmental vein)
O: radiculomedullary veins of the cauda equina
P: anterior internal (epidural) vertebral venous plexus
Q: posterior internal (epidural) vertebral venous plexus
R: ascending lumbar vein
S: basivertebral vein
T: intravertebral body venous plexus
U: anterior external vertebral venous plexus
V: posterior external vertebral venous plexus

Schematic drawing of the spinal venous vasculature (derived from http://neuroangio.org)
APPENDIX III

RESEARCH ETHIC BOARD APPROVAL LETTER

University Health Network
Research Ethics Board
10th Floor, Room 1056
700 University Ave
Toronto, Ontario, M5G 125
Phone: (416) 581-7849

Notification of REB Approval for Access to Retrospective Data for Research Purposes

Date: June 13th, 2012
To: Dr. Richard Farb
Rm 3MCL430, 3rd Floor, McLaughlin Pavilion, Toronto Western Hospital, 399 Bathurst St.
Toronto, Ontario, Canada M5T 2S8

Re: 12-5145-AE
Spinal Dural Arteriovenous Shunt: Accuracy of CEMRA for Identification and Localization

REB Review Type: Expedited
REB Initial Approval Date: June 13th, 2012
REB Expiry Date: June 13th, 2013

Documents Approved:
- Protocol Synopsis
  Received on: June 6th, 2012
- Data Collection Form
  Received on: June 6th, 2012


Best wishes on the successful completion of your project.

Sincerely,

Anna Gagliardi, PhD
Co-Chair, University Health Network Research Ethics Board