Guidelines to be followed by centres, services and units in order to be designated as Reference Centres, Services and Units of the National Health System, as agreed by the Interterritorial Board

48. CARE FOR SPINAL CORD VASCULAR PATHOLOGIES

Spinal cord vascular malformations (arterial and venous) represent a heterogeneous and rare group of vascular anomalies that affect the spinal cord and nerve roots either directly or indirectly, frequently causing acute, subacute and chronic myelopathies resulting from the compression and the hemorrhagic or ischemic complications of these injuries.

They are usually classified into:
1) Spinal aneurysms, which hare very rare;
2) Arteriovenous lesions, composed of arteriovenous fistulas – usually of acquired nature- and arteriovenous malformations – considered to be congenital.

The most complicated spinal cord vascular conditions to treat are the fusiform aneurysms of the Adamkiewicz artery, intradural dorsal and ventral arteriovenous fistulas with multiple feeding arteries, arteriovenous malformations located intramedullary or at the medullary cone, and mixed arteriovenous malformations extradural-intradural located.

Treatment for most of the spinal cord vascular malformations is provided through superselective catheterization and embolization of the malformation. Microsurgery may be chosen for certain cases or use to complement embolization in other cases. More recently, radiosurgery has been introduced to the treatment of spinal vascular malformations.

Given the rare frequency of these conditions, Reference Centres, Services and Units are required for better diagnosis and treatment of the most difficult cases.

A. Rationale for the proposal

► Epidemiological data on spinal cord vascular pathologies.	The incidence is estimated in 7-10 cases/million population and year. 30% of the cases correspond to intramedullary arteriovenous malformations (AVMs) and perimedullary

	arteriovenous fistulas (PAVFs) and 70% to dural arteriovenous fistulas (DAVFs) with perimedullary draining. There is no data on prevalence of these conditions. For DAVFs, which are 70% of these anomalies, the average age when clinical symptoms appear is 55 years, and the distribution in terms of sex for life expectancy is men/women: 4/1; hence, a maximum number of 175 patients per million population may be inferred, with only 25-30% of the cases being diagnosed. The rest of conditions (PAVFs and AVMs), the remaining 30%, have an average age at clinical presentation of 22 years, and average survival of 15, therefore the prevalence may be estimated in 45 patients per million population.
► Data on the use of diagnostic and therapeutic procedures.	Nowadays, in clinical practice 30-40% of the cases are diagnosed. Therefore, for all the population included in the National Health System, there could be 350-500 cases per year. Given the current conditions, in the best case scenario only 105-200 cases will be diagnosed. According to the Spanish Registry for Interventional Radiology, in 2007 76 spinal vascular malformations were treated with endovascular methods. There is not data on surgical treatment use.

B. Guidelines to be followed by Centres, Services and Units in order to be designated as Reference Centres, Services and Units for the care of spinal cord vascular pathologies

Experience of the Reference Centres, Services and Units: - Activity: • Number of procedures that should be performed in a year in patients with spinal cord vascular pathologies to ensure	 - An average of 5 selective medullary catheterization /year in the last 3 years. - An average of 5 intramedullary surgical procedures /year in the last 3 years.

an adequate care. - Other data: research on the subject, postgraduate teaching, continuing training, publications, etc.	 - Accredited postgraduate teaching: Unit participation in the internship and residency programme of the Centre. - Participation in research projects and publications in the field[a]. - Continuing training programme standardized and authorized by the centre's board of directors. - Clinical multidisciplinary sessions, at least once a month, in order to make decisions and coordinate treatments.
► Specific resources of the Reference Centres, Services and Units: - Human resources required for the adequate care of spinal cord vascular pathology. Basic education of the team members [b].	A coordinator for the unit is recommended. - Continuous care in the specialty areas integrating the multidisciplinary team, 24 hours a day, every day of the year. - Multidisciplinary care provided at least by: ▪ 2 neurologists. ▪ 2 radiologists, neurologist, or neurosurgeons with experience in vascular neorointerventional procedures. ▪ 2 neurosurgeons. ▪ Neurophysiologist. ▪ Nursing staff. - Neurologists with, at least, 2 year experience in spinal cord vascular pathology. - Radiologists, neurologists, or neurosurgeons with, at least, 5 year experience in vascular neorointerventional procedures. - Neurosurgeons with, at least, 5 year experience in spinal cord vascular pathology. - Neurophysiologist, with 5 year experience in spinal monitoring. - Nursing staff with experience in patients with spinal cord vascular pathology.

- Specific equipment required for the adequate care of spinal cord vascular pathology.	- Intravascular embolization materials (Onix, liquid adhesive, microcoils, expandable microspheres, etc.) Catheterization and microcatheterization materials required for closure of AVMs, PAVFs, and DAVFs, as well as for managing periprocedural hemorrhagic or thromboembolic complications. - Digital angiography room: ▪ Angiographic equipment with high resolution digital subtraction and fluoroscopic imaging, with capacity for roadmapping. Monoplane equipment, it is recommended the use of biplane equipment. Ability to perform rotational angiography with 3D image reconstruction programme. ▪ Monitoring system, anaesthetic and cardiopulmonary resuscitation equipment. ▪ Disposable material for selective catheterization and supraselective medullary microcatheterization ▪ Embolization material. - Operating theatre available for programmed neurosurgery, with at least: ▪ Adequate instruments for vascular and medullary microsurgery techniques: microdissectors, microscissors, thin aspirators, micro forceps for dissecting and biopsy, vascular clips and applicators, matching clips, temporal clips and microsurgery ports. ▪ Bipolar coagulator and radiofrequency coagulator available, with their own forceps. ▪ Surgical microscope and ultrasonic aspirator. ▪ Digital radiological equipment in operating theatre allowing performing angiographic techniques. ▪ Intraoperative doppler equipment available. ▪ 3D imaging manipulation equipment available. - Digital intraoperative monitoring system, with evoked potentials. - Outpatient care for follow-up of patients with complex spinal cord vascular pathology. - Hospitalization beds available for treatment of spinal cord vascular pathology.

► Resources from other units and services besides those belonging to the Reference Centres, Services and Units required for the adequate care of spinal cord vascular pathologies.	- Anaesthesia services/unit with experience in patients with spinal cord vascular pathology. - Intensive medicine services/unit with experience in patients with spinal cord vascular pathology. - Diagnostic imaging services/unit with experience in patients with spinal cord vascular pathology, at least having: ▪ CT scanner with multislice system and CT angiography programme. ▪ 1.5 Tesla MRI, with MR angiography programme. - Rehabilitation services/unit with experience in patients with spinal cord vascular pathology. - Radiosurgery services/unit available with experience in patients with spinal cord vascular pathology. - Research laboratory available for training with animals.
► Procedure and clinical results indicators of the Reference Centres, Services and Units [c]:	**The indicators will be agreed with the Units that will be designated.**
► Existence of an adequate IT system (Type of data that the IT system must include to allow identification of the activity and evaluation of the quality of the services provided)	- Filling up the complete MBDS of hospital discharge. - The unit must have a *registry of patients* with spinal cord vascular pathology which at least must include: - Medical record number. - Date of birth. - Sex. - Patient's habitual region of residence. - Admission date and discharge date. - Type of admission (Emergency, planned, other). - Type of discharge (Home, hospital transfer, voluntary, death, transfer to a healthcare centre, other.). - Service in charge of patient's discharge. - Main diagnosis (ICD-9-CM). - Other diagnosis (ICD-9-CM).

	- Diagnostic procedures provided to the patient (ICD-9-CM): Type of procedure and date when it was provided. - Therapeutic procedures provided to the patient (ICD-9-CM): Type of procedure, date when it was provided and results of the procedure. - Complications (ICD-9-CM). - Patient monitoring: Clinical results at discharge, six months and a year after. Neuroimaging follow-up with MRI six months after and if needed with follow-up medullary catheterization by indication of the neuroradiologist. The unit must have the required data which should be sent to the Spanish National Health Service Reference Centres, Services and Units Appointment Commission Secretariat for yearly reference unit monitoring.

[a] *To be assessed by the Appointment Commission.*
[b] *Experience will be accredited by certification from the hospital manager.*
[c] *Clinical results standards, agreed to by the experts group, will be assessed, initially by the Appointment Commission, while in the qualification process, as more information from the Reference Centres, Services and Units is being obtained. Once qualified by the Appointment Commission, the Quality Agency will authorize its compliance, as for the rest of guidelines.*

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