PROTOCOLE NATIONAL DE DIAGNOSTIC ET DE SOINS (PNDS) [NATIONAL DIAGNOSTIC AND CARE PROTOCOL]

SYRINGOMYELIA

Intramedullary clefts

Rare Diseases Referral Centre
SYRINGOMYELIA
Neurosurgery
Department Hôpital
Bicêtre

In consultation with:

The Hôpital Ambroise Paré Assessment and Treatment Centre for Pain

The Hôpital Tenon Perineal Neurology and Perineal Scan Department

The Hôpital Pellegrin-Tripode Bordeaux Department of Neurosurgery A, Adult and Paediatric

OVERVIEW

LIST OF ABREVIATIONS

CC	Chronic Condition
MA	Marketing Authorisation
APAISER	Association Pour Aider et Informer les Syringomyéliques Européens Réunis [Association to assist and inform European Syringomyelics]
ASIA	American Spinal Injury Association
BPI	Brief Pain Inventory
GH	General Hospital
UH	University Hospital
CRMR	Centre de Référence Maladies Rares [Rare Diseases Referral Centre]
EFT	Emotional Freedom Technique
QRST	Quantified Review of Sensitivities and Thresholds
PMSAS	Perimedullary Subarachnoid Space
VAS	Visual Analogue Scale
HAS	Haute Autorité de Santé [French Health Authority]
MRI	Magnetic resonance imaging
CSF	Cerebrospinal fluid
FIM	Functional Independence Measure
PMR	Physical Medicine and Rehabilitation
MRS	Modified Rankin Scale
NPSI	Neuropathic Pain Symptom Inventory
SEP	Sensitive Evoked Potential
PNDS	Protocole National de Diagnostic et de Soins [French National Diagnosis and Care Protocol]
QDSA	Questionnaire Douleur Saint Antoine [Saint Antoine Pain Questionnaire]
OTR	Osteo-Tendon Reflex
MS	Multiple sclerosis
TENS	Transcutaneous Electrical Nerve Stimulation

SUMMARY FOR THE ATTENDING DOCTOR

Syringomyelia is defined by the presence of intramedullary fluid formations distributed over at least 2 myelomeres and composed of a liquid in all respects identical to cerebrospinal fluid (CSF). But not all cystic intramedullary images are syringomyelia, which is a rare, chronic disease and most often develops into a severe neurological deficit and dysfunctions by chronic neuropathic pain. Nowadays, given the availability of MRIs, we are seeing more and more patients with a more or less extensive intramedullary cavity image discovered either accidentally or after various clinical signs, including more or less intense pain. These are either intramedullary syringomyelic clefts (subject of this PNDS) or dilation (persistence) of the centro-ependymal canal. The crucial issue is being able to distinguish a real syringomyelia from a dilatation of the ependymal canal and, where appropriate, identifying the origin of the symptomatology in this image in order to adapt care.

Clinical, electrophysiological, urological and psychological examinations are essential tools both to assess the severity and stage of the illness and to determine scalability as well as its regularity to more effectively adapt therapeutic management.

As soon as the MRI diagnoses syringomyelia, cleft or dilation of the ependymal canal, the patient must be referred to a Specialised Centre to refute or confirm the diagnosis and establish follow-up and treatment. Follow-up is shared with the attending doctor, the neurologist, and the pain-treatment centre. Appropriate clinical and radiological monitoring is essential to determine whether this cleft is becoming a progressive syringomyelia, and thus changing category. (Specific PNDS to be published later).

The medical treatment is established by the multidisciplinary team. Syringomyelic clefts generally do not require surgical treatment. The Bicêtre Referral Centre supports these patients in consultations, day hospitalisation and can give advice by email, telephone, remote transmission (e.g. ORTIF), mail etc.

Useful contacts for the attending doctor:

CRMR Syringomyelia at Hôpital Bicêtre:
Referring Physician: Dr. Silvia MORAR
Telephone: 01 45 21 24 55
Email: sylvia.morar@aphp.fr

Coordinating doctor: Pr. Fabrice PARKER
Telephone: 01 45 21 23 80

Secretariat: Marie Annick HUIN
Telephone: 01 45 21 24 55
Fax: 01 45 21 26 00

Consultations:
Telephone: 01 45 21 22 88
Email: consultation.neurochirurgie.bct@aphp.fr

Useful websites:

Internet links:

www.syringomyelie.fr
www.maladiesrares-paris-sud.aphp.fr
www.orphanet.fr

Related sites:

www.neurosphinx.fr
www.apaiser.asso.fr
www.hopital-necker.aphp.fr/marep
www.spinareference.org
www.amcvhs.com

1. INTRODUCTION

SYRINGOMYELIA

Syringomyelia is a rare disease characterised by the abnormal formation of one or more cavities inside the spinal cord. This cavity can develop and increase progressively, leading to lesions of the spinal cord. The incidence of syringomyelia is 8.4 per 100,000 in western countries and 1.94 in Japan. This difference is not explained so far. Various symptoms may appear, but they vary greatly from one patient to another. The neurological examination most frequently shows damage to the spino-thalamic sheaths with a suspended thermo-algic syndrome, neuropathic-type pain, ataxia, or, in cases of extension to the anterior cords, motor impairment, sphincter and sexual function disorders, neurovegetative disorders (e.g. sweating).
The term "syringomyelia" was used to describe any longitudinal collection filled with fluid inside the spinal cord (only accumulations outside the centromedullary canal).
The known pathological situations that can contribute to the formation of syringomyelia are the malformations of Arnold Chiari and other malformations of the cervico-occipital joint, spinal cord injuries, tumours, meningeal haemorrhage, meningitis, dysraphisms. Idiopathic syringomyelia refers to the category in which no cause is identified.
The mechanism responsible for the accumulation of liquid significantly varies and a great ease of understanding is given by a first classification in:

1. Communicating syringomyelia, foraminal (Chiari, dysraphism)
2. Non-communicating syringomyelia
   (tumour, arachnoiditis, post-traumatic or post-infectious).
3. Idiopathic syringomyelia where no pathological entity is found.

The fluid accumulations that start in the centromedullary canal (hydromyelia) may be responsible for outbreaks of medullary white matter in areas of weakness. A lesion of the ependyma allows the syringomyelia cavity to expand without constraint. The spinothalamic tract that intersects at this level is particularly sensitive and vulnerable.
Due to the availability of MRI, we often notice the discovery of intramedullary cavities called intramedullary clefts (syringomyelic, fine, intramedullary cavities), especially in paucisymptomatic patients. There is no clear consensus on the incidence of clefts.

INTRA-MEDULLARY CLEFTS

Syringomyelia can present different morphological as well as clinical aspects.
The entity "intramedullary clefts" are small volume syringomyelic cavities that can evolve slowly and progressively. To differentiate from
"dilations of the centromedullary canal" which are an anatomical variant to

usual. In both cases, the clinical symptomatology is poor and does not, generally speaking, contain objective evidence. This growing number of patients poses both specialists and doctors with the problem of a definitive diagnosis and therapeutic management. The central issue is distinguishing true syringomyelia from a cleft and, if necessary, blaming the symptomatology. A thorough clinical and para-clinical scan, including MRI fibre tracking, would separate the true syringomyelic (possibly evolutive) clefts from the ependymal channel residues and predict their progression as well as the most appropriate care. One of the research axes of the Coordinating Centre (ongoing project) is determining the incidence of clefts and their clinical and radiological progression. In these cases, we note a preponderance of painful symptomatology with a neurological examination close to normal. Neurological deficits are rare. Differentiation between a progressive intramedullary cleft and an anatomical variant to normal (visibility of the centromedullary canal) is essential to guide and adapt pain care for these patients. The differential diagnosis is made by a multidisciplinary team consisting of neurosurgeons, neuroradiologists, pain doctors. It is never a matter of emergency.

SYRINGOMYELIA:

Intramedullary cavity slowly and progressively developing into a neurological disability

VISIBILITY OF CENTRO-MEDULLARY CHANNEL:

Anatomical variant to normal

INTRAMEDULLARY CLEFT:

Potentially progressive syringomyelic cavity

2. OBJECTIVES OF THE NATIONAL DIAGNOSTIC AND CARE PROTOCOL

The objective of the National Diagnosis and Care Protocol (PNDS) is to explain to healthcare professionals the optimal care and care for an intramedullary cleft patient.

The PNDS aims to homogenise initial care, and check-ups for adults to improve quality of life for patients and their family. The PNDS cannot, however, consider all specific cases, all comorbidities, all therapeutic particularities, hospital care protocols, etc. It cannot claim completeness of possible support lines, or replace the individual responsibility of the doctor vis-a-vis their patient. This protocol, however, reflects the essential care structure for a patient with an intramedullary cleft. It must be updated on the basis of new validated data.

This PNDS has been developed as per the "Méthode d'élaboration d'un protocole national de diagnostic et de soins pour les maladies rares" ["Development method for the National Diagnosis and Care Protocol for rare diseases" published by the Haute Autorité de Santé [French Health Authority] (methodological guide available on the HAS website: www.has-sante.fr). The content of the PNDS was discussed and validated by a multidisciplinary working group (Composition in Appendix 1).

A more detailed document which has been used as a basis for the development of the PNDS and which notably includes the analysis of identified bibliographic data serving as a scientific argument is available on the referral centre's website, http://www.syringomyelie.fr.

3. DIAGNOSIS AND INITIAL EVALUATION

The symptomatology usually found in syringomyelia consists of the classic syndrome suspended with the loss of thermal sensitivity and presence of pain in the affected area. The symptoms may be more or less severe, with chronic neuropathic pain, amyotrophy, variable sublesional damage (motor deficit, spasticity, other sensory modalities, urinary, anorectal, genito-sexual disorders etc.). In addition to these symptoms, a large majority of patients experience chronic pain, an often irreducible, neuropathic pain. Acute neurological deterioration may occur during Valsalva-like cases: coughing, sneezing, or even exceptionally by ultrasound (extracorporeal lithotripsy for urolithiasis), etc. The mechanism responsible may be the increase in venous pressure in the epidural veins that forces the cavity to dissect the white matter. This can result in the growth of the syringomyelic cavity (symptomatic or not).
In the case of intramedullary clefts, the symptoms described above are exceptional; pains of an atypical nature are generally in the foreground and at the origin of the radiological diagnosis.
The examination of choice is medullary MRI, which, apart from information on the cavity itself, can give useful information regarding responsible or associated pathologies.
A particular entity consists of intramedullary clefts that are differentiated from progressive syringomyelia by the clinical and radiological aspect. These characteristics will influence the diagnosis (less disabling and better prognosis), the tracking rate and care.

3.1. Objectives

It is extremely important to be able to distinguish intramedullary clefts from true syringomyelia at an early stage. For this purpose, it is necessary to combine a very precise clinical (neurological, pain and urodynamic assessment), electrophysiological and radiological assessment in the hope to identify patients who will advance from the others. Electrophysiology, but especially MRI (mainly fibre tracking sequences), allow on the one hand better understanding of the physiopathological substrate of pain in these patients, and on the other hand, to provide prognostic elements and appropriate care.
Anatomical (post mortem) and clinical (imaging) studies have clearly demonstrated that the ependymal duct may remain permeable at least in part for an adult (Roser 2009). This channel derives from the neural crest that appears on the 7th day of embryonic life and later forms the neural tube. This tube closes gradually towards the 14th day to form a channel. (Kasantikul 1979, Milhorat 1994). At birth, its diameter is 0.05-0.1 mm. At the terminal filum, the canal extends during embryogenesis to form what has become known as the terminal ventricle. Before the central canal begins its gradual closure, it contains a small amount of cerebrospinal fluid and is lined with an epithelium of ciliated ependymal cells. Other studies have shown that the ependymal canal is largely open and permeable only in the

foetus, and the new-born then shrinks in size and is gradually obliterated. The ependymal canal is completely closed in the majority of adults (Milhorat 1994).
The occurrence of syringomyelia has often been related to the ependymal canal. Gardner had postulated that obstruction of the 4th ventricle outlet port caused the forced passage of CSF to the ependymal canal through a permeable obex, which would cause progressive dilatation of the canal and thus syringomyelia (Gardner 1965). This theory had been reinforced by the work of Becker, who was able to demonstrate that experimental hydrocephalus induced in the dog by intrathecal injection of Kaolin was systematically followed by dilation of the central canal unless the entry and exit of the canal were obstructed at the level of the obex and the terminal filum (Becker 1972). This theory has been refuted by other authors. Milhorat (Milhorat 1997) and others oppose the theory that the obex is most often closed and that communication between the 4th ventricle and the ependymal canal is quite rare (4% in a series of 285 patients with foraminal syringomyelia, Aghakhani 1999). Moreover, the upper end of the syringomyelic cavity is most often at a distance from the 4th ventricle.
Moreover, Yasui's anatomical work on the modification of the ependymal canal as a function of age has demonstrated that the configuration of ependymal canal residues does not influence the topography of a possible syringomyelia (Yasui 1999).
With the availability of MRI, more and more patients with this type of small cavity are being diagnosed. In a publication, Jinkins et al. report 3 cases of adult patients with an intramedullary cavity revealed by pain (Jinkins 1999). These patients remain clinically and radiologically stable over a period of 2 years. Holly and Batzdorf report a series of 32 patients with an intramedullary cleft. Over an average follow-up period of 38 months (6 to 110 months), six patients improve, seven worsen and 19 remain stable (Holly 2002). No change in cavity size is detected during this period. There is no clear information on the type and kinetics of deterioration, but this is one of the few publications that report clinical deterioration in patients with a cleft while the radiological appearance remains stable (what the authors call
"Slit-like syrinx cavities").
On the basis of a series of 40 patients with an intramedullary cleft, Roser et al reported that in the majority of cases (90%), the cleft was diagnosed on an MRI performed to explore pain, 4 patients being absolutely asymptomatic (Roser 2009). In these patients, they did not observe any clinical or radiological deterioration at the end of an average follow-up of 36 months (from 6 to 93 months). Considering these results, they call these "hydromyelia" clefts and propose a definition that is at the same time clinical, electrophysiological and radiological: hydromyelia is a central and small medial intramedullary cavity in a patient without neurological sign outside of pain, and in whom no abnormality of CSF circulation or spinal cord conduction (SEP) is evident. They consider these clefts as a state prior to the formation of syringomyelia. "The hydromyelia might be predisposed. If in these patients an adequate trauma occurs, the development of a syringomyelia due to pressure changes in the subarachnoid space can take place". This work is important as

it shows that, on the basis of paraclinical examinations, it is possible to separate clefts from true syringomyelia.

3.2. Professionals involved and coordination arrangements

The professionals involved in the diagnosis and care of intramedullary clefts are doctors and paramedical personnel.
The medical team includes: the attending doctor, neurologists, neurosurgeons, neuroradiologists, physical medicine and rehabilitation physicians, pain specialists, urologists. The files are discussed in multidisciplinary meetings.
The paramedical team consists of: nurses, physiotherapists, psychologists, occupational therapists.
In the Referral Centre, there is also an educational therapy team that supports patients either in consultation or during hospitalisation (day of hospital or conventional hospitalisation).

3.3. Background circumstances

Given the availability of MRI nowadays, we are seeing more and more patients in whom an intramedullary cavity has been discovered either accidentally or with more or less intense pain, or, more rarely, with signs various sub-lesions (progressive motor deficit, sphincter disorders etc.). The morphological features of intramedullary clefts are represented by: one or more cavities of 3 to 6 mm in maximum transverse diameter, located at the crossing of the anterior third with the two thirds posterior and in the median position of the spinal cord. The term "syringomyelia" is often used in radiological reports, patients come in to consultation very worried by the information often collected on the internet concerning syringomyelia, a disease of bad reputation and often synonymous with severe disability. Now, as said above, not every intramedullary cavity is a syringomyelia. To differentiate a true syringomyelia, perhaps at a primitive stage, from an ependymal canal residue, it is necessary to regularly monitor these patients from the clinical, electrophysiological and neuroradiological point of view. Therapeutic management consists of symptomatic medical treatment (including pain), neuropsychological care (the disorders are often the consequence of a misdiagnosis and chronic neuropathic pain) and ultimately surgical care if necessary.
For clefts less than 3 mm in size, it is only considered visibility of the ependymal canal, therefore an anatomical variant to normal.

3.4. Diagnosis confirmation/differential diagnosis

The definitive diagnosis is confirmed by magnetic resonance imaging of the entire spinal cord and cervico-occipital joint. Targeted sequences are used to establish the presence of an intramedullary cleft, search for aetiology and assess likely course:

T1 sequence in sagittal and axial sections of the entire medullary cord and cervico-occipital joint. An injection of gadolinium is necessary during the initial assessment to eliminate a secondary syringomyelia (for example associated with a tumour),

T2 sequence in sagittal and axial sections of the entire medullary cord and cervico-occipital joint,

CISS or DRIVE sequence (T2 thin sections) on the targeted zone of intramedullary cleft or syringomyelia, looking for an arachnoiditis, an arachnoidal flange for example,

Flow sequence at the cervico-occipital joint, peripedular subarachnoid spaces (PMSAS) and in the syringomyelic cavity with the determination in cm/s of intra-cystic diastolic and systolic and PMSAS speeds,

Medullary cone diffusion tensor sequence: beam study + fraction of anisotropy (fibre tracking MRI) (detailed protocol in MR Diffusion Tensor Imaging and Fibre Tracking in Spinal Cord Compression, Denis Ducreux et al., AJNR Am J Neuroradiol 26: 1587-1594, June/July 2005)

The morphological sequences will make it possible to analyse the location and the size of the cavity (extension in height and width, regular or irregular walls, shape of the cavity, Vaquero index), the location of the terminal filum, the state of the terminal filum (its thickness, the existence or not of the fatty signal within it) and to monitor the absence of any cause of syringomyelia. In general, the intramedullary clefts are estimated to be about 3-6 mm in diameter and the centromedullary canal visible at 3 mm. In the different publications these figures are controversial. The best differentiation is considered to be done by MRI in fibre tracking.

The flow sequences will allow for dynamic analysis of CSF circulation at the cervico-occipital joint and peri-medullary subarachnoid spaces. They will verify the absence of any obstacle to the circulation of CSF and within the cavity (if it is possible considering the size). These sequences provide, on the one hand, information on the CSF circulation profile (synchronous or non-synchronous nature of the occurrence of systolic and diastolic peaks).

between the cyst and the PMSAS, aspect softened or not by these peaks) and on the other hand allow to measure the speed of this circulation (Brugières 1999).

MRI fibre tracking reveals the organisation of nerve fibres in the marrow and at different levels (cervical, dorsal upper, middle and lower and around the cleft). These sequences can allow the differential diagnosis of a syringomyelic type intramedullary cleft of a visible centromedullary canal, thus making the difference between a potentially pathological situation of an anatomical variant to normal. They may also explain the incidence of neuropathic pain in the clefts because of nerve fibre spread or destruction.

All these elements are studied and analysed during the follow-up to make it possible to check the existence or not of the likely course of the cleft.

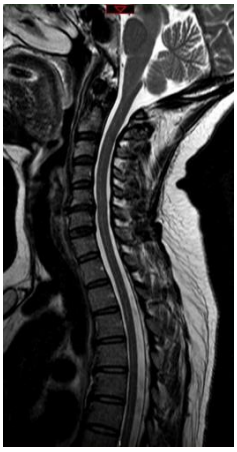


Figure 1. Visible centromedullary canal (less than 3mm in diameter), without pathological characteristics. MRI in sagittal T2 sequence.

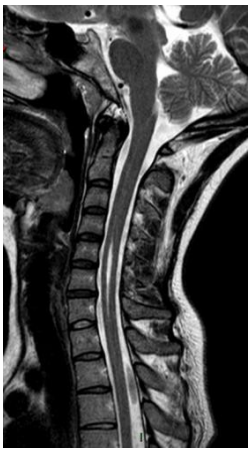


Figure 2. Intramedullary cervical cleft. MRI in sagittal T2 sequence.

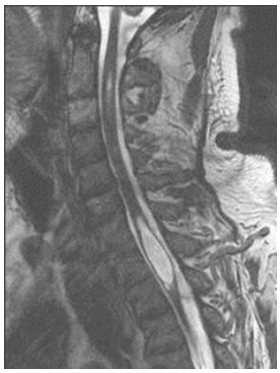


Figure 3. Cervical syringomyelic cavity. MRI in sagittal T2 sequence.

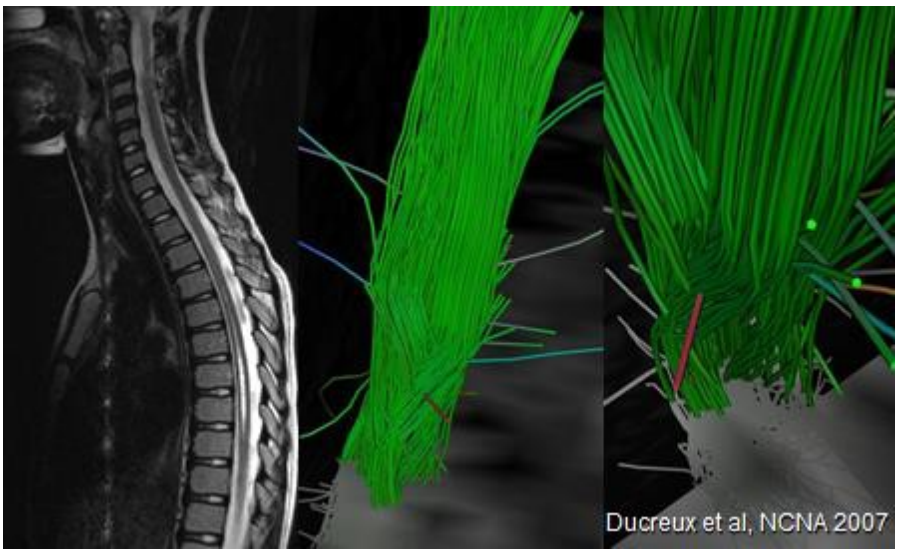


Figure 4. Intramedullary cleft. MRI in sagittal T2 sequence and fibre tracking. Interruption of the fibres on the tracking.

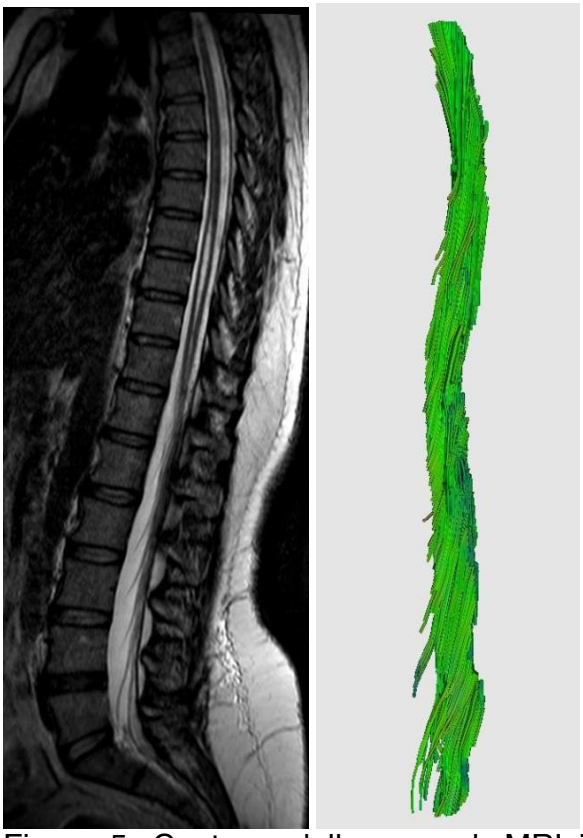


Figure 5. Centromedullary canal, MRI in sagittal T2 sequence and fibre tracking. Anatomical variant to normal (normal tracking).

Differential diagnosis

The differential diagnosis of intramedullary clefts concerns any intramedullary fluid cavity, including post-traumatic, post-infectious, syringomyelia, arachnoiditis (arachnoidal flange, arachnoid cysts, etc.), foraminal and malformative syringomyelia, dysraphisms such as spina bifida, diastematomyelia. Much attention should be paid to aspects of "persistence of the centromedullary canal".

The clinical side involves differentiation with central nervous system diseases such as multiple sclerosis, Devic's optic neuromyelitis, and other inflammatory diseases.

An important aspect of the differential diagnosis is clinically and radiologically non-progressive clefts without aetiology (idiopathic) but with a florid clinical picture including neuropathic pain. The neuropathic aspect can sometimes be surpassed by an inflammatory and/or mechanical character due to other pathologies. In these cases, it is necessary to make the differential diagnosis, particularly with inflammatory systemic pathologies,

Lyme disease, amyotrophic lateral sclerosis, Biermer anaemia, etc. Some patients who have discovered a cleft or even those who fortuitously discovered a visible channel, so a variant of normal, sometimes have a clinical picture of fibromyalgia, often associated with chronic fatigue or even mood disorders. Fibromyalgia can sometimes, in fact, be related to the central dysfunction caused by even minimal spinal cord injury, and not only to the psychological problems of patients, so this type of symptoms can be found in patients with clefts.
Given the long course, sometimes the misdiagnosis and the psychological components of these patients, as well as the secondary benefits of the diagnosis of syringomyelia, it is necessary to ensure the diagnostic announcement in a multidisciplinary setting or during educational therapy sessions.

3.5. Assessment of severity and prognosis

Patients with an intramedullary cleft are most often and mainly in neuropathic pain. The neurological clinical examination does not target the vast majority of cases of neurological deficit. If over the course of the progression in signs other than pain syndrome appear, it is imperative to extend research to eliminate another pathology as well as prevent a possible transformation of the cleft into a real developing syringomyelia. Even if the examination is still rather poor, repercussion in the personal and social life of these patients is important. The size of these intramedullary clefts usually remains stable for years, but there may be a modification by an contributing external factor (such as trauma, meningitis). These factors can disrupt circulation of CSF and trigger the appearance of a syringomyelic cavity.

The severity of the disease lies mainly in the importance of neuropathic pain that may be responsible for a disability.

Patients' advancement over time will be evaluated clinically, electrophysiologically, psychologically, in pain centre and in imaging. The regularity of the evaluation is to be adapted case by case with an average of 3-9 months during the first checks. If the cavity remains stable, electrophysiological and radiological monitoring may be spaced out.

1/ Clinical assessment

The clinical assessment includes the patient interview, the standard clinical and neurological examination. This assessment can be completed using the validated and reproducible scales.

Klekamp-Samii's Scoring System. This scale, proposed and validated by Klekamp and Samii in 1993, is widely used in neurosurgical literature for chronic spinal diseases. It is the only one that allows for a quantified evaluation of the clinical situation. (Klekamp J, Samii M. Introduction of a score system for the clinical evaluation of patients with spinal processes. Acta Neurochir (Wien)1993,123:221–223). It ranges from 0 to 25, with 25 being the score of a person without a disability (Appendix 10).

ASIA Score. The importance of sensory and motor deficit is quantified using the ASIA scale. (American Spinal Injury Association. International standards for neurological classification of spinal cord injury, revised 2002. Chicago). This classification allows for a very precise and quantified evaluation of the neurological deficit (motor and sensory) and therefore the ability to follow the development of this deficit over time (Appendix 4).

Aminoff-Logue Grading Score, Karnofsky Scale (Appendix 14), Modified Rankin Scale (MRS) (Appendix 8). The Klekamp and Samii classification and the ASIA score allow for a detailed analysis of the neurological situation. In addition to some scales, it is useful to have a comprehensive overview of the patient's neurological state and independence. For this we use the modified classification of McCormick (Aghakhani 2008) (Appendix 12), the Modified Rankin classification (Appendix 8) (mRS, Bonita 1988) and the Karnofsky scale (Karnofsky 1949) (Appendix 14), and the Functional Independence Measure (FIM) (Appendix 7).

Moreover, it is important to take into account patient quality of life with either syringomyelia (possibility of neurological disability and pain) or an intraspinal syringomyelic cleft (dysfunction due to chronic neuropathic pain). This assessment is essential to gain insight into how patients value their own progression.

Sensitivity, including thermal sensitivity, can be assessed using the quantified examination of sensitivities and thermal thresholds by means of a thermotest (quantitative sensory testing: QST) allowing the study of thresholds of somatosensory sub-modalities (thresholds detection and pain) in response to hot or cold thermal graduated stimulations. This type of standardised evaluation makes it possible to quantify the deficits and follow their development over time.

Pain should also be assessed during consultations. Tools can be used in this way:

The Visual Analogue Scale (VAS) (Appendix 6) (from 0 = no pain to 100 = maximum pain imaginable) and the Likert numerical scale in 11

items (from 0 = no pain to 10 = maximum pain imaginable) allow evaluation of pain intensity at the present time, average, maximum and minimum pain of 24 hours as in the concise questionnaire on pain (Brief Pain Inventory, BPI - Cleeland and Ryan 1994) (Appendix 13).

Mc Gill's Pain Questionnaire (Melzack 1975) and its abridged version (Mc Gill Short Form Questionnaire, Melzack 1986) available in French: the abridged version consists of 15 items including 11 sensory items and 4 affective items, sides each on a categorial scale in 4 items.

The Saint Antoine questionnaire (QDSA - Appendix 3): its abbreviated version with 16 items, explores the sensory-discriminative aspect of pain in the first 9 items, and on the last 7 its emotional side. However, the abridged version is not validated in Anglo-Saxon literature.

The DN4 questionnaire: the DN4 questionnaire (neuropathic pain in 4 questions) (Bouhassira et al 2005) (Appendix 5) is a diagnostic tool for neuropathic pain and not a pain assessment questionnaire. It has its place in the diagnosis of neuropathic pain and the differential diagnosis with other types of pain (nociceptive, inflammatory, visceral, etc.). It is therefore very suitable for diagnosing the nature of pain in syringomyelia. This questionnaire consists of 7 questioning items based on patient symptoms and 3 simple items of clinical examination. A score of 4 out of 10 allows the diagnosis of DN with a sensitivity of 83% and a specificity of 90%.

The NPSI (Neuropathic Pain Symptom Inventory) questionnaire (Appendix 9) is a self-administered questionnaire with 10 symptomatic items (burn, vice, compression, electric shock, pain caused by rubbing, etc.) and 2 temporary items that allow evaluation of five dimensions of neuropathic pain (burning, deep pain, paroxysmal pain, evoked pain, paraesthesia/dysesthesia) (Bouhassira et al 2005).

2/ Psychological care

A psychological interview may sometimes be necessary in these patients who have sometimes had a misdiagnosis and suffering from an invisible disability. This care is often proposed in the context of the Referral Centre to the patient diagnosis with the freedom to use thereafter.

3/ Urodynamic assessment

A neuro-urological assessment is useful in case of complaints, during the initial assessment or during the follow-up. This evaluation has three sides:

I. Clinical: urinary symptoms, sexual score, anorectal symptoms score, perineal examination,

II. Urodynamic: Assessment of sensitivity and bladder motor skills, detrusor hyperactivity or hypoactivity; sphincter performance analysis; search for partial urinary retention.

III. Perineal electromyography (denervation, right/left value sacral latencies, P40 value of cortical potentials of the pudendal nerve).

4/ Electrophysiological scans

Somaesthetic evoked potentials can be carried out during the assessment and/or during the follow-up as per the symptomatology presented by the patient, the clinico-radiological development. They make it possible to evaluate the functioning of the pathways of sensitivity and authenticate a medullary pain or a sensitivity disorder.

The laser evoked potentials study the functioning of the pathways of thermal sensitivity and neuropathic pain so can be very useful in the evaluation of clefts.

3.6. Finding contraindications to treatment

The treatment is medical and consists of pain management. There is no surgical indication for intramedullary clefts. Since there is no specific treatment and the treatment is only symptomatic, the contraindications are those of the therapies used.

3.7. Diagnosis and patient details

The diagnosis is made in consultation with a neurosurgeon or during an educational therapy session.

The patient benefits from detailed information with the results from their examinations and in particular the MRI images. Standard images of normal marrow and syringomyelia can be shown to the patient to better visualise the difference compared with an intramedullary cleft. The tracking rate is also explained as well as the therapeutic management. The possibility of advancing into real evolutionary syringomyelia and the signs that can alert are explained.

For patients who remain in pain despite standard therapeutic proposals, they are usually referred to a pain centre for specialised pain care.

Definitive diagnosis: It is done by clinical evaluation, MRI and paraclinical explorations.

Lack of urgency

The most common complaint: Neuropathic pain

Clinical examination: In general, normal

An intramedullary cleft is a potentially scalable syringomyelic cavity.

4. THERAPEUTIC MANAGEMENT

4.1. Objectives

Intramedullary clefts do not require surgical treatment. Any surgical notes are indicated regarding aetiological conditions such as Chiari, arachnoiditis, tumours etc. The non-surgical treatment of intramedullary clefts focuses on pain management, with analgesics of different pharmacological classes (antidepressants, antiepileptics, even opiates). Added to this is the maintenance of functional abilities, quality of life with physiotherapy and care in order to avoid socio-professional withdrawal.

4.2. Professionals involved (and coordination procedures)

The attending doctor may start and monitor the treatment of pain, but it is desirable that patients receive timely or regular treatment in a pain centre throughout resistant pains.
In rare cases where a surgical decision is possible, it takes place in a multidisciplinary meeting on a case by case basis, in a referral or competent centre where neurosurgeons, neuroradiologists, anaesthesiologists, medical doctors, psychologists, etc. are gathered. The attending doctor is informed of the decisions.

4.3. Therapeutic management

a. SURGICAL TREATMENT

There is no specific surgical treatment for the intramedullary clefts themselves. On the other hand, it is only the intramedullary cleft that can evolve which could justify a surgical intervention (arachnoidolysis, syringoperitoneal derivation etc.). Only in cases where there is an aetiology to this cleft and this aetiology is symptomatic, surgery may be considered (osteodalural decompression for Chiari malformation, excision of a tumour or other spinal cord injury, etc.)

b. MEDICAL TREATMENT

The therapeutic management consists of, on the one hand, the medical treatment of the symptomatology (in particular the pain), the psychological care (often misdiagnosis and consequences of chronic neuropathic pains) then in fine surgical care if necessary.
Neuropathic pain is persistent and often deteriorates over time (Baastrup and Finnerup, 2008, Finnerup, Attal et al Lancet Neurol 2015, Bouhassira and Attal Pain 2015). Neuropathic pain therapies include tricyclic antidepressants (including amitriptyline), mixed serotonin and noradrenaline inhibitors (e.g. duloxetine), antiepileptics

(gabapentin, pregabalin) and opiates (tramadol, morphine, oxycodone) (Taesell et al., 2010). Baclofen and botulinum toxin type A (BTX-A) are administered to relieve nociceptive pain directly related to spasticity (Taesell et al., 2010). These current treatments only lead to a 20 to 30% reduction in pain (Baastrup and Finnerup, 2008, Finnerup, Attal et al 2015).

c. NEURO-UROLOGICAL TREATMENT

The treatment of vesico-sphincteric, anorectal and genito-sexual disorders is important because of the potential risks of chronic vesico-renal damage due to the neurological bladder and of course the impact of these disorders on quality of life . Anticholinergic or beta3-adrenergic agonists may be proposed in case of an overactive bladder (pollakiuria, emergencies, leaks) and in case of failure peripheral neuromodulation or intra-detrusor injections of botulinum toxin. The bladder hypoactivity responsible for retention may require the introduction of self-catheterisation.

d. PSYCHOLOGICAL TREATMENT

Neuropathic pain has a real impact on the quality of life of patients both personally and professionally. They break into the patient's psychic life and upset the established balance.
Psychological care consists of taking them back to their medical path obviously in line with their emotional feelings. Moreover, the invisible disability is not always recognised as real by friends and family, which will provoke many arguments on the family level as well as professional. This generates and/or accentuates anxio-depressive symptoms.
In the therapeutic approach, it is necessary to take into account, for a certain number of patients, the discrepancy between their clinical feeling (neuropathic pain) and the diagnosis. Indeed, their neuropathic pain is often discordant with the morphological aspect of the cleft. We sometimes find tables that are either depressive or more similar to fibromyalgia.
The psychological treatment of patients with clefts consists of:

1. Cognitive-behavioural therapies,
2. Psychotherapy,
3. Complementary therapies can be considered such as hypnosis, auriculotherapy, acupuncture, EFT (emotional freedom technics), sophrology, relaxing massages,
4. Choosing effective coping strategies in consultation or educational therapy session.
5. Yesrequirement, assessment by a psychiatrist for potential prescription of anxiolytics and/or anti-depressants.

Psychological treatment of patients with clefts will vary depending on the patient's personality. Typically, these patients are likely to have experienced a misdiagnosis. Having arrived at the referral centre, patients feel relieved to have found a place where they will be listened to and supported.

e. REHABILITATION

Rehabilitation combines medical treatments with physical therapies to overcome and counter the sensory and emotional dimensions of neuropathic pain (although it is even more useful for other pain associated with nociceptive pain).
The care strategy has three aspects: physical, neurophysiological and cognitive-behavioural. The techniques of the physical and mechanical side are: massages, joint mobilisations, ergotherapeutic advice on the ergonomics of professional and private life activities.
The neurophysiological strategy covers neuropathic pain and includes transcutaneous electrical nerve stimulation (TENS). Other techniques are used without scientific consensus (cryotherapy, thermotherapy, trans-cutaneous mechanical vibration etc.).

4.4. Educational therapy and lifestyle modification (case by case)

Educational therapy is a new aspect of patient care. It must enable patients to live better with chronic illness. It is an integral part of the patient's care. The patient becomes a player in his or health and care.

In the Syringomyelia Referral Centre in Bicêtre, an educational therapy programme has been set up and will be available online in 2017 at www.has-sante.fr and www.syringomyelie.fr. The public supported by the programme is made up of adults and the patient's family and friends. The developed axes are:

1. Self-care skills (understanding illness and care pathways, analysing, measuring, coping, deciding, practising and applying the behaviours to be followed during a crisis situation and readjusting if necessary).
2. Acquire psychosocial skills (informing oneself, expressing one's needs, using the resources of the health care system, asserting one's rights, formulating a project and implementing it).

The themes tackled by the programme concern knowledge of yourself and your body, the relation between actual disability and the lived experience, social integration, the pains, the course in case of surgical treatment or other care and invasive therapy.

Other specific educational therapies are also sometimes useful depending on the associated signs (http://autosondage.jimdo.com). Education programme set up and published at the HAS by the AMARENCO Pr. department of neuro-urology (site constituting the Referral Centre for Rare Diseases - Syringomyelia).

Mainly symptomatic medical treatment,

uncommonly surgical in accordance with the aetiological factor.

4.5. Use of patient associations

It is very useful for patients to use patient associations to help break down isolation caused by rare diseases, to find moral support, practical help, to be informed.
The association APAISER (member of Alliance Rare Diseases, EURORDIS - Rare Disease Europe, of the Rare Diseases Network NeuroSphinx) is an Association approved by the Ministry of Health representing healthcare users. The actions of APAISER are intended to help and inform patients, but also to support research, organise annual conferences with the active participation of the Syringomyelia Coordinating Centre of Bicêtre, regional meetings, with the edition of a newsletter 3 times per year. The partnership with the Coordinating Centre of Bicêtre remains high priority. Regular meetings are held between the president of the association and the Centre to discuss and set up joint actions, thus representing the views of patients. The meetings are intended to make constructive exchanges in both directions: to bring scientific information from the medical profession and to let patients be heard back through the association.

5. FOLLOW-UP

5.1. Objectives

The main objective of the follow-up includes the radiological aspect which must be put in concordance with the clinical evaluation, with the aim of a therapeutic treatment adapted to the case by case. The secondary objectives, but not least, concern the monitoring of medical, psychological and rehabilitation therapy.

5.2. Professionals involved (and coordination procedures)

The follow-up has 4 axes: clinical, neuro-radiological, rehabilitation and psychological. Within the clinical axis, surveillance is shared between the attending doctor, the neurologist, the neurosurgeon and the doctor from the pain centre. MRI can be prescribed by any of these professionals. Rehabilitation care is provided by the physiotherapist of the sector with, if necessary, hospitalisations from time to time in a specialised centre. Psychological support is carried out by the local psychologist who takes over after a first treatment by the psychologist from the referral or competence centre. Sometimes it is necessary to combine treatment in psychiatry in case of depression or major anxiety disorders for example.

5.3. Regularity and content of consultations

Follow-up is shared with the attending doctor. Clinical monitoring is required every 3 months, depending on the magnitude of the symptoms, and radiological monitoring every 6 months during the first two years, then once a year if the situation is stable. Collaboration between the multidisciplinary team and the attending doctor must be close. Certain complications, urinary in particular, require more frequent consultations and evaluations according to the tables presented.

FOLLOW-UP

The first 2 years:

Clinical surveillance: every 3 months

MRI check: every 6 months

Neurosurgery consultation: every 6 months

After the 2nd year, if the situation is stable:

Clinical monitoring: every 6 months

MRI check: every year

Neurosurgery consultation: every 2 years

5.4. Special circumstances

Sports activities: Within the scope of non-progressive, idiopathic intramedullary cleft, there is no contraindication to sports activities and in general patients should have a normal life. In the case of an aetiology (Chiari malformation, trauma, tumours, etc.) the contraindications will be those of the pathology in question (PNDS to come).

Pregnant women: In the case of pregnant women with an intraspinal cleft, there is no formal contraindication to pregnancy or usual birthing techniques or the usual anaesthetic techniques. Thus, vaginal delivery, caesarean section, peridural anaesthesia, spinal anaesthesia or general anaesthesia do not present any contraindications. Uncommonly, we can see advancement of the intramedullary cleft during pregnancy due to changes in intra-abdominal and therefore epidural pressures. It is recommended that epidural anaesthesia or spinal anaesthesia be the subject of multidisciplinary consultation. It is recommended for pregnant women or women planning a pregnancy to connect their gynaecologist and anaesthetist with the neurosurgeon to decide together, based on the symptoms and morphological, dynamic and aetiological characteristics of the intramedullary cleft, the best treatment possible. The specific risks of each patient will be taken into account in order to ideally plan the pregnancy but especially the delivery.

Lumbar puncture: There is no formal contraindication in patients with an intramedullary cleft unless the aetiology of this cleft is in itself a contraindication (Chiari malformation for example). The discussion should be had on a case-by-case basis and it is recommended to contact a neurosurgeon at a Competence Centre or Coordinator.

Anaesthesia techniques: Epidural anaesthesia and spinal anaesthesia are not formally contraindicated in intramedullary clefts, but as far as possible, it is preferable that the situation be the subject of a multidisciplinary discussion.

Other:

- In cases of urinary lithiasis, extracorporeal lithotripsy should be discussed, even for a patient with an intramedullary cleft, because of the known risk of exacerbation of syringomyelia after ultrasound sessions.
- In cases of lumbosacral dysraphism, invasive procedures such as lumbar puncture, epidural anaesthesia and spinal anaesthesia are contraindicated.

Appendix 1. List of participants

This work was coordinated by Dr. Silvia MORAR and Dr. Anne HERBRECHT, Rare Diseases and Syringomyelia Referral Centre Hôpital Bicêtre, under the direction of Professor Fabrice PARKER.

DRAWN UP BY:

Dr Silvia MORAR
Neurosurgery Department
Rare Diseases and Syringomyelia Referral Centre
Hôpital Bicêtre
78 rue du Général Leclerc
94275 LE KREMLIN BICETRE Cedex
Tel: 01 45 21 24 55
Fax: 01 45 21 26 00
Email: sylvia.morar@aphp.fr

Dr Anne HERBRECHT
Neurosurgery Department
Rare Diseases and Syringomyelia Referral Centre
Hôpital Bicêtre
78 rue du Général Leclerc
94275 LE KREMLIN BICETRE Cedex
Tel: 01 45 21 25 15
Fax: 01 45 21 26 00
Email: anne.herbrecht@aphp.fr

Pr. Nozar AGHAKHANI
Neurosurgery Department
Rare Diseases and Syringomyelia Referral Centre
Hôpital Bicêtre
78 rue du Général Leclerc
94275 LE KREMLIN BICETRE Cedex
Tel: 01 45 21 23 80
Fax: 01 45 21 28 63
Email: nozar.aghakhani@aphp.fr

Pr. Fabrice PARKER
Neurosurgery Department
Rare Diseases and Syringomyelia Referral Centre
Hôpital Bicêtre
78 rue du Général Leclerc
94275 LE KREMLIN BICETRE Cedex
Tel: 01 45 21 23 80
Fax: 01 45 21 28 63
Email: fabrice.parker@aphp.fr

Pr. Denis DUCREUX
Neuroradiology
Department at Hôpital
Bicêtre
78 rue du Général Leclerc
94275 LE KREMLIN BICETRE Cédex
Tel: 01 45 21 33 88
Email: denis.ducreux@aphp.fr

Pr. Jean-Rodolphe
VIGNES Neurosurgery
Department at CHU de
Bordeaux
Place Amélie Raba Léon
33076 BORDEAUX Cedex
Tel: 05 56 79 55 43
Email: jean-rodolphe.vignes@chu-bordeaux.fr

Pr. Gérard AMARENCO
Department of Neuro-urology and Perineal Scans at
Hôpital Tenon
4 rue de la Chine
75970 PARIS Cedex 20
Tel: 01 56 01 76 13
Email: gerard.amarenco@aphp.fr

Pr. Nadine ATTAL
Centre for Pain Assessment and Treatment at
Hôpital Ambroise Paré
9 avenue Charles de Gaulle
92100 BOULOGNE BILLANCOURT
Tel: 01 49 09 59 46
Fax: 01 49 09 44 34
Email: nadine.attal@aphp.fr

Pr. Phong DAM HIEU
Neurosurgery Department at
Hôpital de la Cavale Blanche
Boulevard Tanguy Prigent
29069 BREST Cedex
Tel: 02 98 34 76 88
Email: phong.dam-hieu@chu-brest.fr

Prof. Klaus Luc MOURIER
Department of Neurosurgery at
CHRU de Dijon
14 rue Paul Gaffarel
21033 DIJON
Tel: 03 80 29 37 52
Email: klaus-luc.mourier@chu-dijon.fr

Ms Anne ELBAZ
Psychologist
Rare Diseases and Syringomyelia Referral Centre
Hôpital Bicêtre
78 rue du Général Leclerc
94275 LE KREMLIN BICETRE Cedex
Tel: 01 45 21 24 04
Email: anne.elbaz@aphp.frmailto:anne.elbaz@aphp.fr

Dr Corina CROITORU
General practitioner
Rare Diseases and Syringomyelia Referral Centre
Hôpital Bicêtre
78 rue du Général Leclerc
94275 LE KREMLIN BICETRE Cedex
Tel: 01 45 21 24 55
Email: corina.croitoru@aphp.frmailto:corina.croitoru@aphp.fr

Expressions of interest
All participants in the development of the PNDS completed an expression of interest

Appendix 2. Contact information of referral centres, competence and patient association centres

COORDINATING CENTRE

Neurosurgery Department
Hôpital Bicêtre
78 rue du Général Leclerc
94275 LE KREMLIN BICETRE Cedex
Tel: 01 45 21 24 55
Email: sylvia.morar@aphp.fr

CONSTITUENT CENTRES

Centre for Pain Assessment and Treatment at
Hôpital Ambroise Paré
9 avenue Charles de Gaulle
92100 BOULOGNE BILLANCOURT
Tel: 01 49 09 59 46
Fax: 01 49 09 44 34
Email: nadine.attal@aphp.fr

Department of Neuro-urology and Perineal Scans at
Hôpital Tenon
4 rue de la Chine
75970 PARIS Cedex 20
Tel: 01 56 01 76 13
Email: gerard.amarenco@aphp.fr

Paediatric Neurosurgery Department
at Hôpital Necker
149 rue de Sèvres
75015 PARIS
Tel: 01 44 49 42 67
Email: michel.zerah@aphp.fr

APAISER Association
Le Bas Chemin Bigot
35133 JAVENE
Tel: 06 81 79 61 20

Related websites:
http://www.syringomyelie.fr
http://www.maladiesrares-paris-sud.aphp.fr
http://www.orphanet.net

Patient associations:
APAISER (Association Pour Informer et Aider les Syringomyéliques Européens Réunis) [Association To Inform And Help European Syringomyelics]:
www.apaiser.asso.fr
AMCVHS (Association pour mieux comprendre et vivre l'hydrocéphalie et la syringomyélie) [Association to better understand and experience hydrocephalus and syringomyelia]:
www.amcvhs.com

Appendix 3. QDSA SCALE

A	***Beats***	
	Heartbeats	
	Stitches	
	In flashes	
	Discharge	
	Electrical	
	Intervals	

B	***Radiating***	
	Irradiating	

C	***Injection***	
	Cut	
	Penetrating	
	Stabbing pain	

D	***Pinching***	
	Cramps	
	Compression	
	Crushing	
	Vice-like	
	Grinding	

E	***Twinging***	
	Stretching	
	Distension	
	Tear	
	Torsion	
	Extraction	

F	***Heat***	
	Burning	

G	***Cold***	
	Ice	

H	***Itching***	
	Tingling	
	Irritation	

I	***Numbness***	
	Heaviness	
	Deaf	

J	***Tiring***	
	Tiresome	
	Exhausting	

K	***Nauseous***	
	Suffocating	
	Syncopal	

L	***Worrying***	
	Oppressive	
	Agonising	

M	***Exasperating***	
	Annoying	
	Nasty	
	Torturous	
	Tormenting	

N	***Irritating***	
	Unpleasant	
	Annoying	
	Unbearable	

O	***Irritating***	
	Exasperating	
	Infuriating	

P	***Depressing***	
	Suicidal	

Appendix 4. ASIA

Motor assessment

	R	L	
C2			
C3			
C4			
C5			Elbow flexion
C6			Wrist extension
C7			Elbow extension
C8			Middle finger flexion (P3)
T1			Fifth finger abduction
T2			
T3			
T4			
T5			
T6			
T7			
T8			
T9			
T10			
T11			
T12			
L1			
L2			Hip flexion
L3			Knee extension
L4			Ankle dorsiflexion
L5			Big toe extension
S1			Plantar ankle flexion
S2			
S3			
S4-5			

0 = total paralysis
1 = visible or palpable contraction
2 = active movement gravity-free
3 = active movement against gravity
4 = active movement against resistance
5 = normal movement
NT, not testable

"Motor skills" score: /100
Anal contraction: yes/no

Score ASIA

Patient Identity

Date of examination |_|_|/|_|_|/|_|_|_|_|

Neurological level* { Sensitive right ☐ left ☐ / Motor right ☐ left ☐

*Most caudal segment with normal function

Medullary lesion**: Complete or Incomplete

**Incomplete defined by motor skills or sensitivity in S4-S5

ASIA Anomaly Scale: A B C D E

A = complete: no motor skills or sensitivity in S4-S5
B = incomplete: sensitivity but no motor skills preserved below lesional level, especially in S4-S5
C incomplete motor skills is preserved below lesional level and more than half of the muscles tested below this level has a score < 3
D = incomplete: motor skills is preserved below lesional level and at least half of the muscles tested below the level have a score ≥ 3
E = normal: sensitivity and motor skills are normal

Partial Preservation*** { Sensitive right ☐ left ☐ / Motor right ☐ left ☐

*** Caudal extension of the segments partially

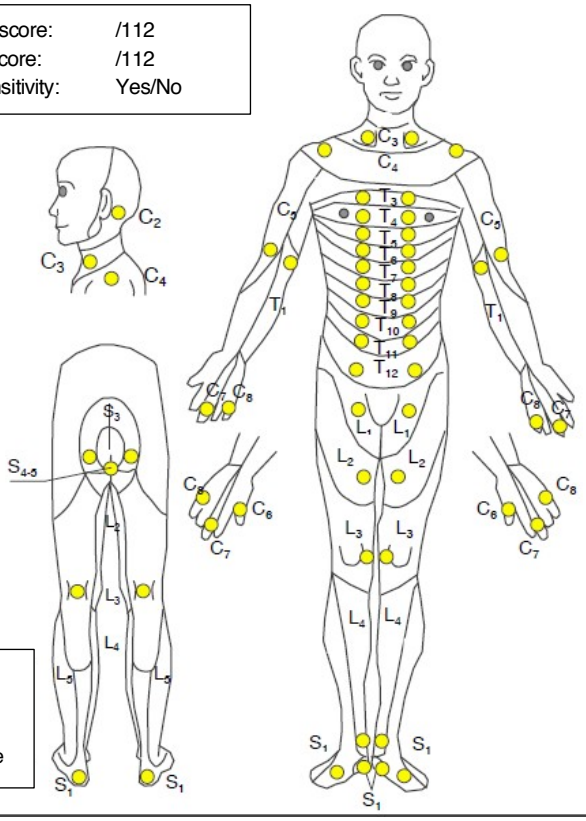
Clinical syndrome:
- Centromedullary ☐
- Brown-Séquard ☐
- Anterior marrow ☐
- Terminal filum ☐

Sensory evaluation

	Touch R	Touch L	Sting R	Sting L
C2				
C3				
C4				
C5				
C6				
C7				
C8				
T1				
T2				
T3				
T4				
T5				
T6				
T7				
T8				
T9				
T10				
T11				
T12				
L1				
L2				
L3				
L4				
L5				
S1				
S2				
S3				
S4-5				

"Touch" score: /112
"Sting" score: /112
Anal sensitivity: Yes/No

0 = absent
1 = decreased
2 = normal
NT = not testable



Appendix 5. DN4 QUESTIONNAIRE

QUESTIONNAIRE DN4 = a simple tool to check for neuropathic pain

To estimate the probability of neuropathic pain, the patient must answer each item of the 4 questions below with "yes" or "no".

QUESTION 1: Does the pain have one or more of the following characteristics?

	Yes	No
1. Burning	☐	☐
2. Painfully cold	☐	☐
3. Electric discharges	☐	☐

QUESTION 2: Is the pain associated in the same area as one or more of the following symptoms?

	Yes	No
4. Tingling	☐	☐
5. Itching	☐	☐
6. Numbness	☐	☐
7. Irritation	☐	☐

QUESTION 3: Is the pain localized in an area where the examination highlights:

	Yes	No
8. Hypoaesthesia to touch	☐	☐
9. Hypoesthesia with stinging	☐	☐

QUESTION 4: Is the pain caused or increased by

	Yes	No
10. Burning	☐	☐

YES = 1 point **NO = 0 points** **Patient Score: /10**

DIRECTIONS FOR USE

When the practitioner suspects neuropathic pain, the DN4 questionnaire is useful as a diagnostic tool.

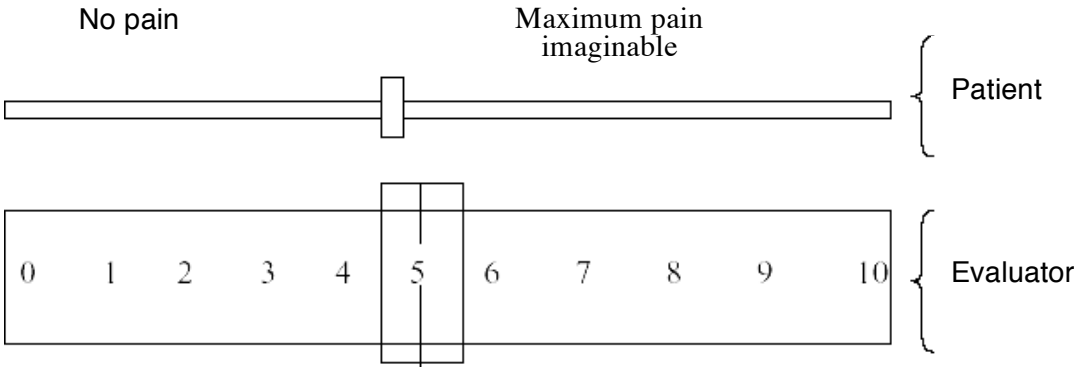
This questionnaire is divided into 4 questions representing 10 items to check :

- √ The practitioner questions the patient and completes the questionnaire
- √ Each item must be answered either yes or no
- √ At the end of the questionnaire, the practitioner counts the answers, 1 for each "yes" and 0 for each "no".
- √ The sum obtained provides the Patient Score, marked out of 10.

If the patient score is equal to or greater than 4/10, the test is positive
(sensitivity at 82.9%, specificity at 89.9%)

According to Bouhassira D et al. Pain 2004; 108 (3): 248-57.

Appendix 6. EVA SCALE



Appendix 7. FIM - MEASUREMENT OF FUNCTIONAL INDEPENDENCE

LEVELS

7 - Complete independence (appropriate to the circumstances and innocuous)
6 - Modified independence (with apparatus, for example)
5 - Observation
4 - Minimum assistance (25%)
3 - Average assistance (50%)
2 - Maximum assistance (75%)
1 - Complete assistance (100%)
If an item is not verifiable, quote level 1

PERSONAL CARE		
A Nutrition		
B Care of appearance		
C Toilet		
D Dressing - upper part		
E Dressing - lower part		
F Use of the toilet		
Sphincter control:		
G Bladder		
H Intestines		
Mobility - Transfers:		
I bed, chair, wheelchair		
J WC		
K Bath, shower		
Transport:		
L Walking (M), wheelchair (F)		
M Stairs		
Communication:		
N Listening comprehension		
N Visual comprehension		
O Verbal expression		
O Non-verbal expression		
Awareness of the outside world:		
P Social interaction		
Q Problem resolution		
R Memory		
Total		**/126**

Appendix 8. MODIFIED RANKIN SCALE

SCORE	DISABILITY
0	No symptoms
1	No significant disability apart from possible symptoms (able to take on roles, carry out activities
2	Light disability (unable to carry out all previous activities, able to carry on own business without assistance)
3	Moderate disability (requires some assistance, can walk without assistance)
4	Moderately severe disability (unable to walk unassisted, unable to attend to own needs without assistance)
5	Severe disability (confined to bed, incontinent and requiring constant attention and nursing care)
6	Death

Appendix 9. NPSI (Neuropathic Pain Symptom Inventory)

1. Does the pain feel like a burning sensation?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

2. Is your pain vice-like?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

3. Is your pain like a sensation of compression?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

4. During the past 24 hours, you have been in pain:

Permanently

Between 8 and 12 hours/day

Between 4 and 7 hours/day

Between 1 and 3 hours/day

Less than 1 hour/day

5. Do you have painful episodes like electric shocks?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

6. Do you have painful episodes such as stabbing pains?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

7. In the past 24 hours, how many painful episodes have you had?

MORE THAN 20	BETWEEN 11 AND 20	BETWEEN 6 AND 10	BETWEEN 1 AND 5	NO PAINFUL EPISODES

8. Do you have any pain caused or increased by rubbing on the painful area?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

9. Do you have any pain caused or increased by pressure on the painful area?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

10. Do you have any pain caused or increased by contact with a cold object on the painful area?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

11. Do you have any itching?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

12. Do you have any tingling?

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

Appendix 10. KLEKAMP and SAMII SCALE

Grade	Bladder and bladder sphincter	Anal sphincter
5	Normal	Normal
4	Slight distention, no catheterisation	Slight distention, complete control
3	Bladder residue, no catheterisation	Use of laxatives, normal control
2	Occasional catheterisation	Occasional loss of control
1	Frequent catheterisation	Frequent loss of control
0	Permanent catheterisation	No control

Appendix 11. MCGILL PAIN QUESTIONNAIRE

Specify the type of pain usually felt (in the last 8 days).

	0	1	2	3	4
	absent (none)	light (a little)	moderate (moderately)	strong (a lot)	extremely strong
Stitches					
Penetrating					
Electric discharges					
Stabbing pains					
Vice-like					
Twinging					
Burning					
Tingling					
Heaviness					
Tiresome					
Agonising					
Annoying					
Unbearable					
Irritating					
Exasperating					
Depressing					

Appendix 12. MC CORMICK CLASSIFICATION

GRADE I	Zero or minor deficiency not affecting Normal Walking function
GRADE II	Sensory deficit or moderate motor affecting function Moderate difficulty walking Severe pain degrading the quality of life Maintaining independence
GRADE III	Severe deficit Walking with cane(s) and/or significant loss of function Need for occasional help
GRADE IV	Severe deficit with impossible walking Loss of independence

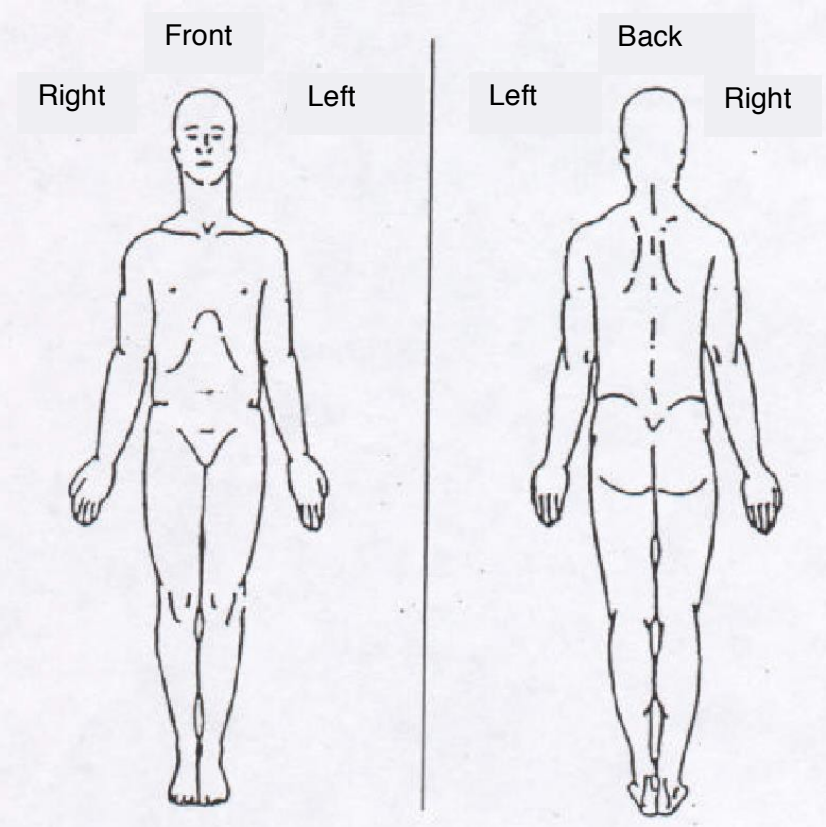
Appendix 13. BPI QUESTIONNAIRE (Brief Pain Inventory)

1. During most of our lives, most of us experience pain (headaches, toothache): over the past eight days, have you experienced pain other than this type of "Familiar" pain?

yes ☐ no ☐

If you answered "no" to the last question, you do not need to answer the following questions. Thank you for participating.

2. Indicate on this diagram where your pain is by shading the appropriate area. Put on the drawing an "S" for a pain near the surface of your body or a "D" for a deeper pain in the body. Also put an "I" where you feel the most intense pain.



3. Please circle the number that best describes the most pain **that you felt last week.**

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

4. Please circle the number that best describes the **lowest** pain that you felt last week.

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

5. Please circle the number that best describes the pain **in general**.

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

6. Circle the number that best describes the pain **at this time**.

No pain	0	1	2	3	4	5	6	7	8	9	10	Maximum pain

7. Which treatments or medications are you taking for the pain?

8. Last week, what relief did the treatments or medications give you: can you indicate the percentage of improvement you got?

No improvement	0%	10%	20%	30%	40%	50%	60%	70%	80%	90%	100%	Complete improvement

9. Circle the figure that best describes how, last week, the pain affected you:

    a. General activity

Unaffected	0	1	2	3	4	5	6	7	8	9	10	Complete hindrance

    b. Mood

Unaffected	0	1	2	3	4	5	6	7	8	9	10	Complete hindrance

    c. Ability to walk

Unaffected	0	1	2	3	4	5	6	7	8	9	10	Complete hindrance

    d. Regular work

Unaffected	0	1	2	3	4	5	6	7	8	9	10	Complete hindrance

e. Relations with others

Unaffected	0	1	2	3	4	5	6	7	8	9	10	Complete hindrance

f. Sleep

Unaffected	0	1	2	3	4	5	6	7	8	9	10	Complete hindrance

g. Zest for life

Unaffected	0	1	2	3	4	5	6	7	8	9	10	Complete hindrance

Appendix 14. KARNOFSKY SCALE

INDEX	DESCRIPTION
100	Normal; no complaints, no signs of illness
90	Able to continue normal activities; minor signs or symptoms of illness.
80	Normal activity, with effort; some signs or symptoms of illness.
70	Independent; unable to continue normal activity or work actively.
60	Occasional need for assistance but ability to provide for basic needs.
50	Need for considerable help for the person, frequent medical care.
40	Disabled; need for specific care and assistance.
30	Fully disabled; indication of hospitalisation, no imminent risk of death.
20	Very ill; hospitalisation required, active treatment or support necessary.
10	Moribund; fatal outcome close.
0	Deceased.

BIBLIOGRAPHIC REFERENCES

Aghakhani N, David P, Parker F, Lacroix C, Benoudiba F, Tadie M. Intramedullary spinal ependymomas: analysis of a consecutive series of 82 adult cases with particular attention to patients with no preoperative neurological deficit. Neurosurgery. 2008 Jun;62(6):1279-85; discussion 1285-6.

Aghakhani N, Parker F, Tadie M. Syringomyélie et l'anomalie de Chiari chez l'adulte [Syringomyelia and the Chiari anomaly in adults]. Analysis and results of the cooperative series of 285 cases. Neurosurgery. 1999, 45 Suppl. 1:23- 36.

American Spinal Injury Association. International standards for neurological classification of spinal cord injury, revised 2002. Chicago (IL): American Spinal Injury Association; 2002

Attal N, Brasseur L, Parker F, Tadie M, Bouhassira D. Caractérisation des troubles sensitifs et des douleurs neuropathiques liés aux syringomyélies [Characterisation of sensory disorders and neuropathic pain related to syringomyelia]. Prospective study. Neurosurgery. 1999, 45 Suppl. 1:84-94.

Baastrup C1, Finnerup NB. Pharmacological care of neuropathic pain following spinal cord injury. CNS Drugs. 2008;22(6):455-75.

Becker DP, Wilson JA, Watson GW. The spinal cord central canal: response to experimental hydrocephalus and canal occlusion. J Neurosurg. 1972 Apr;36 (4):416-24.

Bouhassira D, Attal N, Alchaar H, Boureau F, Brochet B, Bruxelle J, Cunin G, Fermanian J, Ginies P, Grun-Overdyking A, Jafari-Schluep H, Lantéri-Minet M, Laurent B, Mick G, Serrie A, Valade D, Vicaut E. Comparison of pain syndromes associated with nervous or somatic lesions and development of a new neuropathic pain diagnostic questionnaire (DN4). Pain. 2005 Mar; 114 (1-2): 29-36.

Bonita R, Beaglehole R. "Modification of Rankin Scale: Recovery of motor function after stroke." Stroke 1988 Dec;19(12):1497-1500.

Brugieres P, Idy-Peretti I, Iffenecker C, Parker F, Jolivet O, Hurth M, Gaston A, Bittoun J. CSF flow measurement in syringomyelia. AJNR Am J Neuroradiol. 2000 Nov-Dec;2110

C. Sixt, F. Riether, B.E. Will, M.S. Tatagiba, F. Roser Evaluation of quality of life parameters in patients who have syringomyelia, journal of clinical neuroscience, 16 (2009) 1599–1603

Cleeland CS, Ryan KM. Pain assessment: global use of the Brief Pain Inventory. Ann Acad Med Singapore. 1994 Mar; 23 (2): 129-38. Review.

David Facon, Augustin Ozanne, Pierre Fillard, Jean-François Lepeintre, Caroline Tournoux-Facon, and Denis Ducreux, MR Diffusion Tensor Imaging and Fiber Tracking in Spinal Cord Compression, AJNR Am J Neuroradiol 26:1587–1594, June/July 2005

Finnerup NB, Attal N. Tapentadol prolonged release in the treatment of neuropathic pain related to diabetic polyneuropathy--authors' reply. Lancet Neurol. 2015 Jul; 14 (7): 685-6.

Gardner WJ. Hydrodynamic mechanism of syringomyelia: its relationship to myelocele. J Neurol Neurosurg Psychiatry. 1965, 28:247-59.

Holly LT, Batzdorf U. Slit-like syrinx cavities: a persistent central canal. J Neurosurg. 2002 Sep;97(2 Suppl.):161-5.

Jinkins JR, Sener RN. Idiopathic localised hydromyelia: dilatation of the central canal of the spinal cord of probable congenital origin. J Comput Assist Tomogr. 1999 May-Jun;23(3):351-3.

Karnofsky D. The clinical evaluation of chemotherapeutic agents in cancer. Columbia University Press, New-York 1949 :191-205

Kasantikul V, Netsky MG, James AE Jr. Relation of age and cerebral ventricle size to central canal in man. Morphological analysis. J Neurosurg. 1979 Jul; 51 (1): 85-93.

(Klekamp J, Samii M. Introduction of a system score for the clinical evaluation of patients with spinal processes. Acta Neurochir (Wien) 1993, 123: 221-223).

Melzack R. Pain. The McGill Pain Questionnaire: major properties and scoring methods. 1975 Sep;1(3):277-99.

Melzack R, Terrence C, Fromm G, Amsel R. Trigeminal neuralgia and atypical facial pain: use of the McGill Pain Questionnaire for discrimination and diagnosis. Pain. 1986 Dec;27(3):297-302.

Milhorat TH, Capocelli AL Jr, Kotzen RM, Bolognese P, Heger IM, Cottrell JE. Intramedullary pressure in syringomyelia: clinical and pathophysiological correlates of syrinx distension. Neurosurgery. 1997, 41(5):1102-10.

Milhorat TH, Kotzen RM, Anzil AP Stenosis of central canal of spinal cord in man: incidence and pathological findings in 232 autopsy cases. J Neurosurg. 1994 Apr;80 (4):716-22.

Milhorat TH, Kotzen RM, Anzil AP. Stenosis of central canal of spinal cord in man: incidence and pathological findings in 232 autopsy cases. J Neurosurg. 1994, 80(4):716-22.

Roser F., Ebner FH., Sixt C., Müller J., Hagen V., Tatagiba MS. Defining the line between hydromyelia and syringomyelia. A differentiation is possible based on electrophysiological and magnetic resonance imaging studies, Acta Neurochir (Wien). 2009 June 16

Teasell RW, Mehta S, Aubut JA, Foulon B, Wolfe DL, Hsieh JT, Townson AF, Short C; Spinal Cord Injury Rehabilitation Evidence Research Team. A systematic review of pharmacologic treatments of pain after spinal cord injury. Arch Phys Med Rehabil. 2010 May;91(5):816-31.

Yasui K, Hashizume Y, Yoshida M, Kameyama T, Sobue G. Age-related morphologic changes of the central canal of the human spinal cord. Acta Neuropathol. 1999 Mar; 97 (3): 253-9.