

Summary for the attending doctor

**Protocole National de Diagnostic et de Soins
(PNDS) [National Diagnostic and Care Protocol]
Name of the rare disease**

SYRINGOMYELIA
Intramedullary clefts

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 every way 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 doctor: Dr. Silvia
MORAR Telephone: 01 45 21 24 55
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Coordinating doctor: Pr. Fabrice PARKER
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Consultations:
Telephone: 01 45 21 22 88
Email: consultation.neurochirurgie.bct@aphp.fr

Useful websites:

Internet links:

- www.syringomyelie.fr
- www.maladiesraresparis-sud.aphp.fr
- www.orphanet.fr

Related sites:

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