Clinical Reasoning:
A 39-year-old man with abdominal cramps

SECTION 1
A 39-year-old lawyer presented with intermittent spasms and pain in his abdominal muscles, particularly the right upper quadrant. These had occurred since his mid-20s and there had been long asymptomatic periods, including 8 years prior to the most recent 4-month exacerbation. Trivial movement triggered a spasm of the abdominal muscles, leading to severe pain, which made breathing uncomfortable and interfered with sleep. The symptoms subsided spontaneously after 4 to 5 days, leaving him with a sore abdomen for several weeks. Past attacks had also been precipitated by specific forms of repetitive exercise such as jogging. He described ill-defined numbness in the left leg, but denied any muscle twitching, weakness, back pain, or sphincter disturbance. There was no significant past medical or family history.

On examination, cranial nerves were unremarkable. Tone and power were normal in upper and lower limbs. Tendon reflexes were brisk throughout, particularly in the lower limbs, where they were brisker on the left than the right; plantar responses were flexor. Abdominal reflexes were brisk on the right and absent on the left (video on the Neurology® Web site at www.neurology.org). No fasciculations or myokymia were seen throughout. There was no demonstrable sensory asymmetry or loss to any modality in the lower limbs. Gait and cerebellar function were normal.

Questions for consideration:
1. Can you interpret the sign demonstrated in the video?
2. What is the differential diagnosis for this presentation?
3. What is your next investigation of choice?
SECTION 2

Superficial abdominal reflexes are not elicitable in all individuals and become less prevalent with age. They may also be absent in obesity, after multiple pregnancies, or after abdominal surgery. One study characterized the reflexes in each abdominal quadrant of normal young adults. In approximately half the subjects, symmetrical reflexes could be elicited in all quadrants; the remainder showed a variable extent of asymmetrical or absent reflexes. However, in no subjects were the abdominal reflexes consistently present on one side and consistently absent on the other. Such findings in our patient are therefore likely to be significant and—in the absence of sensory loss—suggest a lesion of the upper motor neurons in the ipsilateral thoracic cord, the corresponding lower motor neurons, or both. Both intrinsic and extrinsic lesions of the spinal cord could produce this picture. The differential diagnosis therefore includes neoplasia (e.g., ependymoma, meningioma, glioma), arteriovenous malformation, syrinx, lateral intervertebral disk prolapse, inflammatory myelitis, and infection (e.g., segmental zoster paresis).

The next investigation of choice is an MRI scan of the thoracic spine (figure 1).

Questions for consideration:
1. What is the abnormality on MRI?
2. What further investigations would you consider?
SECTION 3
The MRI shows a central fusiform cavity in the mid-thoracic cord, extending from T7 to T10 and measuring 5 mm in diameter. There was no abnormal gadolinium enhancement. This radiologic description would be compatible with either idiopathic syringomyelia or hydromyelia. Hydromyelia is considered to be a congenital, static persistence or enlargement of the central spinal cord canal without secondary cause. While there may be disturbed CSF flow, dilation of perivascular spaces, and subependymal cavitation, hydromyelia is not associated with tissue necrosis and neuronal injury. By contrast, syringomyelia is a progressive condition associated with intramedullary ischemia and tissue necrosis causing cavitation.

A filiform, or slitlike, appearance on MRI is said to distinguish hydromyelia from syringomyelia, but in some series 20% of patients with filiform dilations of the central spinal cord canal show progression consistent with syringomyelia. The presence of objective neurologic deficits and certain features on MRI (spinal cavity >6 mm in diameter and >5 segments in length; abnormal contrast enhancement; spinal cord expansion; cavity enlargement over time) or electrophysiologic testing (abnormalities on EMG, somatosensory evoked potentials [SSEPs], or motor evoked potentials [MEPs]) allows patients with syringomyelia to be distinguished more reliably from those with hydromyelia.

In the present case, the objective neurologic abnormalities were limited to reflex asymmetry and further investigations should be directed at differentiating between syringomyelia and hydromyelia. Structural features associated with syringomyelia (e.g., Chiari malformation type I [CM-I], spinal dysraphism, tethered cord, neoplasms) were excluded radiologically. SSEPs were normal. MEPs showed a prolonged central motor conduction time (CMCT) to the left but not the right lower limb (abductor hallucis CMCT was 24.8 ms on the left and 15.2 ms on the right; normal 16.0 ± 3.3 ms, mean ± SD). Needle EMG showed neurogenic change with fibrillation potentials and positive sharp waves in the right T8–T12 paraspinal muscles.

Questions for consideration:
1. What are the anatomical limits of the syrinx as defined electrophysiologically?
2. What additional radiologic investigations might help in terms of prognostication?
Syrinxes located centrally on axial MRI are usually asymptomatic. Only a minority produce clinical signs such as spasticity, hyperreflexia, sphincter disturbance, or sensory changes, and these tend to be of limited localizing value. By contrast, clinical signs are common if the cavity has a paracentral extension or is located eccentrically, and in such cases the signs are usually segmental and point to the location of the syrinx.6

Here, MRI demonstrated a centrally located syrinx; in keeping with previous reports, clinical signs were limited. However, electrophysiologic data reveal the syrinx to be functionally eccentric. It involves the thoracic ventral horn on the right (as demonstrated by the EMG) and the lateral corticospinal tract on the left (as demonstrated by the reflex asymmetry and corroborated by MEPs) (figure 2). While not clearly defined on examination, the sensory symptoms are likely to represent involvement of postsynaptic spinothalamic neurons crossing the midline anteriorly to ascend in the right anterolateral funiculus.

Beyond structural MRI sequences, cardiac-gated cine MRI can demonstrate abnormal CSF dynamics at the cranio-cervical junction and at the level of the syrinx.

DISCUSSION Syringomyelia classically presents with a centromedullary syndrome, manifesting as pain (burning, electric-shock like, radicular) and dissociated sensory loss with temperature insensitivity. Spasticity, autonomic dysfunction (including Horner syndrome), and sphincter dysfunction are also recognized.

The most common etiopathogenic association of syringomyelia is CM-I. In addition to the structural causes discussed in section 3, syringomyelia may arise as a result of trauma (including iatrogenic trauma), arachnoiditis/meningitis, and inflammatory myelitis. Such structural pathology alters the CSF dynamics, prompting CSF to be forced into the cord tissue and causing intramedullary venous congestion and cord edema. These result in macrocystic or microcystic changes.3 Where an obvious structural cause cannot be identified, the term idiopathic syringomyelia is applied.

Management is dictated by etiology and neurologic status. Secondary causes such as CM-Is, tumors, and tethered cords are usually amenable to surgery. Stable idiopathic syringomyelia with minimal neurologic deficits should be monitored radiologically and electrophysiologically at intervals of 3–6 months; significant progression should prompt consideration of surgical exploration. Syrinx shunting is rarely appropriate as it does not address any underlying etiology and is associated with high failure rates.7 In all cases, symptomatic treatment should be offered. Central pain often responds to carbamazepine, pregabalin, or gabapentin. Cramps can also be managed with gabapentin or potentially with diltiazem. Spasticity may be controlled with baclofen, tizanidine, or diazepam. Syrinxes can be exacerbated by activities involving a Valsalva maneuver, and patients should be counseled to avoid heavy lifting, to minimize coughing, and to ensure regular and soft bowel motions through increased fluid intake and use of laxatives if required.

Our patient was managed conservatively; carbamazepine proved ineffective and he opted not to try alternative drugs. Serial assessments at 6-month intervals demonstrated no functional or radiographic changes.

There are limited data regarding the natural history of syringomyelia. Several case series report that idiopathic syringomyelia with minimal neurologic symptoms only progresses in a minority of conservatively managed cases.7 Hence, the risks of nonintervention and regular monitoring appear to be limited in this patient group.

AFFILIATIONS AND DISCLOSURE
S. Jaiser is a Wellcome Trust clinical research fellow. M. Baker is a NIHR Clinical Lecturer and is supported by grants from UK Medical Research

Figure 2  Schematic cross-section of the midthoracic spinal cord

Small sensory fibers supplying pain and temperature sensation enter the dorsal horn, synapse in the substantia gelatinosa, and decussate in the ventral white commissure to ascend in the lateral spinothalamic tract (blue, illustrated for right-sided primary fibers only). Pyramidal neurons descend in the lateral corticospinal tracts to enter the ventral horn, where they synapse with lower motor neurons, which leave in the ventral nerve root (green). The asymmetrical lower limb hyperreflexia and the delayed motor evoked potentials to the left lower limb suggest a lesion of the left lateral corticospinal tract (lesion 1). A lesion of the right ventral horn is implied by the neurogenic changes on EMG of the right paraspinal muscles (lesion 2). The sensory symptoms probably represent a lesion of the postsynaptic spinothalamic neurons crossing the midline in the ventral white commissure (lesion 3).
Council, Wellcome Trust, and Academy of Medical Sciences. R. Whittaker is supported by a grant from the NIHR. D. Birchall reports no disclosures. P. Chinnery is a Wellcome Trust Senior Fellow in Clinical Science and receives funding from Parkinson’s UK, the MRC Translational Muscle Centre, and the UK NIHR Biomedical Research Centre in Ageing and Age-related Disease. Go to Neurology.org for full disclosures.

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Clinical Reasoning: A 39-year-old man with abdominal cramps
Neurology 2013;81:e5-e9
DOI 10.1212/WNL.0b013e31829a335e

This information is current as of July 8, 2013

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