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The diagnostic capabilities of modern magnetic resonance (MR) imaging means that once a decision has been made to investigate a case of suspected myelopathy or radiculopathy, few cases of syringomyelia will be undetected. A delay in diagnosis can still occur if the initial clinical assessment is incorrect, requiring a patient to be tolerant of the difficulties of identifying the less common neurological disorders (see Chap. 18). More often, nowadays, we are confronted with the opposite problem, that of over diagnosis, in cases of small cavities that may be unrelated to the presenting symptoms.

It is, therefore, sobering to consider that, within the lifetime of some clinicians still in practice, the diagnosis of syringomyelia depended upon invasive methods such as computer tomography (CT) and air myelography. The unpleasant nature of some of these tests, together with their attendant risks, meant that they were applied selectively. As a consequence patients were usually only diagnosed when their disease had reached a more advanced stage than that which we generally see today. At the same time, we can only admire the diagnostic acumen of the clinicians and the technical skills of the radiologists who correctly identified the condition in patients, in the pre-MR era. In this context, following account of the history of imaging of syringomyelia makes for interesting reading. (GF)

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21.1 Introduction

By the end of the nineteenth century, the structure and functions of the spinal cord were largely defined. Even so, the diagnosis of syringomyelia and other myelopathies could only be suspected on clinical grounds (Turney 1908). At the beginning of the twentieth century, invasive examinations were developed, to localise spinal cord diseases. Thereafter, the application of radiological examinations, in combination with clinical assessment, was better able to suggest the diagnosis of spinal cord lesions. For almost seven decades thereafter the diagnosis of syringomyelia was most likely to be made by myelography but even then the lesion may have been missed in many instances because myelography only defines the subarachnoid space and does not visualise the cord directly. By the end of the 1970s, it became possible to make the diagnosis of syringomyelia more reliably, using a less invasive method in the form of CT myelography. The subsequent development of magnetic resonance imaging (MRI) meant that the diagnosis could be made reliably, in most cases, by an entirely non-invasive imaging technique. This method has now largely replaced all other techniques, except when it is contraindicated for specific reasons or when implanted metalwork degrades the images.

21.2 Plain Radiographs

With syringomyelia, plain radiographic images of the spine may show widening of the spinal canal, in both the sagittal and coronal planes. The cervical spinal canal in particular is often large; in some cases the anteroposterior diameter is seen to be increased, in other cases the transverse diameter and in some instances both diameters are expanded. Cervical or thoracic scoliosis or kyphoscoliosis may also be a signs of underlying syringomyelia, particularly in children. The diagnosis of syringomyelia may also be suggested by the presence of other bone anomalies, such as segmentation abnormalities at the craniovertebral junction and subaxial levels. In some circumstances plain films can also reveal the

aetiology of a syringomyelia, in particular bony changes resulting from an old spinal injury or previous osteomyelitis or spinal tuberculosis (McRae and Standen 1966).

21.3 Myelography

Prior to the advent of CT, the definitive radiographic procedure used to diagnose syringomyelia was myelography, initially with positive contrast media. The earliest of these were oil based but later water-soluble compounds were developed. Subsequently, negative contrast studies were developed, using intrathecal air, injected either at lumbar or cisternal puncture. The latter technique was initially developed in the 1920s, as a means of obtaining samples of cerebrospinal fluid (CSF) (Ayer 1923).

21.3.1 Air Myelography

As early as 1918, Dandy developed the technique of outlining the spinal cord by using an intraspinal injection of air (Dandy 1919). The first reports of its use to localise intraspinal tumours came from Jacobus in 1921 and Dandy in 1925 (Dandy 1925). The technique was later refined to show the entire spinal canal; this involved complete replacement of the CSF with air and distension of the spinal subarachnoid space (Jirout 1958). The technique was difficult to perform and potentially hazardous for the patient, not least because the cisternal puncture required could cause medullary injury. Moreover, it was difficult to demonstrate a craniovertebral block with subarachnoid air. It was also desirable to avoid passage of air into the cranial spaces because, when this occurred, severe headache would ensue and the patient could not then easily lie still, which led to images of poor quality. Further, with air myelography, spinal roots were not shown, and many types of pathology, such as vascular malformations or arachnoiditis, were either not revealed, or the radiological appearances were easily misinterpreted (Heinz and Goldman 1972).

21.3.2 Oil Myelography

In 1922, two French investigators found, by accident, that iodised oils could be moved through the spinal subarachnoid space under the influence of gravity. They then proposed the intraspinal injection of lipiodol as a new diagnostic test, a method referred to as “myelography” (Sicard et al. 1923). Oil-based contrast media proved easier to use than air, and this quickly established oil myelography as the technique of choice, especially in the lumbar spinal canal. It was not until 1932 that the tilting radiographic table was developed and so the contrast was at first introduced by cisternal puncture. Lipiodol did, however, produce some unpleasant side effects, including pain and sensory disturbances in the legs. In 1940, the University of Rochester developed an alternative, ethyl iophenylundecilate, or iophendylate, known as Myodil in the UK and Pantopaque elsewhere. This medium was less viscous and therefore easier to use than the earlier Lipiodol, and it gave better demonstration of the spinal cord. It was very opaque to x-rays and special techniques such as tomography were not required (Peacher and Robertson 1946). It could be left in the spinal canal and rerun postoperatively, to check the adequacy of any surgical procedure (Dandy 1925; Copleman 1946). It gave even greater impetus to the application of oil-based, positive contrast myelography and was widely adopted in clinical practice. Because of its immiscibility with CSF, however, it tended to break up into globules, forming a layer in the spinal canal, which made it difficult to demonstrate both anterior and posterior surfaces of the spinal cord, unless large amounts were used. Nevertheless, for nearly 40 years, Pantopaque was generally the agent of choice, with air being reserved for special situations. Unfortunately, leaving oil-based contrast medium in the subarachnoid space resulted in inflammatory reactions in the meninges, leading to the formation of arachnoidal adhesions (Kendall et al. 1991). As early as 1941, Kubik proposed the removal of the iodised oil after completion of the myelogram, but it was some years before this practice became widely adopted (Kubik and Hampton 1941).

21.3.3 Water-Soluble Myelography

Ionic, water-soluble contrast agents were first used for myelography in the United States, in 1931. Compared to oily contrast media, they provided superior images of the cauda equina and root sheaths. Unfortunately, because of their initial irritating effects on the meninges, they never became popular (Almen 1969). Matters improved – indeed a revolution occurred in myelography – when metrizamide, a new, nonionic water-soluble medium, appeared in 1976. This was far less neurotoxic than were previous agents. It could certainly be used safely around the spinal cord, although its inadvertent passage into the head, when the contrast was being run into the cervical region, could cause generalised seizures (Skalpe and Amundsen 1975). This risk was reduced by introducing the contrast by lateral puncture, at C1-2 (Robertson and Smith 1990). Metrizamide was not entirely nontoxic and a second generation of nonionic agents, such as iohexol (Omnipaque) and iopamidol (Isovue), eventually replaced metrizamide. These agents did not cause arachnoiditis in the concentrations used in clinical practice (Sortland and Skalpe 1977). Pantopaque was withdrawn in 1987, a few years before law suits for chronic arachnoiditis began to be brought before the law courts.

21.3.4 Myelographic Study of Syringomyelia

Both positive and negative contrast studies (air myelography) were needed to make the diagnosis of syringomyelia (Ayer 1923). Myelographic findings, using positive contrast alone, either Pantopaque or metrizamide, were variable because the size of the cord varied with the position of the patient and with how much CSF was still present to support the cord. Myelography would initially demonstrate a non-specific swelling of the cord, which could just as easily have been due to an intramedullary tumour (Batnitzky et al. 1983). During injection of air, the supporting CSF was removed and, when the patient was subsequently tilted upright, the fluid inside the

syrinx would run downwards, leading to collapse of the upper portion of the cavity. The cervical cord shadow on the myelogram was then seen to be narrowed and this came to be known as the “collapsing cord” sign. It was regarded as being pathognomonic of syringomyelia (Heinz et al. 1966). In the case of intramedullary tumours, no change in the appearance of the expanded cord would occur with a change in the patient’s position. With syringomyelia, when the patient was moved back into the horizontal position, the fluid in the cavity ran back up into the cervical cord, causing an enlarged cervical cord shadow once again (McRae and Standen 1966).

21.4 Computed Tomography

The computed tomography (CT) scanner, designed initially for use on the head, was first described in the medical literature in 1973 (Hounsfield 1973). Soon afterwards, the method was applied to imaging of the spine. It provided valuable, cross-sectional perspectives of the spinal canal, formerly very difficult to achieve with conventional radiography. Its further use went on to revolutionise the radiological diagnosis of syringomyelia, and the first article on the use of CT in the diagnosis of syringomyelia appeared in 1975 (DiChiro et al. 1975). The cystic nature of a syringomyelia cavity could sometimes be recognised on standard CT scans, as a distinct area of decreased attenuation within the spinal cord, although this was by no means a constant finding. Administration of intravenous contrast media allowed neural tissue to stand out somewhat better, against CSF and syrinx fluid, but intrathecal administration of contrast media provided excellent visualisation of the cord, as well as other intradural structures within the spinal canal.

21.4.1 CT Myelography

CT myelography using metrizamide was soon developed, as a simple, safe and accurate technique for demonstrating the intrathecal contents of the spine (Di Chiro and Schellinger 1976). This technique was particularly helpful in the diagnosis of syringomyelia and remains so today,

for some patients who cannot undergo MR scanning. In many instances, however, a swollen cord still cannot be distinguished from any other intramedullary lesion. In other cases of syringomyelia, the cord appears normal in size or even atrophic. Moving the patient from the supine to the lateral decubitus position may be helpful, by demonstrating a change in the shape and size of the cord when a syringomyelia cavity is present (Ayer 1923). The appearance of the normal cord does not alter with changes in position on CT myelography.

The most useful feature of CT myelography, however, is the demonstration of contrast agent in the syringomyelic cavity within the spinal cord and this is a pathognomonic finding (Resjö et al. 1979). Although, in some instances, the cavity fills soon after the intrathecal injection of contrast agent, in most cases contrast fills the cavity after several hours, by diffusing through the cord parenchyma or indirectly via the fourth ventricle (Foster et al. 1980). Delayed CT scans are, therefore, often required, to demonstrate such filling. In cases, where the intrathecal metrizamide opacifies the cyst immediately, it suggests the presence of a direct communication between the syrinx and the spinal subarachnoid space.

21.5 Magnetic Resonance Imaging

In the 1980s, during a period when myelography was being greatly improved, MRI was introduced as a diagnostic tool. Within just a few years myelography was pushed almost into obsolescence. Although the first MRI images were of low quality, they were still able to distinguish relatively pure fluids, like CSF, from more inhomogeneous fluid collections. This made it possible to differentiate between the cavities seen in syringomyelia and those seen in neoplastic cysts. Subsequent developments have seen the introduction of better magnets, coils and sequences, and the price of the equipment has also decreased. With modern, high-resolution MRI, almost all intradural features, previously demonstrable by myelography, can usually be better shown by MRI. The development of fast T2 sequencing, at the beginning of the 1990s, meant that nerve

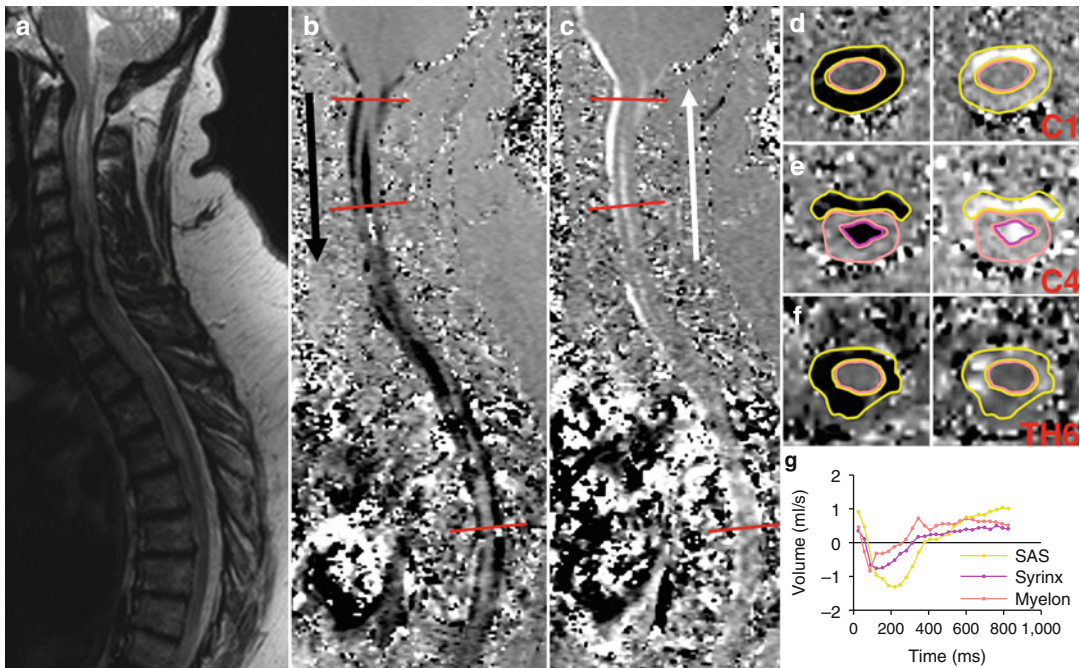


Fig. 21.1 MR flow imaging. (a) T2 sagittal MR image of craniovertebral junction and spinal canal. (b) Corresponding phase contrast MR images demonstrating caudal displacement of CSF (seen as *black*), during systole. (c) The same image during diastole, with return of

CSF in a cephalad direction. The three *red*, horizontal lines on images “a” and “b” show the levels of the transverse image pairs “d”, “e” and “f”, respectively. The graph “g” shows the flow volume, with time, across the cardiac cycle

roots could be visualised routinely. Myelography is now indicated only when MRI is contraindicated, such as when patients have cardiac pacemakers or are claustrophobic. Even then, open scanners can be used and can usually demonstrate syringomyelia cavities, albeit without such high resolution. Cardiac pacemakers which are MR compatible are also now available. Myelography may still have an occasional role, in the detection of arachnoid webs, that may elude detection by MR imaging. If there is a drawback to MR imaging, it is the increasing detection of dilated or persistent central canals, which are reported as abnormalities but which may not always be related to the patient’s symptoms (see Chap. 12).

21.5.1 MR Flow Studies

Phase-contrast magnetic resonance imaging is a powerful, non-invasive method for fluid flow analysis. In the past, this technique has been widely used for the investigation of cardiovascu-

lar blood flow, and it is well suited to the analysis of CSF flow at the craniocervical junction and in the spinal canal. The examination of pulsatile CSF flow requires synchronising the measurement of MR imaging data with the heart beat, thus obtaining velocity data of different points in time during the cardiac cycle. Phase shifts are measured in degrees, and their values should be within a range of ± 180 . In a phase image, the grey value of each pixel represents the velocity information in that voxel. Dark pixels show a high flow velocity in a caudal direction (Fig. 21.1b); bright pixels show a high flow velocity in a cranial direction (Fig. 21.1c). In-plane flow images thus provide *qualitative* visualisation of pulsatile CSF flow, whereas accurate *quantitative* evaluation of flow parameters can only be achieved using through-plane images, which have to be oriented perpendicular to the flow (Fig. 21.1d–f).

Although many studies of the CSF flow in Chiari malformation have been performed, the impact such measurements in clarifying the cause of syringomyelia is still a matter of debate.

Martin et al. summarised previous theories and their own results (Martin et al. 2010). We have performed quantitative pulsatile CSF flow measurements in patients with Chiari 0 malformation and Chiari type I malformation, without and with syrinxes of different sizes (Bogdanov et al. 2004). We compared CSF flow patterns in the subarachnoid channels, spinal cord and syrinxes in Chiari type I malformation patients and controls (Haughton et al. 2003). Our measurements cannot decide if CSF is driven into spinal cord or is generated in the spinal cord itself, but we can demonstrate the hydrodynamic steps in the development of the hindbrain hernia and the syrinx. Depending on the degree of flow obstruction at the foramen magnum, we see a significantly increased pulsatile CSF flow volume and velocity in the subarachnoid channels and spinal cord, beginning just below the craniocervical junction. Lower down, enlargement of the spinal cord by the syrinx impedes pulsatile CSF flow in the subarachnoid channels and allows the flow velocity to increase further. The fluid in the spinal cord, however, maintains nearly the same pulsatile flow volume and velocity as in the section above, just below the foramen magnum, due to the low hydrodynamic fluid resistance of the syrinx. The pulsatile hydrodynamic power is thereby transferred from the subarachnoid channels to the spinal cord and from the spinal cord to the syrinx. In the section of spinal canal below the syrinx, pulsatile flow power of the cavity itself is redistributed to the subarachnoid space, which provokes a transient increase in the velocity in the subarachnoid CSF channels. With progression of the disease, this hydrodynamic power decreases continuously in the subarachnoid CSF channels and increases correspondingly in the syrinx.

In our measurements, the subarachnoid space, spinal cord and syrinx appear to be communicating compartments that are in a hydrodynamic equilibrium. The subarachnoid channels have the lowest resistances and therefore allow the highest flow velocities. The diameter of the subarachnoid space decreases at the location of the syrinx. This leads to higher resistance and thus a decrease in the flow volume while the flow velocity increases or is maintained. The spinal cord has a high

hydrodynamic resistance, because of the higher tissue density of the extracellular space. Inside the syrinx, depending on the properties of the cavity, the resistance is similar or slightly higher than in the subarachnoid space. The transfer of the pulsatile flow wave from the subarachnoid channels to the spinal cord and syrinx leads to a time lag between the flow peaks of subarachnoid space and syrinx. The pulsatile force of the increased flow volume is transferred to the perivascular and extracellular fluid of the spinal cord and to the syrinx. The pathological CSF flow hydrodynamics appears to become self-reinforcing and transfers more and more hydrodynamic energy from the subarachnoid space to the syrinx.

It is a characteristic for the pathophysiology of syringomyelia in Chiari type I malformation that development of the syrinx does not start immediately below the foramen magnum but mainly somewhat more distal, from where it spreads upward and downward as the disease progresses. The accumulation of extracellular fluid decreases the cross-section volume of the subarachnoid space, which intensifies the already pathologically high subarachnoid flow disturbances. Under these conditions the accumulation of fluid in the spinal cord merges in a pre-syrinx and finally syrinx, which again increases the flow disturbances, by reducing the volume of the subarachnoid space. The subarachnoid channels, spinal cord, pre-syrinx and syrinx behave as communicating compartments, with different hydrodynamic resistances. Thus, the hydrodynamic power is more and more transferred or changed from the subarachnoid space, via the spinal cord, into the syrinx.

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