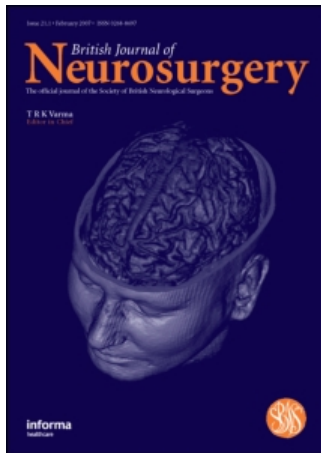


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PROCEEDINGS

Proceedings of the International Symposium Syringomyelia 2007
Keynote Presentations

Syringomyelia and the anatomical pathology of interstitial and cerebrospinal fluid drainage

Roy Weller (Clinical Neurosciences, University of Southampton School of Medicine, UK)

Syringomyelia from whatever cause and hydrocephalus are characterized by the accumulation of fluid within cavities in the CNS, which suggests that there is a failure of fluid drainage systems. This presentation reviews the anatomy and microscopic structure of fluid drainage pathways in the CNS and assesses why they may fail to clear accumulated fluid, in both syringomyelia and hydrocephalus.

Extracellular fluid in the CNS consists of cerebrospinal fluid (CSF) and Interstitial Fluid (ISF). CSF in the human CNS has a volume of 140 ml with 30 ml in the ventricular system and 110 ml in the subarachnoid space (SAS). The majority of human CSF is produced by the choroid plexuses in the ventricular system, at a rate of 350 $\mu\text{l}/\text{min}$, passes through the SAS to drain directly into the blood through arachnoid granulations and villi in the walls of the venous sinuses. Pia mater and the glia limitans separate the underlying brain from the CSF in the subarachnoid spaces. In rodents, sheep and other non-primate species, CSF drains through the cribriform plate of the ethmoid bone into nasal lymphatics to cervical lymph nodes. Such lymphatic drainage plays an important role in immune reactions in the brain in these species, but direct drainage of CSF to lymph nodes from the human brain appears to be minimal.

ISF is produced at rate of 0.1–0.3 $\mu\text{l}/\text{min}/\text{g}$ in the rat partly from the blood and partly from metabolic activity in the brain. Initially, ISF diffuses through the narrow extracellular spaces in brain tissue and then drains from the brain along the basement membranes in the walls of capillaries and arteries. In humans, ISF may drain to cervical lymph nodes along vessel walls, but in other species, ISF may join CSF to drain to cervical lymph nodes. The restricted nature of the ISF drainage pathways in blood vessel walls is emphasized in the elderly with the development of cerebral amyloid angiopathy in which ISF drainage pathways become blocked by the deposition of amyloid- β ($A\beta$) in vascular

basement membranes; this may also be a factor in the accumulation of $A\beta$ in the brain in Alzheimer's disease.

In the acute stages of hydrocephalus, CSF is forced into the periventricular white matter causing oedema, axonal destruction and reactive gliosis, but the grey matter areas do not develop CSF interstitial oedema. This suggests that CSF from the ventricles does not enter the tightly controlled extracellular spaces of grey matter, whereas it readily infuses into the white matter and expands the extracellular spaces when the pressure in the ventricles is raised. In both hydrocephalus and syringomyelia it seems that ISF drainage pathways do not have the capacity to clear excess fluid either from the cavities (ventricles and syrinxes) or from the surrounding CNS parenchyma.

In summary, there are two fluid drainage systems in the CNS, a high flow CSF system and a low flow, restricted ISF drainage system. When flow of CSF is blocked, the ISF drainage system does not seem able to compensate and fluid accumulates in hydrocephalic ventricles and in syringomyelic cavities.

CSF dynamics: from clinical practice to mathematical modelling and back again

Marek Czosnyka (Reader in Brain Physics, University of Cambridge, UK)

The most widely studied disorder of CSF circulation is hydrocephalus. Historically, this condition has been diagnosed using clinical and psychomotor assessment plus brain imaging. The role for physiological measurement to aid the diagnosis has become increasingly appreciated in a present-day clinical practice. This has been reflected by recently formulated guidelines for the management of normal pressure hydrocephalus [*Neurosurgery*, 2005;57(Suppl. 3)].

The use of physiological measurement in clinical neurosurgical practice is mainly related to recording intracranial pressure (ICP) and cerebral blood flow (CBF). Early studies, in the 1960s, defined various waves of ICP, the impact of added volume on mean ICP (pressure–volume curve) and the rate of re-absorption of CSF. These data were used to form mathematical models of CSF circulation. The first model incorporated formation, absorption and

storage of CSF fluid. More sophisticated models were then proposed, reflecting several compartments of CSF or interaction between cerebral blood flow and ICP. Identification of selected parameters then became useful in the diagnosis of hydrocephalus. Such variables as resistance to CSF outflow (R_{csf}), baseline pressure, its pulsatile component and elastance of the system are nowadays used in decision making of many disorders of CSF circulation, including hydrocephalus, craniospinal stenosis and idiopathic intracranial hypertension.

Disturbances of CSF circulation may be expressed quantitatively by an elevated resistance to CSF outflow. The upper limit of normal resistance is reported to range from 13 mmHg/ml/min in younger patients to 18 mmHg/ml/min in the elderly (Boon *et al.*, 1997). Elevated resistance to CSF outflow in NPH is taken as an evidence of impaired CSF circulation, which may be corrected by implantation of a shunt or third ventriculostomy. Elevation of this resistance is not, however, always correlated with a good outcome after shunting, particularly in idiopathic NPH in the elderly. At present, after years of disagreement, almost all centres consider R_{csf} as the most potent predictor for improvement after shunting, although its predictive powers are far less than 100%. High amplitude of pressure pulsation also seems to have predictive power for improvement after shunting. The significance of elasticity has never been studied in this context, but there are some data suggesting that increased elasticity (indicating a less compliant brain) correlates positively with better outcome after third ventriculostomy.

Whether a similar approach and the application of such physiological measurements can be adopted usefully with studies into syringomyelia, remains to be demonstrated. It is, nevertheless, clear that syringomyelia is a disorder of CSF circulation and that understanding CSF dynamics is as important with this condition as it is with hydrocephalus and other CSF disorders. Furthermore, in order to understand the pathogenesis of syringomyelia, it is important to take into account CSF movement in the craniospinal axis as a whole, as well as considering hydrodynamic disturbances locally, within both normal CSF compartments and within abnormal collections, such as syrinx cavities.

Pathogenesis of syringomyelia: implications for treatment

Edward Oldfield (National Institute of Neurological Disorders and Stroke, Bethesda, USA)

The pathophysiology underlying the development and origin of syringomyelia has been controversial, to some extent because of limited physiological information. Recent investigations using anatomic and cine MRI, combined with intraoperative ultrasound (IOUS), in patients with syringomyelia

associated with a Chiari I malformation, have shed some light on the mechanisms involved. They indicate that occlusion of the subarachnoid space (SAS) at the foramen magnum limits the free pulsatile movement of CSF between the cranial and spinal SAS, causes partial entrapment of the CSF in the spinal SAS and the transmission of excess pulsatile pressure waves in the spinal SAS. These result in syrinx formation.

To investigate this further, we performed a series of clinical protocols. Pre-operatively we carried out clinical assessment and anatomic and cine MRI. We also conducted pressure measurements, simultaneously from the syrinx and the cervical and lumbar SAS, at rest and with Valsalva and Queckenstedt maneuvers. We then recorded pressure measurements intra-operatively, from the same sites and also from a cerebral ventricle, together with IOUS monitoring of the spinal cord, the syrinx and the tonsils. Post-operatively we performed MRI and pressure testing in a manner similar to the preoperative testing. The pressure waves in the syrinx and SAS at the various sites were recorded digitally and the cine MRI and IOUS were related to the cardiac cycle (EKG), so that the physiological and anatomic changes that occur over the course of the cardiac cycle could be related and compared.

Similar studies were performed to assess the pathophysiology of recurrent syringomyelia and primary spinal syringomyelia.

The results, which confirm the hypothesis of the study, and their clinical significance will be presented and discussed in relation to other existing theories and in relation to the various types of syringomyelia that occur.

Clinical investigation of syringomyelia

Tatsuya Nagashima (Department of Neurosurgery, Kobe Children's Hospital, Japan)

Syringomyelia is associated with congenital anomalies of the spine or craniocervical junction and with trauma, tumours and arachnoiditis. Syrinx morphology is strongly influenced by the structure of the spinal cord and the underlying pathology. Signs and symptoms depend on the anatomical status of the central myelopathy. Scoliosis is a particularly important and common finding in children. Dysaesthetic pain is the most disabling sensory disturbance caused by syringomyelia.

Magnetic resonance (MR) imaging is the diagnostic modality of choice with syringomyelia. T1-weighted images are most useful for evaluating the anatomic characteristics of the syrinx and cerebellar tonsils. Axial images demonstrate extension of the syrinx into the dorsal horn of the spinal cord, on the same side and at the level of dysaesthetic pain. This provides a model for examining anatomic correlates of central pain. The presyrinx state is a condition of

reversible spinal cord oedema that precedes development of a well-defined syrinx. This state could provide a clue to answer the question of how fluid initially accumulates inside the cord. A recent biomechanical study demonstrated stress concentration at the same area of the presyrinx state in Chiari I malformation.

The cerebral spinal fluid (CSF) flow-void sign, representing non-uniform areas of decreased signal intensity in the CSF cavity, is due to spin rephrasing secondary to pulsatile CSF motion. Phase-contrast MR demonstrates abnormal velocity and pattern of CSF flow through the foramen magnum and pulsatile motion of the cerebellar tonsils in Chiari I malformation. MR CSF flow studies are useful in the intraoperative and postoperative evaluations of foramen magnum decompression for craniocervical abnormalities, as is evaluation of impaired CSF flow around the spinal cord associated with arachnoiditis. Hydrodynamic factors are considered to be associated with the pathogenesis of syringomyelia, and quantitative measurement of CSF flow has become the method of choice for elucidating the pathophysiology of Chiari I malformation, although application of the technique can be limited for a number of technical reasons.

MR imaging may show, incidentally, asymptomatic cavities in the spinal cord or mildly descended cerebellar tonsils. The incidence of a focal fusiform enlargement of the central canal is reported to be 1.5%. Asymptomatic tonsillar ectopia of less than 5 mm appears to be of no clinical significance in the majority of patients.

The role of myelography and CT myelography in the MR era is limited but these techniques are useful when MR imaging is unavailable or contraindicated due to metallic instrumentation. The syrinx cavity may be visualized by 4–24 h delayed images of CT myelography.

Intraoperative ultrasonography demonstrates pulsatile downward systolic movements of the cerebellar tonsils and synchronous constriction of the spinal cord surrounding the syrinx. These movements diminish after dural opening. Intraoperative ultrasonography has also been used to evaluate the effectiveness of foramen magnum decompression and to guide the exact placement of shunt tubes into syrinx cavities.

While the diagnosis of syringomyelia has been improved by MR imaging, theories regarding its pathogenesis remain mostly hypothetical. Further imaging and physiological studies are needed to elucidate key parameters of pathogenesis.

Chiari and hindbrain-related syringomyelia

Thomas Milhorat (Chiari Institute, New York, USA)

Chiari malformations comprise a heterogeneous group of hindbrain abnormalities that are characterized by

herniation of the cerebellar tonsils and association with syringomyelia in 30–65% of reported cases. There is accumulating evidence that Chiari I malformation (CMI), the most common type, is a disorder of the para-axial mesoderm that results in underdevelopment of the posterior cranial fossa (PCF), overcrowding of the normally developed hindbrain, and downward displacement of the cerebellar tonsils through the foramen magnum. Obstruction of CSF flow between the cranial and spinal compartments is the proximate cause of spinal cord cavitation. The resulting syrinx does not communicate with the fourth ventricle (non-communicating syringomyelia) and fills from the spinal subarachnoid space as a consequence of arterial pulsations that drive fluid through anatomically continuous extracellular spaces into the central canal of the spinal cord.

Since the advent of MRI, virtually every example of chronic tonsillar herniation that is not related to myelodysplasia (Chiari II) or cervical encephalocele (Chiari III) has come to be classified as CMI. This practice has made it difficult to arrive at a precise definition of CMI, and fails to weigh potentially significant aetiological factors that influence the clinical features, inheritance patterns and surgical outcomes of patients with the disorder. In this presentation, we summarize selected findings from a prospectively collected cohort of 2813 patients with tonsillar herniation of 5 mm or greater:

Association of CMI and hereditary disorders of connective tissue (HDCT): The diagnostic criteria for Ehlers–Danlos syndrome and related HDCT were met by 357 of 2813 patients (12.7%). In this subset, morphometric evidence of hypermobility of the atlanto-occipital and atlanto-axial joints, posterior gliding of the occipital condyles, and cranial settling was linked to retro-odontoid pannus formation and symptoms referable to basilar impression.

Association of occult tethered cord syndrome (TCS) and acquired tonsillar herniation: In a retrospective review of children and adults undergoing section of the filum terminale for occult TCS, traditional clinical criteria such as thickness of the filum terminale and low position of the conus medullaris (CM) were found to be of limited value in diagnosis. Morphometric evidence of elongation and downward displacement of the brainstem proved to be the defining feature of spinal cord tethering and was reliable to the 98th percentile for establishing the diagnosis and predicting short-term surgical outcome. Postoperative MRI's at three months demonstrated upward migration of the CM, ascent of the cerebellar tonsils, and reduction of brain stem length.

Pathogenesis and classification of CMI subtypes: Based on a morphometric analysis of brain and bone structures of the PCF in adult patients with CMI, the following mechanisms of chronic tonsillar herniation were identified: (1) cranial constriction occurring with classical CMI, craniosynostosis and

osteopetrosis; (2) downward traction of the spinal cord occurring with TCS and CMII; (3) cranial settling and cerebellar ptosis occurring with HDCT; (4) intracranial pressure coning occurring with hydrocephalus and space-occupying lesions; and (5) intraspinal hypotension occurring with CSF leaks and lumboperitoneal shunting.

Surgical treatment of CMI and syringomyelia:

The identification of CMI subtypes has been found to be indispensable in directing surgical strategies, avoiding complications and understanding the causes of failed Chiari surgery. Stratification of subtypes is also essential for the design of meaningful outcome studies. In patients with under-development of the PCF, optimal treatment consists of adequate decompression of the cervicomedullary junction, relief of the CSF block and re-establishment of normal CSF flow between the cranial and spinal compartments. No one operative technique is suitable for all cases and it is desirable to tailor surgical steps according to patient-specific variables. Intraoperative colour Doppler ultrasonography is an invaluable aid in assessing anatomical variables prior to opening dura, guiding the steps of surgery, and determining whether or not the goals of surgery have been met.

Scoliosis and Syringomyelia

Dieter Grob (Schulthess Klinik, Zürich, Switzerland)

Spinal deformities are observed in a number of conditions involving malformation of neural structures. Prominent amongst these is the scoliotic deformity seen with syringomyelia and Chiari I malformations. The relationship between these two entities is, however, still little evidenced in the literature, although there is a reported frequency of spinal deformities of 40–50%. The association of these conditions nevertheless means that two specialties need to be involved, to manage both the neural and the skeletal pathology.

Orthopaedic treatment aims to reduce pain, caused by asymmetric loading of the thoracolumbar and lumbosacral areas, and increase mobility by balancing the spine. These goals can be achieved by correcting the deformity. General orthopaedic principles of three-point correction, rotation and, with rigid curves, osteotomy of the spine (columnotomy) are applied. With appropriate fixation techniques the achieved reduction of the deformity is maintained, until bony fusion occurs.

These neuromuscular abnormalities produce some specific surgical problems. Associated curves have a tendency to rapid progression and a reduced capacity for “self correction”. This raises the question of the timing of surgery. Close observation of the patient is therefore needed. In cases where posterior decompression is contemplated, with its potential for producing skeletal instability, early consultation with a deformity surgeon is recommended.

Spinal fixation needs to cover the entire curve(s), in order to reduce the risk of adjacent decompensation and the appearance of deformities in the non-fused areas. Bony anatomy is often altered because adaptation to the deformity, occurring during growth, produces asymmetric pedicles which are often unilaterally hypoplastic. Osteoporosis and missing posterior elements are also seen. Associated intraspinal pathology increases the risk of neural damage occurring during the skeletal corrective manoeuvres. Critical areas are the occipitocervical junction, the level of any diastematomyelia and the lumbar region where there may be a tethered cord. Careful multimodal intraoperative monitoring, including SEP, MEP and EMG, all along the neural pathways, has been shown to be effective in prevention of neural damage.

In conclusion, the following statements can be made. The relationship between syringomyelia and Chiari malformation on one hand and spinal deformity on the other is not entirely clear. Orthopaedic treatment is indicated when progression of the curve and imbalance of the spine are manifest. Iatrogenic instability should be anticipated and treated at the earliest possible time, to prevent the occurrence of deformities. Correction is performed with appropriate instrumentation and, if necessary, columnotomy of the spine. The balanced spine provides improved quality of life by increasing mobility and sitting capacity and prevention of secondary degenerative changes, with their associated pain.

Paediatric aspects of syringomyelia and the Chiari malformations

Jerry Oakes (Department of Pediatric Neurosurgery, University of Alabama at Birmingham, USA)

Several issues currently challenge us with regard to the recognition, evaluation and treatment of hind-brain abnormalities and syringomyelia, each presenting us with a number of exciting new areas of investigation.

As with many issues, the definition of the entity can be much more difficult than it appears on the surface. How much caudal movement of the hind-brain is important to be considered abnormal and justify surgical treatment? Is the degree of caudal descent of the hindbrain the actual entity that should define a Chiari I malformation? From a pathophysiology standpoint, what comes first? Are most involved children initially affected by a small posterior fossa volume or is the ventral compression of the lower medulla the initiating event? The lack of free CSF movement in the spinal subarachnoid space can certainly be a cause of syringomyelia. How is this best demonstrated from an imaging standpoint?

Typical presentations in childhood and adolescence include scoliosis from a syrinx and valsalva-induced occipital pain but what about the third of children that present with unusual or even unique

presentations? The infant with rage attacks, or poor feeding from tongue/palate weakness, the child with constipation or essential hypertension or the teenager with more subtle findings. These challenge the primary physician to consider a structural neurological cause and justify obtaining an MRI, where the yield of surgically identifiable causes may be slim.

With regard to imaging, how useful are the more sophisticated MRI techniques? Is it really necessary to image the entire CNS? How much ventricular enlargement is abnormal? What is the definition of a tethered spinal cord? How does a tethered cord interact with the development of a syrinx?

Questions that will undoubtedly not be resolved at this conference are those that relate to surgical technique. Specifically, is a more minimalist approach to surgical intervention appropriate? What surgical technique gives the highest rate of syrinx resolution, and at what price with regard to complications? Are there ways to lessen the likelihood of serious post-operative complications? How much ventral compression justifies an initial transoral decompression?

There are many exciting areas of investigation with regard to this topic. The role of growth hormone deficiency and congenital rickets appear to significantly increase the likelihood of the development of a hindbrain hernia and syrinx. Patients with ligamentous laxity syndromes (Marfan's and Ehlers-Danlos syndromes) appear to be at risk as well. Within identical twin populations an involved patient easily justifies the investigation of the other twin.

In some ways, the role of the paediatric physician/surgeon is to identify and successfully treat affected children, so that they do not become adult patients.

Where does the cerebrospinal fluid come from in syringomyelia? Everything can be explained by bulk flow

Harold Rekaté (Barrow Neurologic Institute, Phoenix, USA)

The Pathogenesis of syringomyelia is controversial relative to the underlying causes, the source of the CSF in the syrinx, the importance of pulsatility and the proper treatment of the condition. The purpose of this review is to analyze sources of the CSF within the cavity, in an attempt to create a unifying concept of pathophysiology.

Beginning with a multicompartiment model, that has proved useful for the study of hydrocephalus, and assuming that the central canal of the spinal cord is analogous to the cerebral ventricles, it is possible to analyze the experimental evidence and clinical observations, to provide an improved understanding of syringomyelia. Each of the results summarized here are supported by strong, published basic science and clinical studies.

CSF is produced both by the choroid plexuses and the parenchyma of the brain and spinal cord. The

intact ependymal surface is an effective one way valve mechanism and, once inside the container lined by ependyma, CSF does not return to the white matter. The pial surface itself is impermeable to the flow of CSF in either direction, except within the Virchow-Robin Spaces. Therefore, to exit the spinal cord CSF must flow into the fourth ventricle to mix with ventricular CSF and exit the foramina of the fourth ventricle. This occurs either within the central canal or via the white matter tracks in the spinal cord.

Syringomyelia results when there is complete obstruction to flow of CSF between the spinal cord and the spinal subarachnoid space when there is an open central canal. When the central canal is obliterated spinal cord swelling results, a condition that has been called the "pre-syrinx state."

Adding insult to injury: the scourge of post-traumatic syringomyelia

Graham Flint (Queen Elizabeth Hospital, Birmingham, UK)

Syringomyelia remains a rare disease in the community as a whole. In contrast, post-traumatic syringomyelia is common in the population of spinal cord injury victims, with an incidence of about 1 in 20. The condition has the potential to add significantly to existing neurological deficits and handicaps.

Surgery for hindbrain-related syringomyelia is usually effective in terms of the anatomical result and relief of symptoms. Surgery for post-traumatic syringomyelia has a lower success rate. The reasons relate, in part, to the narrow diameter of the spinal canal as compared with the craniovertebral junction. Furthermore, the pathology underlying post-traumatic syringomyelia is different, consisting of obstructive subarachnoid adhesions, secondary to traumatic intradural haemorrhage.

Surgical intervention is by no means essential for post-traumatic syringomyelia. Many cavities enter a state of hydrodynamic equilibrium and patients may remain clinically stable over long periods. The situation is analogous to arrested hydrocephalus.

When surgery is contemplated for post-traumatic syringomyelia there are several options. We cannot be dogmatic about which is the best procedure, but instead we should define the alternatives, state the advantages and drawbacks of each and select the most appropriate in an individual case. Direct shunting of syrinx cavities may work in the short term, but in common with all CSF shunt systems, catheters will block in time. Even if a shunted syrinx remains collapsed a new one may form alongside, if the underlying obstruction to CSF flow persists. In addition, unless the syrinx presents at the pial surface, the myelotomy required for shunt insertion will cause some loss of dorsal column sensation in the victim who retains sensory function in the lower limbs. Furthermore, there is the question of which

receptacle is chosen for the drained syrinx fluid—the peritoneum, the pleural cavity or the spinal subarachnoid channels. Arguably, the optimal result is reconstruction of the spinal CSF channels, creating a conduit for CSF flow and allowing the syrinx to collapse. The problem is that subarachnoid scar tissue may reform postoperatively, blocking CSF flow once again and causing the syrinx to re-fill. Perhaps the simplest, least hazardous surgical treatment for post-traumatic syringomyelia is insertion of a lumboperitoneal shunt. This aims to lower the overall CSF pressure in the spinal canal and, secondarily, within the syrinx cavity. This procedure can be effective, but the overall success rate is not high.

Several questions remain regarding post-traumatic syringomyelia. Why do 19 out of 20 spinal cord injury victims escape this complication? Why do we not see hydraulic cord compression from without instead of CSF accumulation within the cord? What is the explanation for wide latent interval between injury and presentation of the syrinx, anything from 6 months to 30 years? Do all detected syringes need long-term radiological follow-up? Is there a role for operating upon syrinx cavities below the site of injury in a paraplegic patient? What is the place for spinal cord transaction? Finally, does early correction and fixation of a spinal fracture reduce the likelihood of a syrinx cavity developing?

Does idiopathic syringomyelia exist?

Jörg Klekamp (Christliches Krankenhaus, Quackenbrück, Germany)

The term syringomyelia derives from the Greek language and translates as a tubular cyst inside the spinal cord. Ollivier D'Angers introduced this name in 1827. The term 'idiopathic' is also of Greek origin and describes symptoms or diseases of unknown origin. To analyze whether idiopathic syringomyelia exists, we can approach the question from different angles. Does any cystic formation inside the spinal cord warrant the diagnosis of syringomyelia? Do we have a pathophysiological concept for the development of syringomyelia at all? If such a concept or theory exists, does it cover and explain all clinical observations? Can we identify additional pathologies in every patient with syringomyelia and how does the syrinx respond if such pathology is treated?

From a clinical and therapeutic standpoint it appears reasonable to reserve the term syringomyelia for progressive accumulation of fluid inside the spinal cord. This fluid may be localized inside the parenchyma or the central canal. Certain conditions should be differentiated from syringomyelia, including slit-like dilatations of the central canal which have no space occupying effect and are normal variants of no clinical significance. Gliopendymal or ependymal cysts are lined by collagenous tissue and cuboidal cells with no separate basement membrane. They

cause a marked expansion of the spinal cord, have a symmetric shape and do not display septations. Myelomalacia describes a defect inside the spinal cord that is the final stage of a traumatic cord lesion. Such a cavity does not expand with time. Cystic neoplasms contain fluid which has higher protein content than cerebrospinal fluid (CSF).

The pathophysiology of syringomyelia is not fully understood and a widely accepted theory does not exist. The author's current pathophysiological concept describes syringomyelia as a chronic interstitial oedema due to accumulation of extracellular spinal cord fluid.

Syringomyelia may be associated with almost any disease of the spinal canal. Even though we may not understand every pathophysiological aspect of syringomyelia, we can identify a disease process associated with syringomyelia in each and every patient. For most of these pathologies the development of the syrinx is related to obstruction of CSF flow inside the spinal canal, in close proximity to either the upper or lower pole of the syrinx. The only exceptions are intramedullary tumours and dysraphic malformations causing cord tethering. In other words, any patient with syringomyelia displays a region of CSF flow obstruction, an intramedullary tumour or an area of cord tethering.

The gold standard for treating syringomyelia is treatment of the underlying disease - removing intramedullary tumours, decompression of the foramen magnum for Chiari, resection of arachnoid scarring related to trauma, subarachnoid haemorrhages, infections etc, spinal canal decompression for spinal stenosis or disc disease and release of a tethered cord. For patients with a Chiari type I malformation more than 80% describe a stable clinical condition over more than 15 years following surgery. With arachnoid scarring, the prognosis is related to the extent and severity of scarring, between 70% and 25% experiencing clinical stabilization for at least 10 years. With intramedullary tumours prognosis is related to the tumour histology and the amount of resection. With cord tethering the complexity of the malformation is the main factor.

Even though the pathophysiology of syringomyelia is still somewhat controversial, the association of syringomyelia with other diseases of the spinal canal and the favourable response of the syrinx to successful treatment of this associated disease leads to a clear answer of the title question: idiopathic syringomyelia does not exist.

Canine syringomyelia: a painful problem in man's best friend

Clare Rusbridge (Stone Lion Veterinary Centre, London, UK)

Syringomyelia is emerging as a common canine spinal cord disease particularly in toy breeds such

as the Cavalier King Charles Spaniel (CKCS), King Charles spaniel and Griffon Bruxellois. The pathogenesis is not fully understood, but an important contributory factor is a mismatch between the brain and posterior fossa volume—Chiari-like malformation (CM). The CKCS is overwhelmingly over-represented, suggesting a genetic predisposition. At least 95% of CKCS have CM and as many as 50% have syringomyelia with the proportion of affected dogs increasing with age. Compared with unaffected CKCS, those with syringomyelia-related clinical signs have a smaller posterior fossa to total brain volume ratio. Other unidentified anatomical or environmental factors are, however, likely to be involved. Studies comparing intracranial dimensions have not demonstrated a significant difference in posterior fossa size in CKCS, with and without syringomyelia. In other breeds, such as the Griffon Bruxellois, syringomyelia may be present without CM.

Recent studies suggest that 35% of syringomyelia affected dogs exhibit clinical signs of pain. Some signs, such as vocalizing after sudden changes in posture, are likely due to obstruction of CSF flow. Syringomyelia also causes a neuropathic pain syndrome, probably due to spinal cord dorsal horn damage. Affected dogs behave as if they experience allodynia, appearing to dislike touch on certain areas of skin. Dogs with a wider syrinx are more likely to experience such discomforts, but dogs with a narrow syrinx may be asymptomatic, especially if the cavity is symmetrical and not involving the dorsal horn. Animals with a wide syrinx may also scratch at a localized area of skin. It is hypothesized that this behaviour is in response to unpleasant sensations, i.e. dysaesthesia.

Scoliosis is also common where there is a wide syrinx. This is thought to be due to damage to the dorsal grey column and unilateral loss of proprioceptive information.

The main treatment objective is pain relief. Cranial/cervical decompression surgery is successful in reducing pain and improving neurological deficits in approximately 80% of cases and approximately 45% may still have a satisfactory quality of life 2 years postoperatively. The syrinx, however, appears persistent in many cases and the clinical improvement is probably attributable to improvement in CSF flow through the foramen magnum. In some cases fibrous adhesions over the foramen magnum result in re-obstruction and 25–50% of cases eventually deteriorate again.

Medical treatment plays an important role in management of canine SM. The most commonly used drugs are those that reduce CSF pressure, (e.g. furosemide, cimetidine or omeprazole), non-steroidal anti-inflammatory drugs, neurogenic analgesics (e.g. gabapentin, pregabalin and amantadine) and corticosteroids. Prognosis for SM managed medically is guarded especially for dogs with a wide syrinx and/or with first clinical signs before 4

years of age. A small case series found 36% were eventually euthanized due to pain. However, 43% survived to be greater than 9 years of age—the average life expectancy for a CKCS is 10.7 years. Most dogs retain the ability to walk, although some may be significantly tetraparetic and ataxic.

Foramen magnum decompression with cranioplasty for the treatment of Chiari-like malformation in dogs

Dominic Marino (Long Island Veterinary Specialists, New York, USA)

Foramen magnum decompression (FMD) is the preferred mode of therapy for Chiari type I malformation in humans. Surgery is often successful, but 8–30% of patients require re-operation, due to excessive scar tissue formation at the FMD site. This scar tissue results in compression of nervous tissue and abnormal cerebrospinal fluid dynamics, perpetuating the secondary formation of a syrinx and resulting in neurologic deterioration. A FMD procedure for canine Chiari-like malformation (CLM) has been described; the success rate of this procedure is approximately 81%. Similar to humans, about 25% of canine patients require re-operation due to excessive post-operative scar tissue formation. In our experience, clinical signs of worsening associated with scar tissue impingement typically occur within 3 months of surgery.

A cranioplasty procedure developed for Chiari type I patients at the Chiari Institute has substantially reduced the frequency of post-operative scar tissue compression at the FMD site. Twenty-one dogs with MRI confirmed CLM underwent FMD with cranioplasty and had greater than 6 months follow up (median 11.2 months). Seventeen of 21 dogs were Cavalier King Charles Spaniels, the 4 remaining dogs were a Pug dog, Chihuahua, Pomeranian and a Maltese. There were 10 male dogs, and 11 female dogs. In two dogs, the surgery was a re-operation (excessive scar tissue following standard FMD). Mean age was 3.2 years (range, 10 months - 8yrs, 2 months). Following the FMD procedure previously described, guide holes for either 1.5 mm diameter (1.1 mm drill bit) or 2.0 mm (1.5 mm drill bit) titanium screws were made in the occipital bone, around the edge of the FMD defect. Self-tapping titanium screws were inserted into the holes for an approximate depth of 2–3 mm. A skull plate was fashioned using titanium mesh and polymethylmethacrylate (PMMA), and fixed to the back of the skull, using the titanium screw heads as anchor posts for the PMMA. The caudal aspect of the plate extended slightly over the dorsal defect of C1 and curved dorsally to avoid impinging on the medulla or cranial cervical spinal cord.

There were no intraoperative complications. Post-operative complications included a mild head tilt and

ataxia in 1 dog (re-operated cases), and the need for narcotic pain medications for about 2–3 weeks in 3 dogs (persistent neck pain). All but one of the dogs experienced clinical improvement following the procedure, although one has required a second decompression distal to the cranioplasty secondary to scar tissue formation. Mean follow-up time was 1 year (range, 6 to 16 months). Post-operatively, 2 dogs experienced a lack of improvement and 1 was euthanized due to recurrent pain around the head and neck. This latter dog also had severe PSOM and it was undetermined if the pain were due to CLM or the ear disease. One dog was re-operated because of connective tissue impingement of the cord at a site distal to the surgery.

Cranioplasty with titanium screws and titanium/PMMA plates appears to be well tolerated in dogs with CLM. Although cranioplasty appears to represent an improvement in the surgical management of CLM, it has not provided a solution. Intermittent exacerbation of clinical signs associated with excitement can be expected. The impact of cranioplasty on syrinx formation is currently under investigation as well as a review of dural biopsy findings.

The search for the gene(s) predisposing to Chiari I malformation with syringomyelia

Guy Rouleau (Notre Dame Hospital, Montréal, Québec, Canada)

Chiari I malformation (CMI) represents a common congenital abnormality of the craniocerebral junction, characterized by herniation of the cerebellar tonsils into the foramen magnum, often in association with syringomyelia (50–70%). CMI has an estimated incidence of 1 in 1200 with a higher preponderance in females versus males (3:2). The clinical presentation is usually delayed till the third or fifth decade and diagnosis is only confirmed by MRI. With the advance in imaging technology, CMI has been detected in increasing rates in younger population. The most common symptoms of CMI include headache (81%), brainstem or spinal cord dysfunction including scoliosis (30%) and syringomyelia (65–80%), ocular disturbances (78%), otoneurological disturbances (74%) and lower cranial nerve signs (52%). In advanced cases, CMI can lead to severe neurological deficit and permanent nervous system damage. The aetiology of CMI is thought to be multifactorial involving genetic and environmental factors. The developmental defect in CMI is thought to be the result of an underdeveloped occipital bone and posterior fossa originating from the para-axial mesoderm. CMI in humans is similar to a common condition in the Cavalier King Charles Spaniels (CKCS) breed, Chiari-like malformation (CM) that is characterized by occipital bone hypoplasia with foramen magnum obstruction and

secondary syringomyelia. This high incidence in a specific breed as compared with other breeds suggests the involvement of genetic factors in the aetiology of this disease. Pedigree analysis in a large database of over 5500 CKCS has suggested that CM/SM in the CKCS is inherited where all clinically affected dogs share a small number of common ancestors.

To date, no genetic factor predisposing to CMI in humans has been identified. The dog model is the only known naturally-occurring animal model for CMI in humans. Identification of genes defective in the CKCS will provide an entry point for identifying homologues involved in CMI in humans. We constructed a genealogy of more than 10,600 related CKCS dogs spanning 24 generations across three continents (North America, Australia and Europe) from over 600 MRI confirmed dogs. A whole-genome scan in 173 CKCS dogs followed by two-point linkage analysis identified six genomic regions with LOD scores above 1 on chromosomes 4, 5, 9, 11 and 15. We are currently investigating these potential regions for true linkage to CM/SM by additional genetic analysis in a larger sample size. Confirmed linked regions will be further delineated by linkage and association studies with the ultimate goal of identifying the causative mutation(s) using a positional candidate gene approach. Identification of the CM/SM gene(s) will allow the development of a genetic test for the identification of carriers for breeding purposes with the ultimate aim of reducing or eliminating this devastating condition in the dog.

In parallel, we have initiated the recruitment of a cohort of human CMI patients where we will investigate the human orthologue(s) of the dog CM/SM gene(s) for involvement in human CMI. The identification of a gene that contributes to the aetiology of CMI will provide an important step to the understanding of the underlying molecular and cellular pathogenic mechanisms, and the development of better therapeutic strategies. The finding of a predisposing gene may also lead to the development of simple and accurate diagnostic tests for better counselling, and clinical management of CMI patients and their relatives. Furthermore, this study will help unravel some of the cellular mechanisms implicated in the embryonic development of the craniocerebral junction.

Holter prize winning essay, 2007. Venous insufficiency as the cause of syringomyelia

Helen Williams (London, UK)

This presentation draws on existing published evidence to argue that syringomyelia is caused by inadequate venous drainage of the spinal cord. The Chiari malformation can result in increased tissue fluid pressure, which leads to cavitation of the cord.

Chiari malformation causes CSF obstruction at the foramen magnum, which may be intermittent, as described by Bernard Williams. Progressive cord damage occurs because of the influence of physical exertion on the position of the cerebellar tonsils and the spinal venous volume. Cases of syringomyelia are linked by mechanisms which limit the space available in the spinal compartment as it is influenced by physical movement. Elevated spinal pressure causes ischaemia.

The Chiari malformation may be caused by low spinal pressure and posterior fossa hypoplasia. This presentation describes the implications of these differing causes of foramen magnum CSF blockage.

The works used in the compilation of this theory date back to 1935 and include the observations of many contributors. This mechanism for the pathogenesis of syringomyelia may have been described at least in part by Dorothy Russell in 1963. It is consistent with the observations of all those who have published work on the pathogenesis of syringomyelia.

The Bernard Williams memorial lecture

How far have we come and where are we going?

Ulrich Batzdorf (UCLA School of Medicine, Los Angeles, USA)

The earliest descriptions of syringomyelia date back to the 16th century. The term syringomyelia was coined by Ollivier d'Angers in 1824. The relationship between syringomyelia and hindbrain as well as tonsillar descent was described by Cleland (1883) and Chiari (1891). Trauma as a cause of syringomyelia was first described by Struempell (1880).

Over the ensuing years, efforts to treat syringomyelia focused first on mechanical drainage of spinal cord fluid collections but these were at best only moderately successful. New theories of syrinx formation were introduced, and surgical treatment was designed on the basis of these developing concepts. Thus Gardner's hypothesis (1958) that fluid entered the cord by way of an opening from the fourth ventricle into the syrinx cavity led to the technique of obex plugging.

Bernard Williams, a man of extraordinary intellect and energy, was responsible for the beginning of modern concepts of the formation of syringomyelia, both in relation to cerebellar tonsillar descent and to spinal trauma. His unique experimental work with simultaneous measurements of ventricular and lumbar pressures in patients (1977), led to his concept of craniospinal pressure dissociation, which, in turn, provided the rationale for posterior fossa decompression to accomplish "inactivation of the filling mechanism" of the syringomyelia cavity. Posterior fossa decompression for syringomyelia related to hindbrain descent, although since modified, has stood the test of time.

Our understanding of syringomyelia and its treatment has, of course, been greatly enhanced since the introduction of magnetic resonance imaging, which in many ways has validated some of Williams' concepts. Oldfield (1994) subsequently provided a dynamic view of the cerebrospinal fluid pressure relationships at the craniocervical junction in patients with tonsillar descent.

Williams' interests were broad and included post-traumatic syringomyelia, an appraisal of shunting procedures and of terminal ventriculostomy, as well as orthopaedic aspects of syringomyelia. He analyzed his failures as critically as his good outcomes (1991), to the benefit of patients and improvements in surgical technique. There has been an information explosion on the subject of syringomyelia and related topics in the twelve years since Bernard Williams died but hardly a publication exists that does not cite his pioneering work.

The future lies in a better understanding of the genetic and molecular determinants of hindbrain development, of scar tissue formation after spinal injury and of factors affecting cell death or apoptosis. It lies in refinements of our understanding of cerebrospinal fluid pressure control and of flow dynamics at the level of the foramen magnum. Undoubtedly there will be refinements in surgical technique in the direction of minimally invasive procedures, which will not only diminish postoperative pain, but will also yield other benefits.

The Ann Conroy Trust: a catalyst, bringing together support, education and research, for the benefit of patients with syringomyelia and Chiari malformations

Tony Kember (Chairman, The Ann Conroy Trust)

The Ann Conroy Trust (ACT) aims to help victims of Chiari malformations and syringomyelia understand and manage their condition. Hundreds of concerned, newly-diagnosed patients call our helpline every year, desperate for information and seeking reassurance about this rare condition. Our counselors are themselves sufferers and are very positive about managing the condition, giving emotional support to callers, who are encouraged to take on a positive frame of mind. Counsellors do not, however, offer medical advice.

The ACT also provides educational material, in both printed and electronic form. Our publications are intended to help sufferers understand their condition, making it easier for them to comprehend and assimilate what their own specialist says to them.

The Charity has a group of affiliated neurosurgeons, currently based mainly in the UK. Their contact details, as specialists with an interest in syringomyelia and related disorders, are available to patients and general practitioners, on the ACT website. The Charity organizes regular meetings of

this British Syringomyelia Group, so that interesting and difficult cases can be discussed in detail. Currently, this group is also planning to establish a database, to record anonymized case histories and provide material for future clinical research.

The ACT is also setting up the Bernard Williams travelling scholarship, to support UK-based trainee neurosurgeons who wish to study, specifically, aspects of the treatment of syringomyelia or Chiari malformations.

The organization of Syringomyelia 2007 is the Charity's most ambitious project to date. This event will gather and then disseminate the latest knowledge and understanding about syringomyelia, Chiari malformations and other related disorders. We also hope that the event will act as a springboard for us to offer our three principal services—support, education and research—to sufferers and medical professionals worldwide and not just in the UK.