HYDROSYRINGOMYELIA DUE TO CONGENITAL AND ACQUIRED LESIONS, COMMON MECHANISMS, SEARCH FOR A LOGICAL TREATMENT

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The term syringomyelia, meaning a cavity in the spinal cord, was coined by Charles P. Ollivier-D'Angers in 1927 but it was James Gardner who was the first to realize and to preach that this was not simply a degenerative disease but a condition which was due to penetration of CSF into the cord substance as a result of hydrodynamic forces. He no doubt, would have been very pleased to see the ciné MRI pictures which you have just shown me and which illustrate well the pulsations of the CSF in the subarachnoid spaces and the syrinx, as a result of heart action. Gardner felt that the arterial pulses were the main force that pushed CSF into the central canal and formed the syrinx, in cases where hind–brain herniae or arachnoiditis restricted the flow around the cord and medulla at the foramen magnum. Others, and particularly Williams have felt that venous pressure changes resulting from coughing, respiration, straining etc. are perhaps more likely to induce this cavitation than arterial pulsations alone and I have certainly found very good evidence of this in many of my patients who have developed symptoms while lifting heavy objects, coughing, swimming underwater, etc. I should like to review with you the experience of close to 100 cases of various forms of syringomyelia, not due to intramedullary tumours, to see how these cases help us understand the mechanism of this disorder and to devise a logical treatment for it.

From what I have heard this morning, I will not have to convert any of you to the idea that hydrosyringomyelia of the classical type, associated with the Chiari malformation or basal arachnoiditis, results from hydrodynamic forces but I would like to extend this concept to post-traumatic syringomyelia and to post-arachnoiditis syringomyelia and to show you evidence that they also are caused by CSF forced into the spinal cord by pressure gradients between poorly connecting compartments. Other forms of syringomyelia such as the type which develops with intramedullary tumours seem to have a different origin. The fluid within the tumour associated cysts contains large amounts of proteins, to the point that it sometimes jells in the syrinx, as it cools. These intramedullary cavities are, I think, similar to the cysts one finds around cerebellar hemangioblastomas or astrocytomas and are due to seepage of fluid and proteins from tumour vessels. In certain cases of cystic tumours however it has been shown that water soluble contrast,
injected in the subarachnoid space, can penetrate the cavities as it usually does in hydrosyringomyelia. It could be that some of these tumours partially obstruct the flow of CSF causing compartments in the subarachnoid space above and below the lesion and that hydrodynamic forces can play on these and pump CSF into the cavity in the same way as they do with arachnoiditis or in cases of trauma to the cord. Finally, that leaves a small group of "idiopathic" syringomyelias which develop usually as small cavities in the cervical cord without any history of trauma, arachnoiditis, without hind-brain malformations and, so far without any reason.

The classical type of hydrosyringomyelia develops as a result of a partial obstruction of the foramen magnum. The most common cause is a Chiari I or II malformation. We have seen this in approximately two-thirds of the classical cases. In close to one-third the obstruction is due to adhesive arachnoiditis at the foramen and when these patients are questioned, one often finds a history of a difficult birth with application of forceps, cyanosis at birth etc. So that I feel that a large proportion of these cases of arachnoiditis are in fact due to trauma to the cranio-cervical junction at birth. These adhesions are often associated with a Chiari malformation as well. Basilar invagination can also produce syringomyelia. Whereas in the Chiari malformation, the cerebellar tonsils are too low in relation to the foramen magnum, in basilar invagination it is the foramen magnum which is too high. But the net result is the same, a strangling and partial obstruction of the flow of CSF between the cranial compartment and the spinal compartment. We can see the same thing in acquired basilar invagination such as may happen with Paget's disease of the skull. The softened base of the skull invaginates upwards, the tonsils jam into the narrow foramen magnum and syringomyelia develops. In all these cases, the CSF which cannot escape freely out of the IVth ventricle, because the cerebellar tonsils or adhesions are obstructing the foramen of Magendie is forced into the central canal which gradually distends then ruptures into the substance of the cord. The central canal is usually narrow just below the IVth ventricle, before it widens out in the neck and the connection between the syrinx and the IVth ventricle is often missed by the pathologists. With magnetic resonance, which is the investigative tool of choice in this condition, one can see this thin connection in almost every case. The central canal is thinnest there for a number of reasons the first one probably because the pressure of the herniated cerebellar tonsils squeezes it at that level. The second may be because the anatomy of the grey matter changes radically at the junction of the cord and medulla. It no longer has the classical butterfly shape but breaks up into a number of islands because of the decussations of the pyramids and of the posterior columns. The central canal may be more patent when there are no tonsils behind the upper cord but only adhesions and we have been able to fill it easily with Ethiodan from a ventriculogram in some of these cases. In fact, a narrow central canal at the top may be more dangerous to the patient than a patent one since fluid forced into the cord from above may not come back out again so readily if the canal is very thin.

The fluid in the syringomyelic cavity in cases of foramen magnum lesions usually has a very low protein content, 15 to 25 mg %, more like ventricular fluid but I have never seen it at more than 100 mg % unless there was also a tumour.
If one considers that the syrinx-filling mechanism in syringomyelia is the obstruction of CSF flow at the foramen magnum, the most logical treatment would be to try and reestablish a normal flow. We have to think however that there is flow restriction mainly to large volumes of CSF moving quickly, sometimes one way, then the other way, as happens with coughing and straining and not just to the trickle of CSF secreted steadily by the choroid plexus. Anyone who has watched a myelogram done with Ethiodan under fluoroscopy and has asked the patient to cough, has seen the droplets of oil dance up and down very fast at the foramen with this manoeuvre. Hopefully, we may be able to see this CSF movement with coughing on MRI very soon. To accommodate this large flow, a small catheter joining the IVth ventricle to the subarachnoid space may not be very effective and my inclination has been to try to create a new cisterna magna in these patients. If the bony foramen magnum is invaginated, I think the bone has to be removed as widely as possible. If the cerebellar tonsils are long and in the way, they can be taken out by suction under the microscope, taking care not to coagulate the larger tonsillar arteries which continue on to the cerebellar hemispheres. The more medial and inferior portions of the tonsils can be sucked out from within their arachnoid without injuring the larger vessels and taking great care also not to injure the small perforators from PICA to the medulla. When the tonsils are removed, the floor of the IVth ventricle generally becomes visible and the entrance of the central canal at the obex connects no longer only with the IVth ventricle but with the cisterna magna and the cervical subarachnoid space, it now lies in the same compartment as the subarachnoid space of the upper cervical region, and there are no longer pressure gradients between the IVth ventricle and the subarachnoid spaces. To prevent adhesions from forming and closing the IVth ventricle again, the empty arachnoid from where the tonsillar tissue has been removed can be turned up and tacked with one small suture on either side leaving the foramen of Magendie wide open. With a blunt nerve hook one can gently palpate along the floor of the IVth ventricle and feel the entrance of the central canal. If the hook engages in the canal, small pieces of muscle can be pushed in very gently to obstruct it and sometimes, as Gardner suggested, a tiny whisp of cotton may be placed on top of the muscle in the hope that it will cause some adhesions at that point and keep the muscle from floating out again. One has to be gentle with this manoeuvre and the anaesthetists should monitor the pulse which may slow down if one is too aggressive. One of our cases also developed a unilateral XIIth nerve palsy following too vigorous an insertion of muscle. I am not sure if this is an essential part of the procedure but it is easy to do and I have always done it whenever I had easy access to the floor of the IVth ventricle. For many years, one of these pledges of muscle was marked with a clip and these clips have always been visible, in position, on post-operative films even many years after. Whether the central canal was still effectively obstructed or not however, remains a question.

In order to create a large cisterna magna, the dura should not be closed on itself. Some surgeons leave it open, I have preferred to close it with a Lyodura graft which is sutured in a fashion as water-tight as possible. In reviewing the complications and side effects of this procedure, it has been very clear that many of the problems which arose post-operatively were due to blood in the subarachnoid space. The incidence of fever, aseptic meningitis, headaches, nausea, vomiting, hydrocephalus and pseudomeningoceles was markedly reduced when more attention was given
to a water-tight closure and when we started using epidural suction drains routinely. In the few instances where we have returned to the operative site, the Lyodura was still intact and had not formed adhesions at the cisterna magna.

When one has to deal only with a Chiari malformation, which occurs in approximately two-thirds of cases, it is quite easy to achieve the creation of a large cisterna magna but things are more difficult in cases of basal arachnoiditis with or without herniated tonsils. If the band of adhesions is relatively narrow, the adhesions can be dissected or simply bypassed. It is dangerous to dissect adhesions along the dorsal aspect of the cord because of the PICA and their branches which can easily be injured. If it is not possible to dissect safely these adhesions, they should be left alone and other procedures such as introducing a tube in the IVth ventricle or shunting the syrinx can be resorted too. Even though there may still be adhesions left in place, it may be useful to carry out a duraplasty to enlarge the dural tube.

In the case of simple Chiari malformations, the procedure involves usually only a suboccipital craniectomy and removal of the posterior arch of C1. Very often the syrinx is not visible or is very small at the higher cervical region and the procedure remains limited to the structures of the cisterna magna. However, if there are adhesions or if the cyst is clearly seen at the top, we have often also placed a syringo-subarachnoid shunt. We usually use infant size shunt tubing for this. The tube is most of the time introduced at the dorsal root entry zone where the cord is generally thinnest unless the syrinx involves the dorsal columns which may then be reduced only to a thin arachnoid. A single tube or two small tubes placed in an X, one directed above and one directed downwards and tied together at the opening in the cord have been used at times. This has usually required extending the laminectomy down to C4 but on one side only.

Now that magnetic resonance imaging is available for follow-up of these patients, all the ones who come back are submitted to this examination. A cavity is still visible in approximately two-thirds of the cases even if they are stable clinically. The cavity is generally smaller than what it was on preoperative studies although the comparison may sometimes be difficult between myelographic or CT studies done preoperatively and MRI studies done at follow-up. In others, no cavity can be seen at all although one was demonstrated preoperatively either by penetration of metrizamide in the syrinx, by cord puncture at surgery or by the very large size of the cord before. I have not reoperated in the cases where the MRI still showed persistence of the syrinx unless the patients were deteriorating clinically and in these cases, I have carried out syringopleural shunts. Following these, the syringomyelic cavity has disappeared on MRI and the deterioration has stopped but I am concerned that if we have not disabled the filling mechanism of the cord, these tubes in a collapsed cord may obstruct and that a cavity may form again in the same place or next to the old one.

Even when the syrinx disappears on the MRI, some of the symptoms due to the cord cavity usually persist. The suspended dissociated anaesthesia, the abnormal reflexes, the atrophy and weakness may improve but do not disappear completely. The patients may think that they have
better sensation or power, less ataxia, or stiffness but I find a real objective measurable improvement in only about 25% of cases. Approximately 50% are stabilized but not improved objectively and approximately 25% eventually deteriorate again. The symptoms attributable to the Chiari malformation however do better particularly the headaches in the occipital region which used to follow coughing or straining or even laughing usually disappear. The oscillopsia and the nystagmus improve frequently in a way that can be measured. The dysphagia and the dysarthria also tend to improve. The deep boring pain characteristics of many cases of syringomyelia is very unpredictable, it is sometimes improved and may completely disappear even if some cavity remains visible on MRI, it may persist even when the cavity has disappeared completely. It is a sort of deaffrentation pain which sometimes responds to Tegretol but is generally resistant to analgesics.

In cases who also have hydrocephalus or who develop some post-operatively, ventriculo-atrial or ventriculo-peritoneal shunting is of course carried out. Five of my patients had had ventricular shunting done before their suboccipital craniectomy. Only one of these has remained stable clinically with ventricular shunting alone, she still has a rather large somewhat distended syrinx but she has shown no deterioration in her symptoms for almost ten years and still refuses surgery for the Chiari malformation. The four others deteriorated in spite of a functioning shunt with reduction of the size of the ventricles, until the foramen magnum was decompressed.

In cases of post-traumatic syringomyelia and in cases secondary to arachnoiditis, I think we can postulate the existence of similar hydrodynamic mechanisms to explain entry of CSF into the spinal cord and progression of the syrinx as in cases secondary to foramen magnum abnormalities. In trauma, where the spine is fractured and the cord compressed, there is usually pulping and hematoma formation which is most marked in the central part of the cord. Adhesions generally form around the contused cord causing more or less complete obstruction of the CSF pathways. This complete or relative obstructions results in the creation of two CSF compartments, one below and one above the lesion between which pressure gradients can now appear. After the hematoma absorbs, one is left with a mushy soft cord in that area which looks like wet cotton when it is explored. It is easy to see how CSF could penetrate in it through fissures and enlarged perivascular spaces. Paraplegic patients often use a Credé manoeuvre to empty their bladder. This of course raises the CSF pressure in the lumbar compartment below the lesion and fluid can be forced into the cord rather than around it. As more fluid is pumped up in this way, it dissects further up in the grey matter and eventually may reach the cervical region. These paraplegic patients then notice the appearance of weakness and sensory changes in the hands, progressively ascending, abolishing reflexes in the upper extremities and eventually making life very difficult for them since they may lose the power to transfer from their bed to their chair. If the traumatic lesion is high enough in the cord, another cavity may form below the traumatized level as a result of other pressure changes where at one time the pressure was greater in the upper compartment than in the lower one. If one exposes these lesions by opening the dura above and below and keeping the arachnoid intact, it is possible to see that pressures in the two compartments do not rise simultaneously with respiration or pulse but that one lags behind the other. The differences in pressure must be much greater during strenuous voluntary activity or
with coughing than what one can observe during anaesthesia. When these patients are questioned, they often relate the appearance or the increase in their symptoms to such muscular activities.

We have seen similar phenomena in two patients who developed focal arachnoiditis in the thoracic region, one as a result of infection of a dorsal column stimulating electrode which had been placed for pain resulting from failed lumbar disk operations and the second one after a meningitis resulting from a CSF leak at a lumbar level. In both cases, the adhesions were limited to approximately one segment around T9 but cavities had developed in the cord below the lesion causing weakness of the legs and sphincter disturbances and also above the lesion up to the cervical region causing reflex changes in the arms, sensory loss to pin prick and temperature and pain in one arm. It is a little more difficult to explain how the CSF enters the spinal cord in cases of arachnoiditis where the cord is not crushed as in trauma but, because it is adherent circumferentially to the dura, the cord may get teased radially when the dural tube tries to expand with changes in pressure and fluid may enter through clefts or along perivascular spaces and gradually the pressure gradients will pump more fluid into the cord substance above and below the block.

Again, as in classical syringomyelia we have an arrangement of two compartments which communicate poorly with one another. A spinal compartment below the arachnoidal block and another one above. This is similar to what exists in a Chiari malformation, the only difference is that in the Chiari malformation, the fluid enters the cord through the central canal which distends and ruptures into the cord substance whereas in syringomyelias resulting from trauma or adhesions, it enters at the site of the trauma or adhesions and progresses in the cord parenchyma directly.

If the mechanisms are similar to those of classical syringomyelia with a Chiari malformation, then one should try to apply the same therapeutic principles and it is possible to do so at least if the area of adhesions is not too extensive. By doing a laminectomy extending above and below the area of adhesions and opening the dura and dissecting these adhesions, the cord may be allowed to fall to the floor of the canal. A Lyodura graft can then enlarge the dural tube behind the cord allowing fluid to flow freely behind it between the top and the bottom subarachnoid compartment therefore abolishing or reducing the pressure gradients between these compartments. I think we have been able to achieve this in a few cases but this is easier said than done. If the adhesions are extensive over many levels, it is probably futile to attempt this type of treatment and one has to resort to draining the syrinx. Some of the cases on which we have performed this dural graft already had syringo-subarachnoid shunts in place which had given only temporary improvement. Adding a long dural graft to bypass the adhesions and replacing the blocked subarachnoid shunt has brought on improvement still lasting after 5 years in 2 patients with disappearance of the thoracic and cervical cavity on MRI. But in other cases, we were not able to establish this free bypass as proven by follow-up myelograms and the syringo-subarachnoid shunt placed at previous operations had not helped either. We have therefore reexplored the patient placing this time a syringo-peritoneal or syringo-pleural shunt, a straight tube without valves from the syrinx to a low pressure area and follow-up MRI scans have shown a collapse
of the cavity. Follow-up for this type of shunt is still short, less than two years and it is premature to say if this will control the situation permanently.

I now think that an important step in the evaluation of the post-traumatic and post-arachnoiditis syringomyelia is the performance of water soluble contrast myelograms from above and from below. I feel it is essential for the surgeon to be present when these myelograms are done so that he can watch the progression of the contrast in the subarachnoid space and better judge the degree of obstruction, the loculations etc. When X-rays are done or CT scans a few minutes after the injection, the contrast has usually diffused more or less uniformly and one is not able to judge the presence of loculations as well as by watching under the fluoroscope. If the area of adhesions is very long, it is probably simpler to proceed immediately to a syringo-pleural or peritoneal shunt. If there are two cavities above and below the block, it may be necessary eventually to do two shunts one for each cavity. If the area of adhesions is short I would favour a local exploration and dural graft with or without subarachnoid shunting for the same reason that I prefer to decompress the cisterna magna. If this is successful, it disables the filling mechanism and one does not have to rely on the continuing patency of a tube to control the syringomyelia.

In summary, I feel that post-traumatic and post-arachnoiditis syringomyelias are also due to entry of CSF into the spinal cord and could be called hydrosyringomyelias. Like classical syringomyelias caused by the Chiari malformation, basal arachnoiditis or other anomalies of the foramen magnum, they are due to CSF pressure gradients between poorly communicating CSF compartments above and below a zone of arachnoidal adhesions. In order to stop CSF from entering the cord, these compartments have to be more freely connected together and this can be done by dissecting and bypassing the adhesions with a “augmentation duraplasty” or by decompressing and enlarging the cisterna magna. If this proves to be impossible to achieve usually because the arachnoiditis is too extensive or if it fails because adhesions form again, the syrinx should be drained. It will collapse more completely if the shunt is made to a low pressure area such as the pleural or the peritoneal space. Water-tight closure of the dura and epidural suction drain are essential to reduce the amount of blood in the subarachnoid space and reduce the incidence of post-operative complications. In my opinion, it is more logical to prevent the syrinx from filling than to drain it but draining being a simpler procedure can be the best choice in certain cases.