Dynamic Chiropractic

CHRONIC / ACUTE CONDITIONS

Craniocervical Hydrodynamic Failure and NPH

THE ROLE OF CRANIOCERVICAL SYNDROMES, THE AGING DURA MATER AND PRESSURE CONUS IN CHRONIC NPH

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INTRODUCTION: the artificially deformed crania

In the first part of this series of articles I talked about the relationship between the craniosacral primary respiratory rhythm and cerebrospinal fluid (CSF) movement through the brain. In the second article part we talked about the role of the accessory drainage system in upright posture. In this part I will be discussing the potential role of craniocervical syndromes and hydrodynamic failure in chronic normal pressure hydrocephalus (NPH).

While I was at the museum of Natural History in New York, in addition to the normal crania, I also got to study many artificially deformed crania from Peru and Bolivia. With regards to deformed crania, infant crania are sometimes unintentionally deformed such as from contact with cradleboards, or by strapping their heads to papooses. Similar types of deformities are occurring today as a result of keeping infants flat on their backs to prevent SIDS. Many diseases also cause deformation of the cranium such as Padget's disease and hydrocephalus. But the most unusually deformed crania are the artificially deformed crania from Peru and Bolivia. These crania were intentionally deformed into bizarre shapes. The question is why did these people choose to deform their children's heads?

Besides their intriguing shapes, and that fact that they survived such extreme cranial deformation it was also interesting to note that the sutures in the mature deformed crania were wide open. It seemed likely to me that the delayed closure of the sutures was due to cranial hydrodynamic failure and hydrocephalus. Also of note was other crania from this region that had trephinations, which are holes that appear to have been chiseled out by stones.

No one knows exactly why these people chose to deform their children's heads. Perhaps they did it for aesthetic purposes? Many cultures in fact have used body modification including our own. But even if they did shape children's heads for aesthetic purposes, why did they carve holes in mature crania? On the other hand, perhaps these people somehow knew and were attempting to control hydrocephalus and increased intracranial pressure (ICP). Close breeding may have perpetuated an inefficient design of the CSF pathways, the cranial dural sinus, or accessory drainage systems. This could have increased the incidence of obstruction and hydrocephalus.

NORMAL PRESSURE HYDROCEPHALUS

Hydrocephalus is condition in which CSF volume increases as a result of an imbalance between CSF formation and absorption rates. Because the sutures are still open in childhood, hydrocephalus causes both the ventricles and the skull to enlarge. On the other hand, because the sutures are relatively closed in adults, hydrocephalus likewise causes the ventricles to enlarge, but instead of enlarging the skull, in an adult it compresses the brain.

Typically, hydrocephalus is associated with an increase in CSF volume, an increase in the size of the ventricles, and an increase in intracranial pressure (ICP). It is usually caused by blockage of

CSF pathways. NPH, on the other hand, is both unique and an enigma. First, it is unique because in NPH, CSF volume increases and the ventricles enlarge, but in contrast to hydrocephalus ICP, that is CSF pressure, remains normal or just slightly elevated. Second, NPH is an enigma because blockages are rarely found beginning in the ventricles where CSF is produced, through the subarachnoid space and into the arachnoid granulations in the superior sagittal sinus where it is finally absorbed into the drainage system of the brain.

In addition to blockages of the CSF pathways, however others have considered blockages of the dural sinuses from the superior sagittal sinus on down to the drainage outlets at the base of the skull. In an old textbook on neurology by Adams and Victors it states:

"A matter of considerable practical as well as theoretical interest is whether blockage of the dural sinuses into which CSF is absorbed can result in tension hydrocephalus. The problem is that blockages are rarely found."

That is, not only is it rare to find blockages in the CSF pathways in NPH but it is just as rare to find blockages in the cranial dural sinuses. We haven't, however, looked for blockages of the dural sinuses outside the cranium in the craniocervical spine. Furthermore, we haven't considered the effects of hydrodynamic failure rather than outright blockage of cranial dural sinuses and accessory drainage system.

The intracranial dural sinuses extend outside the cranium into the craniocervical spine. The extracranial portion of the dural sinuses includes the VVP and the suboccipital cavernous sinus system. The suboccipital cavernous sinus is located between the atlas and the occiput and is anatomically similar to the cavernous sinus. It has connections with both the cranial dural sinuses and the VVP. Because of the relationship between the suboccipital cavernous sinus and VVP to the upper cervical spine it is possible that craniocervical syndromes may cause blockage of the drainage systems of the brain. Over time this could lead to chronic NPH.

NPH is a distinct clinical entity. The symptoms of NPH, however, are identical to the classical triad seen in early stages of Alzheimer's disease; they are, abnormal gait, urinary incontinence and personality disorders. The difference between NPH and AD is in the pathology. AD is associated with characteristic, permanent pathological changes such as neurofibrillary tangles, beta amyloidosis, and amyloid placques. NPH on the other hand doesn't have these findings. Lastly, the symptoms of NPH are often reversible, whereas the symptoms of AD are not.

In addition to the condition NPH, NPH is also a clinical finding in many other important diseases such as Alzheimer's disease, Parkinson's disease, schizophrenia, manic depression, rheumatoid arthritis, Padget's disease, lupus erythematosis and others. In Alzheimer's disease, NPH is attributed to brain atrophy, that is, the brain first shrinks in size from the pathological processes associated with AD. CSF volume then increases to fill the void. This is in accordance with the Monroe-Kelli principle. But in fact it may be just the opposite. It may be that NPH is the cause, not the result of AD. So far there doesn't appear to be any connection between these diverse diseases and NPH but in the next and last of these articles, however, we will discuss a possible link.

Lastly, rather than outright blockage of the dural sinuses, NPH may be due to craniocervical hydrodynamic failure as a result of aging and degeneration of the craniocervical spine and accessory drainage system. This article looks at the possible roles of craniocervical syndromes, loss of strength in the aging dura mater, pressure conus, and failed cranial hydrodynamics in causing chronic NPH that can eventually lead to neurodegenerative diseases of the brain.

CRANIOCERVICAL SYNDROMES

The dural sinuses of the brain empty into one of two drainage systems. One is the transverse sigmoid sinus system, which empties into the internal jugular veins. This system is primarily used to drain the brain in the recumbent position. The other drainage route is the occipital marginal sinus system, which empties into the vertebral venous plexus (VVP) located inside the spinal canal and through connecting veins to the external vertebral venous plexus, which surrounds the vertebral column. This system is primarily used to drain the brain in the upright position. In addition to the cranial dural sinuses the extracranial suboccipital cavernous sinus is anatomically and physiologically an extension of both the dural sinuses and the VVP. It appears to serve as a rerouting mechanism to alternative drainage routes.

The accessory drainage system in humans uses several routes to deliver blood to the VVP. Three of the primary routes are located proximal to the occipital condyles. The suboccipital cavernous sinus on the other hand has connections with both the transverse sigmoid sinus system and occipital marginal sinus system, as well as the internal and external VVP. It can further reroute blood to several other alternative pathways such as the sinus confluens, secondary cavernous sinus, and petrosal sinus, which may be helpful during head and neck movements. Inherited or acquired craniocervical syndromes may thus interfere with the drainage system of the brain. Among other things, this could cause an increase in venous pressure in the suboccipital cavernous sinus and thus compress its contents, which include the vertebral artery and the C1 nerve root.

There are many types of craniocervical syndromes as well as gender, genetic and racial variations in design of the craniocervical spine and accessory drainage system that may predispose humans to chronic NPH. For example, certain musculoskeletal diseases such as degenerative and rheumatoid arthritis, and lupus erythymatosis have been associated with NPH. These diseases cause spondylosis that could obstruct flow in the VVP of the accessory drainage system. Spondylosis, however, can also occur as a result of injuries or simply from the effects of aging on upright posture.

Degenerative spondylosis is associated with osteophytic formations, osteoporosis, compression fractures, bulging discs, abnormal curvatures, and thickening and hardening of the ligaments and connective tissues of the spine. These conditions invade the spinal canal and could, therefore, also obstruct flow through the VVP. On the other hand, non-invasive conditions such as kyphosis, lordosis and scoliosis have been shown to cause functional stenosis and obstruction to blood flow in the VVP. In addition, there are congenital malformations such as platybasia, basilar invagination, Klippel-Feil, and atlantoaxial dislocation syndromes that can cause obstruction to CSF and venous outflow (Arnold-Chiari syndrome). Lastly, diseases such as rheumatoid arthritis and Padget's disease cause craniocervical syndromes, which can block the outlets to the VVP and suboccipital cavernous sinus of the accessory drainage system.

Genetic, gender, and racial variations in design of the craniocervical spine and accessory drainage system also effect both the capacity and efficiency of the accessory drainage system. They further determine its susceptibility to obstruction from the effects of aging and injury of the craniocervical spine. For example, features such as the size and number of the emissary foramen and diploe, and the shape of the skull can effect the efficiency of the accessory drainage system. There are gender differences also. For example, males have larger crania, foramen, diploe and spinal canals. In addition, females tend to get osteoporosis sooner than males. These differences can likewise effect the capacity and efficiency of the accessory drainage system as well as its susceptibility to obstruction from degenerative conditions and injuries.

The different racial shapes of the crania, such as orthognathic (Asians), prothognathic (Europeans), brachycephalic and dolicocephalic, may also effect the weight distribution and the position of the brainstem over the foramen magnum. It may, therefore, determine the susceptibility of the

brainstem to slippage toward pressure conus during prolonged upright posture, which can further block the outlets to the accessory drainage system. This will be discussed further later in this article.

In addition to inherited craniocervical features, the accessory drainage system likewise shows significant variations. Some have more connections or larger connections between the cranial sinuses and the VVP. About sixty percent of the population appears to have well-developed occipital marginal sinus systems in the cranium. This gives them better drainage capacity for the VVP. Likewise, there are probably variations in the design of the suboccipital cavernous sinus system as well. These features also increase the efficiency of the VVP.

Chiropractors treat craniocervical syndromes and among other things have consistently reported good results in the treatment and prevention of migraine and other types of headaches. This is important because migraine headaches may be early signs of hydrodynamic failure, and will discussed in part four of this series of articles. In fact, chiropractors probably do more to reduce the incidence of vertebral basilar artery syndromes than they are to cause them. Lastly, both chiropractors and osteopaths have long maintained that craniocervical syndromes can affect CSF hydrodynamics. Considering the close relationship of the craniocervical spine to both the suboccipital cavernous sinus and the accessory drainage system this possibility needs further investigation.

In addition to its importance to the drainage system of the brain, the suboccipital cavernous sinus also protects the vertebral artery from compression as it passes through it on its way to the brain. This is similar to the way the cavernous sinus protects the internal carotid artery from compression. An important additional benefit, however, is that they act as heat exchangers so that warm arterial blood entering the brain is cooled by venous blood exiting the brain. This keeps the brain one to two degrees cooler than the rest of the body, which makes it more efficient for ectothermic humans even during high endurance activities in hot weather. Lastly the suboccipital cavernous sinus appears to have a barorecptor similar to the carotid and aortic bodies. It therefore, appears to play a role in maintaining ICP. This makes it similar to the rete mirabile in the giraffe, the vascular plexus that fills when the head is inverted, which I discussed in the previous article.

In summary, the functional integrity of the craniocervical spine is essential to the accessory drainage system and the regulation of intracranial pressure. Genetic, gender and racial variations of the craniocervical spine may predispose certain individuals to chronic NPH and pressure conus. Acquired craniocervical syndromes may further play a role in cranial hydrodynamic failure, chronic NPH and neurodegenerative diseases.

THE AGING DURA MATER AND HYDRODYNAMIC FAILURE

The dural sinuses form the drainage system of the brain. They are located under and between the lobes and fissures of the brain and on the floor of the cranium. The structural integrity of the dura mater is essential in maintaining an open venous drainage system. As stated above, the strength of the dural sinuses also protects the internal carotid artery and vertebral artery from compression where they enter the brain. A further consideration, however, is the ability of the superior sagittal sinus to remain open under negative pressure from upright posture. True veins would collapse. The ability to stay open under negative pressure is essential for the passive production of CSF.

The increase in pressure gradients from upright posture increases both blood and CSF flow, which likewise increases their outflow. This could be a problem during upright posture because a decrease in CSF volume would cause the brain stem to slip downward toward a pressure conus. Therefore, the increase in CSF outflow must be matched by an increase in CSF production. This is

accomplished by the passive production of CSF during upright posture, which depends on an increase in the CSF pressure gradient, which in turn depends on the ability of the superior sagittal sinus to remain open under negative pressure.

If the dural sinuses lose strength with age it could subject them to compression from the weight of the brain and collapse from negative pressure during prolonged upright posture. This would obstruct the drainage system of the brain and decrease the CSF pressure gradient during upright posture causing a decrease in the passive production of CSF. A decrease in the volume of CSF in the cisterns and subarachnoid space would cause the brainstem to sink downwards toward a pressure conus causing congestion around the outlets to the accessory drainage system.

The relative importance of SSVP to CSF flow was demonstrated in an experiment on cats. In this experiment kaolin induced hydrocephalus in control cats was compared to cats in which the container of the brain was enlarged, first by craniectomy and second by duralectomy. In each case after an initial period of adjustment, CSF hydrodynamics returned to its previous steady state. What this showed was that there was a progressive decrease in the resistance to CSF absorption with each alteration of the container. This led the researchers to conclude first, that the relative contribution of SSVP to total CSF pressure was greater with each alteration of the container, and second, for the most part SSVP determines CSF pressure in the steady state. Third, what they further concluded was that the large changes in CSF hydrodynamics had a minimal effect on CSF pressure so long as SSVP remained stable.

The above experiment demonstrated the importance of stable SSVP to intracranial hydrodynamics in reclining cats. In humans, SSVP likewise, becomes stable in different positions but actually varies from about +20 mmHg in the reclining position to about -10 mmHg in the upright position. Furthermore, craniocervical syndromes and aging dura mater may effect SSVP stability in the upright position. This is because negative SSVP in the upright position depends on two things. One is the ability of the superior sagittal sinus to stay open under negative pressure. The other is the amount of head pressure in the superior sagittal sinus.

Head pressure in the SSVP is created by a waterfall effect from upright posture which causes blood and CSF flow downhill back to the heart in the upright position. A simple model of the potential energy of the head pressure in the SSVP in the upright position, would basically be a function of the volume of fluid in the SSVP, gravity, and the height of the fall assuming there are no blockages. If we leave out volume and gravity, what we are left with is the height of the fall. Theoretically, the maximum potential height of the fall is from the top of the head down to the top of the heart. On the other hand, if you ligate the veins at the base of the cranium then the maximum fall would be from the top of the head to the base of the skull. This would represent a significant decrease the CSF pressure gradient. The question is, can craniocervical syndromes similarly obstruct the drainage system of the brain and decrease the CSF pressure gradient. If so then craniocervical syndromes could among other things decrease the passive production of CSF during prolonged upright posture.

Even if the brain doesn't slip into a full blown pressure conus it may sink enough to cause obstruction of the epidural spaces, the cisterns, and the subarachnoid spaces. This would decrease venous and CSF flow. Moreover, it would trap CSF from active production in the ventricles. During sleep cycles active production of CSF would continue to increase CSF volume resulting in an overfilling. During the day the increased volume would be offloaded during upright posture. Again, however, a lack of passive production would cause the brain to start to sink toward a pressure conus again trapping CSF in the ventricles. Over time this could lead to chronic NPH.

CONCLUSION

In summary, craniocervical hydrodynamics depend on the genetic design of the craniocervical spine and accessory drainage system. It is further be effected by both disease processes and may also be effected by acquired craniocervical syndromes. Craniocervical hydrodynamics also depend on the strength of the dura mater to protect the drainage system of the brain and the internal carotid and vertebral arteries from compression, which also allows the superior sagittal sinus to remain open under negative pressure.

The extracranial suboccipital cavernous sinus system appears to be part of the intracranial cranial dural sinuses and the accessory drainage system. Blockage of the dural sinuses has been long suspected of causing NPH but the problem is blockages have rarely been found. There is a possibility, however, that the blockages may occur outside the cranium in the craniocervical syndromes, which has not yet been investigated. Furthermore, craniocervical syndromes may cause hydrodynamic failure rather than outright blockage of the drainage systems of the brain. In either case, craniocervical syndromes may be a potential cause of chronic NPH.

In conclusion, the relationship between craniocervical syndromes (stenosis) and chronic NPH in causing AD may be similar to the relationship between stenosis of the iridocorneal angle and chronic glaucoma in causing blindness. Rather than trying to cure the blindness it is better to treat the glaucoma and prevent the blindness. Similarly, it might be possible to recognize and treat NPH before it does irreparable damage to the brain. Lastly, lifetime chiropractic care of the craniocervical spine may help to prevent or limit the degree of NPH and its associated illnesses. In the next and last section we will discuss how chronic NPH may lead to irreversible neurodegenerative diseases of the brain.

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