# THE CAUSE OF SYRINGOMYELIA AND ITS SURGICAL TREATMENT

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HYDROMYELIA is a dilatation and syringomyelia is a diverticulum of the central canal of the spinal cord. The terms are used interchangeably; the conditions frequently coexist and are clinically indistinguishable.

## The Thesis

Both hydromyelia and syringomyelia originate in embryonal life as a result of distention of the central canal by ventricular fluid, caused by atresia of the outlets of the fourth ventricle. The rational treatment is to open the obstructed foramen of Magendie.

## The Evidence

1. In embryonal life the central canal of the spinal cord is a portion of the closed neural tube. It becomes a vestigial structure after the foramens of the fourth ventricle open into the subarachnoid space. Conversely, if the foramens fail to open (atresia), the central canal of necessity remains a part of the ventricular system.

2. Early forms of hydromyelia and syringomyelia are present in infants having Dandy-Walker** or Arnold-Chiari deformities,[4-10] while severe forms are found in adults having either of these deformities.[11-15]

3. There is evidence that both Dandy-Walker and Arnold-Chiari deformities of the hindbrain are produced by embryonal hydrocephalus due to atresia of the outlets of the fourth ventricle.[3,12]

4. In all patients with hydromyelia or syringomyelia, the foramen of Magendie is occluded.[11,12] The obstruction is due to a membrane, the attachments of which indicate that it is a persisting remnant of the embryonic rhombic roof, or to a cerebellar hernia that squeezes together the cerebellar tonsils, or to both conditions coexisting.

5. In patients having hydromyelia or syringomyelia, the ventricles usually are dilated[15] and air injected into the spinal canal frequently will fail to enter them.[11,12]

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***A bulging diverticulum of the fourth ventricle described by Dandy and Blackfan[2] in 1914 and shown by Taggart and Walker[3] in 1942 to be due to congenital atresia of the foramens of Luschka and Magendie.*

6. Indigo carmine injected into the ventricles may be recovered from the syrinx.[12]

7. Fluid aspirated from the syrinx resembles the product of the choroid plexus.*[11,16]

8. In the experimental animal, inflammatory sealing off of the outlets of the fourth ventricle is followed by acute hydrocephalus and syringomyelia.[17]

9. Pantopaque or India ink injected into the dilated ventricles of the above animals passes downward into the dilated central canal of the cord.[17]

10. In the human being, distention of the central canal by cerebrospinal fluid may occur when the foramen of Magendie is obstructed by a tumor or a cyst.[11,12]

11. The symptoms of syringomyelia frequently are improved by surgically removing the posterior rim of the foramen magnum and opening the obstructed foramen of Magendie.[11,12,18,19]

## The Normal Embryology

In the embryo the neural tube is a closed cavity that constitutes the anlage of the ventricular system and of the central canal of the spinal cord. The ventricular fluid leaves the neural tube by filtering through the permeable rhombic roof whence it expands the developing subarachnoid spaces.[20] The growth of the choroid plexus during the sixth to the eighth week is accompanied by a dilution of the protein content of the ventricular fluid apparently due to an increase in its rate of formation. There is an associated rapid expansion of the subarachnoid spaces and of the lumen of the neural tube during this stage.

At about the fifth month of fetal life, as the result of progressive attenuation of portions of the rhombic roof, the foramens of the fourth ventricle open. Through these openings, the pulsations transmitted to the ventricular fluid by the choroid plexus[21] are diverted from the central canal to the outer surface of the neural tube. The central canal, devoid of choroid plexus, narrows to become a vestigial structure after the development of this shunt.

## The Pathologic Embryology

During the sixth to the eighth week of embryonal life, should the membranous rhombic roof not be sufficiently permeable to permit free escape of ventricular fluid (atresia), the pressure within the obstructed ventricles will rise correspondingly, causing a pressure cone of the soft embryonal hindbrain known as the *Arnold-Chiari malformation*.[12] More rarely this embryonal hydrocephalus will cause a bulging diverticulum of the hindbrain,[3] referred to

**If the fluid aspirated from a cystic cord is yellow and has a high protein content, the responsible lesion is a tumor or hamartoma and not a true hydromeylia or syringomyelia.*

by Benda[1] as the *Dandy-Walker syndrome*. In either case the confined pulsations of the ventricular fluid continue to be funneled toward the central canal and cause it gradually to dilate. Hydromyelia, therefore, has its onset in embryonal life, and though clinically silent in congenital hydrocephalus, it nevertheless constitutes a part of the pathologic picture. Hydromyelia often has been described[4-10] in association with the Arnold-Chiari deformity. Benda,[1] as well as Gardner, Abdullah, and McCormack,[12] has described it in association with the Dandy-Walker deformity.

## The Hydrodynamics of Syringomyelia

If the intraluminal pressure ruptures the ependymal lining of the central canal, a diverticulum (syrinx) may form parallel to the canal. In an article entitled "Cervical Syringomyelia and Syringomyelia-Like States Associated with Arnold-Chiari Deformity and Platybasia," this mechanism was described by Lichtenstein[13] as follows:

> The constriction of the neuraxis by the bony ring prevents adequate communication between the caudal diverticulum of the fourth ventricle and the posterior cistern. The accumulating cerebrospinal fluid dilates the central canal and, in some instances, is associated with perforation of the ependymal lining and the spread of the fluid into the posterior columns.

The dilatation of the central canal which results in symptomatic syringomyelia obviously is a slow process and the disease seldom is diagnosed until adult life. The late onset of symptoms in patients having syringomyelia has led many investigators to consider it an acquired disease, although 80 years ago Leyden[22] pointed out that syringomyelia in the adult is a "rest" of a congenital hydromyelia that "cuts itself off" from the central canal.

The local manifestation of atresia in the adult is a membranous remnant of the embryonic rhombic roof which occludes the foramen of Magendie and most commonly produces a mild Arnold-Chiari malformation or a cerebellar hernia.[12] Under the steady pounding of the pulse wave of the ventricular fluid, the occluding membrane eventually may perforate; but since meanwhile the surrounding structures have become impacted in the foramen magnum, obstruction of the foramen of Magendie persists. From then on, regardless of whether the foramens of Luschka open, there will remain a partial block between the cranial and spinal subarachnoid spaces. The block is more complete during systole as the impaction is rendered more pronounced by the accompanying downward excursion of the cerebellar tonsils into the funnel-shaped foramen magnum. In response to the friction of the pulsating structures incarcerated in the foramen magnum, a fibrous thickening of the meninges may develop at this level (pachymeningitis cervicalis).

Although in patients having syringomyelia there is no increase in intracranial pressure, there frequently is ventricular dilatation.[15] This dilatation represents hydrocephalus that became compensated during prenatal or post-

natal life, thus permitting the patient to survive to adulthood. The mild form of embryonal atresia that results in mild Arnold-Chiari malformation and eventuates in the clinical picture of syringomyelia in the adult, seldom produces recognizable symptoms or signs early in life.

## The Living Pathology

Although the upper portion of the central canal connecting the fourth ventricle with the hydromyelic sac is patent, it seldom is involved in the dilatation because of the resistance offered by the interlaced decussating tracts in the medulla as well as by the impaction of the herniated structures in the foramen magnum. This absence of dilatation of the upper portion of the central canal, together with the damage to this area which occurs during the standard postmortem removal of the brain, has caused investigators to overlook the connection that exists between the fourth ventricle and the syrinx.

The force of the pulse wave of the ventricular fluid (shown by Bering[21] to consist of a steeply rising gradient and a gradual fall) causes the narrow upper portion of the central canal to act like a one-way valve, so that part of the fluid that enters it during systole remains trapped below during diastole. This hydrodynamic mechanism is aided by the fact that the spinal portion of the dural sac is distensible by virtue of its compressible epidural venous plexus, whereas, the cranial portion is not. When the obstruction of the foramen of Magendie is released at operation, the pulse wave of the ventricular fluid, previously funneled toward the central canal, is shunted into the subarachnoid space on the outer surface of the cord as nature intended. There its force tends to collapse the dilated central canal. This hydrostatic mechanism, so clearly apparent during operation on the living patient, can only be suspected on the basis of the findings at necropsy.

### References

1. Benda, C. E.: Developmental Disorders of Mentation and Cerebral Palsies. New York: Grune and Stratton, 1952, 565 pp.

2. Dandy, W. E., and Blackfan, K. D.: Internal hydrocephalus, experimental, clinical and pathological study. Am. J. Dis. Child. **8**: 406-482, 1914.

3. Taggart, J. K., Jr., and Walker, A. E.: Congenital atresia of foramens of Luschka and Magendie. Arch Neurol. & Psychiat. **48**: 583-612, Oct. 1942.

4. Ingraham, F. D., and Scott, H. W., Jr.: Spina bifida and cranium bifidum; Arnold-Chiari malformation; study of 20 cases. New England J. Med. **229**: 108-114, July 15, 1943.

5. Chiari, H.: Ueber Veränderungen des Kleinhirns, des Pons und der Medulla oblongata in Folge von congenitaler Hydrocephalie des Grosshirns. Deutsche med. Wchnschr. **27**: 1172-1175, 1891.

6. Cameron, A. H.: Arnold-Chiari and other neuro-anatomical malformations associated with spina bifida. J. Path. & Bact. **73**: 195-211, Jan. 1957.

7. Chiari, H.: Ueber Veränderungen des Kleinhirns, des Pons und der Medulla oblongata in Folge von congenitaler Hydrocephalie des Grosshirns. Denkschr. d. k. Akad. d. Wissensch. Math-naturw. Kl. **63**: 71, 1895.

8. Arnold, J.: Myelocyste, Transposition von Gewebskeimen und Sympodie. Beitr. path. Anat. **16**: 1-28, 1894.

9. Schwalbe, E., and Gredig, M.: Über Entwicklungsstörungen des Kleinhirns, Hirnstamms und Halsmarks bei Spina bifida. Beitr. path. Anat. **40**: 133-194, 1907.

10. Russell, D. S.: Observations on Pathology of Hydrocephalus, Medical Research Council, Special Report Series No. 265. London: His Majesty's Stationery Office, 1949, p. 19 (138 pp.).

11. Gardner, W. J., and Goodall, R. J.: Surgical treatment of Arnold-Chiari malformation in adults. Explanation of its mechanism and importance of encephalography in diagnosis. J. Neurosurg. **7**: 199-206, May 1950.

12. Gardner, W. J.; Abdullah, A. F., and McCormack, L. J.: Varying expressions of embryonal atresia of fourth ventricle in adults. Arnold-Chiari malformation, Dandy-Walker syndrome, "arachnoid" cyst of cerebellum, and syringomyelia. Presented at the meeting of the Harvey Cushing Society, Detroit, Michigan, April 27, 1957; J. Neurosurg.: **14**: 591-607, Nov. 1957.

13. Lichtenstein, B. W.: Cervical syringomyelia and syringomyelia-like states associated with Arnold-Chiari deformity and platybasia. Arch. Neurol. & Psychiat. **49**: 881-894, June 1943.

14. Ogryzlo, M. A.: Arnold-Chiari malformation. Arch. Neurol. & Psychiat. **48**: 30-46, July 1942.

15. Netsky, M. G.: Syringomyelia. A.M.A. Arch. Neurol. & Psychiat. **70**: 741-777, Dec. 1953.

16. Wetzel, N., and Davis, L.: Surgical treatment of syringomyelia. A.M.A. Arch. Surg. **68**: 570-573, April 1954.

17. McLaurin, R. L.; Bailey, O. T.; Schurr, P. H., and Ingraham, F. D.: Myelomalacia and multiple cavitations of spinal cord secondary to adhesive arachnoiditis; experimental study. A.M.A. Arch. Path. **57**: 138-146, Feb. 1954.

18. Chamberlain, W. E.: Basilar impression (platybasia); bizarre developmental anomaly of occipital bone and upper cervical spine with striking and misleading neurologic manifestations. Yale J. Biol. & Med. **11**: 487-496, May 1939.

19. Gustafson, W. A., and Oldberg, E.: Neurologic significance of platybasia. Arch. Neurol. & Psychiat. **44**: 1184-1198, Dec. 1940.

20. Weed, L. H.: Development of Cerebro-Spinal Spaces in Pig and in Man. Washington: Carnegie Inst., 1917, 116 pp., 4°.

21. Bering, E. A., Jr.: Choroid plexus and arterial pulsation of cerebro-spinal fluid: demonstration of choroid plexuses as cerebrospinal fluid pump. A.M.A. Arch. Neurol. & Psychiat. **73**: 165-172, Feb. 1955.

22. Leyden, E.: Ueber Hydromyelus und Syringomyelie. Arch. path. Anat. **68**: 1-26, 1876.