EMBRYONAL ATRESIA OF THE FOURTH VENTRICLE
THE CAUSE OF "ARACHNOID CYST" OF THE CEREBELLOPONTINE ANGLE

W. JAMES GARDNER, M.D., LAWRENCE J. MCCORMACK, M.D.,
AND DONALD F. DOHN, M.D.
Departments of Neurological Surgery and Anatomic Pathology,
The Cleveland Clinic Foundation, and The Frank E. Bunts
Educational Institute, Cleveland, Ohio
(Received for publication August 28, 1958)

In the description of many acquired diseases the adjectives "early" and “mild” are as synonymous on the one hand as are “late” and “severe” on the other. The opposite is true, however, of congenital lesions such as the Arnold-Chiari and the Dandy-Walker malformations, inasmuch as in these conditions the mild cases are encountered late in life while the severe instances are obvious early. Cases illustrating this were first described by Chiari for the condition which bears his name, and by Dandy and also by Walker for that which now bears theirs.

It is a medical axiom, that, to acquire a correct concept of a disease process one should begin with the study of mild cases and then progress to the more severe and complicated forms. A previous study of mild (i.e., adult) cases of the Dandy-Walker, and the Arnold-Chiari malformations, showed that each is caused by atresia of the outlets of the fourth ventricle. The embryonic forerunner of such atresia, we believe, is inadequate permeability of the rhombic roof. The rising pressure within the neural tube then results in dilatation of the occluded foramen if the hindbrain is balloonsined in the Dandy-Walker malformation, and in narrowing of the foramens if it is compressed by the downward displacement of the tentorium in the Arnold-Chiari malformation described recently by Cameron. In the foregoing communication it was shown that embryonal atresia also may express itself as a loculated cyst occluding the foram of Magendie, or as a band of thickened meninges encircling and squeezing the cerebellar tonsils against the medulla. These four hindbrain expressions of atresia occur in adults in varying degrees and combinations and frequently are accompanied by congenital scoliosis, basilar impression and syringomyelia. In this presentation, cases will be described showing that persisting portions of the embryonic rhombic roof at the foramen of Luschka may behave in the same manner as those at the foramen of Magendie; namely, they may stretch to form a diverticulum that communicates with the fourth ventricle or their two layers may separate to form a loculated cyst.

An appreciation of the behavior of permeable membranes, together with a review of the embryology of the cerebrospinal fluid spaces is essential to an understanding of these conditions.
EMBRYONAL ATRESIA OF FOURTH VENTRICLE

PROPERTIES OF MEMBRANES

The essential difference between a permeable membrane and a perforated membrane is simply a matter of the size of the holes. To add the prefix "semi" to "permeable" is redundant, since any hole will hold back an object that is larger than itself. The holes of a perforated membrane are of visible or particulate size, while in a permeable membrane they are of submicroscopic or molecular size. The holes in a permeable membrane, although too small to be seen with a microscope, may be relatively speaking so large and closely spaced that water, and even protein molecules pass through almost as freely as though the holes were of visible size. Such a membrane may enclose an opening as a screen encloses a window and not obstruct it to any significant degree. When a membrane is stretched, its holes enlarge and may coalesce. Because of the shape of the hole or its electrical charge, particles or molecules may pass through more freely in one direction than in the other, thus constituting a valve effect.

If protein-containing fluid passes through a membrane, the pores of which filter out some of the large protein molecules, these molecules will pile up against the proximal surface of the membrane, like tumbleweed against a wire fence, causing it to thicken and thus retard the passage of smaller molecules that would readily pass through the membrane in its naked state. This fact was demonstrated by Butt and Keys for collodion membranes, by Chambers and Zweifach for the endothelial lining of living capillaries, and by Gardner et al. for the absorbing areas in the subarachnoid space.

Since permeability of cell membranes is essential to cell nutrition, it follows that all living membranes are permeable to some degree. This is true of the walls of the ventricles as was demonstrated by Bering’s studies on hydrocephalus in which he used heavy water as a tracer. Embryonal membranes enclosing the outlets of the fourth ventricle may be so permeable, and, as a result, the flow of fluid through them so unimpeded that pressure within the ventricles is virtually the same as in the subarachnoid space. However, should the enclosing membrane be less permeable so that a significant head of pressure is necessary for the rate of filtration to approach the rate of elaboration of the fluid, then one of three things may happen. If the membrane is sufficiently fragile, it will rupture. If it is sufficiently yielding, it will respond by stretching until, by a process of attenuation plus increased surface area, the rate of filtration increases to equal the rate of elaboration. If the membrane is unyielding, some yielding portion of the neural tube will stretch and attenuate until equilibrium is established by the increasing permeability of this attenuating area. Yielding, of course, will occur more readily in embryonic than in older tissues. Since the persisting membranes that constitute embryonal atresia of the fourth ventricle consist of two layers of differing physical characteristics, the pulsations of the ventricular fluid may drive fluid through the proximal layer at a rate faster than it can escape through the distal layer. This pumping effect will cause