Atresia of the Foramina of Luschka and Magendie: The Dandy-Walker Cyst

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As of 1942, when Taggart and Walker reported three patients with atresia of the foramina of Luschka and Magendie,24 only six previous cases had appeared in the literature,5,4,7,8,17,22,26 and only two of these cases had undergone surgical intervention. In the majority of cases published since then, the diagnosis was made either at operation or at postmortem examination.

Treatment consists of cyst resection after posterior fossa exploration, or shunting from the lateral or third ventricle.5,7,10,15,27 In 1956, Matson outlined the plain skull film, ventriculographic, and dural sinographic characteristics of prenatal obstruction of the fourth ventricle and emphasized that "intelligent and successful treatment of hydrocephalus depends upon an accurate knowledge of the location and type of obstruction to normal cerebrospinal fluid movement."16

Both the embryologic background and the specific pathological changes associated with this particular clinical entity are controversial. Taggart and Walker maintained that failure in development of the foramina of Luschka and Magendie, in the fourth month of fetal life, produced hydrocephalic enlargement of the fourth ventricle.24 This, in turn, precluded the development of the inferior vermis and prevented the normal descent of the torcular Herophilus and lateral sinuses.5,12 Brodal and Hauglie-Hanssen pointed out that the cerebellar anlagen fuse long before the fourth fetal month, and that thus the cerebellar agenesis could not be the result of the later foraminal atresia.2 They concluded that the entire process was precipitated by a previously existing hydrocephalus of unknown cause. The description of cerebellar embryological development, as outlined by Dow and Moruzzi, supports this concept.9

Benda suggested that atresia of the developing fourth ventricular foramina was only a part of the syndrome and not its cause, because in some cases reported, the foramina were found to be patent.1 He stated that the main pathological process was the "meningomyelocele" sac-like dilatation in place of the posterior medullary velum which results in a cleft cerebellum and hydrocephalus. Gibson, through careful examination of the cystic membranes, concluded that there is no true cerebellar agenesis or aplasia and, therefore, that the whole process may well be secondary to persistent closure of the foramina of Luschka and Magendie.11 D'Agostino, et al., in a review of their pathological material, came to the same conclusion.6

Gardner included the Dandy-Walker cyst among many dysgeneses, all the result of persistent fetal hydrocephalus secondary to impeded flow of cerebrospinal fluid through the attenuated roof of the fourth ventricle.10 The foraminal atresia mentioned by others, according to him, was therefore merely a more remarkable example of the impermeability of the rhombic roof.

For the purposes of this paper we shall avoid this controversy and consider the basic clinical problem to be the presence of a posterior fossa cyst which is entirely within the confines of the fourth ventricle, and which is caused by complete occlusion of the foramina of Luschka and Magendie. We shall refer to this clinical entity as the Dandy-Walker cyst.

Between the years 1963 and 1968, of 212 hydrocephalic infants diagnosed at Cook County Hospital, eight were found to have

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Dandy-Walker cysts. This group constitutes our clinical series, together with one case of arachnoidal cyst of the cisterna magna which we have added for comparison. We shall describe the salient clinical, neuroradiological, and anatomico-pathological features of these cases, and emphasize their correlation.

Case Reports

Case 1. A diagnosis of Dandy-Walker cyst was made on this white baby girl at 2½ months of age. Plain skull films revealed that the squamous occipital bone was bulging and that the lambdoidal sutures were maximally split. Ventriculography revealed symmetrical hydrocephalus with a massive fourth ventricle, and free communication between the third and fourth ventricles. The aqueduct was enlarged. A right ventriculoadal shunt was performed; the lateral ventricle pressure was 220 mm H2O.

At 16 months of age she began to develop posterior fossa signs: bilateral nystagmus on lateral gaze, explosive speech, and truncal ataxia. At 18 months she had a broad-based, anteropulsive gait. Revision of the shunt at 19 months did not significantly alter the progression of posterior fossa symptomatology.

At 4 years of age, because of the progressive development of posterior fossa signs despite a well-functioning shunt system, the child underwent another complete neuroradiologic evaluation. These studies demonstrated moderately enlarged lateral ventricles, but the fourth ventricle was now enormous. Air, Conray 60,19 and Pantopaque* would not pass from the third into the fourth ventricle or vice versa. Since contrast studies performed at birth had revealed free passage of air between the third and fourth ventricles, it was concluded that she had developed a secondary aqueductal occlusion. Therefore, a fourth ventricle peritoneal shunt was inserted; pressure in the fourth ventricle at this time was 150 mm H2O, and the fluid was xanthochromic. The lateral ventricles contained clear fluid at a pressure of 130 mm H2O.

Subsequently, the child made a dramatic recovery. She became stable on her feet, able to walk without anteropulsion, and steady enough with both hands to feed herself without spilling liquids. Her speech became understandable. Subsequent contrast studies demonstrated that air passed freely from the third ventricle through the patent aqueduct into the encysted fourth ventricle.

Case 2. This Negro girl was admitted at the age of 2 months with a history of progressive enlargement of the head. Examination revealed split sutures, bulging fontanelles, and a prominent occipital bone. Cerebral angiography (Figs. 1–4) demonstrated changes characteristic of a Dandy-Walker cyst and of agenesis of the corpus callosum. Lateral ventriculography, performed after a lateral ventricle peritoneal shunt, confirmed these diagnoses. The pressure in the lateral ventricle was 210 mm H2O, and the fluid was clear. No fourth ventricle recording was made.

Nine months later, another shunt revision was performed. The lateral ventricle pressure at this time was 160 mm H2O. The child has done well during the past 8 months.

Case 3. This white girl was born with an occipital encephalocele. Her parents refused surgery until she was 6 months old when a ventriculoadal shunt and closure were accomplished without antecedent angiography or pneumoencephalography. Over the following 3 months, her head continued to get larger.

She was admitted to Cook County Hospital when she was 9 months old because of progressive enlargement of her head. She was found to have bilateral optic atrophy, searching nystagmus, the “setting sun” phenomenon, and a markedly enlarged head. The squamous occipital bone was bulging, and the lambdoidal sutures were widely split. Angiographic and pneumographic diagnosis of a Dandy-Walker cyst was made. The lateral ventriculoadal shunt was replaced by a ventriculoperitoneal shunt. The fluid in the lateral ventricles was clear, and the pressure was 270 mm H2O.

Postoperatively, she developed persistent vomiting in spite of a sunken anterior fontanel. We then removed the lateral ventricle

* Generic and trade names of contrast media: Conray 60 = Methylglucamine lothalamate 60%; Hypaque 50% = Sodium diatrizoate 50%; and Pantopaque, Myodil = Ethyl iodophenylundicylate.