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Cystic malformations of the posterior cranial fossa, current state of classification, CT and MR appearance

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Summary

The cerebellum has the longest development in comparison with other large structures of the brain, and this may be the cause of many developmental abnormalities. [1,2]. They are divided into two groups: cystic malformations and disturbances of cerebellar histiogenesis process. The paper focuses on the former of the two large groups mentioned above [3,4,5]. Cystic malformations leading to hypoplasia of the vermis and cerebellar hemispheres often cause enlargement of the posterior cranial fossa.

The aim of this study is to summarize and systematize the contemporary knowledge of cystic lesions of the posterior fossa, especially their imaging.

Imaging of posterior fossa malformations is practically based on computed tomography (CT) and magnetic resonance imaging (MRI). On CT studies, the posterior fossa is imaged with 5mm or 2-3 mm slices to gain more precise visualization. However, CT is often followed by MRI, mainly in T1-weighted, sagittal and axial images, which allow for the best evaluation of the posterior fossa structures. 2D-FISP imaging technique may be also useful.

Thorough analysis of the literature allowed to put forward the following conclusions: classification of these cystic malformations is frequently changed; it is necessary to evaluate disorders both in CT and MRI; one should use the descriptive term – retrocerebellar cyst, if an image is not clear-cut; the radiologist's goals include finding a lesion, evaluation of its topography and effect on the brain, but not necessarily establishing its particular type.

Key words: Cystic malformations • posterior fossa • CT • MRI

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Background

The human cerebellum develops since the early embryonic period up to the second year of life. This prolonged development, the longest among all the large structures of the brain, makes the cerebellum susceptible to a wide range of developmental abnormalities [1, 2].

Developmental malformations of the cerebellum are usually divided according to the time of their onset and radiological presentation into two large groups [3, 4, 5]. The first group is represented by cystic malformations, which are the topic of this study. They are caused by anomaly in the process of ventral induction, which occurs between the 5th and 10th week of embryonic life [3, 4, 5]. The second group includes malformations caused by disturbances of cerebellar histiogenesis process (beginning in the third month of embryonic life and lasting till the end of infancy), with cerebellar hypoplasia being one of the most common. In contrast to cystic malformations, they do not cause expansion of the posterior cranial fossa [5].

Cystic malformations lead to hypoplasia of the vermis and cerebellar hemispheres and formation of cyst-like structures

resembling cyst within the posterior fossa. They are typically accompanied by enlargement of the posterior fossa and high position of the tentorium [3, 4, 5].

Embryonic background of developmental malformations of the cerebellum

The contemporary model of developmental malformations of the posterior cranial fossa is based mostly on morphogenetic analysis of development of posterior fossa brain structures. The stage essential for formation of cystic malformations during cerebellar development is ventral induction, which starts at the beginning of the 5th week of embryonic life [3, 5]. At this time, the cerebral vesicle – rhombencephalon is the largest, and the floor of primordial fourth ventricle begins to resemble a rhomboid. The upper apex of the primordial rhomboid fossa is directed to the mesencephalon, whereas the lower to the myelencephalon. In the 6th week of embryonic life, the primordial meninx invaginates into primordial fourth ventricle, thus creating a primordium of future ventricular choroidal plexus – the choroidal fold. The choroidal fold runs across the roof of the rhomboid fossa and divides it into two triangle areas, with the same basis – cranially located area membranacea superior (AMS) and caudally located area membranacea inferior (AMI) [5]. In this period, primordia of the cerebellum are also created. The further development leads to formation of the vermis from medial-superior part of the cerebellum primordium, and the cerebellar hemispheres from its lateral parts, also called winged laminae. Neuroblast proliferation of the winged laminae leads to formation of so-called rhomboid lips, which grow very intensively. At the later stage, fusion of the rhomboid lips along the medial line and area membranacea superior occurs. The fusion progresses gradually up to the 15th week of embryonic life, and when this process ends the cerebellar vermis is completely formed. As the result of this process, the AMS is incorporated into the basis of developing cerebellum, and then, by the end of the 8th week of embryonic life, it disappears [3, 5].

In case of a developmental anomaly of the lateral parts of cerebellar primordium – (winged laminae), the AMS is not incorporated into the cerebellar basis and bulges posteriorly between rostrally displaced cerebellar vermis and caudally displaced choroidal plexus, whereas the cerebellar hemispheres are dislocated laterally and ventrally. The markedly widened cyst-like fourth ventricle causes elevation of the tentorium and enlargement of the posterior fossa. All that leads to complete Dandy-Walker syndrome [3, 5].

The area membranacea inferior (AMI) is not directly involved in the process of cerebellar formation. When AMI bulges and expands dorsally under the developing cerebellum, it forms an embryonic recess of the fourth ventricle, called also Blake's pouch. In normal conditions, Blake's pouch regresses by the end of 8th week of embryonic life, forming the inferior medullary membrane and outflow foramina of the fourth ventricle – Magendie's foramen precursor and laterally located two Luschka's foramina [3].

As a result of developmental anomaly of AMI and consequently lack of primary Magendie's foramen, Blake's pouch persists and expands leading to cystic malformation of the posterior fossa named persistent Blake's pouch or Blake's pouch cyst (BPC). BPC causes elevation of the tentorium and widely communicates with the fourth ventricle. It is separated from the subarachnoid space and is always accompanied by hydrocephalus. The cerebellum is not morphogenetically associated with the AMI, therefore in cases of BPC there is no true dysplasia of the cerebellum [3, 6, 7, 8, 9, 10, 11].

As the result of AMI developmental anomaly and late perforation of Magendie's foramen, a large cerebellomedullary cistern (mega cisterna magna) is formed. It is considered a developmental variant. It always communicates with the fourth ventricle, as well as with the subarachnoid space, and it is not accompanied by hydrocephalus [3, 4, 12].

Abnormalities of the further stage of cerebellum development – histiogenesis – do not cause expansion of the posterior cranial fossa; on the contrary, they are often accompanied by the small posterior fossa size as well as low position of the tentorium and confluence of the sinuses. The most common among these malformations is hypoplasia of the vermis and/or cerebellar hemispheres. The abnormalities of histogenesis are beyond the scope of this paper [3].

Imaging methods used in diagnostics of the posterior fossa cystic malformations.

Diagnostics of developmental malformations of the posterior fossa is generally based on computed tomography (CT) and magnetic resonance imaging (MRI), and to much lesser extent on sonography (which is beyond the scope of this study).

In abnormalities of organogenesis, the aim of neuroimaging studies is mainly precise visualization of malformation topography. In CT of the posterior fossa, 5 mm thick slices are usually used, but in order to gain a better image 2-3 mm thick slices may be applied [13, 14]. CT has a limited value in the evaluation of posterior fossa malformations due to the bone artifacts and possibility of obtaining directly only axial views [15]. Hence, CT may be treated only as a preliminary study, followed by MRI. In MRI, structural anomalies of the posterior fossa are best represented on sagittal and coronal T1-weighted images. Axial views are also important, particularly if there is need to compare MRI with previously performed CT. T2-weighted, proton density (PD) and FLAIR sequence images have higher value in search for accompanying histiogenesis, cell migration and myelinization disturbances. Paramagnetic contrast enhancement has little importance in diagnostics of these malformations, with the exception of lesions with associated vascular malformations [1, 15]. Techniques of imaging of cerebrospinal fluid flow such as 2D-FISP sequence (cine option) have recently begun to play an important role in diagnostics of posterior fossa cystic malformations. They show pulsatile real-time flow of the cerebrospinal fluid in the intracranial fluid spaces and spinal canal. This imaging method facilitates differentiation of enlarged posterior fossa fluid spaces caused by cerebellar hypoplasia and atrophy from cystic malformations. It also allows for assessment of communication between the posterior fossa cyst and the fourth ventricle and paracerebellar

fluid spaces. It is considered an alternative for CT-cisternography, and may be of key importance for making decisions concerning treatment [16, 17].

Classification of posterior fossa cystic malformations.

Cystic developmental malformations of the posterior fossa have been debated since the first publication by Dandy in 1914 and introduction of the "Dandy-Walker malformation" term in 1954 to describe coexistence of various degree hydrocephalus, big cisterna magna and hypoplasia of the cerebellar vermis [3]. Different definitions of this malformation have been proposed since that time; however, the following three elements were the most common: cystic enlargement of the fourth ventricle, dysgenesis of the cerebellar vermis and high tentorium position [1, 3, 4, 15, 18, 19]. Later on, definitions of a new associated disease entity have been introduced: Dandy-Walker variants and mega cisterna magna. Modifications of Dandy-Walker and mega cisterna magna definitions have been proposed, as well as abandoning of this terms in favor of vermian-cerebellar hypoplasia [1, 3, 20, 21]. Nowadays the term: "Dandy-Walker complex" is commonly used [21]. The following forms of Dandy-Walker complex, based on axial CT scans are distinguished:
– „classic" Dandy-Walker syndrome with enlarged posterior fossa, agenesis or hypoplasia of the vermis, high position of the tentorium, hydrocephalus,
– Dandy-Walker syndrome variant, which features various grades of hypoplasia of the cerebellar vermis with or without extension of the posterior fossa;
– mega cisterna magna – enlarged cisterna magna with preserved, normal cerebellar vermis and normal fourth ventricle [15, 20, 21].

With the advent of MRI in routine diagnostics of the central nervous system, the classification described above became unsatisfactory and incomplete, because axial CT does not offer complete and reliable evaluation of the cerebellar vermis [15]. Besides, recent studies have proven that hydrocephalus considered previously as a component of Dandy-Walker syndrome, is absent in the majority of patients in perinatal period [1, 15].

Modern classification of cystic malformations of the posterior fossa should include:
– Dandy-Walker syndrome and a variant, originated from abnormalities of development of the area membranacea superior (AMS),
– mega cisterna magna, originated from abnormality of regression of the area membranacea inferior (AMI),
– persistent Blake's pouch, a new nosologic entity introduced by Tortori-Donati, which results from AMI regression abnormality, but has a different clinical course and different MRI appearance,
– posterior fossa arachnoid cyst [3, 6, 7, 10, 22, 23, 24].

Additionally, vermian-cerebellar hypoplasia (VCH) should be mentioned. Although cerebellar hypoplasia is not classified as a cystic malformation, it may also cause secondary enlargement of posterior fossa fluid spaces. They may mimic posterior fossa cysts [1, 20, 23] and could be misdiagnosed as Dandy-Walker syndrome [1, 15]. The feature

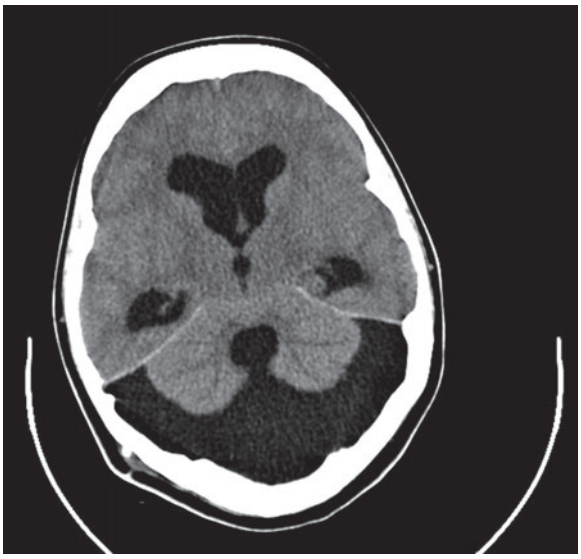

Figure 1. Dandy-Walker malformation (DWM). Axial CT scan through the posterior fossa reveals marked hypoplasia (absence) of the vermis and cystic transformation and extension of the fourth ventricle, which bulges posteriorly and fills almost the entire cranial posterior fossa which is significantly enlarged.

that allows distinguishing both malformations is the size of the posterior fossa – in case of VCH the posterior fossa is small [1, 4, 23].

The greatest diagnostic difficulties are caused by cerebellar histiogenesis malformation – rhombencephaloschizis. If accompanied by characteristic clinical signs (abnormal eye movement, ataxia, intellectual retardation, recurrent respiratory disturbances) rhombencephaloschizis is called Joubert malformation. The fusion of both parts of the vermis is incomplete, which causes a formation of the midline cleft. Cerebellar hemispheres have normal size and are separated by a narrow fissure. Joubert malformation has been often erroneously described as a Dandy-Walker variant [3, 15, 25, 26].

It should be emphasized, that all the aforementioned entities may partly overlap both in terms of clinical and radiological appearance, and possibilities of differentiation between them only by means of neuroradiological techniques are limited [8].

Dandy-Walker malformation (DWM)

The incidence of DWM is estimated to be 1 of 25000-3000 births [3, 4]. It constitutes 14% of cystic malformations of the posterior fossa [4], and it is more common in girls [27]. "Classic" or "true" Dandy-Walker malformation (Fig. 1) consists of the triad of symptoms:
1. dysplasia or hypoplasia of the vermis (lower part) (and the cerebellar hemispheres),
2. cystic transformation and extension of the fourth ventricle, which bulges posteriorly and fills almost the entire posterior cranial fossa,
3. posterior fossa enlargement with upward displacement of the dura mater sinuses and cerebellar tentorium [1, 3, 4, 13, 19, 28] (Fig. 2 A, B).

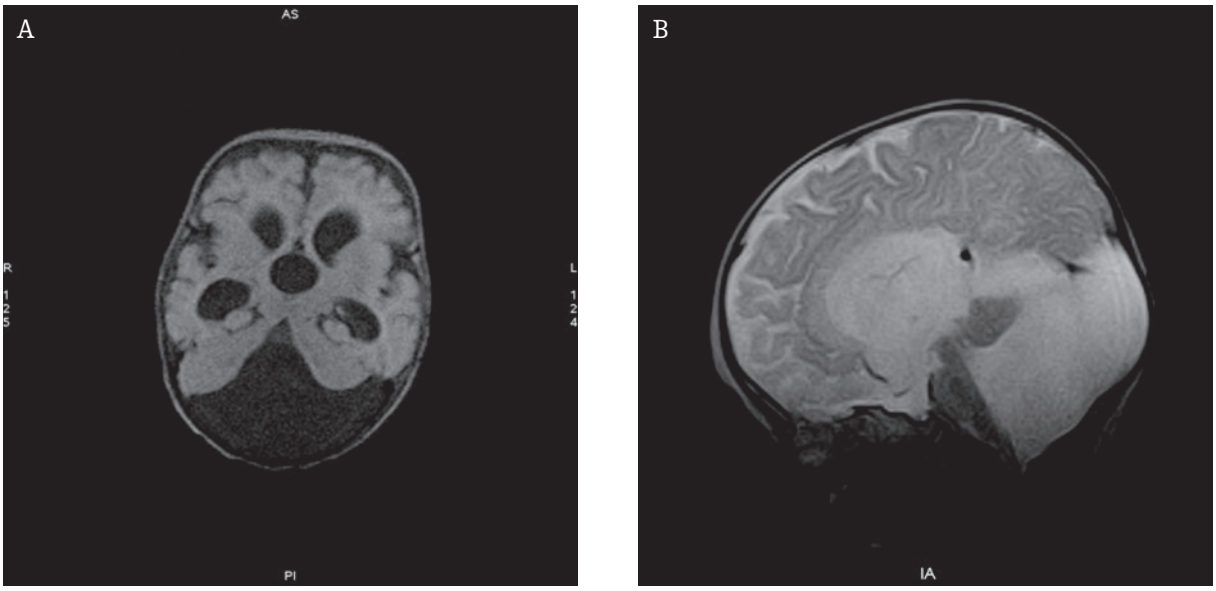

Figure 2. Dandy-Walker malformation (DWM). Axial MRI FLAIR (A) and saggital T2-weighted (D) images show the classic triad of symptoms: marked hypoplasia of the vermis, cystic transformation and extension of the fourth ventricle (which bulges posteriorly, displaces and elevates the cerebellar vermis and fills almost the entire posterior fossa of the cranium) as well as posterior fossa enlargement with accompanying upward displacement of the dura mater sinuses and cerebellar tentorium.

The definition should not include hydrocephalus, as the majority of patients (over 80%) in pre- and perinatal period have a normal ventricular system. Hydrocephalus most frequently develops during the first three months of life, as a complication of the posterior fossa cyst [19, 29, 30]. The degree and location of the impairment of cerebrospinal fluid outflow may differ significantly [4].

Patency of outflow foramina of the fourth ventricle in DWM is assessed to be preserved in 7-39% [3]. Communication between enlarged cystic fourth ventricle and the pericerebellar cisterns is preserved in over 80% of cases [29]. It has also been proven, that in the majority of patients with DWM dimensions of the ventricular system at birth are normal. These facts cast doubts on the theory of fourth ventricle outflow obstruction as a cause of DWM [3, 29, 30].

Apparent changes in imaging examinations in patients with DWM involve the skull, dura mater and sinuses, fourth ventricle, brainstem, cerebellum and supratentorial structures. Both X-rays and CT reveal cranial deformities and enlargement of the skull (dolichocephaly), thin and prominent occipital squame, widened lambdoid suture and deformities of the petrous bone. In addition, in some cases occipital squame cleft and encephalomeningocele may be visualized [3, 19, 31].

The tentorium and the confluence of the sinuses are displaced upward, above the lambdoid suture (lambdoid-torcular inversion) [1, 19]. Sagittal MRI images allow to visualize significant widening of the angle between the superior sagittal sinus and the straight sinus, which may reach 90°-150° (normal range 50°-75°). Both MRI and CT demonstrate widened tentorial incisure and absence of the cerebellar falx [3, 4].

Axial views of CT and MRI present significant cystic enlargement of the fourth ventricle, which bulges posteriorly and fills the posterior fossa. The wall of the cyst adheres anteriorly to residual, dysplastic cerebellar vermis, laterally to the hemispheres and inferiorly to the medulla oblongata. Besides, MRI demonstrates lack of fourth ventricle choroid plexus [10], less commonly it is present, but dislocated to the lateral recess or inferior wall of the fourth ventricle [3].

Sagittal MRI sections reveal various degrees of vermian dysplasia, in 25% of cases complete aplasia, whereas in 75% hypoplasia or aplasia of the lower vermis [3, 4, 23].

Both CT and MRI present hypoplastic cerebellar hemispheres, often asymmetric, displaced anteriorly and laterally. Shunt implantation into the cyst leads to medial displacement of the hemispheres, so that they lie close to each other, just under the hypoplastic vermis [3, 23]. It may result in false diagnosis [31].

MRI visualizes much better than CT accompanying changes within posterior fossa structures. The brainstem is usually hypoplastic [4] and heterotopic foci in the cerebellar cortex as well as various dysplasias of the hindbrain nuclei may be seen on T2-weighted and FLAIR images [19].

Supratentorial structures anomalies accompany classic DWM in 50-70% of cases: corpus callosum dysgenesis in 20-25% of cases, microgyria and gray matter heterotopy in 10%, occipital encephalomeningocele in 3-5%, single ventricular forebrain in 25% [3, 29].

Supratentorial hydrocephalus should always necessitate evaluation of patency of the cerebral aqueduct as a part of MRI analysis, which is best assessed on median MRI sagittal images [32]. Accompanying aqueduct atresia requires implantation of two separate shunts – into the posterior fossa cyst and the supratentorial ventricular system

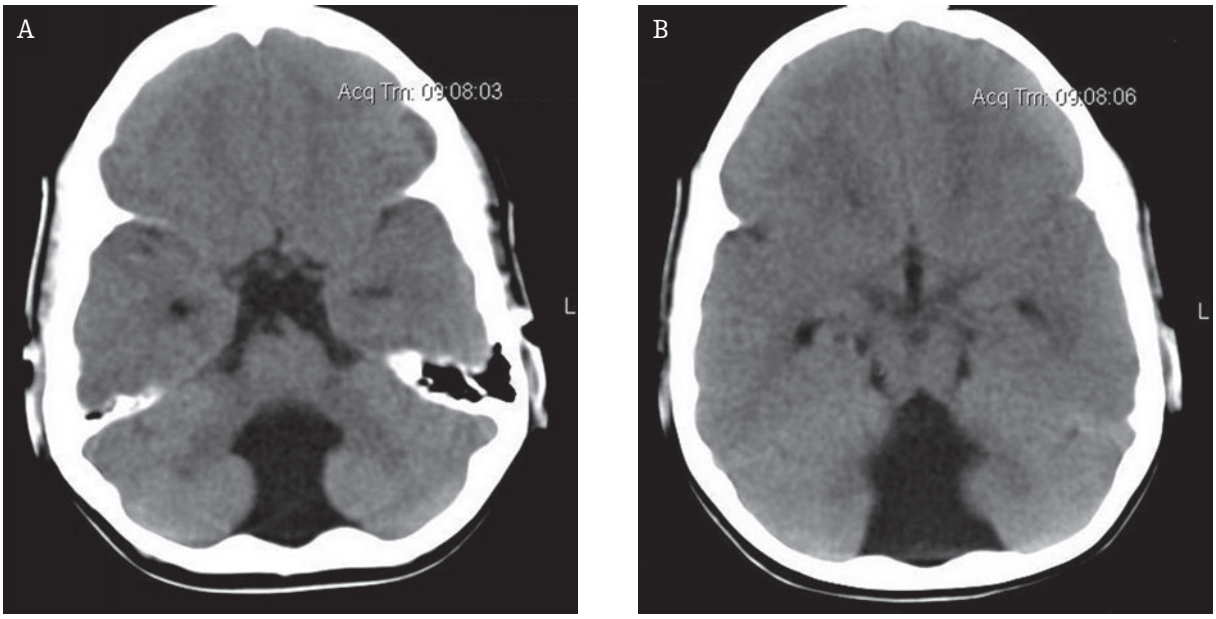

Figure 3. Dandy-Walker variant (DWV). On CT axial scans (**A**, **B**) hypoplasia of the cerebellar vermis (absence) is well seen. The fourth ventricle is moderately enlarged and the posterior fossa is only slightly increased in size.

[3, 19, 29, 33]. Shunting of the posterior fossa cyst only may lead to downward herniation of brain through tentorial incisure. Implantation of the shunt only to the supratentorial ventricular system may cause upward herniation of Dandy-Walker cyst through the tentorial incisure, which produces characteristic sign of "snowman" on sagittal MRI images [19].

Dandy-Walker syndrome variant (DWV)

The term "Dandy-Walker syndrome variant" (DWV) refers to cases resembling classic Dandy-Walker syndrome which are less intensive, and do not have every typical feature of the full syndrome (Fig. 3 A, B). Various definitions of DWV were invented in the last 30 years [3]. It was also proposed to abandon the term DWV and replace it with a new one – vermian-cerebellar hypoplasia [23]. In view of recently conducted patomorphological studies [3, 28] differences between Dandy-Walker syndrome and its variant in terms of patency of the fourth ventricle outflow foramina have lost their importance [3, 29].

On the overall, in the heterogeneous group of cases described as DWV the most common features on imaging include:
– hypoplasia of the cerebellar vermis and/or hemispheres, milder than in classic DWM [19, 30],
– large fourth ventricle, but lesser than in "true" DWM [4],
– the posterior fossa size is usually normal, or only slightly increased; the confluence of the sinuses is usually not displaced superiorly, therefore there is no characteristic lambdoid-torcular inversion [4, 29],
– in the majority of cases the brainstem is normal, accompanying supratentorial malformations are much less common (circa 20%) than in classic DWM [4],
– hydrocephalus is present in only 25% of cases [3, 4, 28].

The absence of features, which allow to distinguish classic DWM from its variant, has been emphasized in several studies [1, 22]. They are presently considered the same cystic developmental malformation with different intensity of individual features (DWV as a mild form of DWM), rather than two separate nosological entities [21].

Dandy-Walker Complex (DWC)

Some authors maintain that a cystic developmental malformations of the posterior fossa is a continuum of disturbances of different degree of severity. The spectrum would consist of mild (mega cisterna magna), moderate (heterogeneous group of Dandy-Walker variants) and severe ("true" classic Dandy-Walker syndrome) malformations [1, 3, 4, 7, 8].

In 1989 Barkovich et al. suggested to change the classification and assign these malformations as Dandy-Walker Complex (DWC). Type A of the complex would correspond to „true" DWM, whereas type B would include a heterogeneous group of malformations formerly called Dandy-Walker variant [1, 3, 4, 7].

DWC would not include discreet retrocerebellar fluid collections, without communication with the fourth ventricle and fluid spaces of cerebellar fissure, which are described as posterior fossa cysts corresponding to arachnoid cysts located behind and above the vermis [4, 7, 21].

Cases of hypoplasia of the vermis and the hemispheres with extended retrocerebellar fluid spaced *ex vacuo* mimicking cystic lesion have been left outside DWC definition.

Blake's pouch cyst (BPC)

Persistent Blake's pouch, also known as Blake's Pouch Cyst (BPC) (Fig. 4 A, B) is a new independent nosological entity introduced by Tortori-Donati [6]. It should be differentiated with Dandy-Walker syndrome, as it has different radiological appearance (especially in MRI) and different embryonic

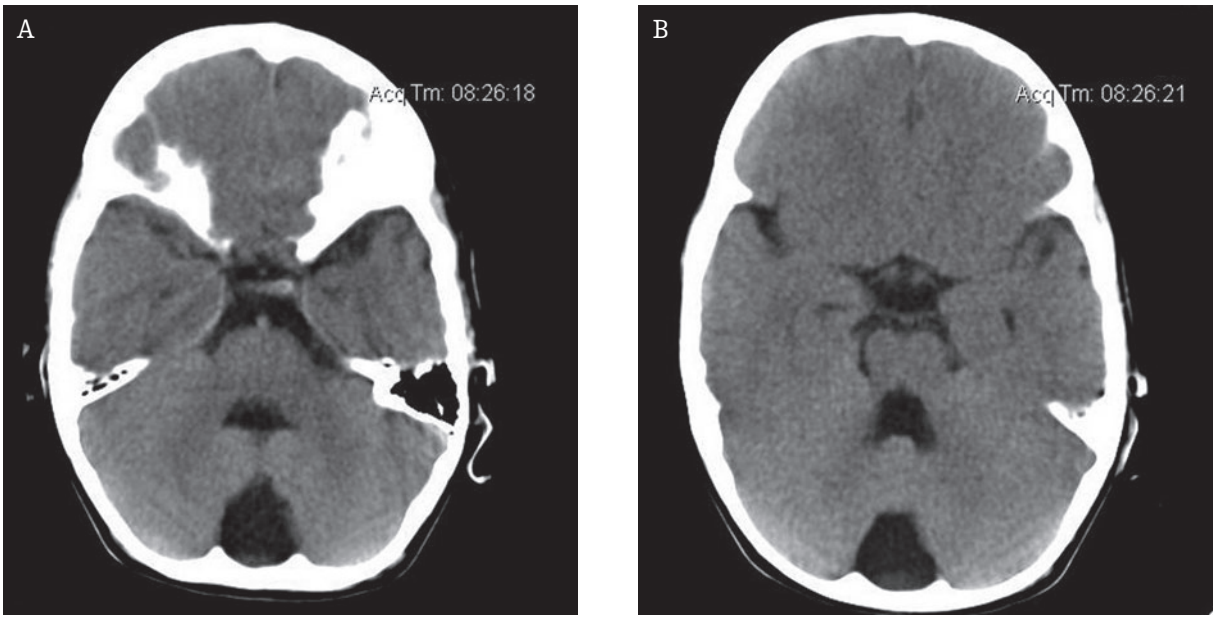

Figure 4. Blake's Pouch Cyst (BPC). On CT axial scans (**A**, **B**) show cystic fluid space localized posteriorly to the normal vermis.

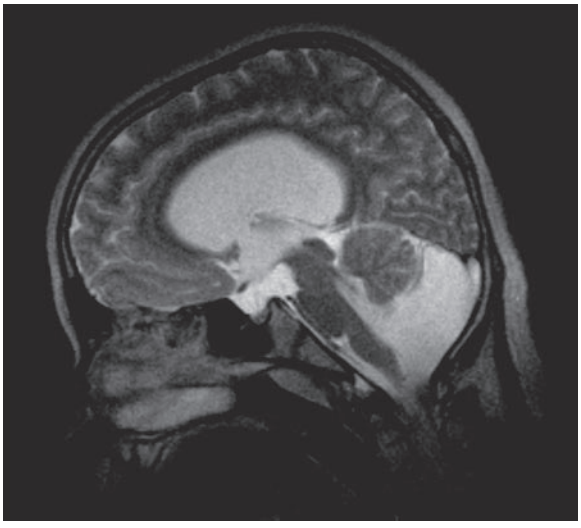

Figure 5. Blake's Pouch Cyst (BPC). MRI saggital T2-weighted image reveals a large cystic fluid space which is localized inferiorly and posteriorly to the vermis and communicates widely with the fourth ventricle. Supratentorial hydrocephalus is present, while the fourth ventricle is only mildly dilated. There is no lambdoid-torcular inversion, typical of DWM.

origin [3, 5, 6, 7, 9, 10, 11]. Continuity between BPC and DWC has also been proposed [21]. It has been argued, that these malformations may overlap each other in terms of radiological and clinical presentation. They present with different degree of development of the medullary veli, cerebellar vermis, posterior fossa cisterns and surrounding meninges [21].

BPC is a posterior bulging of the inferior medullary velum into the cisterna magna, posteriorly to the cerebellar vermis, often also above the vermis [7, 9]. BPC communicates with the fourth ventricle and is separated from subarachnoid fluid spaces of the posterior fossa. It is always associated with hydrocephalus [3, 6]. There is no dysplasia of the cerebellar vermis [3]. BPC requires implantation of a ventriculo-peritoneal shunt, or preferably endoscopic third ventricle vetriculostomy [11].

Radiological criteria of the persistent Blake's Pouch Cyst (BPC) include:
- cystic fluid space is localized posteriorly, inferiorly or above the normal vermis (Fig. 5) and cerebellar hemispheres (occasionally lower part of the vermis may be secondarily hypoplastic) [1, 7]. It creates mass effect directed posteriorly and inferiorly, which can be best visualized on MRI T1-weighted sagittal views. The occipital squame may be molded and thinned [1],
- wide communication between the fourth ventricle and BPC, through markedly dilated cerebellar vallecula [7],
- supratentorial hydrocephalus is always present, however, the fourth ventricle is usually only mildly dilated or normal [7],
- the confluence of the sinuses is displaced superiorly, but there is no lambdoid-torcular inversion, typical of DWM.

In the majority of cases, BPC is diagnosed in the early childhood, because its signs are present since birth (hydrocephalus). Rarely, if patency of Luschka's foramina is preserved, it is diagnosed in older children [7].

On imaging, BPC may mimic other cystic malformations. The key to distinguish these conditions is the position of the choroid plexus in relation to the fourth ventricle lumen. In case of retrocerebellar arachnoid cyst, the plexus is correctly located within the ventricle. There is usually no plexus in DWM, and in case of BPC it is displaced toward the upper wall of the cyst. When the appearance of malformation is not unequivocal and the pathology of the wall unknown, it is suggested to use a descriptive term – retrocerebellar cyst [10].

Cine-MR imaging of pulsatile flow of the cerebrospinal fluid may reveal the lack of communication between the fourth ventricle and the cerebellomedullary cistern.

Mega cisterna magna (MCM)

In physiologic conditions, the cisterna magna (cerebellomedullary cistern) is a space separating the leptomeninx and arachnoidea, and it is located between the lower surface of the cerebellum and medulla oblongata and the spinal cord, in the lower part of the posterior fossa and the upper part of the spinal canal [3, 6].

Mega cisterna magna (MCM) is considered a mild malformation, or a developmental variant with large cistern. It is because cerebello-medullary cistern varies greatly in size in every patient. As the development of the cerebellum is not complete, the cistern may be relatively large [3, 23].

MCM occurs in about 0.4% of the population and constitutes approximately half of the cases of posterior fossa cystic developmental malformations [4, 12].

MCM is best visualized on sagittal and axial T1-weighted MRI scans (Fig. 6).

It is presented as cerebrospinal fluid collection located posteriorly and inferiorly to the normal cerebellar vermis. The vermis is often displaced anteriorly. Occasionally the lesion may be accompanied by molding of the occipital squame, but there is no significant mass effect and hypoplasia of the cerebellar vermis [1] (Figs. 6, 7).

The size of MCM is various, rarely it may reach even the level above of the vermis, up to the straight sinus and laterally to the region of both cerebello-pontine angles. In the majority of cases, MCM is divided in sagittal plane by the cerebellar falx, which is frequently asymmetric, double or multipartite [1]. Rarely, in 12.5% of cases, the confluence and the straight sinus may be positioned higher, but there is no lambdoid-torcular inversion which is typical of DWM [3, 1]. The size and shape of the fourth ventricle are usually normal, less commonly it is slightly enlarged [1]. MCM by definition communicates with the normal fourth ventricle and other cisterns of the subarachnoid space; therefore, there is no associated hydrocephalus [3, 4, 6]. The posterior fossa has normal size or is only slightly enlarged [4].

Associated developmental disturbances and neurological abnormalities are present in 60% of MCM cases [12]. Neurological abnormalities are usually caused not by MCM itself, but by accompanying supratentorial malformations, such as dysgenesis of the corpus callosum or holoprosencephaly [4]. For that reason, MCM is considered an indicator of abnormal brain development or developmental malformations [12].

MCM should be differentiated mainly with DWM and DWV. Sagittal MRI, particularly T1-weighted images, are crucial in this aspect. In cases of MCM, they show normal size or slight narrowing of fourth ventricle and normal structure of the cerebellar vermis [1, 34, 35].

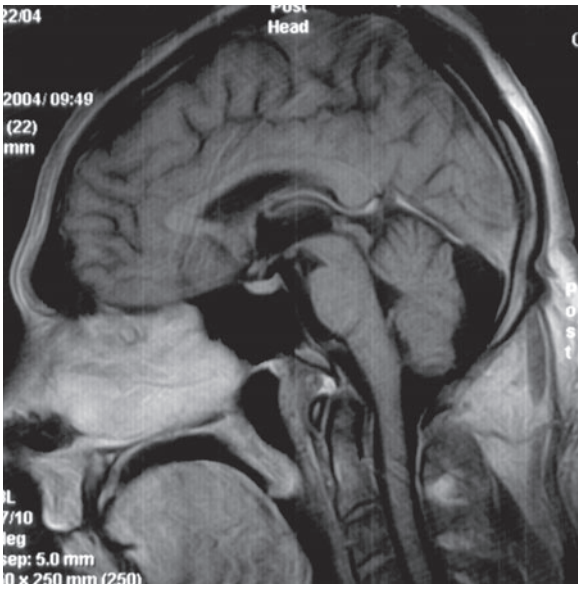

Figure 6. Mega Cisterna Magna (MCM). MRI saggital T1-weighted image shows small fluid collection localized posteriorly to the normal cerebellar vermis. The fourth ventricle and posterior fossa present normal. There are no signs of hydrocephalus.

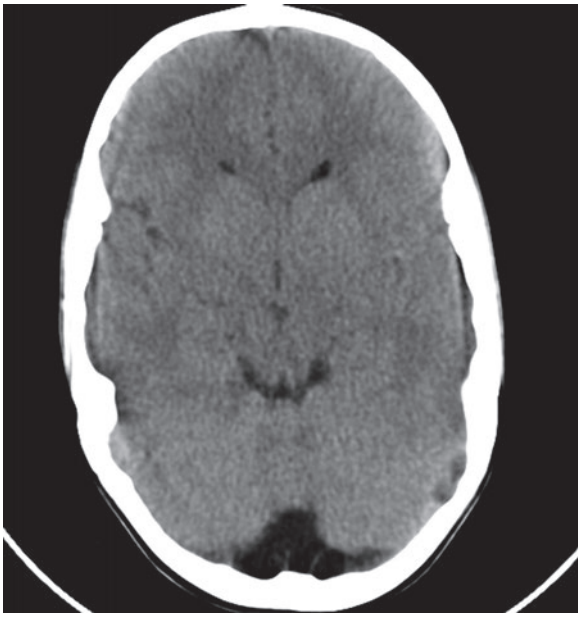

Figure 7. Mega Cisterna Magna (MCM), axial CT scan of another patient exhibits fluid collection located posteriorly to the cerebellar vermis and hemispheres. In the fluid collection, bifid cerebellar falx is seen.

Arachnoid cyst of the posterior fossa (AC)

The term arachnoid cyst (AC) refers to a mild, fluid-filled developmental cavity within the subarachnoid space with no communication with the ventricular system. Usually AC is not associated with abnormal development of the brain or other systemic malformations [4, 32]. Predominantly, it is a single compartment lesion; however, internal septa may occur. It is filled with fluid of

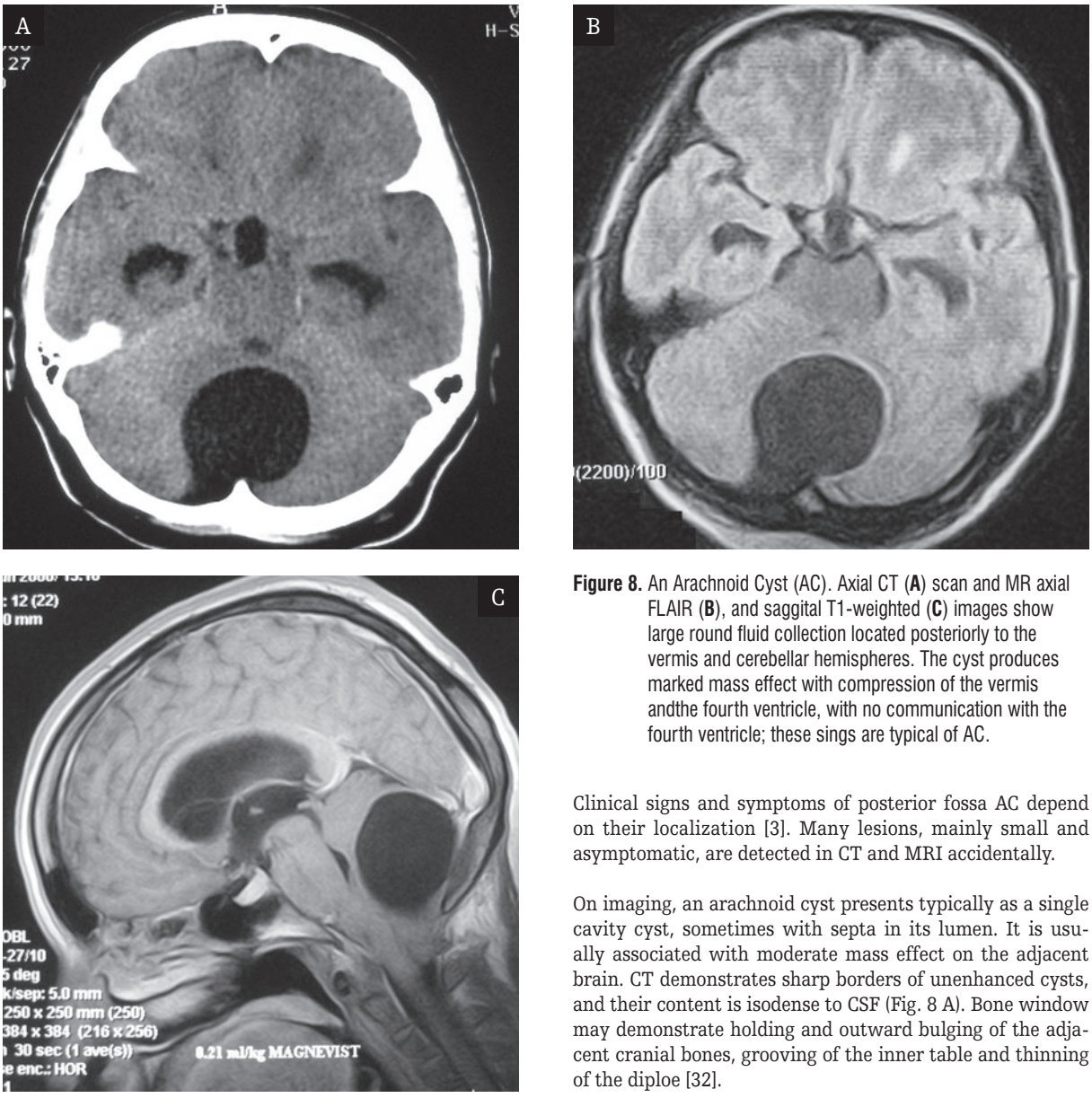

Figure 8. An Arachnoid Cyst (AC). Axial CT (**A**) scan and MR axial FLAIR (**B**), and saggital T1-weighted (**C**) images show large round fluid collection located posteriorly to the vermis and cerebellar hemispheres. The cyst produces marked mass effect with compression of the vermis andthe fourth ventricle, with no communication with the fourth ventricle; these sings are typical of AC.

similar composition to cerebrospinal fluid (CSF) [3, 32]. A small percentage of AC may contain fluid with elevated protein level [32].

The degree of separation of AC from CSF subarachnoid space cisterna is debated. In the majority of cases AC does not communicate with the subarachnoid space [1]. In a substantial percentage of cases (up to 30%), an application of MRI techniques of visualization of pulsatile CSF flow (2D-FISP sequence, cine option) allows to present fluid pulsation and proves the connection with the subarachnoid space [16, 17].

AC is the most common in the middle cranial fossa (50-60%) and relatively common infratentorially – 16 to 23% [3, 32]. In the posterior cranial fossa, AC has predilection to: the retrocerebellar space (9-16%), cerebello-pontine angle (2-11%), vicinity of the quadrigeminal cistern (to 10%) [3, 4, 32, 36].

Clinical signs and symptoms of posterior fossa AC depend on their localization [3]. Many lesions, mainly small and asymptomatic, are detected in CT and MRI accidentally.

On imaging, an arachnoid cyst presents typically as a single cavity cyst, sometimes with septa in its lumen. It is usually associated with moderate mass effect on the adjacent brain. CT demonstrates sharp borders of unenhanced cysts, and their content is isodense to CSF (Fig. 8 A). Bone window may demonstrate holding and outward bulging of the adjacent cranial bones, grooving of the inner table and thinning of the diploe [32].

MRI better visualizes location of the lesion, its dimensions and relation to the adjacent structures (Fig. 8 B,C). Retrocerebellar AC is presented in this examination as a well-delineated, unenhanced round or oval space, located outside the cerebellum [3, 32]. In general, the content of the AC is isodense to CSF in all sequences [3, 32]. AC containing fluid with high protein level or blood products may cause diagnostic difficulties – their signal is not suppressed in FLAIR sequence and is hyperintense to CSF [32]. In diffusion weighted imaging (DWI), due to not restricted diffusion, AC shows low signal, which is a valuable sign in differential diagnosis with other cysts.

The AC wall is free of parietal nodules and calcifications and it does not enhance after gadolinium administration. Occasionally, with associated inflammatory changes, there may be enhancement of the wall, and in this case the differential diagnosis should always include a cystic neoplasm [1].

If AC is localized posteriorly to the cerebellum, it must be always differentiated with enlarged cerebello-medullary cistern (MCM). MCM and AC have similar signs both on T1- and T2-weighted images and in FLAIR sequence (cerebrospinal fluid signal). Sometimes differentiation may be very difficult. Generally ACs tend to cause mass effect, whereas MCM should not produce significant mass effect [3, 32]. Moreover, AC does not communicate with the fourth ventricle, whereas MCM does [4].

Differential diagnosis of these two lesions may be greatly facilitated by CSF flow imaging technique (2D-FISP sequence). This examination allows to evaluate pulsatile flow of the cerebrospinal fluid in the ventricular system, basal cistern and within the cyst itself. In the majority of cases, there is no flow in AC; however, in some cases the flow may be observed, proving communication with the CSF space. In MCM cases, there is intensive, pulsatile CSF flow originating from central CSF spaces – that is from the fourth ventricle [16, 17].

If AC produces significant mass effect on the adjacent brain or causes hydrocephalus, it requires surgical treatment – implantation of a shunt placed in the cyst or fenestration of its wall [4, 36].

Conclusions

On the base of reviewing the current literature of cystic developmental malformations of the posterior fossa the following conclusions may be drawn:

1. Since the first Dandy's publication in 1914, definitions and classification of the cystic developmental malformation have been changed and modified many times. This is caused by weak, superficial understanding of the problem among physicians, including radiologists.
2. Contemporary classification of posterior fossa cystic malformations is based on embryogenesis of the hindbrain, cyst pathology and the appearance on the multiplane MRI. It distinguishes the malformation caused by area membranacea superior (AMS) defect: Dandy-Walker syndrome and its variant, as well as malformations caused by a defect of the area membranacea inferior (AMI): mega cisterna magna and persistent Blake's pouch. Posterior fossa arachnoid cyst should be distinguished as a separate entity.
3. Cerebellar hypoplasia, although not belonging to cystic malformations, should also be mentioned. As it causes the enlargement of the paracerebellar spaces which mimic the cysts, it may lead to misdiagnosis.
4. Clinical and radiological appearances of individual malformations may partially overlap, therefore differentiation only on the base of neuroimaging, especially on axial CT is insufficient.
5. If the appearances of the lesion is not unequivocal, and the wall pathology unknown, it is suggested to use a descriptive term – retrocerebellar cyst.
6. It should be understood, that the radiologist's task is finding the malformation, detailed description of its topography and the effect on the brain, diagnosis of accompanying developmental anomalies, but not necessarily determination of a definite type.
7. In diagnostics of cystic developmental malformations of the posterior fossa, MRI techniques for imaging of cerebrospinal fluid pulsatile flow (2D-FISP sequence, cine option) gain increasing importance. These techniques efficiently replace CT cisternography, and may play an essential role in decision making in terms of indications and methods of neurosurgical treatment.

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