Imaging examinations in children with hydrocephalus

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Abstract

Hydrocephalus is characterized by an imbalance of cerebrospinal fluid (CSF) formation and absorption. It is manifested as a dilatation of the ventricular system. About 55% of all hydrocephalus cases have congenital origin. There are two types of hydrocephalus: communicating and non-communicating with subarachnoid space and the diagnosis depends on the computed tomography (CT) and magnetic resonance (MR) images. The treatment is different for each type of the hydrocephalus. Causes and symptoms of hydrocephalus are changing with the patient's age. Before the age of two we can observe progressive enlargement of the head and widened anterior fontanel. Ophthalmological examination reveals optic nerves atrophy. Children older than two years with hydrocephalus and obliterated anterior fontanel have normal head circumference. They may often present clinical symptoms such as the atrophy of optic nerves and papilloedema of optic disc. The most common reason of hydrocepahalus in children before two years of age is intraventricular haemorrhage in the perinatal period whereas in children older than two years is inflammatory process. Imaging examinations are needed not only to diagnose hydrocephalus but also to assess enlargement of the ventricular system during the therapy.

Key words: hydrocephalus, brain, congenital malformation central nervous system (CNS).

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Diagnosis of the hydrocephalus is based on a correlation between clinical symptoms of elevated intracranial pressure and the image of dilated ventricular system. Hydrocephalus is a result of an imbalance of cerebrospinal fluid (CSF) formation over its absorption [1]. Hydrocephalus is not a disease but a dynamic process which proceeds with changes of the ventricular system size [2]. From the diagnostic standpoint and the therapeutic methods the hydrocephalus can be divided into; communicating hydrocephalus whereas narrowed place of CSF outflow exists outside ventricular system and non-communicating hydrocephalus whereas the location of obstruction lies intraventriculary. In the second case it is accompanied by narrow subarachnoid space [3]. Hydrocephalus can accompany ventriculomegaly as a secondary process. Ventriculomegaly is a result of brain tissue atrophy and malfunction of CSF circulation [4]. In some cases it is very difficult to identify primary hydrocephalus which leads to regressive changes and secondary hydrocephalus. Causes and clinical symptoms of hydrocephalus are changing with the patient's age but 55% of all cases have congenital origin [3]. Children before the age of two present with symptoms of hydrocephalus like large head circumference, wide anterior fontanel with dilated scalp veins. Increased muscle tone and paralysis of upward gaze are frequent at clinical examination. In ophthalmological examination atrophy of the optic nerves are presented.

Children older than 2 years with obliterated fontanel tend to present with neurological symptoms of the increased intracranial pressure while normal head size. When dilated third ventricle compresses hypothalamic structures hormonal dysfunctions such as sexual maturation abnormalities, gigantism, diabetes mellitus can be the first symptoms of hydrocephalus. Dilated ventricular system coexists with slightly dilated subarachnoid space in infants up to six months of age [4]. This condition may result from transitory immaturity of choroid plexus. The most common cause of ventriculomegaly in children before age of two is atrophy of brain tissue as well congenital anomalies of corpus callosum, holoprosencephalon and lissencephalia [5].

Figure 1. **CT examination of the head. Massive subependymal calcifications with dilated ventricular system**

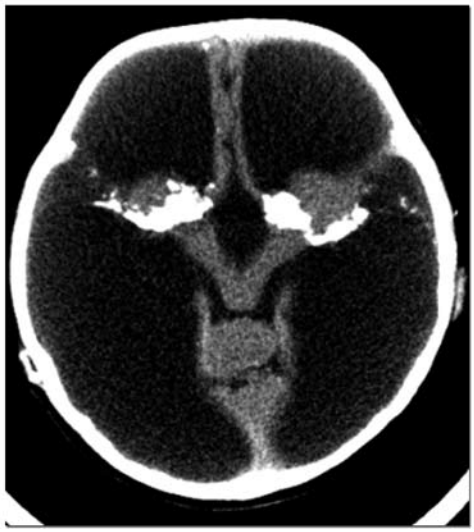
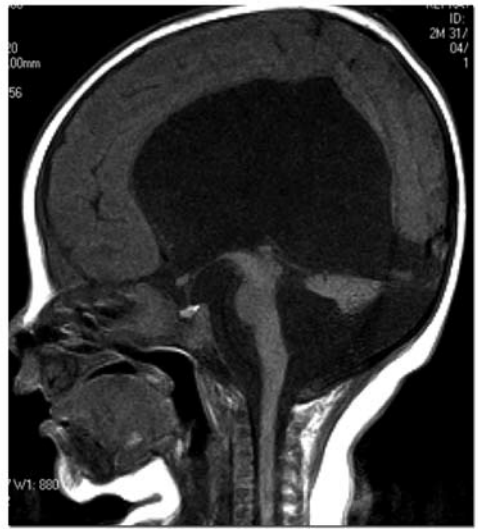


Figure 2. **MRI of the head. Sagittal T1-weighted (a), axial (b) T2-weighted images. Agenesis of the vermis of cerebellum with hypoplastic cerebellar hemispheres. Agenesis of corpus callosum**

a)



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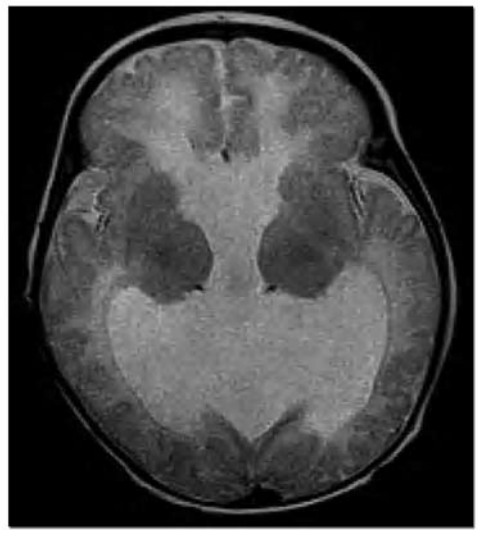


The most frequent cause of hydrocephalus with elevated ICP (intracranial pressure) in children before age of two is intraventricular haemorrhage in perinatal period [2]. The next causes are the congenital inflammatory process of the CNS such as TORCH (toxoplasma, rubella, cytomegaly, herpes simplex) which lead to brain damage (*Fig. 1*). Hydrocephalus as a symptom can coexists with such developmental anomalies like Dandy-Walker syndrome, myelomeningocele or narrowed Sylvian aqueduct (*Fig. 2*) [5]. Hydrocephalus is often the result of head injury with intracranial haemorrhage in this group of age.

One of the causes of hydrocephalus are CNS toumors (the second common neoplasm disease in children beside leukemia) filling the ventricular system [6,7]. The level of ventricular system enlargement depends on the tumour localisation. Metastases in the subarachnoid space and inside the ventricular system may occur in children treated from primary CNS neoplasm. They often block CSF outflow and may result in hydrocephalus.

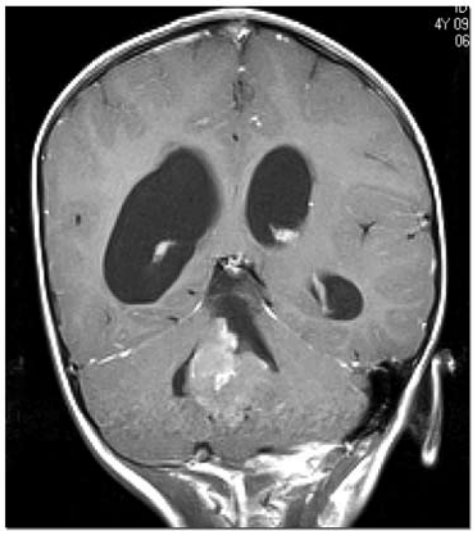
Children older than 2 years have another sequence of causes. First of all we should list inflammatory process of the brain which can include meningitis. Hydrocephalus can be an outcome of complicated or improper treated inflammation [8]. It can develop as a result of secondary scar lesions in a subarachnoid space. The next cause of hydrocephalus is subtentorial neoplastic process [7]. Posterior fossa tumours such as medulloblastoma, ependymoma and astrocytoma result in symmetric supratentorial ventricular system enlargement due to their localisation (*Fig. 3*).

Among the causes of hydrocephalus in the group of elderly children congenital anomalies of CNS such as arachnoidal cyst which gives delayed neurological symptoms of elevation of the intracranial pressure can be found [9]. Imaging examinations make possible both diagnosis and establishing causes of hydrocephalus. Performing a computed tomography (CT) examination or magnetic resonance imaging (MRI) we can estimate the degree of ventricular system dilatation [10]. Some measurements should be done; the width of the temporal horn in relation to the width of the body of lateral ventricle [2]. Enlargement of the temporal horn commensurately with the bodies of the lateral ventricles is a sign on differentiation of hydrocephalus to atrophy. Reduced ventricular angle made by line going through the medial wall of the frontal horn to the long axis of the brain is a sign of hydrocephalus. Imaging examinations differentiate the normal pressure and the elevated ICP hydrocephalus [11]. They reveal zone of periventricular brain tissue oedema which can be seen at the level of frontal horns of lateral ventricles. Another symptom of dilated ventricular system is smoothing out of cerebral cortex [4]. On sagittal MR images we can measure the distance between mammilary bodies and brainstem or the size of recesses of the third ventricle.

Treatment of hydrocephalus is based on two surgical methods; ventriculoperitoneal shunt insertion in the case of communicating hydrocephalus and endoscopic third ventriculostomy

***Figure 3*. MRI of the head. Sagittal (a), coronal (b) T1-weighted contrast-enhanced images. Tumour filling the fourth ventricle with supratentorial hydrocephalus**

a)



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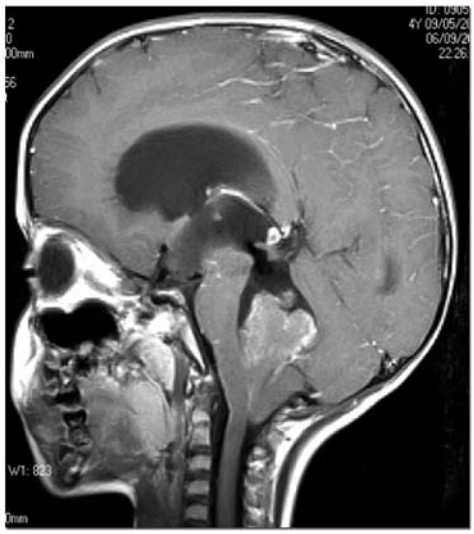
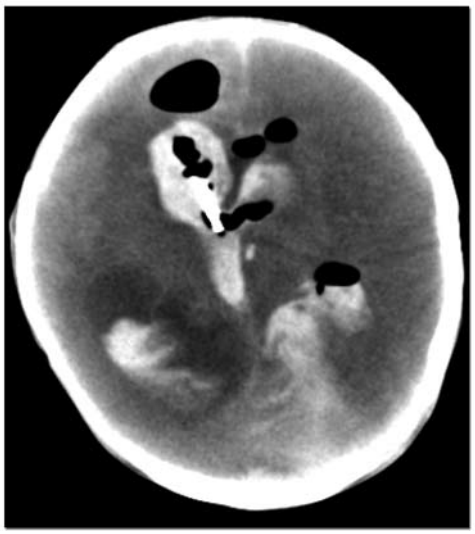


Figure 4. CT examination. Blood and air pockets in the ventricular system as a complication after the ventriculoperitoneal shunt insertion. Cerebral oedema



in the case of non-communicating hydrocephalus [12]. The most often place of ventriculostomy is perforation the floor of the third ventricle just anterior to the maxillary bodies [8]. It makes possible gradual decrease in size of the ventricles. To control shunt function or ventriculostomy condition imaging examinations are performed [13].

Beside assessing the size of ventricular system we can assess route and location of the intracranial proximal segment. Insertion of the ventricular shunt may result in large numbers of complications which present within six month from surgery.

The overall rate of shunt malfunction is 40% for patients before the age of two and 30% in older age group [2]. These complications can concern both shunt tips; intracranial and distal shunt catheters [3].

The most severe complication of ventricular tip insertion is intracranial haemorrhage as a result of procedure or rapid decompression of the ventricular system (*Fig. 4*) [4].

During the insertion of the proximal segment anatomical structures can be damaged or focal neurological deficits can occur such as hemiparesis if the catheter transverses the internal capsule. Meningeal fibrosis may occur as a reaction to chronic subdural hygromas which can be the consequence of shunt insertion [1]. Inflammatory changes around the proximal intracranial segment of shunt catheter penetrating into ependyma and subependymal zone of ventricular system are observed with rate of 5 to 10% of all children with chronic ventricular shunt (*Fig. 5*) [8]. Chronic shunt placement can cause craniosynostosis in the youngest children with separated sutures. A slit ventricle syndrome can become a problem in a small subset of 1% to 5% of older children chronically shunted [4]. These patients present symptoms of intracranial hypertension although they have small ventricles in CT findings. Another complication in shunted patients with non-communicating hydrocephalus is the isolated fourth ventricle [14].

There is no physicodynamic condition for CSF to outflow through Sylvian aqueduct due to shunting of the lateral ventricles. Cerebrospinal fluid produced in the fourth ventricle can outflow neither from obstructed aqueduct of Sylvius nor through subarachnoid space [4]. It leads to cystic dilatation of the fourth ventricle observed on imaging studies.

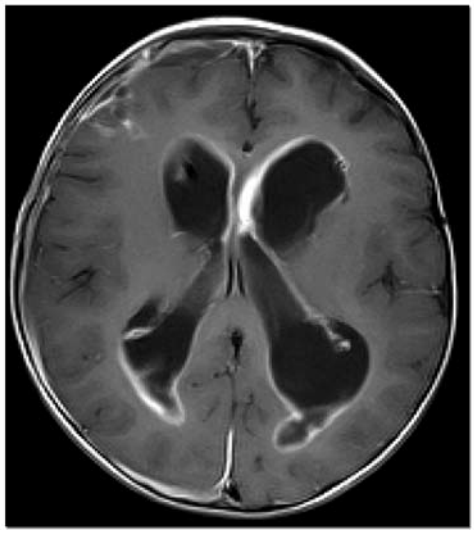
Among the complications of distal catheter are observed: limited fluid cysts, peritonitis, hepatic abscesses, fistulas and perforations of gastrointestinal tract [3].

In the end we need to mention about other radiological examinations carried out to assess hydrocephalus treatment results [11]. Transcranial Doppler ultrasonography sometimes is performed in infant with hydrocephalus [15]. This method allows to evaluate maturity of autoregulation of blood flow in the cerebral vessels. These vessels could be partly or completely damaged in a long lasting treatment with ventricular shunt.

Children with shunt insertion may have MR ventriculography performed [16]. Inserting the needle into the catheter and injecting paramagnetic contrast agent we can observe the flow

Figure 5. **MRI of the head. Axial (a), sagittal (b) T1-weighted contrast-enhanced images. Inflammatory changes of the brain tissue located around the zone of the inserted catheter and ependyma of the ventricular system**

a)



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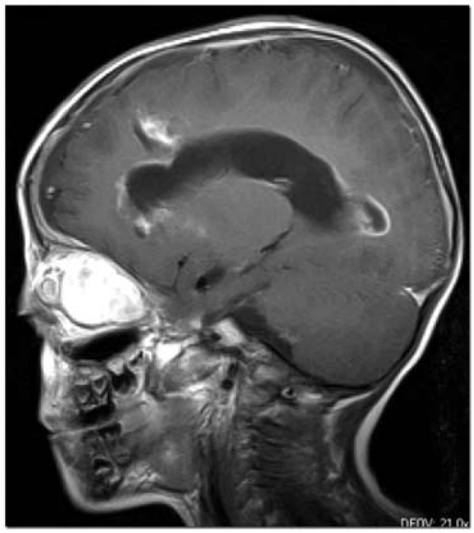
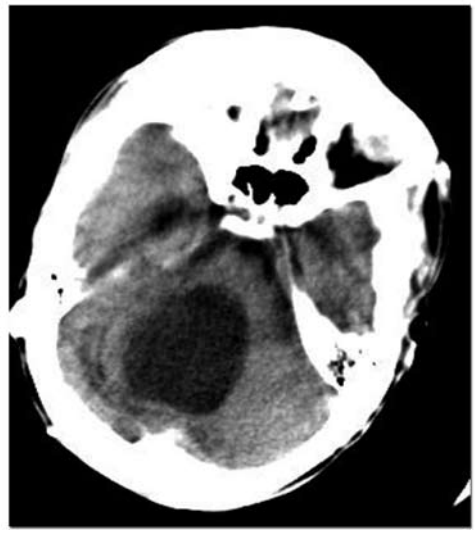


Figure 6. **CT examination. Cystic enlargement of the fourth ventricle**



of this agent in every image. We can also inject isotopic agent (99 m) Tc into ventricular system and evaluate CSF circulation with single photon emission computed tomography (SPECT), [17]. Among all examinations MR imaging has the best diagnostic value due to excellent quality images and no evidence of harmful effect on the living organism.

Using rapid sequences technique we can eliminate the need for child sedation and reduce examination to the one projection only in follow-up studies.

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