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НЕЙРОФІБРОМАТОЗ У НЕВРОЛОГІЧНІЙ ПРАКТИЦІ (З ОПИСОМ КЛІНІЧНОГО ВИПАДКУ НЕЙРОФІБРОМАТОЗУ ДРУГОГО ТИПУ)

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Анотація. Нейрофіброматоз являє собою комплексне захворювання генетичної природи з безліччю симптомів і значною фенотипічною варіабельністю. Найбільш важким є другий тип нейрофіброматозу. Він характеризується розвитком пухлин центральної і периферичної нервової систем. У статті описаний клінічний випадок нейрофіброматозу другого типу. Діагностика і лікування цього захворювання вимагає комплексного мультидисциплінарного підходу.

Ключові слова: нейрофіброматоз другого типа, клінічний випадок, симптом, фенотипічна варіабельність, діагностика та лікування.

NEUROFIBROMATOSIS IN NEUROLOGICAL PRACTICE (DESCRIPTION OF A CLINICAL CASE OF NEUROFIBROMATOSIS 2ND TYPE)

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Summary. Neurofibromatosis is a complex genetic disorder with multiple symptoms and significant phenotypic variability. The most severe is second type of neurofibromatosis. It characterizes by tumors of central and peripheral nervous system. The article describes a complex clinical case of neurofibromatosis type 2. Such a clinical case in neurological practice can be found infrequently. The complexity of diagnosis and treatment of these diseases requires a coordinated multidisciplinary approach.

Key words: neurofibromatosis type 2, clinical case, symptoms, phenotypic variability, diagnosis and treatment.

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HYDROCEPHALUS FROM PAST TO NOWADAYS

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Summary. The study and treatment of hydrocephalus all time excited the physicians. Understanding the reasons of appearance and development of this diseases and also accomplish it anomalies of nervous system help create a system of adequate treatment. Untreated hydrocephalus has a survival rate of 40–50%, with the survivors having varying degrees of intellectual, physical, and neurological disabilities. Prognosis for treated hydrocephalus varies, depending on the cause. Early diagnostic and qualified treatment can improve the outcomes and prognosis of these patients.

Key words: hydrocephalus, anomalies of development nervous system, cerebrospinal fluid, ventricles of brain, shunt system.

Introduction in history development

Hydrocephalus comes from the Greek words «hydro» meaning water and «cephalus» meaning head. Hystory studying of hydrocephalus was long.

The study and treatment of hydrocephalus, over the centuries, passed three stages of evolution. During antiquity, middle ages and renaissance, hydrocephalus was not understood. Prior to the late 19th century, treatment for «water on the brain» included more observation than intervention. Medical treatment was useless; surgery was hopeless. The second stage extends from the 19th century to the end of the first half of the 20th century [1, 2]. Cerebrospinal fluid (CSF) circulation was not understood; surgery however, remained inefficient, but some patients survived with arrested hydrocephalus. The third stage begins in the nineteen fifties with the development of

silicone shunts with a valve. Surgery transforms the prognosis of hydrocephalus, but the number of post-operative complications creates new problems. They made different attempts to solve these problems. As a result was a reduction of the mechanical and infectious complications. CSF overdrainage has been minimized. Percutaneous ventriculo-cisternostomies have in some cases replaced shunts. In the future for improvement result in hydrocephalic should be performed surgery as early as possible.

Problem of hydrocephalus always exited many physicians since ancient time.

Hippocrates (5th century B. C.), the father of medicine, was the first physician who attempt and document the treatment of hydrocephalus, performed ventricular punctures, drained the subdural or subarachnoid space, recommended trepanning for the treatment of epilepsy, blindness and possibly hydrocephalus.

In the works of Galen (130–200 A. D.) indicated this condition as caused by an extraaxial accumulation of cerebrospinal liquid rather than enlargement of the ventricles. This belief led to many erroneous diagnoses and treatments. He recounted examples and described the thinness of the brain and skull associated with this condition. He found the connection of ventricles [2].

Most detailed descriptions of hydrocephalus including the surgical treatment are extant in the encyclopaedic works on medicine of the physicians Oreibasios and Aetios from Amida since the 4th and 6th centuries A. D.

Due to the lack of autopsies in ancient times, the hydrocephalus was never linked to the pathology of the ventricles. All forms of hydrocephalus were believed to be caused by improper handling of the head by the midwife during delivery.

In the Middle Ages, the Arabic surgeon Albucasis (936–1013), wrote a medical encyclopedia which was taught at Muslim and European medical schools until the 17th century, he touched on many aspects of neurosurgery, including the diagnosis and treatment of hydrocephalus. Abu Ali al-Husain ibn Abdallah ibn Sina, is often known by his Latin name of Avicenna, separated the traumatic hematomas outside the skull from the term hydrocephalus. But Avicenna, had not linked hydrocephalus with the ventricular system.

Andreas Vesalius (1514–1564), a Flemish anatomist, revealed as a single pathology an extremely dilative ventricular system filled with water-like fluid which allow it necessary to change completely the ancient concept of hydrocephalus. In 1664 Thomas Willis (1621–1675), neuroanatomist, considered by many to be the father of neuroscience was the first to suggest that the choroid plexuses produced CSF. In 1701, Antonio Pacchioni (1665–1726), an Italian scientist and anatomist, described the arachnoid granulations, which he falsely believed were the source of CSF production. In 1761, Giovanni Battista Morgagni (1682–1771), an Italian anatomist, wrote in Seats and Causes of Diseases that hydrocephalus could occur without accompanying head enlargement; however, he did not know the source of the excess fluid in this disease process. Monro illustrated the presence of the paired intraventricular foramen. Robert Whytt (1714–1766) first described hydrocephalus as a disease, illustrating several cases of internal hydrocephalus caused by tuberculous meningitis. François Magendie (1783–1855), a French physiologist in 1825 described the circulation of CSF within the brain. A German anatomist, Hubert von Luschka (1820–1875), in 1859 confirmed the presence of the foramina of Magendie and described two additional lateral foramina. Heinrich Irenaeus Quincke (1842–1922) first described the lumbar puncture as an effective treatment for hydrocephalus in 1891. In 1881 Carl Wernicke (1848–1905), a German physician, anatomist and neuropathologist, inaugurated sterile ventricular puncture and external CSF drainage. In the early 20 th century, Lewis Weed described the embryology of the choroid plexus and confirmed the absorptive capacity of the arachnoid villi. Johann von Mikulicz-Radecki (1850–1905) first attempted drainage from the lateral ventricle to the subgaleal, subdural and subarachnoid spaces. It was simultaneously a ventriculostomy and a drainage into an extrathecal low pressure compartment. Between 1898 and 1925, lumboperitoneal and ventriculo-peritoneal, -venous, -pleural and ureteral shunts were invented, but these had a high failure rate due to insufficient implant materials in most cases. Harvey Williams Cushing (1869–1939), an American neurosurgeon devised a technique in which the lumbar subarachnoid space was connected to the peritoneal cavity or retroperitoneum by using silver cannulas passed through apertures through the L-4 vertebral body. In 1914, Walter Edward Dandy and Kenneth D. Blackfan developed a technique of producing experimental obstructive hydrocephalus in dogs by placing cotton pledgets at the distal aqueduct of Sylvius, thereby causing proximal ventricular dilation. Dandy to introduce, in 1918, bilateral choroid plexectomy as a means of reducing CSF production. Charles Putnam Symonds and

John Edwin Scarff expanded on this technique by including endoscopic cauterization of the choroid plexus in the late 1930s and early 1940s. The placement of intracranial shunts was also investigated. Third ventriculostomy was introduced by Dandy to bypass aqueductal stenosis, and this technique was later refined by Stookey and other scientists. This technique was further improved with the use of endoscopes. Arne Torkildsen devised a procedure in which a shunt was placed from the lateral ventricle to the cistern magna (ventriculocisternostomy); initially the success rate was high but so too was surgery-related morbidity, which was subsequently reduced. The results study of CSF summarized Russian scientist D. Shamburov (1887–1963), in monographs «Cytology liquor» and «Cerebrospinal fluid». V. Khoroshko (1881–1949) headed the neurological clinic Moscow in 1931–1949 the first time in Moscow began to apply spinal puncture, he has developed a method pneumoencephalography. A. A. Arendt (1948) studied hydrocephalus and its surgical treatment [3]. After 1980 in former USSR a significant place in the treatment of hydrocephalus began using endoscopic surgery. The development that declared in the modern era of hydrocephalus surgery was the introduction of valve regulated shunts and biocompatible synthetic materials in 1952. In 1952 use of a ventriculo-jugular shunt regulated by a spring and ball valve. In 1970, Thomas Herrick Milhorat illustrated the increase in periventricular permeability and the concept of transependymal absorption in experimental hydrocephalus. In the 1980s and 1990s the use of an endoscope again found a role in neurosurgery, the benefits of which include more accurate placement of ventricular catheters and a resurgence of the third ventriculostomy for aqueductal stenosis. In the 1990, there has been a renaissance of endoscopic ventriculostomy as a method of first choice in adult patients with aquired or late-onset, occlusive hydrocephalus. Development of prenatal ultrasonography, diagnostic of hydrocephalus in utero in the late 1970s and early 1980s has led to attempts with intrauterine fetal surgery. Such procedures as ventriculoamniotic shunts and serial cephalocenteses were attempted to curb the ventriculomegaly. Fetal therapy for spinal dysraphism – associated hydrocephalus has seen an emergence in recent years, spurred on by advances in neuroimaging, better understanding of the pathophysiological nature of the disease, and the refinements of surgical techniques including endoscopy. Bruner have reported the first cases of intrauterine closure of a myelomeningocele in 1997. The suspected benefits of this early intervention include decreased hindbrain herniation, improvement of lower extremity function and the decreased need for shunts.

Peculiarity of the brain circulation

Nowadays known that in normal CSF is produced mainly within the lateral and third ventricles by the choroid plexus. The ventricles are connected by narrow passageways. CSF flows from the lateral ventricles through two narrow passageways into the third ventricle. From the third ventricle, it flows down another long passageway known as the aqueduct of Sylvius into the fourth ventricle. From the fourth ventricle, it passes through three small openings called foramina (Luschke and Magendie) into the subarachnoid space surrounding the brain and the spinal cord. Usually CSF is primarily absorbed through tiny, specialized cell clusters called arachnoid villi near the top and midline of the brain. The CSF then passes through the arachnoid villi into the superior sagittal sinus, v.jugularis internae and is absorbed into the bloodstream and filtered by the kidneys and liver and also absorbed into lymphatic channels. The anatomical structure of ventricular system reflect to the **fig. 1, 2.**

CSF has many functions, including a protective barrier against injury, nourishment, chemical messengers and transfer waste products away from surrounding tissues. Our body produce approximately 150 ml of CSF every day, continuously replacing it as it's absorbed. Normal conditions when exists a balance between the amount of CSF produced and the rate at which it is absorbed.

Demographics

Hydrocephalus affects a wide diapason of people, from infants and older children to young, middle-aged and older adults. On data of different authors (UK, USA) about 1–2 in every1000 babies is born with hydrocephalus [4, 5]. Over 1,000,000 people in the United States currently live with hydrocephalus. Hydrocephalus is the most common reason for brain surgery in children. It is estimated that more than 700,000 Americans have Normal Pressure Hydrocephalus (NPH), but less than 20 % receive an appropriate diagnosis [1]. The prevalence of congenital hydrocephalus in Russia and Ukraine on data different references from 1 to 4 per 1,000 live births [6–8]. In developed countries, the incidence of neonatal hydrocephalus ranges from 3 to 5 cases per 1000 live births, with a male predominance, but little is known about the frequency of hydrocephalus in Africa. In Mozambique, there is no primary information related to this disorder, but using the above data, the

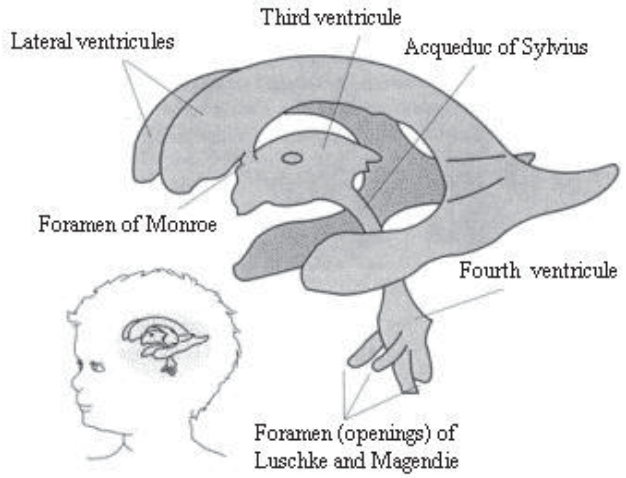


Fig. 1. CSF circulation system (from http://www.hydroassoc.org/)

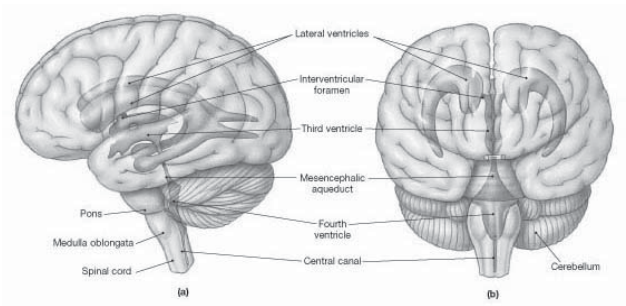
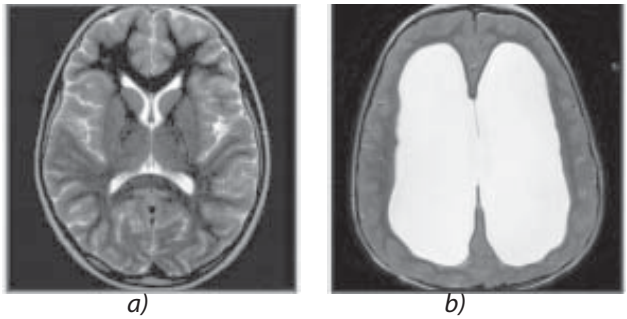


Fig. 2. Ventricles of the brain (a)-lateral view; (b)-anterior view (from thesalience.wordpress.com)

expected incidence of neonatal hydrocephalus would range from 2900 to 4800 cases per year [9]. In the MRI-pictures (**fig. 3**) present normal a) and enlarged ventricles b) with hydrocephalus.

In the **fig. 4** represent clinical case baby 10 months with hydrocephalic syndrome (saphenous veins of the scalp, sparse scalp, tonic gaze deviation down, the retraction of the upper eyelid syndrome («sunset»).

Hydrocephalus may exist as an isolated congenital condition or it may be a common accompaniment to other congenital anomalies such as spina bifida, meningocele, myelomeningocele. Babies with different localization of meningocele produce **(fig. 5)**.



a) *b)*

Fig. 3. MRI-pictures normal a) and enlarged ventricles b) with hydrocephalus (from www.hydroassoc.org)

When cases of spina bifida are included, congenital hydrocephalus occurs in two to five births per 1,000 births [1, 4, 5]. In the case of spina bifida aperta (myelomeningocele), hydrocephalus is present in 85–90 % of the cases. Hydrocephalus may also arise secondary to intraventricular hemorrhage in the premature neonate, infection (e. g. bacterial meningitis), head trauma, or a brain tumor. With appropriate and timely treatment, hydrocephalus is completely relieved, and children in which hydrocephalus is an isolated problem can be normal. When left untreated, the results are devastating [10]. The incidence of acquired hydrocephalus in adults is not known because it occurs as a result of injury, illness, or environmental factors. Normal pressure hydrocephalus was found to be significantly more prevalent in males and can occur in adults of any age group. [1, 4, 5].

Clinic and classification of hydrocephalus

Traditionally, hydrocephalus has been described as a disease characterized by increased intracranial pressure (ICP) and CSF volume, dilatation of the CSF spaces known as cerebral ventricles. Hydrocephalus is an

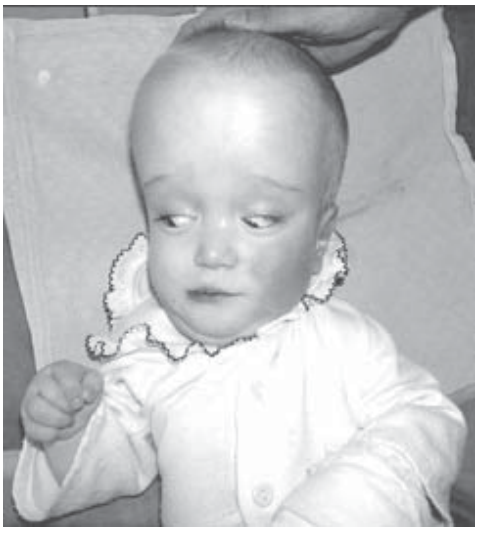
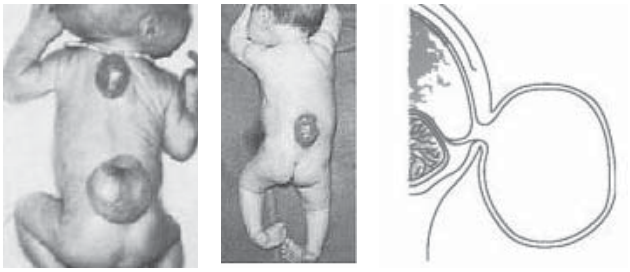


Fig. 4. Hydrocephalic syndrome [8]



a) *b)* *c)*

Fig. 5. Different localization of meningocele a), b), c-scheme of meningocele (contain meninges and CSF) (from A. S. Petruxin, 2009)

abnormal accumulation of cerebrospinal fluid (CSF) within cavities in the brain called ventricles. It occurs when there is an imbalance between the amount of CSF that is produced in the ventricles and in the choroid plexus and the rate at which it is absorbed. Hydrocephalus is defined as abnormal dilatation of the ventricular system. It occurs because of disturbances of CSF circulation and/or resorption [1, 11]. The symptoms of untreated hydrocephalus vary. During pregnancy, routine ultrasound can detect enlarged ventricles or spaces within a baby's brain. In an infant, the typical an abnormal enlargement of the baby's head, which will most likely be noticed at birth or within the first 9 months of life. Other symptoms in an infant may include irritability, excessive sleeping, vomiting, and poor feeding. In children symptoms relating with high pressure include nausea, vomiting, headache and vision problems. In young and middle aged adults typical symptoms of dizziness and vision problems. In older adults with NPH accomplish the symptoms loss of function in three main areas: walking, thinking and bladder control. Infants and children symptoms are: abnormal head enlargement, tense, bulging fontanel, prominent scalp veins, skull bones may feel separated, vomiting, sleepiness, irritability, headache, nausea, vomiting, vision disturbance, downward deviation of eyes. Young and middle aged adults has such symptoms as chronic headaches, difficulty walking/gait disturbances, cognitive challenges or complaints, urinary urgency or incontinence. Older adults (NPH) has: difficulty walking/gait disturbances, cognitive challenges/mild dementia, urinary urgency or incontinence [1, 4, 12]. Neurological examination revealed pyramidal insufficiencies or paralysis, pathology of cranial nerves, cerebellar ataxia, speech disturbance, neuropsychological and emotional disorder and symptomatic epilepsy. Hydrocephalus due to a blockage of the CSF pathway within the ventricular system is called noncommunicating or obstructive hydrocephalus. Hydrocephalus due to impaired CSF resorption at the arachnoid villi is called communicating or malresorptive hydrocephalus (sometimes this type of hydrocephalus corrects itself). Both non-communicating and communicating hydrocephalus can be either «congenital» (existing before or at birth) or «acquired» (developing after birth due to trauma, infections such as meningitis, bleeding, injury, or a tumor) [1, 4]. Acute hydrocephalus is characterized by ventricular dilatation with acute intracranial hypertension (after subarachnoid hemorrhage, obstructive process posterior fossa cranial, tumor).

Hydrocephalus with normal ICP and without progression of ventricular dilatation is called arrested, compensated or chronic hydrocephalus. Normal-pressure hydrocephalus usually develops in people who are age 55 or older. It is a potentially treatable cause of dementia. This type of hydrocephalus often occurs after head trauma, infections, and bleeding within the brain. Sometimes the cause of NPH is known – but most often it is idiopathic, which means the cause is not known.

Clinic NPH

The drainage of CSF is blocked gradually, and the excess fluid builds up slowly. The slow enlargement of the ventricles means that the fluid pressure in the brain may not be as high as in other types of hydrocephalus.

The symptoms NPH

Very often disease affect the legs, the bladder, and the «cognitive» mental processes, causing disorder of memory, reasoning, problem solving, and speaking. This decline of mental processes, if it is severe enough to interfere with everyday activities, remind dementia. It is memory loss, speech problems, apathy (indifference) and withdrawal, changes in behavior or mood, difficulties with reasoning, paying attention, or judgment.

Other symptoms include abnormal gait (difficulty walking, unsteadiness, leg weakness, sudden falls, shuffling steps, difficulty taking the first step, as if feet were stuck to the floor, «Getting stuck» or «freezing» while walking) and urinary incontinence, frequent urination, urgency to urinate, inability to control the bowels [13].

These symptom of NPH can be similar to those of Alzheimer disease, the walking problems are similar to those of Parkinson disease. Experts believe that many cases of NPH are misdiagnosed as one of these diseases. At first, the symptoms in NPH are usually very subtle. They worsen very gradually. The symptoms connected with increasing ICP are: headache, nausea, difficulty focusing eyes [13]. For diagnostic used as usually MRI, CT (**fig. 6**). T2-weighted MRI showing dilatation of ventricles out of proportion to sulcal atrophy in a patient with normal pressure hydrocephalus.

CT head scan of a patient with normal pressure hydrocephalus showing dilated ventricles (**fig. 7**).

Making the distinction is very important because the treatments for these conditions are quite different. Normal pressure hydrocephalus generally cannot be cured. It is a long-term condition. However, many people with the condition obtain substantial relief through surgical treatment. The treatment in these

cases is a shunt operation. Early diagnostic and treatment all types of hydrocephalus are important to minimize or prevent long-term problems.

Ex-vacuo hydrocephalus(ventriculomegalia), which occurs when there is damage to the brain caused by stroke or traumatic injury. This type of hydrocephalus may not be a health danger for some people, in which case treatment is not needed.

External hydrocephalus is a dilatation of the subarachnoid space enlargement of them but no more than mild enlargement of the ventricles. Internal hydrocephalus connected with enlargement of the ventricles [1, 11]. In very rare cases the brain tissue makes too much CSF and the body can't properly absorb this fluid and severe damage if not treated. This is called overproduction hydrocephalus. When CSF production and absorption are in balance, hydrocephalus is considered «compensated»; when out of balance, complications associated with elevated pressure or overdrainage occur – causing the signs of a malfunctioning shunt. Congenital hydrocephalus may result from either genetic or other causes, such as prenatal hemorrhage or trauma during fetal development, also mother`s infections such as toxoplasmosis, rubella, syphilis, mumps or influence alcohol, many drugs and medicines, some metals, such as lead or mercury, radiation [4]. It may be associated with other birth defects, that affect the spine, especially neural tube defects (NTD), which formed within the first 25 to 28 days of pregnancy. NTD may be prevented if a woman takes folic acid before becoming pregnant and during the first 6 weeks of pregnancy. But often a woman does not know she is pregnant until and after the first 6 weeks of pregnancy [1, 9]. Folic acid also is a useful as nutrient component in some green, leafy vegetables, nuts, beans, citrus fruits and fortified cereals that can help reduce the risk of NTD. If it necessary for woman childbearing age doctor admit appropriate amounts of folic acid [14, 15]. Anomaly of development often accomplish hydrocephalus and may be predictors of this diseases. Among them are spina bifida, meningocele, myelomeningocele, Chiari II malformation and others. Spina Bifida is caused by the failure of the neural tube to develop correctly. Related defects are anencephaly (the absence of a brain) and encephalocele (a malformation of the brain and skull), hydrocephalus and others.

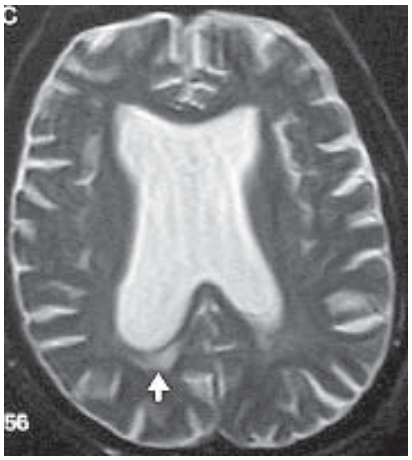


Fig. 6. MRI of patient with NPH.
The arrow points to transependymal flow.
(from http://www.emedicinehealth.com/normal_pressure_hydrocephalus/page14_em.htm#multimedia)

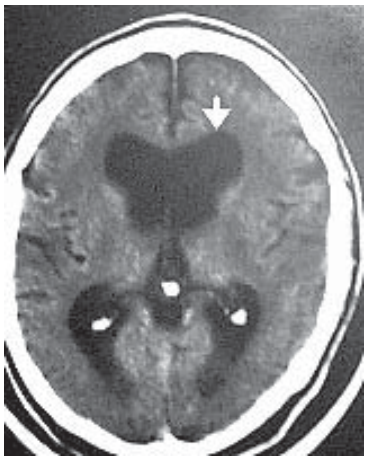


Fig. 7. CT of patient with NPH.
The arrow points to a rounded frontal horn
(from http://www.emedicinehealth.com/normal_pressure_hydrocephalus/page14_em.htm#multimedia)

Spina bifida occurs within the first four weeks of pregnancy. Infants born with spina bifida may have an open lesion on their spine where significant damage to the nerves and spinal cord occurs. Although the spinal opening is surgically repaired shortly after birth, the nerve damage is permanent. This results in varying degrees of paralysis of the lower limbs, depending largely on the location and severity of the lesion. Even without visible lesion, there may be improperly formed or absent vertebrae, and accompanying nerve damage [16]. Each year, there are an estimated 200,000 new cases of infant hydrocephalus in sub-Saharan Africa alone, and 100,000 neural tube defects in India alone. [14, 15] As other causes of death and disability precede, data suggest that spina bifida and hydrocephalus are gaining a larger share of mortality in young children [9]. In Uganda, he's documented that 60 percent of pediatric hydrocephalus cases from infections of neonates [15]. Spina bifida occurs in three major types, differing each others: spina bifida occulta, meningocele, myelomeningocele(spina bifida aperta). Spina bifida occulta (hidden), the mildest form, in which there is no obvious sign

of a malformation. The spinal cord does not protrude through the skin, there may be no obvious signs or symptoms. In many cases, this anomaly isn`t detected until an X-ray is done for another case. However, there may be a small, hairy patch, dimple or birthmark on the lower spine.

Meningocele. In this case the meninges protrude through a defect in the spinal column. The spinal cord typically remains intact, which means that these meninges can be removed surgically with little risk of nerve damage.

Myelomeningocele. There are a sac-like mass of nerves and spinal cord that protrudes from the back. This mass consists of meninges, portions of the spinal cord, and nerves. At birth, babies with myelomeningocele have an open spinal defect which requires urgent surgery to close the skin and protect the spinal cord and nerves. Such children are often fully or partially paralyzed below the defect and usually have difficulty with bladder and bowel control (incontinent). Nerve damage and other severe neurological impairments are also common. Children with spina bifida may have associated health problems, including hydrocephalus (occurs in up to 90 percent of children with myelomeningocele), Chiari II malformation, orthopedic (bone) problems (imbalance of muscle strength, scoliosis, kyphosis, hip dislocation, joint deformities, clubfoot, others) [12, 14, 15].

Chiari I malformation occurs in around 3 percent of the general population. Usually individuals with this condition often don't show symptoms and go undiagnosed, haven`t signs or symptoms and are only diagnosed when they get a magnetic resonance imaging (MRI) scan for another problem. Chiari I malformation is characterised by inferior herniation of the cerebellar tonsils through the foramen magnum. Chiari I needs to be distinguished from tonsillar ectopia, which is an asymptomatic and incidental finding in normal individuals, whereby the tonsils protrude through the foramen magnum by no more than 3-5 mm. If the degree of downward descent of the tonsils more than 5 mm - it is Chiari 1 malformation, between 5 and 10 mm malformation as a rule asymptomatic, if the patients have greater than 12 mm of descent of the tonsils appear pathological symptomatic, sometime forming syrinx [17, 18].

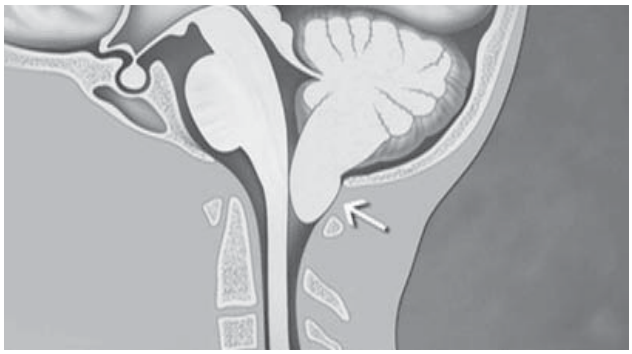
Chiari II malformation: an abnormality that occurs when part of the cerebellum and the fourth ventricle to push downward through the opening at the base of the skull into the spinal cord area, blocking CSF flow out of the fourth ventricle and causing hydrocephalus. This condition lead to such symptoms as vocal cord weakness, swallowing disturbances, leg weakness and spinal deformities, usually accomplish spina bifida. Chiari II malformations meet commonly with an incidence of ~1:1000 live births. When a child is born with a myelomeningocele the vast majority (~95%) have an associated Chiari II malformation [17–19].

Next illustration shows the cerebellar tonsils descending from the skull toward the spinal column, creating pressure (**fig. 8 a), b)**). [20].

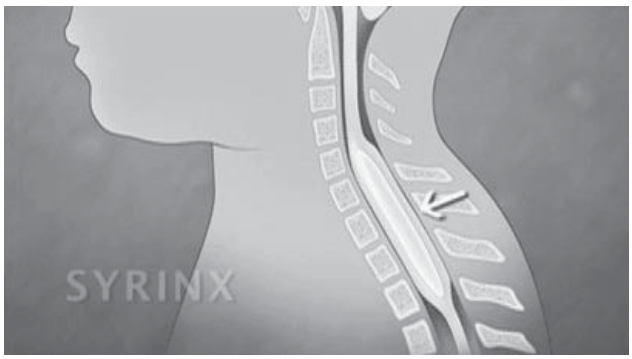
These MRI scans **(fig. 9)** on the right shows that the cerebellum has returned to a normal position, and the red arrows show how the CSF surrounding the cerebellum has returned to normal.

Aqueductal stenosis is the most common cause of congenital hydrocephalus caused by obstruction. The aqueduct of Sylvius, long, narrow passageway between the third and fourth ventricles is narrowed or blocked in connection with infection, hemorrhage, or a tumor. Fluid accumulates «upstream» from the obstruction, producing hydrocephalus. [1, 4, 5, 11]. Magnetic resonance imaging (MRI) before operation [21] show the obstruction the aqueduct of Sylvius (**fig. 10**).

Arachnoid cysts – can also be caused



a)



b)

Fig. 8. a) Chiari malformations, arrow shows extend tonil of cerebellum (from http://weillcornellbrainandspine.org)

b) This illustration arrow shows a syrinx, which is a cyst in the spinal column (from http://weillcornellbrainandspine.org)

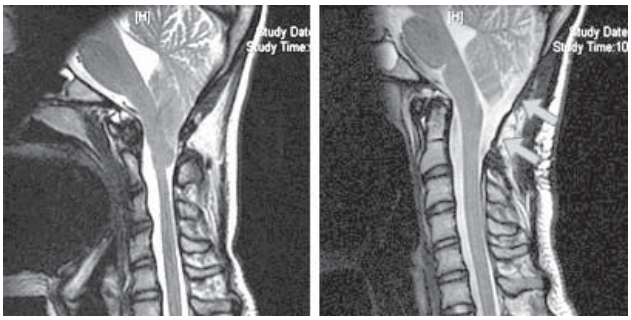


Fig. 9. MRI a patient before (left) and after (right) surgery for Chiari malformation (from http://weillcornellbrainandspine.org)

congenital hydrocephalus anyplace in the brain. In children, they're often located at the posterior fossa and in the area of the third ventricle. These cysts are filled with CSF and cover with the arachnoid membrane. Some arachnoid cysts are autonomous, while others are connected with the ventricles or the subarachnoid space and may block the CSF pathways, causing hydrocephalus [1, 4, 22]. **Fig. 11** reflect present arachnoid cysts of the brain.

Dandy-Walker syndrome – it is another cause of congenital hydrocephalus, the fourth ventricle becomes enlarged because its outlets are partly or completely closed and part of the cerebellum fails to develop. This syndrome may also be associated with abnormal development in other parts of the brain and sometimes leads to aqueductal stenosis. For treatment it case hydrocephalus use two shunts which placed in the child's ventricles – one in the lateral ventricle and another in the fourth ventricle. MRI picture Dandy-Walker malformation (**fig. 12 a), b), c)**) [23]. Show sagittal T1-weighted MRI scan in a 5-year-old girl shows a large posterior fossa cyst elevating the torcular Herophili and sinus rectus (short arrow). The hypoplastic vermis is everted over the posterior fossa cyst (long arrow). The cerebellar hemispheres and brainstem b) are hypoplastic. Thinned occipital squama is seen (arrowheads).

Diagnostic of Hydrocephalus include clinical exams and/or physical neurological

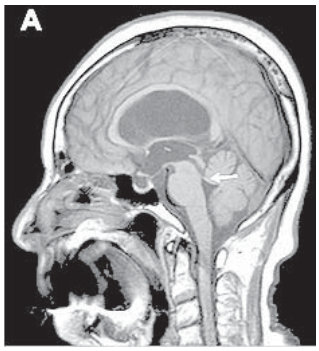


Fig. 10. Triventriculomegaly, empty sella and abrupt narrowing distal to the aqueduct (arrow) [21]

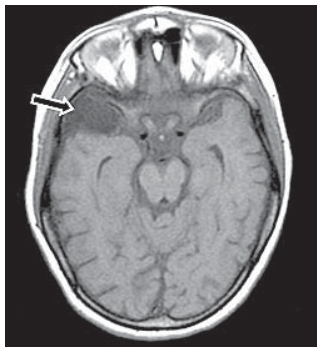
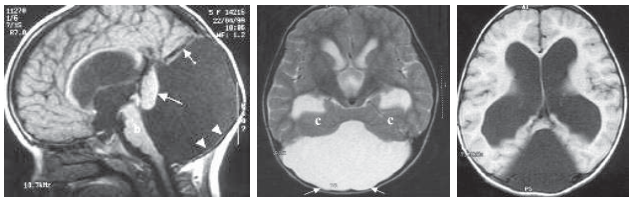


Fig. 11. MRI arachnoid cysts (arrow) [22]

examination, brain images by MRI and computerized tomography (CT) which usually detect enlarged ventricles. CSF tests to predict shunt responsiveness and/or determine shunt pressure include lumbar puncture, external lumbar drainage, measurement of CSF outflow resistance, ICP monitoring and isotopic cisternography. Prenatal diagnostic tests can also be performed during pregnancy to evaluate the fetus for spina bifida. These include:

Blood tests: The American College of Obstetricians and Gynecologists recommends a blood test on the alpha-fetoprotein (AFP) level and other biochemical markers in the mother's blood to determine whether her pregnancy is at increased risk for an open neural tube defect (ONTD) between 15 and 20 weeks of pregnancy for all women. Although the test can't diagnose an ONTD with 100 percent accuracy, it can predict which pregnancies are at greater risk, so that additional testing can be performed as the pregnancy progresses [14, 15].

Amniocentesis: a procedure that involves inserting a long, thin needle through the mother's abdomen into the amniotic sac to withdraw a small sample of the amniotic fluid for examination. The fluid is tested to determine the presence or absence of an ONTD like spina bifida. Although very reliable, this test may not



a) *b)* *c)*

Fig. 12. a) MRI picture Dandy-Walker malformation; b) MRI picture on axial T2-weighted MRI scan shows hydro-cephalus, a large CSF cyst in the posterior fossa, thinned occipital bone (arrows), and hypoplastic cerebellar hemispheres with a winged appearance (c); MRI picture Dandy-Walker malformation; c) MRI picture on axial T1-weighted MRI scan showing ventric-ulomegaly and a superiorly displaced posterior fossa cyst; MRI picture Dandy-Walker malformation

revealed small or closed defects.

Prenatal ultrasound (sonography): a diagnostic imaging technique that uses high-frequency sound waves and a computer to create images of blood vessels, tissues and organs and made detected the spinal defect on the ultrasound study, examine the fetus' other organs and body systems.

Isotopic cisternography

At the beginning of the study performed a lumbar puncture for CSF sampling for analysis and the parallel introduction of radiopaque substance or radioisotope pharmaceutical preparation. Then make certain intervals X-rays or take readings results of sensors, showing the accumulation of isotopes [24].

Treatment anomalies of development.

In most cases, children born with spina bifida occulta do not need treatment. In cases of meningocele and myelomeningocele, treatment depends on the type of spina bifida and its severity. A baby who besides these anomalies also has hydrocephalus will need an operation to relieve the pressure on the brain. This can be done using combined endoscopic third ventriculostomy/choroid plexus cauterization (ETV/CPC).

Other babies with hydrocephalus may require shunt placement, a process in which a small tube is implanted while the child is under anesthesia. This provides continual internal drainage of fluid from the spaces within the brain ventricles.

Common Causes of acquired Hydrocephalus include intraventricular hemorrhage, which the most frequently affects premature newborns and may block or scar the ventricles and the arachnoid villi and cause an acquired form of hydrocephalus because CSF can't be absorbed.

Meningitis caused inflammation of the meninges, disorder the CSF pathway. An acquired form of hydrocephalus may develop if has scarring obstructs the flow of CSF through the narrow ventricles or over the surfaces of the brain in the subarachnoid space.

Head injury can damage the brain's tissues, nerves, or blood vessels and cause the inflammation. Sites of CSF absorption might then be blocked by scarred meninges or by blood cells. The CSF flow is restricted and hydrocephalus develops.

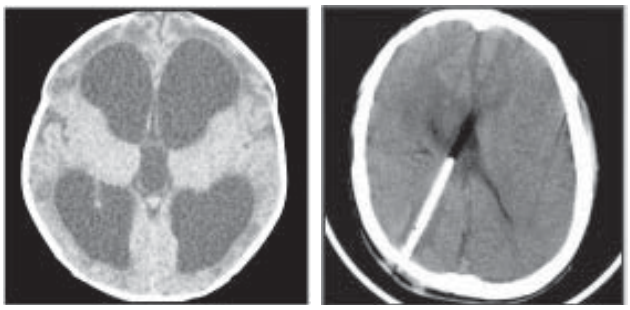
Brain tumor of the children most commonly occur in the posterior fossa. As a tumor grows, it may fill or compress the ventricular system, fourth ventricle, blocking the flow of CSF and causing hydrocephalus.

Treatment of hydrocephalus

Modern technology help in the diagnostic and treatment people with hydrocephalus to lead active lives. There are three forms of surgical treatment currently used to manage hydrocephalus. It is the most common treatment for hydrocephalus. The first type. A shunt is a flexible tube placed into the ventricular system of the brain which diverts the flow of CSF into another region of the body, most often the abdominal cavity, where it can be absorbed. A valve within the shunt maintains CSF at normal pressure within the ventricles. In the **fig. 13** show CN patients with hydrocephalus (enlarged ventricles, a)) and ventricles after shunt placement в).

A second type of treatment option for hydrocephalus is a surgical procedure called endoscopic third ventriculostomy (ETV). This same ETV procedure with the addition of choroid plexus cauterization is available for infants. In the ETV procedure, an endoscope is used to puncture a membrane in the floor of the third ventricle creating a pathway for CSF flow within the cavities in the brain. This approach is an important alternative to shunting for obstructive hydrocephalus and may be useful in other cases as well. This technique is effective for infants, in whom ETV alone is not as successful as in older children. CPC reduces the rate of fluid production, while ETV restores the normal fluid circulation. MRI obtained in a patient treated with ETV for hydrocephalus due to aqueductal stenosis, revealing an open sylvian aqueduct (arrow) [25], (**fig. 14**).

The third type treatment option involves the addition of choroid plexus cauterization with endoscopic third ventriculostomy in infants. The neurosurgeon uses a device to burn or cauterize tissue from the choroid plexus. The success rate for ETV or ETV/CPC depends upon patient factors such as age, cause of hydrocephalus, and whether there is scarring in the fluid space below the floor of the third ventricle. For some patients, the chance for success of the ETV may be up to 90 %; however, for others, ETV – with the addition of CPC for infants – may not be recommended because the chances for success are sufficiently low. [12, 15]. This creates an alternative route for removal of CSF which is constantly produced within the brain and usually restores the physiological balance between CSF production, flow, and absorption when one or more of these functions has been impaired. Valves contained within the shunt pathway act like on-off switches, opening when the differential pressure (DP) – i. e., the pressure difference across the valve – exceeds the valve's opening pressure. Valves are either set to a fixed pressure or they can be adjustable from outside the body. Accessory devices may be added to the shunt to modify valve function;



a) *b)*

Fig. 13. CT patients with hydrocephalus before- a) and after- shunt operation; b) (from www.hydroassoc.org)

for example, to counter gravitational forces an anti-siphon device may be attached in-line with the valve to minimize over-drainage of CSF when a patient stands up. In addition, a bubble-like reservoir can provide external access to the shunt system for evaluation of CSF or measurements of pressure.

Scheme of operation present on the illustrations. (**fig. 15, 16**).

Next picture illustrate critical hydrocephalus baby of 5 months and regress size of ventricles after shunt`s operation through 2, 2 year (**fig. 17**) [26].

Shunts typically consist of three major components.

An inflow (proximal or closer to the inflow site) catheter, which drains CSF from the ventricles or the subarachnoid space; this tube leaves the brain through a small hole in the skull and then runs for a short distance under the skin. A valve mechanism, which regulates differential pressure or controls flow through the shunt tubing; this device is connected to the proximal catheter and lies between the skin and the skull, usually on top of the head or just behind the ear. An outflow (distal or farther away from the inflow site) catheter, which runs under the skin and directs CSF from the valve to the abdominal (or peritoneal) cavity, heart or other suitable drainage site. Other shunt components include reservuar (chambers) for CSF sampling or inject the medications or dyes, on/off devices, anti-siphon or other flow-compensating devices,

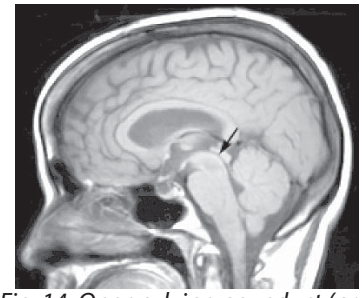


Fig. 14. Open sylvian aqueduct (arrow) after ETV due to aqueductal stenosis [25]

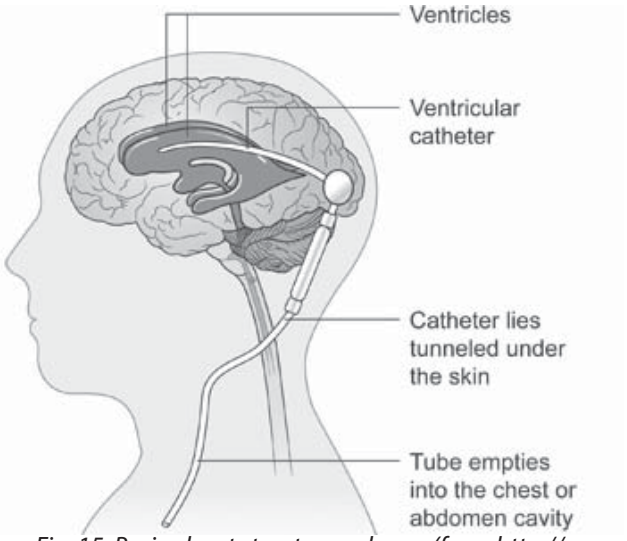


Fig. 15. Brain shunt structure, scheme (from http://commons.wikimedia.org)

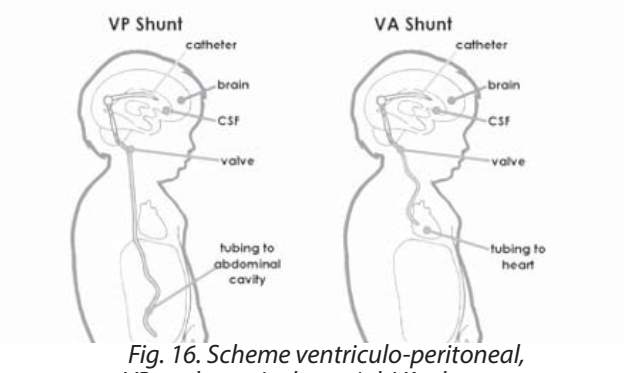
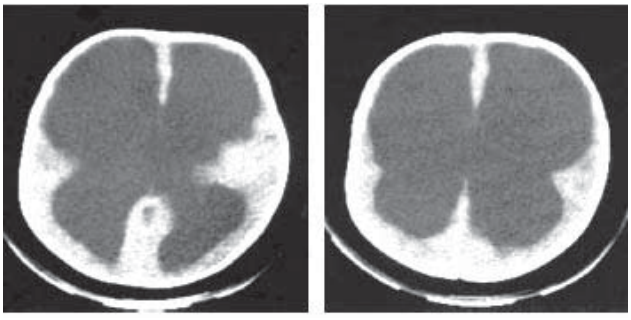
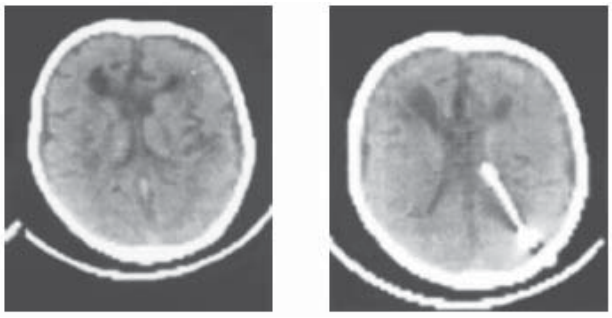


Fig. 16. Scheme ventriculo-peritoneal, VP, and ventriculo- atrial, VA, shunts (from http://pforpeanuts.com/)



a)



b)

Fig. 17. a) critical hydrocephalus A. Before shunt`s operation [26]; b) after shunt`s operation (normal size the brain ventricle, shadow of shunt in the right ventricle) [26]

or accessory catheters to modify performance or adapt the basic system to the patient's specialized needs. The proximal catheter (ventricular or lumbar catheter) drains excess CSF from the ventricles or the spinal lumbar sac through rows of small holes at its origin. Distal catheters are typically placed in the abdominal (or peritoneal) cavity, but may also be placed in the heart, pleural cavity (lungs) and other suitable locations where CSF is drained into the bloodstream. The type of shunt system is named by the inflow and outflow locations, e. g. if the proximal catheter is in the ventricle and the distal catheter is in the peritoneal cavity it is called a ventriculo-peritoneal (VP) shunt. Also exist ventriculo-atrial (VA), ventriculo-pleural (VPL), lumbo-peritoneal (LP) shunts. At times temporary CSF drainage is performed before a full shunt system is implanted; these short-term drainage systems are called external drains (ventricular or spinal) because the distal catheter is open or drains into a bag outside the body. Shunt tubing is made of flexible silicone, with short plastic tubes used at times as connectors to the valve mechanisms. Some shunt tubing is impregnated with antibiotics to reduce the incidence of infection during the post-operative period; examples include the Codman Bactiseal™ catheter and the Medtronic Ares™ catheter. Most valves operate on the principles of change in differential pressure (DP) – the difference between the pressure at the proximal catheter tip and the pressure at the drainage end. Neurosurgeons select a DP valve based upon the age of the patient, the size of the ventricles, the amount of pressure, and other important clinical factors. A number of newer shunts can be adjusted non-invasively (i. e. the DP is changed magnetically from outside the body), while others have self-adjusting flow-regulating mechanisms. When such valves are used, a second surgical operation is avoided as the valve's operating characteristics may be changed non-invasively (programmable valves) or adjusted automatically (flow-regulated valves) [27–29].

Most commercially available fixed DP shunts are provided in three to five ranges: low, medium or high pressures (and very low and very high), depending on their response to the pressure differential between the shunt's upper and lower ends [29].

The following illustration shows examples of shunt valves (**fig. 18**), designed by companies Aesculap and Miethke which have been working together since 1999 to develop better solutions for the complex treatment of hydrocephalus [30].

Managing a hydrocephalus patient's condition can be very challenging. One very important aspect is effectively maintaining a patient's CSF drainage to keep ICP within normal physiological ranges. CSF tends to drain faster when a patient is standing, causing a decrease in ICP and in results appear headaches, nausea and other debilitating symptoms. This necessitates the need for higher valve resistance in upright positions. The taller patient, the higher the necessary resistance. When the pressure setting is increased in a conventional regulable valve system, resistance is increased for all positions, not just the upright position. This results in increased ICP in the lying position causing headaches, nausea and other symptoms. Miethke gravitational valves allow surgeons to provide distinctly different pressures for lying and standing positions, reducing the symptoms that are caused by over and under drainage and providing better patient comfort. Miethke valves don't just function differently, they also look different because they are made using titanium. Titanium was selected because of its excellent MRI and bio-compatibility. The strength of titanium also allows the valves to be made extremely small, but with large flow paths. The rigid housing also makes the valves insusceptible to subcutaneous pressure [31].

Such way, ideal shunt pressure-flow characteristics must match the patient's specific needs. Exist devices with hydrostatic regulated mechanisms (gravitational devices), flow-regulating devices. Last valves have a lower incidence of early obstruction; the time to revision appears to be longer than that with conventional differential pressure valves [27]. CSF shunts using for treatment of hydrocephalus in the case of imbalance can lead to elevated ICP and variety of complications. Treatment with a shunt system often includes complications such as malfunction and infection. An estimated

Fig. 18. Shunts for treatment of Hydrocephalus (from https://www.aesculapusa.com/products/neurosurgery/hydrocephalus-shunts)

50 % of shunts in the pediatric population fail within two years of placement and repeated neurosurgical operations are often required [29].

Shunt Malfunction

Shunt malfunction is a partial or complete blockage of the shunt lead to CSF accumulates and can result in symptoms of untreated hydrocephalus. A shunt blockage from blood cells, tissue or bacteria can occur in any part of the shunt. Both the ventricular catheter and the distal part of the catheter can become blocked by tissue from the choroid plexus or ventricles. The distal part of the catheter is more often blocked in adults. Shunts are very durable, but their components can become disengaged or fractured as a result of wear or as a child grows. Sometimes they dislodge from where they were originally placed. More rarely, a valve will fail because of a mechanical malfunction. The most common infection is Staphylococcus Epidermidis, which is most likely seen one to three months after surgery, but can occur up to six months after the placement of a shunt. People with ventriculoperitoneal shunts are at risk of developing a shunt infection secondary to abdominal infection. Those patients treated with ventriculoatrial shunts may develop generalized infection, which can quickly become serious. Over drainage causes the ventricles to decrease in size creating slit-like ventricles as a result of the brain and its meninges pulling away from the skull. Slit-like ventricles sometimes called slit-ventricle syndrome (SVS), are most commonly a problem in young adults who have been shunted since childhood. A particular symptom of SVS is severe intermittent headaches that are often relieved when lying down. Imaging studies help to determine SVS, which is typically indicated by smaller than normal ventricles. Under drainage causes the ventricles to increase in size and can fail to relieve the symptoms of hydrocephalus. To restore a balanced flow of CSF it may be necessary to place a new shunt with a more accurate pressure valve. For those who have externally adjustable or programmable valves, the balance of flow can be restored by re-setting the opening pressure. Subdural hematoma is most common in older adults with NPH and requires surgery to correct. Multiloculated hydrocephalus is an isolated CSF compartment in the ventricular system that is enlarged and not in communication with the normal ventricle. It may be caused by birth trauma, neonatal intraventricular hemorrhage, ventriculitis, shunt related infection, over drainage or other conditions. This complication may be difficult to identify because it is typically seen in infants and children who may be neurologically compromised. Surgical treatments include multiple shunt placement, ventricular catheters with multiple perforations or openings, craniotomy and fenestration (opening) of the interventricular loculations.

Seizures sometimes present among people with hydrocephalus. There is no correlation between the number of shunt revisions or the site of shunt placement and an increased risk of developing seizures. Past studies [27] have shown that children with hydrocephalus who have been treated with a shunt and who also have significant cognitive delay or motor disorder are more often to have seizures than those without cognitive or motor delays. Studies also indicated that most likely explanation of seizure disorder is the presence of associated malformations of the cerebral cortex. Abdominal complications can occur in people with hydrocephalus treated with a shunt. The peritoneum or abdominal area is the most popular site for distal catheter implantation. Although VP – shunts do not have fewer complications than VA – shunts, the complications are less severe and have a lower mortality rate. Shunt complications that develop in the peritoneum or abdominal area include peritoneal pseudocysts, lost distal catheters, bowel perforations and hernias. The most common complication with these two procedures is closure of the pathway that is surgically created with ETV and infection. Sudden closure of the pathway created using Endoscopic Third Ventriculostomy can be sudden and life-threatening. Pathway closure occurs in 20 to 50 % of patients within five years of the procedure, especially failures occurring within the first six months of the operation. Infection with ETV and ETV/CPC occurs in up to less than one percent of patients who was treated. Fever and Bleeding with ETV and ETV/CPC may be as result of damage of ventricular walls or perforation of the basilar artery. Large bleeds due to vessel injury under the third ventricle can be catastrophic, but they are rare. Other complications from ETV include short-term memory loss, if the procedure may affect the hypothalamus and the areas of the mammillary body which are responsible for memory. Endocrinologic irregularities can occur following ETV and ETV/CPC as a result of the small opening in the area of the third ventricle which is responsible for some hormonal function. This complication is also often short lived. Symptoms of shunt malfunction or ETV closure vary considerably from person to person. When an abrupt malfunction occurs, symptoms can develop very rapidly potentially leading to coma and possibly death. In infants

and toddlers, it's important to be aware that medication with a side effect of drowsiness can mimic or mask signs of shunt malfunction or ETV closure and should be used with caution in those with hydrocephalus, especially infants and young children.

Prognosis

Untreated hydrocephalus has a survival rate of 40–50%, with the survivors having varying degrees of intellectual, physical, and neurological disabilities. Prognosis for treated hydrocephalus varies, depending on the cause. If the child survives for one year, more than 80 % will have a fairly normal lifetime. Approximately one-third will have normal intellectual function, but neurological difficulties may persist. Hydrocephalus not associated with infection has the best prognosis, and hydrocephalus caused by tumors has a very poor prognosis. About 50 % of all children who receive appropriate treatment and follow up will develop IQs in the near-normal or normal range [5, 32]. Early diagnostic and qualified in time treatment can improve the outcomes and prognosis of these patients.

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ГИДРОЦЕФАЛИЯ ОТ ПРОШЛОГО К НАСТОЯЩЕМУ

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Аннотация. Изучение и лечение гидроцефалии всегда интересовало врачей. Понимание причин возникновения и развития этого заболевания, а также сопутствующих ему аномалий нервной системы помогло создать систему адекватного лечения. Нелеченый энцефалит в 40-50% случаев приводит к развитию соматических, неврологических или психических расстройств. Прогноз пролеченного энцефалита зависит от его этиологии. Ранняя диагностика и квалифицированное лечение могут улучшить результаты и прогноз этих пациентов.

Ключевые слова: гидроцефалия, аномалии развития нервной системы, спинномозговая жидкость, желудочки мозга, шунтирующая система.

ГІДРОЦЕФАЛІЯ ВІД МИНУЛОГО ДО СЬОГОДЕННЯ

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Анотація. Вивчення та лікування гідроцефалії увесь час цікавило лікарів. Розуміння причин виникнення та розвитку цього захворювання, а також супутніх йому аномалій нервової системи допомогло створити систему адекватного лікування. Енцефаліт, що не лікували, у 40-50% хворих призводить до розвинення соматичних, неврологічних або психічних розладів. Прогноз енцефаліту, що лікували, залежить від його етіології. Рання діагностика та кваліфіковане лікування можуть поліпшити результати і прогноз цих пацієнтів.

Ключові слова: гідроцефалія, аномалії розвитку нервової системи, спинномозкова рідина, шлуночки мозку, шунтуюча система.

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КЛІНІЧНІ ТА НЕЙРОВІЗУАЛІЗАЦІЙНІ ЗМІНИ ПРИ ЗЛОВЖИВАННІ ПРЕПАРАТАМИ ЕФЕДРИНОПОДІБНОЇ ДІЇ

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Анотація. Стаття присвячена проблемі вживання кустарних психостимуляторів, виготовлених з препаратів ефедриноподібної дії. За даними МРТ виявлено, що застосування цих речовин призводить до безповоротних змін у головному мозку. Дані клініко-неврологічного обстеження показали наявність у хворих синдрому паркінсонізму, пірамідних, мозочкових, окорухових і вегетативних порушень, а також зниження когнітивних функцій та депресивні розлади. Необхідна особлива уважність лікарів при виявленні згаданої вище симптоматики в осіб молодого віку відносно прийому наркотичних засобів з метою надання своєчасної допомоги та адекватної корекції порушень.

Ключові слова: психостимулятори, ефедриноподібні препарати, МРТ-зміни, неврологічні порушення, синдром паркінсонізму.

Вступ

В останні десятиліття наше суспільство буквально захлеснула хвиля наркоманії та токсикоманії, набувши характеру розгорнутого епідемічного процесу. З кожним роком кількість наркозалежних збільшується, вражаючи всі верстви населення незалежно від матеріального статку і соціального становища, завдаючи руйнівної дії на психічне та соматичне здоров'я і величезної економічної та моральної шкоди суспільству.