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Prevalence, Risk Factors and Demographic of Pediatric Hydrocephalus in RSUP H. Adam Malik Medan 2019-2020

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Abstract

Introduction: Hydrocephalus is a disorder of the central nervous system which manifests as excess cerebrospinal fluid found in the head either inside the ventricular system or the subarachnoid space. The prevalence and demographics of congenital hydrocephalus remain poorly defined, in part because the definition of "congenital hydrocephalus" varies between studies; depending on the clinical criteria used to define congenital hydrocephalus, the prevalence has been reported between 1 and 32 per 10,000 live births. Previously known several risk factors include infection during pregnancy (toxoplasmosis, CMV, meningitis), family history hydrocephalus (less than 2%), premature pregnancy and low birth weight (which may increase the incidence of intraventricular hemorrhage).

Method and Result : This Study was a descriptive study, we collected data from 2018-2019 at Haji Adam Malik General Hospital. From the data we found 58 cases of pediatric Hydrocephalus, with hydrocephalus obstructive 24 cases (41%) and communicans hydrocephalus 34 case (59%). Based on age, most samples were <6 months of age in both types of hydrocephalus. Most of the gender is male. Most of the GCS conditions at admission were GCS 13-15. The most common etiology in obstructive hydrocephalus cases was aquaduct stenosis in 13 cases (54%), while the most common etiology in cases of hydrocephalus communicans was infection in 22 cases (65%). Infection during pregnancy, low birth weight and premature pregnancy are thought to be the most risk factors found

Discussion : Hydrocephalus is described as non communicating when it is caused by lesions that obstruct CSF flow through the ventricular system, such as fourth ventricular tumors. Communicating hydrocephalus is present when the lesion is at the level of the subarachnoid spaces, arachnoid granulations, or the venous system. Previously known several risk factors include infection during pregnancy, family history hydrocephalus, premature pregnancy and low birth weight.

Keyword: communicans, hydrocephalus, obstructive

Introduction

Hydrocephalus is a disorder of the central nervous system which manifests as excess cerebrospinal fluid found in the head either inside the ventricular system or the subarachnoid space. The disorder causes the fluid to increase which will further suppress the surrounding brain tissue, especially vital nerve centers. The effect of hydrocephalus is an increase in intracranial pressure which can ultimately be life threatening.

The prevalence and demographics of congenital hydrocephalus remain poorly defined, in part because the definition of "congenital hydrocephalus" varies between studies; depending on the clinical criteria used to define congenital hydrocephalus, the prevalence has been reported between 1 and 32 per 10,000 live births. Some studies have suggested that the prevalence of congenital hydrocephalus increased between the 1960s and 1990s, possibly as a result of the increased survival of extremely preterm infants.

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Others have found that the prevalence of congenital hydrocephalus diminished in recent years. This more recent decline in diagnoses of congenital hydrocephalus has been thought to result from improved care of extremely preterm infants and the advent of folic acid enrichment of food, but it may also be due to an increasing rate of fetal termination after prenatal diagnosis of congenital hydrocephalus.

The incidence of hydrocephalus in Indonesia occurs between 0.2 – 4 out of 1000 births, which is found around 40 – 50% of the entire medical visits or neurosurgery. Risk factors associated with hydrocephalus in the fetus are still uncertain. Previously known several risk factors include infection during pregnancy (toxoplasmosis, CMV, meningitis), family history hydrocephalus (less than 2%), premature pregnancy and low birth weight (which may increase the incidence of intraventricular hemorrhage).[1] [2]

Cerebrospinal Fluid

The brain normally contains areas that are devoid of cells but are filled with CSF. These areas are collectively known as the ventricular system. From rostral to caudal the components of this system include the lateral ventricles, third ventricle, mesencephalic aqueduct, and fourth ventricle. Cerebrospinal fluid leaves the fourth ventricle through the lateral apertures and enters the subarachnoid space around the brain. The fourth ventricle is continued into the spinal cord via the central canal. The ventricular system is lined by specialized columnar cells with microvilli known as ependymal cells. These cells are important as a partial barrier between the CSF and the brain parenchyma.

Three primary compartments exist in the cranial cavity: the brain parenchyma, blood, and CSF. The blood brain barrier exists between the plasma and the extracellular fluid (ECF) and consists of nonfenestrated, tightly joined endothelial cells of the blood vessel surrounded by a relatively complete layer of astrocyte foot processes. The blood-CSF barrier exists at the choroid plexus and consists of two cell layers separated by a thin basement membrane. This is a semipermeable membrane between the CSF and the plasma. The CSF-ECF barrier occurs over the outer surface of the brain and in the ventricles. This barrier is formed primarily by the ependymal cells. On the surface of the brain, the pial-glial membrane also acts as a barrier ·between the CSF and the ECF.[3]

Classification of Hydrocephalus [4] [5]

Hydrocephalus has been classified as obstructive or nonobstructive, which is a somewhat misleading nomenclature because all forms of hydrocephalus involve some form of CSF obstruction under the bulk flow model. The only exception is hydrocephalus ex vacuo, which is not true hydrocephalus but rather ventriculomegaly caused by brain atrophy. A more commonly employed classification describes hydrocephalus as either communicating or noncommunicating. With modern neuroimaging, this test is rarely performed; hydrocephalus is now described as noncommunicating when it is caused by lesions that obstruct CSF flow through the ventricular system, such as fourth ventricular tumors. Communicating hydrocephalus is present when the lesion is at the level of the subarachnoid spaces, arachnoid granulations, or the venous system demonstrates several common sites of obstruction with common lesions that occur in those locations. Rackate has proposed a classification system that defines different forms of hydrocephalus by the site of anatomic obstruction: foramen of Monroe, aqueduct of Sylvius, outlets of the fourth ventricle, basal cisterns, arachnoid granulations, venous outflow, or none (eg, choroid plexus papilloma-related CSF overproduction).

Lateral Ventricles

Choroid plexus tumors compose 1.5% to 3.9% of pediatric brain tumors, an incidence that is three times higher than in the overall population.27- 29 They are particularly common in children under the age of 3 years, and the majority are benign choroid plexus papillomas. CSF production rates three to four times normal have been

described in children with these lesions, 30 and surgical resection cures the hydrocephalus in approximatdy two-thirds of cases. Choroid plexus tumors and other intraventricular masses can also cause monoventricular hydrocephalus by obstructing CSF flow within the lateral ventricle, causing CSF to become "trapped" in the temporal horn. Surgical resection of the tumor typically leads to resolution of the obstruction, but in some cases hydrocephalus can persist and require long-term treatment. These cases of persistent hydrocephalus are likely caused by inflammation and obstruction of the basal cisterns or arachnoid granulations by blood products released intraoperatively.

Radiographs demonstrate a massively enlarged choroid plexus in the trigones of the lateral ventricles, and there is usually concomitant ventriculomegaly. The condition can usually be treated by endoscopic cauterization or surgical removal of the choroid plexus.[31]

Obstruction at the level of the foramina of Monro can also cause hydrocephalus, although this is less common. The foramen may be congenitally occluded by a membrane or it may be stenotic because of atresia. Intraventricular hemorrhage (IVH) and ventriculitis may cause gliosis or neomembrane formation, leading to stenosis or occlusion of the foramen. Affected children may develop ipsilateral ventriculomegaly, which can result in asymmetrical enlargement of the cranial vault if left untreated (Fig. 8.4). Endoscopic fenestration of the septum pellucidum creates an alternative pathway for CSF drainage through the contralateral lateral ventricle and may adequately treat the condition in the absence of a shunt.

Third Ventricle

Cysts and neoplasms in the region of the third ventricle can cause noncommunicating hydrocephalus. In pediatric patients, ependymal and arachnoid cysts of the third ventricle may occur. These lesions have signal characteristics similar to CSF on radiographic studies, and high-contrast, high-resolution MRI sequences, such as constructive interference in steady state (CISS), may demonstrate a thin cyst wall. Endoscopic fenestration of the cyst is one treatment option, but shunt placement with one or more ventricular catheters positioned to drain the cyst and lateral ventricles may be required. 36• 37 Hypothalamic and optic pathway gliomas such as those commonly seen in neurofibromatosis type I (NF 1) may cause hydrocephalus if they enlarge sufficiently to impinge on normal CSF pathways. Patients with these lesions are frequently poor candidates for surgical resection because of the tumor location, and thus CSF diversion is generally required.

Colloid cysts are rare neoplasms located at the anteriorsuperior aspect of the third ventricle that occur more commonly in adults than in children, although they have been reported in children as young as 2 months.[38] These cystic structures are thought to be present at bitth and become symptomatic only when they grow large enough to occlude the foramina of Monro. They are typically treated by stereotactic aspiration of the cyst or, more commonly, by resection of the cyst via either an open craniotomy or an endoscopic approach.[39-42] Long-term placement of a CSF shunt is occasionally required. Hydrocephalus may also occur because of the upward extension of suprasellar masses such as craniopharyngiomas. Although patients with these lesions may need temporary CSF diversion in the form of an external ventricular drain, the hydrocephalus usually resolves with resection of the tumor.

Aqueduct of Sylvii

Obstruction at the level of the Sylvian aqueduct causes a characteristic "triventricular" hydrocephalus, with enlargement of the third and lateral ventricles. The foutth ventricle is generally normal in size (Fig. 8.5). In normal infants, the aqueduct is 12 to 13 mm in length and only 0.2 to 0.5 mm in diameter.[43] It is thus prone to occlusion, because of either intrinsic abnormalities of the aqueduct or extrinsic compression. Intrinsic aqueductal abnormalities include stenosis, forking, the presence of a septum, or

subependymal gliosis (Fig. 8.6). Of patients with obstruction here, only 4% to 8% will have true stenosis of the aqueduct.44 Subependymal gliosis is more common and may occur because of hemorrhage, intrauterine infections such as toxoplasmosis, or mumps encephalitis.[45] As many as 10% of males with isolated congenital hydrocephalus have a mutation in the x-linked gene LCAM1, which codes for a neural adhesion molecule.[46.47] In these children, the severity of the mutation appears to correlate with the degree of hydrocephalus.[47]

Extrinsic compression of the aqueduct is typically the result of a neoplastic process. Low-grade glial tumors of the tectal plate can expand the btain parenchyma surrounding the aqueduct, leading to stenosis or occlusion (Fig. 8.7). These neoplasms are more common in children with NF 1, and they frequendy follow a benign clinical course, provided the associated hydrocephalus can be treated.[48,49] Pineal region neoplasms occur more commonly in children than in adults,[50] and these tumors may compress the midbtain tegmentum, leading to occlusion of the aqueduct. Many of these tumors are radiosensitive, and the associated hydrocephalus frequently resolves if surgical resection or radiotherapy is successful in removing mass effect.

Fourth Ventricle

Neoplasms are a common cause of hydrocephalus resulting from obstruction at the fourth ventricle. In children, approximately one-quarter of all btain tumors are located in the posterior fossa,[51] where they have the potential to obstruct the CSF pathway through the fourth ventricle. Indeed, many of these patients initially present with hydrocephalus, and some require long-term treatment of hydrocephalus even after resection of the mass.

Basal Cisterns, Arachnoid Granulations, and the Venous System

Obstruction of CSF flow at the level of the basal cisterns or impaired absorption caused by pathology in the arachnoid granulations or venous system can cause communicating hydrocephalus. Common causes of communicating hydrocephalus in children include meningitis, subarachnoid hemorrhage, and trauma. Teenage girls on oral contraceptives have an elevated risk for venous sinus thrombosis, which has been reported to cause hydrocephalus.[59] In some young children, enlarged subarachnoid spaces may be seen over the convexity, a finding that has been called (benign) external hydrocephalus. These children occasionally have concomitant mild ventriculomegaly and may present with rapidly enlarging head circumference. In contrast to other causes of hydrocephalus, benign external hydrocephalus is selflimited in the majority of cases and usually resolves without intervention by the age of 2 years. Rarely, placement of a shunt may be required. The etiology of this uncommon condition is not dear but may be related to delayed maturation of the arachnoid granulations.

Case Report

This Study was a descriptive study, we collected data from 2018-2019 at Haji Adam Malik General Hospital. From the data we found 58 cases of pediatric Hydrocephalus, with hydrocephalus obstructive 24 cases (41%) and communicans hydrocephalus 34 case (59%). Based on age, most samples were <6 months of age in both types of hydrocephalus. Most of the gender is male. Most of the GCS conditions at admission were GCS 13-15. The most common etiology in obstructive hydrocephalus cases was aquaduct stenosis in 13 cases (54%), while the most common etiology in cases of hydrocephalus communicans was infection in 22 cases (65%). Infection during pregnancy, low birth weight and premature pregnancy are thought to be the most risk factors found.

Table 1. Type of Hydrocephalus

Type Hydrocephalus	Cases
Obstructive Hydrocephalus	24
Communicans Hydrocephalus	34
Total	58

Table 2. Demographc of sample

	Obstructive Hydrocephalus	Communicans Hydrocephalus
Age		
< 6 Months	16	8
6-12 Months	2	9
1-5 y.o	2	4
5-10 y.o	3	7
10-15 y.o	1	6
Gender		
Male	14	21
Female	10	13
GCS		
3-8	3	4
9-12	2	2
13-15	19	28
Total	24	34

Table 3. Etiology Of Obstructive Hydrocephalus

Hydrocephalus Obstructive Etiology	Cases
Aquaduct Stenosis	13
Dandy Walker Malformation	8
Tumor	3
Total	24

Table 4. Etiology of communicans Hydrocephalus

Hydrocephalus Communicans Etiology	Cases
Infection	22
Post Hemorraghic	12
Total	34

Table 5. Risk Factors Sample

Risk Factor	Obstructive Hydrocephalus	Communicans Hydrocephalus
Infection During Pregnancy	6	7
Family History of Hydrocephalus	2	-
Premature Pregnancy	5	6
Low Birth Weight	5	8
Maternal Diabetes	1	-
No Risk Factor Above	5	13
Total	24	34

Treatment [6] [7]

The principal goal in treating hydrocephalus is the prevention of neurologic decline. Although medications such as the carbonic anhydrase inhibitor acetazolamide have been used in the management of hydrocephalus, they are not effective in obviating the need for shunt placement.[67-68] Children with dangerously elevated I CP due to acute hydrocephalus or shunt failure can and should be temporized with hyperosmolar agents such as mannitol and hypetonic saline until definitive surgical management is performed.

Non-Surgical

Diuretics (frusemide and acetazolamide) and steroids are known to decrease CSF production. Diuretics are still often used in neonatal patients with post-haemorrhagic hydrocephalus (PHH) despite reports of side eVects such as acidosis, CO2 retention, electrolyte disturbance, etc. A recent multicentre randomised controlled trial actually showed a higher rate of shunt placement and increased neurological morbidity in the group receiving diuretics.

CSF Shunts

The most common type of shunt diverts CSF from the ventricles to the peritoneal cavity (ventriculo-peritoneal shunt [VPS]), although other distal sites such as the right atrium of the heart and the pleural cavity are occasionally used. Shunts generally consist of silastic tubing that runs subcutaneously from the head to the abdomen, with a valve between the ventricular and distal catheters. Differential pressure (with fixed or programmable settings) or flow-regulating valve mechanisms are often paired with antisiphon or gravitational devices to prevent CSF overdrainage from posture-related siphoning. However, despite technological progress, valve design seems to have little if any effect on shunt efficacy or failure rates.

External Ventricular Drain

An external ventricular drain (EVD) is a temporary catheter that diverts CSF from the ventricles to a bedside collection system. Typically, a burr hole is created through a small incision behind the hairline, and the catheter is passed through the brain and into the frontal horn of the lateral ventricle. The distal end of the catheter is tunneled out through the scalp and connected to a bedside collection system. EVDs can be placed rapidly at the bedside in emergent situations, or they may be inserted during surgery and left in place after the operation to drain debris and blood products from the ventricles to reduce the risk of shunt dependence. The amount of CSF drained can be controlled by raising or lowering the collection system. When the drain is no longer required, many neurosurgeons elect to "wean" the drain by progressively raising it and ultimately clamping it closed, although there is some evidence in adults that this stepwise approach to EVD removal is unnecessary. 69 If the patient tolerates closure of the EVD without recurrent symptoms of hydrocephalus or radiographic evidence of enlarging ventricles,

the drain is removed; if it is not, the hydrocephalus usually requires more definitive treatment.

Endoscopic Third Ventriculostomy And Choroid Plexus Cauterization

In the 1990s, endoscopic third ventriculostomy (ETV) emerged as an effective alternative treatment for hydrocephalus, particularly in patients with noncommunicating hydrocephalus,65 and is now routinely carried out at most major pediatric neurosurgical center in high-income countries. The procedure involves passing an endoscope into the frontal horn of the lateral ventricle, then through the foramen of Monro, and into the third ventricle. An opening is then made in the floor of the third ventricle, enabling direct communication into the prepontine cistern. Although ETV is successful in many patients, there is a high rate of early failure, particularly in infants.66 Beginning in the early 2000s, however, choroid plexus cauterisation (CPC) was added to ETV to improve efficacy of ETV alone in very young patients.[67]

In the early twentieth century results of small series in which CPC alone was used to treat hydrocephalus showed some success in patients with communicating hydrocephalus,68,69 but with the available techniques, mortality and morbidity were substantial, and any longterm collateral effects of CPC were, and remain, unknown. The modern use of CPC has mostly been in combination with ETV, especially in sub-Saharan Africa.67 According to the bulk flow model, ETV bypasses an obstruction and CPC reduces CSF production. In the hydrodynamic model, ETV acts to create a pulsation absorber and CPC reduces the intraventricular pulsation amplitude.61,70 As described, the ETV and CPC procedure involves use of a flexible endoscope to cauterise the entire choroid plexus throughout both lateral ventricles.

Discussion

Hydrocephalus is described as non communicating when it is caused by lesions that obstruct CSF flow through the ventricular system, such as fourth ventricular tumors. Communicating hydrocephalus is present when the lesion is at the level of the subarachnoid spaces, arachnoid granulations, or the venous system. Previously known several risk factors include infection during pregnancy, family history hydrocephalus, premature pregnancy and low birth weight.

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