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REVIEW ARTICLE

Antenatal Hydrocephalus : An Emerging Neurosurgical Venture- A Literature Review

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Abstract

Hydrocephalus is the term used to describe pathologic dilation of the brain's ventricular system due to increased CSF pressure. The term antenatal hydrocephalus denotes when ventricular dilatation measures >15 mm or an obstructive etiology is associated with increased CSF pressure during intrauterine period. Advances in fetal diagnostic techniques have opened up many areas to prenatal anatomical scrutiny. Intrauterine hydrocephalus and ventriculomegaly can be diagnosed by prenatal ultrasound between 15 and 35 weeks of gestation. This paper reviewed the current understanding of diagnosis and neurosurgical management of prenatal hydrocephalus.

Keywords: antenatal hydrocephalus, ventriculomegaly, neurosurgical management

Introduction

In the earlier decades, the medical professionals could not deal with the cases of neonatal hydrocephalus in ideal way. With the advancement of medical knowledge, recent neuroscientists has the endeavor to deal the cases of antenatal/fetal hydrocephalus in a better way. It's a combined effort of perinatologist, obstetrician, neurosurgeon and geneticist. Congenital hydrocephalus is caused by a complex of neurological disturbances that increase the amount of cerebrospinal fluid and the size of the ventricles of the brain and/or in the subarachnoid space [1]. A strong correlation exists between the biochemical mechanisms of the folate metabolism and the development of the nervous system. An abnormal folate concentration could be the cause of hydrocephalus [2]. According to Garne et al, the incidence of congenital hydrocephalus is 1–4.65 for every 10,000 newborns representing one third of all congenital abnormalities of the nervous system [3]. The causes of hydrocephalus vary with the age of onset of the disease, and the most common cause include congenital malformations, intraventricular hemorrhages, neoplasms, infections and others [4,5]. Advances in real time imaging techniques have enabled more frequent diagnoses of congenital anomalies of the central nervous system in the prenatal period. However, the diagnosis of prenatal hydrocephalus always involves some uncertainty since the fetus is in utero. Diagnosis can be established by clinical signs, supported by sonographic examination, computed tomography (CT) scan or magnetic resonance imaging (MRI). Performing serial accurate ultrasound examination, hydrocephalus can be monitored for therapeutic interventions and also counseling for the parents related to the baby's prognosis. Therefore, CT and MRI have an important role in both the diagnosis and the assessment of therapeutic efficacy and in the monitoring of the disease [6]. The prognosis of both prenatal hydrocephalus and fetal ventriculomegaly depends on several factors. The size of the ventriculomegaly, pathogenesis and the time of onset might influence outcome. The survival rate of overall fetal ventriculomegaly is 40–50%, while some cases being terminated at the request of the parents when the diagnosis is made in the early fetal period. The peri- and postnatal mortality rate is combined 10–20%. The developmental outcomes of fetal ventriculomegaly vary depending on the degree of ventriculomegaly and the presence of associated abnormalities with or without the central nervous system. Normal development is reportedly seen in 80–90% of cases of isolated mild ventriculomegaly. Even in postnatally treated prenatal hydrocephalus, normal development is expected in about 20% of cases [7].

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Historical background:

Hydrocephalus has been recognized for centuries. Accumulation of fluid in various intracranial compartments was recognized by Hippocrates (BC 460–377) and Claudies Galen (130–200 AD). The studies of Thomas Willis (1621–1675) facilitated the understanding of ventricular system and CSF pathways. Franciscus Sylvius (1614–1672), Alexander Monroe (1733–1817) and Francois Magendie (1783–1855) have made important anatomical contributions for the CSF pathway. Finally, Key and Retzeus (1876) established the modern concept of CSF circulation. The so-called fetal neurosurgery was not introduced until 3 years later when Barke et al. (1966) confirmed the diagnosis of fetal hydrocephalus through gas ventriculography. The first therapeutic neurosurgical procedures were carried out by Birnholz and Frigoletto (1981), who used repeated cephalocentesis in a hydrocephalic 25-week fetus, performing 6 ultrasound-guided transabdominal punctures , and by Clewel et al. (1982), who described the first ventriculoamniotic shunting of a 24-week-old fetus, which was effective until the 32nd week of gestation. Fetal neurosurgery did not advance until 1999 when Tulipan et al demonstrated a reversal of Chiari II malformation after the intrauterine correction of fetal myelomeningocele using open fetal surgery, when performed before the 26th week of gestation. In 2003, Cavalheiro et al performed the first fetal endoscopic third ventriculostomy for the treatment of hydrocephalus by aqueduct stenosis in a fetus at 26 weeks of gestation [8].

Pathogenesis:

Neuroscientists are engaged relentlessly to find out the cause of antenatal hydrocephalus. Understanding the pathology is very important to reach the target point of perfect remedy. Fetal Hydrocephalus is a complex multifactorial disease. Congenital and neonatal hydrocephalus can be caused by a wide variety of developmental abnormalities or insults, the primary culprits are neural tube defects, infection, intraventricular hemorrhage, trauma, and tumors. In children, this condition is especially damaging because the expanding ventricles accompanied by increasing CSF pressure, cause the flexible skull to enlarge; this in turn both compresses and stretches adjacent brain tissue. A relatively slow progression of ventriculomegaly over weeks and months may allow cellular plasticity to occur and thus promote intrinsic repair mechanisms. Likewise, preferential expansion of the occipital horns of the lateral ventricle may impact the optic radiations and thus cause selective visual deficits without involving locomotion and motor function. Ventriculomegaly becomes chronic and/or progresses to more severe forms: gliosis and neuroinflammation, periventricular edema, demyelination, axonal degeneration, slow axoplasmic transport, metabolic impairements, stagnant CSF flow, altered blood brain barrier, dendritic and synaptic deterioration resulting in altered connectivity, eventually cell death. The role of neuronal cell death in the overall pathophysiology of hydrocephalus is interesting because apoptosis and necrosis of cortical neurons seem to occur only after prolonged hydrocephalus. The oligodendrocytes appear to be vulnerable during early stages of hydrocephalus and undergo significant apoptosis in the priventricular white matter. Thus myeline formation in the developing hydrocephalic brain can be impeded by multiple simultaneous events: stretch, compression, interstitial edema, hypoxia and oligodendrocyte death [9] (Figure:1).

Investigations:

Advancement of technology is one of the most important armamentarium to have a concrete diagnosis. Recent investigating tools are well equipped to find out the exact pathology and to have a clean cut diagnosis in fetus. For evaluation of antenatal hydrocephalus following common investigations are done.

Comprehensive sonographic evaluation: it helps in diagnosing ventriculomegaly and determining other structural abnormalities.Identification of these structural abnormalities helps in determining the cause of ventriculomegaly and guide the prognosis. Additional imaging of the CNS to be performed. Such imaging would include a detailed evaluation of the lateral, third, and fourth ventricles, corpus callosum, thalami, germinal matrix region, and cerebellum and cerebellar vermis . The sonographic findings of congenital fetal infection are intracerebral and periventricular calcifications, hepatic calcifications, hepatosplenomegaly, ascites, and polyhydramnios. In addition, a detailed fetal sonographic anatomic survey should be performed to look for non-CNS abnormalities since ventriculomegaly can be a component of several genetic syndromes (eg, trisomy 21). Although some experts suggest a fetal echocardiogram to be performed routinely, usually it is not practiced if the heart appears normal on a detailed fetal sonogram.

Magnetic resonance imaging(MRI): Fetal magnetic resonance imaging (MRI) can be used to identify underlying CNS abnormalities not detected by sonography. It is suggested to consider MRI in cases of isolated ventriculomegaly (with a normal karyotype and unknown etiology) because cortical malformations and other potentially significant defects

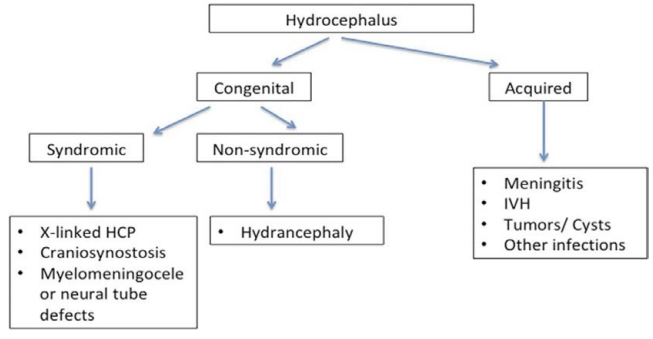

Figure1: Aeito- pathogenesis of prenatal hydrocephalus

cannot be easily detected with ultrasound eg, migrational abnormalities and porencephaly. MRI study has tremendous impact on counseling and management decision.

Amniocentesis: Amniocentesis is done at ≥15 weeks of gestation for determination of the fetal karyotype. Chromosomal microarray (CMA) should be offered to patients with isolated mild ventriculomegaly and recommended when additional abnormalities are detected. The amniotic fluid is also tested for alpha-fetoprotein and acetylcholinesterase to exclude an occult open neural tube defect, which can often lead to ventriculomegaly. PCR for CMV and toxoplasmosis should also be obtained when amniocentesis is performed. Maternal serology is used to identify an infectious etiology. However, serology is neither as sensitive nor as specific as PCR on amniotic fluid, thus amniotic fluid PCR is the preferred method of evaluation for infection.

Treatment:

There are several treatment options: termination of pregnancy, intrauterine shunting, early delivery and neonatal shunting.

After several years of treating and monitoring fetuses with fetal hydrocephalus, it is concluded that an evolving acute hydrocephalus without other associated malformations, intrauterine procedures can be beneficial.

The following requirements have been proposed to select patients eligible for fetal treatment:

1. Hydrocephaly should be diagnosed at an early stage of gestation
2. It should not be associated with other malformations
3. Normal karyotype
4. Ventricular dilatation should be progressive
5. The treatment should be managed by amultidisciplinary team comprising specialists in perinatology, ultrasonography, obstetrics, neurosurgery and genetics.

The ventriculoamniotic shunt placement through a hysterotomy in the second trimester of pregnancy is treatment for isolated obstructive hydrocephalus. Surgery should be planned by serial ultrasonographic examinations and an ultrafast magnetic resonance imaging to confirm isolated aqueductal stenosis. A normal fetal karyotype and negative polymerase chain reaction or culture of the amniotic fluid for cytomegalovirus and toxoplasmosis were obtained. Serial enlargement of the lateral ventricles >1.5 mm/week and fetal macrocephaly were documented. Using epidural anaesthesia a standard ultra small ventricular catheter and valve were inserted via a hysterotomy. The distal catheter, rather than being inserted into the fetal peritoneum, exited between the fetal scapulae [10] (Figure: 2).

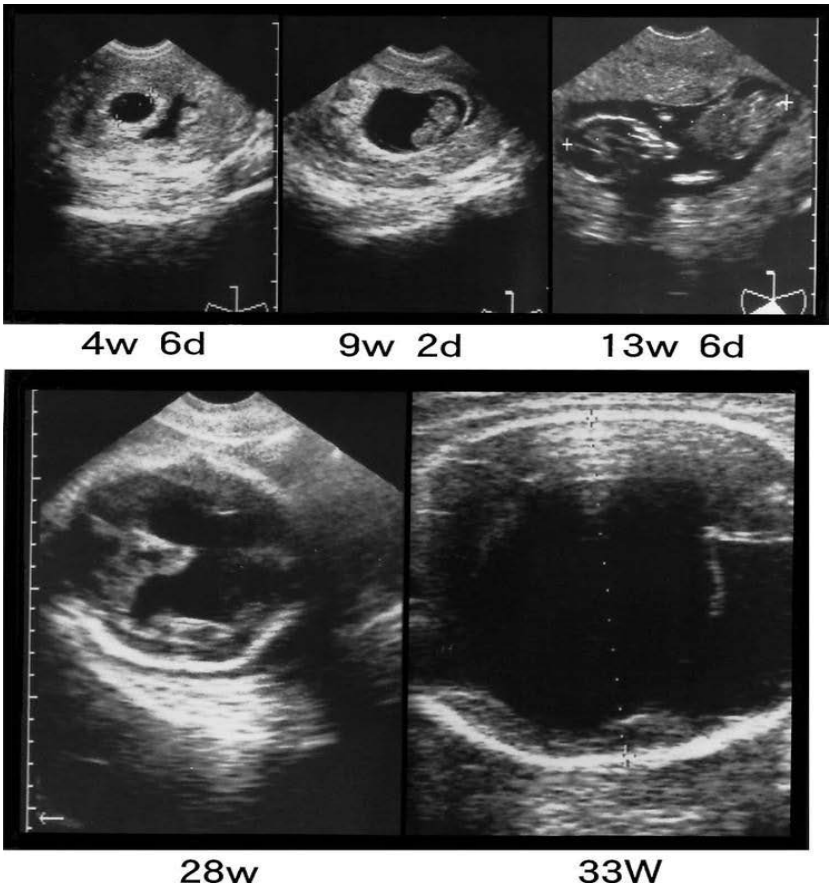

Figure 2: Ultrasonographic findings of prenatal hydrocephalus in different gestational period

Counseling:

The range of potential outcomes should be discussed with the family. If the etiology of ventriculomegaly has been determined (eg, trisomy, CMV) or associated malformations are identified, the parents can be given more specific information than in cases where counseling is only based on ventricular width. Before viability, pregnancy termination is an option and should be offered. In those patients who elect to terminate, evaluation to confirm or determine the etiology is warranted, as identifying a cause can be helpful in determining recurrence risk in future pregnancies.

Discussion:

Systemic review and detail analysis of literature made our understanding of antenatal hydrocephalus more clear. In the past, this was a problem where neurosurgeons were unable to tackle the crisis. Advanced knowledge showing a light of hope. Recent developments in prenatal ultrasonography and MRI neuroimaging techniques have enabled more frequent diagnoses of congenital anomalies of the central nervous system in the prenatal period. The frequency of prenatal hydrocephalus is reportedly 0.2–0.8/1000 births [11]. Robroch et al claimed that the anomaly has a male preponderance [12]. Differentiation from fetal ventriculomegaly, the frequency of which is 0.5–3.8/1000 births, is indispensable but not straightforward in some cases. Prenatal hydrocephalus is highly suspected if the ventriculomegaly is progressive. Such cases of progressive ventriculomegaly occur at a rate of 10–20%, while the frequency of fetal ventriculomegaly remains stable at 50–60%, with spontaneous resolution observed in about 30–50% of cases. According to Hannon et al the incidence of the major forms of ventriculomegaly is 3.6 per 10,000 births [13].

For a definitive diagnosis, MRI, TORCH test (toxoplasmosis,other infections, rubella, cytomegalovirus and herpes simplex virus), genotype and karyotype tests are needed in addition to fetal echocardiography [14]. The prenatal diagnosis of hydrocephalus allows clinicians to provide parents with information about the future of the fetus particularly cases with isolated ventriculomegaly [15]. According to Xie et al cases with an expansion of the lateral ventricle with a transverse diameter ≥12 mm with an intrauterine progression are generally associated with a poor prognosis and should therefore be closely monitored [16].

Antenatal shunting has been considered in a small proportion of selected cases. Criteria for fetuses to be considered for ventriculoamniotic shunts were gestational age of less than 30 weeks, progressive hydrocephalus, and no associated congenital abnormalities seen on sonography. During the early 1980s, the results of intrauterine surgery were not satisfactory owing to an improper distinction between the types of hydrocephalus. Nowadays, with the advancement in imaging and intrauterine surgical techniques, the efficacy of intrauterine therapy in properly selected fetuses, especially progressive hydrocephalic cases has been improved [17]. However, the clinical results are not impressive even in the best surgical hands, although techniques such as cranio-cervical decompression and autologous durosplasty produce satisfactory results in isolated cases of Arnold-Chiari type II malformation [18, 19].

The prognosis of both prenatal hydrocephalus and fetal ventriculomegaly is multifactorial. The size of the ventriculomegaly and pathogenesis and the time at onset may influence outcomes. The survival rate of overall fetal ventriculomegaly is reportedly 40–50%, with some cases being terminated at the request of the parents when the diagnosis is made in the early fetal period. The peri- and postnatal mortality rates reach 10–20%. The developmental outcomes of fetal ventriculomegaly vary depending on the degree of ventriculomegaly and the presence of associated abnormalities within and without the central nervous system. Normal development is reportedly seen in 80–90% of cases of isolated mild ventriculomegaly. Even in postnatally treated prenatal hydrocephalus, normal development is expected in about 20% of cases [11]. A poor outcome seems unavoidable in the majority of genetic, post-infectious, and syndromic hydrocephalus.

Conclusion

The prognosis for fetal hydrocephalus is not as favorable as for neonatal hydrocephalus. Further progress in intrauterine therapy and aggressive obstetric management (e.g., premature induction of labor with early neonatal shunt placement) may improve the morbidity and mortality of fetal hydrocephalus.

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