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RESEARCH ARTICLE

PEDIATRIC OCULAR CHANGES IN HYDROCEPALUS: REVIEW

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ABSTRACT

Hydrocephalus is a condition in which abnormal accumulation of cerebro spinal fluid within the ventricles or subarachnoid space of the brain. It develops when there is an imbalance between the amount of CSF production and the rate at which it is absorbed. This can be caused by a blockage in the pathways through which the fluid travels or from an overproduction of fluid. This disturbance then causes the ventricles to enlarge (i.e. ventriculomegaly) or subarachnoid space and intra cranial pressure to increase, resulting in an enlarged head along with other symptoms i.e. headache, irritability, lethargy, fever, vomiting, visual changes like setting sun sign, visual field defect, strabismus, papilledema etc.. Cranial Ultra Sound, CT and MRI play an important role in the diagnosis of Hydrocephalus. CT can assess size of ventricles and other structures. MRI can find malformation, tumors and other causes. Patients who progress to more severe forms will have to either undergo a shunt placement or endoscopic third ventriculostomy. Thorough eye examination is important to detect ocular changes and it will be useful to stop further vision loss. Further research is required in order to early diagnosis which helps in reducing severity of disease.

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INTRODUCTION

Hydrocephalus is a condition where an abnormal accumulation of cerebro spinal fluid (CSF), causes an increase pressure in the ventricles or subarachnoid space of the brain. It can be caused by either the blockage or by inadequate re-absorption of CSF fluid. The term Hydrocephalus comes from the two Greek words 'hydro' which means water, and 'cephalus' which means head. The shape of the head may also indicate the location of an obstruction. Big forehead is seen in aqueductal stenosis, an occipital prominence of skull is seen in Dandy Walker malformations. Other signs include an enlarged fontanelle and full anterior fontanelle, headache, sun setting eyes, irritability, lethargy, fever, vomiting etc. The evaluation of understanding and treatment of hydrocephalus is summarized. Hippocrates (476-377 BC) and Galen (130-200 AD) believed that Hydrocephalus was caused by extracerebral accumulation of water. Morgagni (1682–1771) described the pathology of hydrocephalus. In 1768, Whytt distinguished internal and external hydrocephalus.

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Early treatment included bleeding, purging, surgical release of the fluid, puncturing the ventricles to drain the fluid, injection of iodine or potassium hydriodate into the ventricles, binding of the head, application of a plaster of herbs to the head, application of cold wraps to the head, lumbar puncture, and diuretics. Surgical treatment attempts were made during the 10th century with evacuation of intraventricular fluid. Treatment of hydrocephalus by ventriculo-atrial shunting was introduced in the 1960s and was followed by ventriculo-peritoneal shunting during the following decade (Forrest 1968, Cinalli 1999). Before the shunting era, there was a very high mortality among children with hydrocephalus, and other conditions than vision was the focus of interest (Hadenius *et al.*, 1962). Nowadays more than 90% of affected children survive. A large proportion of the brain function is devoted to visual tasks, and since hydrocephalus causes multiple impairments of brain function, the visual system is commonly affected. During the pre-shunting era and from the beginning of the shunting era, in the 1960s, the most commonly reported ophthalmologic findings were the setting sun sign, optic nerve atrophy and strabismus in children with hydrocephalus (Duke-Elder 1964, Walsh and Hoyt 1969).

Pathophysiology and Aetiology

Cerebrospinal fluid (CSF) is a clear, watery substance, produced within the lateral ventricle that flows into the subarachnoid space. It is absorbed by means of hydrostatic gradient. CSF contains small molecules, salts, peptides, proteins, enzymes, etc. It nourishes the brain and carries waste products away from the brain. The normal rate of CSF production in infants and children is about 0.33 ml/kg/hr. Normal newborns have about 5 ml and adults have about 125 ml of total volume of CSF, with about 20 ml located within the ventricles. Hydrocephalus leads to changes of the brain, not only of the morphology but also on the circulation, biochemistry, metabolism and maturation. The white matter, especially the periventricular region is the brain region that is most affected in hydrocephalus. Also the cerebral cortex undergoes gross changes with the onset of hydrocephalus, a great thinning of the cortex and extension. Histological and biochemical changes have been noted in the neurons affected by hydrocephalus. Retrograde neural degeneration has been seen in retinal ganglion neurons and cortex (Kriebel *et al.,* 1993). Different aetiological factors have causing hydrocephalus in different parts of the world. The most common prenatal causes are malformations of the CNS and genetic factors. Intra ventricular haemorrhage and infections are the most frequent causes in the perinatal period as are tumours, trauma and infections during the postnatal period. Infections in the CNS may cause development of hydrocephalus due to obstruction to different parts of the CSF pathways. The most common causative infections are: toxoplasmosis, cytomegalovirus infection and bacterial infections (Persson *et al.*, 2005, Warf *et al.,* 2010, Rashid *et al.,* 2011).

Types and Terminology

On Etiology it can be grouped as congenital or acquired. Congenital hydrocephalus is caused by any condition that existed before birth. The hydrocephalus may or may not be present at birth. It results from a complex interaction of genetic and environmental factors. Acquired hydrocephalus is hydrocephalus resulting from a condition that did not previously exist in the patient. Causes for acquired include mass lesions or tumors (medulloblastoma, astocytoma), cysts, abscesses, hematoma, hemorrhage (Chauvet *et al.,* 2009) (Oertel *et al.,* 2009). On Pathology, Hydrocephalus can also be described as communicating or non-communicating. Communicating Hydrocephalus results when the arachnoid villi are unable to adequately reabsorb the CSF then it blocked after it exits the ventricles. Noncommunicating or obstructive hydrocephalus is a condition that occurs when the ventricular system does not communicate with the arachnoid villi to reabsorb due to some obstruction in the pathway. Internal hydrocephalus refers to ventricular dilation and the associated pathophysiology. External hydrocephalus refers to the accumulation of spinal fluid in either the subarachnoid or subdural spaces. Additionally, there are Normal Pressure Hydrocephalus (NPH) occurs in the sixth or seventh decade of life and is characterized with specific symptoms: gait disturbance, cognitive decline and urinary incontinence (i.e. Adam's or Hakim's triad). Preferred treatment for NPH is often shunt surgery.

Visual Symptoms of Hydrocephalus

Children with hydrocephalus are gradually develops eye problems. High cerebro spinal fluid (CSF) pressure can damage vision, the resulting problems ranging from mild deterioration to marked loss. The vision changes in hydrocephalus are sun setting sign, Abnormal pupil reaction to light, Double vision (Diplopia), Swelling of the optic nerve (papilledema), Diminished vision (Visual acuity < 20/20 vision), Grayouts or fuzzouts of vision, Reduced visual field, Reduced color vision. Strabismus is a common complication in children with hydrocephalus. A majority of these children have horizontal deviations, where esotropia is more common than exotropia (Rabinowitz 1976, Aring *et al.,* 2007). Nystagmus is also common and may have various causes and appearances (Rabinowitz 1976, Gaston 1991, Aring *et al,.* 2007). A number of studies have addressed several aspects of vision in patients with myelomeningocoele and have shown that vision is compromised frequently and in diverse ways. These include visuoperceptual disturbance, (Ito *et al.,* 1997), strabismus and amblyopia, papilloedema, impaired visual fields and optic atrophy (Gaston H. 1991).

DISCUSSION

High intra cranial pressure can produce neuro-ophthalmic problems like disturbed ocular motility, visual field defects of optic disc, chiasmal and retrogeniculate nature and lass of visual acuity (Osher *et al.,* 1978., Chattha *et al.,* 1975, Sinclair *et al.,* 1931, Hughes, 1946). The sun setting sign is characterized by the child's inability to look upward, as the eyes are displaced downward, Upper Scleral portion is exposed due to the pressure on the cranial nerves controlling eye movement. As a result, the infant appears as though it is looking at the bottom. Pressure changes in CSF which flows in the meninges surround the optic nerve can also produce pressure on the optic nerve. This pressure reduces the supply of nutrition and oxygen to the optic nerve. It causes to Swelling of the optic nerve (papilledema). This high CSF pressure leads to damage to the optic nerve and results as diminished vision, color vision defect, and visual field loss. In young children esotropia or in older children exotropia may be the result of visual loss due to optic atrophy from long-standing papilledema. (Wybar K. 1974.). (Harcourt RB. 1968.). IIIrd nerve palsy leads to exotropia, IVth nerve palsy lead to vertical deviation, VIth nerve palsy leads to esotropia. Unilateral or bilateral lateral rectus paresis is the most frequent ocular motility disturbance. It can force a child to assume an uncomfortable head posture, with a tilt or turn (to avoid the resulting double vision) can cause pain in head and neck. With this adults may experienced as double vision, while children may or may not noticed double vision. Unilateral and bilateral proptosis have been reported as a feature of Hydrocephalus (Osher *et al.,* 1978) (Gardner, 1948) (Shapi *et al.,* 1976). Visual acuity is often reduced in children with hydrocephalus (Rabinowicz, 1974, Ghose, 1983, Mankinen-Heikkinen *et al.,* 1987; Biglan, 1990) due to lesions at various levels of the visual system. The anterior visual pathway may be affected as in optic atrophy or optic nerve hypoplasia or the posterior visual pathway as in periventricular leucomalacia.

Some authors have reported reduced visual acuity with repeated shunt dysfunctions (Arroy *et al*., 1985, Gaston 1991). In comparison with healthy children, children with hydrocephalus are more often hypermetropic, a result which has also been reported for other children with cerebral damage (Saunders 2002). Impaired emmetropisation related to abnormal visual input and processing is a potential contributory factor. Astigmatism is also common in children with hydrocephalus while myopia has been reported to occur with the same frequency as observed in healthy children (Mankinen-Heikkinen *et al.,* 1987). Ventricular size (measured at the levels of the anterior horn and collateral trigone) correlates inversely with visual function in hydrocephalic children. (Odebode *et al.,* 1998) Later it confirmed this relationship by demonstrating that visual function is impaired in direct proportion to the degree of ventricular enlargement in both control and HLM groups of patients (Shokunbi *et al.,* 2002). Visual loss is a common and well-recognized complication of both children and adults suffering from hydrocephalus. Unilateral or bilateral blurring of vision, and progressive decline of Vision were the commonest visual complaints (Harrison *et al*., 1974). Visual loss in these patients may occur with undetected amblyopia due to eso or exotropia or can be caused by papilledema. Sometimes Vision may also be affected in advanced hydrocephalus due to compression of the optic chiasma as a result of a dilated 3rd ventricle. Visual Loss can also occur with rapid rise in CSF Pressure and Shunt Malfunction/ failure and sudden visual loss occurred with rapid drop in CSF Pressure. Cushing and Walker believed that chiasmal bitemporal visual loss was very rare if it occurred at all as a result of 3rd ventricle dilation (Cushing *et al.,* 1912). Direct trauma to visual pathways by shunts may account for some visual loss.

Diagnosis

Hydrocephalus can be diagnosed before birth with Prenatal Cranial ultra sound (CUS). The computed tomography (CT) or magnetic resonance imaging (MRI) scan can be performed assess the cause of hydrocephalus, may reveal many structural changes like enlarged ventricles, thinning of the cortical mantle, distortion of structures, and possible transependymal flow of CSF. The half-Fourier single-shot turbo spin-echo (HASTE) MRI is a limited T2 image that shows ventricular size. It is an alternative to CT, does not expose the child to radiation, and requires no sedation as it is a short study (Penzkofer *et al.,* 2002). Head circumference should be routinely measured in infants. X-rays of the head may provide further evidence such as craniofacial disproportion, or elongated interdigitations of suture lines. Measuring intracranial pressure (ICP) can be done using an intraventricular or lumbar catheter. Other procedures are Cisternography, CSF tap test, Continuous CSF drainage etc. CSF drainage is one the best available test.

Management

Acetazolamide, a carbonic anhydrase inhibitor was used to reduce CSF production. It cannot be used as a long-term treatment modality. Communicating hydrocephalus is often treated with shunt surgery while noncommunicating hydrocephalus suggests treatment with ETV. Visual aids can be advised in vision impairment. Patching treats poor vision from amblyopia. A shunt is a thin, soft, flexible catheter placed for drain the extra CSF from the ventricles of the brain or in the lumbar subarachnoid space to another area (where it can be absorbed) i.e. the peritoneal cavity (Ventriculo peritoneal shunt), right atrium (ventriculo atrial shunt), or pleural space (Ventriculo pleural shunt). Shunt has three basic components Catheter, which is inserted into brain ventricles, Valve, which regulates flow of spinal fluid and Long catheter that carries CSF from head to whatever CSF is diverted. Endoscopic third ventriculostomy (ETV) surgery is developed to avoid potential shunt complications (e.g. shunt failure, infection rates, etc.). A neuroendoscope enters through a precoronal burr to visualize the anatomy of the ventricles and the floor of the 3rd ventricle. It is guided through the cerebral mantle, through the lateral ventricle and the foramen of Monro into the 3rd ventricle. Forceps and a balloon are used to perforate a hole downward and widen a stoma in the floor of the 3rd ventricle, anterior to the mammillary bodies and bifurcation of the basilar artery, creating a passage to divert excessive CSF into the prepontine space. This diverted fluid will be absorbed through subarachnoid space.

Conclusion

Many studies have demonstrated that various ocular changes developed in hydrocephalus due to pressure variation, cerebral changes and shunt failure. Currently further research can be done, in the absence of a true gold standard diagnostic tools and treatment for the hydrocephalus.

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