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Clinical Presentation of Hydrocephalus

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1. Introduction

The two key determinants of clinical presentation of hydrocephalus are the age of onset and the acuity of the rise in intracranial pressure.

In fetus, while minor degree of hydrocephalus often goes undetected, severe cases of obstructive variety present with the following features: 1 .the head is felt larger, globular and softer than the normal head 2.the head is high up and difficult to push down into pelvis 3.fetal heart sound is situated high up above the umbilicus 4.internal examination during labor may reveal widening of sutures and tense fontanelle. In breech presentation however, the diagnosis is not made until the after-coming head is arrested at the brim. In the communicating variety, often the head is of normal size at birth and its enlargement starts only at 6-12 weeks of age.

In Infancy and early childhood (prior to 2 years of age), progressive enlargement of the head is the commonest manifestation of hydrocephalus as sutures have not united firmly. Occipitofrontal circumference should be measured after 24 hours of birth when moulding and overriding of sutures have disappeared. The head continues to enlarge and appears to be disproportionately larger than the rest of the body. The head is quite heavy and the child is not able to hold it without support. The child starts becoming less playful and does not feed properly. Milestones tend to get delayed. On examination, the child's head is found to be large in proportion to the body with bossing of frontal bones giving an inverted triangular appearance to the head. Serial head circumference should be taken to identify whether it is progressive (active) or arrested hydrocephalus. The fontanelle are widely open, bulging, tense and non pulsatile and the scalp veins are engorged, strikingly so when the infant cries. Sutures are widely separated. Cracked pot sound may be heard on percussion of the head due to separation of sutures and is called "Macewen's sign".Bruits over fontanels on auscultation of skull with the bell of the stethoscope indicates vascular origin of hydrocephalus (vein of Galen aneurysm or other vascular malformation).Transillumination of skull should be done in all cases. Normally, the halo of light around the rim of the illuminator extends upto 1 cm in the occipital region and upto 2 cm in frontal region in term babies. Excessive transillumination indicates abnormal collection of fluids as in hydranencephaly where the whole skull may glow with light or Dandy Walker syndrome where posterior part of skull transilluminates owing to the fluid accumulation in the posterior fossa.

The limbs may show increased tone and brisk reflexes "spastic paraparesis". This results from stretching and distortion of paraventricular corticospinal tracts arising from leg area of motor cortex. These fibres have a longer distance to travel around the ventricles than those supplying the face and the upper limbs. However, mild spasticity and weakness of upper limbs is not uncommon particularly in advanced cases. Spine examination should be performed in all cases to look for presence of spina bifida (commonly associated with Chiari Malformation II).

Downturning of eyeballs with visibility of sclera above the iris is called 'Sunsetting Sign" and is a frequent finding. It is due to pressure on the superior quadrigeminal plate against the free edge of the tentorium causing a supranuclear paresis. It may be intermittent to begin with but later becomes continuous. Other ocular disturbances include unilateral or bilateral abducens nerve paresis, nystagmus, ptosis, strabismus and diminished papillary light responses.Optic atrophy can occur due to compression of the chiasm and optic nerves by a dilated anterior portion of the third ventricle. Papilledema is rare because rising tension is easily buffered by sutural diastasis.

Fig. 1. Congential Hydrocephalus: Downturning of eyeballs with visibility of sclera above the iris (Sunsetting Sign).

In earlier stage, child may be quite playful, pick up the objects, put them into mouth, recognize the parents and follow light and objects. As hydrocephalus progresses , further destruction of cerebral cortex occurs, child tends to become listless , stops taking interest in the surroundings and regression of earlier achieved milestones occurs .There is unusual somnolence, persistent vomiting ,failure to thrive and visual loss. Finally a decerebrate state ensues.

In early to late childhood (2 years and above), neurological symptoms caused by increased intracranial pressure or by focal deficits referable to the primary lesion is the predominant finding and appear before any significant change in head size. The clinical picture of various space occupying lesions depend on their site of origin.A unique but rare hydrocephalic syndrome "the bobble head doll syndrome" is related to obstructive lesions in or around

third ventricle and is characterized by bobbing of head forward and back involuntarily mimicking a bobble head doll. This can be inhibited voluntarily and disappears during sleep. A symptom very closely associated with head bobbing is the presence of ataxia. Several patients were reported to have difficulty in walking, running, and climbing steps because of the bobbing. One likely explanation for such symptom is interruption of the patient's ability to balance which rely on input from various sources namely, the vestibular, ocular and somatosensory due to constant bobbing. The head bobbing is a neurological phenomenon and stems from dilated third ventricle impinging on the medial aspects of the dorsomedial nucleus of the thalamus.

1. Hydrocephalus
2. Hydranencephaly
3. Subdural Effusion
4. Thickened skull bones (achondroplasia, osteopetrosis,pyknodysostosis,craniometaphyseal dysplasia, orodigitofacial dysostosis , rickets, leontiasis ossea,etc.)
5. Cerebral Gigantism
6. Mucopolysaccharidosis
7. Cerebral lipidosis (gangliosidosis)
8. Metachromatic leukodystrophy
9. Fragile X Syndrome
10. Porencephaly
11. Subdural Hematoma
12. Intracranial Tumor
13. Glutaryl-1-Coenzyme A dehydrogenase deficiency

Table 1. D/d of Enlarged Head in Newborn, Infancy and Children

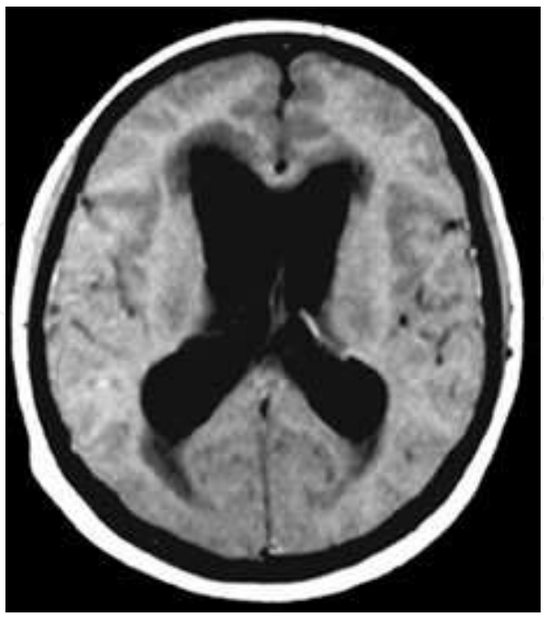

Fig. 2. CT scan of the brain at the level of basal ganglia showing enlarged lateral ventricles suggestive of hydrocephalus.

The features of raised intracranial pressure are evident in almost all instances and includes frontal headache aggravated in the morning, improving with upright posture and associated with nausea and vomiting. The cracked pot sound is prominent on skull percussion. Fundoscopy may show papilledema. Additional features seen in late group are endocrine changes including small stature, gigantism, obesity, precocious or delayed puberty, menstrual irregularities, absent secondary sexual characteristics and central diabetes insipidus. They are probably caused by compression of hypothalamic pituitary axis by an enlarged third ventricle (a particular risk in aqueductal stenosis) resulting in abnormal hypothalamic pituitary function. Spastic diplegia is common. Thought and behavior may be affected adversely. Learning disabilities are fairly common and these children are credited with better verbal IQ than performance IQ. While the severity of hydrocephalus can differ considerably between individuals, some are of average or even above-average intelligence. Patients may develop motion and visual problems, coordination problems, or may be clumsy. Perceptual motor deficits and visual spatial disorganization follow as a result of stretched corticospinal fibres of parietal and occipital cortex due to dilated posterior horns of lateral ventricles. About one in four develops epilepsy.

2. Normal Pressure Hydrocephalus

Normal Pressure Hydrocephalus (NPH) is a clinical symptom complex, characterized by the classic triad of gait abnormality, dementia and urinary incontinence (commonly referred to as "wet, wobbly and wacky").NPH occurs either as idiopathic or secondary condition, roughly in equal proportions. While NPH secondary to an identifiable cause can occur in all age groups, idiopathic NPH is most common in adults over 60 years of age without any sex predilection. These manifestations are believed to arise from dysfunction of periventricular white matter tracts, particularly those sub serving frontal lobe connections .Gait difficulty is often the first clinical manifestation. It is the effect of expansion of the ventricular system (particularly the lateral ventricles) and subsequent traction on the lumbosacral corticospinal fibers arising from the motor cortex. It is also believed to be the most responsive feature to shunting. It is classically described as “magnetic gait”; the patient's feet appear to stuck to the floor, steps are characteristically short with decreased stride length and height and a broad base. This may resemble parkinsonian gait at a glance but is distinguished by a narrow base and absence of tremor or rigidity. Postural stability is impaired, and a history of falls may be reported.

The cognitive disturbance of NPH is likely to be frontal in nature with psychomotor slowing, decreased attention and concentration, and apathy. The patient is slower in timed tasks, performs poorly on tests of divided attention and executive function, has difficulty with fluency tests, and has poor learning and better preserved recognition memory. The dementia is believed to be the consequence of stretching of frontal and limbic fibers that travel in the periventricular region. The Mini Mental State Examination may be an insensitive measure of cognitive impairment in NPH since it exhibits a frontal subcortical pattern rather than a cortical pattern and neuropsychological tests may prove to be a better tool in its characterization as well as diagnosis of coexisting dementia conditions (including Alzheimer’s dementia and vascular dementia which are also highly likely in advancing age).

The third component, urinary incontinence, often begins as urgency and frequency rather than incontinence per se. However, overtime, true urinary incontinence ensues and is

accompanied by a lack of concern to urinary symptoms, reflecting its probable origin in the frontal lobe.

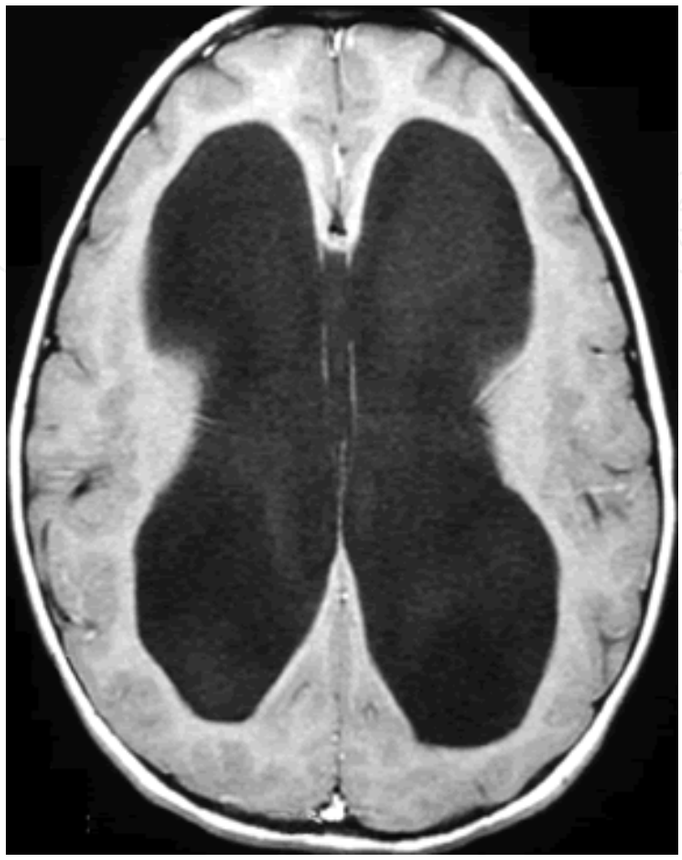

Fig. 3. Enlarged lateral ventricles with thinning out of cerebral cortex in a patient with normal pressure hydrocephalus.

Other features of NPH may include long tract signs with spasticity of lower limbs, hyperreflexia and extensor plantar responses (upper motor neuron signs). In very late stages, frontal release signs, akinetic mutism, and quadriparesis may occur.

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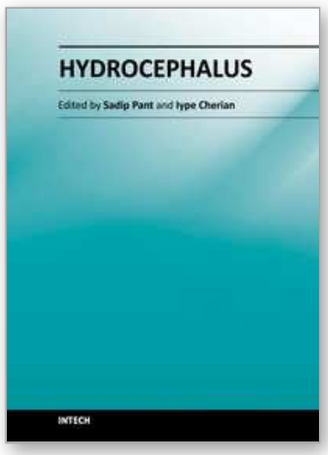

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Description of hydrocephalus can be found in ancient medical literature from Egypt as old as 500 AD. Hydrocephalus is characterized by abnormal accumulation of cerebrospinal fluid (CSF) in the ventricles of the brain. This results in the rise of intracranial pressure inside the skull causing progressive increase in the size of the head, seizure, tunneling of vision, and mental disability. The clinical presentation of hydrocephalus varies with age of onset and chronicity of the underlying disease process. Acute dilatation of the ventricular system manifests with features of raised intracranial pressure while chronic dilatation has a more insidious onset presenting as Adams triad. Treatment is generally surgical by creating various types of cerebral shunts. Role of endoscopic has emerged lately in the management of hydrocephalus.

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