Clinical Features Of Hydrocephalus: Children Vs Adults

M AHMAD M ASIF K SHAFIQUE S MANZOOR A H VOHRA N AHMAD K A BOKHARI I A RAJA
Department. of Neurosurgery , Lahore General Hospital/King Edward Medical College , Lahore .

Correspondence to : Dr. Manzoor Ahmad

Hydrocephalus is defined as an excessive accumulation of CSF within the ventricular system . It can be congenital or acquired and can present at any age . The clinical presentation depends upon the age of the patient , the severity of the underlying cause and the rapidity of progression of hydrocephalus . In infants , hydrocephalus presents as morphological (structural) changes as the predominant feature while in older children and adults , functional disturbances are predominant due to raised ICP. In this prospective study , 100 consecutive patients of all age groups with all types of hydrocephalus are included . The results showed that 80 % patients were children up-to the age of 14 years (group 1) and the other 20 % were adults (group 2) . In the first group , the main clinical features were : macrocephaly (81.3 %), vomiting (18.8 %) , fits (13.8 %) and headache (11.3 %) . Only 5 % patients in this group were found to have papilloedema .In the second group of patients aged more than 14 years , the main clinical features were : headache (90 %) , vomiting (80 %) , deterioration of conscious level (30 %) and deterioration of vision (15 %) . Papilloedema was found in 70 % of these patients . Results are discussed in detail and compared with other studies .

Key Words : Hydrocephalus , Headache , Papilloedema .

Hydrocephalus is the result of an imbalance of cerebrospinal fluid (CSF) formation and absorption of sufficient magnitude to produce a net accumulation of this fluid within the ventricular system[1] . this leads to enlargement of head in children or headache & vomiting due to elevation of the intra-cranial pressure. Hydrocephalus has to be differentiated from cerebral atrophy in which the excessive accumulation of CSF in the ventricles and subarachnoid spaces is due to a loss of cerebral substance rather than a primary defect of CSF formation or absorption .

This differentiation is possible on clinical grounds and by investigations such as CT scan and ICP monitoring[2,3].

Hydrocephalus can be congenital or acquired. It has diverse types of presentations depending upon the age of the patient , the rapidity of onset and severity of progression of hydrocephalus. The clinical course can be discussed as hydrocephalus in :

1. Infants / children .
2. Adults.
3. Normal Pressure Hydrocephalus (NPH) .

Infantile Hydrocephalus:

In a fully established case of hydrocephalus at birth or afterwards there is macrocrania , convex ,tense , non-pulsatile fontanellae , thin and shiny scalp with prominent veins[13] . Cranio-fascial disproportion ,with low-set ears and eyes may be present (Fig.1) .In infants with gross hydrocephalus , Macewen's sign ("cracked-pot" sound on percussion) and transillumination of the head may also be positive[2,4,5,13]. Irritability , vomiting , apathy and delayed milestones may alert the parents or clinician . In advanced cases the infant may develop paraparesis due to the stretching of pyramidal tracts in the corona radiata[6,13].Weakness of upward gaze ("setting-sun sign") due to pressure on the tectum of mid-brain by the dilated third ventricle and unilateral or bilateral sixth nerve palsy may a'so be present[2,4,5,13].

In doubtful cases serial measurements of head circumference and drawing a curve on the centile graph helps in the final diagnosis. Normal head circumference at birth is 33-36 cm. During the first year head circumference increases 2 cm / month during the first 3 months, one cm / month from 4-6 months and 0.5 cm / month from 7-12 months . A diagnosis of hydrocephalus is indicated more strongly if circumference increases across centile curves as compared to the circumferences that are above but parallel to the 95 % centile[13].

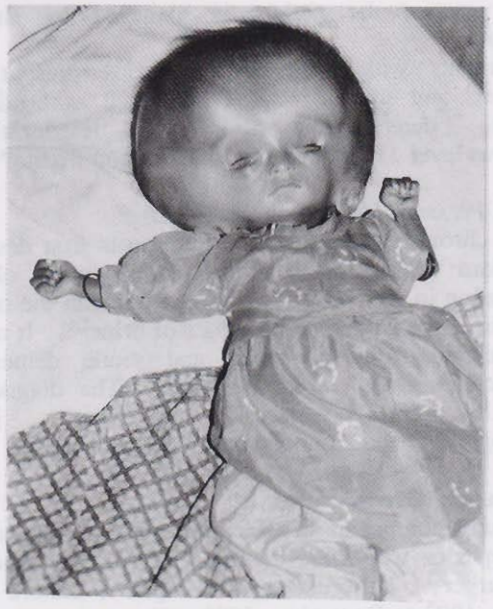

Fig.1: Advanced hydrocephalus with prominent scalp veins, frontal bossing , "setting-sun" eyes , low-set eyes & ears and craniofacial disproportion .

Papilloedema is not a characteristic feature of hydrocephalus in infants[7,13]. The infant should be examined thoroughly to rule out any other congenital malformation such as encephalocele , myelomeningocele (Fig.2), imperforate anus or cardiovascular malformations.

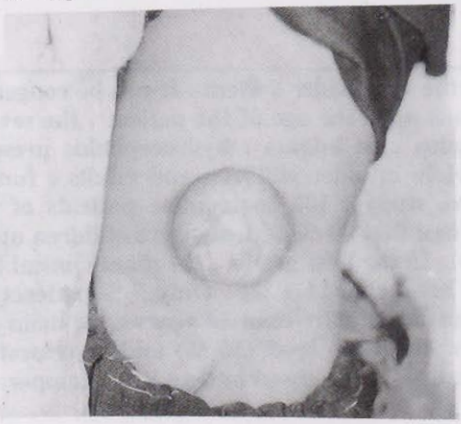

Fig. 2 : Hydrocephalic infant having associated myelomeningocele .

Adult-Type Hydrocephalus :

In older children and adults, enlargement of head is not possible since the fontanellae have closed and sutures have fused . These patients can present with symptoms and signs of acute or chronic hydrocephalus , depending upon the rapidity of progression and the severity of the underlying cause of hydrocephalus . Sometimes the patient may present with arrested hydrocephalus in which the ventricles are dilated but there is no pressure gradient and the patient has no signs of raised ICP . These patients need a regular follow-up so that if there is any deterioration of clinical status, the patient is managed by shunting procedure[2].

Patients with acute hydrocephalus present with symptoms and signs of raised ICP such as headache , vomiting , deterioration of vision and deterioration of conscious level . Papilloedema is a common finding[4,8,13].

Normal Pressure Hydrocephalus (NPH) :

This is chronic hydrocephalus which was first described by Hakim & Adams in 1965[6]. NPH is an old age presentation in which the patient presents with the triad of dementia , apraxia and incontinence of urine[2,14] . It must be differentiated from pre-senile and senile dementia , arteriosclerosis and cerebral atrophy[9]. The diagnosis is made by clinical features , CT scan , isotope cisternography and continuous ICP monitoring[6,9,14].

Material and Methods

This study comprised of 100 consecutive patients with hydrocephalus who were admitted in the Department of Neurosurgery , Lahore General Hospital , Lahore , from January, 1993 to December, 1993 . The study included all patients with all varieties of hydrocephalus . The age ranged from 14 days to 60 years . No patient was excluded from the study . A detailed clinical assessment was done which included a detailed history, thorough clinical examination and appropriate investigations .

History included a detailed history of present illness with special emphasis on neurological symptoms referred to hydrocephalus such as abnormally rapid increase in head size in infants . Raised ICP resulting in headache , vomiting , deterioration of vision and deterioration of conscious level , fits , motor deficits and personality changes in older children and adults . Moreover antenatal , natal and post-natal history along-with family history was noted . Special emphasis was given to cousin marriages , maternal malnutrition , exposure to radiation & drugs , infections during pregnancy and any previous history of congenital malformations in the family in case of congenital hydrocephalus . History of past illness especially any history of head injury , sub-arachnoid haemorrhage or meningitis was taken in cases of acquired hydrocephalus .

Clinical examination included :

- ñ General Physical Examination.
- ñ Examination of the head & spine.
- ñ Systemic examination.

Examination of the head included its circumference and other features of hydrocephalus such as tense bulging fontanellae , dilated prominent scalp veins , thin shiny scalp and "setting-sun" eyes . Spine was examined for any deformities and spina bifida .

Systemic examination included examination of the Central Nervous System and that of other systems. This included examination of higher mental functions , cranial nerve examination (especially papilloedema , visual acuity , extra-ocular movements) , sensory and motor systems and cerebellar signs .

Further systemic examination was done to rule out any other associated congenital anomalies in congenital hydrocephalus (congenital heart disease , imperforate anus , limb deformities etc.)

Analysis of the data collected in all these patients showed that out of these 100 patients , 80 (80 %) were children up-to the age of 14 years and they were kept in one group and other 20 (20 %) were adults .

Out of these 80 children , 65 (81.3 %) presented with large-sized head , 15 (18.8 %) presented with vomiting , 11 (13.8 %) came with fits , 9 (11.3 %) came with headache , 6 (7.5 %) presented with some kind of motor deficit and 5 (6.3 %) came with deterioration of vision . It was also noted that 4 (5 %) children were found to have papilloedema and 4 (5 %) patients were blind due to optic atrophy . Further analysis showed that 2 (2.5 %) children came with deterioration of conscious level . Incontinence of urine and faeces , ataxia , deafness and nystagmus was present in one patient (1.3 %) each (Table 1) .

On analyzing the adult group , out of these 20 patients , 18 (90 %) presented with headache , 16 (80 %) came with vomiting , 6 (30 %) were having deterioration of conscious level , 5 (25 %) had vertigo , 3 (15 %) had fits , 3 (15 %) had deterioration of vision , 2 (10 %) came

with motor deficit , 1 (5 %) with ataxia and 1 (5 %) with loss of memory . It was also noted that 14 (70 %) patients had papilloedema and 1 (5 %) had optic atrophy (Table 2)

Table 1: Clinical Presentation In Children(up-to 14 years of age- Total patients: 80)

Presentation	Frequency	%age
Large-sized head		
Vomiting	15	18.8
Fits	11	13.8
Headache	09	11.3
Motor deficit	06	07.5
Visual deterioration	05	06.3
papilloedema	04	05.0
Optic atrophy	04	05.0
Deterioration of conscious level	02	02.5
Incontinence of urine & faeces	01	01.3
Ataxia	01	01.3
Deafness	01	01.3
Nystagmus	01	01.3

Table 2: Clinical Presentation In Adults(N=20)

Presentation	Frequency	%age
Headache		
Vomiting	16	80.0
Papilloedema	14	70.0
Drowsiness	06	30.0
Vertigo	05	25.0
Fits	03	15.0
Visual deterioration	03	15.0
Motor deficit	02	10.0
Ataxia	01	05.0
Loss of memory	01	05.0
Optic atrophy	01	05.0

Discussion

Hydrocephalus can be congenital or acquired . It has diverse types of presentations depending upon the age of the patient at presentation and the rapidity of onset and severity of progression of the underlying process.

In this study the most common presenting feature in infants and children was macrocrania (81.3 %) followed by vomiting (18.8 %) while in adults headache was the most frequent (90 %), next came vomiting (80 %) . The other important difference was the presence of papilloedema in 70 % hydrocephalic adults while in children it was only 5 %(Table 3) . If the presenting features are compared it is evident that the signs of raised ICP and its effects are more prominent in adults because there is less space to dissipate the increasing pressure while in children head size increases to compensate the pressure effects .

A rare presentation has been reported in which a case of fourth ventricular arachnoid cyst presented as the syndrome of normal pressure hydrocephalus10. Another report from California reported a study of unilateral hydrocephalus in adults due to obstruction of one foramen of Monro11. A very rare presentation has been reported by Filippi, S et al . in which a 50 years old man presented with the complaint of sialorrhea only and there were no other complaints of raised intracranial pressure . The condition improved by CSF shunting[12].

In infants optic discs are often pale . Papilloedema is rarely encountered unless hydrocephalus is due to a tumour[8].

When hydrocephalus is severe and has been present for some years , different types of endocrine disturbances may occur which can result in stunting of skeletal growth, delayed sexual development , obesity , diabetes insipidus, pubertas praecox[8].

In adults, enlargement of the head is occasionally encountered . A large head may be familial, otherwise it indicates arrested hydrocephalus . Convulsions are rare in adults compared to infants and children . Papilloedema is often found . Occasionally , with inconspicuous or no previous symptoms, patients may present in an acute confusional state which may be brief and misdiagnosed as hysteria. Sometimes the underlying process responsible for hydrocephalus is so slow and insidious that the patient never presents with signs of raised ICP. Instead, the clinical presentation shows diffuse organic brain damage leading to deterioration of mental faculties which includes slowing of mental and physical performance, failing memory, lack of initiative and attention, poverty of thought, speech and activity, passing to frank dementia , stupor and coma[8].

Table 3: Clinical Presentation Children Vs Adults

Presentation	Children	Adults
Vomiting	18.8%	80.0%
Fits	13.8%	15.0%
Headache	11.3%	90.0%
Motor deficit	07.5%	10.0%
Visual deterioration	06.3%	15.0%
Papilloedema	05.0%	70.0%
Optic atrophy	05.0%	05.0%
Deterioration of conscious level	02.5%	30.0%
Ataxia	01.3%	05.0 %

Conclusion:

The clinical presentation of hydrocephalus depends upon the age of the patient at presentation and the severity and rapidity of progression of hydrocephalus . The underlying cause may be congenital or acquired .

1. Hydrocephalus is mainly the disease of infants and children up-to 14 years . They are 80 % of the total patients.
2. The main clinical presentation in children is macrocrania (81.3 %) while in adults headache is the commonest symptom (90 %).
3. Papilloedema is a rare finding in hydrocephalic children (5 %) while the majority of adults have papilloedema (70 %).
4. All such patients need shunting procedure at early stage to get optimal results of surgery .

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