Hydrocephalus: Clinical Features

Hydrocephalus is a condition characterized by an abnormal accumulation of cerebrospinal fluid (CSF) within the ventricular system of the brain, leading to increased intracranial pressure. Clinical manifestations vary based on **age of onset, chronicity, and underlying etiology**.

1. Clinical Features in Infants

In infants, the cranial sutures are not yet fused, allowing for compensatory skull expansion in response to increased CSF volume. Key features include:

a) Increased Head Size

- Macrocephaly: Disproportionate increase in head circumference.
- Bulging Fontanelle: Especially in non-sutured infants.
- **Splayed Cranial Sutures**: Due to increased intracranial pressure (ICP).

b) Neurological Symptoms

- Irritability and Poor Feeding: Early signs of increased ICP.
- High-Pitched Cry: Often associated with neurological distress.
- **Sunsetting Sign**: Impaired upward gaze due to pressure on the periaqueductal structures.
- **Hypotonia or Hypertonia**: Depending on severity and chronicity.

c) Developmental Delay

Delayed milestones due to motor and cognitive impairments.

2. Clinical Features in Children

As skull sutures close, symptoms shift from macrocephaly to signs of raised intracranial pressure:

a) Signs of Raised ICP

- **Headache**: Typically worse in the morning, relieved by vomiting.
- Nausea and Vomiting: Due to pressure on the brainstem.
- Papilledema: Blurred vision or visual loss due to optic disc swelling.
- Ataxia & Gait Disturbance: Due to cerebellar involvement.

b) Cognitive and Behavioral Symptoms

- Memory and Attention Deficits
- Irritability & Personality Changes
- Declining School Performance

3. Clinical Features in Adults

Adults with hydrocephalus can present with **acute or chronic** forms.

a) Acute Hydrocephalus (Obstructive, Post-Hemorrhagic, or Post-Traumatic)

- Severe Headache
- Rapid Neurological Deterioration
- Altered Consciousness, Coma
- Cushing's Triad (Hypertension, Bradycardia, Irregular Breathing)

b) Chronic Hydrocephalus (Normal Pressure Hydrocephalus - NPH)

- Gait Disturbance ("magnetic gait")
- Cognitive Decline/Dementia
- Urinary Incontinence

("Wet, Wobbly, and Wacky" triad)

4. Clinical Features in Elderly Patients (Normal Pressure Hydrocephalus - NPH)

NPH is a distinct clinical entity presenting with the classic **Hakim's Triad**:

- Gait Apraxia: Short, shuffling steps, often described as "magnetic" due to difficulty in lifting feet.
- **Dementia**: Subcortical cognitive dysfunction, including executive dysfunction and memory deficits.
- **Urinary Incontinence**: Urgency, frequency, or complete incontinence.

Summary of Key Clinical Features by Age Group

Age Group	Key Features
Infants	Enlarged head, bulging fontanelle, sunsetting sign, irritability, poor feeding
Children	Headache, nausea, vomiting, ataxia, cognitive decline
1/VUIIILC	Acute: Severe headache, Cushing's triad. Chronic: Gait disturbances, dementia, incontinence
Elderly (NPH)	"Wet, Wobbly, Wacky" triad: Gait disturbance, cognitive decline, incontinence

Early recognition and management of hydrocephalus, particularly in pediatric cases, are crucial to prevent long-term neurological impairment.

see Blindness from hydrocephalus.

Hydrocephalus can also cause death.

Infants commonly present with progressive macrocephaly whereas children older than 2 years generally present with signs and symptoms of intracranial hypertension.

One child with headaches, diagnosed with hydrocephalus, who played wind instruments. The patient manifested that their headaches worsened with the efforts made during playing their musical instruments. Martínez-Lage et al. hypothesize that the headaches might be influenced by increases in their intracranial pressure related to Valsalva maneuvers and had serious doubts on if we should advise our young patients about giving up playing their music instruments ¹⁾.

1)

Martínez-Lage JF, Galarza M, Pérez-Espejo MA, López-Guerrero AL, Felipe-Murcia M. Wind instruments and headaches. Childs Nerv Syst. 2013 Mar;29(3):351-4. doi: 10.1007/s00381-012-1920-8. PubMed PMID: 22968210.

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