HYDROCEPHALUS – PATHOPHYSIOLOGY, CLASSIFICATION, AND CLINICAL MANIFESTATIONS

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Abstract: Hydrocephalus is a neurological disorder characterized by abnormal accumulation of cerebrospinal fluid (CSF) within the ventricular system, leading to ventricular dilation and increased intracranial pressure. It affects individuals of all ages, from neonates to the elderly, and may arise from congenital or acquired causes. This article reviews the pathophysiological mechanisms, classification systems, clinical features, and complications associated with hydrocephalus.

Keywords: hydrocephalus, cerebrospinal fluid, intracranial pressure, ventricular dilation, aqueductal stenosis, normal pressure hydrocephalus.

Introduction

Hydrocephalus remains a major challenge in neurology and neurosurgery due to its complex pathogenesis and heterogeneous clinical presentation. The estimated global incidence of congenital hydrocephalus ranges from 1 to 2 per 1,000 live births, while acquired forms are commonly associated with intracranial hemorrhage, infections, tumors, and traumatic brain injury. Persistent elevation of intracranial pressure can lead to progressive neurological impairment and death if left untreated. Timely diagnosis and intervention are critical for favorable outcomes.

CSF Physiology and Pathophysiology

CSF is primarily produced by the choroid plexus at a rate of approximately 0.3–0.4 mL/min and normally circulates from the lateral ventricles through the third and fourth ventricles into the subarachnoid space, where it is reabsorbed by arachnoid granulations. Hydrocephalus develops when this delicate balance between CSF production, circulation, and absorption is disrupted.

Obstruction of CSF flow (obstructive or non-communicating hydrocephalus) occurs in conditions such as aqueductal stenosis, tumors of the posterior fossa, and congenital malformations. In contrast, communicating hydrocephalus results from impaired CSF reabsorption at the level of arachnoid villi, commonly seen after meningitis, subarachnoid hemorrhage, or intraventricular hemorrhage.

Classification of Hydrocephalus

Hydrocephalus can be classified based on etiology and anatomical features:

- **Congenital hydrocephalus**
- **Acquired hydrocephalus**
- **Obstructive (non-communicating)**

- **Communicating**
- **Normal pressure hydrocephalus (NPH)**

Normal pressure hydrocephalus is a distinct entity seen in older adults, characterized by ventricular enlargement without marked elevation of intracranial pressure and presenting with the classic triad of gait disturbance, urinary incontinence, and cognitive decline.

Clinical Manifestations

Clinical presentation depends on the patient's age, rate of ventricular enlargement, and underlying cause. In infants, common signs include rapid head circumference growth, bulging fontanelle, scalp vein distension, and "sunsetting" of the eyes. In older children and adults, symptoms often include headache, nausea, vomiting, papilledema, diplopia, and progressive cognitive impairment.

Untreated hydrocephalus can lead to irreversible brain damage, visual loss, endocrine dysfunction, and severe disability.

Diagnostic Approaches

Neuroimaging plays a central role in diagnosis. Cranial ultrasound is useful in neonates with open fontanelles. Computed tomography (CT) offers rapid assessment in emergency settings, while magnetic resonance imaging (MRI) provides superior anatomical detail and helps identify the underlying cause of CSF obstruction. Phase-contrast MRI can evaluate CSF flow dynamics.

Complications

Complications of hydrocephalus include herniation syndromes, chronic intracranial hypertension, and neurodevelopmental delay in pediatric populations. Long-standing ventricular dilation may lead to periventricular white matter injury and permanent neurological deficits.

Conclusion

Hydrocephalus is a complex and potentially life-threatening disorder. Understanding its pathophysiology and clinical spectrum is essential for early diagnosis and effective management.

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