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Short Communication

Hydrocephalus and the Need for Continuous Personalized Care

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ABOUT THE STUDY

Hydrocephalus, initially described by Hippocrates around 400 B.C., occurs when Cerebrospinal Fluid (CSF) accumulates abnormally in the ventricles of the brain [1-5]. Worldwide, it is the most common condition that neurosurgeons address, with thousands of new cases arising in the United States each year [5-7]. Hydrocephalus is the most common brain disorder in children, though it can affect people of any age [5,7-9].

While hydrocephalus is accompanied by a variety of symptoms, including headaches, vomiting, and sensory disturbances, certain sequelae of hydrocephalus, such as motor dysfunction, cognitive impairment, and growth deficits, render patients in need of ongoing care [4,7,10,11]. The condition can occur for a variety of reasons, which leads to a diverse and complicated patient population that includes patients who face distinct challenges and prognoses [12,13].

Different forms of hydrocephalus contribute to a heterogeneous patient population

The term hydrocephalus captures several distinct conditions that vary in terms of how or when they lead to clinical dysfunction. Relevant factors include age, clinical drivers, and the presence or absence of obstructions.

Age: Hydrocephalus can present at any age. It is estimated that 1.1 out of every 1,000 infants suffer from hydrocephalus [8]. Though hydrocephalus was once considered a disease that primarily affected children, diagnostic techniques have helped to clarify its relatively high incidence rate in adults as well [9].

Congenital hydrocephalus: Congenital hydrocephalus is present from birth and may result from genetics or from events that occur in utero [13].

Acquired hydrocephalus: On the other hand, acquired hydrocephalus develops anytime in response to disease or injury [13]. These forms of hydrocephalus are often observed in response to hemorrhage, tumors, head trauma, or infection of the central nervous system [10].

Clinical drivers: While all cases of hydrocephalus involve a disruption to CSF homeostasis, the clinical mechanisms can be distinct.

Genetics: It is estimated that about 40% of hydrocephalus cases are linked to a genetic etiology, with changes at the molecular level driving hydrocephalus pathogenesis [11].

Hemorrhage or Trauma: Approximately 15% of preterm infants experience hemorrhaging in the ventricles, which leads to Post-Hemorrhagic Hydrocephalus (PHH), the most common neurological complication for those born prematurely [14]. PHH is also one of the most serious complications of Traumatic Brain Injury (TBI) as well as both subarachnoid hemorrhage and intraventricular hemorrhage in adults [10].

Infection: Post Infectious Hydrocephalus (PIH) occurs following infection and may present similarly to PHH as reparative inflammation damages tissue [6]. However, unlike with PIH, PHH may occur due to hemorrhaged blood and its metabolic products, such as blood clots [10-13].

Obstructions: Regardless of the underlying mechanisms, hydrocephalus is often defined by whether an obstruction is present [13,14]. Though in some cases, excess production of CSF causes hydrocephalus, the condition is most often the consequence of obstructed CSF flow and often leads to ventricle enlargement [15,16]. Acute hydrocephalus can occur when there is sudden obstruction of CSF pathways [17].

Communicating hydrocephalus: Communicating hydrocephalus refers to cases where no obstruction is present, and the lack of

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CSF homeostasis arises from increased CSF production or inadequate absorption [13,18].

Non-communicating hydrocephalus: Non-communicating hydrocephalus is the result of blocked CSF flow that leads to ventricle enlargement and enhanced pressure inside the skull. In the case of Normal Pressure Hydrocephalus (NPH), which is more often observed in the elderly, dilated ventricles occur despite no obstruction or enhanced pressure and are accompanied by normal hydrocephalus symptoms such as slow recall [13,19]. This form of hydrocephalus represents the first and one of the only forms of dementia that is treatable [20,21].

Conventional treatments are limited in their value

While shunting and surgery are the most common approaches to hydrocephalus, there are three ways that hydrocephalus can theoretically be treated [4,22]. First, the excessive CSF can be reversed by reducing CSF production through the deactivation of the choroid plexus, which generates CSF. Such deactivation could occur through surgical removal, radiation, or pharmacotherapy. Second, CSF resorption could be enhanced to reduce excess CSF, which could be accomplished by shunting CSF toward areas with low pressure. Third, the blocked pathways resulting from CSF accumulation in the brain can be unblocked through bypass, lesion, or surgical removal.

Unfortunately, conventional therapies for hydrocephalus are associated with complications, including infection and drainage tube obstruction [10]. Shunt complications are common, and shunting is limited in that it can only partially reverse the damage caused by hydrocephalus [16]. As hydrocephalus progresses, its effectiveness diminishes. Within one yar, more than 30% of shunts have failed, and within 10 years, more than 66% of shunts do not survive without revision [23,24]. In addition to the associated health-related challenges, shunt complications and comorbidities, also lead to problems with social functioning [25-28].

Appropriate ongoing care is required to reduce pain and suffering in hydrocephalus patients

Patients are never cured of hydrocephalus, and each patient faces a unique clinical course. It is thus imperative that each patient is matched with the appropriate type and level of care to address their individual clinical needs and reduce the risk of complications.

While surgeries themselves are relatively safe, between 5% and 15% of patients die within 10 years of surgery [29]. A significant proportion of these deaths are attributed to failures in diagnosis and treatment of shunt malfunctions. However, when rapidly identified, shunt malfunction complications can often be overcome successfully [13]. Timely intervention across the lifespan of those with hydrocephalus therefore improves patient outcomes and is also associated with significant cost savings [5,29-31].

CONCLUSION

In addition to the potential for life-saving interventions, ongoing care for those with hydrocephalus is also needed to address the challenges these patients experience. For example, most hydrocephalus patients have neurological deficits, with roughly 60% suffering motor impairments and 25% experiencing visual or auditory disturbances. As emerging data open possibilities for new therapies for hydrocephalus, such as stem cell therapies and gene therapies, it is important to think about how care will need to adjust to a new therapeutic landscape to support the safety and comfort of all hydrocephalus patients.

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