

Paradigm shift in hydrocephalus research in legacy of Dandy's pioneering work: rationale for third ventriculostomy in communicating hydrocephalus

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Abstract

Objective This study aims to question the generally accepted cerebrospinal fluid (CSF) bulk flow theory suggesting that the CSF is exclusively absorbed by the arachnoid villi and that the cause of hydrocephalus is a CSF absorption deficit. In addition, this study aims to briefly describe the new hydrodynamic concept of hydrocephalus and the rationale for endoscopic third ventriculostomy (ETV) in communicating hydrocephalus.

Critique The bulk flow theory has proven incapable of explaining the pivotal mechanisms behind communicating hydrocephalus. Thus, the theory is unable to explain why the ventricles enlarge, why the CSF pressure remains normal and why some patients improve after ETV.

Hydrodynamic concept of hydrocephalus Communicating hydrocephalus is caused by decreased intracranial compliance increasing the systolic pressure transmission into the brain parenchyma. The increased systolic pressure in the brain distends the brain towards the skull and simultaneously compresses the periventricular region of the brain against the ventricles. The final result is the predominant enlargement of the ventricles and narrowing of the subarachnoid space. The ETV reduces the increased systolic pressure in the brain simply by venting ventricular CSF through the stoma. The patent aqueduct in communicating hydrocephalus is too narrow to vent the CSF sufficiently.

Keywords Cerebrospinal fluid · Compliance · ETV · Hydrocephalus · Normal-pressure hydrocephalus · Pathophysiology · Pulse pressure

Introduction

The modern era of hydrocephalus research began with the experimental studies of Dandy and Blackfan [3] in 1914, which, until today constitutes an unsurpassed contribution to this field. By plugging the aqueduct, they produced hydrocephalus in dogs. The division of hydrocephalus into the obstructive and communicating type originates from their work. The bulk flow theory and the concept of cerebrospinal fluid (CSF) malabsorption as the cause of hydrocephalus are also based on their experiment [3]. However, Dandy strongly opposed the view that idiopathic communicating hydrocephalus is caused by an obstruction at the arachnoid villi. Such an obstruction cannot cause a higher pressure in the ventricles than in the subarachnoid space but would rather dilate the subarachnoid space. He also objected to the view that the CSF is absorbed by the arachnoid villi and instead proved that the CSF is absorbed by the capillaries [3, 4]. If this is true, the modern era of CSF and hydrocephalus research started and ended with Dandy, who left the pathophysiology of idiopathic communicating hydrocephalus unexplained for future exploration.

By going back to Dandy and following his lead of a capillary CSF absorption, a new understanding of hydrocephalus may be found. O'Connell [19] and Bering [1] suggested that increased pulse pressure in the ventricles is the cause of communicating hydrocephalus. In 1978, Di Rocco and Pettorossi [5] verified that an intraventricular pulsating balloon causes communicating hydrocephalus in sheep. In 1993, Greitz re-validated the physiology of the

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CSF circulation and hydrocephalus by using flow sensitive magnetic resonance imaging (MRI) and radionuclide cisternography [9]. He suggested that the capillaries absorb the CSF and that the distending force in the production of chronic hydrocephalus is an increased systolic pulse pressure in brain tissue [14].

The hydrodynamic concept of hydrocephalus

Classification of hydrocephalus The classification is based on capillary absorption of the CSF [9–14]. Hydrocephalus is divided into two main groups, acute hydrocephalus and chronic hydrocephalus [14]. Acute hydrocephalus is caused by an intraventricular CSF obstruction in accordance with the conventional view. As opposed to that view, it is suggested that chronic hydrocephalus is caused by decreased intracranial compliance.

Causes of decreased intracranial compliance Although the etiology of the decreased intracranial compliance such as CSF obstructions at the foramen magnum and adhesions in the subarachnoid space after intracranial bleedings, trauma, operations and infections is well known, the etiology remains unknown in many idiopathic and congenital cases of hydrocephalus. However, the final stage of chronic hydrocephalus, independent of its etiology, is uniform. The final stage is characterised by narrow capacitance vessels and reduced cerebral blood volume. This reduces cerebral blood flow, reduces intracranial compliance and enhances the intracerebral pulse pressure.

Intracerebral pulse pressure theory of chronic hydrocephalus Chronic hydrocephalus consists of two subtypes, communicating hydrocephalus and chronic obstructive hydrocephalus [14]. The associated malabsorption of CSF is not involved as a causative factor in chronic hydrocephalus. The theory is based on one basic mechanism, i.e. that chronic hydrocephalus is caused by decreased intracranial compliance increasing the systolic pressure transmission into the brain parenchyma [9, 10, 12–14]. The increased systolic pressure in the brain is the cause of the distension of the brain and ventricles. The systolic expansion of the brain distends the brain towards the skull and simultaneously compresses the periventricular region of the brain. The rationale result is the predominant enlargement of the ventricles and a narrowing of the subarachnoid space.

Rationale for ETV in communicating hydrocephalus As described above, communicating hydrocephalus is caused by decreased compliance increasing the systolic pressure transmission into the brain. The systolic force compresses the brain including the intracranial capacitance vessels, i.e.

the cerebral veins and capillaries. This results in a vicious cycle with narrow capacitance vessels and significantly reduced cerebral blood volume that causes further decrease in intracranial compliance and further increase in intracranial pulse pressure. It is obvious that an endoscopic third ventriculostomy (ETV) may interrupt the vicious cycle and reduce the systolic pressure in the brain simply by venting ventricular CSF through the stoma. The patent aqueduct in communicating hydrocephalus is too narrow to vent the ventricular CSF sufficiently. By reducing the pulse pressure, the ETV also reduces the compression of the capacitance vessels thereby restoring some degree of venous compliance, which in turn reduces the intracerebral pulse pressure further.

ETV versus shunting The proposed concept opens a new avenue in that ETV may be an effective treatment also in communicating hydrocephalus [8, 16–18]. It thus constitutes an interchangeable alternative to shunting. The primary aim of the treatment of chronic hydrocephalus is to restore intracranial compliance, which is achieved by both treatments [14]. Effective shunting is based on a slight over-drainage of CSF that ultimately causes a direct and forced dilation of the compressed veins [14]. Therefore, shunting is more effective in increasing intracranial compliance than ETV where the dilating effect on the veins works indirectly by reducing the systolic compressing force on the veins. Because ETV provides a more physiological treatment and has less late complications than shunting, it may be of benefit to increase the number of patients finally treated by ETV [2, 6–8, 15, 17, 18, 20]. In patients shunted under 1 year of age, ETV should be considered as an alternative treatment when they grow older. However, the final answer as to which is the optimal treatment must be based on randomized clinical studies.

Discussion

The aim of this paper is to briefly describe the hydrodynamic concept of hydrocephalus, which is based on Dandy's observation of capillary absorption of the CSF. Oi and Di Rocco [20] recently suggested that a minor CSF pathway through the brain parenchyma is the dominant CSF absorption site in the embryo, foetus and infant. The theory described here suggests that the minor pathway in the developing immature brain as well as the major pathway in adults are through the brain capillaries. It is beyond the scope of this brief communication to present the numerous physiological (and self-evident) evidences that the brain capillaries have the capacity to absorb fluids such as the CSF and its constituencies [3, 4, 9, 11, 12, 14]. This

contrasts to the total lack of physiological evidence that the arachnoid villi can absorb the CSF. The CSF dynamics has been described in earlier published papers [9, 11, 12, 14].

The proposed concept indicates a paradigm shift in our view on hydrocephalus. It is suggested that chronic hydrocephalus is caused by decreased intracranial compliance rather than a CSF absorption deficit [14]. The associated CSF malabsorption is a secondary phenomenon to the decreased intracranial compliance. The only residue of the conventional view is acute hydrocephalus, which is caused by a CSF absorption deficit (of the periventricular capillaries). Normal-pressure hydrocephalus, aqueductal stenosis as well as communicating hydrocephalus in infants and adults are typical manifestations within the spectrum of chronic hydrocephalus. It should be emphasised that chronic hydrocephalus is an unstable hemodynamic disorder occurring at normal or slightly increased mean intracranial pressure with superimposed vascular pressure waves that can be fatal.

The theory may thus have identified decreased intracranial compliance as a new and potential universal cause of chronic hydrocephalus. Decreased compliance is common to all different types of chronic hydrocephalus whatever their etiology. If the theory is correct, there is hope for improved treatment of chronic hydrocephalus of unknown etiology. How come then that treatment failure rather is the rule than the exception of this devastating condition? The reason may be that shunting and ETV essentially represents symptomatic treatments of hydrocephalus. To improve therapy, we have to find treatments that are more effective in increasing compliance in cases with unknown etiology or are directed against the cause of the decreased compliance in cases with known etiology.

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