Idiopathic intracranial hypertension pathogenesis

see also Intracranial hypertension pathogenesis.

Despite expanding research, idiopathic intracranial hypertension treatment remains challenging.

The failure to develop effective treatment strategies is largely due to poor agreement on a coherent disease pathogenesis model.

Conceptual Review

Fargen et al. provide a hypothesis of a unifying model centered around the internal jugular veins (IJV) to explain the development of IIH, which contends the following: (1) the IJV are prone to both physiological and pathological compression throughout their course, including compression near C1 and the styloid process, dynamic muscular/carotid compression from C3 to C6, and lymphatic compression; (2) severe dynamic internal jugular vein stenosis with developments of large cervical gradients is common in IIH-spectrum patients and significantly impacts intracranial venous and cerebrospinal fluid (CSF) pressures; (3) pre-existing IJV stenosis may be exacerbated by infectious/inflammatory etiologies that induce retromandibular cervical lymphatic hypertrophy; (4) extra-jugular venous collaterals dilate with chronic use but are insufficient resulting in impaired aggregate cerebral venous outflow; (5) poor IJV outflow initiates, or in conjunction with other factors, contributes to intracranial venous hypertension and congestion leading to higher CSF pressures and intracranial pressure (ICP); (6) glymphatic congestion occurs but is insufficient to compensate and this pathway becomes overwhelmed; and (7) elevated intracranial CSF pressures triggers extramural venous sinus stenosis in susceptible individuals that amplifies ICP elevation producing severe clinical manifestations. Future studies must focus on establishing norms for dynamic cerebral venous outflow and IJV physiology in the absence of disease so that we may better understand and define the diseased state [1].

1)
Fargen KM, Midtlien JP, Margraf CR, Hui FK. Idiopathic intracranial hypertension pathogenesis: The jugular hypothesis. Interv Neuroradiol. 2024 Aug 8:15910199241270660. doi: 10.1177/15910199241270660. Epub ahead of print. PMID: 39113487.