Disproportionately enlarged subarachnoid space hydrocephalus

- Idiopathic Normal-Pressure Hydrocephalus Revealed by Systemic Infection: Clinical Observations of Two Cases
- Deformation of brain in normal pressure hydrocephalus is more readily associated with slow vasomotion rather than heartbeat related pulsations of intracranial pressure
- Evaluation of disproportionately enlarged subarachnoid-space hydrocephalus in progressive supranuclear palsy
- Radiographic Evaluation of Normal Pressure Hydrocephalus
- Management of Normal Pressure Hydrocephalus
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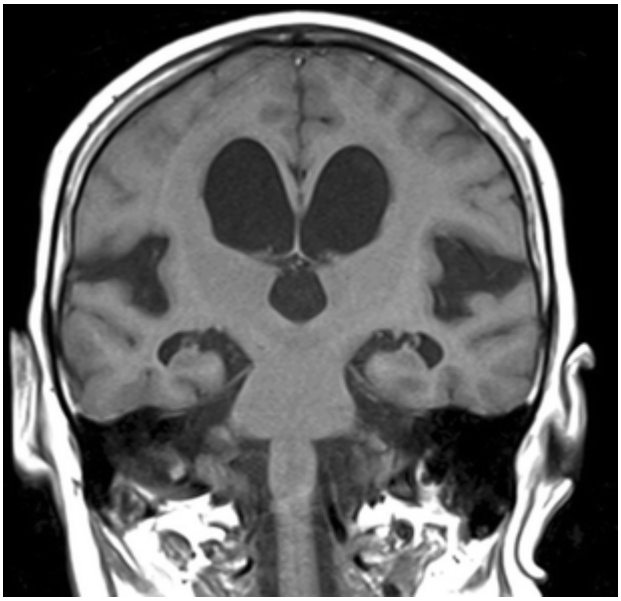
- Prevalence of idiopathic normal pressure hydrocephalus in patients with degenerative cervical myelopathy
- Regional brain volume changes in Hakim's disease versus Alzheimer's and mild cognitive impairment

see also see also Callosal angle, Cingulate sulcus sign, Evans index.



The presence of disproportionately enlarged subarachnoid space hydrocephalus (DESH) on brain imaging is a recognized finding of idiopathic normal pressure hydrocephalus (iNPH), but the features of DESH can vary across patients.

A high response rate to treatment can be achieved by good patient selection. Positive prognostic markers for therapeutic success include Disproportionately Enlarged Subarachnoid Space Hydrocephalus (DESH), short disease duration, predominant gait disturbance, and few comorbidities [1)]

Results indicate that findings of enlarged basal cisterns and sylvian fissures and of focally dilated sulci support, rather than exclude, the diagnosis of shunt-responsive idiopathic NPH and suggest that this condition is caused by a suprasylvian subarachnoid block [2)].

Tight high-convexity and medial subarachnoid spaces, and enlarged Sylvian fissures with ventriculomegaly, defined as disproportionately enlarged subarachnoid-space hydrocephalus (DESH),

are worthwhile for the diagnosis of iNPH [3].

Lipocalin-type prostaglandin D synthaseL (PGDS) might work as a surrogate marker for DESH features, white matter damage, and frontal lobe dysfunction [4].

Score

see DESH score.

Case series

Data from 1,356 individuals were analyzed, and 25 (1.8%) individuals had DESH. Regarding the relationships between the volume of each CSF space and age, ventricles [VS], and Sylvian fissure [SF] volumes increased with age, whereas subarachnoid spaces at high convexity and midline [SHM]) volume did not increase. VS and SF volumes increased as the whole brain volume decreased, whereas SHM volume did not increase even if the whole brain volume decreased; that is, SHM did not expand even if brain atrophy progressed. Moreover, lower Mini-Mental State Examination scores were significantly associated with lower SHM volume and higher VS volume. These associations remained significant even when individuals with DESH were excluded.

This study showed that the volume of high-convexity and medial subarachnoid spaces did not expand and tended to decrease with age; the human brain continuously progresses toward a “DESH-like” morphology with aging in community-dwelling older persons (i.e., DESH might be an “accelerated aging stage” rather than an “age-related disorder”). Our results indicated that brain atrophy may be associated with the development of “DESH-like” morphology. In addition, this morphological change, as well as brain atrophy, is an important condition associated with cognitive decline in older adults. Our findings highlight the importance of investigating the aging process of CSF dynamics in the human brain to preserve brain health in older people [5].

Subjects aged 60 and over in a memory clinic and a community-based cohort were assessed for the presence of ventriculomegaly, Sylvian fissure dilatation, and high convexity tightness by neuroimaging, and a clinical triad of iNPH symptoms, i.e. cognitive, gait and urinary symptoms.

In the memory clinic-based study (548 subjects), the prevalence of DESH was 1.1% and increased with age. The clinical triad was significantly more frequent in subjects with DESH (50%) compared to those with normal images (none), Sylvian dilatation (7%), and ventriculomegaly (12%). Gait disturbance was also significantly more frequent in DESH (83%) compared to those with normal images (2%), Sylvian dilatation (14%), and ventriculomegaly (26%). In the community-based cohort (946 subjects), the prevalence of DESH was 1.0% and increased with age. The clinical triad (11%) was significantly more common in subjects with DESH compared to those with normal images (none), Sylvian dilatation (2%), and ventriculomegaly (7%). Gait disturbance was also significantly more common in DESH (33%) compared to those with normal images (1%), Sylvian dilatation (4%), and ventriculomegaly (10%).

The reported prevalence of DESH was approximately 1% and increased with age. DESH and high

convexity tightness were specifically associated with the clinical triad of iNPH. Of the triad, gait disturbance was associated to DESH and high convexity tightness [6].

2016

Radovnický et al., analysed 1.5-T MRI scans of patients fulfilling the criteria of probable or possible iNPH and positive supplementary tests before and after surgery (ventriculo-peritoneal shunt). FA was measured in the anterior and posterior limb of the internal capsule (PLIC) and in the corpus callosum. Patients were divided into the Disproportionately enlarged subarachnoid space hydrocephalus (DESH) and non-DESH group. These data were also compared to FA values in the control group.

Twenty-seven patients and 24 healthy controls were enrolled. DESH was present in 15 patients and lacking in 12. Twenty-three iNPH patients were shunt responders (85.2 %), and 4 were non-responders (14.8 %). All patients in the DESH group were shunt responders. In the non-DESH group, eight patients were responders (66.7 %). A significant difference between the DESH and non-DESH group was found in the FA of the PLIC. The mean value of FA in the PLIC was 0.72 in the DESH group and 0.66 in the non-DESH group. After the surgery FA decreased in both groups. In the DESH iNPH group FA PLIC decreased to 0.65 and in the non-DESH iNPH group to 0.60. In the healthy controls, the mean FA in the PLIC was 0.58.

DESH on MRI scans is related to a higher FA in the PLIC with a decrease after the surgery. It reflects a more severe compression of the white matter than in non-DESH patients or healthy volunteers. DESH patients had better outcome than non-DESH patients. This study confirmed the importance of DESH as a supportive sign for iNPH [7].

2014

Eight participants with DESH-iNPH (1.6%) and 76 with ex vacuo hydrocephalus (16.1%) at baseline were identified. The mean MMSE in DESH-iNPH, ex vacuo hydrocephalus, and normal MRIs was 26.4, 27.9, and 28.3, respectively, and the mean UPDRSM was 9.75, 2.96, and 1.87, respectively. After a 90-month follow-up, the mortality rates for DESH-iNPH, ex vacuo hydrocephalus, and normal MRIs were 25.0%, 21.3%, and 10.9%, respectively. The perivascular-space widening scores were significantly smaller in the DESH-iNPH cases, particularly at the centrum semiovale, compared to cerebral small-vessel disease and ex vacuo hydrocephalus cases.

The prevalence of DESH-iNPH was 1.6% for participants aged 75 years and revealed significantly lower MMSE and higher UPDRSM scores compared to the ex vacuo hydrocephalus and controls. Moreover, it is suggested that perivascular-space narrowing is a morphological and pathophysiological marker of DESH-iNPH [8].

References

1)
Illies T, Eckert B, Kehler U. What Radiologists Should Know About Normal Pressure Hydrocephalus. Rofo. 2021 Oct;193(10):1197-1206. English, German. doi: 10.1055/a-1425-8065. Epub 2021 Sep 16. PMID: 34530457.
2)
Kitagaki H, Mori E, Ishii K, Yamaji S, Hirono N, Imamura T. CSF spaces in idiopathic normal pressure

hydrocephalus: morphology and volumetry. AJNR Am J Neuroradiol. 1998 Aug;19(7):1277-84. PubMed PMID: 9726467.

3) Hashimoto M, Ishikawa M, Mori E, Kuwana N; Study of INPH on neurological improvement (SINPHONI). Diagnosis of idiopathic normal pressure hydrocephalus is supported by MRI-based scheme: a prospective cohort study. Cerebrospinal Fluid Res. 2010 Oct 31;7:18. doi: 10.1186/1743-8454-7-18. PubMed PMID: 21040519; PubMed Central PMCID: PMC2987762.

4) Nishida N, Nagata N, Toda H, Jingami N, Uemura K, Ozaki A, Mase M, Urade Y, Matsumoto S, Iwasaki K, Ishikawa M. Association of lipocalin-type prostaglandin D synthase with disproportionately enlarged subarachnoid-space in idiopathic normal pressure hydrocephalus. Fluids Barriers CNS. 2014 Apr 15;11(1):9. doi: 10.1186/2045-8118-11-9. PubMed PMID: 24731502; PubMed Central PMCID: PMC3991874.

5) Hidaka Y, Hashimoto M, Suehiro T, Fukuhara R, Ishikawa T, Tsunoda N, Koyama A, Honda K, Miyagawa Y, Yoshiura K, Boku S, Ishii K, Ikeda M, Takebayashi M. Impact of age on the cerebrospinal fluid spaces: high-convexity and medial subarachnoid spaces decrease with age. Fluids Barriers CNS. 2022 Oct 28;19(1):82. doi: 10.1186/s12987-022-00381-5. PMID: 36307853; PMCID: PMC9615391.

6) Akiba C, Gyanwali B, Villaraza S, Nakajima M, Miyajima M, Cheng CY, Wong TY, Venketasubramanian N, Hilal S, Chen C. The prevalence and clinical associations of disproportionately enlarged subarachnoid space hydrocephalus (DESH), an imaging feature of idiopathic normal pressure hydrocephalus in community and memory clinic based Singaporean cohorts. J Neurol Sci. 2019 Oct 25;408:116510. doi: 10.1016/j.jns.2019.116510. [Epub ahead of print] PubMed PMID: 31810041.

7) Radovnický T, Adámek D, Derner M, Sameš M. Fractional anisotropy in patients with disproportionately enlarged subarachnoid space hydrocephalus. Acta Neurochir (Wien). 2016 Aug;158(8):1495-500. doi: 10.1007/s00701-016-2861-x. Epub 2016 Jun 8. PubMed PMID: 27272943.

8) Akiguchi I, Shirakashi Y, Budka H, Watanabe Y, Watanabe T, Shiino A, Ogita M, Kawamoto Y, Jungwirth S, Krampla W, Fischer P. Disproportionate subarachnoid space hydrocephalus-outcome and perivascular space. Ann Clin Transl Neurol. 2014 Aug;1(8):562-9. doi: 10.1002/acn3.87. Epub 2014 Jul 28. PubMed PMID: 25356428; PubMed Central PMCID: PMC4184559.