

Hydrocephalus Classification

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There is no international [consensus](#) on the [classification](#) of [hydrocephalus](#). There are various systems based on the age of onset, [cerebrospinal fluid dynamics](#) and anatomical area of accumulation, the levels of [cerebrospinal fluid pressure](#) and the presence of [symptoms](#).

However, no definitive classification exists comprehensively to cover the variety of these aspects.

Wu et al. proposed a classification Based on Ventricular Pressure ¹⁾.

General information

Hydrocephalus (HCP) is either due to subnormal [cerebrospinal fluid reabsorption](#) or, rarely [cerebrospinal fluid overproduction](#).

● subnormal CSF reabsorption. Two main functional subdivisions:

1. [obstructive hydrocephalus](#) (AKA non-communicating): block proximal to the [arachnoid granulations](#) (AG). On CT or MRI: enlargement of [ventricles](#) proximal to block (e.g. obstruction of [aqueduct of Sylvius](#) → lateral and 3rd ventricular enlargement out of proportion to the 4th ventricle, sometimes referred to as [triventricular hydrocephalus](#)).

see also [Posthemorrhagic hydrocephalus](#)

2. [communicating hydrocephalus](#) (AKA non-obstructive): defect in CSF reabsorption by the AG

● [Cerebrospinal fluid overproduction](#).

[Walter Edward Dandy](#) first described the basic mechanism and classification of [hydrocephalus](#) as:

[Obstructive hydrocephalus](#) or [Non Obstructive hydrocephalus](#).

Despite advances in understanding of the underlying process, current [classification](#) systems still rely upon Dandy's classification scheme ²⁾.

The aim of the study was to evaluate the diagnostic utility of three-dimensional sampling perfection with application optimized contrast using different flip angle evolution (3D SPACE) sequence and Susceptibility Weighted Imaging (SWI) in hydrocephalus and to propose a refined definition and classification of hydrocephalus with relevance to the selection of treatment option.

A prospective study of 109 patients with hydrocephalus was performed with magnetic resonance imaging (MRI) brain using standardized institutional sequences along with additional sequences 3D SPACE and SWI. The images were independently read by two senior neuroradiologists and the etiopathogenesis of hydrocephalus was arrived by consensus.

With conventional sequences, 46 out of 109 patients of hydrocephalus were diagnosed as obstructive of which 21 patients showed direct signs of obstruction and 25 showed indirect signs. In the remaining 63 patients of communicating hydrocephalus, cause could not be found out in 41 patients. Whereas with 3D SPACE sequence, 88 patients were diagnosed as obstructive hydrocephalus in which all of them showed direct signs of obstruction and 21 patients were diagnosed as communicating hydrocephalus. By including SWI, we found out hemorrhage causing intraventricular obstruction in three patients and hemorrhage at various sites in 24 other patients. With these findings, we have classified the hydrocephalus into communicating and noncommunicating, with latter divided into intraventricular and extraventricular obstruction, which is very well pertaining to the selection of surgical option.

Chellathurai et al., strongly suggest to include 3D SPACE and SWI sequences in the set of routine MRI sequences, as they are powerful diagnostic tools and offer complementary information regarding the precise evaluation of the etiopathogenesis of hydrocephalus and have an effective impact in selecting the mode of management ³⁾.

Terms used

[Acquired hydrocephalus](#)

[Acute hydrocephalus](#)

[Adult hydrocephalus](#)

[Arrested hydrocephalus](#) or [Compensated hydrocephalus](#)

[Chronic hydrocephalus](#)

[Communicating hydrocephalus](#) or [Non obstructive hydrocephalus](#)

[Congenital hydrocephalus](#)

[External hydrocephalus](#)

[Focal hydrocephalus](#)

[Hydrocephalus Ex Vacuo](#)

[Idiopathic normal pressure hydrocephalus](#)

[Infantile hydrocephalus](#) or [Pediatric hydrocephalus](#).

[Internal hydrocephalus](#)

[Low-pressure hydrocephalus](#)

[Non obstructive hydrocephalus](#) or [Communicating hydrocephalus](#)

[Normal pressure hydrocephalus](#) for [Idiopathic normal pressure hydrocephalus](#) or [Secondary normal pressure hydrocephalus](#).

[Obstructive hydrocephalus](#).

[Pediatric hydrocephalus](#) or [Infantile hydrocephalus](#)

[Secondary normal pressure hydrocephalus](#).

[Shunt-dependent hydrocephalus](#)

[Unilateral hydrocephalus](#).

With the rare exception of hydrocephalus associated with overproduction of CSF in patients with [choroid plexus papillomas](#) (CPPs), all hydrocephalus is basically [obstructive hydrocephalus](#). That the rare CPP causes hydrocephalus is not debated, but why it does so is the subject of some discussion. CPPs are known to lead to increases in the rate of CSF production and are known to cause hydrocephalus.

Normal CSF absorptive mechanisms can clear the amount of spinal fluid produced in the ventricular system at extremely high rates without producing [ventriculomegaly](#). If CSF production and ICP increase substantially, ventricular size increases ⁴⁾. When CSF flow is obstructed in the context of increased CSF production, there is a great tendency for [ventriculomegaly](#) or hydrocephalus to develop. CPPs, in themselves, can create the only pure form of [communicating hydrocephalus](#). However, that these tumors tend to be large and to restrict CSF flow through the [foramen of Monro](#) or [aqueduct of Sylvius](#), is more likely to account for the severity of hydrocephalus in this context ⁵⁾.

When hydrocephalus is severe, especially in the very young, it may not be possible to define the point of CSF obstruction without introducing tracers into the CSF pathways. In patients treated early in life whose ventricles have become smaller with treatment, it is possible to determine the first site of obstruction to CSF flow on MRI or CT.

Patients with complex congenital anomalies such as hydrocephalus related to a Chiari II malformation and spina bifida often have multiple sites of obstruction ^{6) 7)}. It may not be possible to predict a second or downstream point of obstruction. In these patients, only one point may be obstructed or all of these sites may be restricted.

Based on analyses from a mathematical modeling, of the work on the circuitry of CSF flow, and these potential sites of obstruction, Rekate et al., proposed a classification

Site of Obstruction	Pathology	Treatment
None	Choroid plexus papilloma	Removal
Foramen of Monro	Tumor, congenital anomaly, postshunt ventricular asymmetry	Tumor removal, septum pellucidotomy, ventricle shunt
Aqueduct of Sylvius	Congenital lesion, tumor secondary to extraventricular obstruction	ETV, ventricular shunting
Outlets of fourth ventricle	Chronic meningitis, Chiari II malformation	ETV, ventricular shunting
Basal cisterns	Meningitis, post subarachnoid hemorrhage	ETV, ventricle shunt, spinal thecal shunt
Arachnoid granulations	Hemorrhage or infection in infancy	Ventricle or thecal shunt
Venous outflow	Skull base anomalies, congenital heart disease	Ventricle or thecal shunt, treatment of vascular anomaly if possible

It is generally assumed that endoscopic third ventriculostomy (ETV) is only effective for treating obstructive hydrocephalus, and many assume that obstructive hydrocephalus is synonymous with aqueductal stenosis. The growing number of reports on the efficacy of ETV for treating “communicating hydrocephalus” has generated considerable consternation ⁸⁾.

The “Multi-categorical Hydrocephalus Classification” (Mc HC), was invented and developed to cover the entire aspects of hydrocephalus with all considerable classification items and categories.

Ten categories include “Mc HC” category I: onset (age, phase), II: cause, III: underlying lesion, IV: symptomatology, V: pathophysiology 1-CSF circulation, VI: pathophysiology 2-ICP dynamics, VII: chronology, VIII: post-shunt, IX: post-endoscopic third ventriculostomy, and X: others. From a 100-year search of publication related to the classification of hydrocephalus, 14 representative publications were reviewed and divided into the 10 categories.

The Baumkuchen classification graph made from the round o'clock classification demonstrated the historical tendency of deviation to the categories in pathophysiology, either CSF or ICP dynamics.

In the preliminary clinical application, it was concluded that “Mc HC” is extremely effective in expressing the individual state with various categories in the past and present condition or among the compatible cases of hydrocephalus along with the possible chronological change in the future ⁹⁾.

References

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