The cause is unknown, but many patients have a family history consistent with autosomal dominant inheritance.

Several pathogenic hypotheses have been proposed but the most accredited theory seems to be delayed maturation of the arachnoid villi. There is a consensus that this is a benign entity, correlated to a familial predisposition and, in some cases, inheritance.

Secondary cases of external hydrocephalus are associated with hemorrhage, meningitis, and elevated venous pressure.

Elevated venous pressure has been shown to be a much more common cause of communicating hydrocephalus in children than previously thought.

The absorption of CSF in infants is into the capillary bed of the deep white matter rather than the arachnoid granulations. Absorption into a capillary bed depends on hydrostatic pressure. Similar to older children with communicating hydrocephalus, the infants in this cohort with external hydrocephalus showed evidence of an elevation in venous pressure. Elevated venous pressure may be a much more common cause of external hydrocephalus than previously recognized [1)].

The hypothesis that encephalocranial disproportion is the basic underlying entity for the CT images was proposed [2)].

Transplacental acquisition of anti-Ro antibodies has been associated with external hydrocephalus [3)].

1)
Bateman GA, Napier BD. External hydrocephalus in infants: six cases with MR venogram and flow quantification correlation. Childs Nerv Syst. 2011 Dec;27(12):2087-96. doi: 10.1007/s00381-011-1549-z. Epub 2011 Aug 11. PubMed PMID: 21833725.
2)
Nogueira GJ, Zaglul HF. Hypodense extracerebral images on computed tomography in children. “External hydrocephalus”: a misnomer? Childs Nerv Syst. 1991 Oct;7(6):336-41. PubMed PMID: 1764709.
3)
Edwards RJ, Allport TD, Stoodley NG, O'Callaghan F, Lock RJ, Carter MR, Ramanan AV. External hydrocephalus and subdural bleeding in infancy associated with transplacental anti-Ro antibodies. Arch Dis Child. 2012 Apr;97(4):316-9. doi: 10.1136/adc.2010.194746. Epub 2011 Apr 1. PubMed PMID: 21459880.