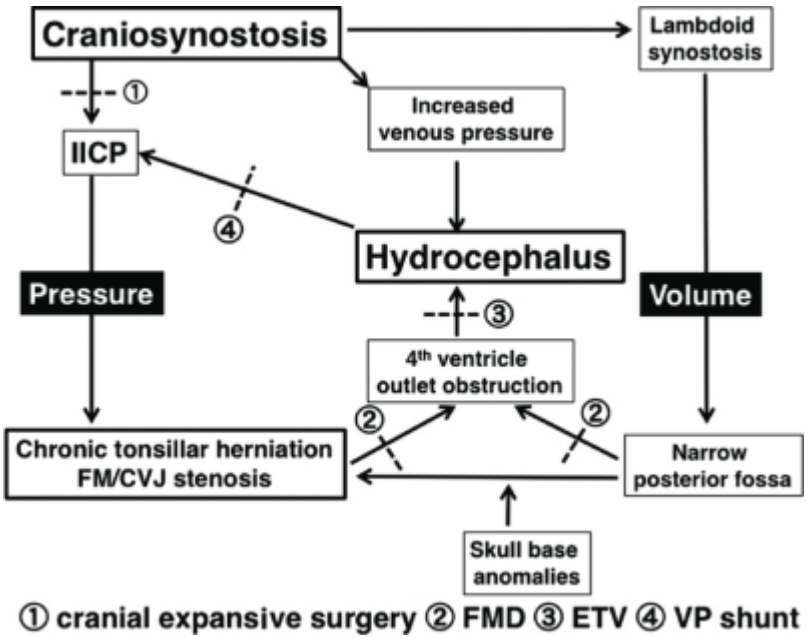
Craniosynostosis pathophysiology



Without adequate evaluation of pathophysiology, cranial vault expansion shows limited ability to compensate intracranial hypertension, as long as there is persistence of other causative factors of the raised ICP, such as hydrocephalus, impairment of CSF circulation or venous sinus circulation and upper airway obstruction [1].

Reduced intracranial volume (ICV) and raised intracranial pressure (ICP) are assumed to be principal pathophysiological mechanisms in childhood craniosynostosis.

A study examined the association between ICV and ICP and whether ICV can be used to estimate the ICP.

Langvatn et al., analyzed ICV and ICP measurements from children with craniosynostosis without concurrent hydrocephalus and from age-matched individuals without craniosynostosis who underwent diagnostic ICP measurement.

The study included 19 children with craniosynostosis (mean age 2.2 ± 1.9 years) and 12 reference individuals without craniosynostosis (mean age 2.5 ± 1.6 years). There was no difference in ICV between the patient and reference cohorts. Both mean ICP (17.1 ± 5.6 mm Hg) and mean wave amplitude (5.9 ± 2.6 mm Hg) were higher in the patient cohort. The results disclosed no significant association between ICV and ICP values in the patient or reference cohorts, and no association was seen between change in ICV and ICP values after cranial vault expansion surgery (CVES) in 5 children in whom ICV and ICP were measured before and after CVES.

In this cohort of children with craniosynostosis, there was no significant association between ICV and ICP values prior to CVES and no significant association between change in ICV and ICP values after CVES in a subset of patients. Therefore, ICV could not reliably estimate the ICP values. The authors suggest that intracranial hypertension in childhood craniosynostosis may not be caused by reduced ICV alone but rather by a distorted relationship between ICV and the volume of intracranial content (brain tissue, CSF, and blood) [2].

References

1)

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2)

Langvatn EA, Frič R, Due-Tønnessen BJ, Eide PK. Intracranial volume versus static and pulsatile intracranial pressure values in children with craniosynostosis. J Neurosurg Pediatr. 2019 Apr 19:1-9. doi: 10.3171/2019.2.PEDS18767. [Epub ahead of print] PubMed PMID: 31003225.