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The story of "asymptomatic ventriculomegaly" – Normal pressure hydrocephalus of infancy and childhood

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Introduction: Ventriculomegaly with or without a shunt or persisting after endoscopic third ventriculostomy (ETV) without obvious signs or symptoms of raised intracranial pressure is most often interpreted as non-treatment dependent hydrocephalus, shunt independency or successful ETV. However, we hypothesize, that in the majority of cases normal pressure hydrocephalus with increased nocturnal ICP dynamics and decreased compliance is present.

Methods: 40 children fulfilling the above named criteria underwent either computerized overnight ICP monitoring after implantation of an ICP sensor or a computerized shunt or reservoir infusion study. Increased nocturnal ICP dynamics, increase ICP peak pressures and amplitudes, indexes of decreased reserve capacity/compliance are findings indicating a pathophysiological situation of low compliance.

Results: In 32 of 40 patients clearly abnormal patterns/results were found. Children either receives shunt implantation or shunt revision. In the majority of cases parents reported an improvement like better cognitive performance, less irritability, better sleep, or loss of headaches of their children after shunt revision/implantation.

Conclusion: Pediatric normal pressure hydrocephalus exists and is associated with ventriculomegaly in the absence of classical signs and symptoms of raised ICP. Under this circumstances the common definition of ETV success = absence of shunt appears in a different light. The persistence of a low compliance situation over years results in subtle motor problems or cognitive decline, or if persistent into adulthood, early NPH development. Pediatric NPH should not be observed but treated as consequently as pressure active hydrocephalus, since modern gravitational shunt systems have largely abolished the problems of overdrainage. Therefore negative affects of untreated hydrocephalus override negative side effects of shunting.