Transtentorial Distortion Syndrome

Complex hydrocephalus affecting lateral ventricle and fourth ventricles separately is occasionally managed with CSF diversion via supra- and infra-tentorial ventricular catheters. The optimal configuration to reduce complications is currently unknown in adults.

Craven et al. describe a consistently similar clinical presentation of patients with complex hydrocephalus and a fourth ventricle separately drained by infra-tentorial shunt insertion.

A retrospective single centre case series. Medical notes were reviewed for clinical presentation, brain imaging and neurophysiological tests results. All patients underwent ICP monitoring (ICPM). Outcomes were determined by ventricular appearance on brain imaging CT and symptomatic improvements post-operatively.

Five adult patients referred to the hydrocephalus service had separate infra and supra-tentorial shunt systems. A common clinical presentation was observed, including lower motor neuron facial palsy (confirmed with electrophysiology), ophthalmoplegia, dysarthria, impaired gait headache and nausea. We refer to this as Transtentorial Distortion Syndrome (TDS). 24-hour ICPM demonstrated clear low pressures. All patients underwent shunt revision connecting the trans-tentorial shunts via a Y-connector and the addition of a distal valve. All subjects had improved ventricular appearance on CT scans post revision and normalisation of ICPM was observed. In the follow up period of 6 months no patient required further shunt revision.

To prevent TDS, supra- and infra-tentorial shunt constructs in adults with encysted fourth ventricles should be similar to the shunt systems widely known in the paediatric population with Dandy Walker syndrome, i.e. joint output to a single valve distal to the connection of the 2 proximal drainage catheters [1].

1)
Craven CL, Baudracco I, Thompson SD, Thorne L, Watkins LD, Toma AK. Transtentorial Distortion Syndrome: A consistent complication following lateral and fourth ventricular shunting in adults. World Neurosurg. 2017 Nov 15. pii: S1878-8750(17)31955-1. doi: 10.1016/j.wneu.2017.11.032. [Epub ahead of print] PubMed PMID: 29155115.