Neurofibromatosis type 1 related hydrocephalus

Neurofibromatosis type 1 (NF1) is associated with hydrocephalus in up to 13%. Currently, there is very little literature describing the actual etiologies and treatment options of NF1-associated hydrocephalus. Roth et al., therefore describe the experience in treating NF1-associated hydrocephalus.

They completed a retrospective data analysis of 1020 NF1 patients treated at the Gilbert’s Israeli International Neurofibromatosis Center (GIINFC) Tel Aviv over a period of 20 years. Patients presenting and treated for related hydrocephalus were included. Clinical, radiological, and surgical data are presented.

Twenty-two patients (2.1% of entire NF1 patient cohort) were included, with 17 under 19 years of age. Twenty had obstructive hydrocephalus. Most common etiologies included aqueductal or third ventricular obstruction. Fifteen patients underwent endoscopic procedures (14 third ventriculostomies), and 7 underwent shunt procedures. Failure rates (including a need for additional CSF procedures) were 60% and 71% respectively.

Hydrocephalus in the context of NF1 is caused mostly by obstructive etiologies. A tailored treatment approach is recommended, addressing the specific etiology. Regardless of the treatment approaches, a relatively high rate of failures is described.