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ORIGINAL ARTICLE

Endoscopic third ventriculostomy in children with third ventricular pressure gradient and open ventricular outlets on MRI

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Abstract

Objective Patients with non-communicating hydrocephalus due to aqueductal stenosis are often successfully treated with endoscopic third ventriculocisternostomy (ETV). In hydrocephalus, due to other locations of obstruction of the major CSF pathways, endoscopic treatment may also be a good option. We investigated our cohort of patients treated by ETV with patent ventricular outflow but pressure gradient signs at the third ventricle in a single-center retrospective study.

Methods We retrospectively reviewed records and imaging studies of 137 patients who underwent an ETV in our department in the time period of June 2010 to March 2018. We included patients who showed the following findings in MRI: 1st: open Sylvian aqueduct, 2nd: open outlets of the 4th ventricle, 3rd: open spinal canal, 4th: intra-/extraventricular pressure gradient seen at the 3rd ventricle and excluded patients with history of CSF infection or hemorrhage. Perioperative clinical state and possible complications or reoperations were recorded. Shunt dependency and changes in ventricular dilatation were measured as frontal and occipital horn ratio (FOHR) before surgery and during follow-up.

Results A total of 21 patients met the defined criteria. During the mean follow-up time of 40.7 ± 30 months (range; 5–102 months), two children had to undergo a re-ETV, and six children (all < 1 year of age) received a VP shunt. ETV shunt-free survival was 100% for children > 1 year of age. The ventricular width measured as FOHR was significantly reduced after ETV 0.5 ± 0.08 (range 0.42–0.69; $p < 0.05$). FOHR was significantly reduced at last follow-up shunt independent patients (0.47 ± 0.05; range 0.41–0.55; $p < 0.001$)

Conclusion We conclude that ETV seems to be a successful treatment option for patients with MRI signs of intra-/extraventricular pressure gradient at the 3rd ventricle and patent aqueduct and fourth ventricular outlets in children older than 1 year of age. This condition is observed only rarely and warrants further research on a multicenter basis in order to get more solid data of its pathophysiology.

Keywords Endoscopic third ventriculocisternostomy · Neuroendocopy · Panventriculomegaly · Extraventricular intracisternal obstructive hydrocephalus

Introduction

The standard therapy for communicating hydrocephalus is the implantation of a ventriculoperitoneal (VP) shunt while non-communicating type of hydrocephalus with, e.g., occlusion at the Sylvian aqueduct or the outlets of the fourth ventricle, may be treated by endoscopic third ventriculocisternostomy (ETV) [1–7]. Both surgical interventions are well established for decades [2, 8–12]. Obvious advantages of ETV include preventing foreign material implantation, which are more often associated with long-term complications such as overdrainage or occlusion [13–18]. ETV in non-communicating hydrocephalus has the aim to bypass the point of obstruction by fenestration of the floor of the third ventricle.

In rare cases in which all ventricular pathways are patent towards the subarachnoid space, still an intra-/extraventricular pressure gradient may be seen as bulging of the membranes at the level of the third ventricle including the lamina terminalis,

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the floor of the third ventricle, or the pineal recess. Kehler et al. presented five pediatric hydrocephalus patients with similar characteristics who were treated successfully with ETV and assumed an extraventricular, intracisternal obstruction [19]. Kageyama and colleagues described this condition as panventriculomegaly with wide ventricular outlets in a cohort of 28 patients, including five children [20]. Further genetic investigation in positive family history for hydrocephalus was positive in three from 17 patients in this cohort.

The aim of this study was to evaluate our cohort of patients with panventriculomegaly and an obvious pressure gradient at the level of the third ventricle on magnetic resonance imaging (MRI) treated by ETV with respect to clinical data and treatment outcome.

Methods and patients

Inclusion criteria and radiological findings

The study received approval from the institutional ethics committee (EA2/132/17). We retrospectively reviewed the records and analyzed the MRI studies of all patients who underwent an ETV in our department from June 2010 to March 2018. Thus, the MRI datasets of 137 patients were analyzed for the following findings and therefore met the inclusion criteria for this study: [1] intra-/extraventricular pressure gradient at the level of the 3rd ventricle, [2] open Sylvian aqueduct, [3] open outlets of the 4th ventricle, [4]. open CSF pathway at the foramen magnum towards the spinal canal. The imaging signs for a pressure gradient would include outward bulging of any 3rd ventricular membranes such as lamina terminalis, floor of the third ventricle, or pineal recess. In addition, the presence of an enlargement of the cisterna magna as well as the 4th ventricle was recorded (Fig. 1). Especially, the patent 4th ventricular outlets and the free communication towards the spinal canal were seen as signs to exclude a Blake´s pouch or any other variant of non-communicating hydrocephalus.

In midline sagittal image data sections, standard T2-weighted or preferably heavily T2-weighted gradient echo sequences with high spatial resolution (e.g., CISS, FIESTA, bFFE) possible arachnoid membranes especially located in the prepontine cisterns were recorded. All characteristics were identified by a neurosurgeon and a neuroradiologist (AT) independently reviewed and finally rated all findings [21, 22].

We identified patients who met the radiological signs as mentioned above and underwent an ETV in our institution. Patients with previous history of CSF infection or hemorrhage or previous intervention for hydrocephalus were excluded. All other children were included, regardless of the comorbidities and other previous surgical interventions.

Surgical intervention

All children underwent an ETV, in which the 3rd ventricular floor was perforated to establish a CSF flow between the 3rd ventricle and the basal cisterns. The optimal entry point was defined by measuring the trajectory parallel to the clivus through the floor of the third ventricle and the foramen of Monro extending to the right frontal calvarial convexity. Measurements were taken relative to the nasion and the midline to re-identify the entry at the patient's head [23]. Endoscopic guidance to the ventricle was either performed by using a navigation system or by using a ventricular guiding instrument in which the coronal angulation towards the calvarial convexity was previously determined on MRI and applied by the guiding tool [24]. The third ventricle was approached by a rigid endoscope and the floor of the third ventricle was identified. The floor's membrane was perforated by blunt instrument, usually a monopolar probe without cautery, flowed by the enlargement of the opening by a dilating forceps or a Fogarty balloon catheter to a diameter of approximately 6 mm pass the membrane with the endoscope. Any prepontine membranes in the basal cisterns were also perforated after performing the fenestration of the floor of the third ventricle (Fig. 1b). If the ETV was not successful during follow-up in terms of persisting clinical signs of increased intracranial pressure, a re-ETV was done or VP shunt implantation surgery was performed depending on individual decisions. The VP shunt was implanted in a standard manner using the same frontal entry point for the ventricular catheter and placing a borehole reservoir followed by a subcutaneous catheter to the ipsilateral retroauricular region. Here, usually a gravitational assisted adjustable differential pressure valve was placed, connected to a subcutaneous catheter which was tunneled from a contralateral periumbilical incision and placed intraperitoneally by using a mini laparotomy.

Follow-up investigations

The ventricular dilatation was measured using the frontal and occipital horn ratio (FOHR) [25] on axial T2-weighted MRI images at the level of the foramen of Monro. The FOHR was recorded before surgical intervention, in the first available MRI postoperatively, and at the end of individual follow-up.

Clinical symptoms were recorded before and after surgery. All surgical interventions after ETV were evaluated during follow-up. Every re-ETV after the first one was documented as revision operation. If shunt implantation was becoming necessary during follow-up, it was rated as ETV failure. Shunt-free survival as implemented in Kaplan–Meier curves is presenting temporal course and rate of shunt implantations after the first ETV.

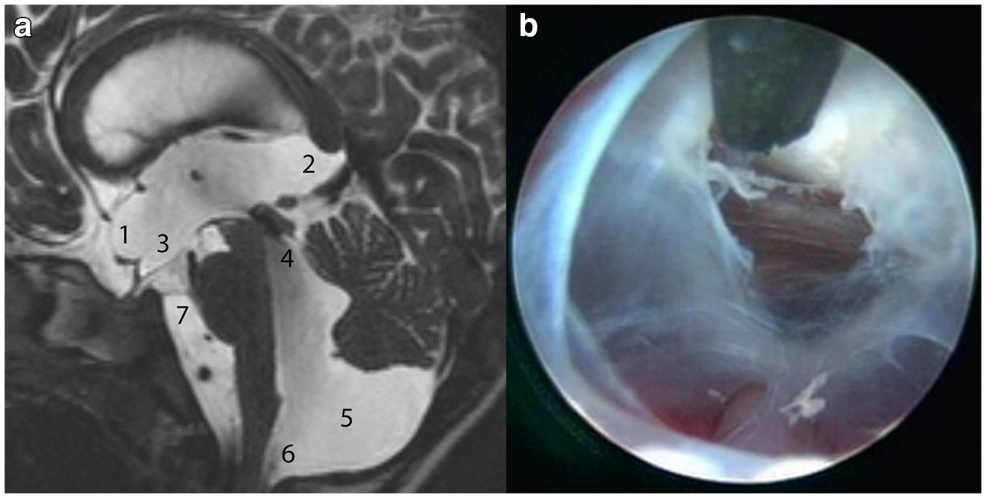

Fig. 1 **a** Radiological signs of extraventricular non-communicating hydrocephalus were the pressure gradient at the level of the third ventricle observed by forward bulging of the lamina terminalis [1], enlargement of the pineal recess [2] or downward bulging of the floor of the third ventricle [3], the open Sylvian aqueduct [4], enlargement of the cisterna magna [5], free communication seen as a flow void towards the spinal canal [6], and the upward bulging prepontine membrane [7]. **b** Intraoperative endoscopic view during the opening of the prepontine membrane below the floor of the 3rd ventricle in the basal cistern. The basilar artery is visualized behind the membrane

Statistical analysis

Data were collected and organized with Excel (Microsoft Office 2016, Alburquerque, NM, USA). The descriptive statistics and the Kaplan–Meier curves were performed and created with SPSS 25 (IBM, Armonk, NY, USA) and Prism 7 (GraphPad Software, San Diego, USA). All values are presented as mean ± standard deviation (SD) if not defined otherwise. Comparison of Kaplan–Meier curves was performed with the log-rank test. The Wilcoxon test was performed to compare the distribution of two paired group values, while Mann Whitney test was used for unpaired comparisons. For multiple comparisons of paired values, the one-way analysis of variance (ANOVA) and post hoc Bonferroni multiple comparison was used. A p value of less than 0.05 was respected as statistically significant.

Results

Baseline characteristics of the population

A total of 21 patients were included (11 males, 10 females) as they matched the inclusion/exclusion criteria. The mean age of the patients at the time of their first ETV was 3.2 ± 3.1 years with a range of 1 month to 11 years and 9 months. The mean follow-up time was 40.7 ± 30 months (range 5–102 months). Thirteen patients were born at term and seven patients at preterm (birth before the 37th week of gestation). In one patient, this specific information was not available. Patient characteristics of our cohort are given in Table 1. None of the patients had a previous history of intracranial hemorrhage, CNS infection, tumor, or trauma as a cause of hydrocephalus. Thus, we assume all patients present with a congenital etiology of hydrocephalus. Patients presented with symptoms of increased intracranial pressure (ICP), such as percentile crossing, increased head circumference, headaches, and nausea. Eleven patients had macrocephaly (head circumference at > 97th percentile), three presented with a bulged fontanelle, and eight patients showed agitate behavior or a delay of mental and/or motoric development. Two patients showed a spastic diaparesis of the lower extremities.

Patients were closely monitored for re-occurrence of their clinical symptoms. Clinical symptoms of increased ICP were improved immediately after ETV in all patients. In the case of clinical deterioration in further follow-up, a re-ETV was performed or a VP shunt was implanted, which was always preceded by a new MRI. One of our patients died during follow-up due to comorbidities, including bronchopulmonary dysplasia, myelodysplastic syndrome, and primary Addison's disease, independent of the history of hydrocephalus treatment.

MR imaging characteristics

In all patients, the neuroradiologist confirmed the four inclusion criteria as visualized on T2- weighted paramedian sagittal sections (Fig. 1; Table 2). The pressure gradient at the level of the 3rd ventricle was seen in 100% of patients represented by either a downward bulging of the floor of the third ventricle in 90.5%, a forward bulging of the lamina terminalis in 95.2%, and a backward extension of the pineal recess in 95.2%. As additional observations, an enlarged 4th ventricle could be seen in 95.2% and an enlarged cisterna magna in 76.2% of patients. A midline prepontine membrane as possible intracisternal obstructing structure could clearly be visualized in 38.1%, was unclear, but suspected in 47.6% and was not

Table 1 Children with PPOHC: clinical characteristics and follow-up outcome

Patient	Sex	Age	Comorbidities	Symptoms	Revision ETV	Shunt?	Follow-up (mo)
1	f	10 yrs 9 mo	None	Headaches		No	38
2	m	6 mo	None	Macrocephaly, bulged fontanelle		Yes	37
3	f	9 mo	VACTERL Association	Mental and motoric developmental delay		Yes	29
4	m	1 y 4 mo	None	Macrocephaly		No	29
5	m	9 mo	None	Macrocephaly		Yes	23
6	m	1 y 1 mo	None	mental development delay, muscular hypotonia,		No	16
7	f	7 yrs 3 mo	Lateralization of the Atlas	Macrocephaly, headaches, nausea	1x	No	20
8	m	2 yrs 4 mo	None	Macrocephaly		No	5
9	f	10 mo	HC in family history	Bulged fontanelle, macrocephaly	2x	Yes	84
10	m	2 yrs	None	Developmental delay		No	5
11	m	8 mo	None	Macrocephaly, developmental delay, vomiting		Yes	69
12	f	2 yrs 1 mo	PDA and PFO	Macrocephaly, Spastic diaparesis of the legs		No	44
13	f	1 y 9 mo	PVLM	Macrocephaly, Spastic diaparesis of the legs		No	68
14	f	2 yrs 6 mo	Lack of white matter	Macrocephaly, developmental delay, ataxia		No	70
15	f	8 yrs 11 mo	None	Headaches		No	86
16	m	3 yrs 2 mo	None	Macrocephaly, developmental delay		No	102
17	m	4 yrs 4 mo	None	Macrocephaly, headaches, agitative behavior		No	39
18	f	11 yrs 9 mo	None	Headaches		No	65
19	m	3 ys 6 mo	None	Macrocephaly, intention tremor, ataxia		No	5
20	f	5 mo	Various	Macrocephaly, Bulged fontanelle		Yes, death due to MDS	16
21	m	1 mo	None	Macrocephaly		NA	NA

MDS, myelodysplastic syndrome; *yrs*, years; *y*, year; *mo*, months, NA, not applicable

visualized in 14.3%. In the latter two categories, no high-resolution T2-weighted gradient echo MRI sequences were available (Table 2).

Surgical revisions

A total of 32 neurosurgical interventions were performed in 20 patients (21 ETVs, 3 re-ETVs, 6 VP shunt implantations, 1 untethering, 1 subduro-ventriculoperitoneal shunt). In one patient, no follow-up data according further surgeries or the need of shunt implantation was available and thus excluded from further analysis. During the entire follow-up period, we observed a total shunt rate of 28.6%. The shunt-free survival of the entire group was 73.8% at 12 months and 49.2% at the end of follow-up (8 years and 6 months). The shunt-free survival for all patients older than 1 year was 100%. All infants, children under 1 year of age at first ETV, finally received a shunt ($n = 6$). Five of 6 patients received the shunt during the first year of follow-up, while one patient received a shunt after 7 years and 5 months (Fig. 2). Two of our patients had to undergo re-ETV, of whom one had two revisions. In one patient, re-ETV was performed at 11 months of follow-up and a shunt was not necessary subsequently. The other patient was revised with an ETV at 6.37 years and again after additional 3 months, however, finally receiving a VP shunt.

Ventricular width

The evaluation of hydrocephalus relief after ETV was evaluated by measuring changes in ventricular width as FOHR. For all patients, the preoperative mean FOHR was 0.55 ± 0.09 (range 0.40–0.69; $n = 21$). In two patients, MR images were not available postoperatively and could not be included for follow-up measurements. Thus, the mean FOHR for 19 patients after ETV was 0.5 ± 0.08 (range 0.42–0.69) at a mean follow-up of 7.87 ± 5.8 months (median 3.3 months; range 0.37–71 months). At the end of follow-up time of 43.22 ± 30.43 months (median 39 months, range 5–102 months)

Table 2 Radiological analysis of the patients' MRI (n = 21)

Radiological sign		Number of patients
Open Sylvian aqueduct		100%
Open 4th CSF outlet		100%
Communication with spinal cord		100%
Any pressure gradient at 3rd ventricle		100%
Downbulging 3rd ventricular floor		n = 19 [90.5%]
Pineal recess bulging		n = 20 [95.2%]
Forward bulging of the lamina terminalis		n = 20 [95.2%]
4th ventricle dilatation		n = 20 [95.2%]
Enlargement of Cisterna magna		n = 16 [76.2%]
Prepontine membrane?	Visualized	n = 8 [38.1%]
	Suspected*	n = 10 [47.6%]
	Not detected*	n = 3 [14.3%]

*No high-resolution T2-weighted gradient echo MRI sequences available

FOHR measurement was available for 18 patients with a mean value of 0.48 ± 0.05 (range 0.41–0.58; n = 18).

Evaluating the early effect of ETV on ventricular width in 19 patients with comparable MRI data sets a significant decrease of mean FOHR before ETV with 0.54 ± 0.08 (range 0.4–0.68) and 0.5 ± 0.08 (range 0.42–0.69) after ETV was observed (p < 0.01; Fig. 3a). Looking at the differences in preoperative FOHR for patients who remained VP shunt free to those who subsequently received a VP shunt in follow-up preoperative ventricular width showed a significant difference (no VP shunt 0.51 ± 0.07 versus subsequent VP shunt in FU 0.62 ± 0.05; p < 0.01). Patients who did not receive a VP shunt showed significant decrease in FOHR postoperatively (preOP 0.51 ± 0.07 versus postop 0.48 ± 0.05; p < 0.01), which was not the case in patients who subsequently needed VP shunt implantation (preOP 0.62 ± 0.05 versus postOP 0.55 ± 0.11; p = 0.31; Fig. 3b). All of the postoperative FOHR measurements were performed on MRI at a mean follow-up time of 7.87 ± 5.8 months (median 3.3 months; range 0.37–71 months) when still none of the patient received a VP shunt.

The long-term effect of ETV on ventricular width was evaluated in 13 patients with comparable MRI data sets and who remained shunt free (Fig. 4). FOHR before ETV was 0.52 ± 0.07 (range 0.44-0.66), which showed a significant decrease after ETV 0.49 ± 0.05 (range 0.42- 0.55, at mean 10.4 ± 8.7 months; median 4.16 months; range 2.26–71.4 months; p < 0.01) and was significantly further decreased at a last follow-up of 43.15 ± 31.67 months (median 39 months, range 5–102 months) with a FOHR of 0.47 ± 0.05 (range 0.41-0.55; p < 0.001; Fig. 4).

Discussion

The present study retrospectively describes our experience with ETV in patients with imaging signs of a pressure gradient between the 3rd ventricle and extraventricular subarachnoid space but still patent Sylvian aqueduct, outlets of the 4th ventricles as well as free communication towards the spinal canal. All patients treated initially at an age older than 1 year remained shunt free during follow-up; clinical condition was improved or stabilized and ventricular width decreased during follow-up. All infants diagnosed and treated under the age of 1 year became shunt dependent.

Shunt dependency is associated with a relevant rate of shunt complications during long-term follow-up [8, 11, 26, 27], which should be avoided if justified. Typical MRI finding indicating an intra-/extraventricular pressure gradient at the level of the 3rd ventricle may serve as relevant justification to treat hydrocephalus patients with ETV in order to re-establish the communication towards the CSF spaces of the intracranial convexity. This became a standard treatment option in aqueductal stenosis patients. In our study, we included

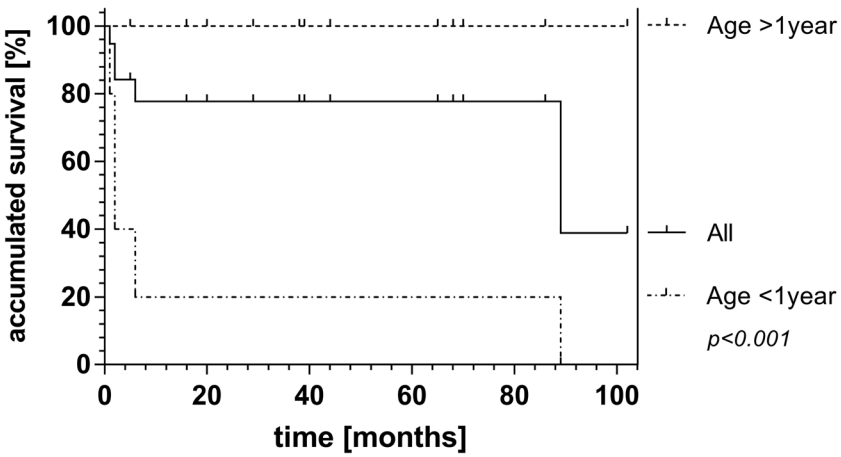

Fig. 2 Kaplan–Meier curves presenting the shunt-free survival of the entire study cohort (middle curve). In infants younger than 1 year of age (lower curve), all ETVs needed conversion to a VP shunt, while in patients older than 1 year of age, no patient received a shunt (upper curve) during follow-up (p < 0.0001)

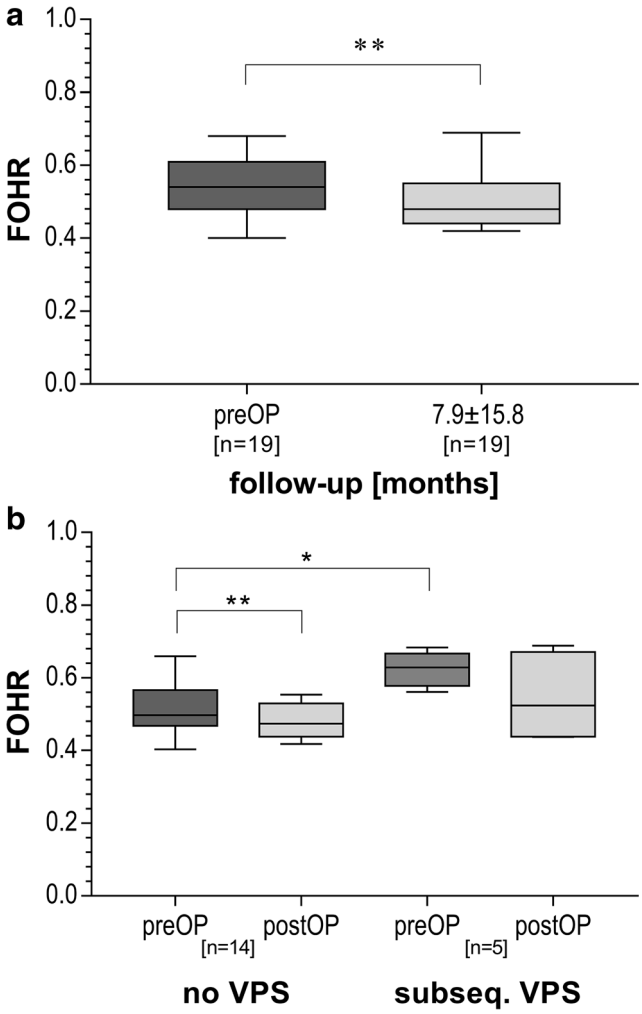

Fig. 3 **a** Measurement of the FOHR before intervention (pre OP; mean FOHR of 0.54 ± 0.08 (range 0.4-0.68)) and after intervention (post OP; mean FOHR of 0.5 ± 0.08 (range 0.42-0.69)) in patients with comparable available MRI data sets before shunt implantation was needed ($n = 19$; $p < 0.01$). $^{**}p < 0.01$ versus preoperative FOHR. **b** Differences in FOHR in preoperative MRI for patients who remained VP shunt free (0.51 ± 0.07) to those who subsequently received a VP shunt (0.62 ± 0.01) showed a significant difference ($p < 0.01$). Patients without a VP shunt showed significant decrease in FOHR postoperatively (0.51 ± 0.07 versus 0.48 ± 0.05; $p < 0.01$), which was not the case in patients later needed a VP shunt (0.62 ± 0.05 versus 0.55 ± 0.11; $p = 0.31$)

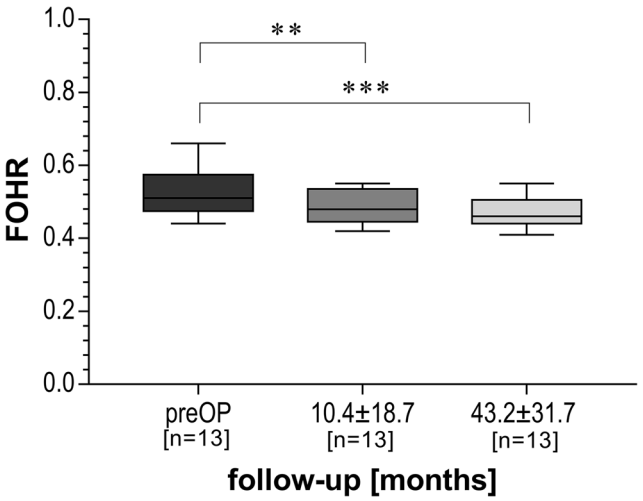

Fig. 4 FOHR measurements from consistently available MR images from non-shunted patients ($n = 13$). FOHR was 0.52 ± 0.07 (range 0.44–0.66) before ETV (pre OP), 0.49 ± 0.05 (range 0.42–0.55, 10.4 ± 8.7 months; $p < 0.01$) postoperatively after ETV and was significantly decreased at a mean follow-up of 43.15 ± 31.67 months with a FOHR of 0.47 ± 0.05 (range 0.41–0.55; $p < 0.001$). $^{**}p < 0.01$; $^{***}p < 0.001$ versus preoperative FOHR

the MRI findings of an outward bulging of the lamina terminalis, the floor of the third ventricle, and/or the pineal recess in conjunction with an open Sylvian aqueduct and open outlets of the fourth ventricle as well as a patent communication to the spinal canal in congenital hydrocephalus. Kehler et al. described previously a similar condition as extraventricular intracisternal obstructive hydrocephalus and hypothesized an obstruction in the basal cisterns [19, 28]. In a significant number of our patients, a prepontine membrane was clearly visualized or suspected, which often showed an upward bulging phenomenon. Thus, we hypothesize that the intraventricular space is communicating with the cisterna magna and the spinal canal, but communication is blocked towards the supratentorial subarachnoid space leading to the pressure gradient at the level of the third ventricle. This condition must be distinguished to any hydrocephalus due to restricted CSF resorption (post-infectious or post-hemorrhagic), where the obstruction is located more distally and imaging signs of a pressure gradient between CSF spaces are missing.

The prepontine membranes are parts of the arachnoid trabecular system, which normally traverses the subarachnoid space. The basal cisterns are built as a fine network of these arachnoid trabeculae, contributing to a directional CSF flow [29]. One of the most well-known components of these cisternal arachnoid membranes is the Liliequist's membrane (LM), an important surgical landmark in ETV procedures and previously discussed as a cause for failed ETVs [21, 30, 31]. The LM is described as a highly variable structure located in the basal cisterns, composed of one to three leaves, which originates from the dorsum sellae and which mainly separates the interpeduncular, prepontine, and chiasmatic cisterns [32–34]. The diverticular expansion of LM layers during embryological development is likely to play an important role in the formation of suprasellar, interpeduncular, mesencephalic, or prepontine arachnoid cysts [35–37]. Indicating the heterogeneitity of arachnoidal architecture, these differently located cystic malformations have their origin of arachnoid membranes at different levels of the basal premesencephalic and prepontine membranes [29, 38–42].

We assume an anatomical variant of these membranous structures with subsequent intracisternal obstruction in our patients. If the prepontine membrane can be visualized on MRI, this condition may specifically be called "prepontine

non-communicating hydrocephalus." However, if this membrane could not be verified, it may be due to the fact that imaging quality was insufficient or the membranes were rather located laterally apart from the midline. Prospective image data collection in those patients is needed in order to better characterize this rare condition of hydrocephalus. Kageyama et al. described a similar entity, which they term panventriculomegaly (PaVM) in a mixed cohort of adult and pediatric patients. They divided their cohort into two groups depending on presence or absence of a downward bulging 3rd ventricular floor, hypothesizing different CSF pathophysiology in their cohort and thus deciding for different treatment modalities of ETV or VP shunt. Patients with downward bulging of the 3rd ventricular floor were treated successfully by ETV [20], which is in line with our findings.

In the presented cohort of patients, ETV failed in all patients younger than 1 year of age. It is well known that age is an important factor for ETV success in patients with triventricular hydrocephalus and aqueductal stenosis, where failure rates are high in neonates and younger infants [5, 10, 43–45], which is defined in the ETV success score [46]. A contributing factor of ETV failure in infants may be the open fontanelle with lower CSF pulsation velocity at the stoma level but also the high growth potential of any tissue components, which may contribute to re-occlusion of the stoma. Moreover, the early onset of symptoms at young age represents different dynamics of pathophysiology and might be due to other contributing factors of minor CSF pathway disturbances or CSF malresorption. For those patients, ETV in combination with choroid plexus cauterization (CPC) has been proposed and successfully applied in order to increase the rate of shunt independent pediatric patients [47, 48]. Long-term outcome of ETV and CPC versus shunted patients, however, is still missing in order to give clear recommendations for this age group.

Conclusion

In our single-center retrospective pediatric cohort study of hydrocephalic patients with MRI signs of intra-/extraventricular pressure gradient at the level of the third ventricle in combination with free CSF pathways between the ventricles, the cisterna magna and the spinal canal, ETV treatment ameliorated the clinical condition in all children above 1 year of age. This entity of hydrocephalus and its treatment outcome is presented for the first time in a pure pediatric cohort. We hypothesize an extraventricular intracisternal CSF obstruction leading to non-communicating hydrocephalus. Further clinical and especially radiological studies are necessary to understand better the pathophysiology of this rare entity of congenital hydrocephalus.

Conflict of interest The authors have no conflict of interest to declare in relation to the presented content of this study.

Compliance with ethical standards

The study received approval from the institutional ethics committee (EA2/132/17).

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