ORIGINAL ARTICLE

Endoscopic third ventriculostomy in children with chronic communicating congenital hydrocephalus: a single-center cohort retrospective analysis

Laura Baeza-Antón[1] · **María Isabel Martínez-León**[2] · **Bienvenido Ros-López**[3] · **Miguel Ángel Arráez-Sánchez**[3]

Abstract

Objective The aim of this study was to analyze the role of endoscopic third ventriculostomy (ETV) in the treatment of pediatric chronic communicating congenital hydrocephalus (CCCH).

Material and methods This retrospective study comprised a series of 11 children with CCCH treated with ETV. Data were recorded on gender, history, presenting symptoms, age at surgery, complications during surgery, clinical evolution, ETV survival, and follow-up period. Radiological variables including ventricular and cephalic diameters were also recorded to determine a series of ventricular indexes in magnetic resonance imaging (MRI) before and after the ETV procedure. The procedure was considered to be successful when there was clinical stability or improvement accompanied by a reduction in the radiological indexes in the postoperative control images, such that there was no need to place an extrathecal cerebrospinal fluid shunt.

Results Over a mean follow-up period of 35.8 months (range: 6–108 months) from the ETV procedure, three patients required shunt placement; one of these was due to early failure in an 8-month old girl, the only patient younger than 12 months in our series. The radiological indexes were reduced in all patients except for one of the cases of ETV failure. The mean ETV survival among the successful cases was 32.1 months (range: 6–108 months), whilst that of the failed cases was 16 months (range: 6–108 months).

Conclusion Although studies with larger sample sizes are needed, ETV appears to be a promising option for the treatment of this type of patient with CCCH.

Keywords Severe idiopathic communicating hydrocephalus · Ventricular size · Key cisterns · Neuroendoscopy

Introduction

Endoscopic third ventriculostomy (ETV) is considered the treatment of choice for patients with non-communicating hypertensive hydrocephalus [1–3], though controversy exists for its use in other indications, in particular chronic communicating hydrocephalus [4–11].

Two problems exist concerning the use ETV in chronic hydrocephalus. The first relates to the prediction of success, given that the prediction scales developed to date are only for use in hypertensive hydrocephalus [7]. The second problem is the assessment of success or failure of the procedure, as most authors consider ETV to be successful if there is then no need to place a shunt, though this criterion can be disregarded in chronic or "long standing" cases with procedure failure but with symptoms that are tolerated by the patient. On the other hand, patients

✉ Laura Baeza-Antón
laubaeza1221@gmail.com

María Isabel Martínez-León
maria.i.martinez.leon@gmail.com

Bienvenido Ros-López
broslopez@yahoo.es

1 Department of Neurosurgery, HGU Doctor Negrín, Las Palmas de Gran Canaria, Spain

2 Pediatric Radiology Section, Department of Radiodiagnosis, HRU Carlos Haya, Las Palmas de Gran Canaria, Spain

3 Pediatric Neurosurgery Section, Department of Neurosurgery, HRU Carlos Haya, Málaga, Spain

with long-term signs or symptoms like macrocephaly, mild headache, psychomotor retardation, or poor sphincter control for example, may not improve obviously or significantly in the immediate post-operative period and require radiological studies to confirm the patency of the ventriculostomy [12]. Other problems also surround the use of ETV in communicating hydrocephalus due to the varying concepts reported about this particular type [8, 13–15]. The aim, therefore, of this study was to analyze our results in a series of children with CCCH.

Methods and patients

Inclusion criteria and radiological findings

This retrospective study involved 11 patients with CCCH. The data were extracted from the endoscopy database of our center, which is constantly updated and records all cases. This report covers the period January 2003 to October 2020. Communicating hydrocephalus was defined radiologically as ventricular dilatation that on MRI shows aqueductal flow artifact, no block in the foramen magnum or in the IV ventricle outflow, and patency of the interpeduncular and suprasellar "key cisterns"; that is, the flow artifact should extend from the prepontine cistern to the interpeduncular and suprasellar cisterns [16–18].

The communicating hydrocephalus was considered to be clinically chronic if it was characterized by lack of symptoms or the presence of long-term symptoms (months), like headache but with no intracranial hypertension (nausea, vomiting, papillary edema, diplopia), macrocephaly, or alterations in cognitive development, gait, or sphincter control [19, 20].

Patients with a previous history of head injury, CSF infection, or hemorrhage or previous interventions for hydrocephalus were excluded. Thus, the MRI datasets of 11 patients who met the inclusion criteria for this study were analyzed for the data described below.

Demographic and clinical variables

The following variables were included: gender, background, age at surgery, clinical symptoms before and after surgery, perioperative complications, clinical outcome, and follow-up time.

Concerning prognosis, improvement was defined as a combination of clinical and radiological data. This group included both patients with significant sustained improvement as well as clinically stable patients provided there was improvement in the ventricular indexes.

Radiological variables

Radiological variables were measured and recorded in both pre- and post-ETV MRI control studies and compared. MRI measurements were obtained using the most recent MRI control before surgical intervention as the pre-ETV control, and the latest MRI as the post-ETV control. Heavily T2-weighted sequences (Philips Intera 1.5 T MRI scanner) and 3D T2 high spatial resolution (sampling perfection with application-optimized contrasts using different flip angle evolution: SPACE) (Siemens Aera 1.5 T MRI scanner) were obtained in midline in sagittal planes, and TSE T2 sequences of the whole brain in the axial plane. The following measurements were made, and classified into four groups according to their characteristics: [21–23].

1. Measurements of the cerebral ventricles and cranium: internal bifrontal diameter (IFD), bifrontal horn width (BFHW), frontal horn width (FHW), internal biparietal diameter (IPD), biocciptal horn width (BOHW), occipital horn width (OHW), and third ventricle width (TVW)
2. Measurements of the third ventricle: third ventricle floor distance (VFD), third ventricle floor bowing (VFB), lamina terminalis distance (LTD), lamina terminalis bowing (LTB), anterior commissure-tuber cinereum distance (ACTC), mamillary body-lamina terminalis distance (MBLT)
3. Cerebral ventricle relative indices: Evan's index (EI), frontal horn index (FHI), frontal occipital horn index (FOHI), third ventricular morphological index (TVMI)
4. Anatomical landmarks: aqueductal artifact (AA), prepontine membrane (PPM), patency of ventriculostomy (PV)

Table 1 gives the definitions of these radiological parameters.

Image analysis and measurement were done by an experienced pediatric radiologist (MIML). For the data analysis, radiological success was considered to be when there was improvement in the radiological parameters relating to the relative indexes (EI, FHI, FOHI, TVMI) (Figs. 1 and 2).

A descriptive qualitative analysis was also done for the presence in the pre- and post-surgical MRI images of the changes in the parameters as well as the visualization of the depth of the convexity sulci, the corpus callosum thickness, the presence or otherwise of periventricular edema, and the morphological change in the third ventricle from "omega" (pre-ETV) to "w" (post-ETV) (Fig. 3) [24]. The existence of benign enlargement of the subarachnoid space (BESS) pre- and post-ETV was also documented.

Table 1 Explanation of radiological measurements (diameter, width, distance, bowing, in millimeter), indices, and anatomical landmarks

Abbreviation	Radiological variable	Definition
1. IFD	Internal bifrontal diameter	Maximum inner skull diameter at frontal level (increase with patient age)
2. BFHW	Bifrontal horn width	The transverse distance defined by a line connecting the anterior corners of the frontal horns (head of the caudate nucleus level)
3. FHW	Frontal horn width	Maximum width between the walls of one frontal horn of the lateral ventricle (head of the caudate nucleus level)
4. IPD	Internal biparietal diameter	Maximum inner skull diameter at parietal level (increase with patient age)
5. BOHW	Bioccipital horn width	The transverse distance defined by a line connecting the trigone of the occipital horns
6. OHW	Occipital horn width	Maximum width between the walls of one occipital horn of the lateral ventricle (trigone level)
7. TVW	Third ventricle width	Maximum width of the anterior recess of the third ventricle (superior to the suprasellar cistern)
8. VFD	Third ventricle floor distance	The line between mammillary bodies and posterior margin of the infundibular recess
9. VFB	Third ventricle floor bowing	The line between the depression of the tuber cinereum and VFD (8). The VFB measurement is not valid in post-ETV studies because it is the location of the ventriculostomy stoma
10. LTD	Lamina terminalis distance	The line between anterior commissure and anterior optic chiasm
11. LTB	Lamina terminalis bowing	The line between the bulging of the lamina terminalis and LTD (10)
12. ACTC	Anterior commissure – tuber cinereum distance	Maximum distance between the anterior commissure and the midpoint of the floor of the third ventricle (midpoint of the tuber cinereum)
13. MBLT	Mammillary body – lamina terminalis distance	Maximum distance between the mammillary bodies and the midpoint of the lamina terminalis
14. EI	Evan's index	BFHW/IPD (2/4)
15. FHI	Frontal horn index	BFHW/IFD (2/1)
16. FOHI	Frontal occipital horn index	BFHW + BOHW / IPD (2 + 5/4)
17. TWMI	Third ventricular morphological index	ACTC + MBLT/IPD (12 + 13/4)
18. AA	Aqueductal artifact	Means open aqueduct
19. PPM	Prepontine membrane	The presence of prepontine membrane
20. PV	Patency of ventriculostomy	Means open endoscopic third ventriculostomy

Numbers in parentheses refer to the items in the first column

Surgical technique

All procedures were performed by two pediatric neurosurgeons, BRL and SIM. Patients underwent ETV with the standard procedure through a right precoronal burr hole. A rigid endoscope AESCULAP MINOP ® with a 0° scope was used. The third ventricular floor was fenestrated between the mammillary bodies and the tuber cinereum to establish a CSF flow between the third ventricle and the basal cisterns. The opening was created by perforating the floor membrane with a blunt instrument, usually a monopolar probe without cautery, followed by enlargement of the opening with a Fogarty balloon catheter to a diameter of approximately 6 mm past the membrane with the endoscope. When the opening was considered insufficient, another contiguous fenestration was performed to ensure CSF flow. Both the ependymal layer and the so-called internal membrane were pierced. The arachnoid of the interpeduncular cistern was also opened. Soon after the fenestration, pulsations of the third ventricle floor were observed intraoperatively in most cases.

Follow-up studies

The first outpatient clinical evaluation after the ETV took place 1 week after hospital discharge. Subsequent revisions were at 1 month and 3 months. The first MRI after the ETV was done during the first week prior to discharge when possible; otherwise, it was done on an outpatient basis during the first month. The minimum follow-up period was 6 months. All surgical interventions after ETV were evaluated during follow-up. If shunt implantation became necessary during follow-up, ETV was rated as a failure.
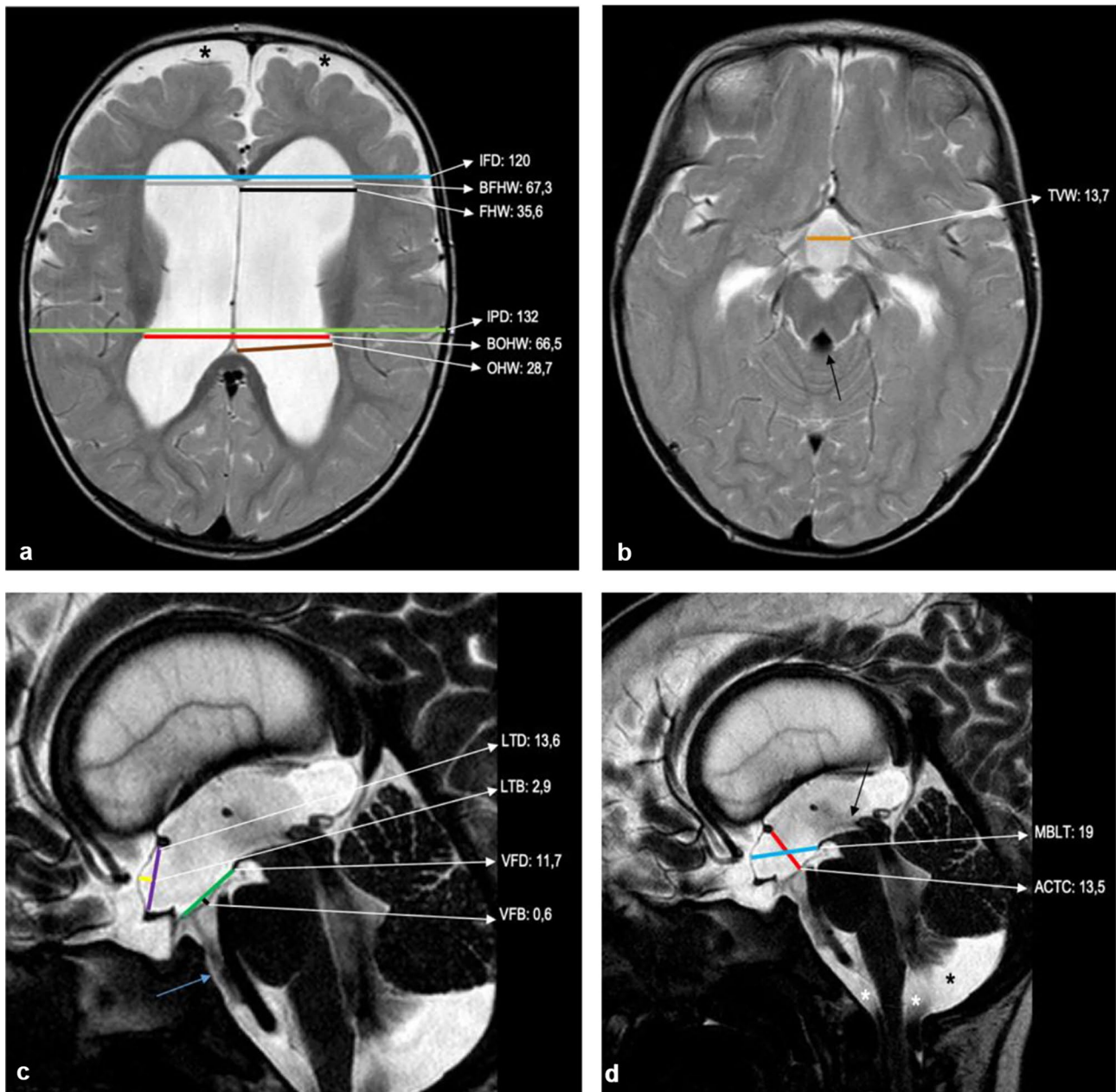
Fig. 1 Patient no. 10, 23 months old. Pre-surgery MRI. **a**, **b** Transverse TSE T2 sequence. Generalized ventriculomegaly, large subarachnoid frontal space (asterisk), open aqueduct (black arrow). Measurements of the lateral and third ventricles, and the skull vault (in millimeter): internal bifrontal diameter (*IFD*, blue line), bifrontal horn width (BFHW; grey line), frontal horn width (*FHW*; black line), internal biparietal diameter (*IPD*; green line), biooccipital horn width (*BOHW*; red line), occipital horn width (*OHW*; brown line), third ventricle width (*TVW*; orange line). Evan's index (*EI*): 0.50; frontal horn index (*FHI*): 0.56; frontal occipital horn ratio (*FOHR*): 1.01; third ventricle morphological index (*TVMI*): 0.24. **c**. **d** Sagittal 3D T2-SPACE sequence. Generalized ventriculomegaly, open aqueduct (black arrow), megacisterna magna (black asterisk), free communication seen as a flow void towards the spinal canal (white asterisk), there is no prepontine membrane (blue arrow). Third ventricle floor distance (*VFD*, green arrow): 11.7; third ventricle floor bowing (*VFB*, black arrow): 0.6; lamina terminalis distance (*LTD*, purple arrow): 13.6; lamina terminalis bowing (*LTB*, yellow arrow): 2.9; anterior commissure-tuber cinereum distance (*ACTC*, red arrow): 13.5; mammillary body-lamina terminalis distance (*MBLT*, blue arrow): 19
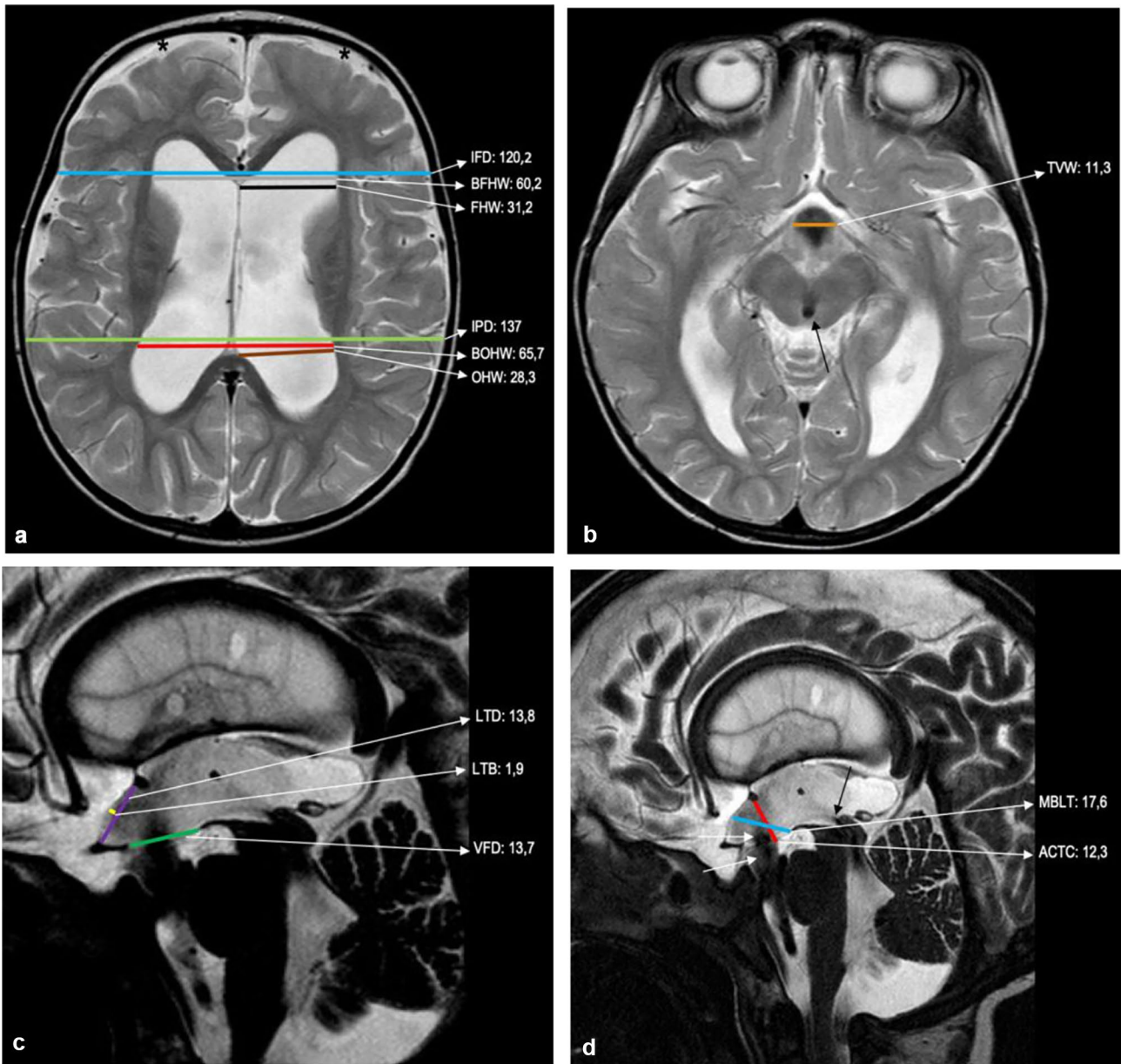
Fig. 2 Patient no. 10. MRI control study one month after ETV. **a**, **b** Transversal TSE T2 sequence. Aqueduct is open (black arrow), subarachnoid frontal space is smaller (black asterisk). After ETV, all ventricular measures have decreased: frontal horn width (*FHW*, black line): 31.2; bifrontal horn width (*BFHW*, grey line): 60.2; occipital horn width (*OHW*, brown line): 28.3; biooccipital horn width (*BOHW*, red line): 65.7, third ventricle width (*TVW*, orange line): 11.3. The skull vault increased in size as the patient is older: internal bifrontal diameter (*IFD*, blue line): 122, internal biparietal diameter (*IPD*, green line): 137. The data showed a decrease in all the relative indices: Evan´s index (*EI*): 0.43, frontal horn index (*FHI*): 0.49, frontal occipital horn ratio (*FOHR*): 0.91, and third ventricle morphological index (*TVMI*): 0.21. **c**. **d** Sagittal 3D T2-SPACE sequence. ETV is open (white arrows); aqueduct is also open (black arrow). Radiological demonstration of flat ventricular walls after successful management, most of the previous third ventricular measures and all the indexes have decreased after ETV: third ventricle floor distance (*VFD*, green line): 13.7; third ventricle floor bowing (VFB not measureable, ETV localization), lamina terminalis distance (*LTD*, purple line): 13.8, lamina terminalis bowing (*LTB*, yellow line): 1.9), anterior commissure-tuber cinereum distance (*ACTC*, red line): 12.3; mammillary body-lamina terminalis distance (*MBLT*, blue line): 17.6)
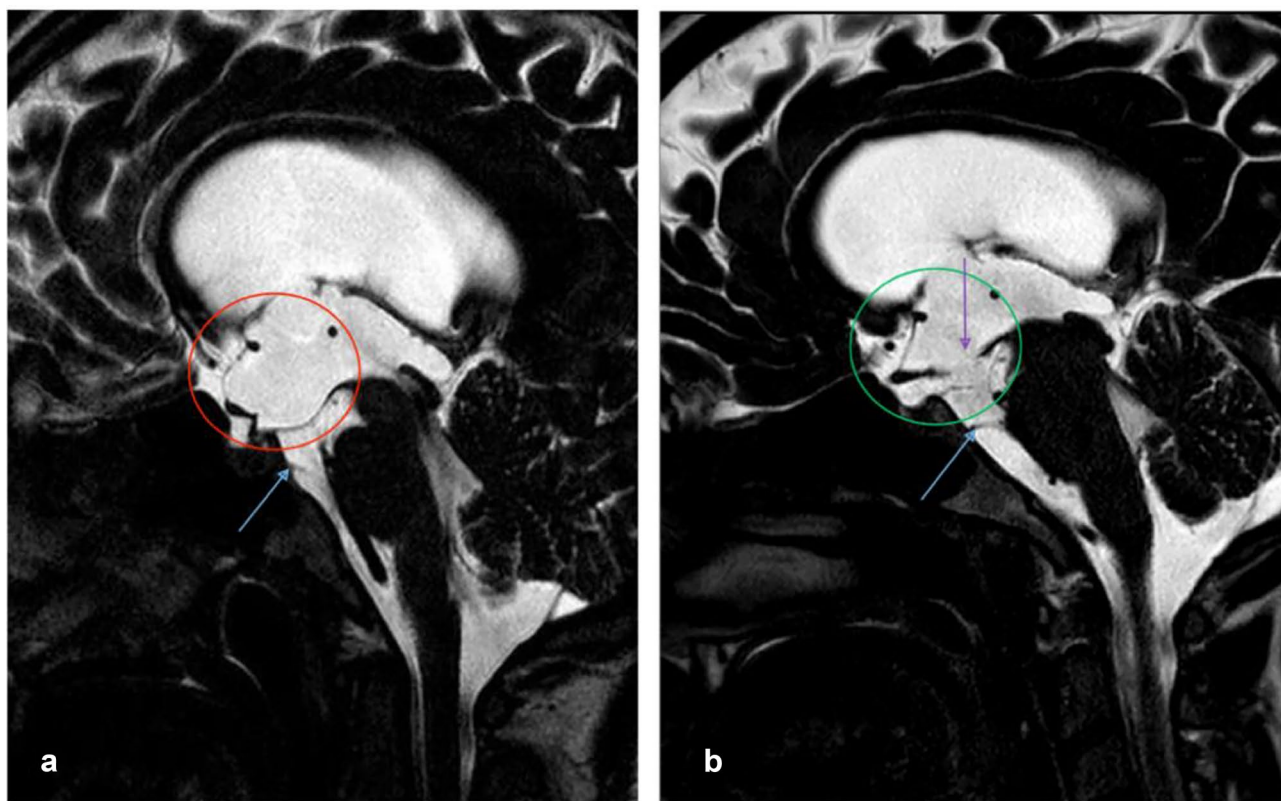
Fig. 3 **a** Pre-surgery MRI. **b** Post-surgery MRI. Incomplete preponttine membrane (blue arrow) without upward bulging, visualized in patient number 4 (patient data in Table 2). "Omega" shape previous to ETV surgery (red circle), and "W" shape after ETV surgery (green circle). Open ETV with a week signal artifact (*purple arrow*)

Results

Baseline characteristics of the population

The study included 11 patients (8 male, 3 female) who matched the inclusion/exclusion criteria. The clinical presentation in eight patients was macrocephaly (head circumference at 98th percentile or greater than two standard deviations above the mean); four showed psychomotor retardation (age matched). Two patients presented headache (mild and chronic), and another two had gait disturbance. One patient experienced urinary incontinence. The patient characteristics of our cohort are shown in Table 2.

The first ETV procedure was performed in 2006 and the rest during the period 2013–2020. The mean age of the patients at the time of their ETV was 77 months (approximately 6.5 years) with a median of 39 months (approximately 3 years) and a range of 8–229 months. In this study, to evaluate psychomotor retardation, gait disturbance, and clinical changes after the ETV, we considered the observations of the children's parents and teachers. Post-ETV improvement was considered to be when there was a cause-effect relation between the ETV and clinical improvement,

i.e., when the parents noted an important sustained improvement during the first post-operative weeks.

MRI characteristics

Analysis of the pre- and post-ETV radiological measurements showed a generalized relative improvement in the ventricular indexes in the successful ETV procedures (Figs. 1 and 2). Tables 3, 4 and 5 give the results of the radiological measurements. An improvement was also noted in the qualitative parameters in three patients after the procedure (Fig. 3) [24].

Assessment of ETV success or failure

Patients were closely monitored for re-occurrence of their clinical symptoms. The mean follow-up period of the patients from the ETV was 35.8 months, with a median of 27.7 months (range 6–108 months). The clinical outcome was stable or showed an improvement immediately after ETV in all patients. Three cases experienced clinical worsening in further follow-up and required shunt implantation. Two of these three also experienced neurological

Table 2 Patients demographics, clinical, and radiological data

Patient n°	Age (mo)	Gender	Macrocephaly	Psychomotor delay	Gate disturbance	Bladder dysfunction	Headache	Postoperative symptomatology	Cerebral ventricle relative indices decrease	ETV clinical success	ETV survival (mo)
1	149	M	No	No	Yes	Yes	No	Asymptomatic	EI, FHI, FOHR, TVMI	Yes	108
2	106	F	Yes	Yes	No	No	No	Initially stable, headache after 18 mo	FOHR	No	18
3	39	M	No	Yes	No	No	No	Seizure control, improved cognitive performance	EI, FHI, FOHR, TVMI	Yes	47
4	229	F	Yes	No	No	No	Yes	Asymptomatic	EI, FHI, FOHR, TVMI	Yes	26
5	74	F	Yes	No	No	No	Yes	Stable, minor headaches	EI, FHI, FOHR, TVMI	Yes	31
6	29	M	Yes	Yes	No	No	No	Improved psychomotor performance	EI, FHI, FOHR	Yes	14
7	37	M	Yes	No	Yes	No	No	Initially stable, impairment of fine motor skills, tremor and headache	EI, FHI, FOHR	No	29
8	124	M	Yes	Yes	No	No	No	Stable	EI, FHI, FOHR, TVMI	Yes	15
9	29	M	No	No	No	No	No	Stable	EI, FHI, FOHR, TVMI	Yes	10
10	23	M	Yes	No	No	No	No	Stable	EI, FHI, FOHR, TVMI	Yes	6
11	8	F	Yes	No	No	No	No	Early failure (pseudomeningocele, risk of CSF leak)	EI, FHI	No	1

Yes: not possible to make specific measurements of the third ventricle due to lack of necessary plane and sequences on MRI (retrospective study)

CSF cerebrospinal fluid, *ETV* endoscopic third ventriculostomy, *Mo* months

Table 3 Radiological measurements of the cerebral ventricles and cranium, pre- and post-ETV

Patient no	FHW pre-ETV	FHW post-ETV	BFHW pre-ETV	BFHW post-ETV	OHW pre-ETV	OHW post-ETV	BOHW pre-ETV	BOHW post-ETV	TVW pre-ETV	TVW post-ETV	IFD pre-ETV	IFD post-ETV	IPD pre-ETV	IPD post-ETV	ETV success	BESS pre-ETV	BESS post-ETV
1	36.2	33.9	76.2	69.4	38.3	15.6	90	86.3	17.5	14.9	137	138	154	156	Yes	No	No
2	26.6	25.3	50.5	54	20.6	24	56.9	55.9	11.1	10.9	113	122	131	136	No	No	No
3	21.1	21.5	41.8	42	19.3	18.5	52.5	50.5	10	9.4	103	110	123	128	Yes	No	No
4	45	44	93	90	36.5	33.6	79	76	14	13.6	133	133.3	141	141.1	Yes	No	No
5	28.1	22.7	55.1	44.7	40.4	26.3	79.7	55.6	15.2	13.6	115	118	129	132	Yes	No	No
6	24.1	23.6	51.2	51.3	17.5	19.6	50.3	54.8	7.6	8.2	117	124	127	139	Yes	Yes	No
7	40.3	36.7	80.9	73.9	40.4	40	94.7	83.2	20.7	19.7	119	128	145	146	No	No	No
8	35.3	34.9	75.1	74.1	43.5	40.2	92.3	88.2	21.5	17.6	127	131	146	146	Yes	No	No
9	20.3	20	51.6	46.9	29	22.2	70	64.6	12.4	8	105	107	130	130	Yes	No	No
10	35.6	31.2	67.3	60.2	28.7	28.3	66.5	65.7	13.7	11.3	120	122	132	137	Yes	Yes	No
11	22.0	20.3	44.1	45.2	17.2	17.9	45	49.7	6.6	5.4	101	108	117	24	No	Yes	Yes

FHV frontal horn width, *BFHW* bifrontal horn width, *IFD* internal bifrontal diameter, *OHW* occipital horn width, *BOHW* biooccipital horn width, *IPD* internal biparietal diameter, *TVW* third ventricle width, *BESS* benign enlargement of the subarachnoid space

Table 4 Measurements of the third ventricle pre- and post-ETV

Patient n°	VFB pre-ETV	VFB post-ETV	LTB pre-ETV	LTB post-ETV	ACTC pre-ETV	ACTC post-ETV	ACTC decrease %	MBLT pre-ETV	MLBT post-ETV	MBLT decrease %	ETV success
1	2.4	*	2.1	0	19.3	15.1	11.8	21.1	19.3	8.5	Yes
2	0	*	0	0	13.3	15.1	None	13.1	15.3	None	No
3	0	*	0	0	14.6	15	None	14.4	14.3	0.7	Yes
4	3	*	3.2	0	19	13.2	31.5	24.2	19.4	19.8	Yes
5	3.5	*	3.1	0	19.5	12.7	34.9	19.9	17.3	13.1	Yes
6	-	*	-	0	-	10.9	-	-	1.5	-	Yes
7	-	*	-	1.2	-	19.9	-	-	21.7	-	No
8	3.5	*	4.5	0	23.7	18.1	33.6	31.2	19.9	36.2	Yes
9	0	*	2.8	0	15.1	13.4	11.3	17.1	12.1	29.2	Yes
10	0.6	*	2.9	1.9	13.5	12.3	8.9	19	17.6	7.4	Yes
11	-	*	-	-	-	11.3	-	-	7.4	-	No

VFB third ventricle floor bowing, *LTB* lamina terminalis bowing, *ACTC* anterior commissure-tuber cinereum distance, *MBLT* mammillary body-lamina terminalis distance.—unknown

*The VFB measurement is not valid in post-ETV studies because it is the location of the ventriculostomy stoma

Table 5 Cerebral ventricle relative indices pre- and post-ETV

Patient n°	EI pre-ETV	EI post-ETV	EI decrease %	FHI pre-ETV	FHI post-ETV	FHI decrease %	FOHR pre-ETV	FOHR post-ETV	FOHR decrease %	TVMI pre-ETV	TVMI post-ETV	TVMI decrease %	ETV radiological success
1	0.49	0.44	10.2	0.55	0.5	9.1	1.07	0.99	7.5	0.26	0.22	15.4	Yes
2	0.38	0.38	0	0.44	0.44	0	0.81	0.8	1.2	0.2	0.22	None	Yes
3	0.33	0.32	3	0.4	0.38	5	0.76	0.72	5.3	0.23	0.22	4.4	Yes
4	0.65	0.63	3.1	0.69	0.67	2.9	1.21	1.17	3.3	0.3	0.23	23.4	Yes
5	0.42	0.33	21.4	0.47	0.37	21.3	1.04	0.75	27.9	0.3	0.22	26.7	Yes
6	0.4	0.36	10	0.43	0.41	4.7	0.79	0.76	3.8	-	0.15	-	Yes
7	0.55	0.5	9.1	0.67	0.57	14.9	1.21	1.07	11.6	-	0.28	-	Yes
8	0.51	0.5	2	0.59	0.56	5.1	1.14	1.11	2.6	0.37	0.26	29.7	Yes
9	0.39	0.36	7.7	0.49	0.43	12.3	0.93	0.85	8.6	0.24	0.19	19.8	Yes
10	0.5	0.43	14	0.56	0.49	12.5	1.01	0.91	9.1	0.24	0.21	12.5	Yes
11	0.37	0.36	2.7	0.43	0.41	4.7	0.76	0.76	0	-	0.15	-	Yes

EI, Evans index, *FHI* frontal horn index, *FOHR* frontal occipital horn ratio, *TVMI* third ventricular morphological index

deterioration at three years of follow-up. The third had an early risk of a leak due to a recurring tension pseudomeningocele after the ETV. All patients experienced improvement in at least one of the ventricular indexes. The combination of the clinical and radiological data showed ETV success in eight patients, with three failures, two late and one early. All the patients in this cohort were aged ≥ 23 months, except for the patient who experienced early failure, who was 6 months of age.

The mean ETV survival in the successful cases was 32.1 months (range 6–108 months), and in the failed cases, it was 16 months (range 6–108 months).

Discussion

Although ETV is considered the treatment of choice in non-communicating hypertensive hydrocephalus [1–3, 38], several articles have reported its use in other types of hydrocephalus, such as chronic communicating hydrocephalus [4–11]. However, two problems exist concerning the use of ETV in chronic hydrocephalus. The first concerns the prediction of success, as the prediction scales developed are only for use with hypertensive hydrocephalus, as for example the Endoscopic Third Ventriculostomy Success Score (ETVSS) of Kulkarni et al. [7], constructed from a series where all the patients had symptomatic high-pressure hydrocephalus. These authors defined ETV success as the absence of ETV failure during the 6 months after the procedure, and ETV failure as the need for any later procedure for CSF shunt or death related with the hydrocephalus within 6 months of the procedure [7].

The second problem concerning the use of ETV in chronic hydrocephalus relates to the evaluation of success or failure. Most authors consider ETV success to be the lack of requirement to place a shunt [25–32], though this can be disregarded in chronic or "long standing" cases with procedure failure but with symptoms that are tolerated by the patients. On the other hand, patients with long-term signs or symptoms like macrocephaly, mild headache, psychomotor retardation, or poor sphincter control for example may not improve obviously or significantly in the immediate post-operative period and require radiological studies to confirm the patency of the ventriculostomy. This problem has been studied in some works that propose the combination of clinical and radiological criteria for the reliable definition of success in all types of hydrocephalus, and particularly in chronic hydrocephalus [12].

Concerning the chronic nature of hydrocephalus, hydrocephalus is considered chronic taking as a reference the "Multi-categorical Hydrocephalus Classification (McHC)," proposed by Shizuo Oi in the seventh category, where hydrocephalus is classified based on its chronology as acute,

chronic, or long-standing, and from the point of view of its progression as progressive or detained [20, 39]. The concept "long standing" was described by Shizuo Oi in 2000 in the work on "long-standing overt ventriculomegaly in adults" (LOVA) and considers that from a clinical viewpoint, this group of patients is characterized by being able to have macrocephaly with or without an intellectual quotient below normal, headache, dementia, gait disturbance, urinary incontinence, vegetative state, akinetic mutism, apathy, or parkinsonism [33].

From the clinical viewpoint in children, the concept of chronic or normal-pressure hydrocephalus in childhood and adolescence was defined by Bret and Chazal, where most patients present at least two elements of the adult triad: psychomotor retardation, gait abnormality, or poor sphincter control. In this series, a shunt was implanted or revised in 16 persons, in 5 of whom the symptoms resolved completely, 7 improved, and 4 failed to improve [19]. The presence of alterations associated with the cerebral parenchyma seemed to play an important role in treatment failure. Psychomotor retardation in this study was considered to be subtle worsening in psychomotor activity, involving difficulty for the families and the physicians when trying to distinguish between clinical changes, as well as highlighting the difficulty to distinguish active dynamic changes in the CSF from an arrested hydrocephalus [19]. In our series, we included patients with no symptoms (patient 9) or with just macrocephaly (patients 10 and 11). The indication for ETV in these cases was based on the conviction that the severe ventriculomegaly, if left untreated, could lead in the pediatric age to a slow but progressive alteration in psychomotor development that might remain silent until it manifested clinically years later and was then irreversible. In these patients, therefore, ETV would have a certain preventive nature. Postoperative "stability" would mean the absence of clinical worsening. It is thus so important to consider radiological variables as well in order to determine the possible success of the endoscopic procedure. The future development and application of standardized tools to evaluate the symptoms and CSF dynamics (ICP monitoring, infusion studies) would be of great help in decision making variables. In the case of patient no. 6, we consider the fact that the FHW measurements experienced an isolated decrease could be related with the long time interval between the pre- and post-ETV MRI studies, undertaken when the patient was 9 months and 19 days and again at the age of 3 years and 6 months, respectively. This idea is reinforced by the fact that both the radiological indexes and the clinical situation improved after the ETV.

Another controversial aspect concerns the indication for ETV in communicating hydrocephalus. The very concept of communication can cause confusion. Dandy's definition of communicating or non-communicating hydrocephalus refers to the presence of CSF passing between the lateral ventricle and the lumbar subarachnoid space, confirmed by injection of contrast in a lateral ventricle and its detection in a lumbar puncture [34]. Once the CSF is found in the subarachnoid space, Russell proposed the concept of obstructive/non-obstructive. According to this, all hydrocephalus are obstructive except those caused by hyperproduction of CSF in a plexus papilloma/carcinoma or in venous sinus thrombosis [9]. All these concepts have recently been recorded by Oi and Di Rocco [35].

Concerning CSF circulation in the subarachnoid space, Dandy had already defined in 1938 the concept of "key cisterns" (interpeduncular and chiasmatic) that distribute the CSF to the interhemispheric and the Sylvian fissures, and where an obstruction anterior to these key cisterns could be treated by opening the third ventricle floor unlike an obstruction posterior to the key cisterns [16].

Al-Hakim et al. recently reported on a cohort of 21 patients with idiopathic hydrocephalus treated by ETV. The MRI showed a pressure gradient in the third ventricle and obstructions of the Sylvian aqueduct, fourth ventricle, or the foramen magnum were ruled out [4]. These authors assumed the presence of a "midline prepontine membrane," that is an extraventricular intracisternal obstruction in the prepontine arachnoid septa, clearly visualized or suspected in 38.1% and 47.6% of the patients, respectively [4]. In our study cohort, we identified one case where an incomplete prepontine membrane was seen, with no signs of "upward bulging" (Fig. 3), which we therefore considered non-obstructive. These membranes were not clearly seen in the other study subjects. The cases reported by Al Hakim could correspond to Dandy's concept of pre-key cistern obstruction, in a type of communicating hydrocephalus (according to Dandy), but at the same time obstructive (according to Russell) [9, 34].

The literature contains many definitions of communication or obstruction, ranging from functional aqueduct stenosis to traumatic or post-infectious hydrocephalus and even those occurring after resection of posterior tumors [8, 13–15]. Other studies, however, like that of Kulkarni et al., define a subtype called "congenital communicating hydrocephalus" for this type of hydrocephalus [17].

Concerning developmental hydrocephalus, Tully et al. studied 236 children, classifying the cases in five groups, one of which comprised communicating hydrocephalus. A subgroup of this latter group, called "severe idiopathic communicating hydrocephalus," was formed by those cases with marked ventriculomegaly, and characteristically in some of these, there was an excessive accumulation of extra-axial fluid in the posterior fossa, seen as the presence of extra-axial fluid collections on MRI [18]. The hydrocephalus in our patients were communicating, chronic, and idiopathic, presenting many similarities to this subgroup. Three patients (pts 6, 10, and 11) had excessive fluid accumulation in the posterior fossa on pre-ETV

MRI. A notable extra-axial space may be present in these patients, thus underlying the communicating nature of the hydrocephalus and not contraindicative of surgery. The existence of BESS in these patients is information that is difficult to interpret, as only one of them had macrocephaly. Although all three were younger than 29 months, patient no. 9 (also in this age range) did not have either macrocephaly or BESS. Finally, patient no. 11 continued to have BESS after the ETV (still younger than 24 months and still with macrocephaly). Further studies are required to establish any relation between the so called "benign subdural effusion of infancy" and a possible evolution towards CCCH in some cases.

CSF absorption shows a specific dynamics in the immature brain, predominating the minor pathway of CSF absorption and lack of development of the major pathway, as proposed in 2006 by Oi and Di Rocco in the evolution theory in CSF dynamics [35, 39]. Concerning the case in our cohort mentioned above with early failure, the patient was 8 months of age at the time of ETV, and was the only case in our series in which ETV was performed before the age of 12 months. This failure can be assumed to be due to the predominance of the minor CSF absorption pathway.

Concerning the patency of the ventriculostomy, in all our patients, it was possible to identify an artifact in the stoma, even in the cases of clinical ETV failure. In one case, the patient experienced clinical improvement for 18 months, after which persistent headache developed with a recording of pathological intracranial pressure. Another case involved a scarce fine artifact, with little improvement in the radiological indexes but with data suggesting clinical improvement during 30 months, after which there was an enlargement of the cranial perimeter, psychomotor deterioration, tremor, and occasional headache. The third failure in the series concerned an early failure in an 8-month-old girl, with early development of a pseudomeningocele with the risk of a leak. All three cases required shunt placement. The presence of an artifact in these cases of failed ETV suggests the need for a combination of clinical and radiological criteria to assess ETV success.

The question therefore remains of whether it is possible to perform ETV in CCCH with no radiological data of noncommunication or obstruction of the subarachnoid space (at least until Dandy's key cisterns). Greitz's hydrodynamic theory and the more modern pulsatile vector theory of Preuss justify the possible performance of ETV in pure communicating hydrocephalus via attenuation of the reflection wave produced after opening a second communication, this time in the third ventricle floor [36, 37]. The improvement in the transmission of the pulse wave in the subarachnoid space could, in part, account for the improvement in the radiological parameters in these patients.

Conclusion

Our data show that ETV is associated with a good result in the treatment of children with CCCH. A combination of radiological and clinical criteria increases the reliability in the definition of ETV success. Pending further studies with larger sample sizes, ETV appears to be a valid alternative for the treatment of these patients.

Declarations

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