Failure of third ventriculostomy in the treatment of aqueductal stenosis in children

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Object. The goal of this study was to analyze the types of failure and long-term efficacy of third ventriculostomy in children.

Methods. The authors retrospectively analyzed clinical data obtained in 213 children affected by obstructive triventricular hydrocephalus who were treated by third ventriculostomy between 1973 and 1997. There were 120 boys and 93 girls. The causes of the hydrocephalus included: aqueductal stenosis in 126 cases; toxoplasmosis in 23 cases, pineal, mesencephalic, or tectal tumor in 42 cases; and other causes in 22 cases. In 94 cases, the procedure was performed using ventriculographic guidance (Group I) and in 119 cases by using endoscopic guidance (Group II). In 19 cases (12 in Group I and seven in Group II) failure was related to the surgical technique. Three deaths related to the technique were observed in Group I. For the remaining patients, Kaplan–Meier survival analysis showed a functioning third ventriculostomy rate of 72% at 6 years with a mean follow-up period of 45.5 months (range 4 days–17 years). No significant differences were found during long-term follow up between the two groups. In Group I, a significantly higher failure rate was seen in children younger than 6 months of age, but this difference was not observed in Group II. Thirty-eight patients required reoperation (21 in Group I and 17 in Group II) because of persistent or recurrent intracranial hypertension. In 29 patients shunt placement was necessary. In nine patients in whom there was radiologically confirmed obstruction of the stoma, the third ventriculostomy was repeated; this was successful in seven cases. Cine phase-contrast (PC) magnetic resonance (MR) imaging studies were performed in 15 patients in Group I at least 10 years after they had undergone third ventriculostomy (range 10–17 years, median 14.3 years); this confirmed long-term patency of the stoma in all cases.

Conclusions. Third ventriculostomy effectively controls obstructive triventricular hydrocephalus in more than 70% of children and should be preferred to placement of extracranial cerebrospinal shunts in this group of patients. When performed using ventriculographic guidance, the technique has a higher mortality rate and a higher failure rate in children younger than 6 months of age and is, therefore, no longer preferred. When third ventriculostomy is performed using endoscopic guidance, the same long-term results are achieved in children younger than 6 months of age as in older children and, thus, patient age should no longer be considered as a contraindication to using the technique. Delayed failures are usually secondary to obstruction of the stoma and often can be managed by repeating the procedure. Midline sagittal T2-weighted MR imaging sequences combined with cine PC MR imaging flow measurements provide a reliable tool for diagnosis of aqueductal stenosis and for ascertaining the patency of the stoma during follow-up evaluation.

KEY WORDS • aqueductal stenosis • hydrocephalus • third ventriculostomy • neuroendoscopy • cine phase-contrast magnetic resonance imaging • long-term outcome
nique from one in which ventriculographic guidance was used to one in which direct endoscopic guidance was used, we have included patients treated earlier in our study to ascertain the long-term efficacy of the technique.

Clinical Material and Methods

Between March 1973 and December 1997, 336 patients underwent third ventriculostomy, 106 in procedures performed using ventriculographic guidance (as described by Guiot and associates7,8) and 230 in procedures in which endoscopic guidance was used (as described by Sainte-Rose and colleagues20,21). Patients with posthemorrhagic and postmeningitic hydrocephalus, myelomeningocele with Arnold–Chiari malformation, Dandy–Walker syndrome, and posterior fossa tumors were excluded from this retrospective study because in these cases the cause of the hydrocephalus and the site of obstruction were multifactorial and the different subgroups contained too few patients to allow for interpretation.

Only patients with obstructive triventricular hydrocephalus due to purely mechanical obstruction of the sylvian aqueduct (primary aqueductal stenosis, toxoplasmosis, tectomesencephalic tumor or hamartoma, pineal tumor, posterior thalamic tumor, and all cases of shunt malfunction presenting with sylvian aqueduct syndrome1 or global dorsal midbrain dysfunction) were included. We performed a retrospective analysis on the following factors: the patient age at presentation, symptoms and signs at the time of the diagnosis, findings of neuroradiological examinations performed at the time of diagnosis and during the follow-up period, operative notes made during the third ventriculostomy, ventriculography x-ray films obtained during the procedures in which ventriculographic guidance was used before September 1987, video recordings of the procedures in which endoscopic guidance was used beginning in September 1987, the patient’s clinical picture, and the results of the radiological examination performed at the last follow-up visit.

Preoperatively, all patients treated before 1978 were studied radiologically by using ventriculography, between 1978 and 1987 by computerized tomography (CT) scanning and ventriculography, between 1987 and 1990 by MR imaging, and after 1991 by MR imaging with a cine phase-contrast (PC) flow study of the aqueduct. The ventriculographic criteria for aqueductal stenosis or obstruction have already been described.19 The MR imaging criteria18 included: absence of the whole length of the aqueduct on midline sagittal T1- and T2-weighted images (Fig. 1 upper left); absence of a systolic/diastolic flow on cine PC MR imaging measurements (Fig. 1 upper right); significant dilation of the lateral and third ventricles; a normal fourth ventricle; and morphological changes at the level of the mesencephalon and floor of the third ventricle, as described by other authors.12,13,17

At surgery, the criteria used to define a successful pro-
procedure were the rapid passage of contrast medium into the subarachnoid spaces of the posterior fossa when the procedure was performed with ventriculographic guidance and clear visualization of both the clivus and the basilar artery when the procedure was performed with endoscopic guidance. In the absence of one of these criteria the procedure was considered a technical failure.

Postoperatively, most patients underwent CT or MR imaging either immediately after surgery or later during the follow-up period. The CT criteria for a successful procedure were the absence of transependymal resorption and the increased visibility of the subarachnoid spaces over the convexity. The MR imaging criteria used to assess the patency of the stoma were the presence of a flow-void artifact on T2-weighted images (Fig. 1 lower left) and the flow measurements obtained using a commercially available cine PC sequence showing the presence of a systolic/diastolic flow through the stoma (Fig. 1 lower right).

In the postoperative period and during follow up, the criteria used to define a successful procedure were complete resolution of all symptoms and signs and disappearance of all radiological signs of active hydrocephalus. In the absence of one of these criteria, an additional surgical procedure was performed and treatment was, therefore, considered a failure.

**Statistical Analysis**

The long-term effectiveness of the treatment was studied by performing Kaplan–Meier survival analysis with the aid of a commercially available statistical software program (SPSS for Windows, version 8.0.0; SPSS, Inc., Chicago, IL). All patients who benefited from a successful procedure were enrolled in the study on the day of the third ventriculostomy and remained in the study until their last follow-up date. In case of treatment failure, which was determined by at least one of the criteria listed earlier, the day of the surgical procedure required to treat the failure was considered as the endpoint.

**Results**

There were 213 patients: 120 boys and 93 girls. The causes of the triventricular hydrocephalus are shown in Table 1. In 94 cases the procedure was performed with ventriculographic guidance (Group I) and in 119 cases with endoscopic guidance (Group II). The details of patient age distribution are shown in Fig. 2. No significant differences in patient age or gender or causes of the triventricular hydrocephalus were observed between the patients treated with the two techniques. Symptoms and signs at the time of presentation are summarized in Table 2. The follow-up review of the whole series (mean 45.5 months; median 23.3 months; range 4 days–17 years), excluding technical failures, showed a functioning rate of 72% at 6 years (Fig. 3). Patients younger than 6 months of age had a higher number of failures overall. This difference was confirmed in patients in Group I, whereas no difference was found between the two age groups in patients in Group II (Fig. 4). Long-term patient outcome and its relation to cause are shown in Fig. 5.
Ventriculography (Group I)

In 12 (12.8%) of the 94 cases in which surgery was performed with the aid of ventriculographic guidance, the surgeon failed to achieve good communication between the third ventricle and the subarachnoid spaces and these cases were classified as technical failures. In three of the cases, a severe hemorrhage led to death of the patient. In four cases no passage of contrast medium was observed through the hole and in three cases hemorrhage after perforation of the floor required external drainage and insertion of a ventriculoperitoneal (VP) shunt some days later. In two cases of severe macrocramium, the trochar proved to be too short and a very small hole was obtained. Most technical failures occurred during the first 5 years in which the technique was used.

In 82 cases third ventriculostomy was successfully performed. The follow-up period ranged from 20 days to 17.4 years (mean 6.32 years, median 4.1 years). In 21 (26%) of these cases a diagnosis of delayed treatment failure was made during follow-up review (range 1 day–4.9 years). Symptoms leading to the diagnosis of failure in most cases (14 cases) included: progressive macrocramium with or without delayed psychomotor development (seven cases), headache (three cases), vomiting (three cases), decreased consciousness level (three cases), gait disturbances (two cases), and seizures (one case). There was a significantly higher failure rate (50%) at 6 years postoperatively in patients who were younger than 6 months of age at the time of the surgery compared with patients older than 6 months (20%).

In 18 cases treatment failure was managed by insertion of an extracranial CSF shunt. In three patients in whom treatment failure was diagnosed at 23, 29, and 51 months, respectively, after the first procedure, repeated ventriculography revealed obstruction of the stoma. A second third ventriculostomy was successfully performed and the patients remained free of symptoms during follow-up periods lasting 8, 15, and 11 years, respectively.

Fifteen patients underwent MR imaging 10 years or later after they underwent third ventriculostomy (range 10–17 years, mean 13.9 years, median 14.3 years). In all cases a flow-void artifact was observed on sagittal T2-weighted images (Fig. 6) and a measurable systolic/diastolic flow was found on cine PC MR imaging sequences.

Endoscopy (Group II)

In seven (5.8%) of the 119 cases in which surgery was performed with endoscopic guidance, the surgeon failed to achieve good communication between the third ventricle and the subarachnoid spaces; these cases were classified as technical failures. In one case a severe venous hemorrhage occurred after perforation of the floor of the ventricle, resulting in transient left hemiparesis and ventricular hemorrhage that required external drainage and, eventually, placement of a VP shunt. In six cases the procedure was aborted because there was mild hemorrhage affecting operative visibility (three cases) or because the anatomy of the floor was not considered adequate for perforation (three cases). In two of these patients, the procedure was repeated successfully a few days later and, on the day of the second procedure, these two patients were entered into the long-term follow-up study. As shown in Fig. 7, most technical failures were observed at the beginning of our experience with endoscopic surgery. The failures were equally distributed among the three surgeons.
who performed most of the procedures; for each surgeon failures occurred in the first 20 cases that were surgically treated (Fig. 7).

In 114 patients a third ventriculostomy was successfully performed (in two cases at the second attempt). Follow up ranged from 4 days to 9 years (mean 2.1 years, median 1.1 years). In 17 (15%) of these patients a delayed treatment failure was diagnosed (according to the criteria already described) during follow-up examination, which occurred 10 days to 5.9 years postoperatively (mean 10 months, median 3 months). Symptoms leading to diagnosis of failure included: progressive macrocranium with or without headache and vomiting (six cases), delayed developmental milestones (two cases), gait disturbances (three cases), decreased level of consciousness (two cases), visual deterioration (one case), and enlarging ventricles in asymptomatic patients (three cases). Although patients younger than 6 months of age experienced treatment failure earlier than patients older than 6 months, no overall long-term difference was observed (Fig. 4).

In 11 cases treatment failure was managed by insertion of an extracranial CSF shunt. Cine PC MR imaging performed in six cases in which there was treatment failure demonstrated no flow through the third ventriculostomy in five cases and good flow in one case.

In two cases in which the failure was diagnosed 3 months and 6 years, respectively, after the first procedure, cine PC MR imaging revealed obstruction of the stoma (Fig. 8 left). A second endoscopic procedure confirmed the obstruction of the stoma. In both cases third ventriculostomy was repeated, but failed to control the symptoms. Shunts were placed in these patients 20 and 30 days, respectively, after the second procedure.

In four cases of treatment failure, which were diagnosed 1, 6, 14, and 25 months, respectively, after the first procedure, cine PC MR imaging revealed obstruction of the stoma (Fig. 8 left). A second endoscopic procedure confirmed the obstruction of the stoma (Fig. 8 center); third ventriculostomy was repeated and the patients were free of symptoms at a follow-up duration of 5, 5, 6, and 40 months, respectively (Fig. 8 right).

Discussion

Since the experience of Guiot and associates in the 1960s, third ventriculostomy has been considered to be a good alternative to VP or ventriculoatrial shunt placement in patients affected by obstructive triventricular hydrocephalus. This is particularly true in cases in which aqueductal stenosis is induced by a mechanical obstruction within the lumen of the aqueduct, such as septations or membranes, or by an extrinsic compression due to a tumor of the tectal plate or pineal gland, with no additional obstacle in the CSF pathways distal to the obstruction.

By forming a communication between the third ventricle and the subarachnoid space, an almost normal physiological CSF circulation should be restored. Moreover, the very low infection rate, absence of foreign material, lower morbidity and mortality rates, absence of all overdrainage-related complications, and absence of the need for repeated revisions are obvious advantages that this technique offers over extracranial CSF shunt placement.

Nevertheless, despite these theoretical advantages and the ability to select patients accurately, several reports and our results show that, in a significant number of these patients, the symptoms and signs of increased intracranial pressure related to hydrocephalus are not controlled by third ventriculostomy. Additionally, technical failures related to the experience of the surgeon occur and a higher failure rate has been reported in neonatal patients.

Finally, before third ventriculostomy will be widely accepted as the treatment of choice for these patients, a detailed long-term follow-up study is required.

Technical Failures

The retrospective analysis of patients who underwent surgery with the aid of ventriculographic guidance shows that this technique should be abandoned because of the lack of visual guidance during the procedure and because of the high mortality rate (3%) encountered in its use.

In patients who underwent surgery in which endoscopic guidance was used, retrospective analysis of the operative films revealed that, in the three cases in which the procedure was aborted because of mild intraventricular bleeding, this probably could have been managed by intraoperative irrigation, which has been systematically used since 1994 in our department. Figure 7 clearly shows the progressive decrease in technical failures over time, with
virtually no failures in the last 72 patients during a 4-year period (1994–1997). The distribution of technical failures according to surgeon shows that the failures seem to be related to surgical experience rather than to the individual surgeon. A near-zero technical failure rate should, therefore, be achievable after reasonable training in endoscopic surgery.

**Early and Delayed Treatment Failures**

After a successful procedure, treatment failure can be diagnosed early (six patients underwent repeated surgery within the 1st month); however, it is usually seen later, after an initial period of complete resolution of all signs and symptoms. Early failures can be explained by a multifactorial etiology of the hydrocephalus, combining an obstructive component at the level of the aqueduct and an additional obstacle to CSF circulation at the level of the subarachnoid spaces. In these cases, third ventriculostomy would allow a transient improvement as a result of the ventricular tapping performed during the procedure, followed by a recurrence of the symptoms of intracranial hyperp-tension in the days following the procedure. In these cases, sagittal T2-weighted MR imaging performed after the procedure usually demonstrates a good flow artifact at the level of the third ventriculostomy and cine PC MR imaging flow measurements reveal a good systolic/diastolic flow at the level of the stoma in spite of a slowly worsening clinical picture.

The pathophysiological mechanisms of delayed failures are probably more complex. Obstruction of the stoma due to proliferation of gliotic tissue or arachnoid membranes seems to be the most probable mechanism; this has already been described by some authors\(^{8-11}\) and was observed both radiologically and endoscopically in our series (Fig. 8 left and center).

In fact, all but one of the neuroradiological studies performed in the patients presenting with delayed treatment failure (three ventriculographies and 12 cine PC MR imaging studies) demonstrated absence of flow through the stoma. The obstruction was visually confirmed in all patients who underwent reoperation with endoscopic guidance and an additional opening in the floor of the third ventricle usually resulted (seven of nine cases) in long-term resolution of the symptoms of intracranial pressure. Therefore, secondary obstruction of the stoma seems to be the most frequent cause of delayed failure of third ventriculostomy in cases of obstructive triventricular hydrocephalus.

In all patients presenting with recurring symptoms of increased intracranial pressure, cine PC MR imaging should be performed to ascertain the patency of the stoma. If the flow-void artifact is not visible on sagittal T2-weighted images or if the flow measurements do not show a systolic/diastolic flow through the stoma, a second endoscopic procedure should be performed to ascertain the diagnosis of obstruction.

**Patient Age Factor**

Our earliest experience in performing third ventriculostomy with the aid of ventriculographic guidance\(^{9,10,19}\) showed that a higher failure rate could be expected when the procedure was performed in patients affected by hydrocephalus in the first 6 months of life. The same concern has been raised in more recent reports.\(^{3,6}\) However, this observation was not confirmed in those patients who underwent third ventriculostomy in which endoscopic guidance was used in the last 10 years (Fig. 4). In this group, the only difference observed was in the timing of the failure, which usually occurred soon after the procedure in patients younger than 6 months of age. Two factors might be responsible for the improved long-term outcome in the neonatal group. First, findings on MR imaging and cine PC flow measurements probably allow the surgeon to make a more accurate patient selection. Second, endoscopy provides direct visualization of the third ventricle floor and the anatomical landmarks of the interpeduncular cistern under magnified conditions, allowing for greater safety and efficacy of the procedure.

![Fig. 8. Magnetic resonance images and endoscopic view obtained in the same patient as shown in Fig. 1. Six months after third ventriculostomy, the patient was readmitted because at follow up a sagittal T2-weighted MR image (left) revealed increased ventricular dilatation when compared with the MR image in Fig. 1 lower left, as well as transependymal resorption and disappearance of the flow-void artifact at the level of the stoma. Center: Operative view of the second endoscopic procedure revealing that the stoma is obstructed by a heterogeneous membrane presenting different degrees of thickness and lucency. A new third ventriculostomy was performed without encountering specific technical difficulties. Right: Sagittal T2-weighted MR image obtained 2 weeks after the second third ventriculostomy. Note the decreased ventricular dilation and the flow-void artifact at the level of the stoma.](image-url)
The larger size and the more precise location of the stoma in the floor of the third ventricle might play a role in young patients in whom the risk of secondary obstruction may be high.

Therefore, all children with neonatal hydrocephalus who have no history of meningitis or intraventricular hemorrhage should be studied by $T_2$-weighted MR imaging in sagittal views with cine PC flow measurements of the aqueduct. In those cases in which there is aqueductal stenosis, endoscopic third ventriculostomy should be performed and close follow-up evaluation should be ensured in the 1st year following the procedure to detect early treatment failure.

**Long-Term Follow Up**

Some authors have already demonstrated that the persistence of significant ventricular dilation after third ventriculostomy does not affect the patient’s intellectual outcome. Concerns about the long-term effectiveness of the third ventriculostomy have been raised because of the lack of reports concerning its application in large numbers of patients with very few data about long-term follow up when compared with the data available in the literature on long-term outcome of patients treated with extracranial CSF shunts. Life table analysis of the present series of patients shows that all treatment failures were observed in the first 5 years following the procedure (Fig. 3). No recurrence of symptoms or signs of intracranial hypertension were seen and no patient required reoperation beyond 5 years. Long-term cine PC MR imaging studies confirm that third ventriculostomy ensures a satisfactory intracranial CSF shunt up to 17 years after the procedure, providing a long-term and probably definitive cure of obstructive triventricular hydrocephalus in the majority of patients.

**Conclusions**

Third ventriculostomy is an effective procedure that allows for a long-term intracranial CSF shunt in more than 70% of children affected by obstructive triventricular hydrocephalus and should be the treatment of choice. Using ventriculographic guidance, the procedure has a higher technical failure rate, a higher mortality rate, and a higher treatment failure rate in children younger than 6 months of age and is, therefore, not recommended. Children younger than 6 months of age who undergo surgery performed with endoscopic guidance achieve the same long-term results as older children and, thus, patient age should no longer be considered to be a contraindication to using the technique. Delayed treatment failures are mainly related to secondary obstruction of the stoma and can usually be managed by repeating the procedure. Midline sagittal $T_2$-weighted MR imaging sequences combined with cine PC MR imaging flow measurements provide a reliable tool for diagnosis of aqueductal stenosis and for confirming the patency of the stoma.

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**References**