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Myelomeningocele: the management of the associated hydrocephalus

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Abstract
Background The pathogenesis of the hydrocephalus associated with myelomeningocele (MMC) has been the subject of an extensive number of studies. The contemporary reduction of the incidence of the Chiari II malformation and of the associated active hydrocephalus after closure of the spinal defect in utero is in line with previous studies suggesting a prominent role of the posterior cranial fossa abnormalities, where even the increased venous pressure might be at least mostly a consequence of the constriction of the posterior cranial fossa structures. Pure absorptive abnormalities however coexist, the main ones documented to be abnormal cisternal spaces and peculiar cerebrospinal fluid chemical features.
Materials and methods We reviewed the pertinent literature concerning the pathogenesis and management of the hydrocephalus associated to MMC. We also reviewed our personal experience in managing the hydrocephalus in such patients through an endoscopic third ventriculostomy.
Results and conclusions The literature review demonstrated an overall reduction in more recent series of children with MMC needing to be treated for the associated hydrocephalus postnatally, questioning the role of the prenatal care of the disease in this context. Less severe conditions and a more conservative neurosurgical attitude have certainly contributed to the reduction of the reported active postnatal hydrocephalus rate. Long-term cognitive evaluation of the children with MMC that we managed with an endoscopic third ventriculocisternostomy (ETV) as primary as well as secondary procedure did not demonstrate significant differences in the outcome compared with non-complicated extrathecally shunted children, favouring ETV as a valuable option in this subset of patients.

Keywords Hydrocephalus · Myelomeningocele · Pathogenesis · ETV

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Introduction

Most of the features which characterise the hydrocephalus associated to myelomeningocele (MMC) were already pointed out in late 1970s of the last century such as, for example, its high incidence and its adverse prognostic significance in terms of intellectual development and survival as well as its multifactorial and complex pathophysiology [52].

It was noticed in fact that only one out of six infants born with MMC presented signs of increased intracranial pressure at birth and that only one out of eight of them had a head circumference (HC) above the 98th percentile. It was also observed how the hydrocephalus became obvious clinically, eventually in some cases after the spinal defect repair, in a further 65 % of the affected children in early postnatal life with a peak in its recognition at 2–3 weeks of age and how irregular its progression could be subsequently. Consequently, it was emphasised that the HC at birth—in most cases inferior to the 50th percentile—did not have any predictive value for the occurrence of the hydrocephalus as well as for its successive evolution.

Some characteristics of this specific type of hydrocephalus, which have relevant implications for its management, were already recognised at that time in spite of the poor available diagnostic tools. Aqueductal stenosis was found in almost three fourths of the cases using air ventriculography with several subjects associating narrowing of the aqueduct

to more distal block to cerebrospinal fluid (CSF) circulation. However, radioisotopic agents and other suspensions were reported to flow through aqueducts which appeared to be occluded on air ventriculograms [8, 50]. Only one fourth of the children with MMC and subsequent hydrocephalus were believed to have a communicating type hydrocephalus. However, such a distinction was not relevant with regard to the surgical treatment as only extrathecal CSF shunting procedures were utilised in that time. The currently increasing role of endoscopic third ventriculocisternostomy (ETV) has led to a renewed interest towards the understanding of the pathophysiological mechanisms at the base of the hydrocephalus associated with MMC in order to select the best candidates and the most appropriate time for this type of procedure.

Pathogenetic mechanisms of the hydrocephalus associated to MMC

In spite of the numerous studies aimed at understanding the pathogenesis of the ventricular dilation accompanying MMC, this peculiar type of hydrocephalus remains still relatively obscure [69]. Most of its pathogenetic interpretations appear to have been influenced by the mere consideration of the associated anatomical abnormalities which could impact on the CSF dynamics rather than be based on objective scientific demonstrations. In such a direction, Raimondi is credited to have coined the definition "constrictive hydrocephalus" by taking into account the overcrowding of developing nervous and vascular structures within an osseous container unable to accommodate the volumetric growth of the brain. In 1989, in Raimondi's laboratory, Mc Lone and Knepper [32] propounded that an insufficient mesenchymal induction during the embryonic life at the level of the future posterior cranial fossa due to the loss of CSF through the spinal defect was the actual cause of the hypoplasia of the structure which in turn accounted for the caudal descend of the hindbrain and the secondary hydrocephalus. Such a hypothesis was confirmed in recent years when the intrauterine repair of the spinal defect was seen to be associated with a minor incidence and severity of the Chiari type II malformation, typically found in myelodysplasic subjects and possibly of the associated hydrocephalus [55, 63].

Chiari type II malformation with its impact at the foramen magnum eventually leading to occlusion of the outlets of the fourth ventricle and impairment of the CSF circulation at the cervico-medullary junction constitutes certainly the main factor accounting for the obstructive nature of the hydrocephalus associated to MMC [34, 54, 59]. However, other possible obstructions in CSF circulation do exist in myelodysplasic individuals. The vertical translation of the brain stem and its caudal dislocation may cause an increased resistance to CSF movements through the tentorial hiatus and within the aqueduct. The last canal may be primarily malformed with commonly occurring focal stenosis and forking [58].

On the other hand, the hypoplastic posterior cranial fossa and cerebellar prolapse commonly result in abnormal tension within the too small bone container with consequent compression and increased resistance to the cerebral venous outflow which may generate a communicant type hydrocephalus (Figs. 1 and 2). Besides the defective CSF absorption due to the increased venous pressure, some experimental observations suggest that other functional/structural anomalies might also contribute to the genesis of a communicating type hydrocephalus in MMC such as, for example, the abnormal presence and distribution of glycosoaminoglycans in subependimal ventricular regions [13, 41].

It is likely that a various combination of obstructive and CSF hypo-reabsorptive pathogenetic factors accounts for the different characteristics and severity of the hydrocephalus associated to MMC in the different individuals. These differences would be also responsible for the different response to the surgical treatment, namely extrathecal CSF diversion procedures or intrathecal CSF shunting.

The mild and slowly progressive hydrocephalus which may characterise a proportion of subjects with MMC is likely to occur when the obstructive pathogenetic factors play a relatively low role. In such an instance, the surgical indication can be difficult for the possible spontaneous evolution towards the arrest of the hydrocephalus or the commonly occurring failures of endoscopic third ventriclecisternostomy due to an excessively low intraventricular pressure unable to maintain patent the surgically created stomy in the floor of the third ventricle. In some of these patients, the successive evolution of the hydrocephalus from the prevalently communicating to the prevalently obstructive type, depending on the progressively increasing disproportion between the hypoplasic posterior cranial fossa and the developing hindbrain may explain the late success of the endoscopic procedure in patients who did not respond to the operation when performed in infancy.

Surgical management of the hydrocephalus associated to MMC

Extrathecal CSF shunts

Currently, a minor proportion of children born with MMC undergo the placement of a CSF shunting device than in the past time. The phenomenon may be explained according to various hypotheses, the most commonly propounded being the impact of prenatal diagnosis leading to interruption of pregnancy in most severe cases and the possible minor severity of the malformation in babies born by mothers whose nutrition is improved by folic acid supplementation.

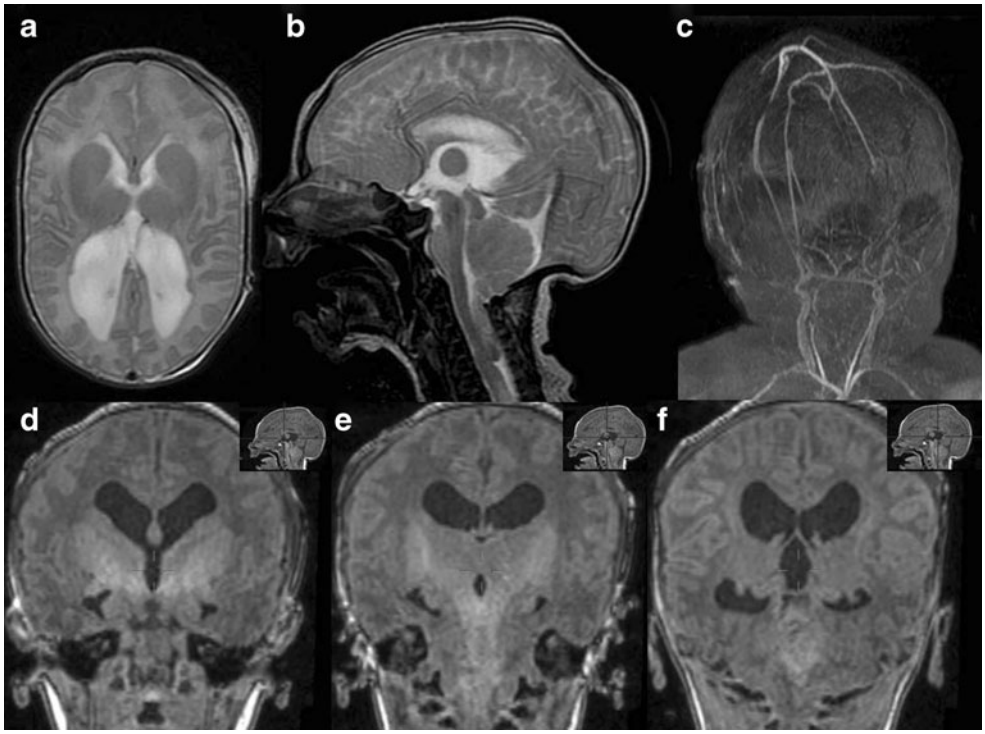

Fig. 1 **a** Axial T2-weighted MR image showing the colpocephalic appearance of lateral ventricles, with dilated posterior portions and slit frontal horns. **b** Sagittal T1-weighted image documenting the small and crowded posterior cranial fossa, associated with Chiari type II malformation and partial agenesis of the tentorium. **c** Angiographic sequence of MR demonstrating several anomalies of the venous sinuses, in particular, stenosis of the superior sagittal sinus, vertical course of the sinus rectus, and very-low-lying transverse sinuses. **d**, **e**, **f** Coronal sections depicting the ventricular anatomy along the anterior–posterior axis (see reference on sagittal *plane at the right-top corner* of each image): The third ventricle is narrow in the anterior portion, almost entirely occupied by the large massa intermedia in the middle portion and largely cystic in the posterior portion

However, the most important cause of this apparently minor incidence of hydrocephalic myelodysplasic infants requiring a CSF shunting procedure should be seen in the changed attitude of the neurosurgeon who has become reluctant to insert a CSF shunt apparatus in this particular condition because of the related high number of complications [16, 42]. Indeed, shunt-related complications and even death have been noticed to be significantly higher in hydrocephalus associated to MMC than in other types of pathological conditions requiring CSF shunting [23, 31, 40, 60]. Actually, up to 40 % of the shunted children with MMC may experience a failure of the inserted CSF shunt device already during the first postoperative year [27]. Shunt-related infections, in particular, are a common complication in hydrocephalic infants with MMC harbouring a ventriculoperitoneal (VP) CSF shunt [33, 39, 47, 60, 67]; their negative influence on cognition was already emphasised by Mc Lone and colleagues in early 1980s [32].

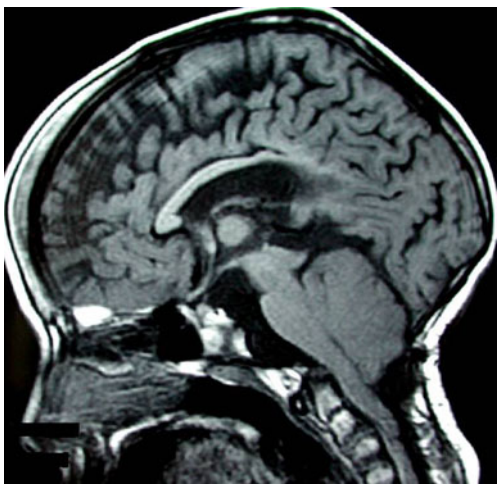

Fig. 2 Sagittal T1-weighted image of the brain showing the main obstructions of the CSF circulation, in particular, the stenosis of the aqueduct and the effacement of the fourth ventricle at the ventricular level, the cystic dilation of the prepontine cistern, the crowding of the posterior cranial fossa, and the obstruction of the foramen magnum at the subarachnoid level

In 2008, Chakraborty and colleagues [10] reported that the shunt placement rate in myelodysplasic children could be lowered to 52 % if a moderate ventricular dilation was accepted. The authors based their conclusion on what observed in a series of 56 infants born with MMC whose spinal defect had been repaired at a mean age of 3 days. These children were subsequently followed by a multidisciplinary spinal dysraphism team which evaluated their psychomotor development accurately and assessed the stability

of their hydrocephalus by means of seriated ultrasounds scan in the first postoperative year. The authors' criteria for the shunt placement were progressive ventricular enlargement, progressive increase in HC, bulging fontanelle, bradycardia, and sunsetting sign. On the other hand, the presence of pseudomeningocele or CSF leak at the closure site was not deemed an indication for shunt placement and eventually treated as local wound complications.

Interesting enough, a similar rate (54 %) of shunt placement was reported in the large series of 116 infants with MMC operated on in utero, a result mainly attributed to the reversion or minor degree of Chiari type II malformation observed following the antenatal repair of the spinal defect [6, 62, 63].

The relatively low rate of shunt depending on children being born with MMC and developing hydrocephalus reported by Chakraborty and colleagues [10] is in contrast not only with the old reports but even with the incidence described in contemporary series which ranges around 80 % of the cases [4, 22, 30, 36, 49, 53, 63, 64] with a minimum of 63 % [15] and a maximum of 91 % [5]. Obviously, this observation seems to rule out the hypothesised variations in the population characteristics while points on possible differences in the criteria adopted for the surgical indication.

It is likely that in the past time the neurosurgeons were prompted to place a shunt immediately after the ventricular dilation was recognised by the experience gained with other types of hydrocephalus, the progression of which could be very rapid and which responded to an early surgical treatment very favourably with an acceptable low rate of complications. In MMC, however, it is not rare that the hydrocephalus may slow down its progression after a transient phase of increased intracranial pressure and reach a spontaneous arrest in a significant percentage of the cases.

A further reason which may account for the high proportion of infants who undergo an early CSF shunt placement is the leakage of CSF at the site of the repair of the spinal defect which is observed in numerous instances. The event is considered by almost all the surgeons the evidence of an active hydrocephalus requiring the implantation of a CSF shunt device. The frequency of the phenomenon and the assumption that nearly all the infants born with MMC develop a progressive hydrocephalus has led to consider the possible advantages of a simultaneous correction of the spinal malformation and a CSF shunt device placement rather than following the traditional sequential approach in which the repair of the spinal defect is carried out in the first and the CSF shunt insertion later on. Those surgeons in favour of the traditional sequential approach emphasised the minor risk of infective complications when the placement of the shunt is performed after the closure of the spinal defect. Such a maneuver would eliminate the way for possible environmental contamination in infants whose clinical evolution and laboratory exams still exclude the presence of an infection. Seven to 10 days after the spinal malformation repair was suggested to be the safe time for shunt placement. Indeed, the main theoretical concern arisen was the risk that the presence of the shunt could allow the possibly contaminated CSF flow at the spinal level to reach the cerebral ventricle in infants whose immature and likely compromised immune function could not provide an effective defense [43, 60, 61]. A further argument propounded in favour of the sequential approach was that the delayed shunt placement could avoid inserting the shunt device in some children which would not develop a progressive hydrocephalus [7]. In our own experience on 170 patients, reported in 1996 [7], we observed a shunt infection rate as high as 29 % of the cases in infants who underwent the simultaneous repair of the spinal defect and CSF shunt placement while the infection rate dropped to 7 % in patients in whom the shunt was inserted late in a separate session. Furthermore, we noticed that 9 % of the patients addressed to the conventional sequential treatment did not actually need of a CSF shunt placement after the initial MMC repair.

On the other hand, those surgeons in favour of the simultaneous approach emphasised the relatively common occurrence of CSF leak from the site of the spinal malformation repair as one of the possible causes of CSF infection. Actually, there is a general agreement that CSF leak represents a major risk of infection, although some authors did not confirm such an association. A further widespread conviction considers the phenomenon to be an early indicator of a progressive hypertensive hydrocephalus. On these grounds the "simultaneous" repair of the spinal defect and placement of a CSF shunting system was advocated by several surgeons in the late 1980s of the last century [3, 9, 17, 21] for both favouring the healing of the wound on the back, that is a prophylactic measure against infections related to CSF leak, and preventing the adverse effect on cognitive function which could result from elevated intracranial pressure in case of excessively delayed shunting. Further advantages were also discussed, namely avoiding a second operation and reducing the duration of the hospitalisation.

As the undeclared but obvious assumption justifying the simultaneous approach was that nearly all the infants born with MMC develop an hydrocephalus which soon or later will necessitate of a CSF shunt operation and provided that only a minority of them show an hypertensive hydrocephalus at birth, the debate on simultaneous versus conventional sequential treatment was particularly intense in the last years of the twentieth century among those surgeons who emphasised the need to prevent damage produced by a late correction of abnormally elevated CSF pressures on a still frail and developing brain and those stressing the importance of avoiding unnecessary shunting in a subject which could not need such a procedure.

Actually, in several clinical experiences, carried out in the late 1980s of the last century, the increased risk for an

infective complication due to the early shunt placement such that performed in the simultaneous approach was not demonstrated [3, 9, 17, 21]. Similar results were reported by Parent and Mc Millan in 1995 [44] and by Miller and coworkers in 1996 [35] and more recently by Tuli and coworkers in a prospective study on a series of 189 children with MMC [61]. The authors did not find the sequential or concurrent surgical treatment to bear significantly different risks for shunt infective complications.

However, in spite of the several reports demonstrating that the CSF shunt placement may be carried out concurrently to the repair of the MMC without an increased risk for shunt malfunctioning or infection, the debate still continued in this century in countries where the incidence of MMC is still high. Actually, while Machado and Santos de Olivera in 2004 [30] and Radmanesh and coworkers in 2009 [48] affirmed that the MMC repair and shunt placement can be performed concurrently without inducing more complications than those associated with the delayed insertion of the CSF shunt device, Arslan and coworkers [1] reported that the rate of shunt infection was markedly higher in children undergoing the simultaneous management of the spinal malformation and the hydrocephalus. These last authors suggested also repeated ventricular taps to treat CSF leakage from the sac to control CSF pressure while waiting for reaching the patient's safe condition for shunt placement, a maneuver, however, which appears to contradict the justification the same authors propound to justify the delayed shunt operation, that is the prevention of infected CSF flowing from the lumbar region to the ventricles.

It is likely that the optimal time for the placement of a CSF shunt device is still far to be established as a recent study by Clemmensen and coworkers [12] appears to demonstrate. In the retrospective study by the authors, the incidence of infective complications was particular high in newborns receiving the shunt in the first 2 weeks of age as well as in infants which underwent a delayed shunt insertion after an excessively long period of watchful waiting in the hope to avoid the shunt operation and the related risk to develop shunt dependency.

Endoscopic intrathecal CSF shunt

According to Sgouros et al., the predominant features of hydrocephalus in children with myelomeningocele may change in time. In infants, the subarachnoid space deformation and immaturity combined with the increased venous outflow resistance would prevail; however, the placement of an extrathecal CSF shunt device in early age could lead to a further constriction and overcrowding of posterior cranial fossa structures, due to cranial bone overgrowth which in turn would worsen the aqueductal stenosis and change the hydrocephalus form, mainly communicating, in a mainly obstructive type [51].

Endoscopic ventricular anatomy in children with hydrocephalus and myelomeningocele

The ventricular anatomy of children with myelomeningocele has received particular attention in the last decade due to the increasing number of publications dedicated to endoscopic third ventriculostomy in the management of hydrocephalus in these patients and the attempt to better understand the reasons for its relatively high failure rate in these patients [2, 11, 14, 19, 20, 26, 28, 29, 38, 65]. In fact, different anatomic variants have been described, and some of them might potentially be relevant for the correct conclusion of an ETV and its success (Figs. 3, 4, 5, and 6); in a personal series of ten pediatric patients, Pavez et al. [45] reported the impossibility of recognising any mammillary bodies in 40 % of the cases (4/10); presence of septations in 5/10 cases; presence of atypical veins in the floor of the third ventricle in 60 % of the cases; the presence of floor umbilications in 50 % of the children (5/10) and the presence of arachnoid adherences in 70 % of them. Mori et al. [37] reviewed MR and CT findings of 21 children who underwent shunt placement for the management of the hydrocephalus related to MMC, looking for possible anatomic variants that could have interfered with the conclusion of an endoscopic third ventriculostomy. Once excluding the patients with insufficient radiological data, they reported the presence of a huge massa intermedia in 63.2 % of the cases, sloping of the third ventricle floor in 30 % of the cases, a narrow anteroposterior length of the third ventricle floor in 20 % of the children, and a narrow prepontine cistern with crowding of the posterior fossa in 38.1 % of the cases. Three of these patients were selected for an endoscopic third ventriculostomy, one as primary procedure, the remaining two at the time of shunt malfunction. The endoscopic anatomy showed as prominent features a huge massa intermedia and an opaque third ventricle floor in all cases, confirming, though on a limited sample, that the endoscopic view might show data not always predictable on radiological investigations. That the endoscopic view might show features contraindicating an ETV, in spite of a preoperative patient selection on the preoperative radiological work-up is confirmed by other authors. Three of the seven patients for whom ETV was abandoned at surgery in the series by Peretta et al. [46] (collecting 355 pediatric patients) were affected by myelomeningocele; similarly, Jenkinson et al. reported that ETV was abandoned because of improper anatomical landmarks in a total of three adult patients (3/190 cases), two of them affected by myelomeningocele [24]. In their preliminary series of 93 ETV on pediatric patients affected by hydrocephalus and open spina bifida, Warf et al. [66] reported as main endoscopic feature hampering the correct conclusion of an ETV the scarring of the interpeduncolar cisterns, described in 11 cases on a total of 74 with a report in this direction, though without specified percentages, they also reported that the basilar apex was usually not recognisable as well as the

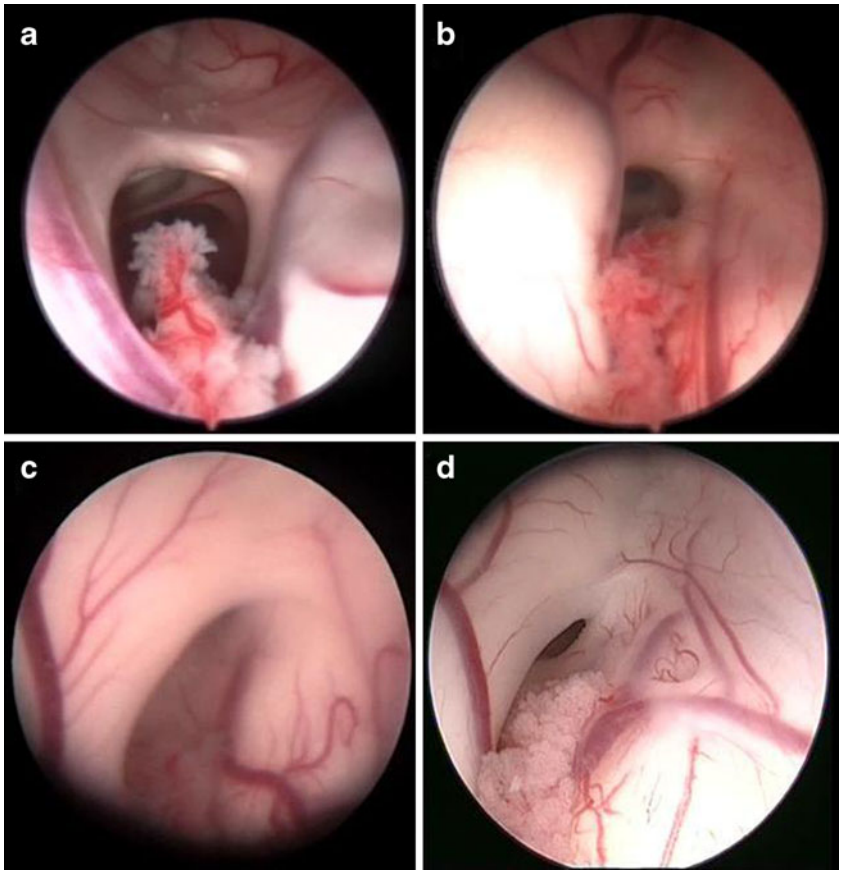

Fig. 3 Endoscopic pictures showing different appearance of the right foramen of Monro, associated to a progressively increasing difficulty to cannulate the third ventricle. **a** Horizontal, **b** oblique, **c** vertical , **d** vertical and small-sized

mammillary bodies, two further factors that might contribute to an incorrect conclusion of a third ventriculostomy in these patients.

Results of endoscopic third ventriculostomy in children with hydrocephalus and myelomeningocele

The first large series of patients with hydrocephalus and myelomeningocele treated through an endoscopic third ventriculostomy is the one of Teo and Jones, published in 1996 [57]. These authors collected the results of ETV performed in a time span of 17 years in 69 children operated on at birth for a myelomeningocele. The overall success rate of the procedure was 72 %. A significant difference however in the results was reported if the procedure was performed in children less than 6 months (success rate 12.5 %) or above 6 months (80 %). There was also a related significant difference in the success rate of ETV performed as primary procedure (success rate of 29 %) if compared with the success rate of the procedure when performed at the time of shunt malfunction (success rate of 84 %). Though not reaching statistical significance, other factors that predicted a favourable result were the presence of a triventricular hydrocephalus, a diameter of the third ventricle >4 mm, and the evidence of normal or slightly reduced subarachnoid spaces. On the other hand, the previous cerebrospinal fluid infection and/or of an intraventricular hemorrhage were negative predictive factors [57]. Similar results were reported by Jones and Kwok in a series of 25 patients operated on at a different institution; only one out of 11 patients had a successful long-term result despite initial good fenestration of the floor of the third ventricle and strict selection of the patients (adequate third ventricular size and relatively slowly progressive hydrocephalus). On the other side, 14 patients had a ventriculostomy performed instead of shunt revision. In 13 of them, the procedure had a long-term success [25]. Mori et al. [37] also observed differences in the outcome in patients aged less than 1 year as compared with their older counterpart (25 % versus 90 % success rate). Other papers in the literature, though based on single case reports or series with limited numbers of patients, have challenged the just-mentioned results, in particular, the role of age and the lower success rate of primary versus secondary ETV. Fritsch et al. [18] reported a 50 % success rate which was independent from children's age at surgery. Similar results were reported in the multicenter study of Portillo et al. [47] who referred an overall success rate of 21 % in a series of 19 myelomeningocele patients, a rate which was not related with age or time when ETV was performed. In a preliminary series coming from our institution [56], we were not able to find significant differences between infants (<6 months) undergoing primary ETV (overall success rate, 70 %) and older patients who underwent third ventriculostomy as secondary procedure at the time of

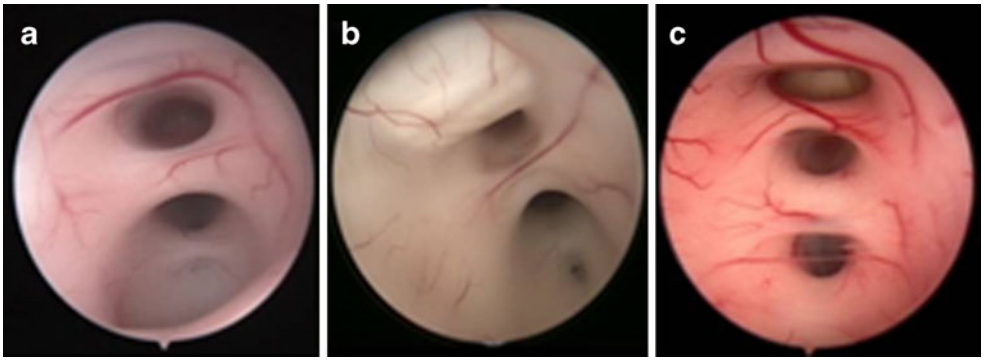

Fig. 4 Endoscopic pictures showing different appearance of the third ventricle, with increasing distortion of the ventricular anatomy. **a** Divided in two portions by the hypothalamic adhesion, **b** divided in two portions by the hypothalamic adhesion with the presence of the aqueduct in the inferior part of the visual field, **c** divided in three portions by two hypothalamic adhesions

malfunction of a previously inserted CSF shunt device (overall success rate, 60 %).

A perspective innovation to the usual performance of a third ventriculostomy in these patients was suggested by Warf et al. [66]. Based on a previous positive experience in children with posthemorrhagic and postinfective hydrocephalus, these authors proposed to combine ETV and cauterisation of the choroid plexus (CP) inside both the lateral ventricles as a primary procedure for the management of the hydrocephalus in a population of 115 children affected by myelomeningocele (mean age, 3 months). Results with a follow-up of at least 1month were available in 93 cases. At a mean follow-up time of 19 months (range, 1–49 months), 71/93 infants had a successful outcome with no need to proceed to further surgery. Of the 22 treatment failures, 14 occurred within 3 months from the operation and only three at a follow-up longer than 6 months. In a multivariate logistic regression analysis of the possible causes of treatment failure, only scarring of the CP (p= 0.026) and scarring of the cisterns (p=0.021) were statistically related to a higher failure rate. Age at the time of surgery, poor flow through the ETV, and the status of the aqueduct were not predictive of treatment failure. Seven of the 22 infants in whom ETV failed as primary procedure underwent a redo third ventriculostomy. In six of them, the ventriculostomy site was found to be obstructed by a

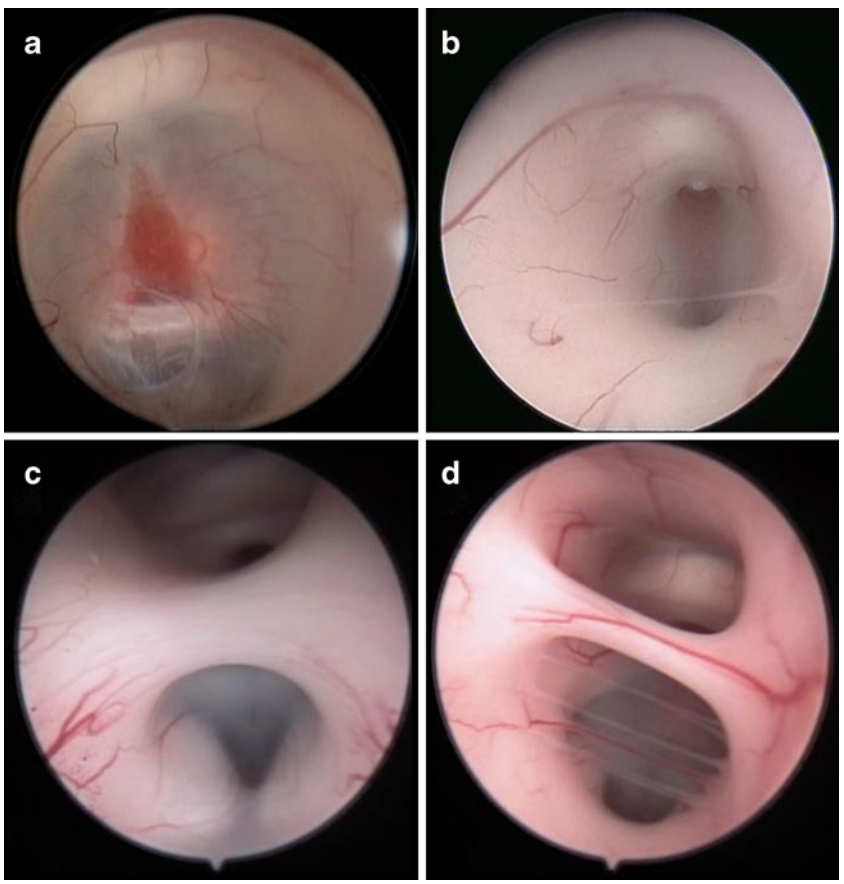

Fig. 5 Endoscopic pictures showing different appearance of the floor of the third ventricle, resulting in increasing difficulty to perform the ETV. **a** Horizontal, translucent, and thin with the well recognisable infundibulum, **b** parenchymatous with a thin hypothalamic adhesion, **c** parenchymatous with a thick hypothalamic adhesion, **d** parenchymatous with a thick hypothalamic adhesion and several vessels

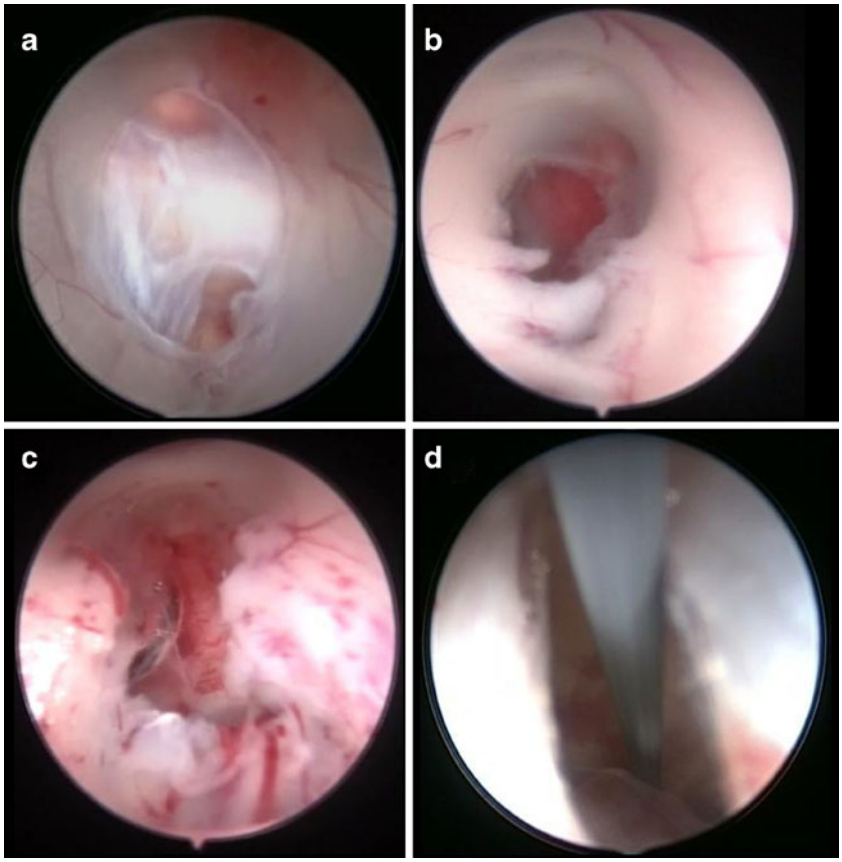

Fig. 6 Endoscopic pictures showing different anatomical aspects after fenestration of the floor of the third ventricle. **a** Multiple membranes, **b** free cisternal space under the parenchymatous floor, **c** multiple arachnoidal sinechiae under the parenchymatous floor, **d** unique arachnoidal membrane down in the cistern, extremely distant from the parenchymatous floor

scarring tissue and underwent a reopening of the ventriculostomy site, succeeding in four of them at a mean follow-up of 16.1 months. Compared with previous series, a definite added benefit resulted in these authors experience combining CPC with ETV [66]. The reduction of CSF production claimed after CPC could compensate for the "communicating" component of the hydrocephalus in these babies. One of the criticisms to the series of Warf et al. is that, as in other hydrocephalus etiologies, dealing with the hydrocephalus through ETV in the first months of life carries the risk of forcing the compensating mechanisms typical of the infants with open sutures at the cost of a chronically increased intracranial pressure with unpredictable consequences on the neurocognitive development of these patients. To address this issue, in a subsequent paper, Warf et al. [67, 68] compared the neurocognitive outcome of 55 infants who underwent an ETV-CPC procedure for the management of the hydrocephalus with the one of 19 patients who underwent the positioning of a VP shunt and 19 patients that did not need any treatment for their hydrocephalus. The modified Bailey scale for infant development (BSID-III) was the adopted method of evaluation. The evaluation was performed at a mean age of 15.6 months. The mean scale scores for untreated patients were no different from normal (all $p>0.27$) in all portions of the BSID (excluding gross motor) and were generally significantly better than those for both VP shunt-treated and ETV/CPC groups. The ETV/CPC-treated patients had nonsignificantly better mean scores than patients treated with VP shunts (all $p>0.06$), except receptive communication, which was significantly better for the ETV/CPC group ($p=0.02$). There was no difference in the size of the ventricles at the time of the evaluation as stated by the mean fronto-occipital ratio that did not correlate with outcome and did not appear significantly different between the untreated group and either the VP shunt or ETV/CPC groups. A limit in these series and in particular in the evaluation of neurocognitive outcome could still be represented by the relatively short term they have been performed.

Long-term evaluation of the results of endoscopic third ventriculostomy in children with hydrocephalus and myelomeningocele: personal experience

We reviewed all the patients affected by myelomeningocele who underwent endoscopic third ventriculostomy at our institution and who reached a minimum follow-up of 5 years. The overall series included 29 children (M/F=18/11; mean age, 16.7 months), operated on between March 2001 and October 2007. Fifteen of these patients (group I, M/F=9/6; mean age=5.4 months) underwent third ventriculostomy as primary management of the hydrocephalus (Group I), whereas 14 of them (M/F=8/6; mean age 31.8 months)

underwent ETV as secondary procedure at the time of a previously implanted shunt malfunction (Group II).

Group I Preoperative symptoms and signs were represented by an abnormal increase of the head circumference (HC increase>2 cm in 1 month) and suture diastasis in ten cases, and an associated tense anterior fontanel in five cases. The mean preoperative Evans Index on preoperative MR was 0.51.

At a mean follow-up of 6.8 years (min, 4.8 years; max, 8.9 years), control of preoperative clinical symptoms and signs was documented in 8/15 cases (53.3 %), none of them requiring further procedures for the management of the hydrocephalus. The remaining seven children underwent ventriculoperitoneal shunt implantation; three of these patients (3/7=42.8 %) needed one (two cases) or two (one case) shunt revisions because of shunt malfunction. MR controls showed a slight reduction of ventricular sizes in all cases (mean postoperative Evans Index=0.41). All patients underwent preoperative and postoperative seriated neurocognitive evaluations through the Griffith and Bailey Mental Development Scales. The mean preoperative Quotient of Intelligence Total (QIT), Quotient of Intelligence Verbal (QIV), and Quotient of Intelligence Performance (QIP) were 80, 86.5, and 71.75, respectively. At last follow-up, the mean QI values were 90.25 (QIT), 89.75 (QIV), and 89.75 (QIP). No significant differences were documented with a comparable group of 15 patients who had undergone VP shunting as primary treatment for their hydrocephalus (p=0.2).

Group II In Group II, preoperative symptoms and signs were represented by acute signs of increased intracranial pressure in 11 cases and by chronic signs (abnormal increase of the head circumference, chronic nuchal headache) in three children. The mean preoperative Evans index on MR was 0.41.

At a mean follow-up of 6.6 years (min, 5.4 years; max, 7.8 years), control of preoperative symptoms and signs was documented in 9/14 cases (64.3 %); only one of these patients underwent a redo endoscopic third ventriculostomy 4.5 years after the initial procedure; none of them required a reimplantation of the VP shunt. The remaining five children underwent shunt reimplantation at a mean time interval of 1.5 months (min, 1 day; max, 3 months). MR controls showed stable ventricular size, compared with preoperative MR examinations, but with flow documented on T2 and Fiesta MR sequences through the stoma in 3/9 cases (33.3 %). A reduction of ventricular sizes was associated in the remaining six cases (6/9 cases=66.6 %). The mean postoperative ventricular index was 0.33.

All patients underwent preoperative and postoperative seriated neurocognitive evaluations through the Griffith and Bailey Mental Development Scales in children aged ≤4 years and the Wechsler Intelligence Scales in children aged >4 years. Mean preoperative and last follow-up QIT, QIV, and QIP were respectively: QIT pre=79.25; QIT post=83.25; QIV pre=73.17; QIV post=94.75; QIP pre=61.67; QIP post=94.75.

No significant difference was documented with a comparable group of 20 patients who underwent VP shunt revision at the time of shunt malfunction (p=0.09).

In summary, though on a limited series of patients our results document that endoscopic third ventriculostomy can be considered both as primary procedure and secondary procedure for the management of hydrocephalus in myelomeningocele patients. Immediate results are almost maintained in the long-term, most of the failures occurring in the first 2 months after surgery. Though the conversion of this form of hydrocephalus from an active one to an arrested form cannot be excluded, the long-term neurocognitive evaluations demonstrate comparable results with children undergoing primary VP shunting or VP shunt revision at the time of shunt malfunction, suggesting that no substantially added damage risk is present in patients undergoing endoscopic third ventriculostomy.

Conclusions

The indication and modalities of the treatment of hydrocephalus associated with MMC are undergoing a process of revision which may be summarised as follows:

1. Nowadays, a minor number of children with MMC undergo a surgical treatment, the main reason being the possibility of evaluating the evolution of the ventricular dilation in non-operated on infants by means of repeated ultrasonographic and neuroimaging studies.
2. In case of necessity to treat an evolutive hydrocephalus, there is an increasing reluctance to place extrathecal CSF shunt devices because of their known high rate of complications, especially the infective ones.
3. Endoscopic third ventriculostomy is gaining an increasing favour due to technical improvement of the operator, more reliable physiopathogenetic interpretations, more timely surgical procedures and, more important, better knowledge of the anatomical substratum.
4. Nevertheless, hydrocephalus in MMC still constitutes the most relevant problem as far as the cognition is

concerned. Indeed, an increasing evidence is cumulating in the literature showing a stepwise probability of cognitive problem from subjects with MMC without hydrocephalus to those with arrested hydrocephalus and those with operated on hydrocephalus

5. While there are no significant differences in late cognitive outcomes between hydrocephalic subjects treated with extrathecal CSF shunt and endoscopic third ventricle cisternostomy, CSF infections, which weight extrathecal shunts nearly exclusively, remain the most dreadful complication of the surgical treatment of the hydrocephalus associated to MMC.

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