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Complex Hydrocephalus

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1. Introduction

Hydrocephalus arising from intraventricular septations is known as complex or loculated hydrocephalus. Many synonyms for complex hydrocephalus have been used in the literature such as compartmentalized or loculated hydrocephalus. Complex hydrocephalus remains a challenging neurosurgical problem. Definitive treatment is surgical, yet the approach remains controversial. Traditional treatment consisted of shunting, often requiring the placement of multiple shunt systems and multiple revisions (Ross et al., 1994; Schultz & Leeds 1973).

Some neurosurgeons advocated stereotactic aspiration of the cysts and communication of the compartments (Mathiesen et al., 1993; Ross et al., 1994), others recommended microsurgery with lysis of intraventricular cysts. Good results are reported after using the transcortical approach (Sandberg et al., 2005) or the transcallosal approach (Nida & Haines, 1973). Early experience with neuroendoscopic management is promising. Endoscopy offers a simple means of communicating isolated cerebrospinal fluid spaces and ventricles by membrane fenestration. This can be done through the same burr hole as that for the placement of a ventricular catheter. In some cases, neuroendoscopy has led to eliminating or avoiding the need for shunting (EL-Ghandour, 2006, 2008).

2. Historical perspectives

Before the advancement of neuroendoscopy, microsurgery was the primary method for fenestration of intraventricular cysts. In 1972, Rhoton and Gomez reported a case of multiloculated hydrocephalus which was operated by microsurgical fenestration (Rhoton & Gomez, 1972). In their case, multiloculated hydrocephalus was converted to a univentricular system via a small corticectomy using the microsurgical technique. The hydrocephalus was then managed by a single shunt with good short term results.

In 1982, Kleinhaus et al reported the first successful endoscopic fenestration procedure in a child who had a ventricular cyst. In their case, the bronchoscope has been used as the neuroendoscope (Kleinhaus et al., 1982). In 1986, Powers performed endoscopic fenestration of ventricular cyst using a flexible steerable endoscope and argon laser (Powers, 1986). In 1992, Powers used the same technique and performed successful fenestration in five out of seven patients (Powers, 1992).

Saline torch was used for ventricular cyst fenestration by Heilman and Cohen in 1991 (Heilman & Cohen, 1991) and by Manwaring in 1992 (Manwaring 1992). In 1993, Nida and Haines reported their experience in treating 6 patients with multiloculated hydrocephalus by transcallosal fenestration of intraventricular septations with significant decrease in postoperative shunt revision rate (Nida & Haines, 1993). Lewis et al in 1995 published the first report including a group of patients with loculated hydrocephalus (21 uniloculated and 13 multiloculated) operated by endoscopy (Lewis et al., 1995).

In 2008, EL-Ghandour reported the largest group of patients with multiloculated hydrocephalus (24 pediatric patients) operated by endoscopic cyst fenestration resulting in avoiding, eliminating or simplifying the use of shunts, with significant decrease in postoperative shunt revision rate. The results reported in this study is comparable to or even much better than those obtained previously by using the microsurgical technique. However, the endoscopic procedure has the advantage of being minimally invasive (EL-Ghandour 2008).

3. Classification

Complex hydrocephalus is classified as either uniloculated or multiloculated. Uniloculated hydrocephalus means the presence of a single cyst inside the ventricular system, whether supratentorial (isolated lateral ventricle) or infratentorial (isolated fourth ventricle). Multiloculated hydrocephalus means the presence of multiple cysts or locules isolated by multiple intraventricular septations. Uniloculated hydrocephalus is generally congenital with unaffected cerebrospinal fluid pathways, whereas multiloculated hydrocephalus is generally postinfectious or postinflammatory with obliterated subarachnoid spaces.

The distinction between both types is important because their pathogenesis, success of treatment and prognosis markedly differ. Consequently, it has been concluded that the 2 divergent types of complicated hydrocephalus should not be included in a single study (El-Ghandour, 2006, 2008).

4. Etiological factors

4.1 Uniloculated hydrocephalus

Many designations for uniloculated hydrocephalus have been used such as unilateral hydrocephalus or isolated lateral ventricle. Many types of cysts can exist within the ventricular system. Uniloculated hydrocephalus occurs if the lateral ventricle becomes trapped due to obstruction of foramina of Monro by noncolloid neuroepithelial cysts such as ependymal, choroid plexus or arachnoid cysts (Abtin & Walker, 1998). Arachnoid cysts although typically extradural, can present within the ventricles, as well as choroid plexus cysts, neoplastic cysts, and parasitic cysts (hydatid and cysticercotic cysts).

It has also been described in shunted myelodysplastic children who were treated by low pressure shunt inserted in the contralateral ventricle. The ipsilateral ventricle continues to overdrain through the low pressure shunt, whereas the contralateral or the remainder of the ventricular system becomes dilated. Most of these patients remain asymptomatic and require no treatment. However, if the patient shows progressive enlargement of the lateral ventricle contralateral to the shunt and symptoms occur, further treatment is necessary. Upgrading the shunt valve pressure or simply adding shunt to the contralateral side, has been recommended (Berger et al., 1990).

The most common presentation of uniloculated hydrocephalus is found in the shunted patients. Inflammatory and reactive changes related to shunt catheters, bleeding, infection and scar tissue formation can lead to obstruction or occlusion at the foramen of Monro, thus providing an isolated ventricle. Congenital anatomical causes may be present as well. (Abtin & Walker 1998).

The fourth ventricle can also become isolated due to obstruction of cerebrospinal fluid at the level of the aqueduct of Sylvius and the fourth ventricle outlets, in patients in whom a shunt has already been placed for postmeningitic or posthemorrhagic hydrocephalus. The cerebrospinal fluid produced from the choroid plexus gradually accumulates, resulting in progressive dilatation of the fourth ventricle followed by compression of the brain stem and cerebellar parenchyma. A well functioning lateral ventricular shunt not only decreases the supratentorial pressure, but also reduces the pressure needed inorder to keep the aqueduct open, which occasionally leads to aqueductal collapse. Consequently, the lateral and third ventricles become decompressed, thus leaving the fourth ventricle dilated.

Conventionally, isolated fourth ventricles have been managed by insertion of a ventriculoperitoneal shunt into the fourth ventricle or by craniotomy and microsurgical aqueductal canalization or microsurgical fenestration of the outlets of the fourth ventricle. With the advent of endoscopy in neurosurgery, isolated fourth ventricle is more often treated by endoscopic procedures such as endoscopic third ventriculostomy and aqueductal reconstruction by aqueductoplasty with or without aqueductal stenting (Mohanty, 2005).

4.2 Multiloculated hydrocephalus

Multiloculated hydrocephalus has many synonyms such as multilocular hydrocephalus, polycystic hydrocephalus, polycystic brain disease or intraventricular septations. The ventricular system may become trabeculated or encysted following bacterial meningitis or germinal matrix hemorrhage (Eller & Pasternak 1985; Schultz & Leeds 1973). Predisposing factors include a low birth weight, premature birth, perinatal complications and congenital central nervous system malformations (Albanese et al., 1981). In a study including 24 pediatric patients with multiloculated hydrocephalis operated by endoscopic cyst fenestration, neonatal meningitis was the most common cause (9 cases), followed by intraventricular hemorrhage (6 cases), postoperative gliosis due to previous shunt infection (6 cases) and multiple neuroepithelial cysts (3 cases) (EL-Ghandour, 2008).

In another study including 34 cases of complex hydrocephalus operated by endoscopy, among 21 cases of uniloculated hydrocephalus, 15 cases (71%) were caused by noncolloid neuroepithelial cysts, 3 cases (14%) by choroid plexus cysts, 2 cases (10%) by postoperative gliosis, and 1 case (5%) by meningitis. Among the 13 cases of multiloculated hydrocephalus included in the same study, 4 cases (31%) were caused by intraventricular hemorrhage, 4 cases (31%) by multiple neuroepithelial cysts, 3 cases (23%) by meningitis and 2 cases (15%) by postoperative gliosis (Lewis et al., 1995).

5. Incidence

Complex hydrocephalus is mainly detected in children especially neonates, without any significant variable incidence between boys and girls. The incidence of neonatal meningitis ranges from 0.13% to 0.37% in full-term infants, and from 1.36% to 2.24% in preterm infants.

Hydrocephalus as a sequela of neonatal meningitis is well described, with an incidence of up to 31% (Albanese et al., 1981; Kalsbeck et al., 1980).

Only few reports exist on the rarer subgroup that develop multiloculated hydrocephalus. Many patients with bacterial meningitis also develop ventriculitis, reported as many as 92% of cases on autopsy and 100% of cases seen clinically (Albanese et al., 1981). Postmeningitic hydrocephalus is often believed to be communicating in nature due to an insult at the arachnoid granulations. Based on the pathological findings, however, components of both communicating as well as obstructive hydrocephalus may be present.

6. Pathophysiology

The pathogenesis is still unclear, but believed to be secondary to inflammatory changes and gliosis after ventriculitis, especially commom when caused by Gram-negative organisms (Kalsbeck et al., 1980). It seems that the common link between neonatal meningitis, shunt infection and intraventricular hemorrhage is the resultant ventriculitis. It is believed that the septations giving rise to multiple loculations probably represent the organization of intraventricular exudates and debris produced by ventriculitis, regardless of whether it is chemical or infectious in origin. An inflammatory response at the ependymal surface could encourage proliferation of subependymal glial tissue, upon which exudates and debris organize and serve as a nidus for the formation of septations that span the ventricles.

The septations not only alter the ventricular anatomy, but also disrupt the normal flow of cerebrospinal fluid leading to its accumulation within a loculated cavity with progressive dilatation and mass effect (Nida & Haines, 1993). Grossly, the usual findings are ventricular dilatation, and compartmentalization by membranes. The membranes appear translucent and vary in thickness. Microscopically the septations are composed of fibroglial elements with round and polymorphonuclear cells. The characteristic findings of chronic ventriculitis are frequently present, such as subependymal gliosis, small areas of denuded ependyma, and glial tufts extending through the denuded ependyma into the ventricular lumen (Schultz & Leeds, 1973).

7. Clinical features

Presenting symptoms are mainly those related to increased intracranial pressure leading to macrocephaly in infants and headache in older children. Other symptoms include seizures, gait ataxia, and hemiparesis. Developmental delay is reported to be more common in multiloculated than uniloculated cases. It has been mentioned that neurological status in this patient population is extremely poor.

Patients with multiloculated hydrocephalus are notoriously difficult to treat, and are often compromised intellectually. In a study including 33 pediatric patients who underwent craniotomies for fenestration of progressive multiloculated hydrocephalus, 29 patients (87.9%) were severely delayed or vegetative. Many patients were nonverbal or minimally verbal, nonambulatory, had significant motor deficits, had significant spasticity and experienced seizure disorders (Sandberg et al., 2005).

In a study including 10 pediatric patients with multiloculated hydrocephalus caused by neonatal meningitis, ventriculitis or intraventricular hemorrhage, who were treated surgically (6 patients underwent craniotomy and trancallosal fenestration and 4 patients

were treated by shunting), most of the patients were found to be seriously affected. The children reported in this study had detectable cognitive deficits, ranging from profound psychomotor retardation to mild learning disability (Nida & Haines, 1993).

In a study including 34 patients with complex hydrocephalus operated by endoscopy (21 uniloculated and 13 multiloculated), the presenting symptoms were headache in 14 cases (41.2%), developmental delay in 11 cases (32.4%), macrocephaly in 6 cases (17.6%), seizures in 5 cases (14.7%), gait ataxia in 2 cases (5.9%), and progressive hemiparesis in 2 cases (5.9%) (Lewis et al., 1995).

In another study including 24 pediatreic patients with multiloculated hydrocephalus (21 cases were infants younger than 2 years of age), operated by endoscopic cyst fenestration, all patients (100%) presented with head enlargement, 18 patients (75%) presented with developmental delay, 4 patients (16.7%) with epilepsy, and 2 patients (8.3%) with hemiparesis (El-Ghandour, 2008).

8. Diagnostic studies

8.1 Ultrasound

The use of ultrasound is a well established diagnostic method for neonates and patients with open fontanel (Machado et al., 1991). It is useful in the evaluation of patients with complex hydrocephalus, often demonstrating the cyst walls and revealing any compartmentalization that has occurred. Its advantages include that it is nonradiating, noninvasive, gives a multiplanar view and patients don't require sedation. Nevertheless, it is operator-dependant and it couldn't be considered a definitive preoperative diagnostic modality.

8.2 Computed tomography scan

Plain CT scan shows disproportionate hydrocephalus, and can be used for screening of patients. However, the cysts usually have a density similar to that of cerebrospinal fluid, and the walls are usually transparent making it usually difficult to visualize the cyst walls accurately. Often CT scan shows transversely oriented septations of varying thickness and nonuniform distribution, changing the ventricular system into one of irregular contour.

Although the septations may not be visible in the early disease process, retrospective temporal review often delineates pattern of progressive compartmentalization and asymmetrical hydrocephalus. The initially documented ventricular pattern gradually becomes unrecognizable, so that ultimately the cerebral mantle encloses a large single multiloculated cavity. In advanced stages, it may be difficult to recognize the ventricles and other anatomical structures of the brain (Nida & Haines, 1993). However, CT scan is unable to identify communication or noncommunication between cavities.

8.3 Magnetic resonance imaging

Magnetic Resonance Imaging with gadolinium is considered to be the diagnostic method of choice for patients with complex hydrocephalus. Its advantages over the CT scan include the multiplanar view providing a detailed picture in three different planes (axial, coronal, sagittal), and it is more sensitive in revealing septations. The difference in protein content of

these cysts may delineate them from the cerebrospinal fluid containing ventricular system. It can also occasionally show the etiological factor as well, such as a neuroepithelial cyst. However, MR imaging fails to provide accurate information on communication or noncommunication of the compartments in multiloculated hydrocephalus.

8.4 Contrast CT ventriculography

Contrast material injected into the ventricular system or the cyst verifies noncommunication of the cyst with the ventricular system and defines the margins of the compartments. It can be done by injecting 1-2 ml of metrizamide directly into the proximal catheter of a pre-existing shunt system, by cannulating the cyst with a 22-gauge spinal needle, or by placing an external ventricular drain. The patient's head is then rotated back and forth inorder to allow the contrast matrial to spread inside the cavity, thereby showing any comartmentalization. A second CT scan is usually done 30-60 minutes after the contrast injection.

Contrast CT ventriculography, provides absolute confirmation of noncommunicating loculations, and allows direct visualization of sequestrated ventricular compartments. However, in multiloculated hydrocephalus, it requires multiple punctures for different compartments inorder to delineate the presence or absence of communication among them. Simultaneous sampling of cerebrospinal fluid for analysis of protein content may also indirectly support the presence of noncommunicating cysts (Nida & Haiens, 1993).

9. Surgical management

The definitive treatment of complex hydrocephalus is surgical. However, a high proportion of patients have a history of prematurity and/or central nervous system infection, and require medical treatment not only for intraventricular hemorrhage but also for any associated co-morbidities. The goals of surgery are to control hydrocephalus, simplify complex shunts (that is, replacing multiple shunts with a single shunt comprising only 1 intraventricular catheter, 1 reservoir, and 1 peritoneal catheter), reducing shunt revision rate, avoiding implanting shunts if possible, and decreasing operative morbidity.

9.1 Ventricular shunting

The basic underlying pathophysiological problem is that ventricular compartmentalization interferes with the proper drainage of the dilated ventricular system, making therapeutic shunting difficult. Consequently, placement of multiple shunt systems should be done inorder to drain loculated ventricular compartments that do not communicate with each other (Kaiser, 1986). The disadvantage of additional proximal shunt catheters is that multiple noncommunicating ventricular compartments are often present, and a second or even a third ventricular catheter may not adequately drain cerebrospinal fluid from all the compartments.

The greater the number of operative procedures and hardware placed, the greater will be the risk of infection. Moreover, when a shunt malfunction or infection is suspected in a patient with complex shunt systems, both diagnosis and surgical treatment can become quite complicated. Consequently, such procedure is usually followed by multiple shunt revisions and is associated with high morbidity and mortality rates. In one series a 54%

mortality rate was reported with the remaining patients severely impaired (Kalsbeck et al., 1980). For these reasons it is better to avoid multiple shunt systems if possible (Sandberg et al., 2005). Insertion of multiperforated ventricular catheter together with puncture of the membranes has also been suggested for control of compartmentalization (Kalsbeck et al., 1980), however, this has not proven to be a reliable method.

9.2 Stereotactic procedure

Many reports stress the operative simplicity and low complication rate of the stereotactic procedure in the treatment of intraventricular cysts. However, the initial enthusiasm for stereotactic aspiration has been tempered by its lack of success in treating some patients. In some patients, a mobile cyst with tough outer capsule may be pushed away by the needle and become resistant to stereotactic puncture (Ross et al., 1994).

Moreover, stereotactic aspiration is considered to be a blind technique (i.e. there is lack of intraoperative visual control) (Hellwig et al., 2008). Most importantly, simple aspiration may lead to high recurrence rate (up to 80%), because the stereotactic procedure fails to devascularize the cyst or to create a large window (>1 cm) in the cyst wall (Mathiesen et al., 1993).

9.3 Microsurgical treatment

Reports of complex hydrocephalus treated by craniotomy and lysis of ventricular cysts have been infrequent and include small groups of patients. Good results were reported in a series of 6 patients operated by transcallosal approach (Nida & Haines, 1993), and in a series of 31 patients operated by transcortical approach (Sandberg et al., 2005). Advocates for using the microsurgical procedure say that adequate hemostasis can be confidently achieved because the intraventricular septations are dissected under direct visualization. Bleeding can be more easily controlled during microsurgery, which permits the bimanual use of the standard instrumentation, such as bipolar cautery, regulated suction, with the ability to apply topical hemostatic agents (Levy et al., 2003).

They also claim that the creation of multiple wide fenestrations can be easily performed during the microsurgical procedure because the surgical microscope provides better visualization of the various compartments and membranes under higher magnification (Sandberg et al., 2005). The stereoscopic view provided by the surgical microscope, gives the surgeon a superior depth perception, particularly during deep dissection. The familiarity of traditional microsurgical techniques makes open procedures more accessible to most neurosurgeons, avoiding the potentially hazardous learning curve needed for endoscopy (Levy et al., 2003).

Nevertheless, there are many drawbacks for this procedure. First, the transcallosal surgery is technically demanding and carries the risks of sagittal sinus thrombosis, venous infarction from division of bridging veins, inadvertant damage to the pericallosal arteries, and injury to the fornix with cognitive sequelae (Jeeves et al., 1979). Second, the transcortical approach is sometimes associated with seizures and may lead to subdural collection postoperatively, because the cortical mantle is often thinned out from hydrocephalus (Kalsbeck et al., 1980). Third, the loss of cerebrospinal fluid during cyst decompression leads to collapse of the

ventricular walls (Powers, 1992). Finally, a second craniotomy may be required to reach additional cysts within the fourth ventricle, or to place a shunt.

9.4 Endoscopic treatment

The significant potential morbidity of microsurgical technique and the high failure rate of ventriculoperitoneal shunting, prompted neurosurgeons to search for an alternative procedure. The ability of the endoscope to bring into communication isolated loculated compartments within the ventricular system with minimal cortical disruption has been recently advocated as an alternative to the use of complex shunt systems or microsurgery. The endoscope is an ideal instrument for exploration of fluid-filled cavities, and the intraventricular location of these cysts makes them accessible for the endoscopic procedure. Endoscopy is a safe and effective treatment option for complex hydrocephalus, it is less invasive and offers greater operative simplicity than microsurgery. Patients recover on the ward after endoscopic fenestration of ventricular cysts and can be discharged the following day if there are no associated medical problems; in comparison, patients spend at least 1 day in the intensive care unit after the microsurgical procedure (Lewis et al., 1990).

Endoscopic fenestration combines both advantages, the minimal invasiveness of stereotactic fenestration, and the effectiveness of microsurgery. A single burr hole provides rapid access to the entire ventricular system and avoids the pitfalls of transcallosal approach. In cases of uniloculated hydrocephalus, ventriculocystostomy offers the best chance for treating these patients. Septum pellucidotomy or septostomy can be also performed endoscopically to treat isolated lateral ventricles. Fenestration of the septum pellucidum to connect the two lateral ventricles in patients with isolated lateral ventricle will preclude the need for two shunts in the majority of patients. Isolated fourth ventricle can be also treated endoscopically by aqueductoplasty and/or aqueductal stenting (Cinalli et al., 2006). In cases of multiloculated hydrocephalus, endoscopic cyst fenestration provides wide communication between the ventricular compartments and offers the greatest possibility of avoiding the need for additional shunt catheters (El-Ghandour, 2008).

9.4.1 Endoscpic tools

Flexible endoscopes have been preferred by some neurosurgeons (Lewis et al., 1995), due to its flexibility and steerability which provides increased maneuverability. However, it requires a significant amount of expertise and its main role in multiloculated hydrocephalus in the author's view is navigating the ventricular system and performing aqueductoplasty in cases with isolated fourth ventricle. The author prefers rigid endoscopes because they have greater light intensity and superior optics which allow better visualization. The limited maneuverability of the rigid endoscopes is usually offset by careful selection of the burr hole placement site and by widening the outer edge of the burr hole which provides greater freedom in maneuvering the endoscope and selecting different trajectories (El-Ghandour, 2008).

9.4.2 Endoscopic trajectory

The approach is individually designated in each case depending on location of the cysts, entry site of the pre-existing ventricular catheter, and the need for the placement of a new shunt. Cysts located anteriorly are approached through a standard midpupillary coronal

burr hole, whereas an occipital or posterior parietal route is used for posterior or temporal loculations. The burr holes are bevelled laterally to allow the endoscope to reach the contralateral ventricle, and the trajectory is planned to fenestrate the maximum number of cysts, this can be achieved by projecting the best angle with the aid of preoperative coronal and sagittal MR imaging. Sometimes it is not possible to fenestrate all intraventricular cysts because of the high risk of damaging important neural structures during septostomies. The ventricular system or the cyst is cannulated with a 14-F peel-away sheath and stylet. Care is taken to prevent the release of cerebrospinal fluid before the endoscope is introduced. After withdrawal of the stylet, a 2 mm-diameter rigid lens (wide-angle, straight-forward, 0 degree) with angled eye piece and working channel diameter of 3 mm is inserted. In severe cases with more areas of hydrocephalus, working through multiple burr holes (multiportal) ensures more successful localization (El-Ghandour, 2008).

9.4.3 Cyst localization

Intraoperative ultrasonography has proven to be extremely useful both in localization, and assisting in directing the endoscope to the desired location providing an ongoing intraoperative orientation. It gives live feedback of the surgical progress. It is also useful in determining depth of the cysts and presence of any solid structures beyond the cyst walls, as well as any shift of internal contents during or after the fenestration. One can make evaluation concerning successful fenestration and communication between various compartments by viewing saline jet bubbles flowing from one cyst to another as seen on ultrasound. However, its disadvantages include its operative dependability, requirement of a window in the cranial vault, and potential crowding of the operating suite (Abtin & Walker, 1998).

Frameless stereotaxy or neuronavigation has increased the accuracy and safety of endoscopy, and it can be used in conjunction with endoscopy inorder to overcome the problem of distorted anatomy. However, the disadvantage of frameless technology is that the patient is often a young child and the head cannot be secured to the operating table by a 3-pin holder. Without rigid fixation, the head cannot remain in a constant position with respect to the stereotactic arc after markers are registered; consequently a high percentage of errors may occur and the patient may need to undergo re-registering. Recently, simultaneous image-guided MR imaging and endoscopic navigation without rigid cranial fixation (pinless frameless stereotactic assembly) has been used in infants. However, errors may still occur as a result of shifting of intracranial structures after ventricular or cystic access. The frameless system, as opposed to intraoperative ultrasound, is not real time. Consequently, both systems can be used during the same procedure inorder to take advantage of the strengths of both (Abtin & Walker, 1998).

There are not many data on the role of neuronavigation in conjunction to neuroendoscopy in the treatment of multiloculated hydrocephalus in the published literature. In a recently published study, 16 children with multiloculated hydrocephalus were operated by endoscopically navigated procedure. In all children, sufficient drainage of the multiloculated ventricular system was reported. In this series, the authors mentioned that they didn't encounter significant problems with the occurrence of intraoperative brain shift. They attributed this observation to 2 reasons: 1) The majority of opened compartments were small, and each by itself constituted only a small portion of the whole compartmented

ventricular system. 2) Continuous irrigation has been used throughout the procedure, thereby maintaining the existing anatomy and dimensions of the penetrated cysts and parts of the ventricular system as much as possible (Schulz et al., 2010)

9.4.4 Operative findings

In most cases, it is difficult to recognize the ventricle and other anatomical structures of the brain due to severe anatomical distorsion. Differentiation between the ependyma and cyst wall is crucial, the latter is usually light blue in color, variable in thickness (translucent in early cases), and usually mobile with cerebrospinal fluid pulsations. A yellow discolouration of the ependyma due to previous intraventricular hemorrhage is sometimes noticed, and glial tufts extending into the ventricular lumen are frequently present. All patients with complex hydrocephalus who presented in an advanced stage, had more ventricular distortion and thicker membranes. Consequently long delays before fenestration is usually accompanied by progressive loculation and worse prognosis (El-Ghandour, 2008).

9.4.5 Endoscopic cyst fenestration

Different methods of fenestrating intraventricular cysts have been reported such as steerable fiberscope, argon laser, Nd:YAG (neodymium:yttrium-aluminum-garnet) laser, saline torch, and bipolar cautery. Cyst fenestration is performed in a relatively avascular segment of the cyst wall. The fenestration is widened to more than 1 cm in diameter to prevent early reclosure. This is achieved sharply by widening the initial hole or connecting multiple holes by using the bipolar electrode. A very wide fenestration has been advocated by some authors (EL-Ghandour, 2008), because of the high incidence of reclosure of small openings due to the low pressure differential across cyst walls as well as the inflammatory origin of the disease (Figs. 1A & B). Devascularization of the cyst wall is done by coagulating its vascular supply to prevent or slow its regrowth. Pulsed irrigation with lactated Ringer solution is used to prevent thermal injury and collapse of the ventricles. Any bleeding from the cyst wall usually stops with irrigation or coagulation. After completion of cyst fenestration, real-time ultrasound can be used while injecting saline into the cyst to confirm successful communication with the ventricular system (Lewis et al., 1995).

9.4.6 Septum pellucidotomy

It is a well recognized, minimally invasive technique that is used in the treatment of isolated lateral ventricle, obviating the need for two shunts, and in some cases, the need for any shunting. Finding and fenestrating the septum in a patient with both lateral ventricles dilated is a simple task, but unfortunately, most patients requiring this procedure have one collapsed ventricle and one grossly dilated ventricle. It is extremely difficult to identify the septum pellucidum in these patients, that frameless stereotaxy or neuronavigation is recommended.

The burr hole is placed more lateral to the standard position, approximately 5-6 cm lateral from the midline. A rigid endoscope is then passed into the nondilated lateral ventricle and through the septum pellucidum under direct vision. Reaching the septum by introducing the endoscope through the dilated ventricle is risky, because the opposite ventricle is often

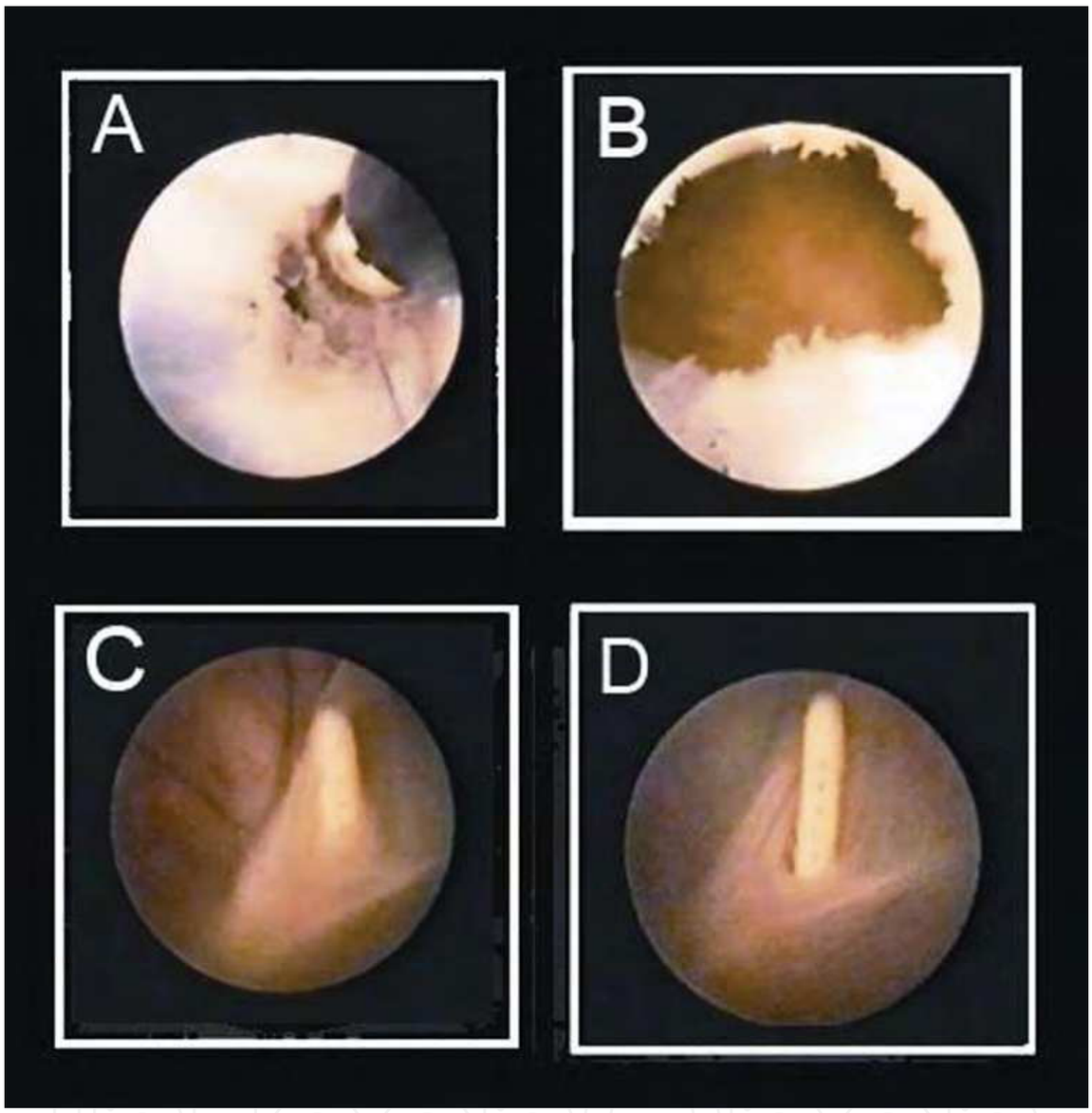

Fig. 1. A, intraoperative photograph showing bipolar electrode coagulating the cyst wall and making initial perforation. B, very wide fenestration is done in order to avoid reclosure. Note yellow discolouration due to previous intraventricular hemorrhage. C, preexisting ventricular catheter is seen obstructed by adherent membranes. D, tip of the catheter after being dissected so that it can be withdrawn safely without risking intraventricular hemorrhage. [Reprinted with permission from El-Ghandour NM (2008). Endoscopic cyst fenestration in the treatment of multiloculated hydrocephalus in children. *J Neurosurg Pediatr,* Vol 1, No. 3, (March 2008), pp:217-222, ISSN 1933-0707].

small and penetrating into the opposite ventricular wall is a potential complication. Significant delay between entering the lateral ventricle and piercing the septum may cause excessive cerebrospinal fluid to escape and this can result in an error due to the theoretical risk of brain shift.

The septum contains relatively avascular tissue, and generous septostomy can be performed safely, taking care to avoid injuring the fornices or corpus callosum. The precise anatomical position of the thinnest part of septum pellucidum is above and infront of the foramen of Monro. The use of lasers or cautery is extremely valuable in performing the septum fenestration, because it allows the surgeon an opportunity to make a larger opening. Large vessels should not be encountered during the procedure. If vessels are seen through the endoscope or if there is significant hemorrhage, this means that the endoscope is passing through or above the corpus callosum (Teo, 1998).

9.4.7 Endoscopic third ventriculostomy

It is a challenge to do endoscopic third ventriculostomy in cases of complex hydrocephalus especially in multiloculated cases, due to the changes in the ventricular architecture to the extent that the third ventricle could not be identified in most of the cases. It has been reported that the role of endoscopy is limited to fenestration, and patients (especially infants) will still need shunts to treat hydrocephalus because of immaturity of the subarachnoid cerebrospional fluid dynamics. Moreover, the pathophysiological mechanism that initially caused the septations are likely responsible for the deficiency of the absorptive capacity of the subarachnoid space due to scarring of arachnoid granulations (Teo, 1998). Nevertheless, performing endoscopic third ventriculostomy and eliminating the need for placing shunts in patients with multiloculated hydrocephalus has been reported recently by some authors (El-Ghandour, 2006, 2008).

9.4.8 Aqueductoplasty

The ideal candidate for this procedure is the premature neonate who has an isolated fourth ventricle. When the fourth ventricle expands through the tentorial incisura, endoscopic fenestration into the trigone or occipital horn of the lateral ventricle is possible. However, when a symptomatic isolated fourth ventricle remains within the posterior fossa, aqueductoplasty with or without a silastic stent becomes the procedure of choice.

The procedure is performed using the flexible endoscope which is introduced through a standard midpupillary coronal burr hole into the lateral ventricle, and hence into the third ventricle. Once the aqueduct is identified, the glial membrane, which invasively covers the orifice, is punctured with a 5-F transluminal angioplasty balloon catheter and dilated to a balloon inflation diameter of 3 mm. Cerebrospinal fluid should be observed flowing through the dilated canal. A small length of silastic tubing may be left in the aqueduct to maintain patency.

However, implanting stents carries the risk of stent dislodgement and migration (Mohanty, 2005). Manipulation of the aqueduct may also traumatize the tegmentum of the midbrain ventrally and the tectal plate dorsally. The clinical manifestation of this traumatization is the development of postoperative dysconjugate eye movements that usually resolve with time (Teo, 1998).

9.4.9 Endoscopic shunt retrieval

Removal of old malfunctioning ventricular catheters under endoscopic guidance is considered to be one of the most important roles of endoscopy in the treatment of complex

hydrocephalus. Ventricular catheters embedded in scar tissue or adherent to the choroid plexus, which previously could not be withdrawn safely without risking intraventricular hemorrhage, can now be removed under direct vision with endoscopic assistance. The YAG laser can be used to coagulate the choroid plexus and to cut along the interface of the scar and the silicone tubing in order to dislodge the catheter. Because pulsed irrigation causes movement of the choroid plexus and cyst wall, irrigation is discontinued when firing the laser. The laser is used in a fluid medium to avoid thermal injury to the surrounding structures. When removing an embedded ventricular catheter that has been placed through an occipital approach, a coronal or anterior frontal approach is preferred because the laser can be used at a trajectory perpendicular to the catheter tubing (Lewis et al., 1995).

In another study including 6 patients with pre-existing malfunctioning shunts who had previously undergone shunt drainage through occipital burr holes, another burr hole was done 2-3 cm above the old one through which the endoscope was introduced parallel to the ventricular catheter with slight inclination towards the area of scarred-in catheter tip. Four out of these 6 patients (67%) had complex shunts and simplification was performed in 3 of them (50%). The obstruction was proximal due to the presence of adherent membranes. The author found it very difficult to retrieve a pre-existing ventricular catheter in 2 out of these 6 patients (33%) which necessitated the use of bipolar diathermy probe to dissect and dislodge the tip of the catheter from adherent surrounding membranes (Figs.1C & D), so that it can be withdrawn gently and safely without risking intraventricular hemorrhage. It has been concluded that retained pre-existing malfunctioning ventricular catheters in multiloculated hydrocephalus could not be withdrawn safely without endoscopic assistance (El-Ghandour, 2008).

9.4.10 New shunt placement

Some neurosurgeons prefer placing an external ventricular drain routinely after endoscopic cyst fenestration and postponing shunting procedure to a later date (Abtin & Walker, 1998). The author doesn't put an external drain routinely except if there is intraventricular bleeding, and prefers to place a new shunt during the same session of endoscopic cyst fenestration. It has been reported that external drainage might lead to cyst collapse and initiate or accelerate early closure of the cyst fenestrations due to interference with the already existing cerebrospinal fluid pressure gradients (El-Ghandour 2008).

After removing the old catheter, the surgeon places a new catheter in the optimum position under direct endoscopic visualization. Both shunt retrieval and placement of new shunt can be performed during the same session of endoscopic cyst fenestration, provided there is no intraventricular bleeding. However, if there is bleeding it is better to insert an external ventricular drain and postpone the shunt procedure until a later date, to avoid its malfunctioning by the bloody cerebrospinal fluid (El-Ghandour, 2008).

There are many techniques which can be used for endoscopic shunt placement. The ventricular catheter may be loaded on an endoscope with the tip of the endoscope exposed for visualization. The shunt may then be placed directly into a ventricle through the appropriate trajectory, while looking through the shunt with the endoscope to confirm the shunt catheter position. If the ventricular catheter tip is in an inappropriate position, the shunt is withdrawn and modification of the trajectory may be made.

Another technique, which applies to neurosurgeons using a rod lens endoscope system, involves placing an appropriate-sized peel-away catheter into a ventricle, then placing the endoscope through the peel-away catheter. In this way, the potential shunt position is confirmed and length of the ventricular catheter can be estimated. Once the peel-away catheter is in a satisfactory position, the endoscope is withdrawn and the ventricular catheter is passed into the desired depth. The peel-away catheter is then removed while the ventricular catheter is held gently with a smooth forceps, leaving the ventricular catheter in the appropriate place (Brockmeyer, 1989).

9.4.11 Repeated endoscopic procedure

In a study including 24 pediatric patients with multiloculated hydrocephalus operated by endoscopic cyst fenestration, a repeat endoscopic procedure was necessary in 8 out of 24 patients (33%), among the mean follow-up period (30 months). At the second procedure persistent cysts unrecognized from the first operation were encountered in 5 patients (21%) and reclosure was found in the remaining 3 patients (12.5%). It was also noticed that patients in whom shunts were placed before endoscopic cyst fenestration had a 7.5 risk of having a repeat endoscopic procedure more than patients in whom endoscopic cyst fenestration preceded shunt placement (El-Ghandour, 2008).

While another contributing factor may exist which is the increased severity of multiloculated hydrocephalus in the former group of patients that might increase the incidence of repeat procedure, the difference in incidence between both groups is still highly significant. The incidence of 13% repeat rate detected in patients where endoscopic cyst fenestration was performed prior to shunting is comparable to the 16% incidence reported in a group treated by craniotomy (Nida & Haines, 1993).

The statistically significant difference between the increased incidence of a repeat endoscopic procedure in patients who were shunted before endoscopic cyst fenestration as compared to patients on whom endoscopic cyst fenestration preceded shunt placement ($p<0.001$, highly significant), makes it clear that early diagnosis and fenestration is critical in obtaining the best results in treating these patients. This better prognosis in the latter group can be explained by the thinner ventricular septations with easier fenestration, less vascular cyst wall and less likelihood for postoperative reclosure (El-Ghandour, 2008).

10. Complications

There is no much information about complications connected with the use of neuroendoscopic procedures in the treatment of complex hydrocephalus in the published clinical data. Major complications of cyst fenestration and septostomy include ventricular hemorrhage, ventriculitis, injury to adjacent neural tissue, and cerebrospinal fluid leakage. Hemorrhage and ventriculitis may lead to further loculations. The risk of injury to adjacent neural tissue can be minimized by careful planning of the surgical approach and by having a knowledge of the neural structures beyond the point of fenestration. Consequently, orientation is critical and the intraoperative use of ultrasound and stereotaxis in conjunction with endoscopy are so helpful (Abtin & Walker, 1998).

It has been mentioned that the surgeon's ability to control unexpected hemorrhage during endoscopic fenestration of ventricular cysts is suboptimal and could lead to catastrophic

complications (Nida & Haines, 1993). However, recent studies didn't report any significant bleeding during endoscopic treatment of complex hydrocephalus. In a study including 24 pediatric patients with multiloculated hydrocephalus operated by endoscopic cyst fenestration, minor intraoperative arterial bleeding was encountered in 2 patients (8%), but terminating the procedure was not necessary, and the bleeding stopped within few minutes with irrigation. None of the patients required craniotomy nor suffered a postoperative neurological deficit or seizures. There was no deaths (0%) and morbidity was minimal. It included cerebrospinal fluid leakage in 2 patients (8%) which stopped spontaneously within 3 days (El-Ghandour, 2008). Among 34 cases of complex hydrocephalus (21 uniloculated and 13 multiloculated) operated by endoscopic procedure, cerebrospinal fluid leakage occurred in 1 case (3%) and central nervous system infection occurred in 1 case (3%) (Lewis et al., 1995).

11. Outcome and prognosis

The outcome depends on the surgical procedure adopted, as well as the type of complex hydrocephalus. The outcome is evaluated by the incidence of improvement of hydrocephalus in postoperative MR imaging, avoiding or eliminating the need for shunting, simplifying complex shunt systems, and reducing shunt revision rate. Uniloculated hydrocephalus is easier to treat endoscopically than multiloculated hydrocephalus and consequently, it carries a better prognosis. It has been reported that 84% of patients undergoing endoscopic cyst fenestration for unilateral hydrocephalus have either remained shunt free or not required additional shunt placement (Abtin & Walker, 1998).

In a series of 6 patients with multiloculated hydrocephalus operated by transcallosal fenestration of intraventricular septations, a significant decrease in the rate of shunt revision per year was reported from a mean of 2.75 shunt revision per year (over an observation period of 44.5 months) to 0.25 per year (over a median follow-up period of 27 months) (Nida & Haines, 1993).

Successful communication between the isolated cavities can be confirmed postoperatively by contrast-enhanced CT ventriculography (Fig. 2). In a series of 24 pediatric patients with multiloculated hydrocephalus operated by endoscopic cyst fenestration, improvement of hydrocephalus was detected in postoperative imaging in 18 patients (75%), where there was an apparent decrease in the cyst size or ventricular size and restoration of normal ventricular architecture (Figs. 3-5). The need for shunt was avoided in 3 patients (12.5%) who were treated by endoscopic cyst fenestration and endoscopic third ventriculostomy (El-Ghandour, 2008).

In a group of 6 patients with malfunctioning pre-existing shunts included in the same study, endoscopy reduced the shunt revision rate from 2.9 per year before fenestration (among a mean observation period of 12.3 months) to 0.2 per year after fenestration (among a mean follow-up period of 27.3 months) ($P<0.01$, highly significant) (Table 1). Four of these 6 patients had complex shunt systems, and simplification was performed in 3 of them (75%).

However, all the 6 patients required a single repeated endoscopic procedure, endoscopic cyst fenestration was repeated in all of them (100%), and shunt revision was done in 3 patients (50%). Patients in whom shunts were placed before endoscopic cyst fenestration, more frequently needed a repeat endoscopic cyst fenestration (6 out of 6) than patients in

whom endoscopic cyst fenestration preceded shunt placement (2 out of 15) with a relative risk ratio 7.5:1 ($p<0.001$, highly significant). In these 2 patients, endoscopic cyst fenestration was repeated without shunt revision. In other words, none of the 15 patients (mean age 9.1 months) in which endoscopic cyst fenestration preceded shunting, required shunt revision among the mean follow-up period of 30.7 months.

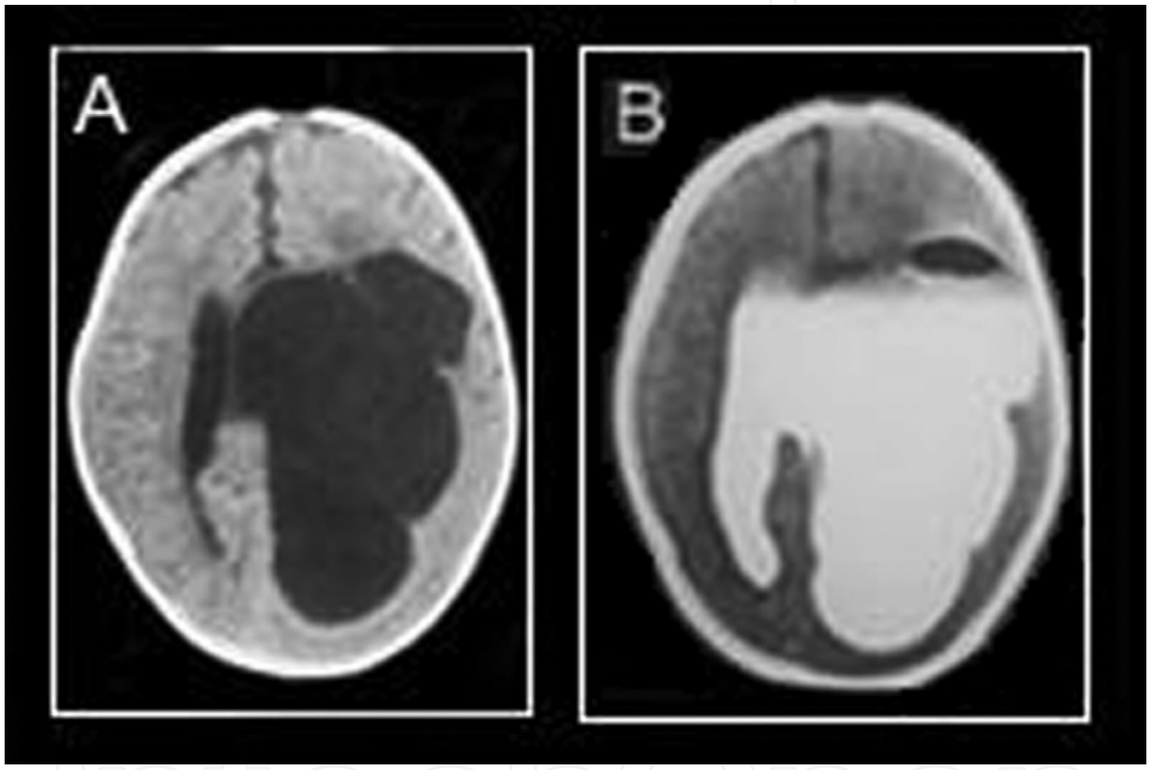

Fig. 2. A, Preoperative CT scan axial view of a 5 months old male patient with multiloculated hydrocephalus. B, postoperative CT ventriculography performed 1 week after surgery showing the contrast material evenly filling the entire ventricular system confirming communication among all cavities. The presence of air within the cyst is an indication of successful fenestration [Reprinted with permission from El-Ghandour NM (2008). Endoscopic cyst fenestration in the treatment of multiloculated hydrocephalus in children. *J Neurosurg Pediatr,* Vol.1, No. 3, (March 2008), pp:217-222, ISSN 1933-0707].

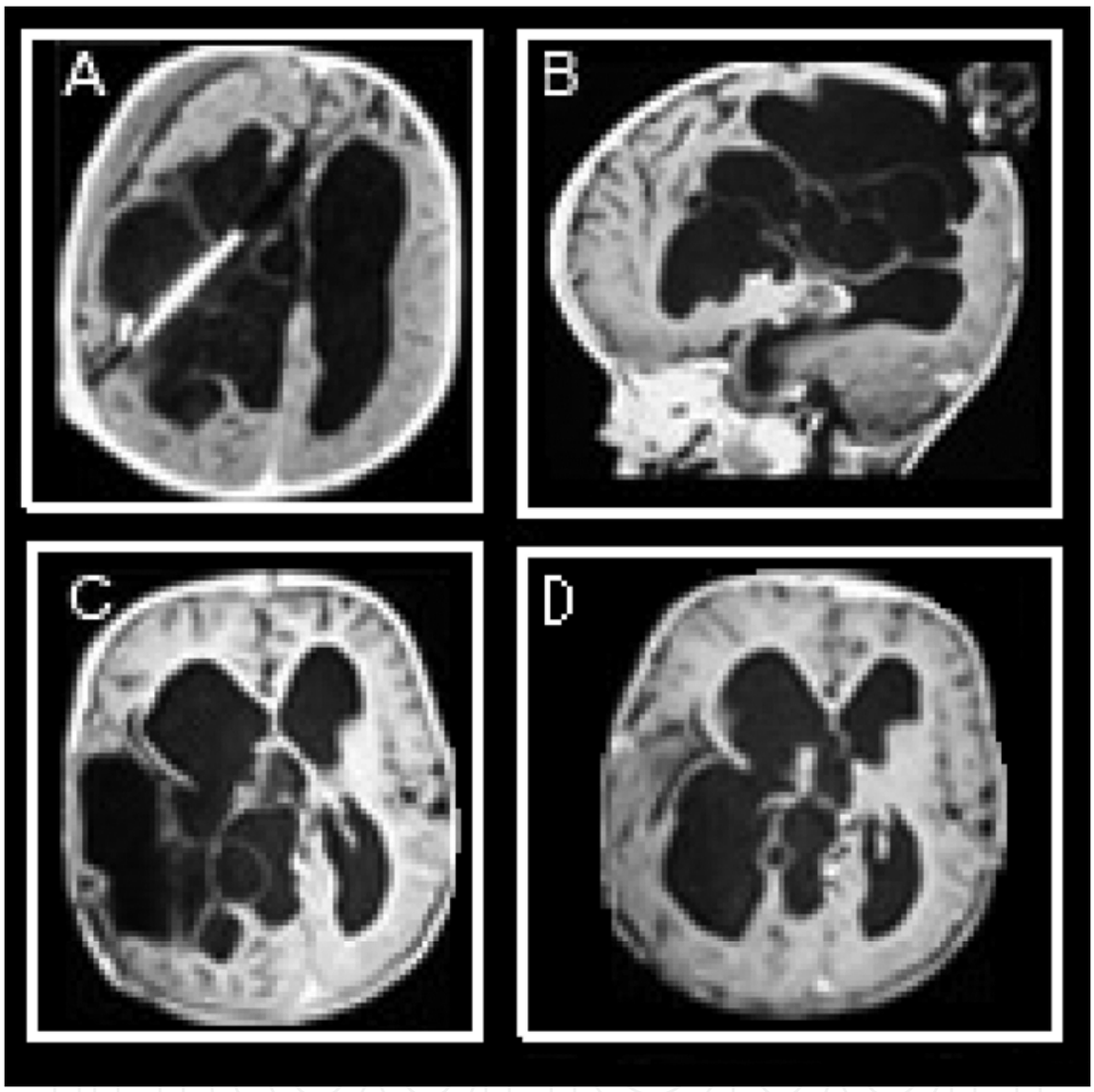

Fig. 3. A, preoperative CT scan of a 1 year old male patient with severe multiloculated hydrocephalus due to gliosis secondary to shunt infection. Note complex malfunctioning shunts. B, preoperative sagittal MR imaging of the same case. C, axial MR imaging performed 3 months after surgery. D, axial MR imaging performed 1 year after surgery showing improvement of hydrocephalus, increase in cerebral mantle, and restoration of ventricular architecture [Reprinted with permission from El-Ghandour NM (2008). Endoscopic cyst fenestration in the treatment of multiloculated hydrocephalus in children. *J Neurosurg Pediatr,* Vol.1, No. 3, (March 2008), pp:217-222, ISSN 1933-0707].

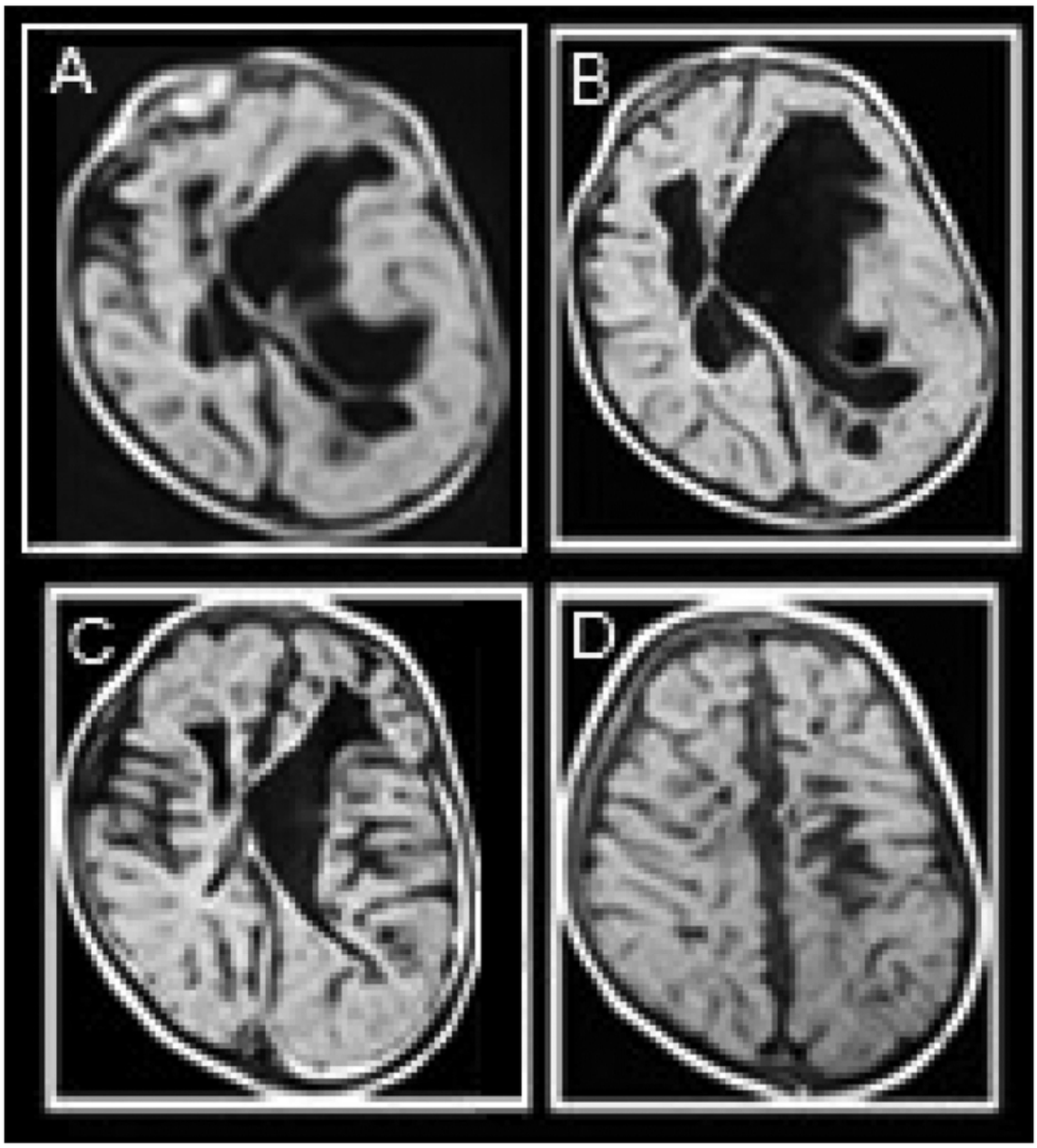

Fig. 4. A, B, preoperative axial MR imaging of a 7 months old male patient with multiloculated hydrocephalus due to intrauterine toxoplasmosis infection. C, axial MR imaging performed 3 months after surgery showing improvement of hydrocephalus. D, axial MR imaging performed 1 year after surgery showing complete resolution of hydrocephalus [Reprinted with permission from El-Ghandour NM (2008). Endoscopic cyst fenestration in the treatment of multiloculated hydrocephalus in children. *J Neurosurg Pediatr,* Vol.1, No. 3, (March 2008), pp:217-222, ISSN 1933-0707].

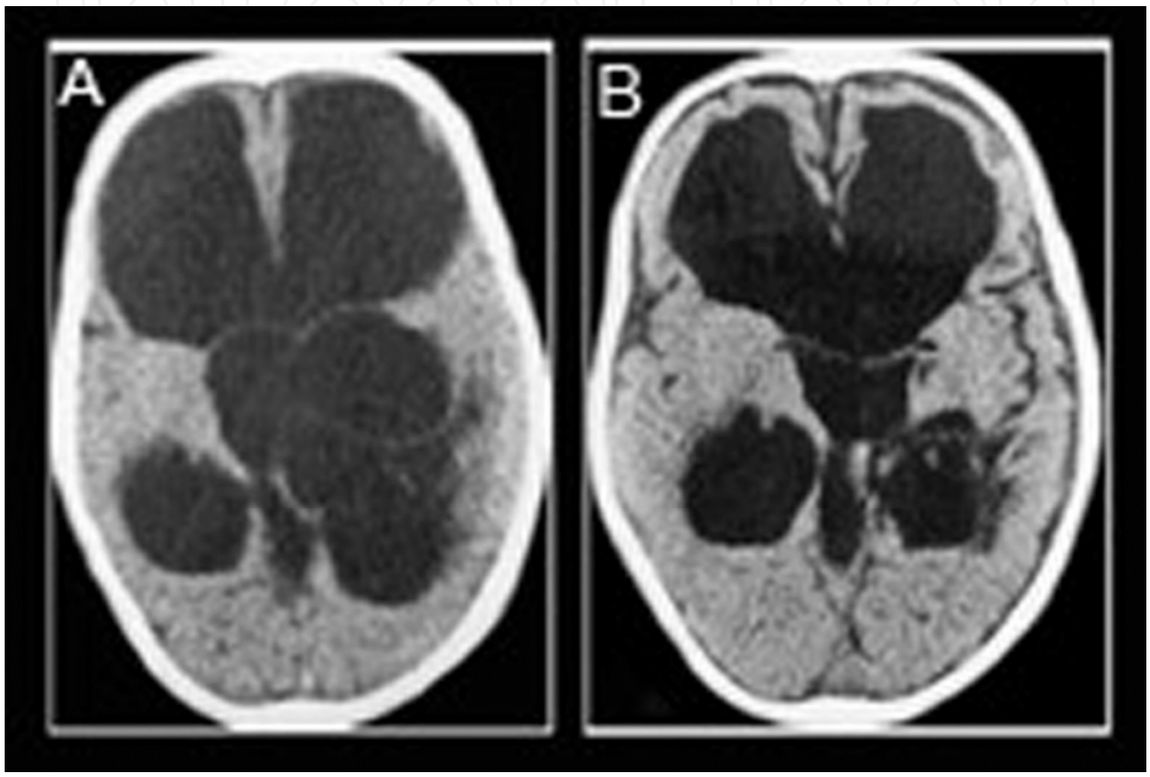

Fig. 5. A, preoperative CT scan axial view of a 1.5 years old female patient with multiloculated hydrocephalus. B, postoperative CT scan performed 3 months after surgery showing improvement of hydrocephalus, increase in cerebral mantle, opening of subarachnoid space, and restoration of ventricular architecture. This patient has been operated through a biportal technique (left coronal + left occipital burr holes) [Reprinted with permission from El-Ghandour NM (2008). Endoscopic cyst fenestration in the treatment of multiloculated hydrocephalus in children. *J Neurosurg Pediatr,* Vol.1, No. 3, (March 2008), pp:217-222, ISSN 1933-0707].

Case No.	Age at ECF (mos)	Observation Period (mos)		No of Revisions		No of Revisions / year	
		Pre ECF	Post ECF	Pre ECF	Post ECF	Pre ECF	Post ECF
1	12	9	36	4	0	5.33	0
2	24	18	21	4	1	2.66	0.57
3	16	9	9	1	0	1.33	0
4	36	15	42	3	1	2.40	0.28
5	8	5	26	1	0	2.40	0
6	36	18	30	5	1	3.33	0.39
Mean	22.0	12.3	27.3	3	0.5	2.91	0.21
S.D.	12.1	5.4	11.6	1.7	0.55	1.34	0.24
Median	20.0	12.0	28.0	3.5	0.5	2.54	0.14

Table 1. Age, observation period in months (mos), and shunt revision rates before and after endoscopic cyst fenestration (ECF), in 6 patients with multiloculated hydrocephalus presented with preexisting shunts. [Reprinted with permission from El-Ghandour NM (2008). Endoscopic cyst fenestration in the treatment of multiloculated hydrocephalus in children. *J Neurosurg Pediatr,* Vol.1, No. 3, (March 2008), pp:217-222, ISSN 1933-0707].

12. Conclusions

Complex hydrocephalus is a challenging problem in pediatric neurosurgery. Early diagnosis and treatment is the key for a better prognosis, therefore a high threshold of alertness is needed among pediatricians in dealing with patients having meningitis or intraventricular hemorrhage or premature infants. Multiplanar MR imaging is the preferred diagnostic modality. The definitive treatment is surgical, yet the approach remains controversial. Cyst fenestration is the main strategy of treatment, and it can be done either microsurgically or endoscopically, aiming at improving hydrocephalus, reducing number of shunts and shunt revision rate. However, the endoscopic treatment has the advantage of being minimally invasive. It is worthy to mention, that multiloculated hydrocephalus carries a worse prognosis than uniloculated hydrocephalus.

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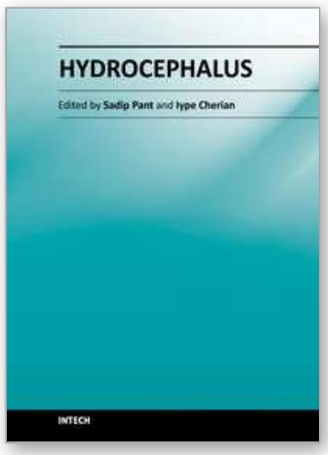

Hydrocephalus

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Description of hydrocephalus can be found in ancient medical literature from Egypt as old as 500 AD. Hydrocephalus is characterized by abnormal accumulation of cerebrospinal fluid (CSF) in the ventricles of the brain. This results in the rise of intracranial pressure inside the skull causing progressive increase in the size of the head, seizure, tunneling of vision, and mental disability. The clinical presentation of hydrocephalus varies with age of onset and chronicity of the underlying disease process. Acute dilatation of the ventricular system manifests with features of raised intracranial pressure while chronic dilatation has a more insidious onset presenting as Adams triad. Treatment is generally surgical by creating various types of cerebral shunts. Role of endoscopic has emerged lately in the management of hydrocephalus.

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