Diagnosis and management of shunt complications in the treatment of childhood hydrocephalus

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Introduction

The history of the treatment of hydrocephalus by using cerebrospinal fluid (CSF) shunt devices is characterized by two main features. The first of them is the great success rate as far as the percentages and, in most cases, the quality of survival are concerned, for a condition which in the past invariably led to the patient's death or severe clinical deterioration. The second characteristic is the number of the complications associated with this type of therapy, which has tempered the initial enthusiasm and, subsequently, the hope arisen by the introduction of any newly designed CSF shunt apparatus. Actually, the overall analysis of the results after treatment clearly indicates that the impact of CSF shunt device improvements on the failure rates seems to be very limited. To better identify the involved mechanisms further basic research into the pathophysiology of some specific shunt complications (e.g. the Slit Ventricle Syndrome) is clearly necessary. Unfortunately, mathematic models as well as animal models have proved to be not sufficient in such a regard. From a practical point of view, prevention and early identification and management of CSF shunt failures remain the main possibilities to assure the quality of the patient's long term outcome. In fact, fifty years of history teach us that the efficacy of new treatments should be scrutinized scientifically, by considering uncontrolled series and testimonial assertions with great skepticism. With these assumptions in mind we can agree with Drake's suggestion that shunt failure rates of about or little less than 5% per year and infection rates of less than 1% per year should be considered the reasonable goals for the next decade in this new millennium (1).

Mechanical Malfunction

In clinical series mechanical shunt complications account for more than half of all CSF shunt failures. Mechanical complications may result from either obstruction or disconnection of the different components of the implanted device.

<u>Obstruction</u>

Shunt occlusion, the most common shunt complication in the pediatric hydrocephalic population, accounts for approximately 50% of all shunt failures. Even though the obstruction may interest the distal end as well as the pumping device or the valve, itself ventricular catheter blockage is by far the most frequent events (1).

Symptoms are generally the same, that is those of intracranial hypertension, whatever the site of obstruction, though the clinical picture may be influenced at least to a certain extent by the etiology of the hydrocephalus and the patient's age. Children with mielomeningocele have the greatest variability of clinical manifestations, because of the number of associated Nervous System anomalies (i.e. Chiari malformation, tethered cord, syringomielia), that can become symptomatic in case of shunt malfunction (2). A bulging fontanel and sutures diastasis are the most frequent presenting signs in infancy and generally precede symptoms like irritability, a decreased level of consciousness, nausea and vomiting, commonly found in the growing child (3). In every case diagnosis should always regard both clinical conditions and imaging results; data from the pediatric shunt design trial indicated that there is a 9-29% of possibilities of underestimating a shunt malfunction only on clinical grounds (2); on the other way radiological findings alone may nevertheless be misleading; according to Iskandar et al. one third of patients with shunt malfunction may have "normal" imaging data (4).

1. Ventricular catheter obstruction

It may result from improper placement, choroidal and ependymal reaction, or secondary malposition due to ventricle coarction. Strict spatial relations between the catheter and the choroid plexus are known to increase shunt failure rates. In order to reduce this risk Tuli et al. suggested the frontal and the occipital horn for the proximal tip position (5). Nowadays endoscopic assisted placement of the ventricular catheter might assure a more proper result than in the past. However, even a proper surgical placement can't avoid the risk of the ventricular catheter being trapped by the choroidal plexus, due to the varying position of the ventricular catheter tip with the patient's body position and, in the course of time, the varying spatial relationships of the ventricle and the skull. It is worth to note that in most cases the analysis of CSF shunt failures on large series failed to statistically demonstrate the significant impact of the surgical ventricular catheter placement on the incidence of its obstruction. According to some authors (6) only catheters within the third ventricle are associated with a lower malfunction rate; in this anatomical space the choroid plexus is minimally developed and the fluctuations of CSF are more intense, thus providing a continuous washing action of the catheter.

Apart from the exposition of the catheter to the action of the choroid plexus, other common sites of improper placement are: within a cisterna, especially the interhemispheric one, or across the midline, within the opposite lateral cerebral ventricle. Even though, in some cases these locations do not exclude a partial drainage of the CSF by the shunt, nearly always they end in failure of the system, and for this reason a prophylactic surgical revision should be taken into consideration. A further cause of ventricular catheter obstruction is the penetration of brain debris during ventriculostomy; this inconvenience may be obviated by using the obturator as well as having the ventricular catheter filled with saline when entering the cerebral parenchyma (7). Finally this type of complication is influenced by the position of the valve along the CSF shunt device, being by far more common in case of proximal than distal valve systems.

a) Diagnosis

In the first year of life sonography plays a major diagnostic role. With this simple and noninvasive procedure it is possible not only to image ventricles size, but also to calculate cortical thickness and to indirectly establish cerebral blood flow parameters. In many instances it may clarify ventricular catheter position and its relations with the choroid plexus suggesting proximal tip obstruction (8). In the absence of an adequate sonographic window CT scan is still the most important tool. It is easily performed and all shunt complications can be ruled out (9,10). Position of the ventricular tip is always recognizable and its connections with the ventricular walls and brain parenchyma are outlined. MRI is necessary only in rare cases; it can be useful to visualize intraventricular septations and their relation with the proximal shunt.

Direct procedures on the implanted device (i.e. CSF withdrawal from a subcutaneous reservoir) are not indicated; they increase the risks of infection; moreover also an apparently normal CSF flow, can't exclude a partial obstruction of the ventricular catheter (11).

b) Treatment

Various procedures have been proposed for the prevention and treatment of ventricular catheter obstruction. An example are the number of catheters suggested since Portnoy first report (12), all designed to decrease contact and adhesions with the choroid plexus. Actually they have not significantly modified shunt failure rates (1,12,13). To improve ventricular catheter position perioperative sonography, neuronavigation and endoscopy have been used; data from the literature indicate that they can be safe and effective and help in reducing improper placement rates (14, 15,16,17). In spite of this, revision of the ventricular catheter is still the most common burden to afford in children with shunt malfunction. Monopolar coagulation through an intraluminal stylet is known to help the extraction of tubes adherent to the choroid plexus or ventricular walls (18). A recent modification of this technique is the employment of a flexible coagulating electrode; a moldable surface allows its use in cases in which the catheter does not follow a straight course (19). According to Hudgins if CSF flow is established during coagulation, the procedure should stop and the old catheter should be left in site; the aim is to decrease hemorrhagic complications (20). It is our opinion that in most cases only a transient success can be obtained this way; difficult cases should be treated with the help of endoscopy; coagulation under direct view of the choroid surrounding the proximal tip allows shunt removal in the majority of patients (15); a further suggestion can be to use intraluminal endoscopes; success rates up to 85% have been described (21).

2. Valve and distal obstruction

Distal catheter obstructions are common when using "slit valves" devices with an incidence of 3-28% in most surgical series. On the other hand shunt malfunction due to mechanical valves occlusion is relatively rare; in fact in several cases proximal differential-pressure valves continue to drain a sufficient amount of CSF, also in the presence of a partial filling (7). Flow - regulated (Orbis-Sigma) systems seem to be particularly prone to this type of complication; obstructions occur mainly in the early post-operative period, and are probably due to debris introduced at surgery (22). The role of high CSF protein concentration on valve and distal shunt obstruction has been discussed; in a recent prospective study Brydon (23) confirmed Mc Laurin (24) first report: an elevated spinal fluid protein content does not influence the functional integrity of the system. On the other hand CSF eosinophilia, a common finding in children with shunted hydrocephalus, is associated with an increased risk of distal obstruction.

Consequently systemic glucocorticoids have been suggested in these patients, in order to reduce CSF eosinophils and prolong shunt survival (25).
Differences exist between children with atrial and those with peritoneal shunts. Atrial systems have a higher risk of distal obstruction (26); factors related are the magnitude of blood flow and the size of the vein where the catheter is lodged; thrombosis within the distal tubing significantly increases when the cardiac end migrates from the atrium to the superior vena cava (x-ray level: T4); an undesired lodging within the subclavian vein is also associated with a raised risk of malfunction (7).

In children with peritoneal shunts, a mechanical bloc can result from a series of causes, such as retraction of the tube outside the peritoneal cavity, bowel adhesions or displacement into visceral structures and through the walls delimiting the abdominal space, kinking of the catheter, and the presence of foreign bodies in its lumen. The incidence of obstruction is inversely related to the amount of fluid drained. In the case of an obstructed peritoneal end by endoluminal material, a yellowish white material is seen inside the tubing on gross examination. The microscopic examination may reveal acute and chronic inflammatory phenomenon, which may occur even when the material cultured by routine methods does not reveal the presence of bacteria. Another cause of obstruction is the formation of a reactive film on the surface of the draining catheter without any endoluminal tissue. Kinking of the catheter may also occur, mainly because of technical failure in placement of the shunt inside the abdominal cavity (7).

a) Diagnosis

In most cases the diagnosis of a distal obstruction is possible only at surgery. An objective sign that may help preoperatively is the collection of subcutaneous fluid along the course of the system; if disconnections have been excluded it may indicate pericatheter CSF flow, a physiological attempt to overcome distal malfunction. The position of the distal tip of the shunting device should be controlled with x-rays in order to rule out kinkings or migrations of the catheter outside the atium or peritoneal cavity. X-rays are also helpful in the diagnosis of visceral perforations. In children with ventriculo-peritoneal shunts ultrasonography is an important tool; it can show the presence of pseudocysts or document ascitic collections (7, 8). CT is only rarely necessary to confirm these data. Suggestions of a distal malfunction may otherwise come from the CT study of the brain. CSF edema along the ventricular catheter or cysts inside the white substance are usually associated with peritoneal shunt obstruction, in patients harboring CSF shunting devices with distal "slit valves" (27,28). Some authors have also proposed radionuclide shuntograms: in some cases a pericatheter CSF flow can explain mild symptoms in children with distal CSF shunt occlusions and no evidence of ventricular dilation on neuroimaging studies (29,30).

b) Treatment

Overt or occult shunt infections should be regarded as the primary cause of early distal obstructions; in such cases the obstruction is associated with fibrous encapsulation of the tip of the shunt (31). CSF should therefore be sent for microbiological examination at surgery. Malpositions and displacements need almost to be correct in emergency. By the way atrial catheters at, or above T4 (superior vena cava), normally combined with a high risk of obstruction, should be repositioned in the right atrium, also in asymptomatic patients (32). In children with visceral perforation the catheter should be externalized, in order to rule out CSF infections before replacement. Change in position of the distal tip should be chosen in cases of pseudocysts or ascitic collections. While patients with abdominal pseudocysts may well tolerate a new abdominal lodging of the CSF shunt devices, patients with ascites should undergo an atrial or pleural shunt. Technical difficulties are almost exclusively met with cardiac tubes revisions. Unyielding adhesions with the vascular walls may be corrected by intravascular manipulation under radiological and electrocardiographic guidance; if this procedure doesn't succeed the catheter should be left temporarily in site to be possibly removed subsequently by thoracotomy (7).

Disconnection

With the advent of one -piece shunts the incidence of this type of complication has decreased. Actually it is mainly seen as a consequence of excessive strain on the pumping device; stretching at this level causes valve damage and may bring with time to the system fracture. Multipiece shunt can disconnect at any attachment site; because of the continuous traction forces junctions at the neck or the abdomen should always be avoided. The weakest points are the interface between silicone and metallic components and the ligatures, which favor the tearing of the plastic material.

a) Diagnosis

Even though disconnections may be accidentally noted in asymptomatic children during routine surveillance of the shunt, most patients exhibit signs of an increased intracranial pressure or evidence of local swelling due to subcutaneous fluid accumulation over the tube. In exceptional cases the complication may lead to acute ventricular dilation and death. Plain radiograms are used to outline the disconnection site. False positives may be due to radiolucent components; in these cases the knowledge of the implanted system may help diagnosis; intervals of less than 1 cm. at connection points should be regarded as normal.

b) Treatment

Reconnection of the CSF shunt device should be done with the attempt of reducing pericatheter strains. In cases of migration of the different components problems may arise in dealing with the "lost" catheters. Generally ventricular catheters are left in place as there is no harm described for catheters lying free within the ventricle. Lost peritoneal tubes are harmless too. In both conditions catheters removal is compulsory only in case of infection: endoscopy is the main surgical choice (1,15).

Misplacement of the atrial catheter in cardiac shunts is actually rare, due to the improvement in shunt design and surgical technique. The most serious complication related is its migration into the pulmonary artery. More commonly however the catheter remains within the right chambers; at this site it may cause extrasystolic disturbances and permanent cardiac arrhythmia; the risk of systemic infections is also enhanced. For these reasons a detached atrial catheter should always be removed, by thoracotomy or endovascular techniques (31).

Infection

Infection is the foremost type of CSF shunt complication after mechanical malfunctions. Even though the incidence of this complication may vary in the literature, most authors report infection rates between 5% and 18% (7). It is also known that infection represents the most expensive burden of CSF shunt implantation; an average of 8-45 thousands of dollars are needed for each treated patient (33). Seventy percent of infections are diagnosed in the first month after surgery; a further 15% occur between the first and the ninth month; altogether they represent the so-called early infections (34). In a recent review Baird et al. suggested differences in etiopathogenesis between early and late appearing cases. In the first group pathogens are more probably introduced with surgery, while in the second one they are more likely seeded from neighboring sites (34). To testify this the most common germs involved in early shunt infections are Staphylococcus Epidermidis (52.8-88.9%) (35,36) and Staphylococcus Aureus (12-40%) (35,37); these bacteria may enter the operating field during surgery, from hair follicles and sebaceous glands opened by the surgical incision; an insufficient asepsis, and a long lasting surgery contribute to this risk (38, 39). In late cases Baird couldn't find any Staphyloc. Aureus infection (34); according to his and other authors experience, germs like Propionibacterium Acnes (40) Enterococcus or Streptococcus Faecalis (9-11.4%) (41,42), which are normally rare agents in CSF infections, are quite usual in delayed contexts. These germs likely infiltrate through superficial wounds or ascend from the peritoneal catheter after contamination from the visceral content. Apart from an improper positioning adherences between the peritoneal catheter and the visceral walls may occur, leading with time to microscopic tearings of the bowels and causing local shunt infection. The subsequent ascending migration to the cerebral ventricles is assured by the germs ability of remaining attached to the catheter surface against the flow of rushing water (34,40,41,42). A number of risk factors have been suggested for CSF shunt infection. Many authors have indicated ages lower than 1 month at operation as significantly increasing infection rates (43,44,45,46); this is probably due to prolonged hospital stay as a result of underlying pathology, combined with the propensity for a high skin bacterial density with more adherent strains (47). A reduction of immunitary defenses may further contribute (44,48), especially in low birth weight and premature infants (35,49). However Davis et al. in a series of 2325 VP shunting procedures didn't confirm these data; infection rates were similar between infants and children. The type of surgical procedure (insertion or revision of the shunt) had a no specific role in this study in which the only elements that significantly increased infection rates were fluid accumulation along the shunt tract and an open CSF leak (50). Similar findings were reported by other authors (35,49). The weight of hydrocephalus etiology is discussed: Dallacasa et al. indicated previous intraventricular hemorrhages or CNS infections as main predictors for a shunt infection (44). It has also been reported that, when compared with other groups, children with mielomeningocele have a proportionally higher risk of infection (45,46). Other factors that have been studied in relation with shunt infection such as an increase in CSF proteins (51,52), the type of implanted system (39,53), previous abdominal surgical operations (54); without, however, finding an evidence of a direct causative role in increasing the infection risk.

a) Diagnosis

1. Symptoms and general laboratory examinations

Symptoms may be different in early and late cases. Early infections are usually revealed by obvious cutaneous manifestations and /or by prolonged postoperative fever, eventually associated with signs of meningitis, sepsis, or peritonitis (in ventriculo-peritoneal shunts). On the other hand the clinical onset of late-shunt bacterial contamination is often subtle. Williams et al. found symptoms of shunt malfunction (33%), wound changes (22%) and abdominal pain (19%) as main manifestations of long-term V-P shunt infections; fever can be absent or mild, yielding sometimes to a diagnostic delay (40).
The warning-signs of lately infected atrial shunts are also usually mild; a low-grade fever precedes for some weeks the appearance of symptoms like malaise, easy sweating, and chilling. A persistent low-grade fever may be accompanied by splenomegaly and anemia; white blood cell counts may be elevated, but also remain inferior to 10000/ mm3. The other infection parameters (i.e. VES, PCR) play a secondary role, as they can change post-operatively without an infection being present. In some instances laboratory examinations reveal hypocomplementemic glomerulonephritis, which on rare occasions can be also suspected on the basis of hypertension and peripheral edema. Even though in such cases, blood cultures in anaerobic media may demonstrate diphteroid or other "nonpathogenic" organisms, in several subjects the blood cultures result as negative (7).

2. CSF

Biochemical examination of the CSF can raise the suspect of CSF shunt infection. A reduced sugar level (below 20 mg./dl.) and an increase in CSF cells (>5 /mm.), are the most common parameters to be taken into consideration. Further information can be obtained from CSF white blood cell count (40).
Spinal fluid culture is more reliable. However in Ronan experience 17.1% of CSF samples were normal on microscopy, in spite of a verified contamination of the implants (55). Vanalocha confirmed this finding: CSF cultures were negative in 49/54 of his cases (90.7%) whereas cultures of the removed shunts were positive in 32/54 (59.2%); percutaneous CSF obtained from "reservoirs" on admission were particularly prone to show false negative cultures (51). Overall misleading CSF results can be due to the fact that coagulase-negative bacteria (Staphyloc Epid., Staphyloc. Aureus), the most common responsibles for shunt infection, produce an excess of mucoid substance which promotes adherence to the smooth surface of the systems and reduce germs circulant levels (7).

3. Other

Ecography of the abdomen may be helpful in children with V-P shunts. Apart from a specific inflammatory findings (i.e. splenomegaly) it may show enclosed fluid collections (pseudocysts). Infected pseudocysts are the result of the reaction of the enteral serosa to the presence of germs. They are seldom multiloculated and commonly have an iperproteic signal (56).