Rare complications of CSF shunts – a single center experience

BLIONAS ALEXANDER¹, MELISARIS SAVVAS¹, ROVLIAS ARISTEIDIS¹

Abstract

Valve mechanisms and CSF drainage in various compartments as well as into the peritoneal cavity (CSF shunting) is a modality for treating hydrocephalus in children and adults and were firstly introduced in 1905 by Kausch. They still remain as important means of tackling hydrocephalus, but they are accompanied by a large number and a variety of complications. It is important that health professionals are familiar with these complications in order to be ready to recognize and manage them promptly. In the present study, we present 10 unusual cases of hydrocephalus shunt complications managed in our neurosurgical department and we subsequently discuss treatment, prevention, etiology and pathophysiology. In conclusion, being aware of all the complications of hydrocephalus shunts is valuable for their diagnosis and treatment, but also increased care is required in order to prevent them.

Keywords: hydrocephalus, valve mechanisms, rare complications, case presentation

Introduction

The installation of cerebral shunts is nowadays one of the most frequent interventions in neurosurgical practice. The utilization of the peritoneal cavity to drain cerebrospinal fluid (CSF) in cases of hydrocephalus requiring a hydrocephalus valve mechanism in order to decompress the lateral ventricular system, was introduced in clinical practice by Kausch in 1905. After the introduction of CSF shunts accompanied by a slit valve mechanism (1967), which result into improved intraventricular pressure control, this method quickly became extremely popular worldwide. Due to the increasing number of surviving patients with hydrocephalus, and with the increase of life span, the amount of CSF shunt complications is increasing. It is very important for physicians to be aware of these rare complications of CSF shunts, and more specifically ventriculoperitoneal shunts which are the most common type of CSF shunting, in order to be prepared for managing them, but also constantly utilize measures of prevention. In this article, we present ten cases of adult patients affected with rare complications of CSF shunts, and the cases are throughoutly discussed along with their pathophysiological mechanisms.

Case presentations

1st case: A 40-year-old man, was admitted in the Intensive Care Unit and in our Neurosurgical Department due to severe head injury (diffuse axonal injury) after a traffic accident. He progressively developed clinical signs of communicating hydrocephalus, managed with the installation of a ventriculoperitoneal shunt. The patient remained in severe neurological status with post-traumatic encephalopathy, however one month later the patient became sluggish and lethargic, developed high grade fever, stiff neck, and abdominal muscle spasm and tenderness in the right hypochondrium. Laboratory testing revealed significant leukocytosis with elevated polymorphonuclears as well as an increase in ESR and CRP. The patient underwent Computed Tomography
(CT) of the upper - lower abdomen, which showed an extensive subdiaphragmatic hypodense collection containing the peritoneal catheter (Figure 1). The subdiaphragmatic abscess was surgically drained, the previous shunt was removed, and a new external ventricular drainage system was installed from a novel burr hole and onto a novel position. The purulent collection contained 2200 WBC/mm³ of a polymorphonuclear type, and cultures revealed staphylococcus aureus. The patient received aggressive intravenous antibiotic treatment with chloramphenicol and teicoplanin, combined with daily intraventricular injections of amikacin (10 mg daily) until culture and inflammatory marker test turned negative. After 10 days a novel luboperitoneal shunt system was installed (an alternative method of CSF drainage from the spinal space to the peritoneal cavity), draining into the right iliac fossa. Intravenous antibiotic treatment was continued for a total of six weeks, and there were no recurrences for the next 5 years of patient follow up.

2nd case: A woman aged 27 years, with a ventriculoperitoneal shunt operated elsewhere in the age of 17, due to communicating hydrocephalus caused by post viral encephalitis. The patient presented to the Surgical Emergency Department due to severe and progressively worsening abdominal distension. Clinical signs included generalized abdominal dullness, positive fluid wave sign, and a large palpable painless mass. There were no symptoms of intracranial hypertension or papilledema. An upper - lower abdomen CT revealed a sizable cystic formation occupying almost the entire peritoneal cavity causing abdominal organ shift. The peritoneal catheter tip was located within this cystic collection (Figure 2). It was a giant peritoneal pseudocyst which had been gradually accumulating CSF. We conducted a small middle supra-umbilical laparotomy, acknowledged the collection and drained about 2000 ml of clear fluid. The catheter tip had normal CSF flow which confirmed the normal functioning of the VP shunt system. There was no true cystic wall recognized, and granulation tissue adhesions with the adjacent intestine were carefully detached. Immediate Gram-staining was negative, and due to the absence of fever, it was decided to install a new peripheral catheter in the right atrium effectively converting the ventriculoperitoneal shunt into a ventriculo-atrial shunt (a CSF shunt system diverting CSF from the ventricular system into the right heart atrium). Final CSF cultures, pseudocyst fluid, peripheral catheter, and adhesion tissue all proved negative for microorganisms. The patient during her follow-up (approximately 15 years) had excellent neurological status without any recurrences.

3rd case: A 36-year-old with post-traumatic hydrocephalus (due to severe traumatic brain injury) two years ago, managed by us with the use of a ventriculoperitoneal shunt. The patient presented in our Neurosurgical department with painless rectal prolapse of the peritoneal catheter tip, and continuous CSF flow. Clinical examination and laboratory testing did not reveal any signs of intracranial hypertension or peritonitis. Orthosigmoidoscopy was performed (Figure 3), which revealed the distal catheter entry site into the sigmoid and the free catheter tip into the rectum. The general CSF exam showed 250 cells/mm³ with a polymorphonuclear cell type and increased protein. Cultures revealed Enterococcus faecium and the patient was administered intravenous penicillin and amikacin. The peritoneal catheter was cut and ligated subcutaneously in the subclavian region, and the rectal distal tip was removed endoscopically. During the following days, the patient showed no signs of intracranial hypertension, remained neurologically stable, and finally was declared non-shunt-dependent. The patient remains asymptomatic until today (more than 10 years of follow-up).

4th case: A 74-year-old male was transferred from a nursing home due to progressive gait instability / gait difficulty and urine loss. The patient was operated elsewhere 15 years ago installing a ventriculoperitoneal shunt system due to normal pressure hydrocephalus (NPH). Brain CT showed extended dilatation of the ventricular system and periventricular oedema, with the central catheter however in a proper position. Plain chest and abdominal x-rays revealed an unusual detachment and migration of the entire peripheral catheter lower into the peritoneal cavity. The upper - lower abdomen CT confirmed the presence of the entire peripheral catheter situated low in the free minor pelvis (Figure 4), thus justifying CSF shunt malfunction. The patient underwent revision surgery with the installation of a new peripheral catheter and removal of the free intraperitoneal segment, and immediately showed clinical improvement. Repeated brain CT showed decrease inventricular size and amelioration of periventricular oedema. Neurological status remained stable for 2 years of follow up until the patient succumbed to an unrelated malignancy.

5th case: A 41-year-old female arrived in the Emergency Department due to intense pleuritic chest pain of three days duration, aggravated by deep breathing. Patient history included the installation (in another department) of a cysto-atrial shunt (a CSF diversion valve system from an brain arachnoid cyst to the right heart atrium) at an age of 16 due to a large arachnoid cyst in the right middle fossa. The physical, neurological, cardiological examination, and laboratory tests were nor-
mal. Chest X-ray revealed a portion of the distal catheter into the left pulmonary artery and cranial radiography showed a disconnection of the distal catheter in the right occipital region (Figure 5). Migration of almost the entire distal catheter into the main shaft and the left main pulmonary artery in a curly manner was confirmed by chest CT (Figure 6). Brain CT did not reveal hydrocephalus or increased arachnoid cyst dimensions. The disconnected distal portion of the catheter was urgently removed by cardiac catheterization through the femoral artery, resulting into immediate symptom relief. Due to tolerable neurological status, the absence of intracranial hypertension, and normal CT findings, it was assumed that the patient was no longer “shunt-dependent” and it was decided not to revisit the CSF shunt system. The patient continues to be asymptomatic for 8 years of follow-up.

6th case: A 32-year-old man was admitted in the Neurosurgical department due to persistent headache, vomiting, fever, and lethargic episodes. At the age of two years he had been operated elsewhere undergoing a ventriculovenous shunt installation due to communicating hydrocephalus, and had no follow-up ever since. The clinical examination revealed moderate mental impairment, asunet eye sign, and subcutaneous inflammation in the cervical region along the peripheral catheter route. Laboratory testing revealed significant leukocytosis with elevated polymorphonuclear cells as well as increased ESR and CRP. Brain CT revealed calcified chronic bilateral subdural heterogeneous high-density collections occupying extensive intracranial space along with universal dilatation of the ventricular system and brain atrophy (armoured brain) (Figure 7). CSF cultures revealed a resistant strain of Enterobacter, sensitive only to colimicin. An external ventricular drainage system was inserted from a novel burr hole and colimicin was administered intravenously and intraventricularly at high doses. Four weeks later, repeated CSF cultures were sterile, inflammatory markers were normal, and therefore a new programmable valve was installed at a pressure setting of 100 mmH2O, resulting in the definitive remission of all initial patient’s symptoms, apart from mental impairment. There was no operation on the calcified chronic subdural hematomas, considering that the patient’s symptoms were solely due to inflammatory malfunction of the CSF shunt system. Clinical status remained good for 3 years of follow-up, with no further follow-up due to patient immigration.

7th case: A 75-year-old woman was operated in our department two years ago due to normal pressure hydrocephalus undergoing the installation of a ventriculoperitoneal shunt system with a programmable valve. Postoperatively the patient showed improvement of her previously ataxic gait along with improved bladder control. Two months later, the patient developed non-typical symptoms from the upper GI tract, including epigastric pain, indigestion, abdominal fullness and vomiting, especially postprandial. Brain CT confirmed reduced ventricular size and proper positioning of the intraventricular catheter. The patient was subjected to further testing with oesophagogastrscopy, which was also normal. Abdominal X-ray in an upright position revealed hepatomegaly, folding of the distal catheter, and wrapping of itself around the stomach and the duodenum (Figure 8). Considering that the patient’s abdominal symptoms were the result of peritoneal irritation from the folded catheter, we revised the distal part of the CSF shunt system by replacing and relocating the peritoneal catheter. The patient experienced immediate relief of epigastric pain and continues to be asymptomatic for 3 years of follow-up.

8th case: A 59-year-old male was transferred elsewhere 8 ago months due to obstructive hydrocephalus caused by a colloid cyst of the third ventricle (ventriculoperitoneal shunt with a programmable valve). The patient had a clinical presentation of acute intracranial hypertension and visual disturbances. Brain CT showed proper positioning of the intraventricular catheter, but great dilatation of the ventricular system and adjunctive periventricular oedema. Valve mechanism palpation revealed shunt malfunction, while X-ray examination revealed an intraperitoneal eight-shaped knot near the distal catheter end tip (Figure 9). The patient underwent emergent laparotomy and surgical exploration. The knot was located surrounded by omentum, while its free end tip did not have any CSF outflow. The previous catheter was removed (bacterial cultures of which proved sterile), and subsequently a new peripheral catheter was installed. The patient experienced immediate postoperative clinical improvement and was ultimately completely relieved of symptoms. Follow-up continues for over 5 years with no relapse.

9th case: A 44-year-old male was transferred because of progressive gait ataxia and urine incontinence. The patient had been operated 12 years ago undergoing the installation of a ventriculoperitoneal shunt in a different department due to post-traumatic hydrocephalus. Brain CT showed moderate dilatation of the ventricular system, and plain radiographs revealed an unusual disruption of the peripheral catheter in the lateral cervical region at the level of the 5th cervical vertebra, thus justifying the valve malfunction and deteriorating clinical picture (Figure 10). The distal catheter stump was located at the level of the 3rd rib, but without apparent lesions in
the continuity of the subcutaneous and intraperitoneal route. With the help of two straight plastic binders and a healthy part from a novel catheter, we managed to bridge the existing gap, restore CSF flow, therefore restored the continuity of the original catheter. The patient gradually improved without any further complications, for 3 years of follow up.

10th case: A 64-year-old male with a medical history of repeated CSF shunt revisions due to of obstructive hydrocephalus caused by cysticercosis, presented with fever and shivering, but no focal neurological signs or neck stiffness, of 5 days duration. Brain CT and x-ray examination did not show any abnormalities, but indicated significant inflammation, subcutaneous cellulitis and skin abscess on the lateral abdomen in the course of the subcutaneous distal catheter route just before the peritoneal entry point (Figure 11). The valve mechanism showed satisfactory function, and CSF sample cultures obtained by transdermal puncturing of the valve’s silicone reservoir proved sterile. Subsequently we proceeded in the incision of this subcutaneous collection and surgical drainage, extracting purulent material for cultures, and subsequently we commenced on daily basis, wound dressings, flushing and surgical debridement. To our great surprise, cultures revealed the rare strain of Leuconostoc spp. and the patient was administered high intravenous doses of Penicillin G and Clindamycin. There was a dramatic remission of the inflammation, along with improved clinical status, and wound healing was successful by secondary intention. The patient remained asymptomatic for 6 years of follow up until he suddenly succumbed to pulmonary oedema.

Case discussion

The use of the peritoneal cavity for CSF drainage has long been a scientifically proven technique for the treatment of hydrocephalus. Installation of ventriculoperitoneal shunt systems is perhaps the most commonly used technique for this purpose. As even more patients with hydrocephalus survive, the number of complications increase. Approximately 25% of these complications are intraperitoneal.

The literature review reveals a number of common complications such as catheter disconnection, obstruction of the peritoneal or ventricular catheter tip, infection, perforation and ileus, and a list or rarer complications—such as subdiaphragmatic abscess, perforation of the small intestine with the formation of CSF-enteric fistula, ascites, migration of the peritoneal catheter in the pleura, the subdiaphragmatic or intracranial space, protrusion of the peritoneal catheter tip in the rectum, and pseudocyst formation. We discuss in detail each presented rare complication, commenting on the pathophysiological mechanisms and possible prevention strategies.

1) The formation of an intraperitoneal abscess after ventriculoperitoneal shunt placement is a rare and severe complication, which according to the literature search usually involves children and less commonly adults. The majority refers to intrahepatic abscesses while the case which we are presenting is one of the few cases of subdiaphragmatic abscesses that has been reported. The subdiaphragmatic abscess is probably caused due to direct transfection of subdiaphragmatic space during implantation of the catheter into the peritoneal cavity leading to reverse expansion of the inflammation in the cerebral ventricular system. Previous patient admission in the ICU may have played a role in bacterial colonization and maybe facilitated foreign body transfection. The rapeutically, incision and drainage of the abscess collection, removal of the infected catheter, and a temporary external CSF drainage system along with antibiotic treatment are the sole way to address such complex case. In general, transfection appears to be related to pathogen entry either intraoperatively or post-operatively through the surgical wound, hence the use of robust antiseptic methods and frequent postoperative wound inspection is the only way to reduce infection probability.

2) Pseudocyst formation is also a rare complication with a reported incidence of 0.8 - 10%. First reported in 1954 by Harsh, it is also a complication most commonly occurring in children, rather than adults. The CSF pseudocyst presented as a thin-walled cyst around the tip of the peritoneal catheter, the wall of which consisted of fibrous connective tissue without epithelium. Usual predisposing factors of its formation include, transfection, repeated revisions, adhesions, increased CSF albumin, CSF malabsorption due to aseptic peritonitis, and local tissue response to material antigens (talc, latex, silicone) or catheter sterilization techniques. The interesting fact about the case described is the time interval between shunt placement and pseudocyst appearance which was 10 years, while the gigantic size and the increased probability of relapse led to our decision of converting the previous ventriculoperitoneal shunt into a ventriculoatrial shunt. It must be noted that there are multiple literature reports of laparoscopic revision techniques for restoring such complications, which however is not appropriate in the case of increased presence of adhesions or increased pseudocyst size, as in our current case. Management strategies are identical with intraperitoneal abscesses since the infection factor seems to play a major role, although in some cases there seems to be an aseptic inflammatory
reaction of unknown aetiology\(^{10}\), which may be associated with previous surgical operations and the development of adhesions.

3) Bowel perforation caused by the peritoneal catheter is also a rare complication of ventriculoperitoneal shunts, the incidence of which varies from 0.01 to 0.07%\(^{11,12}\). This complication was first reported in 1966, and most modern publications involve colon perforation (most often the sigmoid) in children\(^{13-16}\). The most common causes of intestinal perforation include the presence of a foreign body in the peritoneal cavity, the sharp edge of the intraperitoneal catheter, inflammatory fibrous tissue formation around the catheter tip which causes fixation and constant pressure on a part of the intestinal wall, and also poor patient condition resulting in reduced intestinal wall pressure resistance. After initial entry of the peritoneal catheter into the intestinal lumen, constant intestinal peristalsis entrains the catheter peripherally, towards the rectum. When diagnosis of intestinal puncture is established at an early stage, the prognosis is satisfying, while higher mortality applies to patients with peritonitis and bacteraemia, particularly when caused by gram negative strains\(^{13,17-19}\). It is important to be always aware of this type of complication in order to ensure early diagnosis. The use of more non-traumatic catheter material and accurate control of intraperitoneal catheter length are the main methods of prevention\(^{20,21}\).

4) Distal catheter migration into the peritoneal cavity is another rare complication of ventriculoperitoneal shunts, and often but not always requires the detachment and / or rupture of the distal portion, which may occur in a variety of anatomic areas\(^{22-30}\). One of these areas is theoretically the minor pelvis although there are very few references in the literature concerning this area\(^{31}\). The peripheral catheter migration is often associated with intraperitoneal catheter length, therefore precise control over catheter length and relative length restriction is important\(^{21}\). Furthermore, avoiding excessive mechanical loading and the use of more robust materials are both important to avoid such complications\(^{32,33}\). Usually the therapeutic practice in similar situations is the removal of the distal catheter and shunt revision, in cases of CSF shunt malfunction\(^{32,33}\), such as our current case.

5) Catheter migration into the pulmonary artery is a rare complication, and involves cases of ventriculocardiac shunts. In the past, ventriculocardiac shunts were widely utilized in the treatment of hydrocephalus, but were often accompanied with high dysfunction rates and complications such as inflammation, glomerulonephritis, and thromboembolic events. Existing literature references are few, and typically involve concomitant inflammation of the peripheral catheter, pulmonary embolism and / or thrombosis\(^{34,35}\). Distal catheter fracture followed by catheter migration into the pulmonary artery is probably a result of injury that may not be clearly evident inpatient history, or in some cases, material failure\(^{11,32,33,36-40}\). The previously mentioned prevention strategies also apply for these types of complications.

6) The natural history of calcified chronic subdural hematomas is not fully specified. The time interval from initial bleeding to calcium deposition may range from three months up to multiple years\(^{41,43}\). Patients undergoing ventriculoperitoneal shunting during childhood, require close follow up since they can develop chronic subdural collections, which if left untreated can become calcified and result in the notable “armored brain” syndrome. Most authors believe that surgical treatment of calcified chronic subdural hematomas offers no clinical improvement in the context of a chronic and irreversible established symptomatology but can instead lead to catastrophic rebleeding. Considering that our patient's symptoms were solely due to CSF shunt malfunction, we did not operate on the calcified hematomas despite their impressive size. There is a great amount of data linking CSF overdrainage (meaning the over function of the CSF shunt system which usually leads to a reduction of intraventricular pressure and ventricular wall collapse) to the formation of chronic subdural hematomas\(^{44}\), and therefore close monitoring with serial CT brain scans and early diagnosis of overdrainage syndrome along with realistic external valvemechanism regulation, as in modern programmable hydrocephalus valve systems, may help to avoid this unusual complication\(^{43}\).

7) Peritoneal catheter folding is an unusual entity and is probably the result of peristaltic visceral movements, which may even lead to digestive tract obstruction\(^{45}\). The usual treatment is through distal catheter revision. The main method to avoid this complication is to control peripheral and intraperitoneal catheter length, limiting it as much as possible in order to avoid catheter winding, which is referred throughout the literature as an established strategy for avoiding a wide range of CSF shunt complications\(^{20,21}\).

8) As previously discussed, distal catheter coiling may generate a widerange of complications. The creation of a knot is reported in the literature as a rare cause of peripheral shunt catheter occlusion, and management naturally involves peripheral catheter revision surgery (laparoscopy or laparotomy), since this complication may lead to mechanical obstruction, strangulation and / or ischemia of the intestine\(^{45-51}\). The exact mechanism of knot formation is not precisely known and many mechanisms have been implicated such as catheter characteristics, the capacity and configuration of the peritoneal cavity and catheter mobility. Increased infra-
peritoneal length of the distal catheter, decreased catheter diameter, increased catheter flexibility, and strong intestinal peristalsis appear to increase the likelihood of this complication.

9) Spontaneous rupture of the peripheral shunt catheter in multiple regions has been reported in the literature and can be associated with material aging, or increased mechanical loading. Peripheral catheter revision is practically the only method to restore shunt function. A method for reducing the likelihood of this complication is to avoid heavy mechanical loading, which can be achieved with stricter patient education regarding daily activities. Furthermore, rupture risk is primarily a matter of distal catheter material quality and mechanical strength, which could be improved through collaboration between clinicians, biologists and engineers, on the required level of material mechanical endurance and specifications.

10) Contamination of CSF shunt systems is a common complication, but usually involves Staphylococcus aureus or S. epidermidis. Physicians must always be aware of such complications in order to achieve early diagnosis and immediate treatment in order to avoid life-threatening CNS infections. CSF shunt infections with rare pathogens have been reported in the literature and always pose a significant clinical challenge. Surgical revision can be extremely difficult during disseminated CNS infection and the removal of the CSF shunt may be insufficient for definitive infection control. CSF shunt infection with Leuconostoc spp. has never been reported in the literature. This pathogen is found mainly in patients with lymphatic system malignancies, and in such cases, it may affect the CNS, but in our case there was no such history.

Conclusions

Although most complications of CSF shunts are not acute and life-threatening, they can however cause CSF shunt malfunction and raise diagnostic and therapeutic problems.

Familiarity with the complete range of CSF shunt complications is necessary for proper management of this increasing patient population. Modern imaging techniques are necessary utilities for proper diagnostic documentation. Knowledge of these situations by all neurosurgeons, and also by other physicians that might have to treat CSF shunt patients, and timely differential diagnosis can help to gain valuable time and reduce the risk of serious complications.

It is important to summarize preventive measures for these rare complications. It is important to maintain an adequate, but not exaggerated intraperitoneal catheter length, it is also important to focus on vigorous antiseptic protocols intraoperatively and postoperatively, instructing patients to avoid excessive physical activities which can increase catheter mechanical loading, to support the use and development of CSF shunt systems with increased mechanical endurance, as well as the use of softer and more atraumatic materials, and also selecting externally programmable valve systems, and careful patient follow up to avoid the phenomenon of overdrainage with proper regulation of the valve mechanisms CSF opening pressure. Finally it is important for physicians to always have clinical suspicion of CSF shunt infections in order to achieve early diagnosis and treatment.
References

References


Figure 1. Abdomen CT. Highlights a hypodense subdiaphragmatic collection with the peripheral catheter within.

Figure 2. Abdomen CT. The peripheral part appears to be inside a sizable peritoneal collection, which occupies almost the entire peritoneum. Giant peritoneal pseudocyst.

Figure 3. Orthosigmoidoscopy. Shows the entry position of the distal catheter into the sigmoid and rectal prolapse.

Figure 4. Upper and lower abdomen CT. Migration of the distal catheter into the lesser pelvis.
Figure 5. Simple skull X-ray. Shows distal catheter fracture in the right occipital region.

Figure 6. Chest CT. Highlights the migration of almost entire peripheral catheter within the main shaft and the left pulmonary artery in a curly manner.

Figure 7. Brain CT scan. Highlights bilateral chronic high density calcified subdural collections. Ventricular dilatation, cerebral atrophy. Armored Brain.

Figure 8. Abdominal plain x-ray (upright position). It shows hepatomegaly, folding of the peripheral catheter and winding around the stomach and duodenum.
Figure 9. Plain abdominal x-ray. Shows an intraperitoneal eight-shaped knot near the terminal end of the distal catheter.

Figure 10. Plain Neck X-ray. Loss of distal catheter continuity in the lateral cervical region at the level of A5.

Figure 11. Photo of the patients lateral abdomen. Cutaneous inflammation, cellulitis, cutaneous abscesses on the subcutaneous route of the distal catheter.