EANS/UEMS European examination in neurosurgery

Variants of questions with answers (compilation - Vyacheslav S. Botev, Department of Neurology and Neurosurgery, Ivano-Frankivsk National Medical University)

SHUNT MALFUNCTIONS

A ventricular shunt (VS) malfunction is a common neurosurgical emergency. In fact, a shunt revision is one of the most common procedures a neurosurgeon may perform. It is estimated that up to 50% of shunts may fail within 2 years. Despite its apparent simplicity, a shunt revision requires meticulous attention to detail and vigilance in diagnosis and management to ensure the patient is treated in a timely and adequate manner. The workup and surgical treatment of a VS malfunction is fraught with risks and complications even in the most experienced hands.

Common causes of shunt malfunction include mechanical failure (obstruction, disconnection, or migration), hardware failure (valve), infection, functional (underdrainage or overdrainage), or a combination of these aforementioned issues.

Infection is a common etiology of shunt failure and represents the second cause of shunt dysfunction after mechanical malfunction.

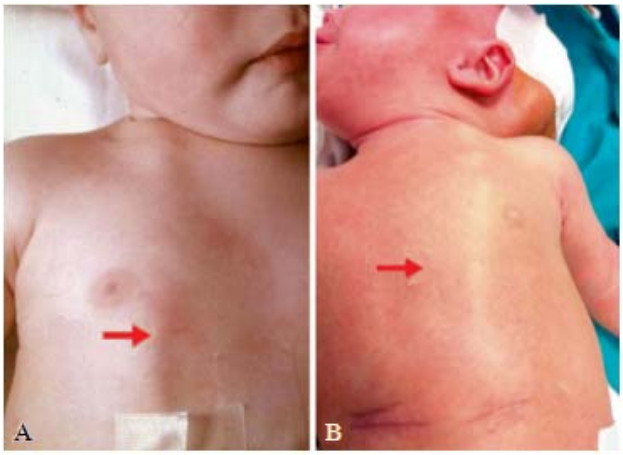
The reported incidence of shunt infection range from 1 to 40 % and average 8.5–15 %. The majority of shunt infections occur in the postoperative period (70 % within 1 month, 85 % within 9 months) and are thought to be mainly due to contamination during the surgery. Intraoperative contamination by skin flora is the primary mode of infection, while other sources of shunt infection are proximal seeding from meningitis and distal seeding from peritonitis and wound infections.

Over 95% of all shunt infections occur within 1 year of the last shunt instrumentation, with the majority of them occurring within 3 months.

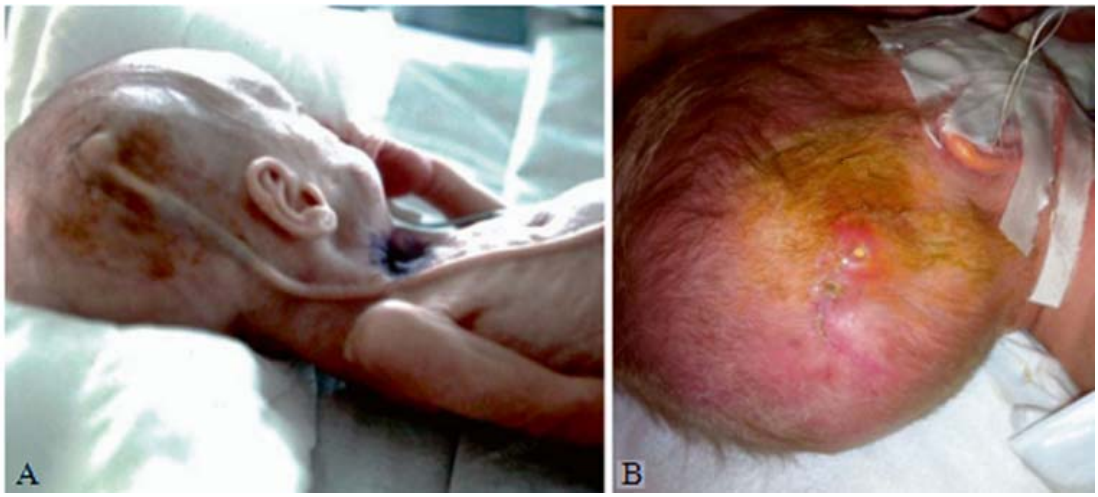
Late infections are less common. In fact, only 10–15 % of overall infectious complications occur 1 year after the shunt implantation and are attributed to contamination of the distal catheter by the visceral content or infiltration of germs through superficial skin wounds. Overall, the incidence of shunt-related infection is reported to be between 0.33 and 41 % with most recent reporting rates of approximately 7–12 % of patients and 3–8 % per procedure.

Additionally, the incidence of infection related to external ventricular drainage (ventriculostomy) is higher that the infection rate secondary to ventriculoperitoneal shunt. Although all shunt implant procedures are associated with a high risk of infection, the rate of infection does not appear to differ greatly between ventriculoatrial, ventriculopleural, and ventriculoperitoneal shunts.

Most infections in shunt systems are diagnosed within 6 months of placement (90 %) with the high incidence observed during the first 2 months. The majority of early infections originated from bacterial contamination introduced at the time of surgery, often from the skin flora. On the contrary, delayed infections can occur many years after shunt surgery and are likely caused by sending off of infection from remote sites such as the urinary tract or lungs.

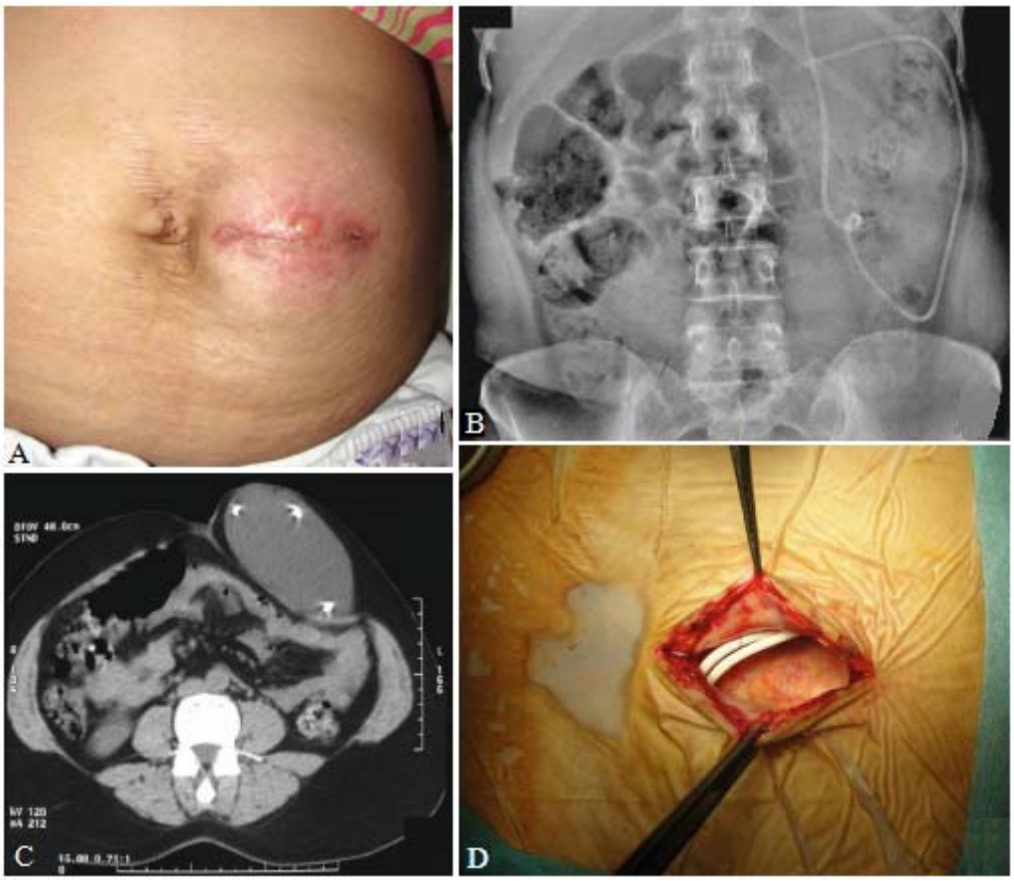


(A) Photograph illustrating skin reddening along the thoracic wall in a child with shunt infection (*arrow*), (B) photograph of the thoracic tract of a VP shunt showing subcutaneous accumulation of fluid (*arrow*).



(A) The image shows the extremely thin skin of a premature infant. The entire trajectory of the shunt can be easily viewed. (B) The photograph shows an exposed part of a valve reservoir.

Photographs of two patients: (A) one with ascites due to infection, (B) the other with abdominal distention due to chronic constipation.



(A). Erythematous paraumbilical abdominal wound with swelling. (B) Plain abdominal radiography without particular abnormalities. (C) Axial abdominal CT scan showing an extraperitoneal subcutaneous pseudocyst with the distal shunt catheter in the middle of a fluid collection. (D) Operative findings showing the subcutaneous enrolled distal catheter.

Questions

1. What is the most common cause of shunt failure?

2. What are the three places that a shunt may be occluded?

3. What is the pharmacologic approach in the case of suspected ventriculitis/meningitis associated with a neurosurgical procedure including an external ventricular drain (EVD), head trauma, or shunt infection?

4. What is a typical clinical presentation of an acute VS malfunction?

5. What are some organisms involved in shunt infections?

6. A patient with a ventriculoperitoneal shunt presents with an acute abdomen. Is this a reason to tap the shunt?

7. Why is it prudent from an infectious disease point of view to schedule shunt surgeries early in the day?

8. When is it appropriate to treat a shunt infection with antibiotics alone?

9. When can a shunt infection be considered cured?

10. What are the major risk factors for shunt infection?

11. Can perioperative CSF analysis help reduce the risk of shunt infection and/or malfunction?

12. What are the symptoms and signs of shunt infection?

13. When tapping a shunt, what is normal CSF pressure as measured from the ventricle?

14. How often does the patient have to pump the shunt?
15. When the shunt tap is indicated?

16. What is the Hydrocephalus Clinical Research Network (HCRN) protocol?

MCQs

1. Which of the following pathogens are most likely to cause early (less than 2 weeks) and late (greater than 6 months) shunt infections, respectively?

A. *Staphylococcus aureus, Staphylococcus epidermidis*
B. *Staphylococcus epidermidis, Streptococcus pneumoniae*
C. *Staphylococcus epidermidis, Staphylococcus epidermidis*

D. *Staphylococcus epidermidis, Propionibacterium acnes*
E. *Staphylococcus epidermidis, Staphylococcus aureus*

2. Which of the following is false with regard to shunt nephritis?

A. It is a well-described complication of VP shunts
B. It is due to deposition of immune complexes in the glomeruli of kidneys
C. The diagnosis is suspected with hematuria, elevated erythrocyte sedimentation rate (ESR), and decreased complement levels
D. Proper treatment entails removing the entire shunt
E. There is an elevated peripheral WBC count

3. With regard to shunt infections, which of the following is false?

A. *Staphylococcus* is the organism implicated in a majority of cases
B. Symptoms include shunt failure, headache, nausea, and vomiting
C. Elevated temperature is a more reliable sign of infection in VA as opposed to VP shunts
D. There is a greater risk of shunt infection with distal revision than proximal revision
E. *Staphylococcus* infections may cause obstruction without fever

4. What are the characteristics of shunt infection?

a. Risk of early infection is _____% — 7% overall
b. Risk of mortality is _____% — 10 to 15%
c. Risk of late is _____% within 6 months — 2.7 to 31% (typically 6%)
d. Organism is _____ _____ — *Staphylococcus epidermidis*

5. What are the characteristics of shunt nephritis?

a. v_____ s_____ — ventriculovascular shunt
b. c_____ l_____-l_____ i_____ — chronic low-level infection
c. i_____ c_____ d_____ in g_____ — immune complex deposit glomeruli
d. p_____ and h_____ — proteinuria and hematuria

6. Gram-negative bacillus (GNB) shunt infection compared with gram-positive bacillus (GPB)

a. morbidity — higher in GNB
b. Gram stain — more than 90% + Gram stain (in contrast to GPB only 50%)
c. protein — ↑ protein
d. glucose — ↓ glucose
e. neutrophils — ↑ neutrophils
f. The reason we must identify GNB infection is because
   i. treatment _____ _____ than for staph and — is different
   ii. there is a higher _____ for GNB — morbidity

7. What is the treatment for shunt infection?

a. Remove _____ — shunt
b. Insert _____ _____ _____ — external ventricular drain (EVD)
c. Administer antibiotics of _____ plus — vancomycin
   _____ — rifampin (change to nafcillin if possible)
d. for _____ days — 14—with CSF sterilization
e. Add i_____ a_____ — intrathecal antibiotics
f. by clamping _____ for _____ minutes — EVD for 30

8. True or False. With respect to a disconnected or nonfunctioning shunt:

a. A disconnected shunt may continue to function by CSF flow through a subcutaneous fibrous tract — true
b. If in doubt, better to watch, not shunt — false
c. Patients with a nonfunctioning shunt should not be followed with serial CT scans but possibly with serial neuropsychological evaluations — false

9. Concerning shunts and hydrocephalus, what type of shunts do you know? Hint: palmt

a. v_____ s_____ — ventriculo*p*eritoneal shunt
b. v_____ a_____ — ventriculo-jugular vein–right cardiac atrial
c. l_____ — lumboperitoneal
d. m_____ s_____ — miscellaneous shunts–ventriculopleural
e. T_____ s_____ — Torkildsen shunt (ventricle–cisterna magna)

10. What is shunt usage priority?
Hint: palmt

a. most often used _____ _____ ventriculoperitoneal shunt
b. abdominal abnormality _____ _____ ventriculoatrial shunt
surgery
peritonitis
morbid obesity

c. pseudotumor cerebri ___ ___ lumboperitoneal shunt–small ventricles
d. alternative _____ _____ miscellaneous shunts
e. acquired obstructive hydrocephalus_____ _____ Torkildsen shunt

11. Which are the miscellaneous shunts?
Hint: gupc

i. g_____ ventricle to *g*all bladder shunt
ii. u_____ ventricle to *u*reter or bladder shunt
iii. p_____ ventriculo*p*leural shunt
iv. c____ *c*yst shunt (arachnoid cyst or subdural hygroma cavity to peritoneum)

12. Name six possible shunt complications.
Hint: odesma

i. o_____ obstruction
ii. d_____ disconnection of shunt parts
iii. e_____ erosion through skin
iv. s_____ seizures–5.5% first year, 1.1% after 3 years
v. m_____ metastases of tumor cells
vi. a_____ allergy to silicone

13. What are ventriculoperitoneal shunt complications?
Hint: h2alo3mvps

i. h_____ hernia–inguinal 17%
ii. h_____ hydrocele
iii. a_____ CSF ascites
iv. l_____ lengthen catheter with growth (preventable)
v. o_____ obstruction by omentum or debris
by peritoneal cyst (infection
or talc from surgical gloves)
severe peritoneal adhesions
malposition of catheter tip

collapsed ventricular wall
choroid plexus

vi. o_____ obstruction or strangulation of intestine
vii. o_____ overshunting
viii. m_____ migration of tip to:
scrotum
perforation of stomach, bladder, diaphragm
ix. v_____ volvulus
x. p_____ peritonitis
xi. s_____ subdural hematoma

14. What are ventriculoatrial shunt complications?
Hint: liverssh

i. l_____ lengthening in children
ii. i_____ infection
iii. v_____ vascular: perforation, thrombophlebitis, pulmonary microemboli
iv. e_____ shunt embolus
v. r_____ retrograde blood flow
vi. s_____ superior vena cava obstruction
vii. s_____ subdural hematoma
viii. h_____ hypertension (pulmonary)

15. What are lumboperitoneal shunt complications?
Hint: Carols

i. C_____ Chiari I malformation (70% made worse)
ii. a_____ arachnoiditis and adhesions
iii. r_____ radiculopathy (from tube - hard to control)
iv. o_____ overshunting (6th and 7th cranial nerve dysfunction)
v. l_____ leakage of CSF
vi. s_____ scoliosis due to laminectomy (14% in children)

16. When do you tap the shunt?
a. to study CSF for
i. i_____ infection
ii. c_____ cytology
iii. b_____ blood
b. or to assess function

i. measure p_____ pressure
ii. instill c_____ contrast
c. inject m_____ medication

17. What are acute symptoms of undershunting?
Hint: salvadib h

a. s_____ seizures
b. a_____ ataxia
c. l_____ lethargy
d. v_____ vomiting
e. a_____ apnea
f. d_____ diplopia
g. i_____ irritability
h. b_____ bradycardia
i. h_____ headache

18. What are complications of overshunting?
Hint: s4i

a. s_____ slit ventricles 12%
b. s_____ subdural hematoma
c. s_____ sylvian aqueduct occlusion
d. s_____ skull change - craniosynostosis or microcephaly
e. i_____ intracranial hypotension

19. Complete the following concerning hydrocephalus and subdural hematomas (SDs):

a. Cause of SD in patients with shunts is _____ of the brain and _____ collapse; tearing of the bridging veins
b. Risk factors
i. b_____ a_____ brain atrophy
ii. l_____-s_____ h_____ long-standing hydrocephalus
iii. n_____ v_____ p_____ negative ventricular pressure

20. If subdural hematoma develops as a shunt complication the subdural is located on

a. the same side as the shunt _____% 32%
b. opposite side of the shunt _____% 21%

c. bilaterally _____% 47%

21. Treatment for subdural hematomas that occurs due to shunting for hydrocephalus could include
Hint: bcdht

a. b_____ burr holes
b. c_____ craniotomy
c. d_____ drainage – subdural peritoneal shunt
d. h_____ higher pressure shunt
e. t_____ tie off shunt

22. True or False. In VP shunt and laparoscopic surgery, abdominal insufflation can increase ICP. true

Answers

1. What is the most common cause of shunt failure?

Mechanical obstruction. About half of implanted CSF shunts fail within 2 years.

2. What are the three places that a shunt may be occluded?

The entry point (proximal occlusion), the valve system (valve obstruction), and the distal end (distal catheter occlusion). A CT scan of the head, a shunt series, and palpation of the valve are important in determining the site of the occlusion.

3. What is the pharmacologic approach in the case of suspected ventriculitis/meningitis associated with a neurosurgical procedure including an external ventricular drain (EVD), head trauma, or shunt infection?

EVD and/or shunt infections frequently involve gram-positive organisms (*S. epidermidis* and *S. aureus*, including methicillin-resistant *S. aureus*). Around 25% of these infections are secondary to gram-negative organisms, such as *E. coli, Klebsiella, Acinetobacter*, and *Pseudomonas species*. Vancomycin plus efepime or meropenem is recommended. Any infected hardware should be replaced, removed, or externalized. Intraventricular and intrathecal administration of antibiotics may be necessary.

4. What is a typical clinical presentation of an acute VS malfunction?

A typical clinical presentation of an acute VS malfunction includes drowsiness, severe headaches, and vomiting. However, the presentation may be quite diverse, from rapid to slow /subtle and chronic. The common signs and symptoms may be as modest and inconspicuous as deterioration in school performance, irritability, increase in head circumference over the 95th percentile, increased lethargy or sleep, clumsiness, chronic malaise, chronic fever, abdominal pain, or swelling around the shunt tract. More impressive presentations include seizure, cranial nerve paresis (III, IV, or VI), decrease in visual acuity, paralysis of upward gaze, papilledema, weakness or paralysis, stupor, coma, or change in vital signs (decreased pulse or increased mean arterial pressure).

5. What are some organisms involved in shunt infections?

Staphylococcus epidermidis and *aureus*, streptococci (*pyogenes, viridans, pneumoniae*), *propionibacterium acnes*, rarely gram-negative organisms (associated with higher morbidity and mortality). Ninety percent of possible infections will occur within the first 6 months of shunt insertion.

6. A patient with a ventriculoperitoneal shunt presents with an acute abdomen. Is this a reason to tap the shunt?

Yes. Rarely, patients with shunt infection may present with signs of an acute abdomen. These patients may be treated with catheter externalization and antibiotics. Not performing these maneuvers may expose the patient to an unnecessary abdominal procedure.

7. Why is it prudent from an infectious disease point of view to schedule shunt surgeries early in the day?

The number of airborne microorganisms increases over the course of the day in the operating room. This is the rationale for performing surgery early in the day, as is practiced at some large pediatric centers.

8. When is it appropriate to treat a shunt infection with antibiotics alone?

When the patient is high surgical risk and/or there is a shortage of alternative shunt sites. The standard of care is to remove the shunt, place an external ventricular drain, and start antibiotics once cultures of the CSF are drawn.

9. When can a shunt infection be considered cured?

When three negative cultures are obtained from the external ventricular drain. Once this is observed, the external ventricular drain can be removed and the shunt can be inserted at a different site.

10. What are the major risk factors for shunt infection?

The major risk factors for shunt infectionare are as follows:

High risks	Young age (especially children younger than 1 year)
	Premature birth
	Immunosuppressive states
	Etiology of hydrocephalus (myelomeningocele, intraventricular hemorrhage)
	CSF leak
	Previous or concomitant systemic infection
	Shunt revision
Low or controversial risks	Long hospital stays
	Prior exposure to antibiotics or steroid treatment
	Poor skin conditions
	Gender (male patient?)
	Disturbance of consciousness
	Former radiotherapy or chemotherapy
	Neurosurgeon experience
	Long duration of shunt surgery
	Use of a single glove
	Intraoperative hypothermia
	Increased CSF protein level
	Location of the ventricular catheter (frontal or occipital)
	Number of people present in the operating room

Some neurosurgeons suggest waiting until myelomeningocele patients are 2 weeks old before shunt insertion.

11. Can perioperative CSF analysis help reduce the risk of shunt infection and/or malfunction?

No

12. What are the symptoms and signs of shunt infection?

The symptoms and signs of shunt infection are as follows:

Symptoms	Signs
Headache	Fever
Nausea/vomiting	Fluid collection along shunt tract
Feeding problems	Erythema along shunt tract
Lethargy	Cellulitis
Irritability	Incisional wound infection/ frank pus discharge
Changes in sensorium	Nuchal rigidity/meningeal signs
Neck/back pain	
Abdominal pain	Abdominal distention/guarding
Diarrhea/constipation	Palpable intra-abdominal mass
Respiratory symptoms: chest pain, cough, dyspnea	Respiratory insufficiency signs/ hypoventilation/tachypnea

13. When tapping a shunt, what is normal CSF pressure as measured from the ventricle?
Less than 15 cm of CSF in relaxed recumbent position.

14. How often does the patient have to pump the shunt?
Patient must not touch the pump unless instructed to do so.

15. When the shunt tap is indicated?

Shunt tap is indicated if the fever is greater than 38°C or there is a positive blood culture in last 48 hours and/or shunt system intervention within 6–12 months, proceed with shunt tap prior to revision. Over 95% of all shunt infections occur within 1 year of the last shunt instrumentation, with the majority of them occurring within 3 months.

16. What is the Hydrocephalus Clinical Research Network (HCRN) protocol?

A standardized protocol to reduce cerebrospinal fluid shunt infection: the Hydrocephalus Clinical Research Network Quality Improvement Initiative.

Quality improvement techniques are being implemented in many areas of medicine. In an effort to reduce the ventriculoperitoneal shunt infection rate, a standardized protocol was developed and implemented at 4 centers of the Hydrocephalus Clinical Research Network (HCRN).

MCQs

1. C. *Staph. epidermidis, Staph. epidermidis*

Staphylococcus epidermidis is the most common pathogen to result in shunt infections in both an early and late fashion.

2. A. It is well-described complication of VP shunts.

Shunt nephritis a well-described complication of VA (ventriculoatrial) shunts. VA shunt complications are much more severe and potentially life threatening than VP shunt complications.

3. D. There is a greater risk of shunt infection with distal revision than proximal revision.

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