TREATMENT OF HYDROCEPHALUS (SHUNT PHYSIOLOGY AND PREVENTION OF INFECTION).

Hydrocephalous

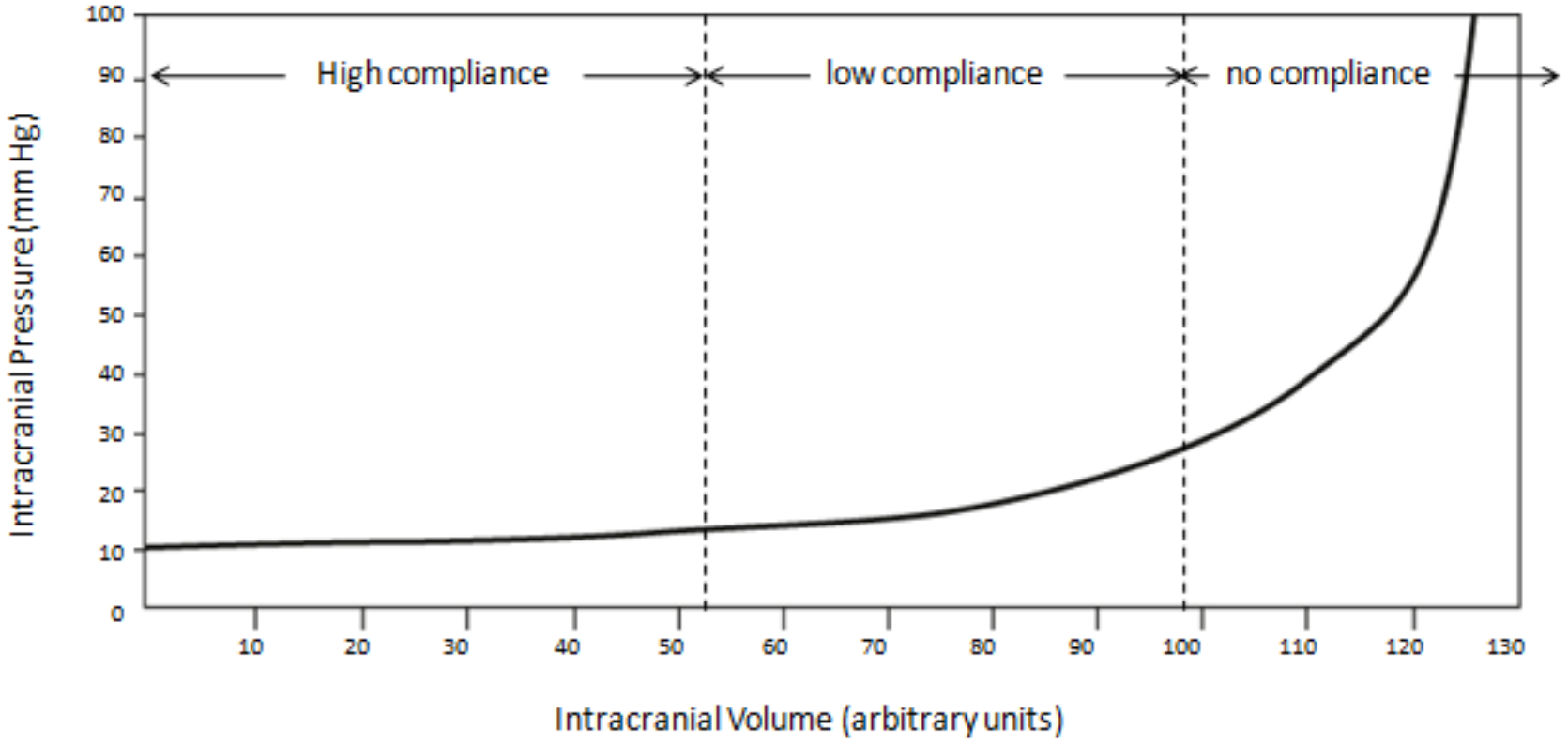
- An excessive accumulation of CSF within the head due to a disturbance of formation, flow or absorption
- If left untreated the patient may develop increased intra-cranial pressure (ICP)
- Result may be brain damage and/or death

Compliance and the Cranium

- The brain and skull contain three primary components:
  - Brain Tissue
  - Blood
  - Cerebrospinal fluid

- A change in any one of these components results in adjustment to the other two which is called **compliance**

Compliance Dynamics



$$\text{Compliance} = \frac{\Delta P}{\Delta V}$$

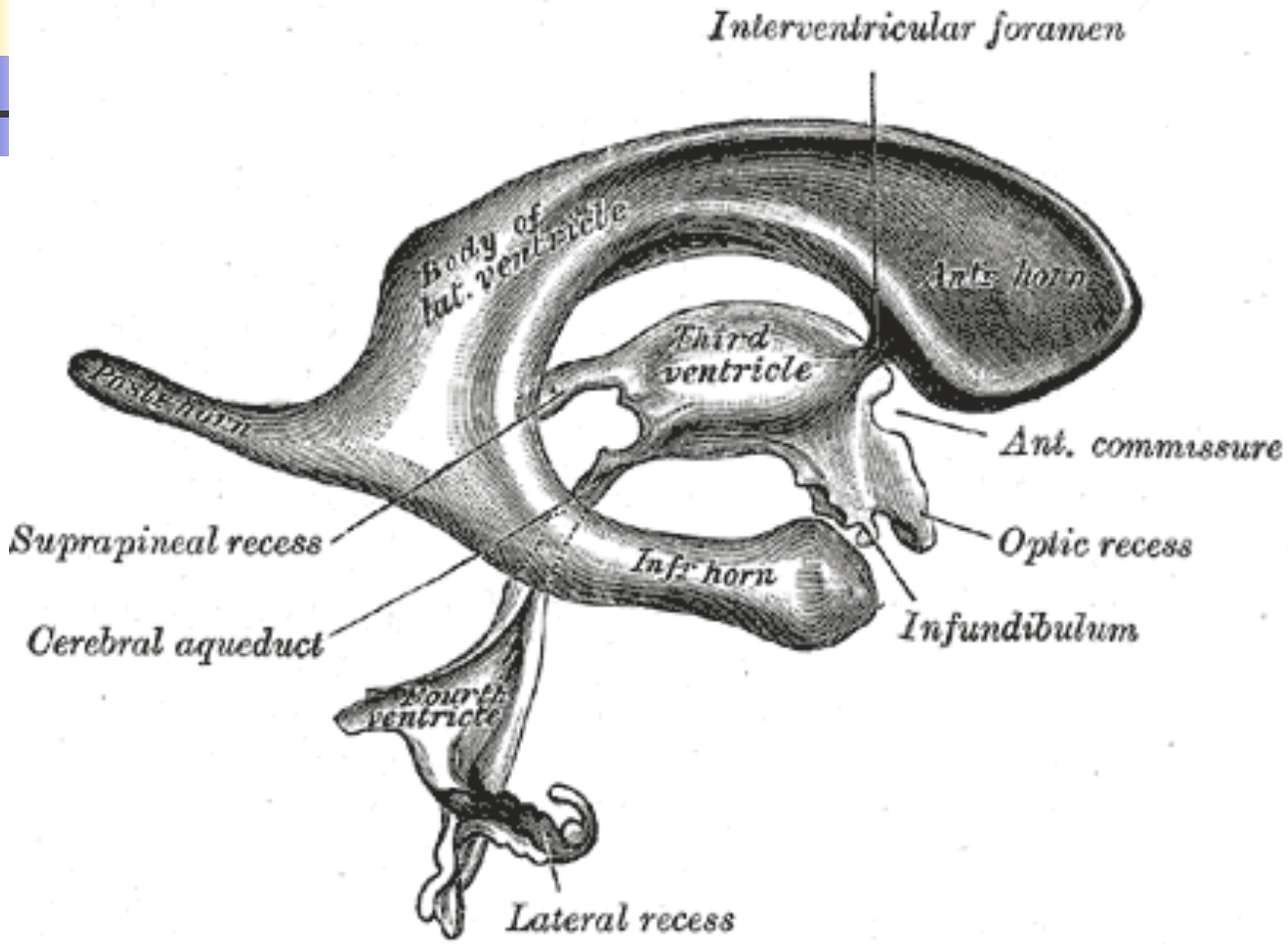
Cerebral Spinal Fluid (CSF)

- CSF is produced mainly in the ventricular system by the choroid plexus and is present throughout the central nervous system

- CSF has many functions:
  - Remove waste
  - Carry nutrition
  - Regulation of brain function
  - Neurotransmitter, paracrine and endocrine effects
  - Cushions the brain

CSF Production & Absorption

- CSF is produced at a rate of 20ml per hour and is normally absorbed at the same rate
- The brain and spinal cord system contain approximately 150ml of CSF (25ml in the ventricles)
- The entire CSF volume is turned over approximately 3 times per day
- CSF is transferred from the subarachnoid space into the superior sagittal sinus (venous system) through small granulations called the arachnoid villa



Gray anatomy 20th ed

2 Forms of Hydrocephalus

- *Communicating*
  - Full communication of CSF between ventricles and the subarachnoid space
- *Non Communicating*
  - CSF cannot flow out of the ventricles due to blockage or malformation

Compliance and Hydrocephalus

- As discussed, the skull is a fixed vault with limited volume to hold brain tissue, blood, and CSF
- If too much CSF exists, the blood and brain tissue are compressed or squeezed out resulting in a possible neurological deficit

Causes of Hydrocephalus

- Acquired hydrocephalus (secondary)
  - Tumors and cysts
  - Inflammation (meningitis)
  - Absorption blockages
    - Sub Arachnoid Hemorrhage (SAH)
    - Normal Pressure Hydrocephalus (NPH)
    - Head injury
    - Aqueductal Stenosis
- Idiopathic hydrocephalus (primary)
  - iNPH

Causes of Hydrocephalus

Foramen of Monro Gliosis Colloid cysts	**Absorptive obstruction** arachnoiditis (posthemorrhagic) (postmeningitic) venous thrombosis
Third ventricle chismal ghiomas craniopharyngiomas arachnoid cysts	**Pineal region** Tumors
Fourth ventricle medullobIastomas ependymomas astrocytomas Dandy-Walker cysts	**Cerebral aqueduct** aqueductal stenosis Aqueductal forking Subependymal gliosis periaqueductal gliomas
Basilar obstruction arachnoiditis Chiari malformations	

Symptoms of Hydrocephalus

- Usually associated with high intracranial pressure (ICP)
  - Headaches, nausea, vomiting, sleepiness, irritability, seizures, downward deviation of the eyes, blurred vision, failing mental function, other problems
- In infants - expanded head or bulging fontanelles, “sunset sign”.

Pediatric Hydrocephalus

- Pediatric Etiologies
  - Intraventricular hemorrhage (IVH) occurs often in premature babies
  - Congenital hydrocephalus
  - Tumor

Adult Hydrocephalus

- Adult Etiologies
  - Normal pressure hydrocephalus (NPH)
  - Sub arachnoid hemorrhage (SAH)
  - Post-trauma, aneurysm
  - Pseudotumor cerebri
  - Adult onset of congenital hydrocephalus

CT/MRI CRITERIA OF HCP

- Hydrostatic hydrocephalus suggested when-

1. The size of both temporal horns is >2mm in width ; sylvian, interhemispheric fissures and cerebral sulci are not visible. Or
2. Both temporal horn are >2mm and the ratio FH/ID>0.5

CT/MRI criteria contd..

- Ballooning of frontal horns of lateral ventricles (Mickey mouse ventricles) and third ventricle.
- Periventricular low density on CT or periventricular high intensity signal on T2w1 on MRI suggesting transependymal absorption or migration of CSF

CT/MRI criteria contd..

- Used alone- FH/ID

  <40% - Normal

  40-50%- borderline

  >50% - HCP

- Evans Ratio- Ratio of frontal horn to maximal biparietal diameter>30%.
- Sagittal MRI may show upward bowing of corpus callosum.

Treating Hydrocephalus

- Medical Management.
- Spinal Tap.
- Surgical Management

Medical treatment

- Diuretic therapy- Tried in infants with bloody CSF to see if there is any resumption of normal CSF absorption.
- Acetazolamide and furosemide started simultaenously.
- To counteract acidosis start alkasol(2meq of K +/ml,no Na +)
- S/E- electrolyte imbalance, lethargy, tachypnea, diarrhoea, paraesthesia.

Spinal Taps

- HCP after intraventricular hemorrhage may be transient serial taps may temporize until reabsorption resumes but LP can only be performed for communicating HCP.
- If reabsorption does not resumes when protein is <100mg/dl then it is unlikely to start as before.

Surgical treatment

- Goal- “Optimum neurologic function and good cosmetic result” not “normal sized ventricles.
- Options-

1. Eliminating the cause of obstruction.
2. Endoscopic methods.
3. Shunting.

Endoscopic - 3rd Ventriculostomy

- Indications-

1. Obstructive HCP.
2. Shunt infection(removal of hardware).
3. Patients with subdural hematomas (shunt removed before TV is performed).
4. Slit ventricle syndrome.

- Contraindication- Communicating HCP.

Endoscopic - 3rd Ventriculostomy contd.

- Complications-

1. Hypothalamic injury.
2. Transient 3rd and 6th nerve palsies.
3. Uncontrollable bleeding.
4. Cardiac arrest.
5. Traumatic basilar artery aneurysm.

3rd Ventriculostomy contd.

- Success rate- overall=56% (range is 60 to 94% for nontumoral aqueductal stenosis). Success rate is lower in infants as they may have under developed sub arachnoid space.
- Lower success rate – if preexisting pathology present like- tumor, previous shunt, previous SAH, WBRT, adhesions.

Endoscopic choroid plexus coagulation

- First done by Dandy(open)
- Indications-

1. Communicating slowly progressing HCP in infants- 64% cured.
2. Choroid plexus papilloma/hyperplasia.
3. Necrotizing enterocolitis.
4. Intractable shunt failure.

- Contraindication- Obstructive HCP.

Endoscopic fenestration

- Septostomy – for U/L HCP
- Multiloculated HCP.
- Aqueductoplasty or aqueductal stenting.
- Cysts with secondary HCP- Arachnoid cyst, Cysticercal cysts (3/4 ventricle)
- Colloid cyst of third ventricle.
- Pineal region tumors- ETV + Biopsy

Types of shunts

- VP shunt
- VA shunt
- Torkildsen shunt- ventricles to cisternal space.
- Miscellaneous– Ventriculopleural, gall-bladder, ureter or bladder.
- LP shunt
- Cyst or subdural shunt

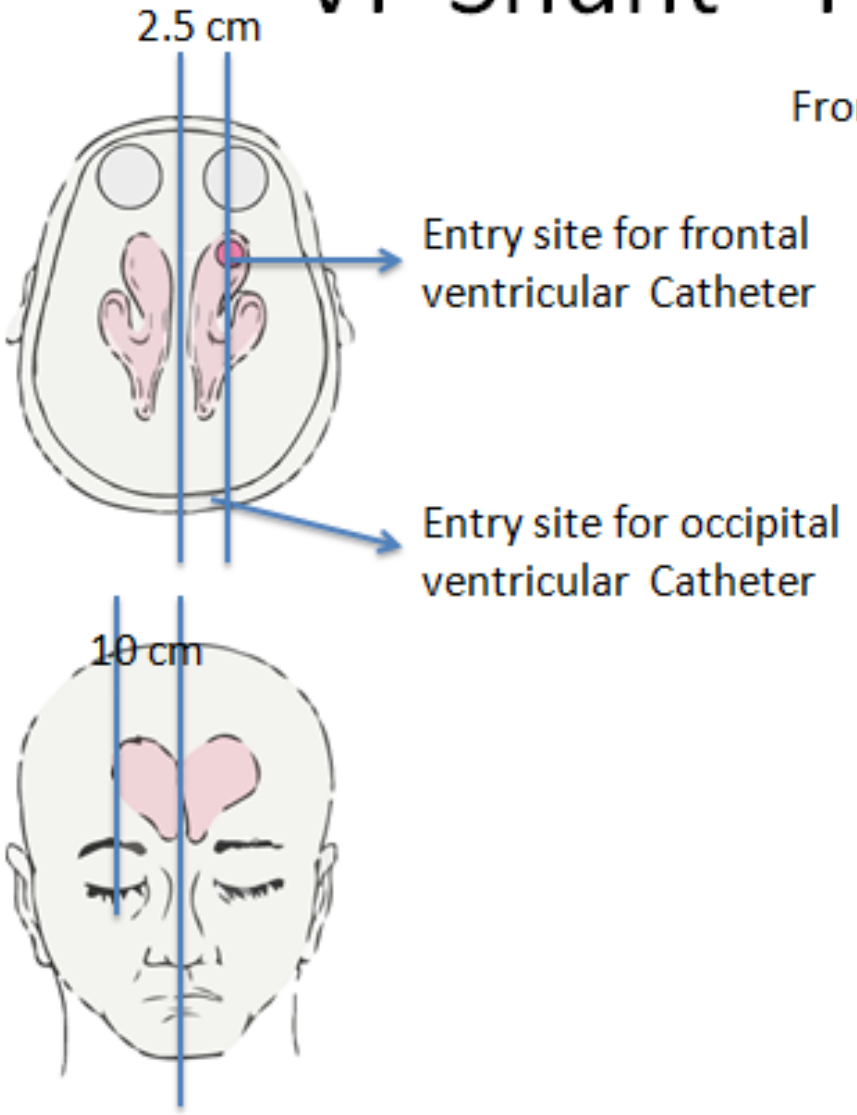
Shunting

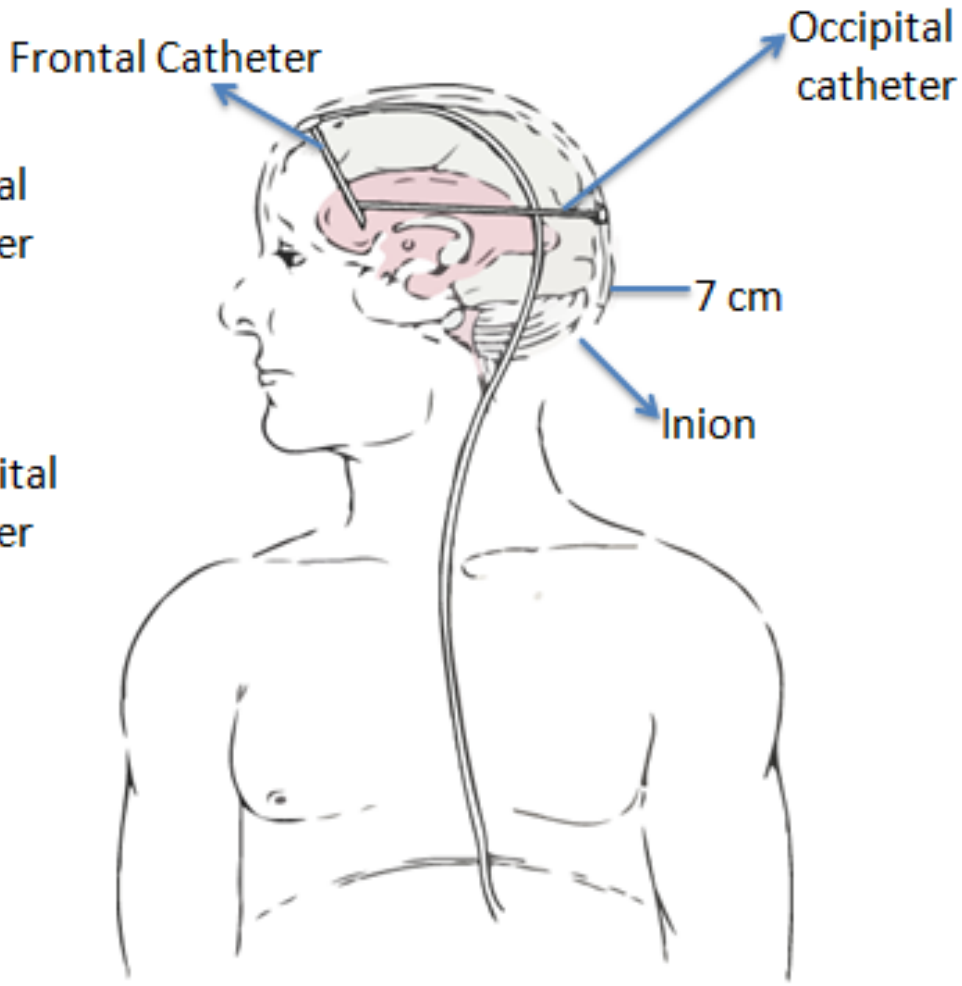
- Surgical Goal
  - Re-direct CSF to another area of the body to normalize ICP
- Shunt Considerations
  - Choose the correct operating pressure (fixed pressure valve)
  - Avoid catheter obstruction
  - Avoid shunt infection
  - Avoid other issues
    - Blood, high protein in CSF
    - Catheter disconnection

Surgical Procedure

- The surgical procedure to place a shunt is relatively short and uncomplicated:
  - Incision in the scalp
  - Small burr hole on the skull (6-9mm)
  - Insertion of the ventricular catheter
  - Incision in the peritoneal cavity
  - Tunneling under the skin
  - Closure

VP Shunt - Positioning





VA Shunt

- **Repeated lengthening required in a child.**
- **Higher risk of infection and septicemia.**
- **Possible retrograde blood flow into valves.**
- **Shunt embolus.**
- **Vascular complications- thrombophlebitis, pulmonary emboli, PHT**

LP Shunt

- **Do not use in child- laminectomy causes scoliosis, risk of progressive scoliosis**
- **Overshunting. May cause 3rd and 6th CN palsies.**
- **Difficult access to proximal catheter for revision.**
- **Lumbar root irritation.**
- **Leakage of CSF around catheter**
- **Pressure regulation is difficult.**
- **Arachnoiditis.**

Torkildsen shunt

- Shunts ventricles to cisternal space.
- Rarely used.
- Effective only in acquired obstructive HCP as children with cong HCP frequently do not develop normal sub arachnoid CSF pathways.

Ventriculopleural shunt

- Viable alternative if peritoneum is not available.
- Risk of hydrothorax requiring relocation of distal catheter.
- Recommended only for >7 yrs of age.

Disadvantages/complications of various shunts

- Those that may occur with any shunt-

1. Obstruction –M.C. , proximal catheter>valve/distal catheter(12-34%)
2. Disconnection
3. Infection
4. Hardware erosion through skin.
5. Seizures-5.5% in I yr, 1.1 %/yr after 3 Yr. (Higher in frontal catheter.)
6. Conduit for extraneural mets.
7. Silicone allergy.

VP shunt complications

- Inguinal hernia– if inserted when processus vaginalis is patent.
- Requires long catheter to compensate for child growth.
- Peritoneal end obstruction-more with distal slit valves, by peritoneal pseudocyst, Peritoneal adhesions may decrease absorptive surface, catheter malpositioning.

VP shunt complications contd..

- Peritonitis
- Hydrocele
- CSF ascites
- Tip migration –Into scrotum, viscus perforation, through diaphragm.
- Intestinal obstruction.
- Volvulus
- Intestinal strangulation- shunt removed forcibly.
- Overshunting.

Shunt Complications - INFECTIONS

- Incidence -1.5 to 38%(International society of pediatric neurosurgeons cooperative study 1994- 6.5%)
- Mortality, morbidity, costs.
- Time to infection- 92% infections occur within three months.

Shunt Complications – INFECTIONS contd..

- Risk factors-

1. Age – most important. (Different skin flora, less immunity and IgG).
2. Reason for shunt placement.
3. Type of shunt.
4. Educational level of surgeon.
5. Presence of spinal dysraphism.(50% children with MMC who were shunted within 1 wk developed shunt infection.)

Shunt Infections contd..

- Presentation-

1. Headache,lethargy,nausea and vomiting.
2. Infants- irritability, severe- apnea and bradycardia.
3. Fever, gait disturbance, seizures, visual disturbances, upgaze palsy, papilledema, abdominal pain-swelling.
4. E.coli-severe abdominal pain & septicemia, Staph- indolent , erythema along tract.

Shunt Infections - Evaluation and diagnosis

- History and examination (D/D specially in children- URI, gastroenteritis, UTI, Appendicitis)
- Imaging-

1. X ray(shows disconnection of the system.)
2. USG – Cranium (ependymal enhancement), abdomen.
3. CT scan
4. Shunt tap- CSF and manometry

Shunt tap

1. Indication-

- Diagnosing infection/cytology/remove blood/check function/inject medication.

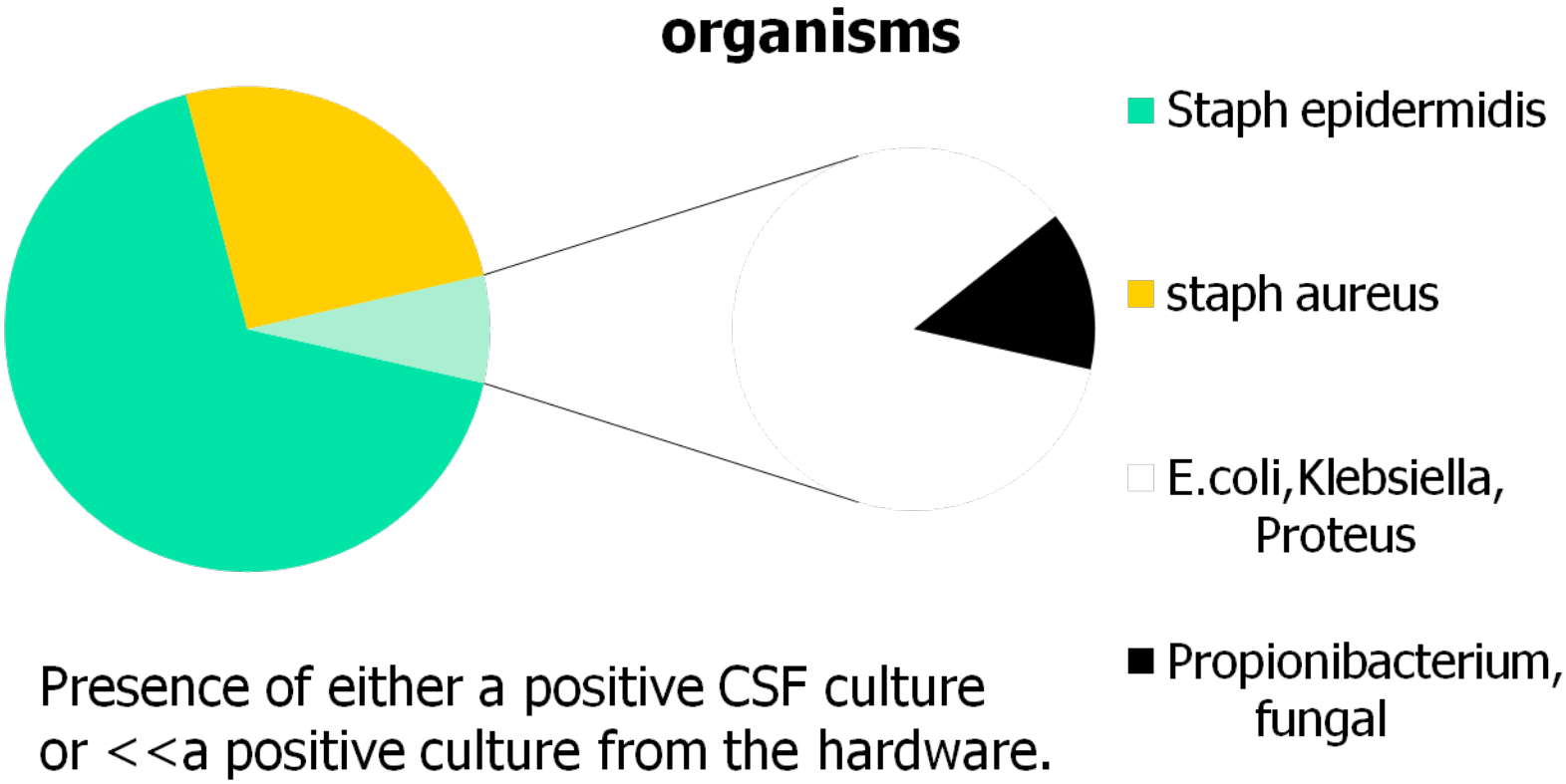
2. Steps-

- Insert a 25 gauge butterfly canula and look for flow. Measure pressure.
- Measure pressure with distal occlude pressed.
- If no spontaneous flow , try to aspirate CSF with a syringe.
- Send CSF sample
- Connect with manometer.
- Repeat measurement after injecting 3-5 ml of saline.

Shunt Infections -Prevention

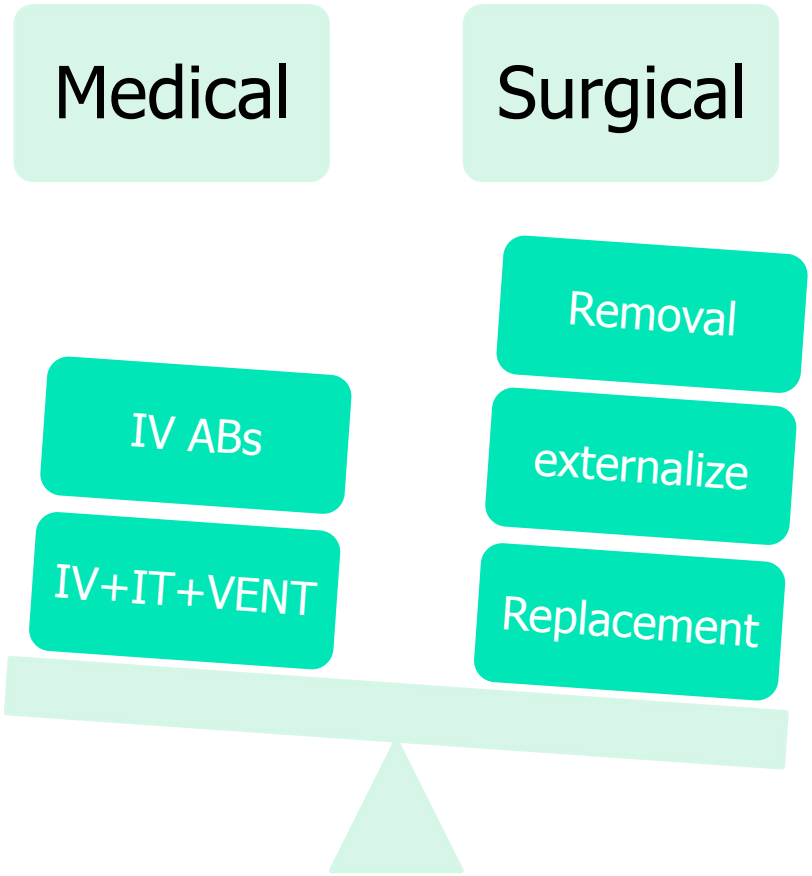
- Sterile surgical technique.
- Haines and Walters have found 50% reduction in infections with use of prophylactic antibiotics.
- Antibiotic impregnated shunt tubing.
- Use of antibiotics before dental procedures, one piece system, biannual screening, hypothermia during surgery.

Shunt Infections -Organisms

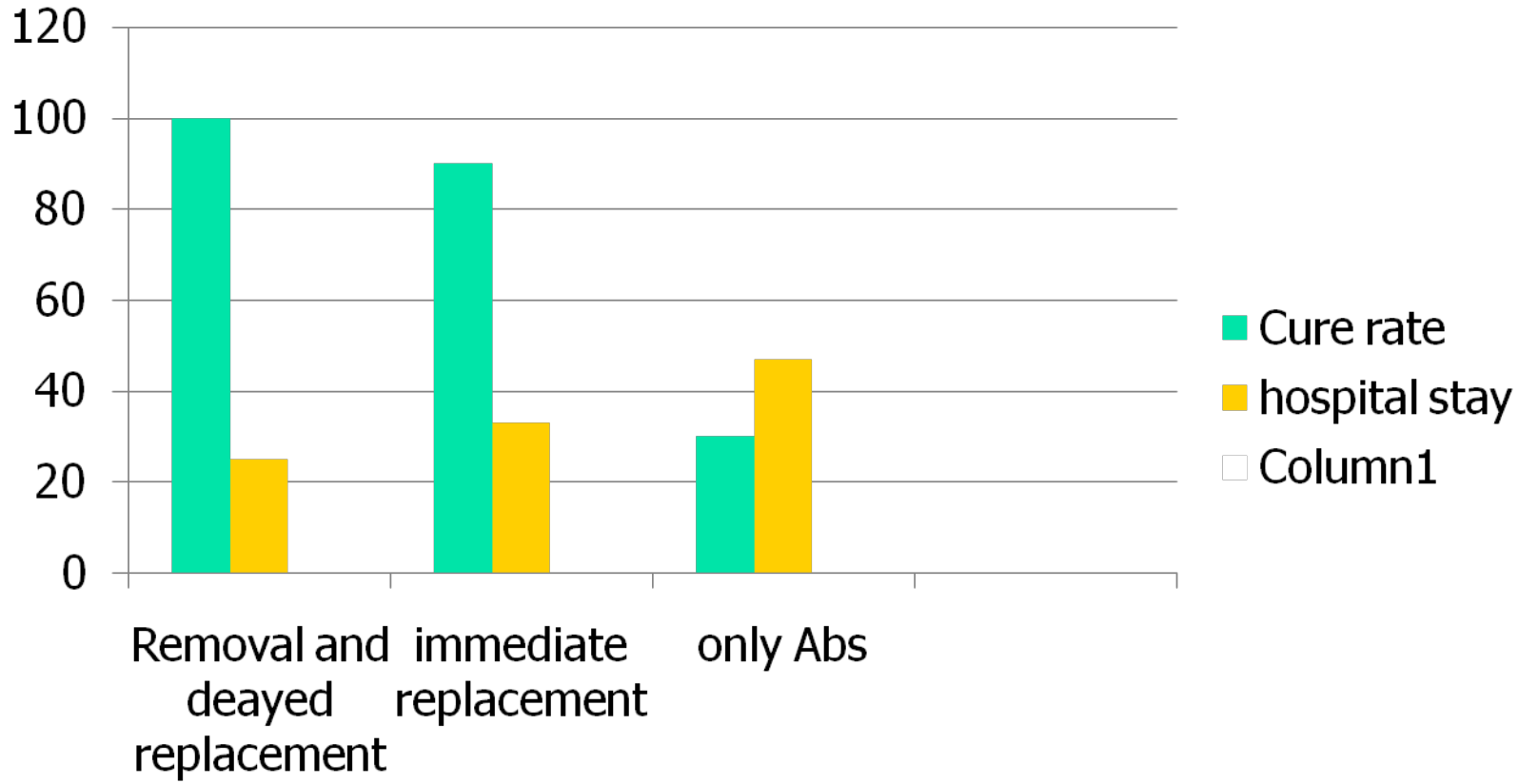


Presence of either a positive CSF culture or <<a positive culture from the hardware.

Shunt Infections -Treatment



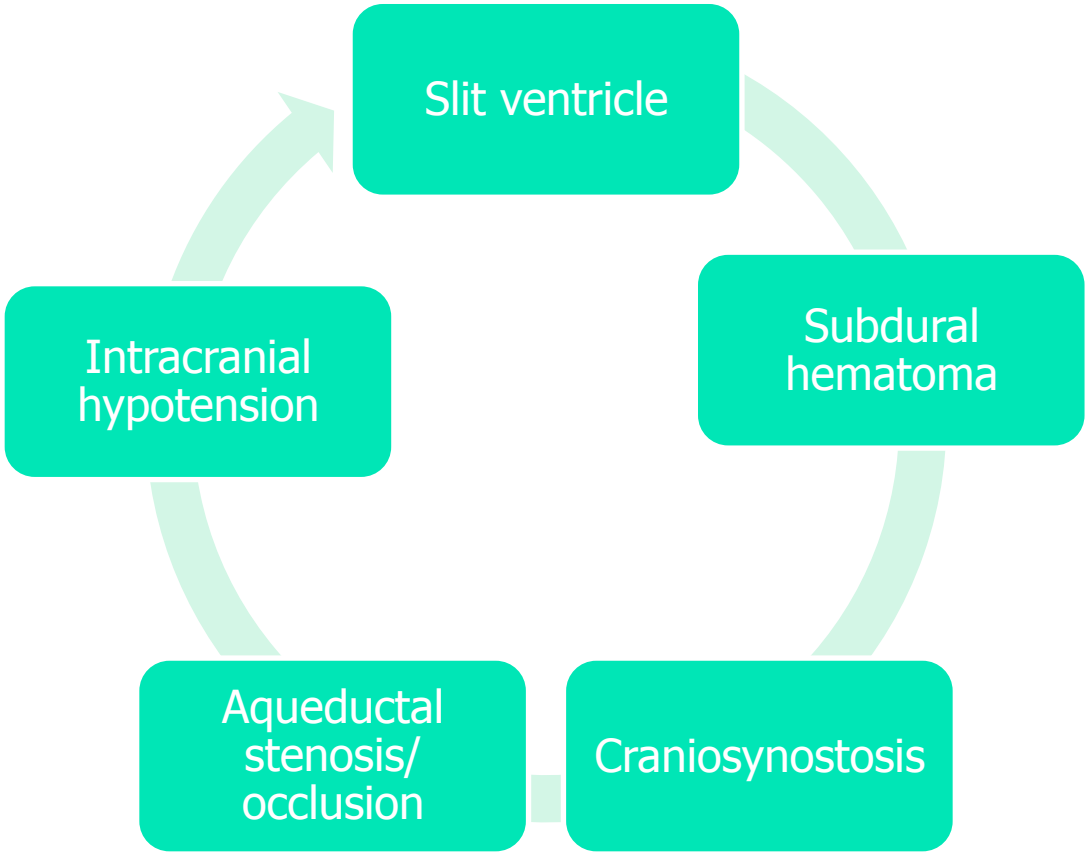
Treatment outcomes



Frame & McLaurin –J neurosurgery.

Shunt Complications-Overshunting

10-12%,VP shunt>VA SHUNT(SIPHONING)



Undershunting

- Shunt malfunction rate 17% in I yr.
- Cause- blockage (choroid plexus/ proteinaceous material/blood); Disconnection.
- Symptoms-H/A,N/V, diplopia, lethargy, ataxia, infants, seizures.
- Signs-upward gaze palsy, 6th CN palsy, field cut, papilledema.

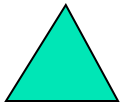
Shunt Complications (continued)

- Disconnection
  A part of the system becomes disconnected. The connections between catheters, valves or accessories are damaged. Sometimes due to growth of the patient.

- Bowel Perforation
  The distal/peritoneal catheter perforates the bowel. Must be revised.

Physiology of shunt devices

- History- hippocrates probably did first ventricular puncture. Nulsen and Spitz did the first ventriculojugular Shunt.
- Hydrodynamics-

1. Flow=  P/R
2. R=8nL/^R4
3. P=IVP+PGH-OPV-DCP.

Shunt physiology-

- Siphoning- Difference in the height of ventricular catheter and that of the distal catheter, causes pressure differential equal to *p*gh.
- Hysteresis- It occurs d/t slight change in the mechanical properties of the valves, depending on whether they are opening or closing. Seen with Slit and miter valves.

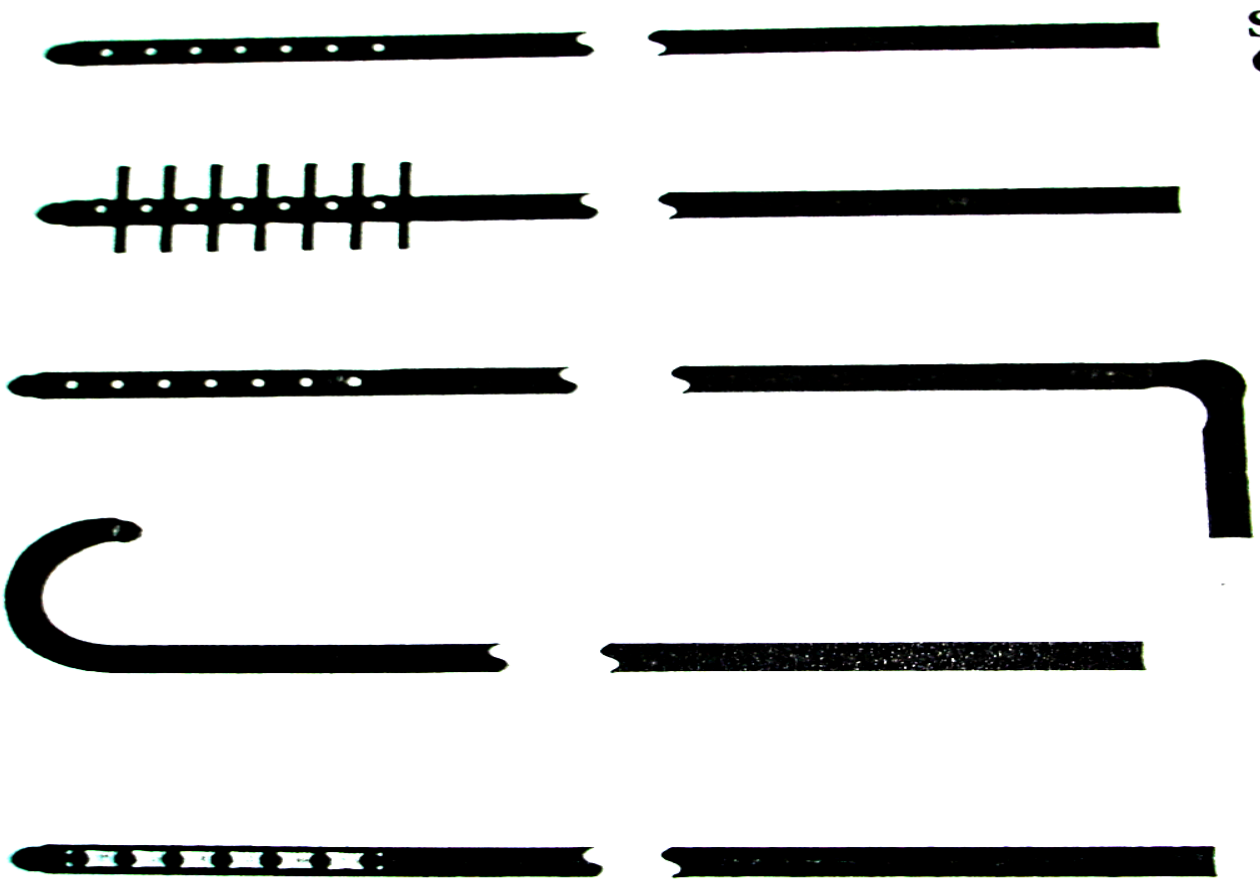
Shunt Systems

- Shunt systems come in a variety of configurations and models but they have similar functional components:
  - Valve Mechanisms – flow or differential
  - Fixed, programmable, or variable settings
  - Catheters
    - Ventricular (proximal)
    - Peritoneal/Atria (distal)
  - Accessories
    - Reservoirs, Siphon Devices
    - Connectors, Filters, Pumping Chambers

Shunt Components - Catheters

- Ventricular Catheters
  - Placed in the ventricles to deliver CSF to the Valve or Distal Catheter
  - A series of holes in the ventricular catheter allows CSF to enter the shunt system

Types of ventricular catheters



Straight Ventricular Catheter

Flanged Ventricular Catheter

Angled Ventricular Catheter

"J" shaped

Recessed Holes

14 1:46

Shunt Components - Catheters

- Distal Catheters
  - The distal tubing is made of silicone and has got slit valves near the distal end with close ended tube or open ended. Valves function when open end gets blocked.

Shunt Components - Valves

- Valves
  - Mechanism which helps regulate the ICP by redirecting enough CSF distally. The valve must provide the optimal balance of CSF diversion, not under drain or over drain the ventricles.
  - The valves are one way and have operating pressure ranges or opening pressure settings.

Valve Basics

- Proximal Valves
  - Valves placed close to the ventricles
- Distal Valves
  - Valves placed in the peritoneum, away from the ventricles

Shunt valves

- Differential pressure valves

A. Slit valves (distal or proximal)
B. Mitre Valves (Hysteris occurs).
C. Diaphragm Valves (Most common).
D. Ball in Cone Valves.

- Defined by their opening pressure.

Flow regulated valves

- Designed to increase the hydrodynamic resistance as the pressure gradient increases. Keep flow rate constant.
- Less likely to be associated with siphoning, but due to small orifice have higher chances of getting obstructed.
- Eg. Orbis sigma valve.

Gravity actuated valves

- Gravity actuated valves attempt to prohibit or reduce siphoning by increasing opening pressure with assistance of gravity when patient sits up.
- Eg. Cordis Horizontal vertical valve for use with LP shunt.

Antisiphon devices

- This device is typically placed under the scalp has a small diaphragm that reduces the rate of CSF flow when pressure inside the shunt falls below atmospheric pressure.

Why a Programmable Valve?

Flow = Δ p / R

200 mmH_2O

Δ p = **ICP - OP + HP - PP**

30 mmH_2O

- Patient dependant (age, physiology,…)
- Time dependant (activity, growth, adaptation…)

Adjustable Valves

- Valve Mechanism allows you to change operating pressure non invasively
- Codman Valve
  - Codman Programmable Valve
  - 18 pressure settings

Valve Components

- Ball & Seat - Synthetic Ruby
- Baseplate - Titanium
- Cam - Polyethersulfone
- Stator – (Nickel alloy)
- Magnets – Samarium Cobalt (SmCo)
- Housing - Silicone
- Spring - Stainless Steel

External Influences

- Investigated During Device Development:
- Vibration & Shock Studies
  - Normal activity levels
- RF Field Studies
  - Household appliances, Cell phones, Airport security gates, ...
- Strong Magnetic Fields Studies
  - MRI units
  - Strong household magnets

External Magnetic Fields

- Magnetic field threshold for deprogramming
- > 80 Gauss (1 step, “optimal” conditions)
- Examples:

Headphone	50 Gauss at the surface
Household appliances	< 10 Gauss
Magnetic Therapy Pillow	>430 Gauss
1.5 Tesla MRI	15,000 Gauss
CHPV Programmer	325 Gauss

=> Unlikely the valve will be affected by everyday sources

Warnings / Precautions

- Valve is supplied without a preset pressure and must be programmed prior to implantation
- Aseptic surgical technique
- Don't flush, fill or pump valve with lint-containing fluid
- Take care to prevent shunt from touching surface
- Don't tie sutures tightly

X-Ray Verification

- Shoot film perpendicular to the plane of the valve
- Shoot film in relation to valve and not patient anatomy
- Non-implanted site of patient's head should rest on plate
- Any angle other than 90 degrees may lead to misinterpretation of pressure setting

MRI Studies

- Safe for use; “MRI Conditional”
  - no movement of valve in tissue pocket
  - no selective heating
  - no effect on valve performance
- MUST Reprogram after each MRI
  - MRI will change the pressure setting

Post-Op Programming

- New pressure setting determined by patient clinical symptoms
- Locating valve mechanism - palpate hard plastic casing
- Mark position of valve mechanism with finger or surgical market
- Transmitter head placed directly over the CAM of valve mechanism.
- Verify setting with VPV, x-ray or Fluoro

X-ray Verification

- As illustrated below, there is a direct correlation between the position of the programming unit control panel pressure selector buttons and the position of the pressure indication on the valves as seen when x-rayed.

- When the valve is programmed to 70, 120, or 170, the pressure indicator aligns with the “X” in the center of the valve.

X-Rayed Valve Information

- The white marker on the valve (1) indicates the right hand side of the valve
  - The marker will not be seen on the x-ray if it is positioned on a 30° angle or more; the valve cannot be programmed if the angle is 45° or more
- The pressure indicator on the white ring (2) denotes the chosen pressure

Non X-Ray Verification

- Clinical Need
  - Minimize patient exposure to X-Rays
- Clinician Performing the Reprogramming
  - Improve ease of use
- Methods Investigated
  - Magnetic verification of valve setting
  - Acoustic verification of valve setting
  - Infrared laser
  - Ultrasound

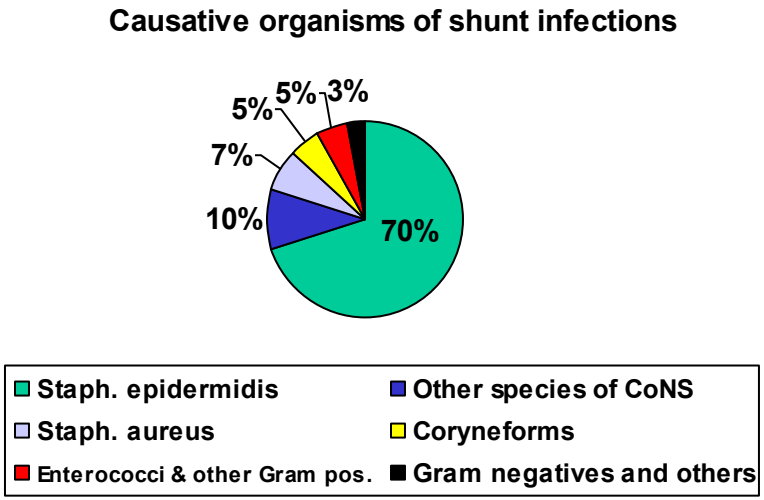
Antibiotic impregnated shunts

»**Bacteria In Shunting**

›Most common bacteria in shunt infections?

Account for approx. 77% of shunt infections.

‹*S. epidemidis*
‹*S. aureus*
‹*Coryneforms*
‹*Streptococci*
‹*Enterococci*



Antibiotic impregnated shunts

- ***Effectivity***
  - *It is not intended to be effective against all causative organisms for shunt infections.*
  - *It is effective against the bugs that are susceptible to rifampicin and clindamycin.*
  - *Rifampicin and clindamycin are effective against most strains of bacteria that cause shunt infections.*

Antibiotic impregnated shunts

- *A shunt infection occurs when a pathogen attaches itself in or on the shunt*
- *Majority of bacterial contamination is introduced at time of surgery*
- *Infection becomes clinically evident in 3-4 weeks post op*
- *Shunt infections can be both internal or external to the shunt*

Internal or External ?

Internal

- *Majority*
- *S. epidermidis or Coryneforms*

External

- *Minority*
- *Wound infection complicated by foreign body*
- *S. aureus*

Antibiotic impregnated shunt- contd

Colonization Process

- *Bacteria adhere to the silicone*
- *The bacteria produce an extracellular slime*
- *This slime adheres to the inner lumen surface of silicone catheters*

Contd..

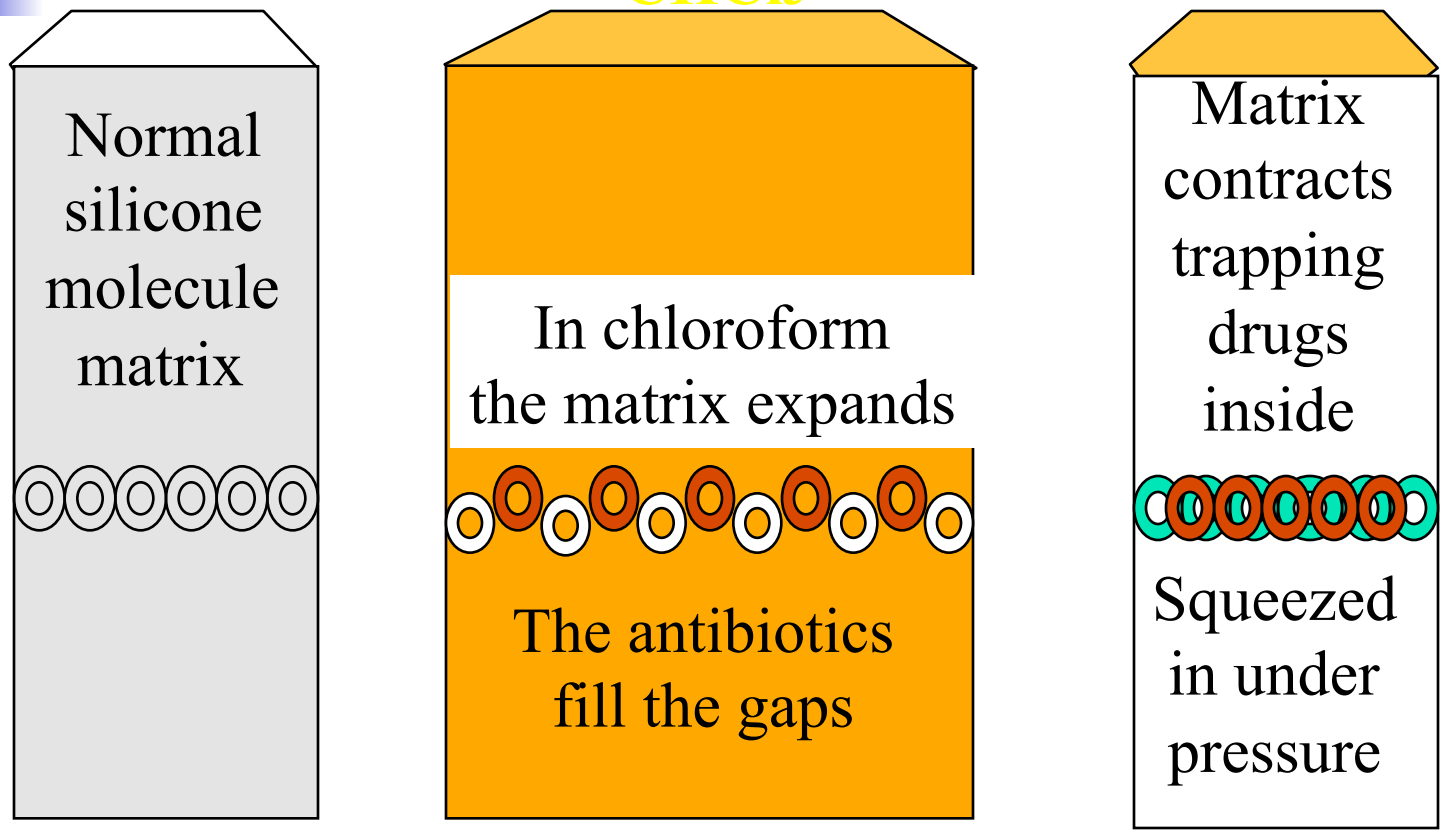
Internal Shunt Infection

- *The organisms start to multiply*
- *And they produce extracellular slime*
- *This can, in time, completely block the shunt*

Antibiotic impregnated shunt

- *Ventricular and Distal Silicone Catheters*
- *Impregnated with Two Antibiotics*
  - *Rifampicin & Clindamycin*
- *And they are ORANGE!!!*

Contd.
How are They Made?



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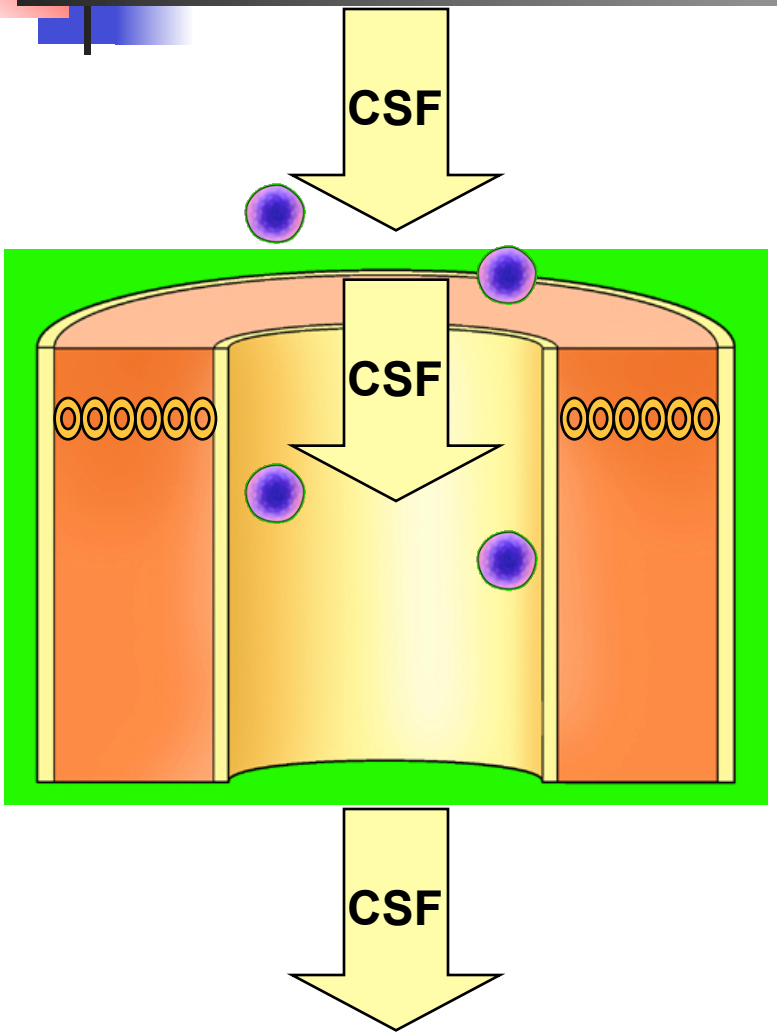
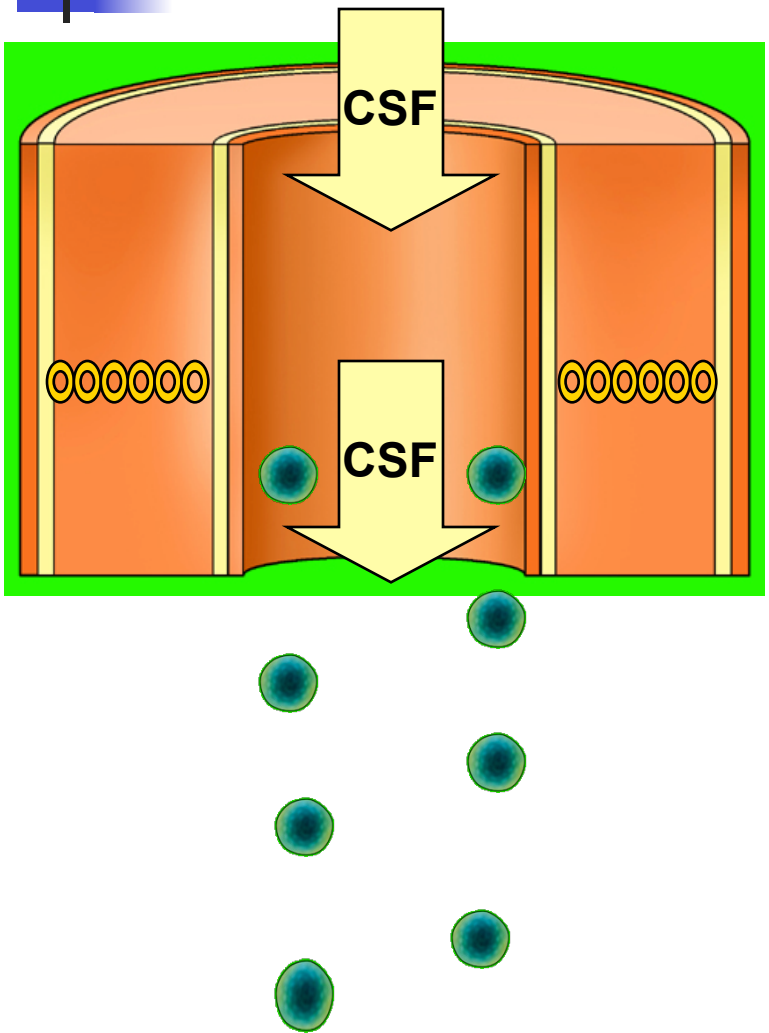
How Do They Work?



Bacteria from the skin, introduced during implantation are carried by the CSF and attach themselves to the catheter surface.

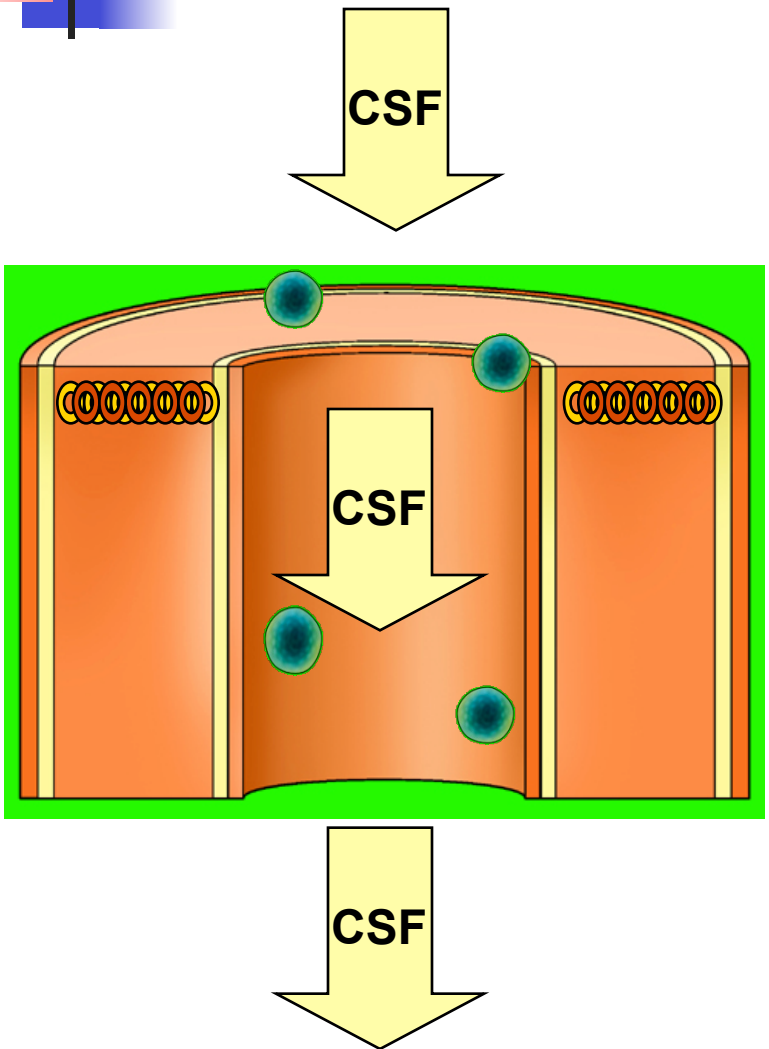
Contd.
How Do They Work?



Laboratory studies have shown that the protective effect is active for at least 28 days protection.

Most shunt infections occur within the first month - having been introduced during implantation.

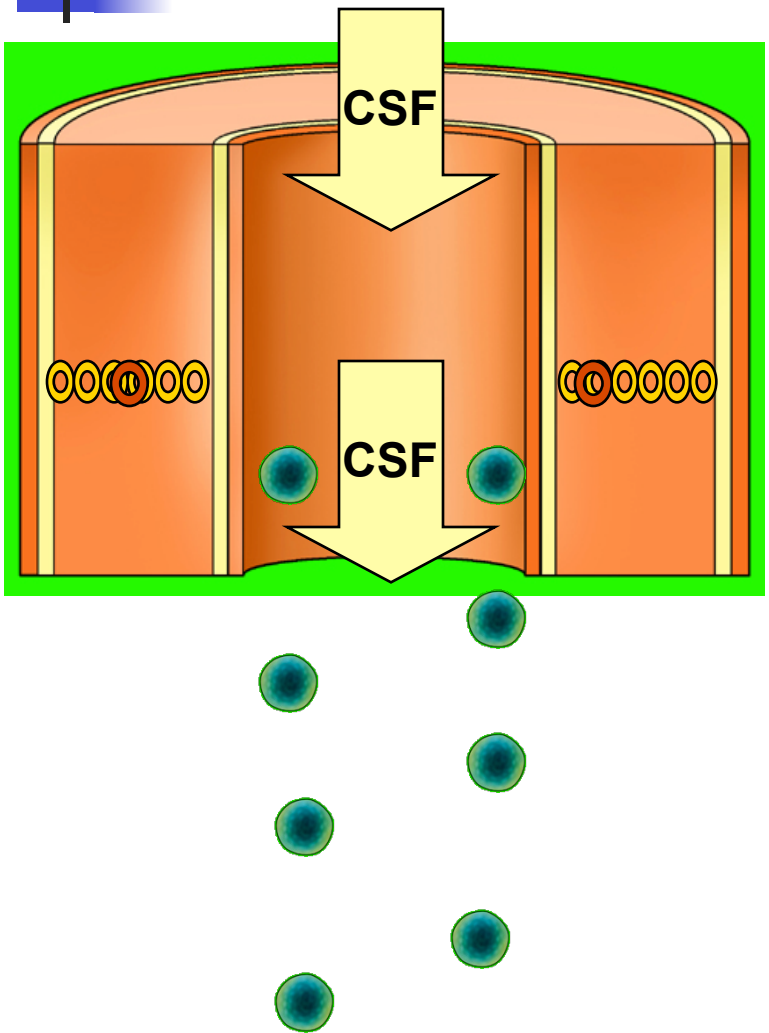
Contd..
How Do They Work?



Due to the concentration difference between the catheter and the external environment, there is a positive diffusion gradient which causes the drugs to slowly diffuse out of the silicone.

The concentration of drugs at the surface of the catheter is high enough to inhibit colonization.

Contd.
How Do They Work?



The concentration of drugs at the surface of the catheter is high enough to inhibit colonization.

Precautions

Pre Implant Technique

- *Surgeon should not pre soak Bactiseal in saline or antibiotic solutions prior to implantation because the diffusion process will be activated.*

Precautions

Packaging

- *It is sterilized by autoclave*
- *It **cannot** be sterilized in the same way as Valves*
- *For this reason it cannot be supplied packed with a valve or unitised.*
- *It must be stored in a temperature controlled environment not to exceed 80°F (27° C)*

Why clindamycin and rifampicin?

These two antibiotics in combination have proven to be effective against the specific organisms that cause the majority of shunt infections.