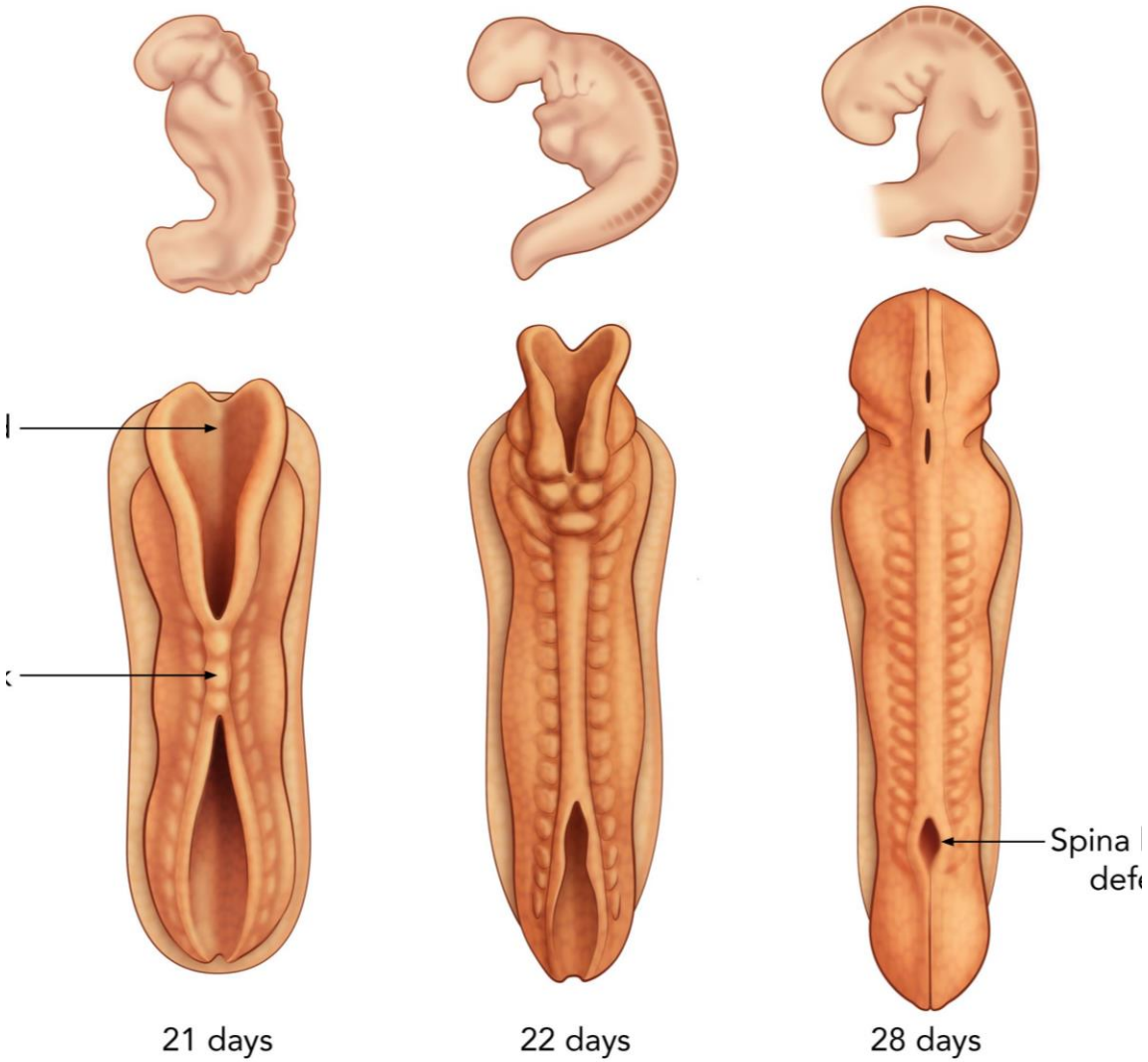
Myelomeningiocele and Tethered cordsyndrome

Neural tube defect

- Congenital malformation of Brain and spinal cord (Dysaphism)
- Incomplete neural tube closure during embryologic development
- Significant reduce incidence with folate supplementation



Types of spina bifida

Occulta

Spina bifida occulta is the least serious and most common type. It is usually discovered only on x-rays or scans. Most people never become aware of their condition.

Meningocele

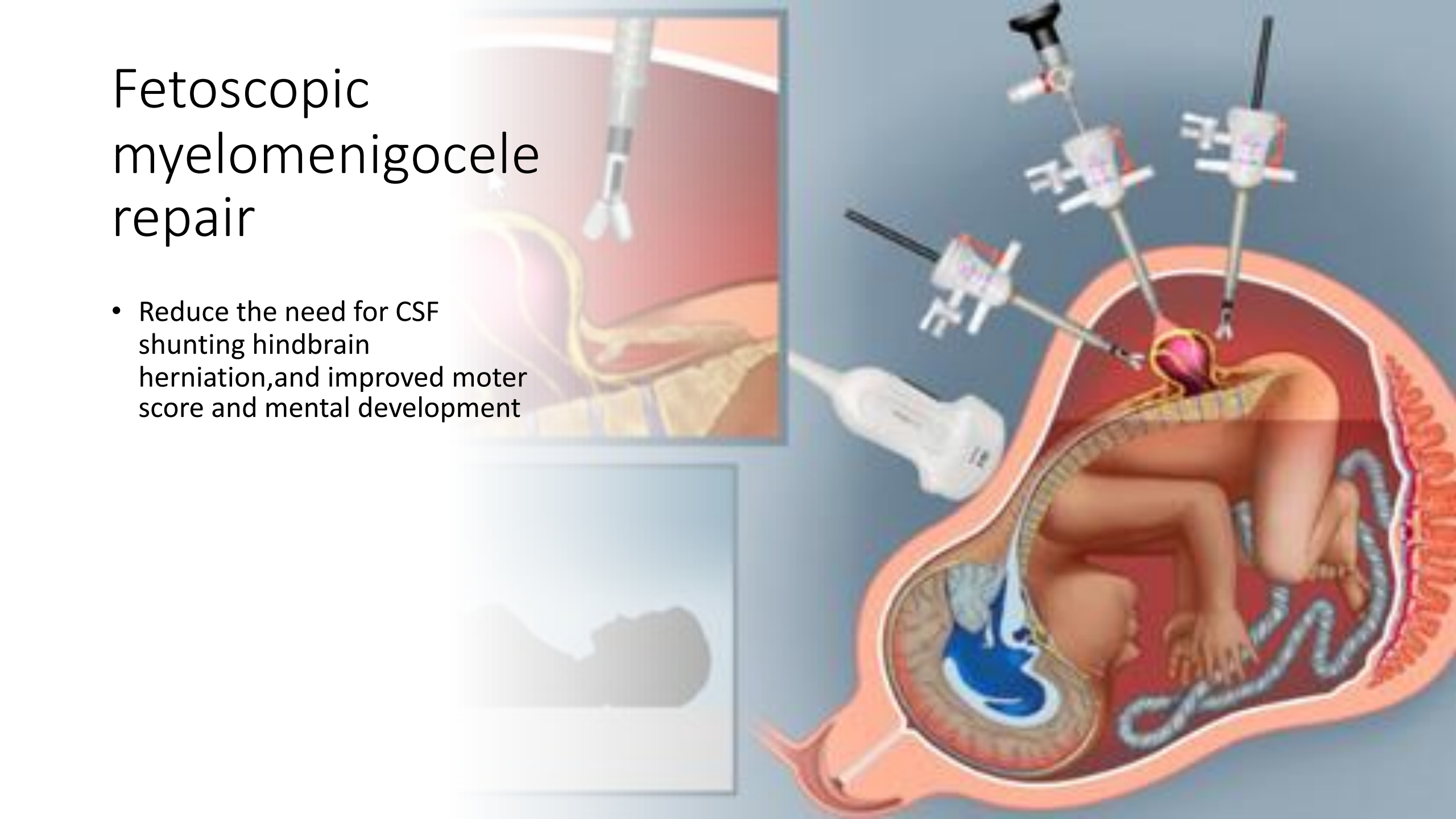
In the next most serious type, meningocele the coverings of the spinal cord (meninges) pass back through the opening in the spine to form a cyst-like swelling.

Myelomeningocele

If the spinal cord is enclosed in the cyst the condition is called myelomeningocele. This is the most serious type of spina bifida.

Fetoscopic myelomenigocele repair

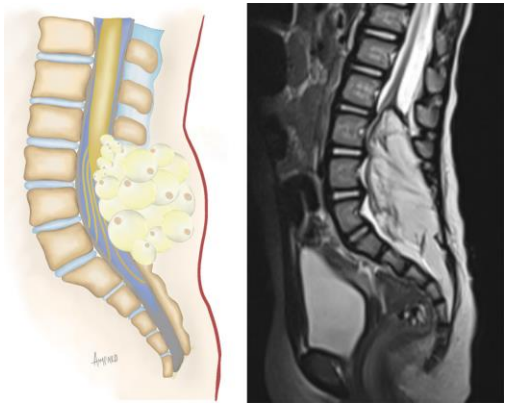
- Reduce the need for CSF shunting hindbrain herniation,and improved moter score and mental development



Procedure/operation

- Myelopmenigocele
- Simple closure
- Repair

- Spinal lipoma
- Lipoma excision

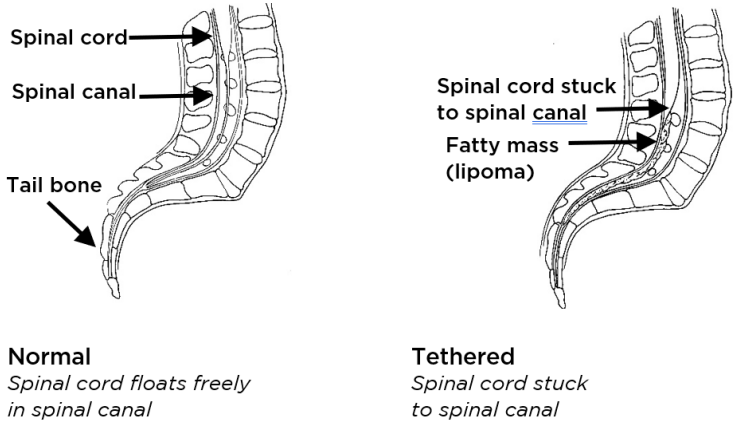


-Tethered cord syndrome

Released tethered cord

Detethered cord

Tethered cord from scar



6 key points

- 1. emergency/ urgency procedure in Neonate

Myelomeningiocele

- Ruptured Myelomeningocele → Urgency in newborn
- Repair within 48-72 hrs after birth → decrease risk of infection and ventriculitis
- Anesthetic Consideration for neonate

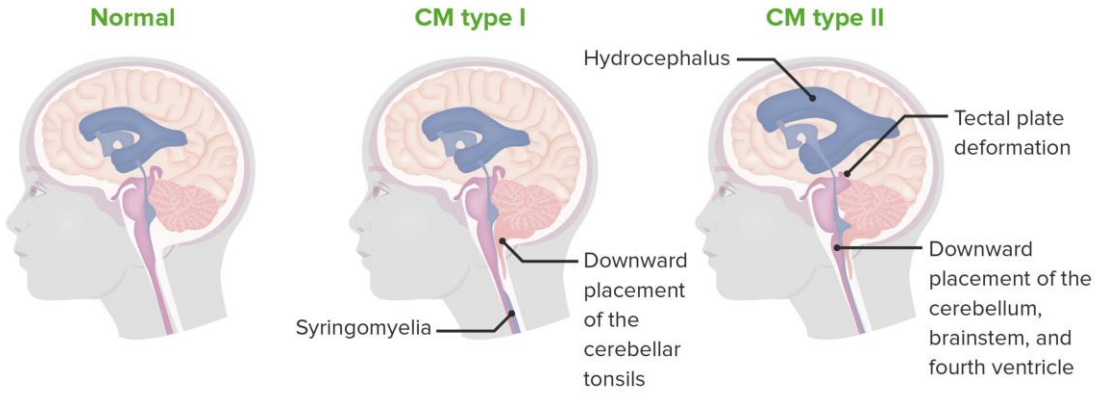
Anesthetic consideration for neonates

- Premature or low birth weight
- Circulatory change at birth

  Transitional pulmonary system,renal and liver immaturity
- Vascular access
- Hypothermia prevention Coagulopathy, SSI,decrease PONV
- Glucose monitoring
- Fluid management
- Lower anesthetic requirement than older children

2. Associted anomalies/diseases

Chari II malformation



- Present in nearly all patient with myelomeningocele

  Downward displacement of cerebellar tonsil and medulla

- Lack of distension of the embryonic ventricular system leads to develop a normal sized cerebellum in a small posterior fossa → CSF flow obstruction → Hydrocephalus
- Crainial nerve and brainstem dysfunction → increase risk of apnea
- Abnormal swallow and gag reflexes → increase risk of aspiration
- Increase risk of vocal cord dysfunction,stridor or bradycardia

Hydrocephalus

85% in myelomeningocele

Hind brain herniation with CSF flow obstruction

Signs of increased ICP in children

- High head circumference
- Positioning during intubation

Sign of intracranial hypertension in infants and Children

Infants	Children	Infants and Children
• Irriabililty	Headache	Decrased conciousness
• Full fontanelle	Diploma	Cranial nerve(II and V) palsies
• Widely separated cranial suture	Papilledema	Loss of upward gate(setting-sun sign)
• Cranial enlargement	Vomiting	Signs of herniation,Cushing's triad,pupillary changes

Myelomeningocele

- Congenital heart defects:ASD,VSD,TOP,coarctation of aorta, hypoplastic left heart syndrome
- Urinary tract complication(91%):incontinence or retention,vesicouteteric reflux

  Prolong urinary catheterization and frequent surgeries
- Bowel dysfuction(87%)

lesion above conus medullaris → increase sphincter tone constipattion

lesion below conus → sphincter flaccity incontinence

- Scoliosis and kyphosis
- Hip and foot deformity

Tethered cord syndrome

- Anorectal anomalies
- VACTERL associate
- Vertebral:hemivertebrae,fused segments
- Atresuas:anal/duodenal atresia
- Cardiac:ASD,VSD ,dextrocardia,TOF
- TE fistula,Esophageal atresia
- Renal:houseshoe kidney,kidney agenesis,ureteropelvic obstruction,hypospadias
- Limb:absent/deformed radius,polydactyly,oligodactyly

During positioning

- Meticulous care and attention during transferring or positioning
- Equipment for prone positioning

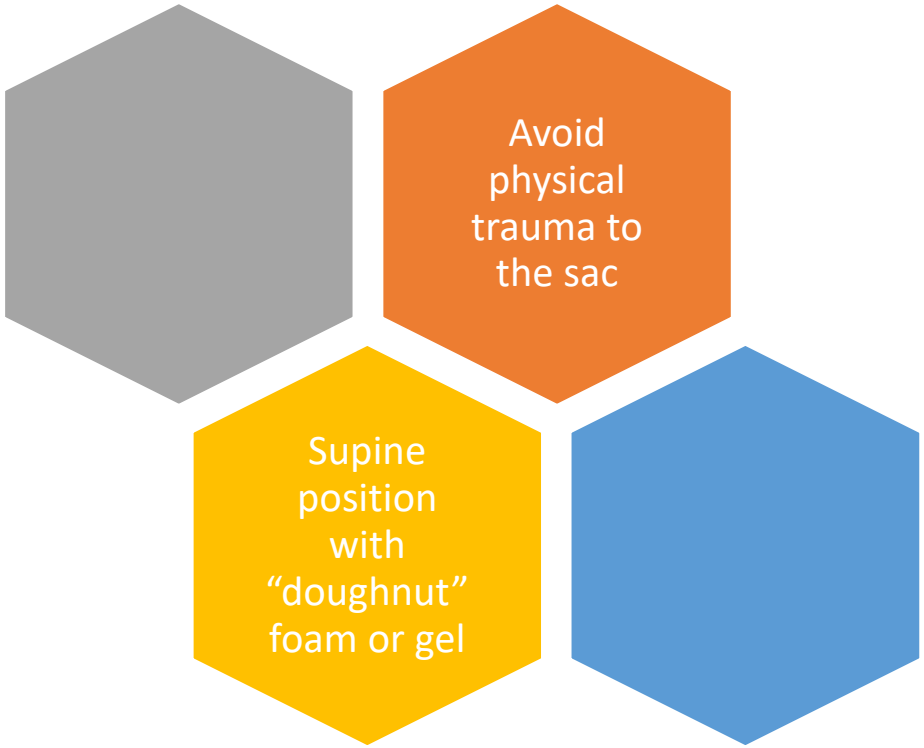
  Silicone pads,gek bolsters

  Carefully checked colostomy or ureterostomy after flip patient

  Included IV line, patency of urinary catheter

3. Intubation

Position during intubation



- Intubation lateral position

  can prevent trauma to defect

- Difficulty ventrilation and intubation:preparation of proper equipment

  Video laryngoscope may help in trained anesthesiologist

Myelomeningocele

- Short trachea in 36% of myelomeningocele patients
- Reduced numbers of tracheal cartilaginous rings
- Be caution of endotracheal intubation
- Check lungs after intubation and positioning

4.Latex allergy

Spinal bifida

- 10-43% of myelomeningocele-history of latex allergy,varying intensity
- Spina bifida patients-Latex Alert(high risk) patients

Produce latex-specific IgE antibiotics with consequent sensitization from disease itselt and the number of exposure to latex-contaioning products.

Frequent exposure to latex from multiple surgeries → increased risk of latex sensitivity

Latex free environment for patients with spina bifida → reduce risk of Latex sensitivity

- Scheduled first on the OR list
- Removed all latex-containing products esp.gloves,urinary catheter
- Anesthetic circuits,IV catheter,IV fluid-Latex free
- Latex-free box:lists of product safe to use,protocol for anaphylaxis

5.Intraoperative Neurophysiologic Monitoring(IONM)

IONM

- Detect and prevent neurological injuries esp.Tethered cord syndrome
- SSEP
- MEP
- EMG
- BCR

Anesthetic agent for IONM

- TIVA propofol and opiod as fentanyl or remifentanil
- Inhalations dose dependent reduction in signal of MEP,SSEP,BCR
- Recommendation from JSA(2020)≤ 0.5 MAC add on TIVA for reduce propofil used in all-aged pediatric case requiring MEP.
- Low dose inhalation can be used additionally,but a prompt switch to TIVA is recommended if the IONM signals cannot be facilitated.
- N2O should be avoided.
- NMBA suppres signals of MEPS,BCR,EMG

MEP,BCR:TIVA
Avoid NMBA

SSEP <1.5 MAC inhalation or TIVA
NMBA use as necessary

EMG:Avoid NMBA

Our Practice in MEP,BCP,pediatric cases

- Induction with IV anesthetic or inhalation
- Intubation with a dose of NMBA
- Bite block to prevent oral tissue injury
- Maintenance with inhalation in the first period of procedure,foley catheterization,Iv catheter,IONM needle insertion
- After turing to prone position:anesthetic technique was switched to propofol-based TIVA with fentanyl(no N2O) for baseline signals

  Propofol 100-170mcg/kg/min,Fentanyl 1-2 mcg/kg/hr
- After neurosurgeon finished risky operation, the final IONM signals were performed.Change to inhalation+/- NMBA to avoid a high dose of propofol and to facilitate emergence.

Physiologic factors affedted IONM

- Hypothermia
- Hypotension
- Anemia
- Hypoglycemia
- Hypoxemia,hypocapnia
- Electrolytes abnormalities;HypoNa,HyperCa

6.Postperative Extubation

- Most of cases: Extubation at end of procedures
- Delayed extubation in comorbidities,massive blood transfusion,airway edema from long duration of surgery,postoperative apnea,from Chiari II Malformation
- Postoperaative apnea of prematurity in preterm neonate