

Spina Bifida

(spinal dysraphism)

by

Dr. Mohamed Al Tamimi

Specialist neurosurgeon

Lecturer in medical college



Spina Bifida

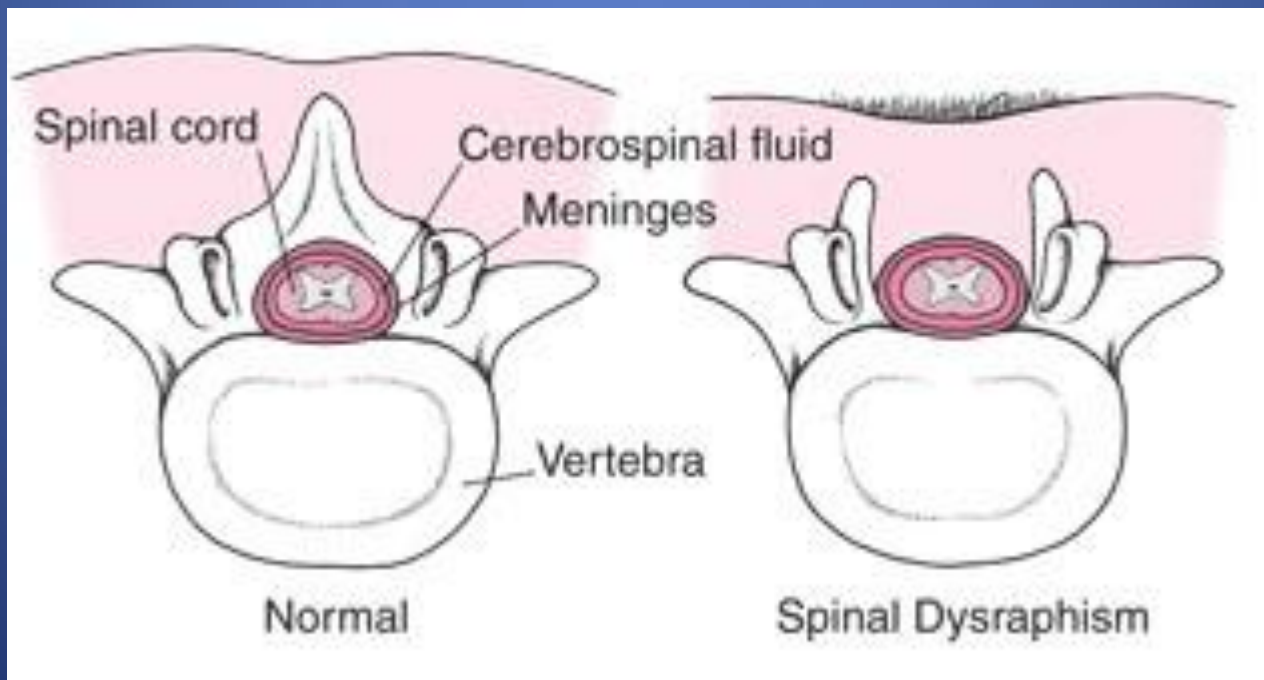
- What is it?
- What causes it?
- How can it be prevented?
- How can it be managed?
- Social impact
- Further information

Spina Bifida

- What is it?
- What causes it?
- How can it be prevented?
- How can it be managed?
- Social impact
- Further information

What is spina bifida?

“Split Spine” caused by incomplete closure of the neural tube, usually in the lumbar or sacral region



What is spina bifida?

- Worldwide incidence is 1-2 cases in 1000 births

 - US incidence is 0.7 per 1000 live births

 - East coast higher than West coast

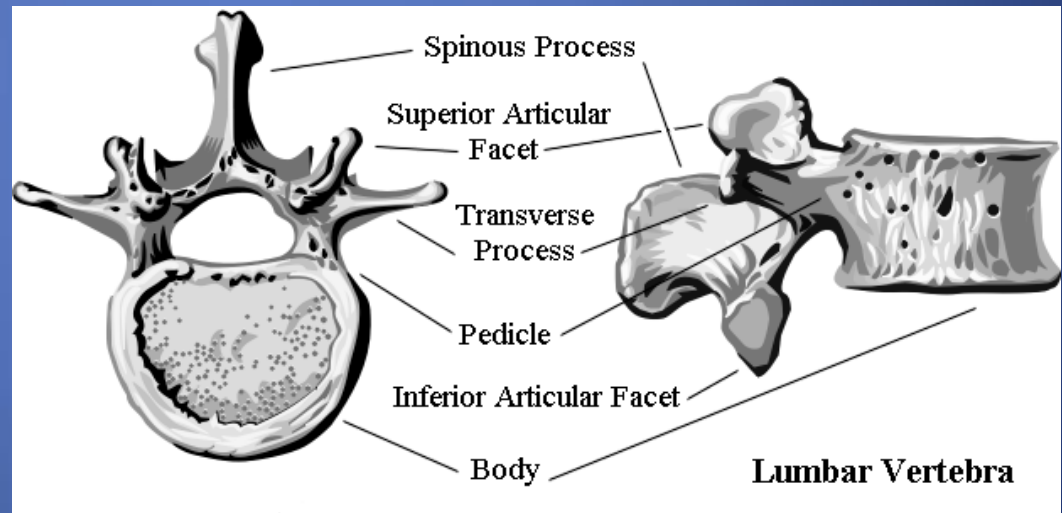
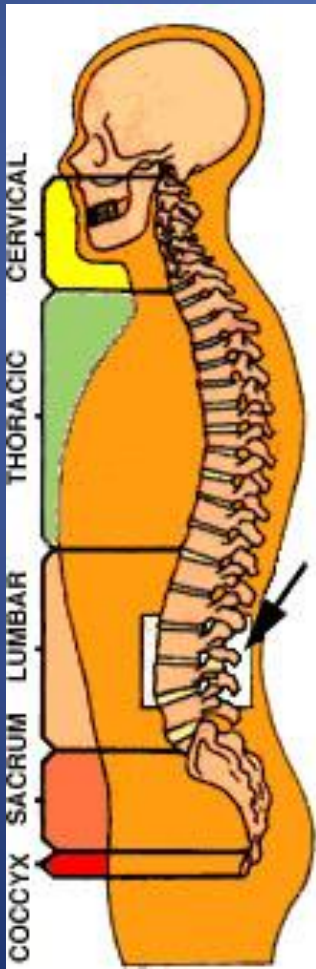
 - Slightly higher incidence in Caucasian population

 - Irish immigrants also have a higher risk

- Seen more in children born in late summer and early fall

What is spina bifida?

Anatomy review



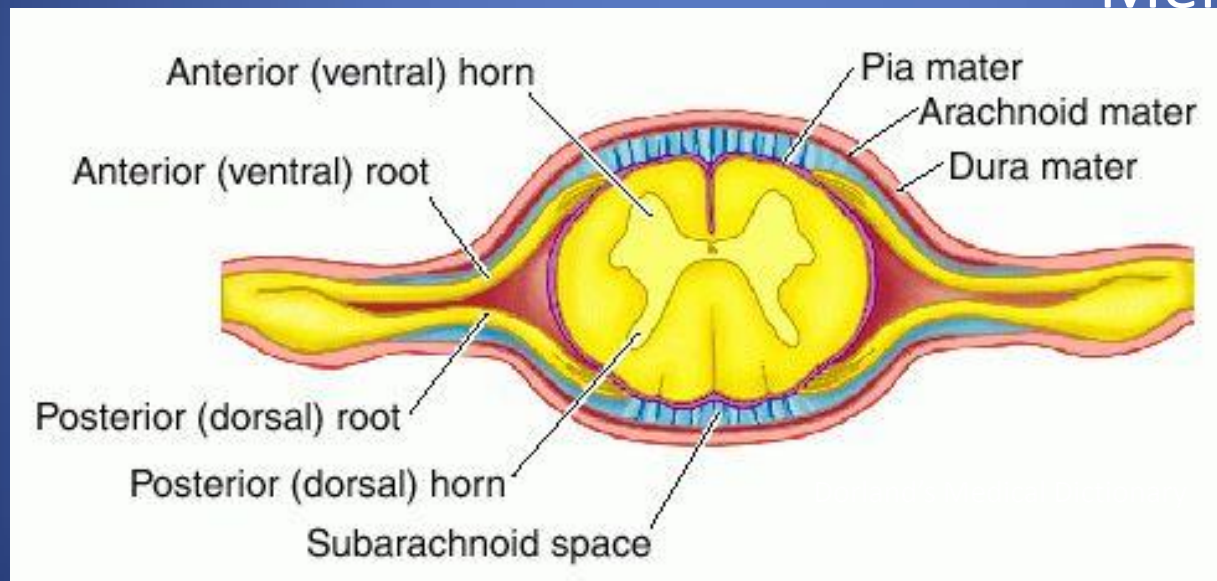
<http://www.fpnotebook.com/LumbarSpineAnatomyVertebra.gif>

http://thespine.net/articles/lumbardecompression_files/image001.gif

What is spina bifida?

Anatomy review

Meninges



What is spina bifida?

Several classifications that vary in severity depending on location and extent of opening

Spina bifida occulta

Spina bifida cystica

meningocele

myelomenigocele

Spina bifida ventralis

What is spina bifida?

Spina bifida occulta – “hidden”

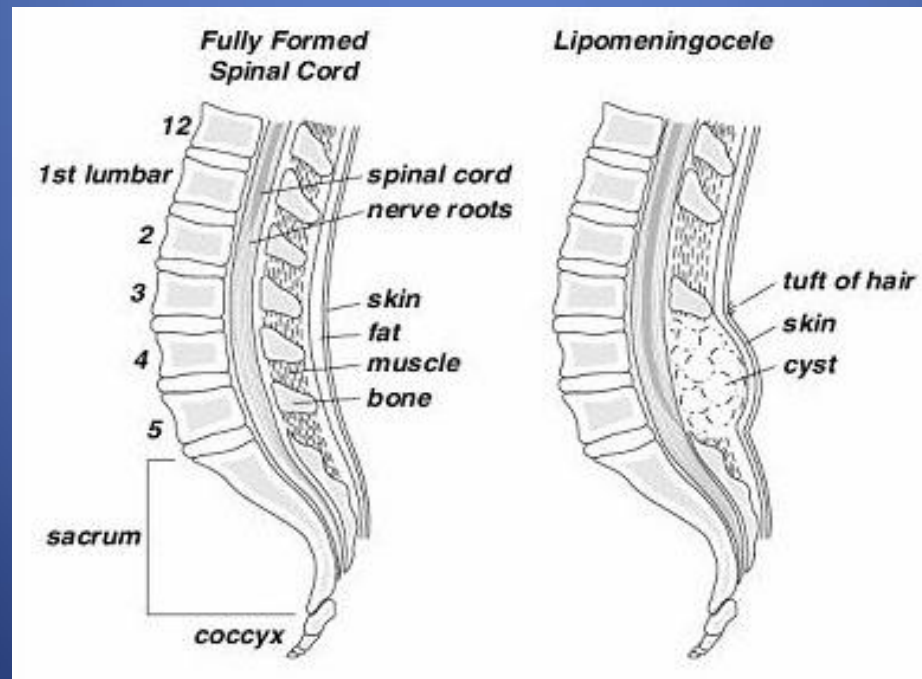
- The bony vertebra is open, but the spine is within the spinal canal
- The skin may have a lipoma (small benign fatty tumor), some discoloration (birthmark), or a small tuft of hair overlying the spinal defect
- Most patients with spina bifida occulta do not know they have it
- There may be tethering of the spinal cord



What is spina bifida?

Spina bifida occulta – tethered spinal cord

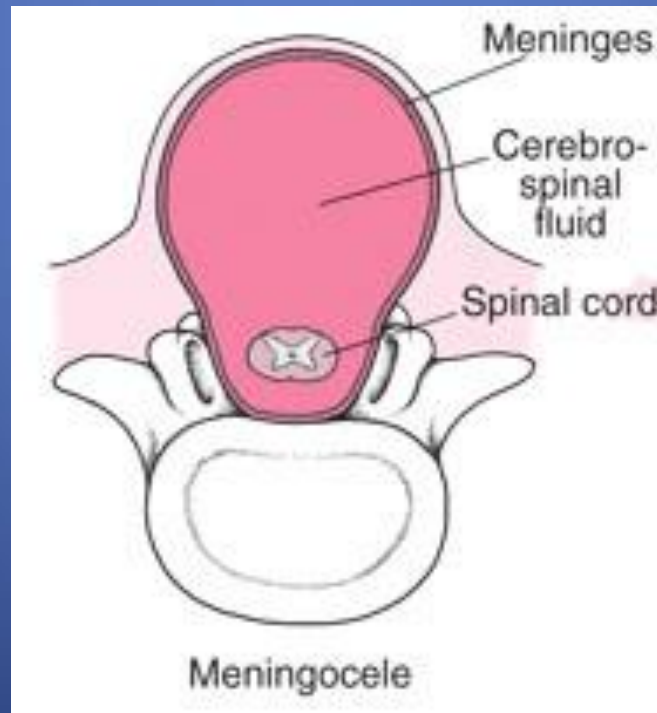
- Often occurs later in life
- Caused by limitations of movement of the spinal cord within the spinal column
- Patients often have low back pain, weakness in the legs, and/or incontinence depending on the site of tethering



What is spina bifida?

Spina bifida cystica – meningocele

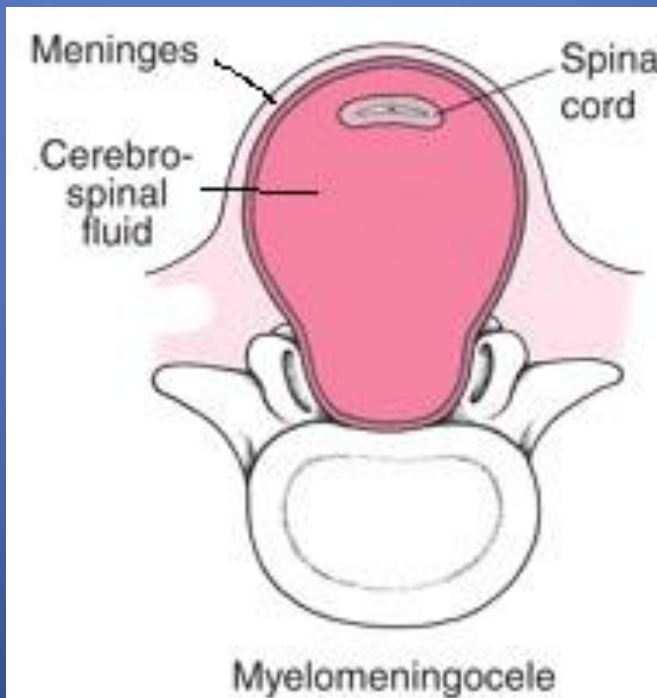
- The bony vertebra is open, part of the meninges is protruding out of the spinal canal
- Since the spinal cord is not protruding, there is often normal function
- Some cases of tethering have been reported



What is spina bifida?

Spina bifida cystica – myelomeningocele

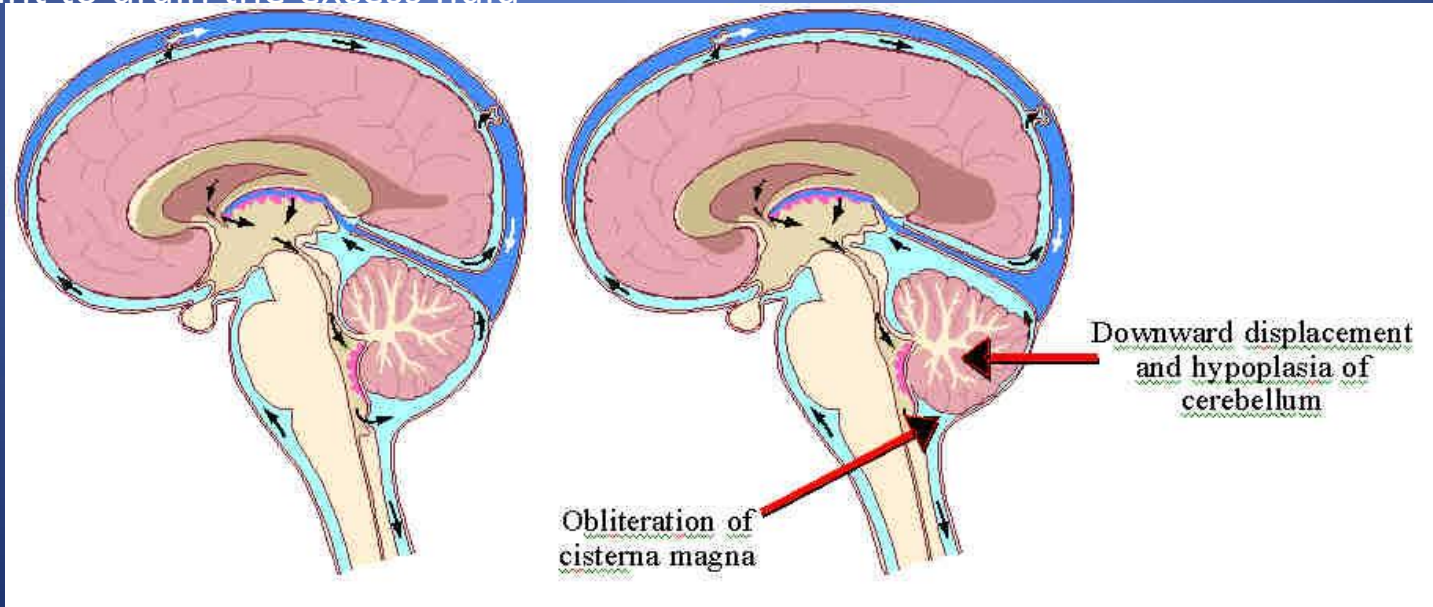
- The bony vertebra is open, part of the meninges and part or all of the spinal cord is protruding out of the spinal canal
- Since the spinal cord is protruding, it is often not fully developed
- Involved nerve roots are often not developed resulting in weakness, pain, and/or paralysis



What is spina bifida?

Spina bifida cystica – myelomeningocele

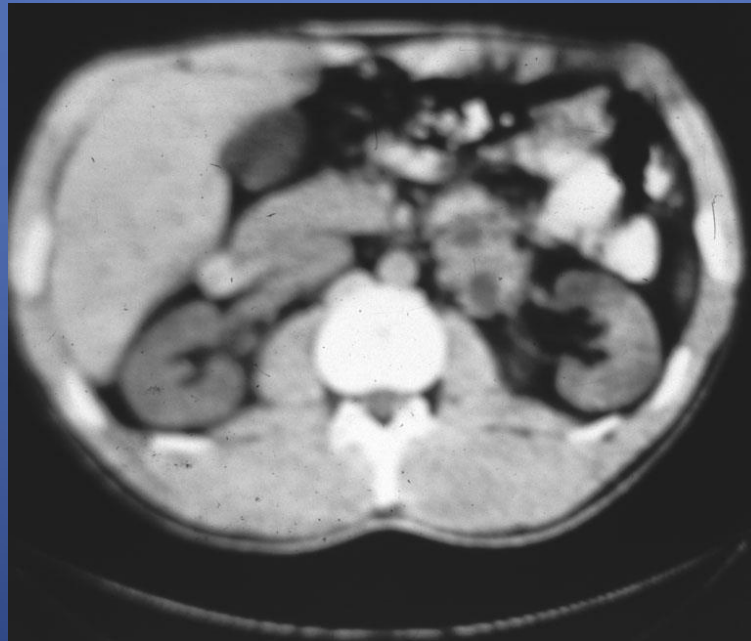
- Arnold Chiari malformation II is often associated with myelomeningocele and occurs when the cerebellum is forced downward
- This can result in life-threatening situations because the build-up of cerebrospinal fluid can cause pressure on the brain
- Patients with Arnold Chiari malformations often require placement of a shunt to drain the excess fluid



What is spina bifida?

Spina bifida ventralis – anterior opening

- Much less common than other forms of spina bifida
- Meningeal sac will protrude into the retroperitoneal space and impinge on retroperitoneal organs such as the duodenum, ascending/descending colon, kidneys, adrenal glands, pancreas, aorta, and inferior vena cava



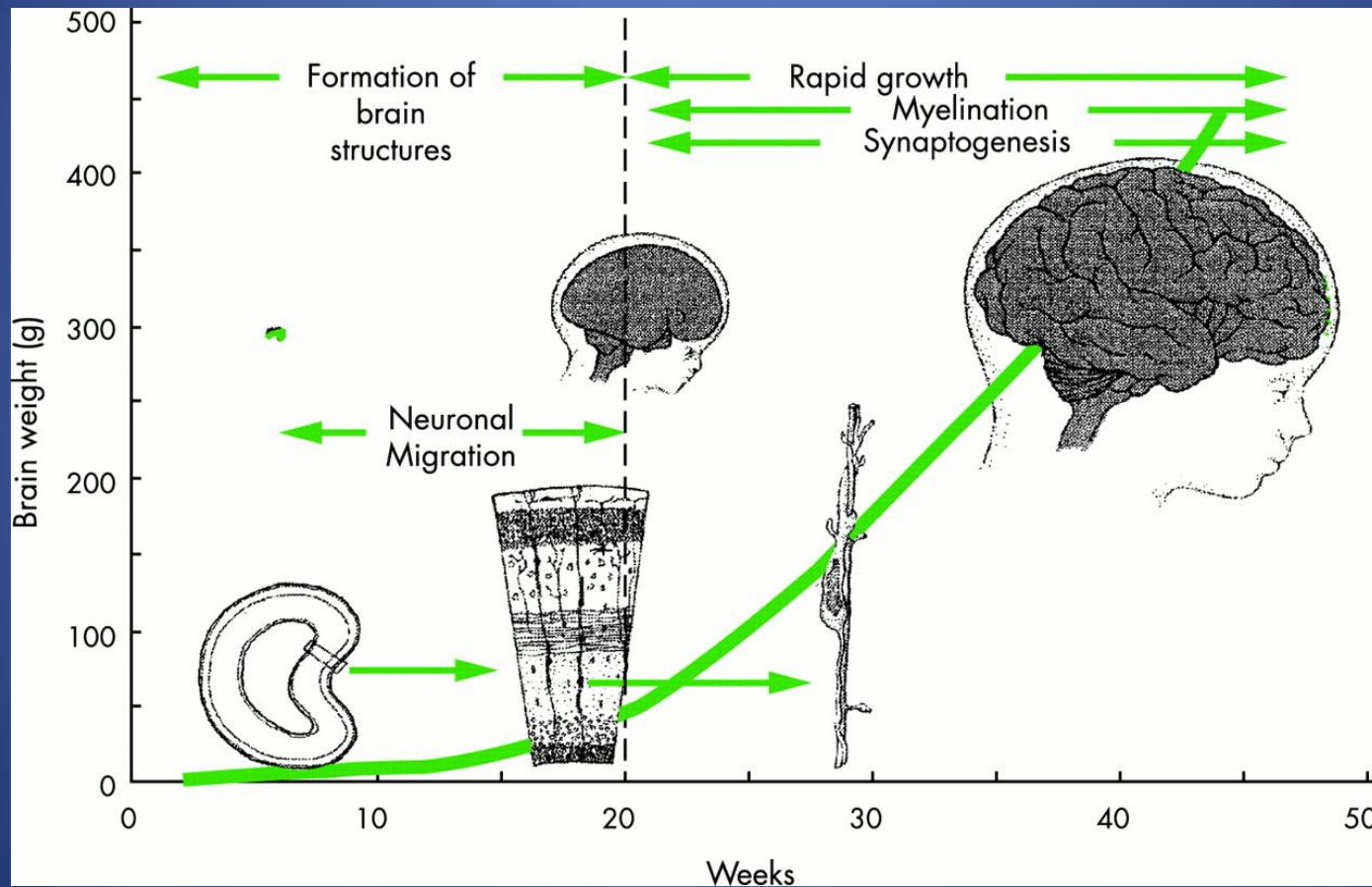
Spina Bifida

- What is it?
- What causes it?**
- How can it be prevented?
- How can it be managed?
- Social impact
- Further information

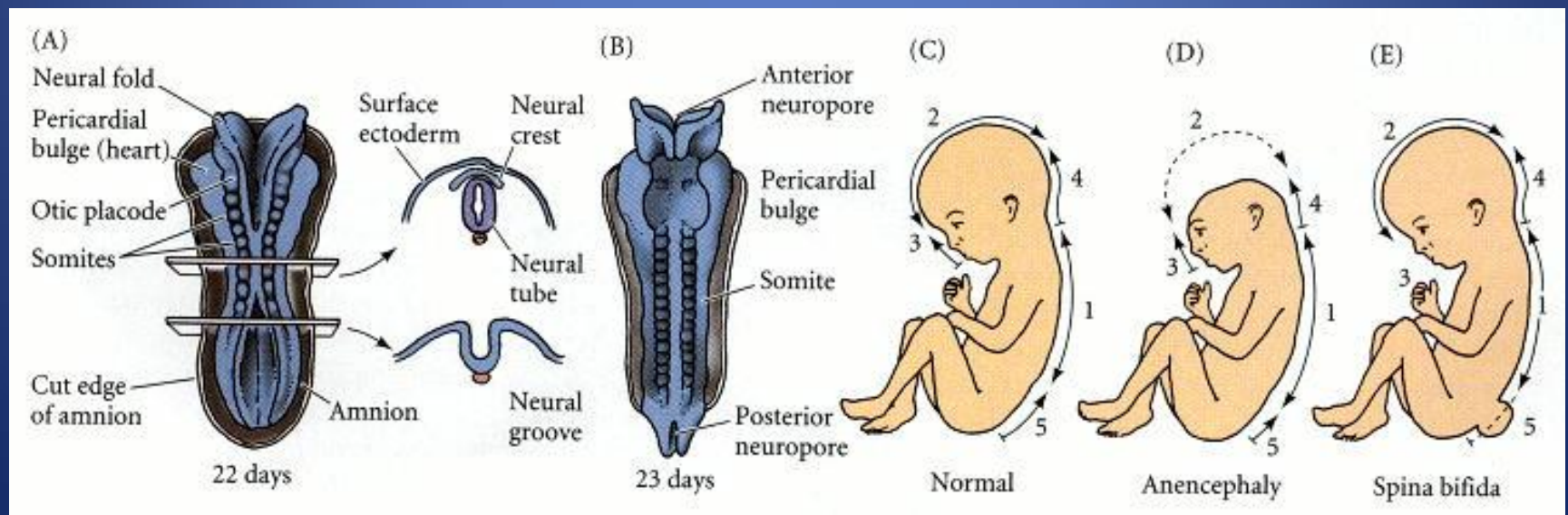
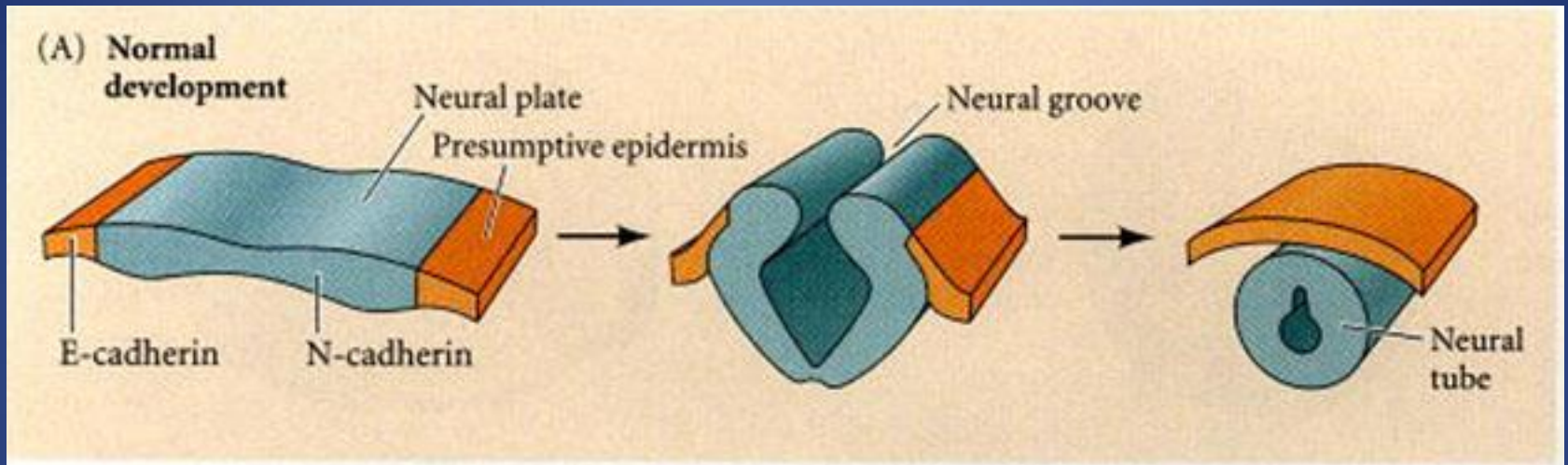
What causes spina bifida?

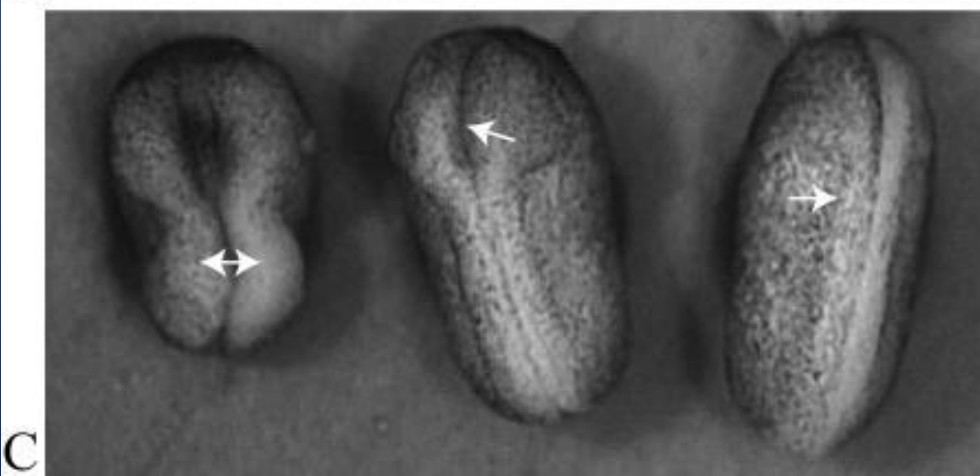
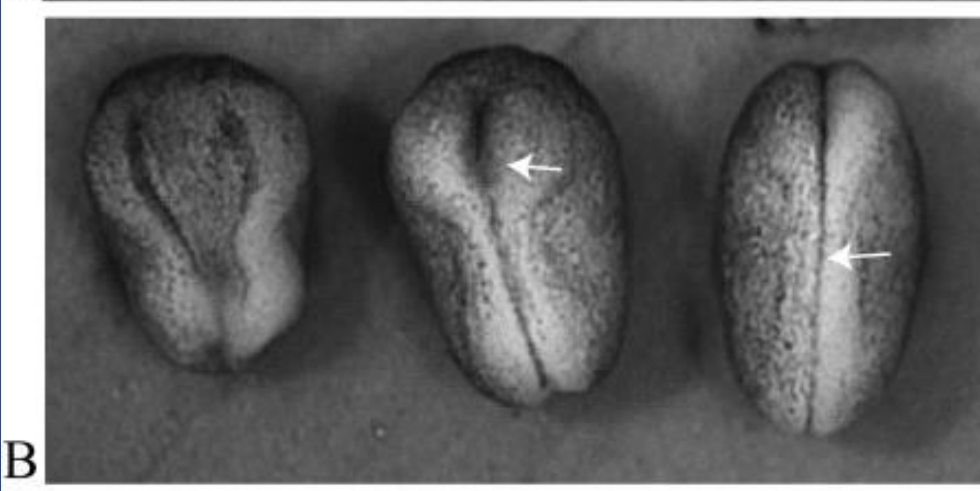
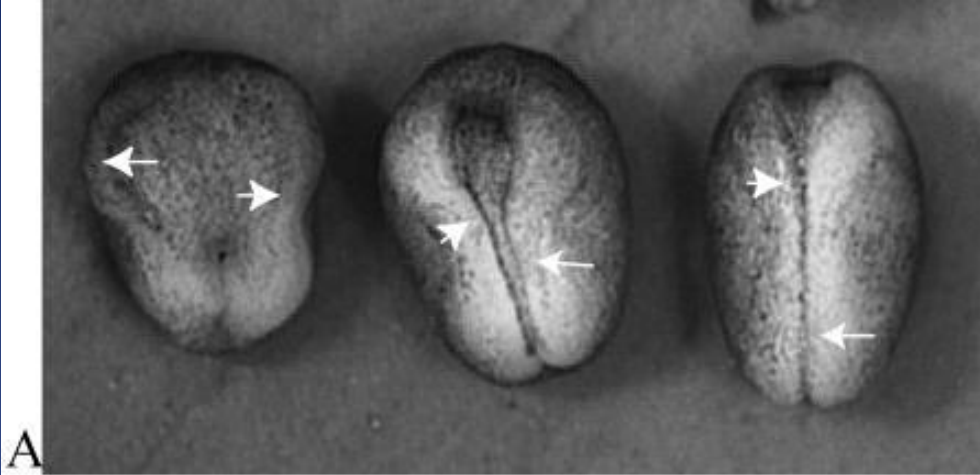
- The exact cause of spina bifida is unknown
- All research to date has indicated both a genetic and environmental influence
- The developmental process that results in spina bifida is well studied

Genetic Neuronal Migration White Matter Injury Cortical gray & subcortical injury



What causes spina bifida?





Stages in the closure of a xenopus neural tube

Video Views and Reviews: Neurulation and the Fashioning of the Vertebrate Central Nervous System
Christopher Watters

What causes spina bifida?

What would prevent the neural tubes from closing properly?

Folate seems to play a large role in the closing of the neural tube – but it is unknown exactly how folate works in this process.

Folate influence was discovered by the increased incidence in spina bifida seen in Irish babies born in late summer and early fall. The lack of leafy green vegetables caused the mother to have low levels of folate during conception.

Genetics also play a role in the development of spina bifida.

- Mothers with one child with spina bifida have an increased risk of additional children having spina bifida
- Studies with folate-resistant mice

What causes spina bifida?

Valproic acid (Depakote)

- Oral medication used to treat seizures/convulsions, migraines, and bipolar disorder
- Mechanism of action – thought to increase GABA levels in the brain
- Pregnant women taking Valproic acid have an increased risk of having children with spina bifida



Spina Bifida

- What is it?
- What causes it?
- How can it be prevented?**
- How can it be managed?
- Social impact
- Further information

How can spina bifida be prevented?

Education

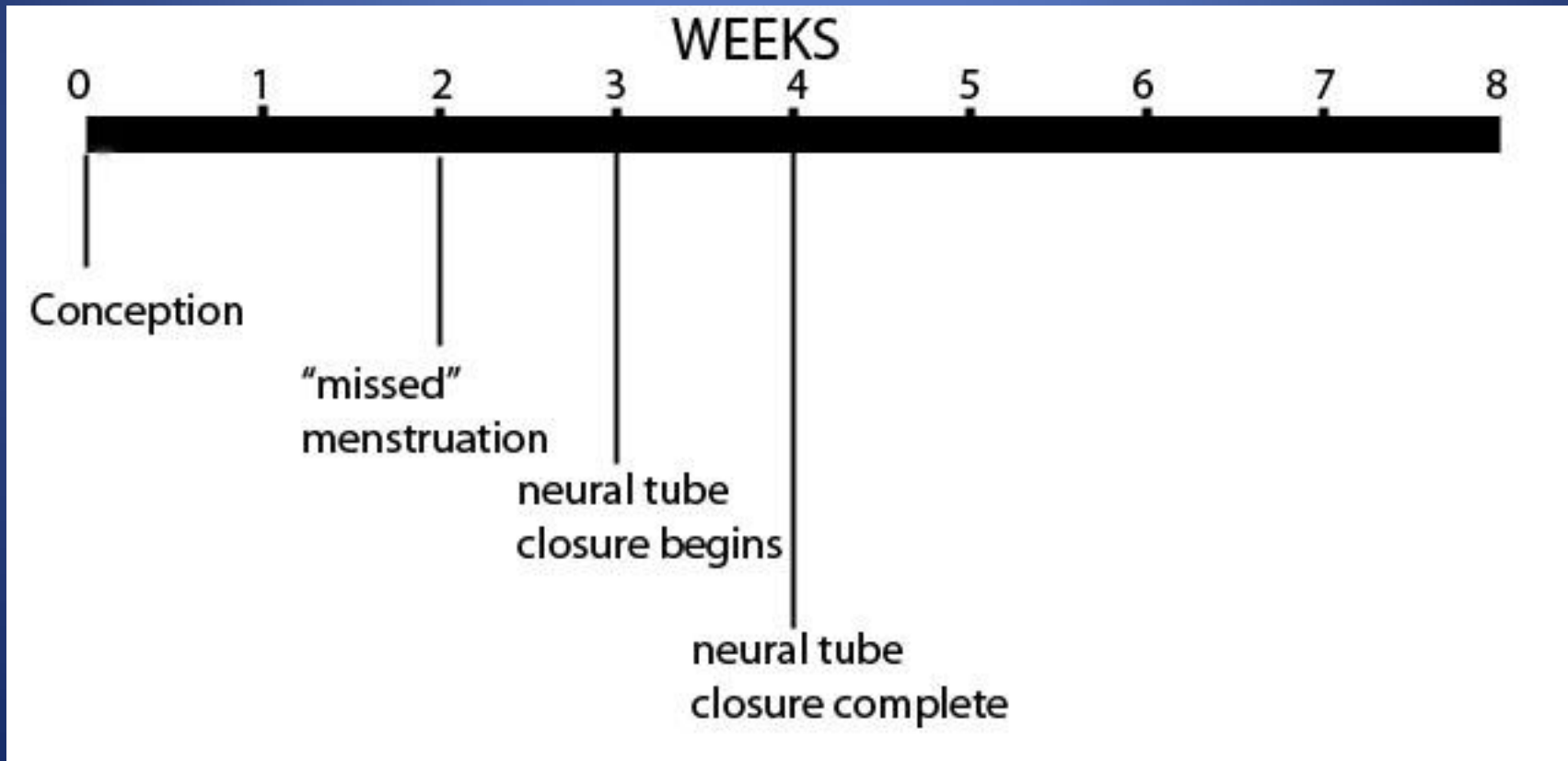
Folate intake

How can spina bifida be prevented?

Education

- Neural tube begins to close at day 22 after conception
- Neural tube is usually fully closed by day 28 after conception

How can spina bifida be prevented?



How can spina bifida be prevented?

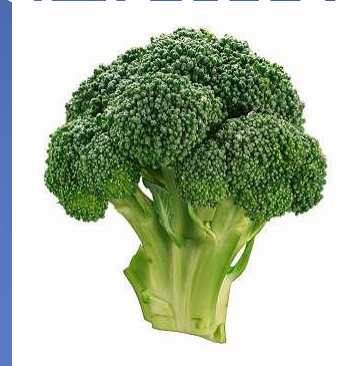
Folate intake

- Recommended that women of child-bearing age take 400 micrograms of folate per day
- Pregnant women should take 600 micrograms of folate per day
- Women with a previous child with spina bifida should take 4000 micrograms of folate per day
- Folate can decrease the risk of spina bifida by up to 75%

How can spina bifida be prevented?

Sources of folate

- Vegetables and grains
- Many foods are now enriched with folate
- Most multi-vitamins contain folate
- Some vitamins are specifically formulated for women



Nutrition Facts			
Serving Size 1 cup (228g)			
Servings Per Container 2			
Amount Per Serving			
Calories	250	Calories from Fat 110	
% Daily Value*			
Total Fat	12g		18%
Saturated Fat	3g		15%
Cholesterol	30mg		10%
Sodium	470mg		20%
Total Carbohydrate	31g		10%
Dietary Fiber	0g		0%
Sugar	5g		
Protein	5g		
Vitamin A			4%
Vitamin C			2%
Calcium			20%
Iron			4%
Folate			30%

* Percent Daily Values are based on a 2,000 calorie diet. Your Daily Values may be higher or lower depending on your calorie needs:

	Calories:	2,000	2,500
Total Fat	Less than	65g	80g
Sat Fat	Less than	20g	25g
Cholesterol	Less than	30mg	300mg
Sodium	Less than	2,400mg	2,400mg
Total Carbohydrates		300g	375g
Dietary Fiber		25g	30g

Spina Bifida

- What is it?
- What causes it?
- How can it be prevented?
- How can it be managed?**
- Social impact
- Further information

How can spina bifida be managed?

Treatment for spina bifida depends on the extent of spinal cord involvement

- Spina bifida occulta usually requires no treatment unless pain from tethering develops
- Meningocele usually requires removal and early management of the cyst
- Myelomeningocele usually requires the most extensive treatment, but treatment is variable

How can spina bifida be managed?

Lesion Level

Spinal-related disability

Above L3

Complete paraplegia and dermatomal para-anesthesia, Bladder incontinence, Nonambulatory

L4 and below

Same as for above L3 except preservation of hip flexors, hip adductors, knee extensors; Ambulatory with aids, bracing orthopedic surgery

S1 and below

Same as for L4 and below except preservation of feet dorsiflexors, and partial preservation of hip extensors and knee flexors; Ambulatory with minimal aids

S3 and below

Normal lower extremity motor function; Saddle anesthesia; Variable bladder-rectal incontinence

How can spina bifida be managed?

- Detection
- Antibiotics
- Surgery
- Careful observation
- Physical therapy



How can spina bifida be managed?

-Detection

-Triple screening

- Maternal blood test for α -fetoprotein

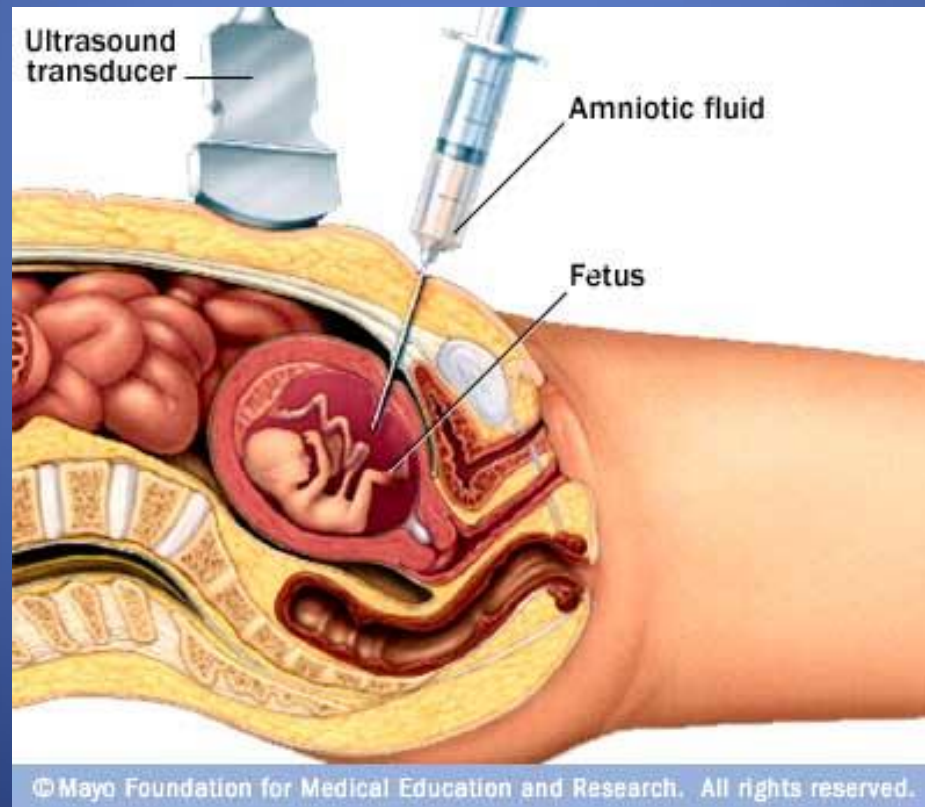
- Ultrasound for bone defects

- Amniocentesis

- α -fetoprotein is elevated in 75-80% of cases of spina bifida (myelomeningocele)

How can spina bifida be managed?

Amniocentesis – using a needle to collect amniotic fluid



How can spina bifida be managed?

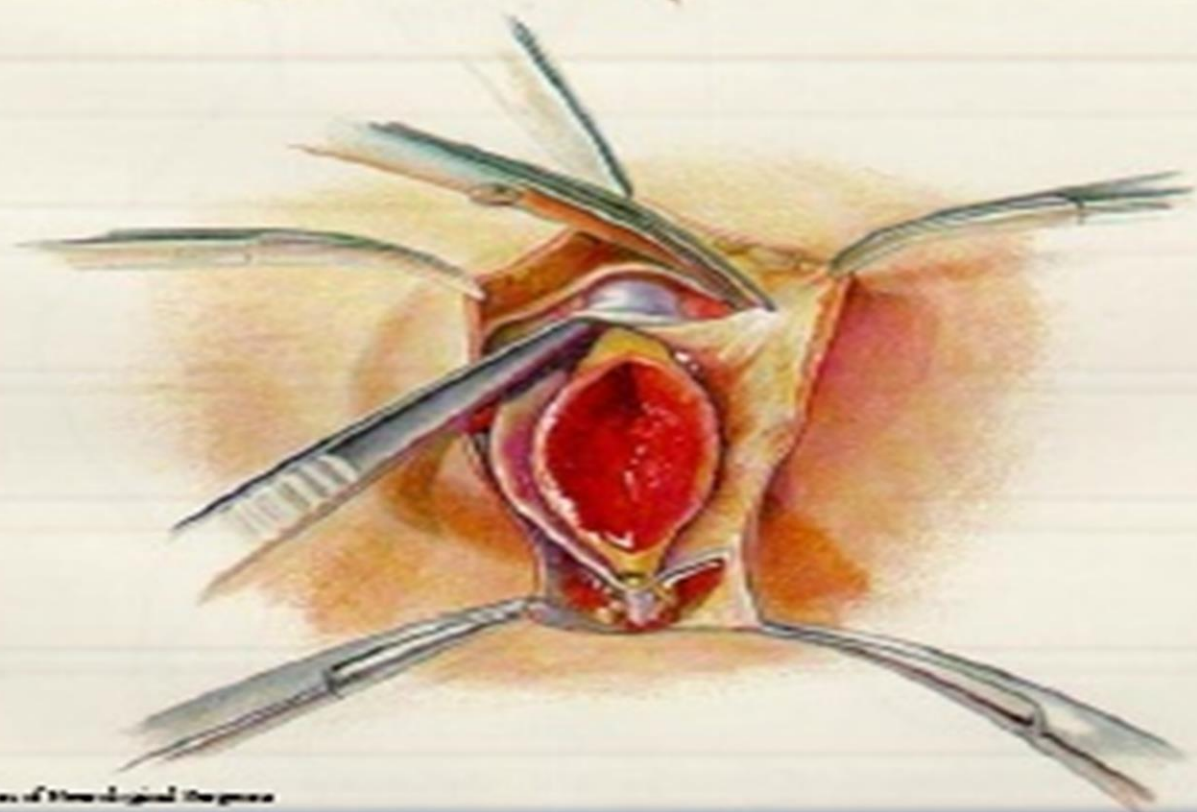
-Antibiotics

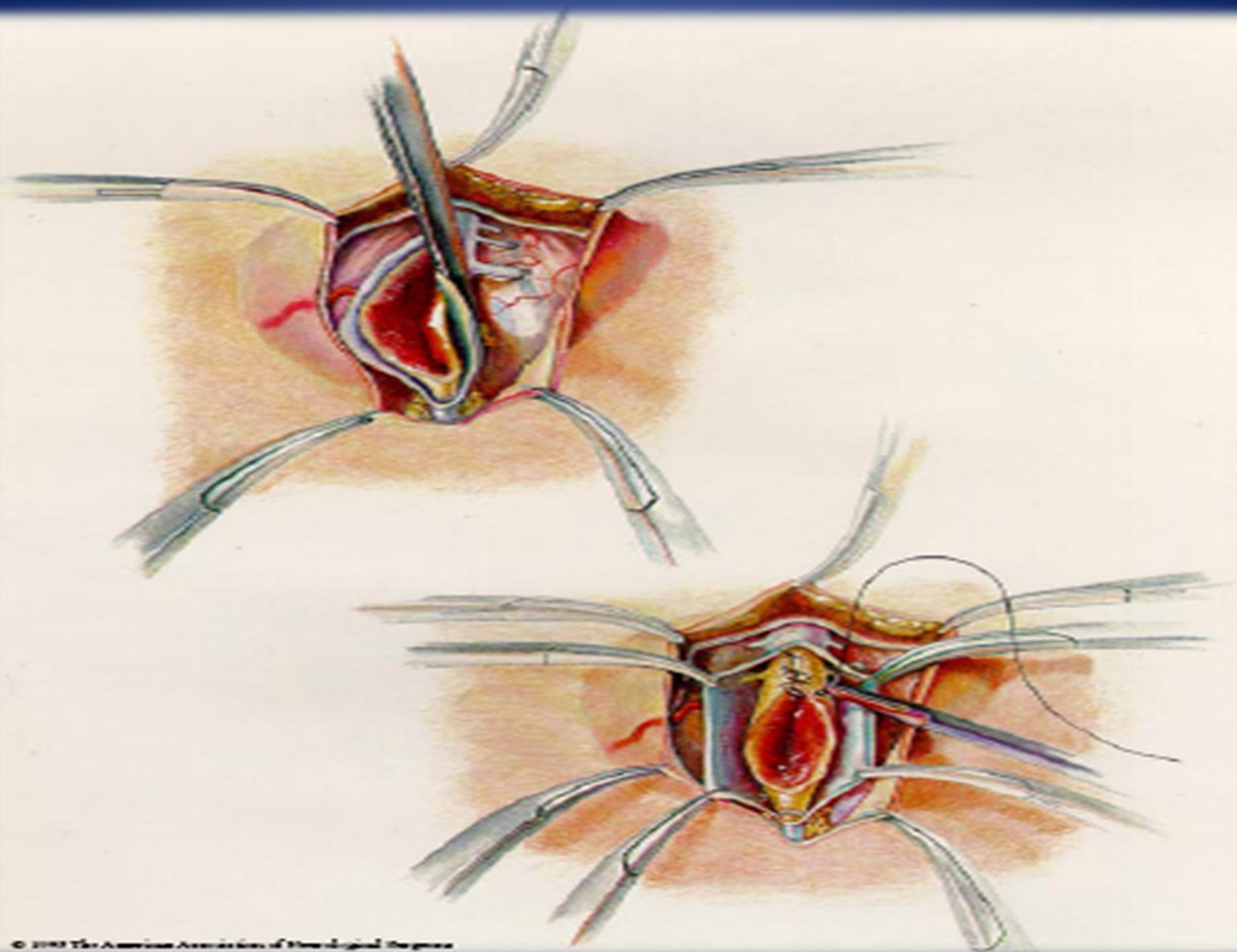
- In some cases the spinal cord is exposed to the environment
- Antibiotics are essential in preventing infection of the CNS

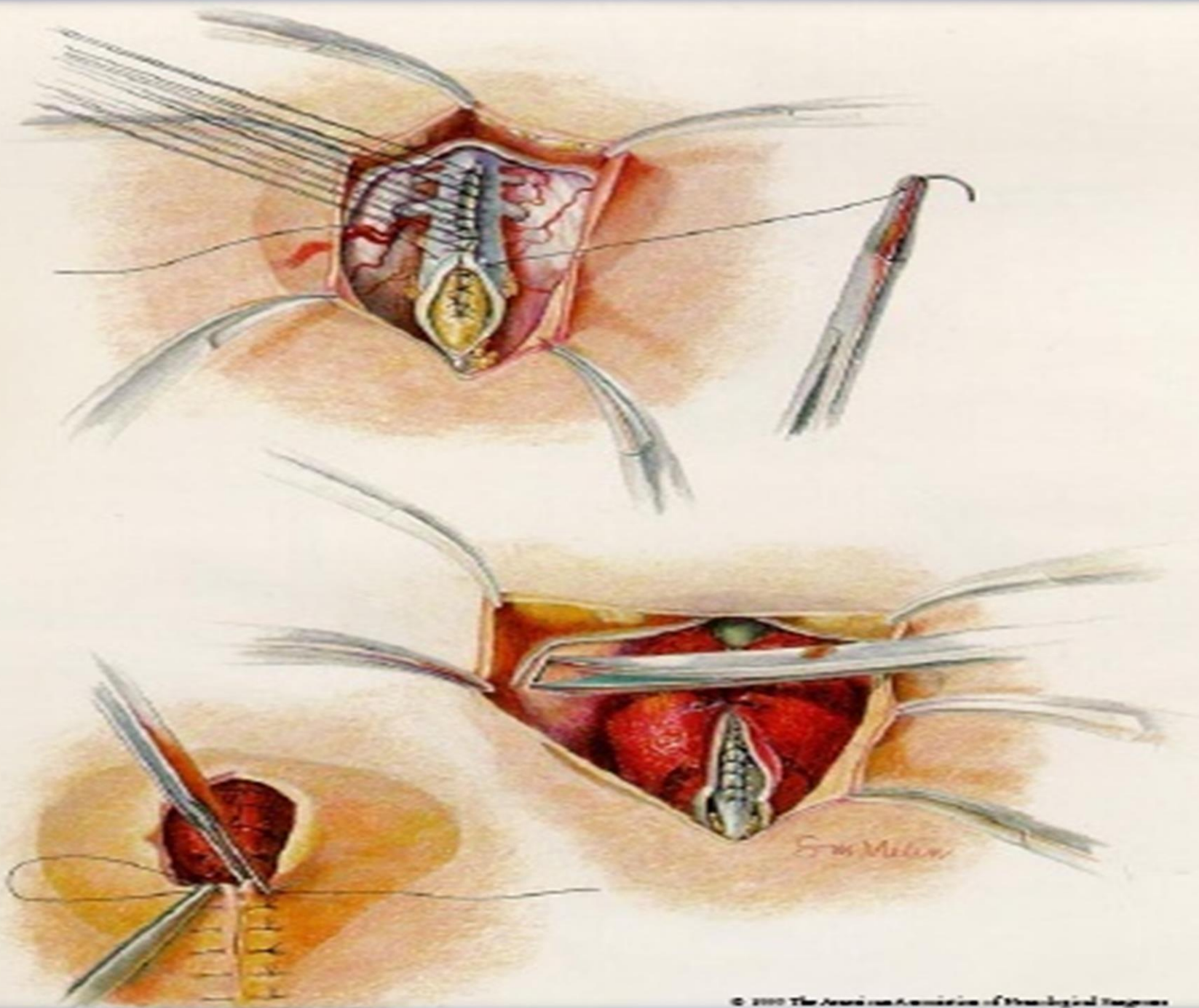
How can spina bifida be managed?

-Surgery

- In some cases the spinal cord is exposed to the environment or tethered
- Surgery is performed in order to cover the spinal cord with muscle and skin or to untether the spinal cord
- *in utero* surgery has also become a viable option for some cases







How can spina bifida be managed?

- Careful Observation

- Children with myelomeningocele often have hydrocephalus (blockage of CSF)
- Children may present with paralysis, blindness, MR, inability to speak, convulsions
- Any changes in mental status or behavior should be quickly brought to the attention of the child's physician(s)

How can spina bifida be managed?

- Physical Therapy

- Spinal cord damage can cause muscle weakening and wasting

- Speech therapy may also be useful for some individuals

Spina Bifida

- What is it?
- What causes it?
- How can it be prevented?
- How can it be managed?
- Social impact**
- Further information

Social impact

Prognosis and any deficits are dependent upon level of involvement

- Estimates from 5-40 % of the world's population may have spina bifida occulta
- Meningocele is not very common and often has minimal impact once the cyst is removed
- Myelomeningocele has the largest impact on patients and their families

Social impact

Changes with time

- Before antibiotics most children with myelomeningocele died because of infections in the CNS; those that survived were unlikely to ever walk
- During the 1990s, the discovery of the role of folate in neural tube closure drastically decreased the number of cases of myelomeningocele
- In the late 1990s, *in utero* surgery was attempted to close neural tube defects

Social impact

Children born with spina bifida today require some special treatment

- Multiple surgeries starting as early as 48 hours after birth
- Physical therapy
- Bowel and/or bladder surgery – helps prevent infection and social stigmatism
- Latex allergies are often present

In many cases, special centers are better equipped to treat children with spina bifida and have a variety of specialists on staff

Social impact

Most children that are treated early will have normal IQ and be able to attend public schools

Mobility is the biggest concern for many patients with spina bifida – lack of mobility can lead to obesity and scoliosis

With proper treatment, individuals will live well into adulthood

