CARE OF THE PATIENT WITH MYELOMENINGOCELE

AKA

SPINA BIFIDA

1

TEAM APPROACH

- Pediatrician
- Pediatric/Neonatal Nurses
- Pediatric Neurosurgeon
- Pediatric Urologist
- Pediatric Orthopedics
- Social Services
- Occasionally- Genetics, Pediatric Neurology, Pediatric PMR, Developmental pediatrics, Radiology, Anesthesia

2

DEFINITIONS

- Spina Bifida Occulta- dimple only
- Meningoceles- Skin covered defect containing only membranes or nonfunctional nerves
  no paralysis
- Lipomyelomeningocele- lipoma in spinal defect connected to neural elements still in canal
- Myelomeningocele- Open spinal cord defect not covered by skin

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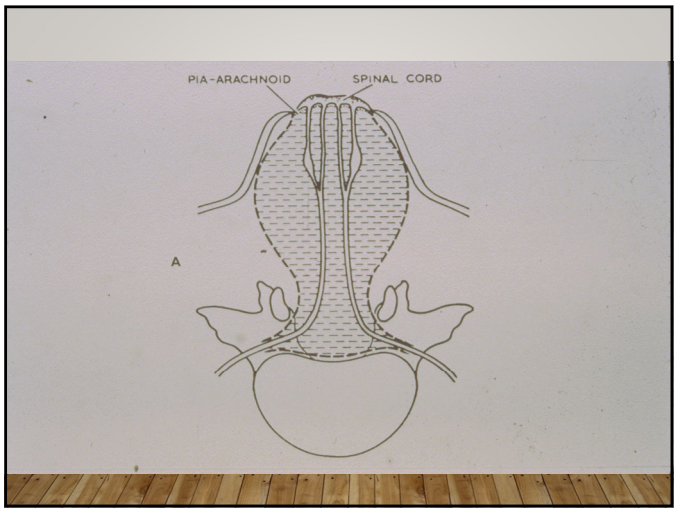
PIA-ARACHNOID
SPINAL CORD
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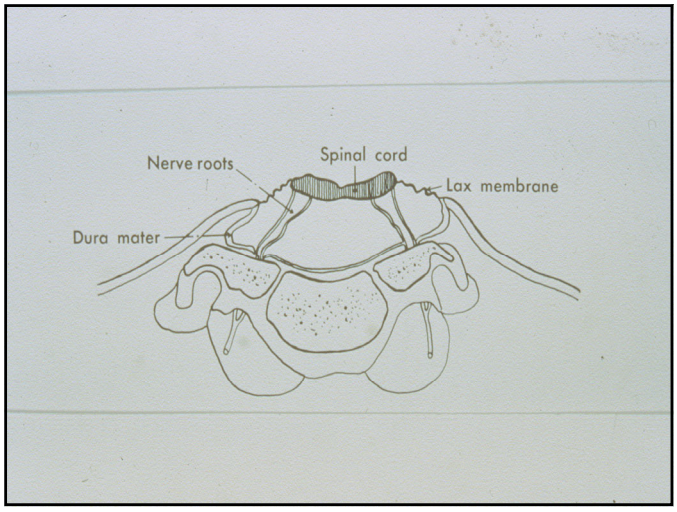
Nerve roots
Spinal cord
Lax membrane
Dura mater

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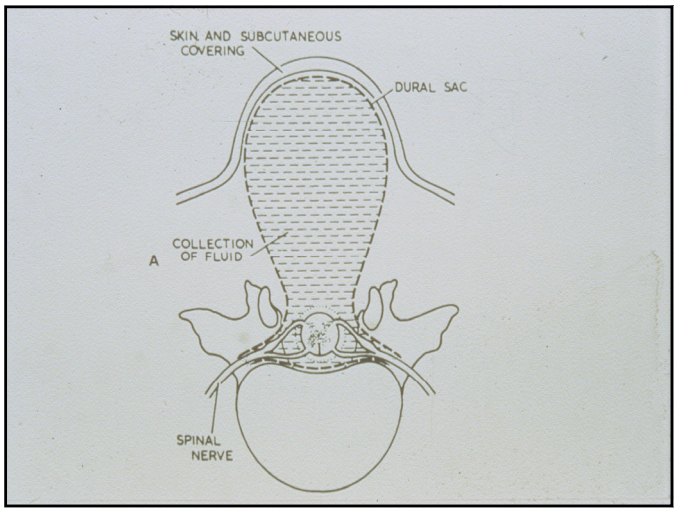

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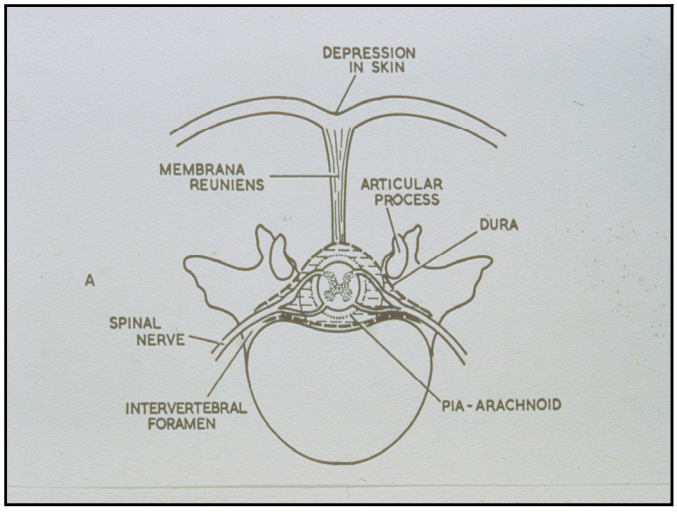

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EMBRYOLOGY

- Location and type of lesion dependent on the timing of a defect in neurulation

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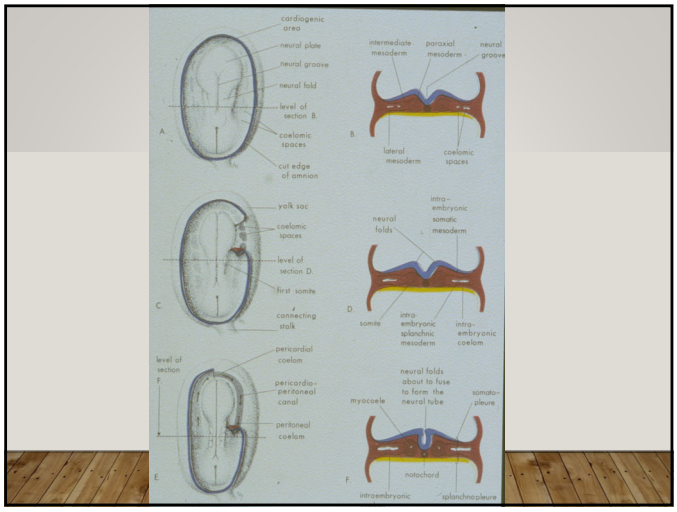

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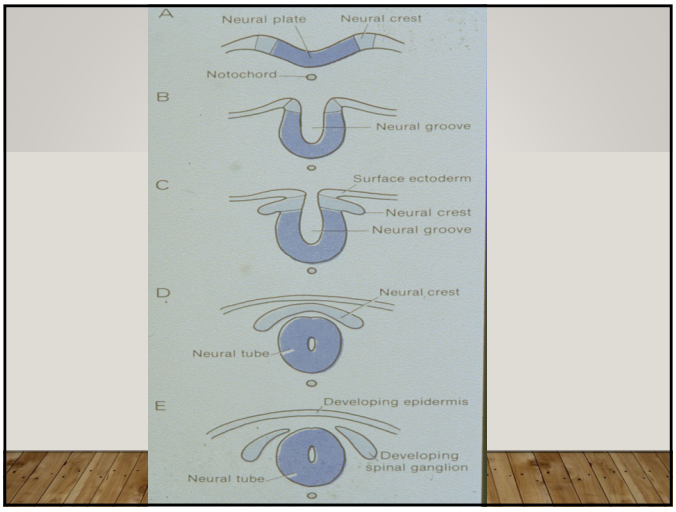

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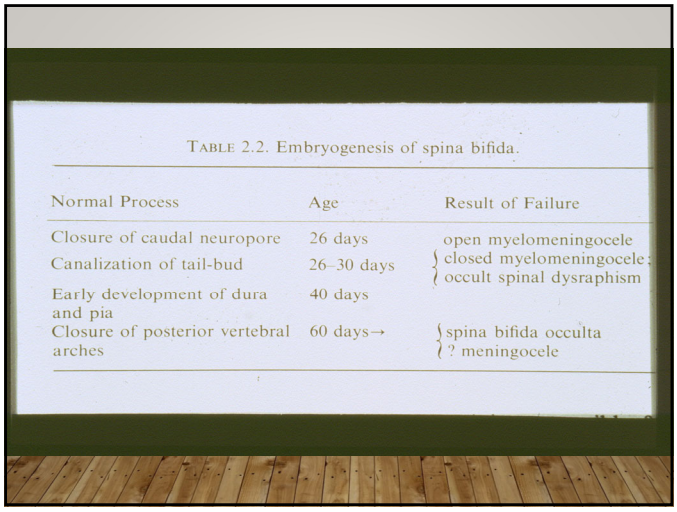

TABLE 2.2. Embryogenesis of spina bifida.

Normal Process	Age	Result of Failure
Closure of caudal neuropore	26 days	open myelomeningocele
Canalization of tail-bud	26–30 days	closed myelomeningocele; occult spinal dysraphism
Early development of dura and pia	40 days	
Closure of posterior vertebral arches	60 days→	spina bifida occulta; ? meningocele

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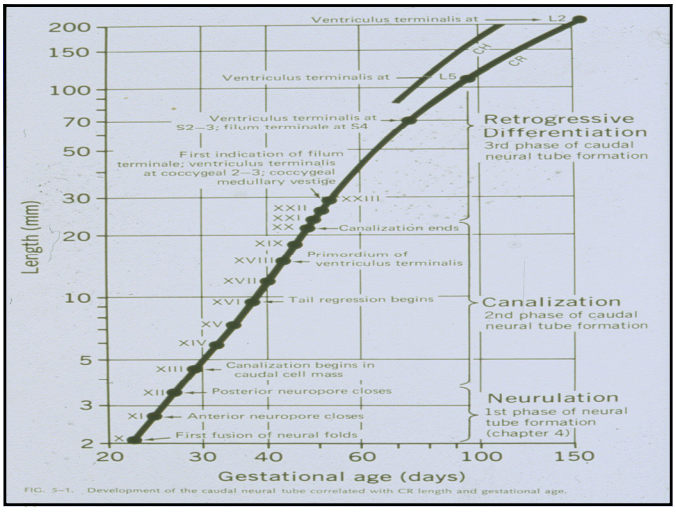

FIG. 5–1. Development of the caudal neural tube correlated with CR length and gestational age.

16

PREVALENCE OF MYELODYSPLASIA

- Previously thought varies by location and race
  - African black-1:10000
  - Celts-1:80
  - U.S.-1:1000
- Now clear that this is affected by folic acid
  - Countries with Mandatory Supplementation
    - 35 per 100 000 live births
  - Countries without Mandatory Supplementation
    - 52 per 100 000 live births

17

SCREENING IN PREGNANCY

- MSAFP- elevated
- 2nd trimester Ultrasound anatomy scans

18

ETIOLOGY OF DEFECT

- Multifactorial
  - Environmental- teratogens-anticonvulsants and ETOH, Obesity, Hyperthermia
  - Vitamins-Deficiency vs. metabolism issues
  - Genetic factors- multiple genes noted in metabolism of folic acid and formation of the spinal cord

19

FOLIC ACID

- B vitamin
  - Must begin before conception(50% unplanned)
  - 400 mcg/day recommended by public health service (most women got 200 or less/day in their diet)
  - Nearly 100% of synthetic form absorbed
    - Dietary form lost in cooking and storage
  - Enriched foods
    - Breads, cereals and rice
    - Since beginning in 1998 there has been a 35% reduction in cases of spina bifida

20

TAKE HOME MESSAGE

- Spina bifida occurs before most mothers are aware of their pregnancy
- Folic acid may (has) markedly reduce the incidence of spina bifida

21

TYPE OF LESION

- Meningomyelocele-75% of lesions noted at birth
- Meningocele-25% of lesions seen at birth

22

LOCATION OF LESION

- Lumbar -42.8%
- Lumbosacral-26.8%
- Thoracolumbar-11.8%
- Sacral-8.4%
- Thoracic-6.7%
- Cervical- 3.5%

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LUMBAR

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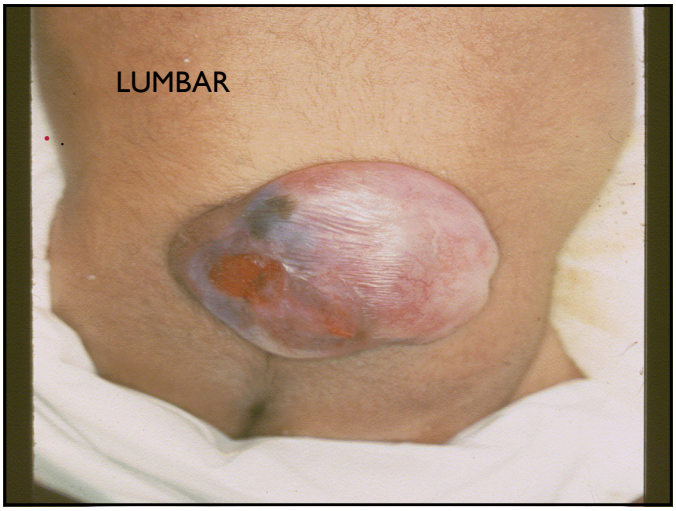

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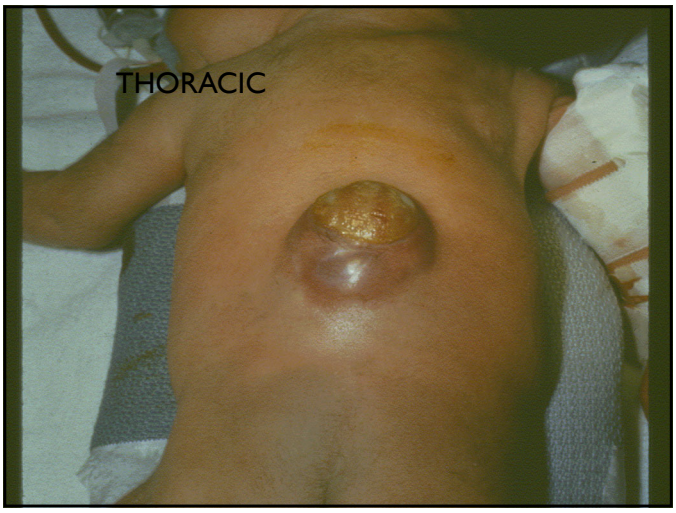

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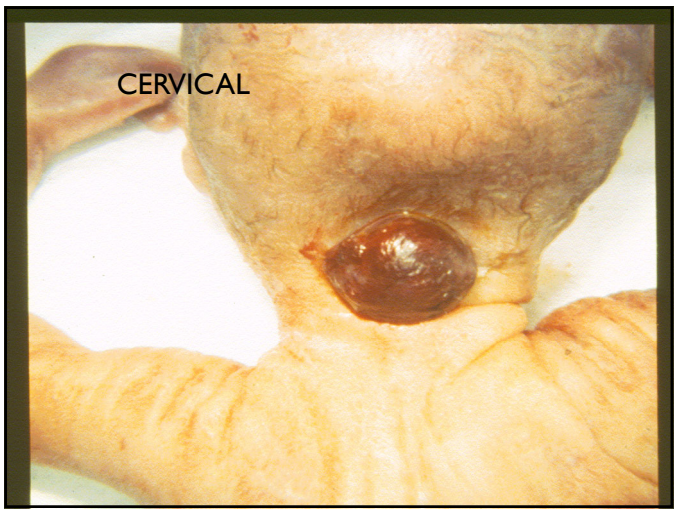

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DELIVERY ROOM MANAGEMENT

- ABC's
- Cover open lesions with sterile cloth soaked in saline.
- Do not use betadine on open neural tissue.
- Funny feet or big head look at the back.

28

TREATMENT AND EVALUATION

- Closure of lesion
- Evaluation
  - CNS
  - Urological
  - Orthopedic
  - GI
  - Cardiac
  - Developmental

29

NEUROSURGICAL CARE

- Closure of lesion – 1st 2-3 days preferred
  - Reduced infection and improved outcome over waiting to repair
- Shunt-as needed consider other options
- Watch for symptomatic Chiari malformation
- Watch for signs of tethering as child grows

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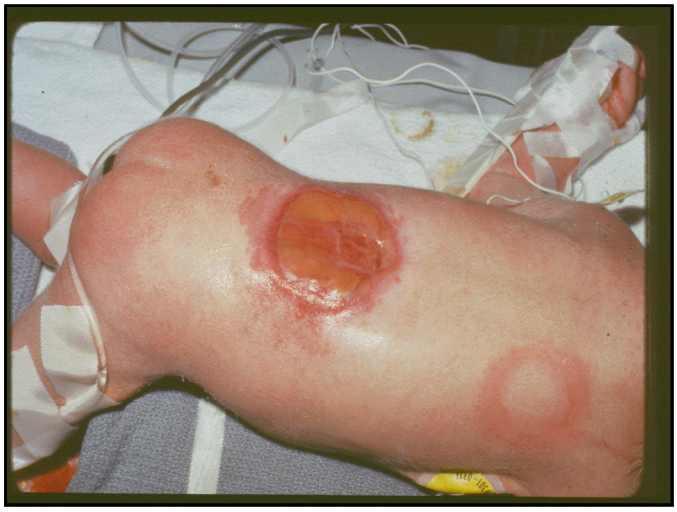

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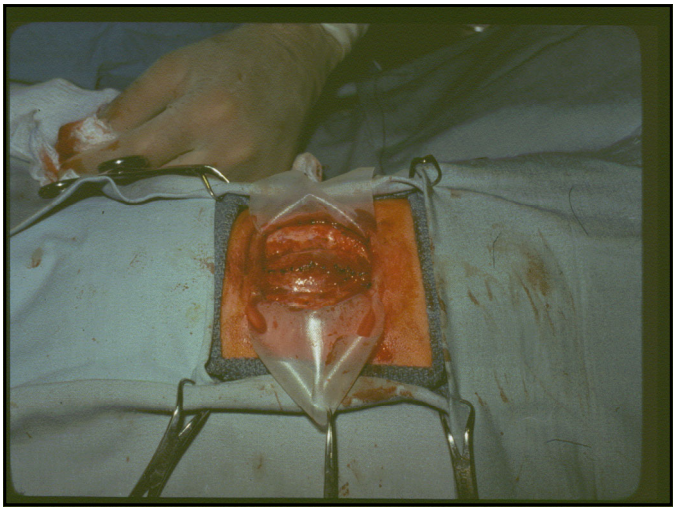

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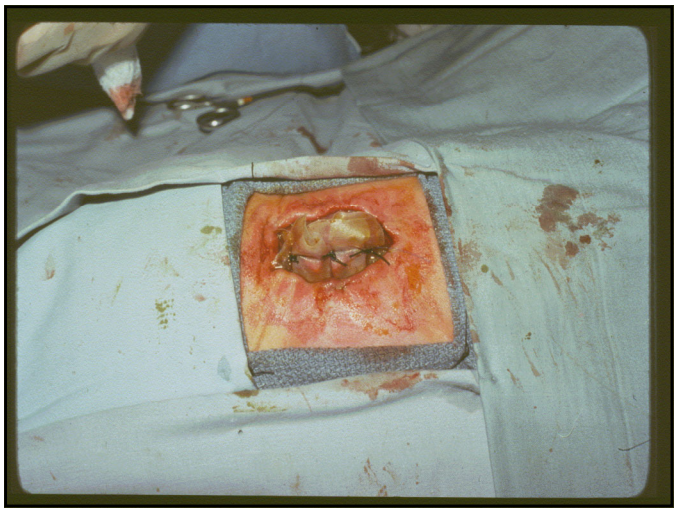

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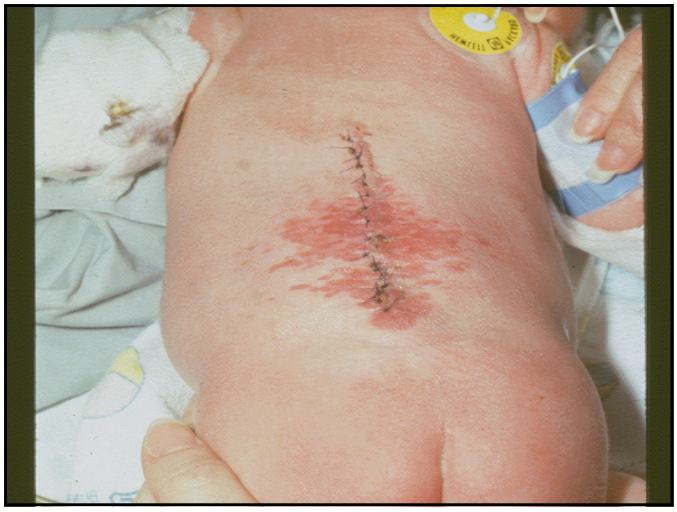

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HYDROCEPHALUS

- Occurs in 80% of cases
- Due to Arnold Chiari type II
- May present with or progress over time

37

ARNOLD-CHIARI TYPE II

- Major Features
  - Caudal displacement of 4th ventricle into cervical canal
  - Medulla and Pons stretched and flexed
  - Caudal displacement of medulla and cerebellum through the foramen Magnum
  - Boney defect of occiput and cervical vertebrae

38

ARNOLD CHIARI TYPE II

- May be symptomatic
  - Respiratory drive problem
  - Vocal cord paralysis
  - Loss of gag/airway protection
  - LEADING CAUSE OF DEATH

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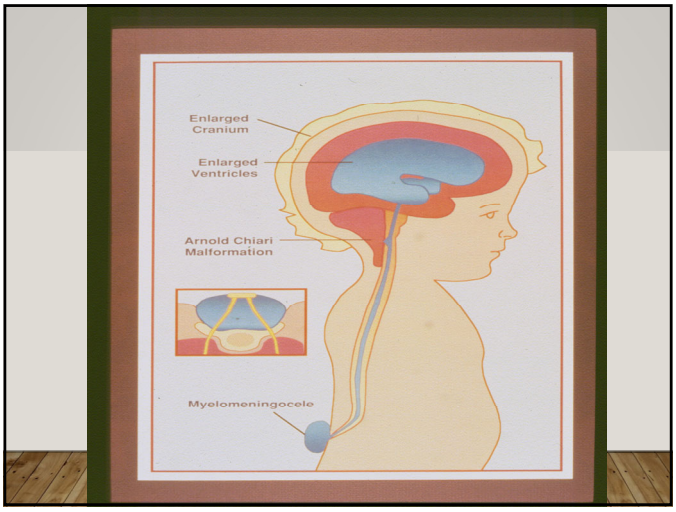

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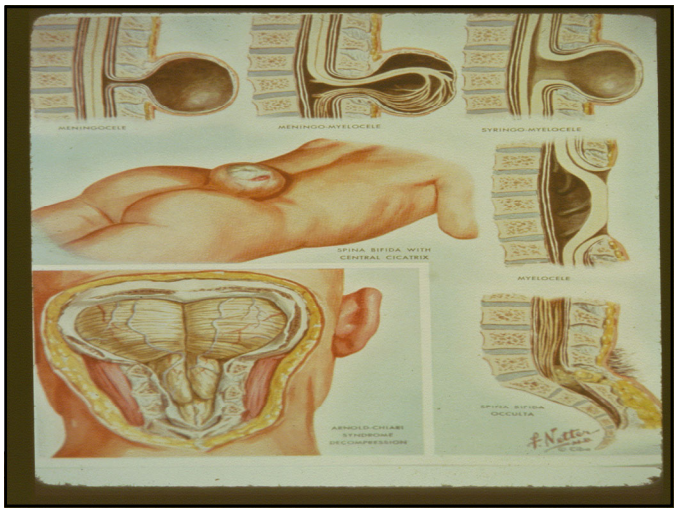

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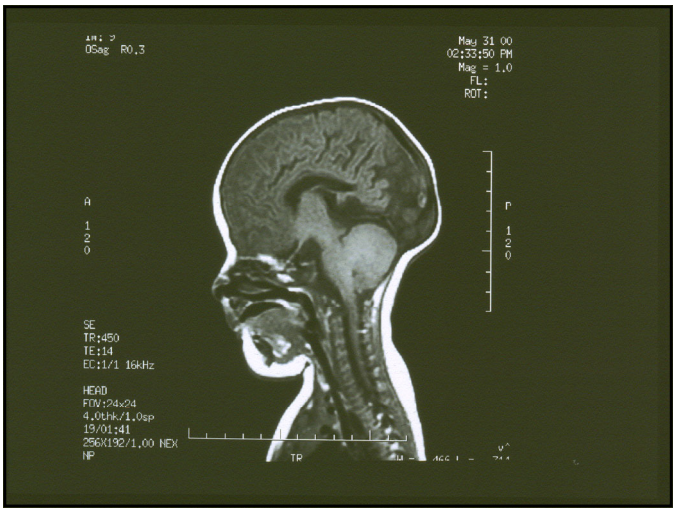

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VP SHUNTS

- 2 main types
  - Fixed valves shunts
    - Fluid flows if reaches certain pressure
  - Programable valves
    - Able to adjust with handheld device
    - Are affected by MRI devices
      - Must be reset after MRI

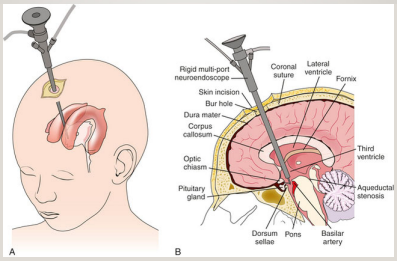
43

NEWER OPTIONS TO SHUNTING

- ETV with/without CPC
  - ETV- endoscopic third ventriculostomy
    - Allows for flow of CSF out of the ventricle
    - Best done when dilated to get scope in to do this
    - Use of laser to puncture the floor of the 3rd ventricle
  - CPC- choroid plexus cauterization
    - Done in conjunction with ETV to limit amount of further CSF production and hopefully keep hydrocephalus from reoccurring. At baseline choroid plexus makes up to 1 oz /hours of CSF.
    - Uses laser to cauterize in 1st and 2nd ventricle
  - Overall success rate 41% in infants

44

ENDOSCOPIC THIRD VENTRICULOSTOMY



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TAKE HOME MESSAGE

- Spina bifida patient with stridor, difficulty feeding, apnea, hoarse cry, or vomiting….Assume symptomatic Chiari and likely hydrocephalus.
- Leading causes of death in under 5 yo has not changed in more than 25 years…symptomatic Chiari/ Urosepsis.
- Leading cause of death in older patients Urologic disorders not hydrocephalus.

46

UROLOGIC ISSUES

- Neurogenic bladder – 90 % of patients
- Complications can include
  - Reflux
  - Hydronephrosis
  - UTI's
  - Stones
  - Social issues

47

UROLOGIC EVALUATION

- Post void residual - <5 cc normal
  - Test after spinal shock resolved.
- Labs- UA, urine culture, BUN, Creatinine
- Imaging- Renal and Bladder U/S, VCUG, Nuclear med scan (Lasix), MRI (when scanning spine helpful)
- Testing – FUDS

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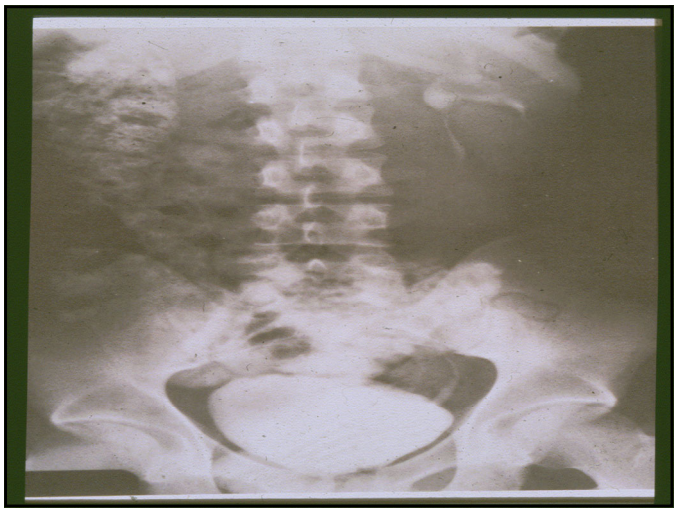


49

GOALS OF TREATMENT

- Preserve renal function
- Treat urinary incontinence
- Allow independence

50

NEUROGENIC BLADDER

- 4 types

Sphincter tight **Flaccid Bladder**	**Sphincter tight** **Spastic bladder**
Sphincter loose **Flaccid bladder**	**Sphincter loose** **Spastic Bladder**

51

UROLOGIC THERAPY

- Catheter-CIC
  - + reduces infections and pressure in the bladder
  - -Time intensive and not patients all are capable
- Anticholinergics-Ditropan
  - - Side effects variable anticholinergic
  - + a means to relax the bladder and the pressure
- Botox
  - + relax a tight sphincter
  - - has to be repeated

52

SURGICAL OPTIONS

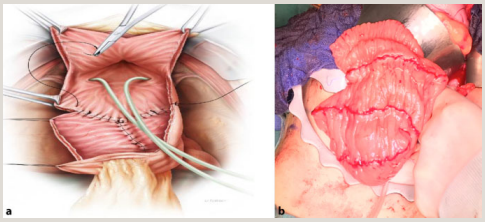
- Vesicostomy
  - - Constant leakage/irritation
  - + does relieve high pressure situations
- Bladder Augmentation
  - + increase bladder capacity to not leaking urine and keep pressure low
  - -Foul urine, stone formation
- Mitrofanoff
  - + means to independence. Patient can cath self and or easier for care giver
  - -Complicated to do, tend to try to close over time

53

VESICOSTOMY

54

BLADDER AUGMENTATION



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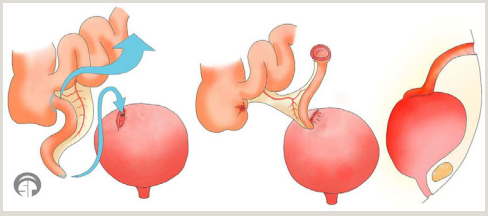
MITROFANOFF



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COMPLICATIONS

- Hydronephrosis
- Reflux
- Scarring of kidney
- Incontinence
- Infection
- Stone formation
- Hypertension

57

TAKE HOME MESSAGE

- UTI is common but if CIC only check urine if
  - Foul smelling
  - Fever
  - Ill patient
- Routine Urine cx in CIC patients will grow bacteria every time.
- Use what ever means to gets the urine out and protects the childs renal function.
- Avoid antibiotics when able.

58

GI COMPLICATIONS

- Constipation- Neurogenic bowel
- Fecal incontinence
  - Huge concern for preteens and older
- Associated Issues
  - Malrotation, omphalocele, imperforate anus
  - Inguinal hernia

59

BOWEL MANAGEMENT

- Multiple ways to manage.
- The better the bowel program the better the quality of life and fewer UTIs.
- Goals similar in all cases- managing stool burden as well as timing of removal
- Most common
  - Suppositories
  - Stool softeners
  - Enemas- Retrograde and antigrade

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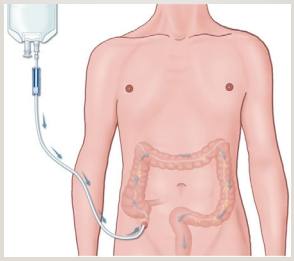
ACE (MACE)
(ANTEGRADE CONTINENCE ENEMAS)



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RETROGRADE ENEMA

62

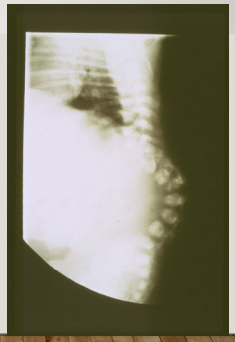
ORTHOPEDICS

- Spine- Scoliosis
  - Multiple causes
- Extremeties- Gait
  - Hips
  - Knees
  - Feet

63

CAUSES OF ORTHOPEDIC DEFORMITIES

- Paralysis- thoracic lesion
- Unopposed muscle action- club feet, dislocation of hips
- Congenital malformations- Hemi vertebrae



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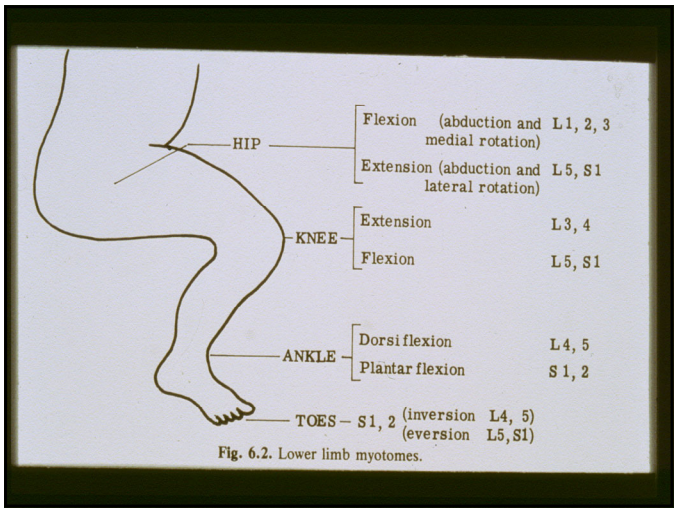

Fig. 6.2. Lower limb myotomes.

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Table 9-1 Joint Deformities at Birth

Last intact root	Hips	Knees	Feet
L_1	Flexion and external rotation	Completely paralyzed. Deformities are dependent on the intrauterine position	Completely paralyzed. Deformities are dependent on the intrauterine position
L_{2-3}	Moderate flexion and adduction	Extension or slight flexion	Dependent on intrauterine position
L_4	Flexion, abduction, external rotation	Extension or recurvatum	Calcaneovarus
L_5	Moderate flexion	Extension or slight flexion	Marked calcaneus
S_1	No deformity	No deformity	Calcaneovalgus; rocker bottom, foot convex pes planus, clawing of toes
S_2	No deformity	No deformity	Cavovarus and clawing of toes

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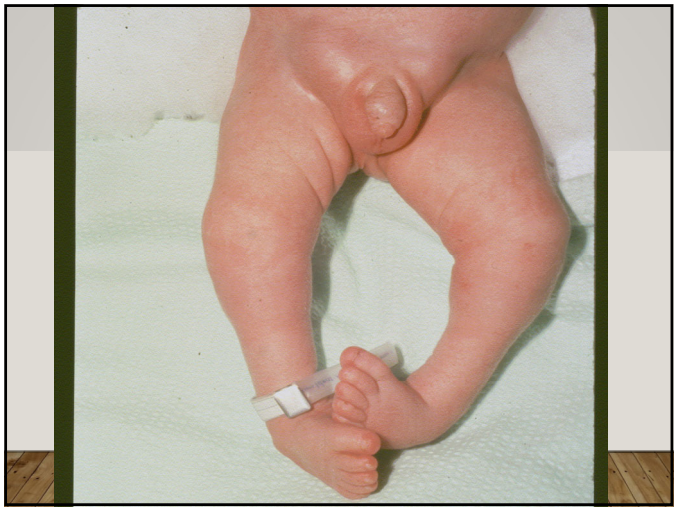

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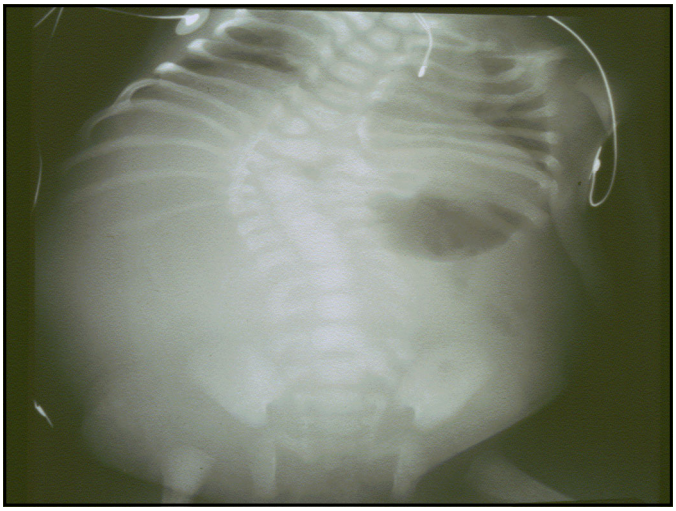

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71

GOALS OF TREATMENT

- Keep joints mobile and pain free
- Maintain truncal balance and stability- spine
- Prevent ulcers
- Maximize opportunity for ambulation.

72

ORTHOPEDIC TREATMENT

- Spinal Fusions for scoliosis- anterior and posterior
- Osteotomies
- Tendon and/or muscle transfers
- Contracture releases
- Braces as needed to assist ambulation

73

NEW TECHNOLOGIES IN ORTHO

- Casting and tenotomies – Club feet (Ponseti)
  - Able to do most in outpatient area
  - Braces for lower extremities common
- Scoliosis- Occurs in 50% of patients with Spina Bifida
  - Treatments
    - Braces- challenging
    - Surgery
      - Fusion in skeletally mature child
      - Growing Rods vs MAGEC rods in younger children

74

PONSETI FOR CLUB FEET

75

AMBULATION PROGNOSIS BASE ON LEVEL OF LESION

- T12 level- No lower limb control– wheelchair bound
- L1 level- Pelvic movement only- Exercise ambulation
- L 3 level- Hip movement – Household ambulation
- L4 level- Knee movement- Community ambulation

76

SENSORY DEPRIVATION

- Dependent on level of lesion
- IV issues
- Pressure sores
  - Heels, coccyx, back, surgical sites
- Fractures
  - Swelling, fever redness
- Burns- water mostly

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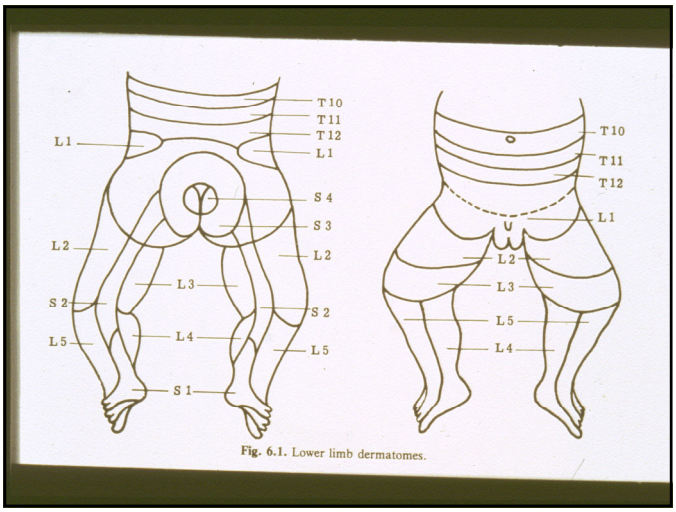

Fig. 6.1. Lower limb dermatomes.

78

TAKE HOME MESSAGE

- After initial repair most surgery is orthopedic
- Fever and swelling of limb think fracture
- Ambulation best if below than L3 level
- Sensation lacking but variable
  - No IV in lower limbs
  - Protect and cover as able

79

OTHER ORGAN SYSTEMS

- Cardiac- ASD, VSD, PDA, Coarctation
- Dev. Delay/ seizures-structural anomalies of brain
- School-spacial orientation, hand eye coordination, ADD
- Short Stature
- Premature pubertal dev- Females with Hydrocephalus most common.

80

LATEX ALLERGY

- Previously common in Spina Bifida-28-67%
  - Seem predisposed…why?
  - Seen much less now than 20 years ago as now limited latex exposure
- Health care workers-2.6-16%
- General population-1%
- VATER, imperforate anus, Cerebral Palsy, etc.

81

LATEX PRECAUTIONS

- Avoid medical use
- Avoid exposure in environment- less of issue in current times as not sensitized.
- Rare cross reactivity with foods
  - Kiwi, Bananas, chestnuts, avacado

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TETHERED CORD

- All myelo patients s/p repair
- Lipomyelomeningoceles
- Occult spinal lesions

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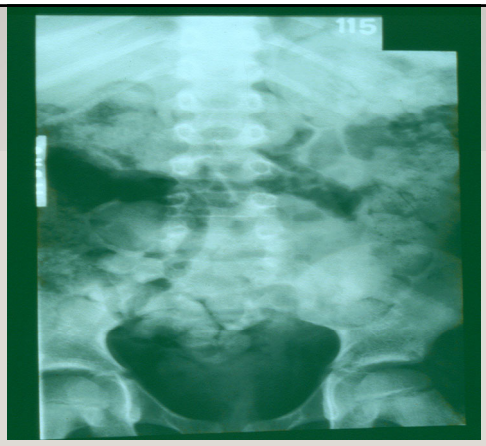

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OCCULT SPINAL DYSRAPHISMS

- Hypertrichosis
- Dimples
- Lipomas
- Hemangiomas
- Dermoid cyst or tract
- Cutis aplasia or scar

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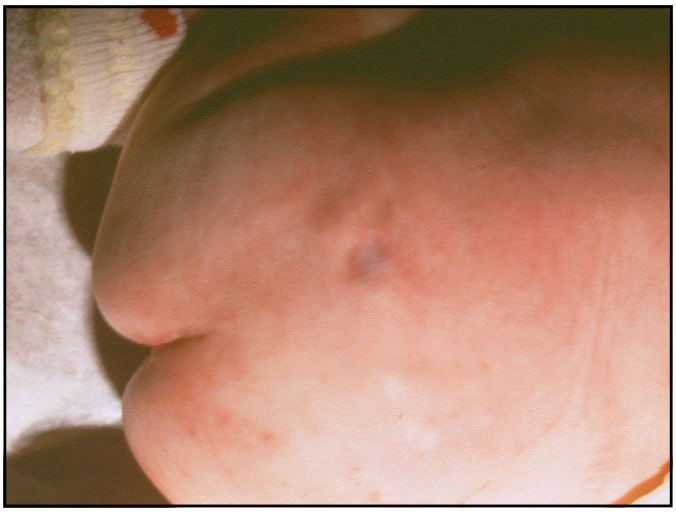

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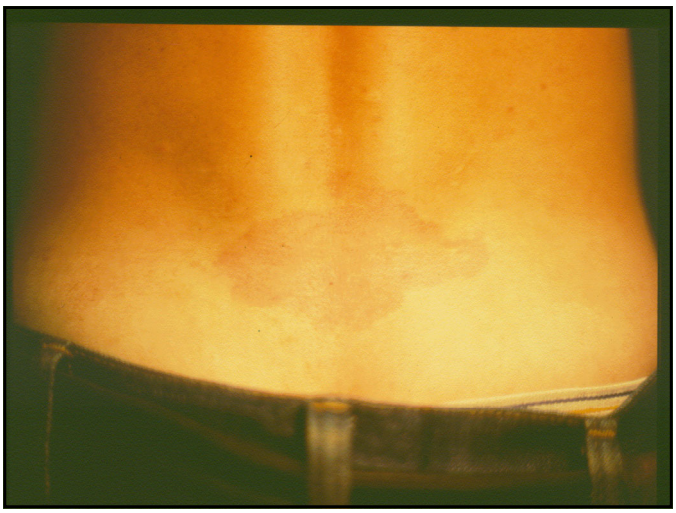
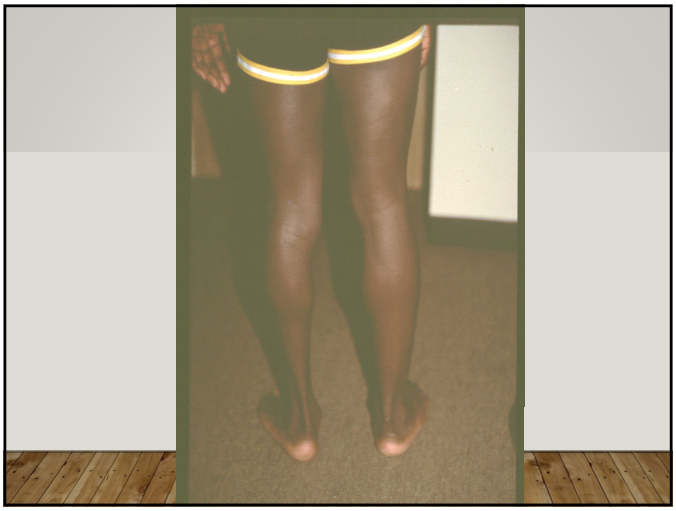


92

TETHERING SYNDROME

- Gait Abn.
  - Weakness
  - Contractures
  - Scoliosis
- Urinary / Bowel symptoms
  - Incontinence, UTI's, Changes form baseline

93

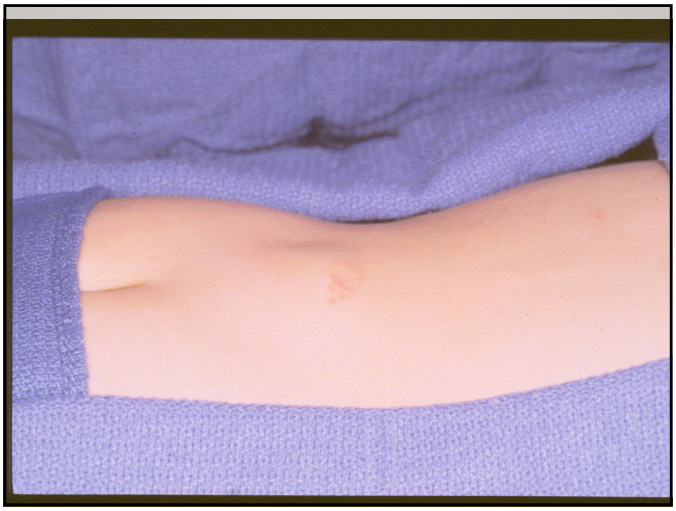


94

TREATMENT FOR TETHERING

- Eval based on age and symptoms
  - U/S may help in infant
  - FUDS- can be normal in <18 mo
  - MRI- GOLD STANDARD
- Treatment surgery
  - Optimal <6 mos but can reverse symptoms up to 18 mos.
  - Must follow for retethering

95



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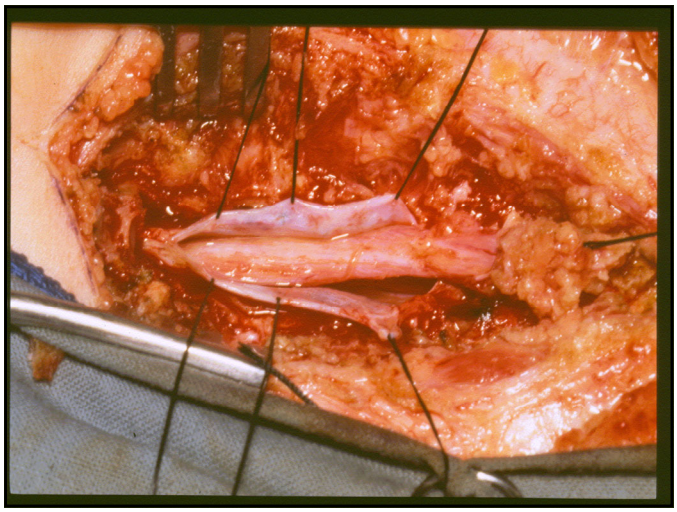

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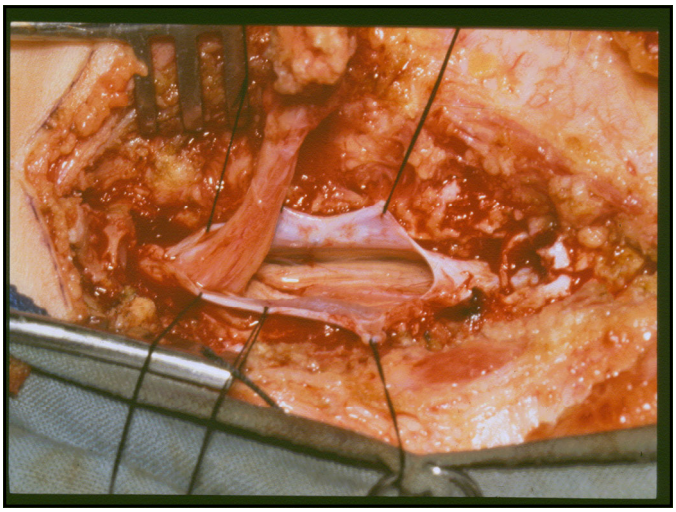

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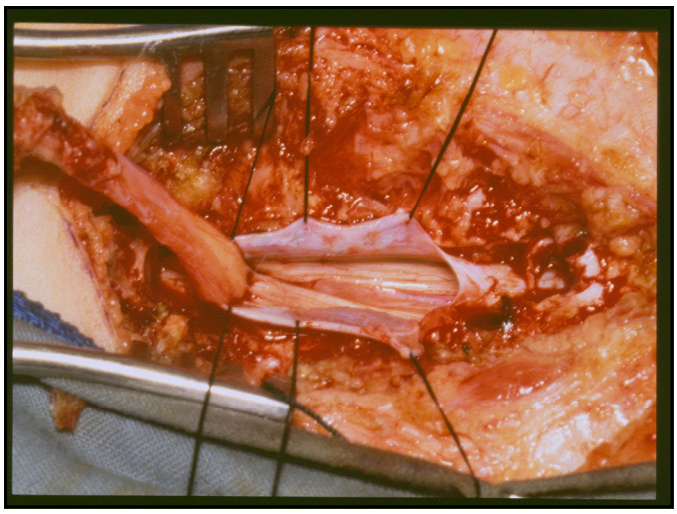


99

IN UTERO REPAIR

- First attempted in 1990s via endoscopic approach
- First successful open repair in 1998
- Management of Myelominigocele Study (MOMS)

100

101

ADZICK ET AL: A RANDOMIZED TRIAL OF PRENATAL VERSUS POSTNATAL REPAIR OF MYELOMENINGOCELE. NEJM 2/2011

- MOMS trial (2003-2010)
  - NICHHD randomized trial comparing prenatal vs postnatal repair
  - Repair at one of three centers
  - Plan for 200 participants (183 enrolled)
  - Primary outcome of fetal or neonatal death or need for shunt at 12 months
  - Trial discontinued early secondary to efficacy

102

ADZICK ET AL: A RANDOMIZED TRIAL OF PRENATAL VERSUS POSTNATAL REPAIR OF MYELOMENINGOCELE. NEJM 2/2011

- MOMS trial
  - Primary outcome of need for shunt
    - Postnatal Repair: 98% vs Prenatal Repair: 68%
    - Actual shunt rate: 82% vs 40% (RR of 0.48)
  - Primary outcome of mental development and motor function also significantly better
    - Bayley Psychomotor Developmental Index significantly higher in prenatal repair group
    - 42% in the prenatal repair vs 21% in the postnatal repair were walking independently at 30 months

103

ADZICK ET AL: A RANDOMIZED TRIAL OF PRENATAL VERSUS POSTNATAL REPAIR OF MYELOMENINGOCELE. NEJM 2/2011

- MOMS trial risks
  - Preterm delivery
    - Postnatal Repair: 37.3 weeks vs Prenatal Repair: 34.1 weeks
    - 3039 g vs 2383 g
    - 46% deliver prior to 34 weeks
    - 13% deliver prior to 30 weeks
  - Uterine dehiscence
    - 25% show a very thin hysterotomy site at delivery
    - 10% have area of dehiscence or complete dehiscence

104

ADZICK ET AL: A RANDOMIZED TRIAL OF PRENATAL VERSUS POSTNATAL REPAIR OF MYELOMENINGOCELE. NEJM 2/2011

- Both prenatal repair and postnatal repair groups had 2 deaths
  - Prenatal repair group had 1 in utero demise and one 23 week delivery death. Both occurred within 5 days of the surgery.
  - Postnatal repair group had 2 patients die both of a symptomatic Chiari malformation despite shunting.

105

MOMS TRIAL FOLLOW UP AGE 5.9 -10.3 YEARS

- More prenatal patients walked without orthotics or assisted devices
  - 29% vs 11%
- Lower rate of hind brain herniation
  - 60% vs 87%
- Fewer shunts for hydrocephalus
  - 49% vs 85%
  - fewer revisions 47% vs 70 %
- No improved cognitive function

106

OUR EXPERIENCE

107

PATIENT #1

108

MILD VENTRICULOMEGALY

109

STEPS

- Amniocentesis performed
  - Positive acetylcholinesterase
  - Karyotype normal
- MRI confirms findings

110

PRE-OPERATIVE EVALUATION

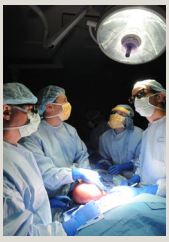
- Prospective surgery work up includes:
  - MFM consultation
    - Detailed ultrasound, amniocentesis, MRI
  - Pediatrics consultation
  - Neurosurgery consultation
  - Ob Anesthesia consultation
  - Pediatric Cardiology consultation
  - Social worker evaluation

111

OR TEAM

LARGE NUMBER OF FOLKS

EVERYONE GETS THEIR TURN



112

OR TEAM

- Obstetrical Anesthesia (2)
- Maternal Fetal Medicine (2)
- Pediatric Cardiologist (2)
- Pediatric Surgeon (1)
- Pediatric Neurosurgeon (1)
- Pediatric Plastic Surgeon (1)
- Scrub tech (2), Circulators (2), OB nurse (1)
- On-lookers (minimum of 6)

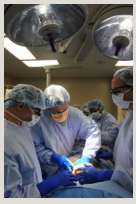
113

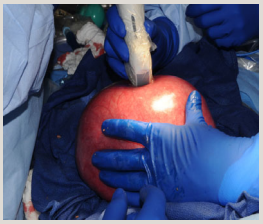
PROCEDURE DETAILS

- Anesthesia
- Abdominal incision
- Uterine incision
- Fetal repair
- Uterine Repair
- Post-Op care

114

UTERUS "EXTERIORIZED"



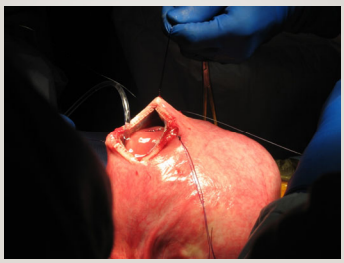


115

PROCEDURE
STAY SUTURES AND HYSTEROTOMY

116

HYSTEROTOMY WITH STAY SUTURES

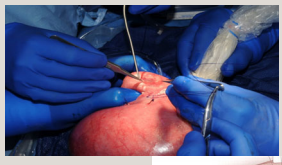


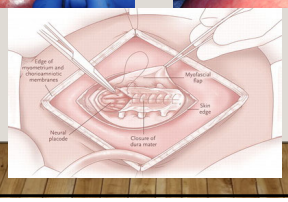
117

FETAL EXTRACTION

118

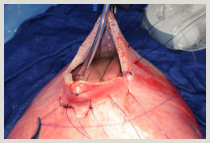
FETAL REPAIR





119

UTERINE REPAIR



120

RETURNING THE UTERUS

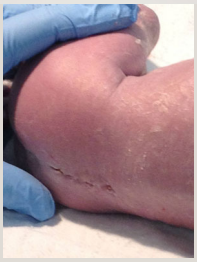
121

POST OP

- Post operative care
  - ICU admit
  - Fluid restriction
  - Aggressive tocolysis
  - Pain control
- Prolonged tocolysis with bedrest
- Ultrasound surveillance
- Delivery by c-section at 37 weeks

122

PATIENT 1
DELIVERY AT 35 6/7 DUE TO PPROM



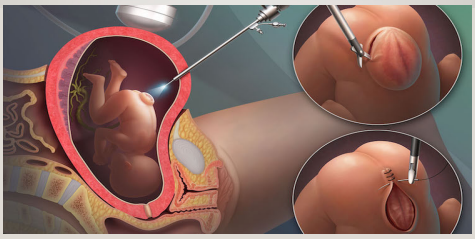
123

CASES DONE HERE

- 9 other cases.
- 4 shunt placed out of the 9
- No longer done here
- Most done at specific centers.
  - Closest to us Texas Children's, Vanderbilt
  - Others we have seen were done at Children's Hospital of Philadelphia

124

FETOSCOPY SURGICAL REPAIR



125

FETAL SURGERY OUTCOMES

- No C section required.
- 50% deliver vaginally
- Average gestation 37-38 weeks
- Rate of treatment for hydrocephalus needed for both types of fetal surgery 37%

126

LIFE EXPECTANCY

- Mid 20th century- 80 % died in 1st year of life.
- 1980's – 14-18% alive at 5 years
- Today mean age expectancy 40 years

127

TAKE HOME MESSAGE

- If midline lesion on back above coccyx evaluate
- Myelo infant with stridor, feeding problems, tongue fasiculations, hoarse cry- Chiari is symptomatic
- Known patient with headache, vomiting , altered LOC - think hydrocephalus
- Fever without obvious source
  - Shunt, UTI, fractures.

128

TAKE HOME MESSAGE

- Don't check routine urine if CIC
- Keep bowels moving
- Changes in bowel, urinary, gait, scoliosis – think tethered cord
- Most spina bifida patients have normal intelligence

129

OUTCOMES

- Infant mortality rates with Spina Bifida
  - US/ Canada was 23/100,000 live birth in 1981-83
  - US/ Canada now 2.3/100,000 live birth 1990-2007
    - PLoS One. 2021; 16(5): e0250098.
    - Published online 2021 May 12.

130

CHALLENGES

- Expertise- most all in pediatric providers
- Insurance
  - All can get Medicaid with waiver
  - Families Helping Families- Great support group
  - Limited options as adult for health care

131