Spinal Dysraphism Imaging with Postnatal Correlation: Where Are We Now?

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Disclosures

None
Learning Objectives

• Understand the classification of spinal dysraphism.
• Discuss advances in prenatal US/MR in the evaluation of spine lesions.
• Review the criteria for fetal meningomyelocele surgery.
Embryology: Neurulation

Neural plate begins to develop at 3rd gestational week.

Lateral edges thicken forming the neural folds.
Conus Medullaris

L2-3 or higher

Tethering may be due to

- Failed migration of mesodermal tissue over neural tube
- Failure of neurulation/migration of mesodermal structures
Spinal Dysraphism

term used to describe nearly all spinal anomalies

**Dysraphism** = Defect of closure of neural tube (primary neurulation)

- Tubulation of neural plate
- Separation from ectodermal elements
- Disjunction of superficial from neural ectoderm
Spinal Dysraphism

Open

Closed

Subcutaneous mass?

YES

NO

Simple

Complex

Tortori-Donati, Rossi, 2000 Neurorad
Spinal dysraphism

Open (ONTD)
- Myelomeningocele
- Myelocele

Closed (CNTD)
Subcutaneous mass?
- Yes
  - Meningocele
  - Lipomyelomeningocele
  - Lipomyeloschisis
  - Myelocystocele
- No
  - Simple
    - Spina bifida
    - Lipoma
    - Tethered cord
    - Persistent terminal ventricle
  - Complex
    - Split cord malformation
    - Caudal regression
    - Neurenteric cyst

A
- Spinal cord
- Skin
- Meninges

B
- Spinal cord
- Vertebra

C
- Spinal cord
Open Spinal Dysraphism

OSD /ONTD - 30%

translucent membrane or neural elements exposed through bone defect

• **NO SKIN COVERING**

Myelomeningocele
Closed Spinal Dysraphism

CSD/CNTD - 60%

- mass
- hemangioma
- hairy patch
- or NO mass

+ Skin covered

Spina bifida occulta  Meningocele
Open Spinal Dysraphism

Classified according to the position of placode w/ respect to skin surface

**Myelomeningocele:**
Placode elevated by expansion of SAS  Covered by a membrane

Courtesy Heron Werner
Open Spinal Dysraphism

**Myeloschisis/Myelocele:**
Plaque of neural tissue lies exposed at same plane as skin

*Courtesy Heron Werner*
Open Spinal Dysraphism

Associated with **Chiari II malformation**

- Rhombencephalic vesicle does not distend
- Hindbrain herniates due to CSF leak at level of spinal defect
Closed Spinal Dysraphism

Mass

**Meningocele:**

Extension of dura/ arachnoid through spinal bifida (no neural tissue in sac)

/+ - tethered/hydromyelia

**Skin Covered**

**No Chiari malformation**
Closed Spinal Dysraphism
No mass
Tethered Cord
Closed Spinal Dysraphism
Mass

**Lipomyelomeningocele**
premature disjunction of cutaneous ectoderm allows mesenchyme to be induced to become fat which interferes w/ neurulation

**Lipomyeloschisis** – flat

*Skin Covered*  
*No Chiari malformation*
Closed Spinal Dysraphism Mass

Terminal Myelocystocele:

- Dilated terminal ventricle herniates through spina bifida
- Assoc cloacal anomalies
- May have herniated posterior fossa in third trimester.
Closed Spinal Dysraphism
No mass

Lipoma
Tethered Cord
BEWARE!!

Prenatal differentiation between CNTD and ONTD may be difficult at times

Hindbrain herniation may be minimal with a low MMC in second trimester

Hindbrain herniation may develop in the third trimester w/ terminal myelocystocele

*Husler et al  Prenat Dx 2009;29*
Open Neural Tube Defects

MMC 0.5-1/1000 pregnancies

Prevention - Folate metabolism

Screening - AFP, acetylcholinesterase
  - Glycoprotein secreted by yolk sac and fetal liver
US Screening

Nontargeted US accuracy variable

Depends on

- operator experience,
- fetal lie
- maternal body habitus
US SCREENING of targeted exams up to 100% accurate

Cranial findings

- Banana sign
- Lemon sign
- Hydrocephalus
US - Banana Sign
US “Lemon Sign” overlap frontal/parietal bone
US Cranial findings

Ventriculomegaly - 80-90%
Small BPD in second trimester
May develop macrocephaly later
MRI Cranial Findings
Small Subarachnoid Space
Frontal concavity
Ventriculomegaly
Chiari II Malformation

Small posterior fossa
Small cisterna magna
Small/absent fourth ventricle
MRI - Hindbrain Herniation

Normal
MMC- US spine findings

60% lumbosacral

• Dysraphic spine defect
• Absent overlying skin
• Tethered cord
US - Transverse views key
3D sweep to assess level of defect
MRI - Spine
MRI – MMC thin membrane sac
MR - Tethered cord
Additional Findings  equino varus
Trisomy 18
US typically can make the diagnosis

MRI useful for difficult cases
Low defect
Oligohydramnios
Obese pts
Outcome

Postnatal surgery to cover exposed cord
VP shunt (86%) –
  • 95% at least 1 shunt revision

Tethered cord, scoliosis, incontinence, urologic complications, spasticity
Outcome

24% mortality
23% seizures
80% social bladder continence
IQ>80 in 2/3 of patients
Ambulation:
23% with open defects
70% ambulation with closed defects

Bowman et al 2001
Outcome

Prognosis depends on anomalies ventriculomegaly +/- level

Level doesn’t independently affect VM, outcome

AJR 1997:169
Options
TOP
Continue
Fetal surgery
• MOMS trial
Background

Apparent benefits

• Improve hindbrain herniation - reverses
• Reduce shunting
• Conflicting results on neuromotor function
Inclusion Criteria

- MMC T1-S1 with evidence of hindbrain herniation
- Singleton pregnancy 19° to 25° wks
- Normal karyotype
Exclusion Criteria
hemorrhage
30 ° scoliosis
level below S1
Dissection of Placode
SO.....

In the era of fetal surgery for MMC it is important to reliably differentiate closed from open dysraphic spinal defects.
Diagnosis?
?Candidate for fetal surgery?
Review US –
NO hindbrain herniation
Transitional MENINGOCELE
MUST confirm presence of Chiari Malformation
Exclude other anomalies
Open neural tube defect = thin membrane
Chiari Malformation
Closed neural tube defect =
Thick overlying skin
No Chiari Malformation
Conclusions

• Wide variety of spinal anomalies
• US important screening tool
• MRI can improve prenatal diagnostic and prognostic accuracy.
• Precise fetal diagnosis becomes more important as advanced fetal management evolves.
Thank you!