Spinal dysraphism

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Presented by
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Spinal dysraphism

- A distinct group of congenital anomalies characterized by a failure of midline structures of ecto- and mesodermal origin to fuse

- Nicolas Tulp (1651)  
  spina bifida

- Virchov  
  spina bifida occulta

- Lichtenstein  
  spinal dysraphism

- Von Recklinghausen (1866)  
  classification

- Ruysch 1691 “with respect to cure little or nothing can be done”
Management historical

- Puncture of the sac
- Ligation of the sac suture, enterotome (Rizzoli)
- Sclerosants
- Bayer Flap closure (1892)
- Marcy Surgical closure (1895)
- Hautin closure at 18 M if IQ normal (43)
- Lorber, Matson, Sharrad closure in 1st week
Embryology

- Neural tissue development in 3rd week of gestation by induction by anteriormost part of the primitive streak (Hensens node) during gastrulation
- Neural groove formation and folding to form the neural tube
- Neurulation develops faster than embryonal axis and neural tube closes before axis extension is complete (26 days in humans)
- The remnants of the primitive streak in the caudal pole form the caudal cell mass which forms a lumen communicating with the lumen of the neural tube and gives rise to neural tissue distal to S2

MARK S. DIAS, MICHAEL PARTINGTON: Embryology of myelomeningocele and anencephaly. Neurosurg Focus 16 (2): 1-16
Embryology

- The first part of the tube to close is the cervical part
- At least 5 waves of closure in mammals
- Posterior neuropore closes at 25-27 days
- Posterior neuropore located at S2 level
Embryology

- Prolonged exposure of neural tissue to amniotic fluid leads to destruction of neural tissue
- Open neural tube- loss of CSF- low pressure in 4th ventricle- low attachment of tentorium. When cerebellar primordium grows, insufficient place to accommodate it and it herniates to the cervical canal

CLASSIFICATION (Tortori-Donati)

OPEN SPINAL DYSRAPHISM (95%)
- Myelocele
- Meningocele

CLOSED SPINAL DYSRAPHISM (5%)

With subcutaneous mass
- Lipomeningocele
- Lipomyeloschisis
- Myelocystocele (cervical, terminal)
- Meningocele, cervical myelomeningocele

Without subcutaneous mass

Simple dysraphic states
- Posterior spina bifida
- Lipoma (intradural, intramedullary, filum terminale)
- Tight filum, abnormally long cord
- Persistent terminal ventricle

Complex dysraphic states
- Dorsal enteric fistula/neurenteric cysts
- SCM
- Dermal sinus
- Caudal regression
- Segmental spinal dysgenesis

Epidemiology

- Wide variation 0.3-3.8/1000 live births
- Lowest in Asians
- Slight female preponderance
Epidemiology

- Genetic
  - Trisomy 13,18
  - Family history of NTD 3-4% (triples with each affected sibling)
  - Syndromic (acrocallosal, CHILD, Waardenberg, Fraser, Meckel - Gruber)
  - MTHFR, Folate synthetase, Pax 1,3
- Folate deficiency
- Obesity
- Diabetes with hyperinsulinemia 1% risk
- AED (valproate, carbamazepine) 1-2% risk
- Febrile illness early in pregnancy
- Pesticide, solvents, radiation exposure
- Tobacco use
Ante natal diagnosis

- MSAFP
  - 16-18 weeks
  - 2.5-3 multiples of median warrant a second USG

- USG
  - High resolution USG 100% prediction of level
  - Banana sign Chiari (93% of MMC)
  - Lemon sign hydrocephalus (80% of MMC)
  - Demonstration of defect in posterior elements

- Amniocentesis
  - Amniotic fluid acetylcholinesterase
  - Amniotic fluid AFP
  - Rise also seen with Turners and omphalocele

- MRI
  - Foetal MRI in the second trimester might be a clinically valuable adjunct to ultrasound for the evaluation of CNS anomalies, especially when ultrasound is inconclusive due to maternal obesity

Acta Obstet Gynecol Scand. 2010 Dec;89(12):1571-81
Counseling

- Ideally in the ante natal period
- It should not come as a surprise
- Parents should know what to expect
- The course of action already discussed and charted
- If detected post birth for the first time
  - Realistic expectations from therapy
  - The parental hope a determinant in health related quality of life
  - Screening in subsequent pregnancies
Ethical issues

- Termination of pregnancy
  - Legal up to 24 weeks in India
  - Right of fetus Vs maternal right
- To treat or not to treat (¿ historical )
- Quality of life?
  - 10-15% will die <6 Years despite therapy
  - 20-25% have subnormal IQ
  - 38% require bowel programme for continence
  - Social isolation, low self esteem 80% require psychiatric counseling
  - 10-15% require custodian care
  - < 10% are economically independent
Foetal surgery

- Reduced Amniotic fluid exposure of exposed neural tissue
- Birth trauma to exposed neural tissue
- Reduced shunt requirement
- Reduced incidence of Chiari
- No maternal mortality
- Perinatal foetal mortality 4%
Associated anomalies

- VACTERL (vertebral, anal, cardiac, tracheoesophageal, renal, limb)
- OEIS (omphalocele, extrophy, imperforate anus, spinal defects)
- Curriano triad (partial sacral agenesis, sacrococcygeal teratoma, anorectal malformation)
- CTEV
- Renal (90% have neurogenic bladder)
Peripartum management

- ? LSCS for all (after lung maturity before labour)
- 20-30% have latex allergy (latex precautions)
- Nurse lateral/ prone
- Avoid enteral feeding
- Closure 24-72 Hours (before colonization)
- 85-90% have HCP
- Most have Chiari malformation
Preoperative workup

- Imaging
  - Renal, GIT, cardiac
  - Spinal MRI (with fat suppressed sequences)
  - Cranial CT, USG
  - Only essential imaging to assess fitness for surgery and extent of surgery

- Hb, glucose

- Counseling parents

- Neurological assessment
Neurological assessment

- 2/3 have myelopathic features due to incomplete functional cord lesion
- Stimulus applied from distal to proximal till the infant grimaces
- Kyphoscoliosis, limb asymmetry may be due to SCM
- Obstructive apnoea, stridor, opisthotonus due to Chiari
Post operative management

- Hypothermia, hypoglycemia, hypocalcaemia
- Hb, coagulation
- Analgesia
- Infection avoidance
  - Frequent dressing
  - Antibiotics
- Nurse prone with head low
  - Avoids pressure on wound
  - CSF drainage (reduces leak)
  - Reduced tethering
Complications

- CSF collection/ Leak
  - Poor dural / wound closure
  - 4-8 days
- Wound breakdown / infection
  - Catabolic state
  - Poor feeding
  - Tension in closure / kyphosis
- Meningitis
- Pneumothorax
- Ileus / NEC
- Neurological deterioration (10%)
- Retethering
Open spinal dysraphism

- Meningocele, meningomyelocele, hemimeningocele, hemimyelomeningocele
- Posterior bony defect with prolapse of meninges and the cord
- 85% caudal thoraco lumbar spine, 10 % in the thorax and the rest cervical
- Multisegmental in 1%
- 80 90% associated with Hydrocephalus requiring shunt
- Most have Chiari II malformation
Open spinal dysraphism

- Delay in repair increases infection
- If delayed confirm negative cultures from placode
- 15% require shunt in the same sitting
- Avoid betadine contact with the placode
- Any tandem lesions dermoid, lipoma, SCM, neururentic cysts, meningocele manque should be addressed
Open spinal dysraphism (repair)

- **Aim**
  - Protect functional neuronal tissue
  - Prevent CSF loss
  - Reduce risk of meningitis by reconstruction of cord and coverings

- **Technique**
  - Isolation and tubulation of placode
  - Watertight dural closure
  - Closure of lumbodorsal fascia
  - Subcutaneous Undermining and tensionless skin closure
  - Plastic closure with flaps may be required
Closed spinal dysraphism

- Congenital spinal defects covered by intact skin
- *lesions seen*
  - Lipomeningocele
  - Lipomyeloschisis
  - Myelocystocele (cervical, terminal)
  - Meningocele, cervical myelomeningocele
  - Spina bifida
  - Lipoma (intradural, intramedullary, filum terminale)
  - Tight filum, abnormally long cord
  - Persistent terminal ventricle
  - Dorsal enteric fistula/ neurentric cysts/ Dermal sinus
  - SCM
  - Caudal regression
  - Segmental spinal dysgenesis
## Manifestations of occult spinal dysraphism

<table>
<thead>
<tr>
<th>Cutaneous stigmata</th>
<th>Orthopedic deformities</th>
<th>Urologic problems</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asymmetric gluteal cleft</td>
<td>Foot or leg deformities</td>
<td>Neurogenic bladder</td>
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<tr>
<td>Capillary hemangioma</td>
<td>Scoliosis</td>
<td>UTIs</td>
</tr>
<tr>
<td>Subcutaneous lipomas</td>
<td>Sacral agenesis</td>
<td>Incontinence</td>
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<tr>
<td>Hypertrichosis</td>
<td></td>
<td>Delay in toilet training</td>
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<tr>
<td>Dermal sinus tract</td>
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<tr>
<td>Cutis aplasia</td>
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</table>
# Neurological signs and symptoms in different age groups

<table>
<thead>
<tr>
<th>Infants</th>
<th>Toddler</th>
<th>Older children</th>
<th>Young adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>Decreased spontaneous leg movements</td>
<td>Delayed walking</td>
<td>Asymmetric motor/ sensory</td>
<td>Back pain</td>
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<tr>
<td></td>
<td></td>
<td>development</td>
<td></td>
</tr>
<tr>
<td>Absent reflexes</td>
<td>Abnormal gait</td>
<td>Back/leg pain</td>
<td>Leg cramping/pain</td>
</tr>
<tr>
<td>Leg atrophy</td>
<td></td>
<td>UMN signs</td>
<td>Spasticity</td>
</tr>
<tr>
<td>Foot asymmetry</td>
<td></td>
<td>Painless ulceration</td>
<td>Hyperreflexia</td>
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<tr>
<td>Decreased urinary stream</td>
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<td></td>
<td>Bowel/bladder incontinence.</td>
</tr>
<tr>
<td>Structure</td>
<td>Findings</td>
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<td>-----------------------------------</td>
<td>--------------------------------------------------------------------------</td>
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<tr>
<td>Lamina</td>
<td>Fusion defects, midline defects, abnormal spinous processes</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vertebral bodies</td>
<td>Hemivertebrae, Butterfly vertebrae, Block vertebrae, Midline cleft defects, canal stenosis</td>
<td></td>
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</tr>
<tr>
<td>Disk space</td>
<td>Congenital narrowing</td>
<td></td>
<td></td>
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<tr>
<td>Pedicles</td>
<td>Flattening, thinning</td>
<td></td>
<td></td>
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<tr>
<td>Widening of spinal canal</td>
<td>Interpedicular widening, scalloping of posterior border, Midline bony spur.</td>
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<td></td>
</tr>
<tr>
<td>Failure of development</td>
<td>Reduced number of vertebral bodies, Absence of parts of vertebrae, sacral dysjunction</td>
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<tr>
<td>Spinal curvature</td>
<td>Scoliosis, kyphosis, lordosis.</td>
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</tbody>
</table>
Lipomeningocele

- Lipoma with lipoma cord interface posteriorly distracted out of the spinal canal
- Congenital lumbosacral lipomas the entire spectrum
- Dysjunction in timing of closure between the surface ectoderm and neural tube allowing mesenchyme to migrate/differentiation of caudal mass cells into lipoma
- Symptoms due to tethering/compression
- 1:4000 births F>M
- Defecits start by 1-2 years
**Classification**

- **Type I dorsal**
  - Attached to posterior cord
  - Distinct cord lipoma interface
  - Cord and coverings distally normal
  - Lipomeningocele/ lipo MMC

- **Type II transitional**
  - Interface continues beyond myeloschisis
  - No normal cord distally
  - Roots enmeshed in lipoma

Type III caudal
- Arise from conus/filum
- Largely/completely intradural
- Interface diffuse

Chaotic (Pang)
- Caudal portion ventral to placode
- Fusion lines blurred distally
- Location of DREZ and roots unpredictable

Surgery

• Indications
  • SC lipoma in infants > 2M with cord lipoma attachment
  • Fresh / progression of deficits
  • Undergoing scoliosis correction
  • Severe dysesthetic pain

• Goals
  • Detach cord tethering structures
  • Debulk as much lipoma as possible
  • Close myeloschisis and reconstruct tubular cord
Surgery

• Technique
  • Expose 1-2 vertebral level above and 1-2 cm below
  • Wide laminectomy
  • Detach dura from lipoma
  • Identify and separate fusion line (medial to DREZ and roots)
  • Sharp dissection rostro caudally in the white plane
  • Neural tissue after 3 sacral roots may be sacrificed
  • Pia – pia sutures with non absorbable sutures
  • Expansile duroplasty
Hypertrophic Filum terminale

- 50% Associated with sacral/ gluteal cleft dimples
- Leg weakness, numbness, sphincter weakness
- MRI conus below L2 and filum > 2 mm
- Fibrous / fatty
- VACTERL, arthrogryphosis
- TCS Symptoms may be mimicked by non functional shunt
- Surgery
  - All symptomatic
  - ? Asymptomatic
  - Identify filum/ ensure no roots on it ( nerve stimulator)
  - Cauterize and divide filum
Split cord malformation

- Commoner in females 3:1
- Average presentation 4-6.5 years
- Cutaneous lesions in 71%
- Associated with MMC 6%
- Location of split
  - Lumbar 47%
  - Thoracolumbar 27%
  - Thoracic 23%
  - Cervical / sacral 1.5%
- Formation of accessory neurentric canal with communication between yolk sac and amnion
Split cord malformation

- **Type I**
  - 2 Cords in separate dural sleeves separated by bony/cartilaginous septum

- **Type II**
  - 2 cords in single dural sleeve separated by fibrous septum

### Split cord malformation

<table>
<thead>
<tr>
<th>Type of Deficit</th>
<th>No. of Cases (%)</th>
</tr>
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<tbody>
<tr>
<td>motor</td>
<td>68 (26.77)</td>
</tr>
<tr>
<td>sensory</td>
<td>12 (4.72)</td>
</tr>
<tr>
<td>autonomic</td>
<td>18 (7.08)</td>
</tr>
<tr>
<td>motor &amp; sensory</td>
<td>50 (19.68)</td>
</tr>
<tr>
<td>motor, sensory, &amp; autonomic</td>
<td>34 (13.38)</td>
</tr>
<tr>
<td>motor &amp; autonomic</td>
<td>21 (8.26)</td>
</tr>
<tr>
<td>asymptomatic</td>
<td>38 (14.96)</td>
</tr>
</tbody>
</table>

**Management**

- Detethering of the cord
- Excision of septum/ fibrous septae
- Look for and divide ventral tethering elements
- Conversion into single dural tube
- Associated defects addressed

Myelocystocele

- Occult spinal dysraphism with a localized, cystic dilation of the central canal of the spinal cord that is herniated through a posterior spina bifida
- 4-8% of LS occult dysraphism
- Terminal / non terminal
- No familial, sex preponderance
- Associated with OEIS
Myelocystocele

- Cystic, skin covered LS mass, in gluteal cleft
- Deficits due to associated lesions
- Theories
  - Steinboch and Cochrane: LDM in a hydromyelic cord
  - McConnell and Naidich: dilatation of terminal ventricle due to closure of terminal neourethral
- MRI trumpet like flaring of distal central canal
- Surgery for all

Closure of the dural layer (2) and reinforcement of the dural closure (3) with mobilized pedicled muscle on a lumbosacral terminal lipomyelocystocele. The terminal end of the dilated cystocele (1) is left open to communicate with the subarachnoid space distally.

Mahapatra AK, Gupta DK: Terminal myelocystoceles: a series of 17 cases. Neurosurg (pediatrics) 103: 344-352, 2005
Neurenteric cysts

- Can occur along the entire neural axis
- Male predominance
- Complicated lesions present later in life
- Pain myelopathy, meningitis acute neurological deterioration
- MRI high protein content without haemorrhage

Neurenteric cyst management

- Complete excision is the goal
- Approach anterior Vs posterior
- Aspiration reduces the cyst size and eases dissection
- Dense adhesions if previous meningitis
- Post op complication rate 23%
- Recurrence rate 11%
Dermal sinus tract

- Tracts reaching from the skin to varying structures
  - 10% subcutaneous
  - 20% between fascia and dura
  - 10% subdural
  - 60% subarachnoid space

- Location
  - 90% lumbosacral
  - ≈ 1% cervical
  - ≈10% thoracic

- The tract may traverse several vertebral levels before penetrating dura
Dermal sinus tract

- **Theories**
  - Abnormal separation of cutaneous and neuroectoderm
  - Splitting of notochord and persistence of mesenchymo cutaneous fistula

- **Presentation**
  - Majority detected by 5 years on cutaneous stigmata
  - TCS
  - Infectious, Neurological, urological, orthopedic

- **Management**
  - Surgery for all
    - If infected after infection subsides
    - To relieve mass effect in presence of infection
Meningocele

- Prolapse of meninges and CSF through a defect
- Posterior, anterior, lateral
- Absence of Chiari, Hydrocephalus, Limb anomalies Vs MMC
- Concomitant neurocutaneous lesions which may cause tethering (posterior)
- Elective surgery at 4-6 months
Meningocele (anterior, lateral)

- Commoner in females
- LS location commonly presacral
- Curriano’s triad
- Anterior approach to close dural defect and detether
Thank you