Incidence : 1: 250

Now Decreasing with antenatal care
How to pick up

- Maternal Alpha-fetoprotein – 80-90%
- Ultrasonography –
- Amniotic Fluid for –
  - acetylcholine esterase, AFP,
  - chromosomal analysis (Trisomy 13 & 18)
- Terminate
Neural tube defects
wide spectrum

- **Occulta** – Cutaneous stigmata
  Diastematomyelia

- **Aperta** - Meningocele,
  Meningomyelocele
  Rachischisis
  Lipomeningomyelocele

- **Anencephaly** – ? terminate
Why does it occur:

Failure of midline fusion

When does it occur:

- Open MMC: 12-13 days
- Covered defects: > 28 days
Etiology -

Mostly Not known

Radiation, Environmental, Consanguinity Counseling ?

Folic Acid def.
Anticonvulsants ?
Alcohol intake ?
Single / multiple Defects

Higher the MMC - Lesser the deficits
Lumbo- Sacral MMC with gross deficits

**Deficits:**
- Paresis
- Neurogenic bladder
- Neurogenic bowel
- 80% hydrocephalus

**Ethical Issue:** Operate or NOT
Girl with hanging MMC
(r/o S.C.Teratoma - calcification, solid)
Large defect: Rachischisis

- Extensive damage
- No surgery advised
- Antenatal repair improves the neurologic deficit
Associated anomalies

- Club feet - commonest
- Arnold Chiari Malformation (80-90%)
- Hydrocephalus,
- Muskulo-skeletal defects
- Exstrophy UB, UDT, HN.
- Vesico ureteric reflux
- Dislocation of hip
- Anorectal Malformation
- CHD, umbilical hernia, prolapsed uterus
Clinical Assessment

- For single / multiple lesions
- Associated anomalies
- Sensory – distal to proximal
- Motor  Hip flexion L1-3, Knee flexion L5-S1
- Bladder - ?compressible
- Bowel - anal reflex, patulous anus
Investigations for

- X-ray - Spine, in Newborns
- MRI, UGS for Kidneys for occulta group
- Associated hydrocephalus, fundus
How to Manage

?? selection criteria, ?? Operate all
Immediate Surgery

- No neurological deficit
- Meningocele, MMC
- Limited weakness of legs
- Impending rupture, leak
- Recent leak (after 24hrs antibiotic)
Delayed Surgery

- Good skin cover
- Atretic MMC (? MRI)
- Infected lesions, Leak > 24hrs
Surgery Contraindicated

- Open neural tube defect (apparent deficit)
- Infected and leaking lesions with deficit
- MMC with neurological deficit –
  - Bladder, bowel, Limb paralysis,
  - Gross hydrocephalus
Postnatal outcome

Surgery

Well – 95%
Postop. Deficit - 5%
Orthopedic support

No Surgery

80 % die – 1 Yr
10% for surgery
10% crippled
Late results

- Paresis due to ? Tethered cord
- Assoc. spinal pathology on MRI
- UTI, Neurogenic bladder, bowel
- Scoliosis, Trophic ulcers,
- Non healing wounds
- Psycho-social adjustments
Encephalocele

- 1 in 5000, girls
- Frontal, Occipital,
- Atretic
- Associated Microcephaly
- USG for contents – Surgery
- Poor outcome: if cerebellum, brain, HC, sinus
- Mental retardation, ataxic gate, microcephaly
Lipo MMC usually with NO deficits
(Late - Abnormal gait, UTI, trophic ulcers)

MRI for detailed anatomy
Diastematomyelia
(Thick filum, spur, double sheaths)
Newborn with antenatally diagnosed Spina Bifida Occulta, with a discharging sinus

MRI for detailed anatomy
Neuroenteric cysts:
Large MMC, with int. mucosa, deficit of legs, Early hydrocephalus
NB with Neuro-enteric Malformation
Post op. Hydrocephalus

- Increase in head size
- Medical Therapy
  - diamox/Glycerol
  - 50% CSF is reduced
  - 50% pts. respond
Indications for Shunt surgery

- Evidence of raised ICP
- Uncontrolled with medical therapy
- Cortical mantle <2 cm in an infant < 6 mo
- Follow-up showing - Evidence of thinning of cortical mantle, delayed milestones, falling MPQ
- Parental informed consent
Monitoring for Head size during treatment
Hydrocephalus: AIIMS Exp.
(> 2200 cases in HC Clinic)

- Congenital hydrocephalus - 46%
- Post-MMC hydrocephalus - 28%
- Post-Meningitic hydrocephalus - 21%
- Post shunt infection - 7%
Best is if Shunt can be avoided
Postop complications – too many

- Shunt malfunction,
- Infection -skin level,
- Shunt colonization)
- Shunt fracture, extrusion
- Subdural hematoma
- Slit ventricle syndrome
- Cranial synostosis
- Seizures
Stem cell therapy in Spina Bifida

Indications:
Spina Bifida with neurological deficits, and family insisting for surgery

Aim:
Try if SCT can help recovering the neurological deficit - as a Pilot study
Procedure for SCT in Spina Bifida

- **Repair**: Of spina Bifida (selection criteria)
- **Special**: consent for MMC repair & SCT
- **Source**: Bone marrow from Tibia
- **Dose**: 4 ml, 4 million / ml
- **Route**: Local injection, caudal space
- **Sites**: Spinal Cord, muscles, bone

Mainly in Distal half of defect, in Caudal space (1 ml)
Material: (Aug. 2005-07*)
22 Pts. (age 2 days – 6 yrs)

- MMC repair & SCT: 17
- Only SCT: 4
- Re-do case: 1
  (4 yr old now, neurologic deficit, tethered cord, with diastematomyelia, Syrinx, dermoid cyst)

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>NB</td>
<td>7</td>
</tr>
<tr>
<td>1-5 m</td>
<td>11</td>
</tr>
<tr>
<td>1-6 yr</td>
<td>4</td>
</tr>
<tr>
<td>(Ruptured)</td>
<td>7</td>
</tr>
</tbody>
</table>

Total 22
(antenatal – 1)
Postop. Monitoring

- Incidence of postop. hydrocephalus
- Improvement in leg movements
- Monitoring by evaluation of neurological status – bladder / bowel
- Comparing with the preop. status
Results: FU 1-24 months (N=22)

- Improved power - 9 /13
  (Dramatic recovery -6)
- Improved sensory loss: - 1
- Status quo (already good) - 5
- No improvement - 4
Summary: Spina Bifida

- Very Common & serious malformation
- Selection Criteria – judicious treatment
- Postop. HC (80%), needs decompression
- Post Shunt: High complications
- A composite management is essential
Prevent if you can

- Folic Acid therapy
- Antenatal diagnosis - Termination
- Stem cell Role - undecided
Qutab minar

Red Fort

Jama Masjid

Bahai Temple
January 2005

NICH Symposium, Karachi
MMC – Large, with all deficits