A 50 Year Experience with Management of Spina Bifida Aperta: Myelomeningocele

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Etiology of Neural Tube Defects

Chromosomal Anomalies
- T13; T18; Triploidies, others?

Gene Determined
- Dominant - ?
- Recessive (Encephalocele)
  - Roberts Syndrome
  - HARD Syndrome
The Spinal Fluid Shunt Era

- Holter Valve 1955
- Heyer Shunt 1957
- Ventriculsar Atrial Systems
# Meningomyelocele Current State of Management

<table>
<thead>
<tr>
<th>Postnatal Management</th>
<th>Much improved</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newer CSF shunts</td>
<td></td>
</tr>
<tr>
<td>Ultrasound, CAT Scan, MRI</td>
<td></td>
</tr>
<tr>
<td>Newer more powerful antibiotics</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Prevention vs Termination</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Prenatal Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cesarean Section</td>
</tr>
<tr>
<td>“Intrauterine Myelomeningocele Repair”</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Management of “Normal Development”</th>
</tr>
</thead>
</table>
NEURAL TUBE DEFECTS SCREENING

Maternal Serum
Alpha-fetoprotein
Ultrasound
Salad Signs
19-YEAR FOLLOW-UP OF FETAL MYELOMENINGOCELE BROUGHT TO TERM

Sharon Liu, David Shurtleff MD, Richard Ellenbogen MD, John Loeser MD, Robert Kropp MD

Children’s Hospital and Medical Center Seattle, WA USA
Meningomyelocele

Intact Amniotic Membranes During Labor
Meningomyelocele
Sac > 1 cm

Ruptured Amniotic Membranes During Labor
MENINGOMYEOCELE
Sac < 1 cm

Ruptured Amniotic Membranes During Labor
FETAL CONTRA

INDICATIONS FOR
PRELABOR C/S

- Abnormal chromosomes
- Fatal fetal anomaly
- Hydroanencephaly
- Flat or depressed MM lesion
- ? Gibbus - Kyphosis
Intrauterine Myelomeningocele Repair - IUMR

Joseph Bruner, MD Vanderbilt
October 2003

- Fewer urinary tract infections
- Less GE reflux
- Improved leg function
- Less need for CSF shunts
- Improved cognition
I UMR and Chiari II

- Improved anatomy - V + CHOP

Symptoms

referring literature V + CHOP centers
6/22 (27%) 11/63 (17%)* ?
5 - 30%*
+ 6 ? - Vent
55% *

*X^2, P=1
## IUMR - Hydrocephalus CSF Shunts

<table>
<thead>
<tr>
<th>V + CHOP Centers</th>
<th>Controls</th>
<th>Referring</th>
</tr>
</thead>
<tbody>
<tr>
<td>57/104 (55%)</td>
<td>162/189 (86%)</td>
<td>18/22 (82%)</td>
</tr>
<tr>
<td>X^2, P=0.15</td>
<td>X^2, P=0.03</td>
<td></td>
</tr>
<tr>
<td>P=0.003</td>
<td>20/22(91%)*</td>
<td></td>
</tr>
</tbody>
</table>

*includes 2 lost to follow-up
<table>
<thead>
<tr>
<th>Complication</th>
<th>V + CHOP Referring</th>
<th>V + CHOP Referring</th>
<th>Seattle + UCSF Centers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Death</td>
<td>11/190 (6%)</td>
<td>-</td>
<td>1/31 (3%)</td>
</tr>
<tr>
<td>Dehiscence</td>
<td>1/10 (10%)</td>
<td>5/22 (23%)</td>
<td>0/31</td>
</tr>
<tr>
<td>Infection</td>
<td>?</td>
<td>5/22 (23%)</td>
<td>0/31</td>
</tr>
<tr>
<td>Hydromyelia</td>
<td>?</td>
<td>2/22 (9%)</td>
<td>0/31</td>
</tr>
<tr>
<td>Tethered Cord</td>
<td>3/49 (6%)</td>
<td>1/22 (5%)</td>
<td>0/31</td>
</tr>
</tbody>
</table>

Fisher Exact, P < 0.0001
Maternal-Fetal Surgery

Neural Tube Defects:

The Management of Myelomeningocele Study

MOMS
THE DEFECTIVE INFANT

URINARY INFECTION
SHUNT OBSTRUCTION

BIRTH OF DEFECTIVE
BACK CLOSURE
SHUNT
FOOT CASTS OR OPERATIONS

PERMANENT CRIPPLES
MENTAL RETARDasION

PARENTAL ANXIETY CURVE

[Diagram depicting stages and outcomes related to a 'defective infant']
Myelomeningocele

TRANSITION

Begins in infancy
- Mobility
- Self Care

Adolescence
- Asexual to sexual person

Recurrence Risk
Move effectively and efficiently through space
Multiple Modes of Mobility
<table>
<thead>
<tr>
<th>Surface Type</th>
<th>Parapodium (%)</th>
<th>Wheelchair (%)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hard Floor</td>
<td>0</td>
<td>0</td>
<td>insig</td>
</tr>
<tr>
<td>Rug</td>
<td>34%</td>
<td>94%</td>
<td>0.01</td>
</tr>
<tr>
<td>Level Turf</td>
<td>69%</td>
<td>90%</td>
<td>0.05</td>
</tr>
<tr>
<td>Rough Ground</td>
<td>8%</td>
<td>38%</td>
<td>0.05</td>
</tr>
<tr>
<td>Curbs/Ramps</td>
<td>42%</td>
<td>69%</td>
<td>0.05</td>
</tr>
<tr>
<td>Snow/Ice</td>
<td>0%</td>
<td>38%</td>
<td>0.05</td>
</tr>
</tbody>
</table>
# Activities of Daily Living

## Ease of Care

<table>
<thead>
<tr>
<th>Activity</th>
<th>Parapodium</th>
<th>Wheelchair</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female Cath</td>
<td>3.3</td>
<td>1.6</td>
<td>0.01</td>
</tr>
<tr>
<td>Bowel</td>
<td>4.0</td>
<td>2.0</td>
<td>0.001</td>
</tr>
<tr>
<td>Underwear</td>
<td>3.0</td>
<td>2.0</td>
<td>0.002</td>
</tr>
<tr>
<td>Dressing</td>
<td>2.7</td>
<td>1.7</td>
<td>0.005</td>
</tr>
<tr>
<td>Into Device</td>
<td>2.6</td>
<td>2.0</td>
<td>0.05</td>
</tr>
<tr>
<td>Out of Device</td>
<td>2.3</td>
<td>1.7</td>
<td>0.04</td>
</tr>
</tbody>
</table>
COMPARISON OF TOTAL POPULATION MEAN SCORES TO EXPECTATIONS OF NORMAL

Points

Social interaction
Appliance care
Bowel program

Age, years

1/2/4 2 3 4 5 6 7 8 9 11 13 15 17 18+
Toilet training

Little changed from 1976 and now in 2006
Despite CIC and MACE


OUR PUBLISHED DATA FROM 1986 follows
Quality of Urinary Independence
Levels L2 & Above

- Independent
- Dependent
- Poor Quality

0% - 100%

0 - 36 MOS
3 - 4 YRS
5 - 6 YRS
7 - 8 YRS
9 - 10 YRS
11 - 12 YRS
13 - 15 YRS
16+ YRS
Quality of Urinary Independence
Sacral level Lesions

Quality of Urinary Independence

Independent
Dependent
Poor Quality
STAGES OF GENITOURINARY DEVELOPMENT - 1

Infancy
- Hold and release urine
- Exploration of genitalia

Early Childhood
- Observation of other’s and demonstration of own genitalia
STAGES OF GENITOURINARY DEVELOPMENT - 2

Early School age

- Discard Diapers
- Rx odor and soiling
- Fallibility of one’s nose
- Privacy of one’s genitalia
STAGES OF GENITOURINARY DEVELOPMENT - 3

Late Childhood
- Prostatitis/epididymitis
- Pregnancy

Adolescents
- Sex and reproduction
MALE FERTILITY

25% OR LESS AS ESTIMATED BY INCOMPLETE STUDIES

Shurtleff et al. Myelodysplasia: Western Journal of Medicine 122:199, 1975


85% OR MORE FOLLOWING TESTICLE BIOPSY AND IN VITRO FERTILIZATION

Penile Sensation for men with myelomeningocele

Three successful cases

- When at what age
- Effect on development of Sexuality?
- Effect on development of male motivation?
Learned Helplessness

Teaching the Student with Spina Bifida Text
Thank you for your attention