SPINA BIFIDA

Dr Kevin DUNNE
Developmental Medicine
Rehabilitation Service
2010

Aetiology/Classification
- Malformation of spinal cord and brain
- 28 days of development of embryo
- Failure of fusion of neural folds during neuralation
- Cause unknown?
- Genetic ? Metabolic ? Environmental
- 75% isolated
- 25% associated with other defects

Partial paralysis of lower limbs
- No weakness
- Partial paralysis
- Wheelchair dependence

Joint deformity
- Spinal abnormalities
  - Scoliosis
  - Kyphosis/lordosis

Neurogenic bladder
- Urinary incontinence
- Renal and bladder abnormalities

Neurogenic bowel
- Faecal incontinence

Skin
- Anaesthetic – Pressure sores
Spina Bifida

- Hydrocephalus
  - Arnold Chiari Type 2
  - CSF shunt
  - Learning problems

- Upper spinal cord mild abnormalities
  - Mild upper limb problems

Spina Bifida - types

- N
- Dysraphism
- Meningocele
- MM

Myelomeningocele

- spina bifida occulta
  - 10% adult spines

- meningocele
  - 6% cases
  - 11% survivors

- myelomeningocele
  - 94% cases
  - 89% survivors

Encephalocele

Lipomeningocele

Spina Bifida Thoracic

10% adult spines
6% cases
11% survivors
94% cases
89% survivors
Meningocele

Spina Bifida

Severe

Incidence

- Australia: 0.95/1000
- Western Europe: 1.5-3.0/1000

Sex Incidence
- Male: 58%
- Female: 42%

Spina Bifida- incidence

- Varies from country to country
- Range 1-5/1000 live births
- High in Celts (Ireland, Wales)
- Low in Northern Europe.
- Encephaloceles higher in SE Asia

Foetal neural tube

- Conception - D18
  - 3 germ layers which make the different tissues and the neural plate forms along length of fetus
- D19 - D28
  - Neural tubes fold and should close along length of fetus

Spina Bifida
Spina Bifida - Classification

- Neural tube defect
  - Anencephaly: 50% (lethal)
  - Spina Bifida Cystica
    - Myelomeningocele: 45% (2/3 - 90% live)
    - Meningocele: 5% (all live)
    - Encephalocele
- Lipomyelomeningocele
  - Clinically similar but no Hydrocephalus
- Spina Bifida Occulta
  - 710% back pain, constipation

Myelomeningocele

- Cervical: 0.5% mild
- Thoracic only: 0.5% Severe
- Low thoracic: 27% Severe
- High Lumbar L1-2: 23% WChair
- Low Lumb, Upper sacral: 45% Braces
- Lower sacral: 4% Independ

Lipomyelomeningocele

- Lipoma
  - Grows in the spinal canal or outside
  - Press on the spinal cord
  - Nerves traverse and become nonfunctional
  - Partial weakness of lower limbs
  - Neurogenic bladder and bowel
  - Spine abnormality
  - No hydrocephalus - normal cognition

Lipomeningocele

- MRI of Underlying Abnormality (Lipoma)

Spina Bifida - Aetiology

- Unknown - multifactorial
  - Abnormal gene interacts with other genetic loci and
    or environmental factors to modulate the incidence or
    severity of the defect.
- Genetic
  - Increased in chromosomal abnormalities i.e. trisomy
    13 and 18
  - Increased in siblings and other relatives
  - Abnormal genes
    - SOM
    - VANGL1 (mouse, 3/166) (NEJM 355,(14)2007

Genetics

- Multi-factorial
  - 1 affected child: 1:50 Recurrence Risk
  - 2 affected children: 1:10 RR
  - 3 affected children: 1:4 RR

Note

- Risk of any child born with a major defect 1:30 (3%)
- If an adult with Spina Bifida has a child risk is 1:25
Spina Bifida - aetiology
- Drugs
  - Valproate (1%)
  - Methotrexate
  - Clomiphene
  - Folic acid antagonists
  - Trimethaprim, Anticonvulsants
- Environment
  - Maternal diabetes, Hyperthermia,
  - Paternal - Agent orange

Incidence
- Australia 0.95 /1000
- Western Europe 1.5-3 /1000
- Male 58%
- Incidence has decreased significantly in Victoria, largely due to termination, secondarily due to folate.

Spina Bifida - Prevalence
- NTD conceptions in Victoria
  - 40% decline 1995 to 2000
- Due to folic acid supplementation
  - 1995 2005
  - NTD 19.2 -12.2/10000
  - Spina Bifida 8.3 -7.9/10000
  - Increase in encephalocele (increased Asian)
- Reduced births due to TOP

Spina Bifida Common Birth Defect
Top most common birth defects, 2005

<table>
<thead>
<tr>
<th>Defect</th>
<th>N/10,000</th>
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<tbody>
<tr>
<td>Hydropsphalus</td>
<td>45.7**</td>
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<tr>
<td>Obstructive defects of the renal pelvis</td>
<td>36.4</td>
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<tr>
<td>Ventricular septal defect</td>
<td>28.5</td>
</tr>
<tr>
<td>Trisomy 18</td>
<td>17.5</td>
</tr>
<tr>
<td>Congenital dislocated hip</td>
<td>16.1</td>
</tr>
<tr>
<td>All NTDs</td>
<td>12.2</td>
</tr>
<tr>
<td>Trisomy 19</td>
<td>9.1</td>
</tr>
<tr>
<td>Myelodysplasia</td>
<td>8.8</td>
</tr>
<tr>
<td>Cleft palate</td>
<td>8.4</td>
</tr>
<tr>
<td>Cystic kidney disease</td>
<td>7.3</td>
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</table>

*This figure was not included in the original image.*

Spina Bifida

Number of new cases/year
Royal Children's Hospital

<table>
<thead>
<tr>
<th>Year</th>
<th>Number new SB cases</th>
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<tbody>
<tr>
<td>1970</td>
<td>30</td>
</tr>
<tr>
<td>1990</td>
<td>28</td>
</tr>
<tr>
<td>2000</td>
<td>10</td>
</tr>
<tr>
<td>2006</td>
<td>5</td>
</tr>
<tr>
<td>2008</td>
<td>2</td>
</tr>
</tbody>
</table>
Spina Bifida

Incidence

- So why have the numbers dropped
- Antenatal diagnosis with ultrasound
- Termination of pregnancy
- Folate – small impact
- Small drop in prevalence

Spina Bifida - Prevention

- 1960's Laurence in Wales ?Vitamin Deficiency.
- 1970's Smithells
  Periconceptual multivitamins
  Reduced conception rate if previous NTD
  Methodological flaws
- MRC Study 1992
  Multi centre trial
  Stopped at 1400 (n = 2000)
  76% reduction recurrence risk for NTD
  Folate rather than other Vitamins the cause

Folate

- Folate supplementation reduces recurrence by up to 72%  
  _Lancet 1991_
- Recommend -
  1 month pre- and 3 months post-conception
  - Routine 0.5mg daily
  - High risk pregnancy 5mg oral daily
    - Parent with SB
    - Parent with a previously affected child
    - Mother on Anti-Epileptic Drugs
    - Mother with Diabetes Mellitus

SB - Folate Prevention

How does it work – unknown

Generalizable?

- Berry NEJM 2004
  130000 Chinese women 0.4 mg folate, 117000 controls
  - 41% reduction NTD
  - Risks - nil reported ? Twins

How much?

- 0.4mg – 36%
- 5mg – 85%

Peri-conceptual folic acid
Awareness post 1996,1999

<table>
<thead>
<tr>
<th></th>
<th>Before</th>
<th>1997</th>
<th>2000</th>
</tr>
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<tbody>
<tr>
<td>15-24 yrs</td>
<td>5%</td>
<td>6%</td>
<td>4%</td>
</tr>
<tr>
<td>25-34 yrs</td>
<td>19%</td>
<td>24%</td>
<td>29%</td>
</tr>
<tr>
<td>34 yrs+</td>
<td>12%</td>
<td>16%</td>
<td>26%</td>
</tr>
</tbody>
</table>

NTD Recurrence Risk

- One child 2%
- Two Children 10%
- Parent NTD 4%
- Blood related 1%
- Remarry 0.6%
Spina Bifida

- 12.2 NTD per 10000 conceptions
- 85 NTD for 70000 conceptions Vic pa
- 5 new case treated
- 1% blood relative risk

Multidisiplinary Team

- Allied Health
- Medical
- Stomal therapist
- Urologist
- Physiotherapist
- Neurosurgeon
- Occupational therapist
- Orthotist
- Neonatologist
- Paediatrician

Spina Bifida Management

Multidisciplinary team

- Neuropsychologist
- Social Worker
- Clinical Psychologist
- Education
- General practitioner
- Spina Bifida Association - social

SB – Folate to flour

- Folate added to flour
- Average daily consumption
  - Supplement to 0.4 mg
- USA and 38 other countries
- 36 % reduction in NTD
- Australia about to start

Multidisciplinary team

- Ultrasound
- Lesion vertebral column
- Hydrocephalus - lemon sign

- Amniocentesis
- AFP, open lesion only
- Maternal Serum Screening
- AFP non specific (twins, down etc)
Spina Bifida
Antenatal Ultrasound
- Can visualise at 15-16 weeks
- Vertebra have 3 ossification centres

Spina Bifida
Antenatal Ultrasound
- Ultrasound – “sac”

Spina Bifida
Antenatal ultrasound
- Lemon and banana sign

Spina Bifida
Foetal Surgery
- Does foetal surgery improve outcome
- Randomized trial USA
- Ongoing at moment
- No data yet

Termination
- Prenatal Ultrasound
  - 18-20 weeks
  - Detailed foetal anatomy
  - Feto-maternal obstetric outpatients for discussions and counselling
  - Termination at 20-22 weeks should the parents choose

Spina Bifida
Mode of Delivery
- Improved outcome with elective LUCS
- Use if leg movement seen on US
- Rationale
  - Reduces damage to neural plaque
  - 2 neurosegmental levels better
  - Bladder and bowel status unchanged
  - (LUTHY et al NEJM 1989)
Spina Bifida
Initial Assessment
- Refer specialist centre
- Neurosurgical assessment
- Paediatric assessment
- Discussion with family
- Facts not biases
- If severe - non treatment option
- Family’s wishes respected
- Palliative care - 10% survive

Spina Bifida
Initial Assessment
- Clinical exam – chromosomal/other
- Extent of lesion – can it be closed
- Motor level
- Hydrocephalus cranial US
- Musculoskeletal Hip US
- Joint deformity
- Spine deformity
- Renal Renal US

Spina Bifida
Initial Assessment
- “to treat or not to treat”
- Prior 1955 most died
  - Hydrocephalus, Meningitis, Renal
- 1955 CSF shunt (VA), Ileal conduits
- 1955-70 many with severe abnormalities
- Treatment Criteria
  - Lorber 1971
  - Smith and Smith 1973

Spina Bifida
Initial Treatment
- Back closure within 24 hours (infection)
- Prophylactic antibiotics
  - Delay closure for 72 hour
- Monitor for hydrocephalus
  - Back closure head circumference increases
- Shunt insertion about 1 week later
- Hospital 1-3 weeks

SB - Neurosurgical
- Back Closure
  - Within 24 hours
  - Preserve nerves
  - Motor deterioration post surgery
- Investigations US, CT MRI
- Back closure - hydrocephalus
Spina Bifida Hydrocephalus

- 80-90% develop significant hydrocephalus
- Increased with higher level
- 20-30% present at birth and by 1 month of age in 77%
- Rarely develops after 6 months
- Frequently progress after back closure
- Most shunts in 1st month

SB - Hydrocephalus

- Arnold Chiari type 2
- VP shunt
- Complications
  - Infection, obstruction, disconnection
  - Low pressure

SB - Hydrocephalus

- CSF Shunt

Arnold Chiari Type 2

- Tethered Cord Causes
  - Myelomeningocele
  - Post operative
  - Tight filum
  - Lipoma/lipomeningocele
  - Split Cord (diastematomyelia)
  - Dermal sinus
  - Tight or fatty filum terminale
Tethered Cord Symptoms
- Pain
- Weakness- evolving
- Sensory Loss
- Incontinence
- Scoliosis
- Cutaneous markers
- Investigate Muscle chart, MRI

Spina Bifida Tethering

Lipoma - tethering

Diastematomyelia

Neurosurgical Clinical
- Epilepsy
  - 10 - 15%
  - Increased with shunt obstruction, Infection, ventriculitis
- Arachnoid Cysts
- Sphynx
Spina Bifida

- Orthopaedic

SB Orthopedic Aims

- Pattern of motor development near normal
- Maximize mobility
- Stable posture if standing
  - Centre of gravity over feet
- Prevent pelvic obliquity
- Correct spinal deformity
  - “stability of skin”

SB Orthopaedics Principals

- Developmental knowledge
- Natural history of condition
- Monitor progress
- Time interventions
- Investigations
  - Imaging
  - Muscle Charts
  - Gait analysis

SB Orthopaedic Hip dislocation

- Thoracic L1, L2, L3 53%
- L4 33%
- L5 20%
- Bilateral
  - Operative benefit marginal
  - Pain
  - Prevent hip flexion contracture - or fit an orthosis (RGO)
- Unilateral
  - Leg length discrepancy, pelvic obliquity
  - Ducubitis
  - Operate on low lesion

SB Orthopaedics Knee

- Valgus deformity
- Flail undeformed knee
- Undeformed knee with reduced quadricep
- Fixed flexion
- Treatment
  - Cast
  - Braces
  - Muscle transfers
  - Osteotomies
Deformities in Ankle Feet

- The deformities include:
  - Equinus deformity
  - Clubfoot or talipes equino varus deformity.
  - Calcaneal deformity.
  - Cavus or cavus-varus deformity.
  - Plano-valgus deformity.

Clubbed Feet

Spine deformity

- Scoliosis
- Kyphosis
- Lordosis

- Monitored by X-ray
- Worsen with age
- Adolescence
- Pain, decubiti sitting
- Brace
- Surgery

Scoliosis Surgery

SB Orthopaedics Techniques

- Casting
- Orthosis
- Soft tissue releases
- Muscle transfers
- Bony procedures
  - Osteotomies
  - Arthrodesis
  - Ilizarov frames

AFO
KAFO
- Knee and ankle stability
- Mobilize with crutches
- Lock to enable sitting

RGO
Reciprocating gait orthosis

Muscles and movement 3
- Physiotherapist
  - In community provide regular therapy
  - Equipment assessment and funding
  - Independent skills - transfers/muscle strength
  - RCH service 6-12mly muscle mapping
- Orthopedic surgeries
  - Major reason for admission

Spina Bifida
Urology
- Bladder
- Bowel
- Skin

SB
Neurogenic Bladder
- 90% plus affected
- Inervation S2-4
- Associations
  - VU reflux
  - Renal Anomalies (horseshoe kidney)
  - Bladder wall trabeculation
  - UTI/pyelonephritis

SB Urology
Treatment
- Principles
  - Protect the kidney’s from damage
  - Social continence
- Management
  - Evaluate renal tract (imaging, urodynamics)
  - Regular imaging (deterioration silent)
  - Prevent infection (Bactrim, keflex, macrodantin, cranberry)
  - Treat infection
  - Investigate deterioration
  - Surgery
**SB Neurogenic Bladder**

- **Types**
  - Areflexic (acontractile)
    - Enlarged
    - Urine dribbles
    - Sphincter incompetent
    - common
  - Contractile (Spastic)
    - Small Volume
    - Thick wall

**Neuropathic Bladder 2**

<table>
<thead>
<tr>
<th>Bladder type</th>
<th>Contractile</th>
<th>Intermediate</th>
<th>Acontractile</th>
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</thead>
<tbody>
<tr>
<td>Detrusor contraction</td>
<td>Strongly</td>
<td>Continuous weak</td>
<td>Doesn't contract</td>
</tr>
<tr>
<td>Sphincter tone</td>
<td>High</td>
<td>Weak/ ineffective</td>
<td>Weak</td>
</tr>
<tr>
<td>Outflow</td>
<td>Obstructed</td>
<td>Obstructed</td>
<td>Mild resistance (incomplete relaxation)</td>
</tr>
<tr>
<td>Effects</td>
<td>High residual volume, small bladder capacity</td>
<td>Small capacity and continuous dribbling</td>
<td>Continuous dribbling</td>
</tr>
<tr>
<td>Upper tracts</td>
<td>At risk</td>
<td>At risk</td>
<td>Rarely at risk</td>
</tr>
<tr>
<td>Options</td>
<td>C.I.C, Sphincterotomy</td>
<td>C.I.C, Sphincterotomy</td>
<td>Anticholinergic, Botulinum ABD, C.I.C</td>
</tr>
</tbody>
</table>

**Neuropathic bladder 4**

1. Preserve renal function (ensure emptying and prevent infection)
2. Achieve continence

- **Medical**
  - Clean intermittent catheterisation
  - Anticholinergics (reduce detrusor hyperreflexia)
  - Manage infections
- **Surgical**
  - Botulinum toxin
  - Sphincterotomy
  - Bladder augmentation
  - Artificial urinary sphincter
  - Vesicostomy, urinary diversion/ undiversion
  - Mitrofanoff procedure

**SB Neurogenic Bladder Investigations**

- **Renal Ultrasounds**
  - Yearly
- **MCU**
  - prn
- **Nuclear Scans**
  - Baseline prn
- **Urodynamics**
  - Prn
- **Cystoscopy**
  - prn

**SB Urology Surgery**

- **Reflex surgery**
- **Vesicostomies**
- **high pressure, infection**
- **Bladder Augmentation**
  - Enlarge the bladder (mucus, rupture, cancer)
  - Flap from Ureter, stomach, bowel, cultured cells
  - Mitrofanoff - appendix for anterior access
  - Artificial Sphincters
  - Select candidates
- **Ileal Conduits**
  - Bleed, Stenosis, stones, Cancer, Last 10 years
SB Augmentation cystoplasty

SB Artificial Sphincter

SB Urology
Recent Developments
- Botox
  - High pressure bladder
  - Detrusor instability
- Nerve Transplants
  - Chinese research
  - Improved function
  - Trials underway in USA

SB Urology
Continence
- Initially nappies
- Prophalactic antibiotics
- Clean intermittent catheterization 3-5/day
- Pads
- Bokka Pants
- Condom drainage
- Medication eg oxybutinin
- Surgery

Spina Bifida
Neurogenic Bowel
- 90% + have faecal incontinence
- Sensory nerve to colon, rectum, anus
  - S2-4
- Autonomic nerves
- Major social problem

Neuropathic bowel 1
- 25% of adults with Spina Bifida are bowel continent
- Most who are incontinent have poor sensation
AND either
- Increased bowel outlet resistance
  - Constipation and overflow diarrhoea
- Decreased bowel outlet resistance
  - Frequent stools throughout the day
SB Neurogenic bowel

Types
- Patulous anus and constipation
- Diarrhoea
  - Diet sensitive
  - 10-20%
  - Loperamide
- Patulous anus and constant leakage
  - Seen in mobile (abdominal compression)

SB Neurogenic Bowel

Principles
- Aim for social continence
- No single program that works
- Bowel management starts from birth
- Try to have a program by school
- Teenagers and adults work it out

SB Neurogenic Bowel

Treatments
- Nappies initially
- Diet
- Laxatives often worsen but used.
  - Movicol
  - Timed Toileting
  - Pressure
  - Digital stimulation
  - Combinations
- Suppositories
- Microlax enema’s
  - Give 4-5 hour clean - good for school
- Bowel washouts
  - 2 or 3 per week
- Malone antegrade enema
- Anal Plugs
  - Work well in some 30%
  - Expensive, last 6 hrs
- Buttock strapping eg for swimming

SB Neurogenic Bowel

Malone procedure

SB Neurogenic Bowel

Malone/ Anal plugs
Malone Antegrade Washout

Bowel Washout Peristeen System

SB - Bowel Management Experimental
- Gracilis slings
- Electrical stimulation
- Cuff similar to Artificial Sphincter.

SB Pressure sores
- Skin Level
- dermatomes
- Mapped using pin prick
- Can be different to motor level
- Problem anaesthetic skin
- Pressure areas sitting braces
- Burns
- friction

SB Decubitus

Decubitus
### Spina Bifida Pressure Sores
- **Prevalence**: 20 – 25%
- **80 – 90% will have at some stage**
- **Cause**
  - Anaesthetic skin
  - Excessive pressure: 42%
  - Orthosis: 23%
  - Urine, faecal soiling: 23%
  - Friction: 10%

### SB Pressure sores
- **Prevention**
  - Pressure care training, regular lifts
  - Correct fitting orthosis
  - Pressure cushions
- **Site**
  - Low Lumbar, Sacral
  - Feet, buttocks
  - Thoracic, high lumbar
  - Spine, buttocks, feet

### Spina Bifida Latex Allergy
- Recognized since late 1980's
- 7 Due to intra abdominal surgery
- Most Sensitivity
  - Rash, lip swelling
  - Rubber gloves, catheters, balloons
- Some Anaphylaxis
- Risk during surgery
- **Prevalence 33% on RAST**
- **Treatment**
  - Test
  - Latex free theatre

### Spina Bifida Other Medical Issues
- **Obesity**
  - Especially in adolescence
  - Stop walking - wheelchair
- **Endocrine**
  - Precocious puberty, osteoporosis
- **Ophthalmology**
- **Psychological**
  - Adults suicidal ideation 10%

### Cognitive function
- Intellectual disability strongly associated with hydrocephalus
- General intelligence “normal range”
- Skewed to the lower end
- Specific Learning difficulties
- Verbal IQ > Performance IQ
- Relative Deficit Increases with age
  - Primary school 5 points down - overestimates
  - Secondary 10-15 points down
- Factors
  - Hydrocephalus, shunt blockage, infection
Spina Bifida
Cognitive profile
- Executive Functions
  - Difficulty completing task
  - Difficulty organizing task
  - Overwhelmed by complex new information
  - Impulsive
  - Difficulty applying new knowledge
- Higher order Language
- Information Processing (memory)
- Attention Memory, New learning
  - Poor sequential and working memory
  - Sequencing

Spina Bifida
Educational
- Most attend normal school
  - Special on cognitive grounds (Yooralla)
- Parent should visit school
- Usually require an integration aide
  - Continence
  - Cognitive
  - Safety
- School modifications (access, toilets)
- Program Support group
- School therapy

Spina Bifida
Secondary School
- Cognitive Difficulties more apparent
- Mobility/incontinence affect relationships
- Peer group issue – isolation
- Poor body image
- Sexuality issues
  - Impotence in males
  - Misery, low self esteem
  - Depression

Spina Bifida
Therapy
- Developmental program
  - Mainly physiotherapy
  - OT many have upper limb/ cognitive issue
  - Speech
- RCH initially or
- Specialist Children Services (home)
- Centre Based from 3y
- School based therapy

Spina Bifida
Cognitive profile
- Visuo motor Integration
  - Delayed laterality
  - Poor motor planning
  - Poor hand control
  - Slow mastery of writing
  - Untidy and disorganized presentation or written work
### Spina Bifida Therapy
- Motor development depends on level
- L3 and quads for walking
- Motor developmental program
  - Standing frames
  - Walking frames
  - Braces
  - Crutches
  - Wheelchair training

### Continence
- Continence nurses
  - Advise: management of the bladder and bowels
  - Training (CIC, washouts): parents, carers, aides and patients
  - Applications for CAAS funding
  - Sourcing equipment (catheters, pads, nappies, specialised underwear)
- Also Stomal Therapists and Wound Care!

### SB Social problems
- Friendship difficulties
- Realization that they are different 7-8y
- Exclusion by peers
- Unable to keep up in playground
- Reduced self esteem
- Body image problems

### Spina Bifida Post School
- Unrealistic expectations re careers
- Tertiary few
  - University a few.
  - TAFE
- Employment
- Independent Living

### Spina Bifida
- CLINIC STAFF
- Monitoring
  - Medical 6 monthly
  - Orthopaedic varies
    - Physio annually
  - Neurosurgery - annually
  - Urology 6 -12 monthly
    - Ultrasounds 1-2 years

### Transition
- When complete high school
- Sometimes ongoing orthopaedics or urology
- MECRS
- Monash
- Bendigo, Ballarat, Geelong
Spina Bifida Outcome
- Medical
- Psychological
- Employment
- Housing

Spina Bifida Adults Medical
- Urological
  - Recurrent UTI 38%
  - Hypertension 11%
  - Stomal Problems 58%
  - Review recommended 36%
- Orthopaedic
  - Backache 25%
  - Knee Instability 15%
  - Hip pain 14%
  - Shoulder Pain 10%
  - Arthritis 6%

Spina Bifida Adults Psychological
- Depression 5%
- Anxiety 3%
- Psychosis 2%
- Suicide attempts 7%
- Substance Abuse 7%

Spina Bifida Adults Education
- 114 Seen in 1992
- Tertiary 8%
- Completed High School 16%
- Special school 28%
- England 42% completed high school compared to 75% Controls (Tew 1984)
Spina Bifida Adults
Employment - Home
- Competitive Employment 33%
- Not Working 54%
- Living with parent 62%

Spina Bifida Adults
Pregnancy
- 17 women 23 pregnancies
- Less complications if Vaginal delivery
- Recurrent UTI
- Pyelonephritis
- Back pain
- Pressure sores

The End

i. Which tissues are involved?
- Spina bifida occulta (bone only)
  10% adult spines
- Meningocele (bone and meninges)
  6% SB, 11% survivors
- Myelomeningocele
  (bone, meninges, nerves)
  94% SB, 89% survivors

ii. Where is the lesion?
- 1% Cervical only (Level 1)
- 1% Thoracic only (Level 1)
- 6% Lower thoracic (Level 2)
  and Upper lumbar (Level 3)
- 92% occur at L3 and below
  42% Lumbosacral jct (Level 4)
  Lower sacral (Level 5)
Neuropathic Bowel 2
- Diet, fluids
- Regular sit, cough, push on toilet
- Laxatives, enemas, washouts
- Anal plugs
- Biofeedback
- Malone Procedure
(exclude other causes diarrhoea)

Myelomeningocele
Functional Ambulation
- Non Ambulator
- Household
- Indoor or outdoor
- Thoracic
- Wheelchair

- Community
- Thoracic - L3
- L3 - Sacral
- Wheelchair long distances

Sensation
- Sensory loss below level of lesion
  - Patchy
  - Dense
- High risk of pressure areas/burns
  - Slow healing (poor blood supply)
  - Historically, common cause of death

Occupational therapists
- Seating advise (includes car)
- Pressure care
- Tools for function/home modification
  (houlds, rails, handles, grips)
- Upper limb function
- Driver’s Licence

SB Orthopaedic
Ilizarov frame

SB - Prevention
Peri-conceptual folic acid
- Folic acid 0.5mg
- 2 month pre conception
- Family history 5mg
- Campaigns 1996, 1999

Spina Bifida
Initial Assessment
- Treatment considerations
- Severe paralysis
- Severe hydrocephalus
- Kyphosis
- Other congenital abnormalities
Spina Bifida - Numbers

- Treated at the RCH
  - 1970: 27
  - 1980: 23
  - 1990: 15
  - 1995: 5
  - 2000: 4
  - 2005: 4

Spina Bifida Embryology

[Diagram showing stages of development of the spinal cord and head at 21 days, 22 days, and 28 days.]