



Infants with Spina Bifida - What can you offer?

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What can you (we) offer?

Prevention

Prenatal diagnosis

Fetal surgery(?)

Neurosurgical

Orthopaedic

Urological

Medical (Neuro / Nephro)

Rehabilitative

School & learning support

Adolescent issues

Adult Issues

AIMS

- Minimize morbidity and improve survival
- Maximize functional independence
- Full and meaningful societal participation
- Better quality of life

Neurosurgical Repair

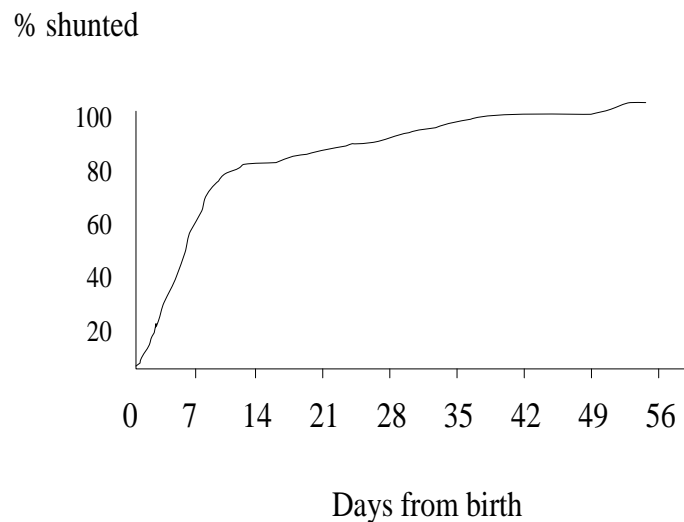


- Early repair of spinal defect
- Urgent if there is CSF leak
- Insertion of CSF reservoir at the same time

- Provide emotional support

Hydrocephalus and MMC

Timing of VP Shunting



- Hydrocephalus is often not apparent at birth
- 80-85% developed hydrocephalus that requires VP shunts
- CSF tapping from reservoir may reduce need for VP shunt

Managing the associated deformities



- CTEV
- Hip dislocation
- “Will my child walk?”



Management of Foot Deformities

- CTEV, Calcaneal and Equinus deformities
- Postnatal care
 - Careful manipulation to keep foot supple
 - Supple foot may be splinted
 - Meticulous skin care
- Orthopaedic surgery
 - Soft tissue release at 6/12
 - Tendon release, muscle transfer, etc
 - Maintain foot in corrected position with splint
 - Needs orthosis as soon as child starts to walk



“Will my child walk?”



Neurosegmental Level At Birth

Thoracic	Complete paraplegia
Upper Lumbar (L 1-2)	Some hip flexion and adduction
Mid-Lumbar (L 3-4)	Quadriceps (knee extension) with muscle power grade 3 or better
Lower Lumbar (L 4-5)	Foot dorsiflexion grade 3 or better No plantar-flexion
Sacral (S 1-2)	Normal dorsiflexion Moderate plantar-flexion Weakness of intrinsic foot muscles
S 3 or below	Normal motor and sensory function



Ambulation (± aids)

Level	% Walked	Age at walking	% stopped walking	Age stopped walking
Thoracic	20	4 yr 6 mo	43	6 yr 9 mo
High Lumbar	50	5 yr 2 mo	60	6 yr 11 mo
Mid Lumbar	60	5 yr	33	7 yr
Low Lumbar	84	3 yr 10 mo	13	9 yr
Sacral	100	2 yr 2 mo	0	NA

William et al, DMCN 1999



Chances in Malaysian Children

	Bowman et al (2002)	Ong LC et al (2002)
High (thoracic and L1/2)	0%	0%
Mid lumbar - L3/4	57%	65.6%
Low lumbar - L5	91%	
Sacral	100%	100%



Subsequent Care and Follow Up

- Best catered for by dedicated combined / multidisciplinary team (spina bifida clinic)
- Team members
 - Paediatricians (**neurologist, nephrologist**, community paediatrician)
 - **Paediatric urologist** / surgeon
 - Orthopaedic surgeon / rehabilitation physician
 - Continence or community nurse, etc



Assessment of Neurogenic Bladder

- Normal
- Detrusor hyperreflexia
- Acontractile bladder



Renal Failure

- Common cause of death
- Risk of renal failure is strongly related to sensory level
- Rare with sensory level at or below L4
- Common at or above T10
- However, it may occur even with spinal bifida occulta



Current Recommendation on Urological Management

- Aims: Preserve renal function and treat urinary incontinence
- Start CIC and oxybutynin soon after birth (one week after closure of spinal defect)
- Urodynamic study at 3-4 months old
- Continue oxybutynin if detrusor is overactive
- Continue CIC if sphincter is overactive



Urological Treatment

- Bladder augmentation
- Artificial sphincter
- Perineal urethrostomy
- Intravesical Treatment
- etc



Continence

Management of neurogenic bladder

- Should be started when a child normally attains bladder control.
- Depends on type of dysfunction.
- Managed by medication
 - toilet timing / training
 - clothing / pads / shields.
 - operations.



Bowel

- Avoid constipation
 - Regular + predictable timing
 - Managed by - high fiber diet
 - toilet timing / training
 - medication (laxative / enema)
 - manual evacuation
 - exercise
- Malone Antegrade Colonic Enema



Malaysian Children with Spina Bifida

Urinary Continence

- 24.5% dry all the time
 - 13.3% dry by day only
 - 62.2% wet all the time
-
- Low lesions were more likely to achieve continence

Faecal Soiling

- 51.1% continued to soil regularly

Diaper Use in 82.2%

- High lesion (90%)
- Intermediate (100%)
- Low lesion (59%)

Tethered Cord Syndrome



- New onset weakness of lower limbs
- Sensory loss
- Changes in gait
- Increasing spasticity
- Bowel/bladder function deterioration
- Back pain \pm radiation to the legs
- Progressive neurogenic scoliosis



Outcome After Detethering

	Pre-op	Post-op		
		Improved	Stable	Worsen
Scoliosis	10	5	5	-
↓ Muscle strength	3	3	-	-
Gait changes	8	6	1	1
LE contractures	3	1	2	-
Spasticity	6	5	1	-
Pain	5	5	-	-
Urinary changes	3	2	1	-

Bowman et al, 2001



Complications of Cord Detethering

~30% of patients experienced complications

- Loss of functional neurological levels
- CSF leak
- Wound infection
- Re-tethering of cord



Use of Orthosis / Walking Aids to support ambulation

- Ankle Foot Orthosis (AFO)
- Parapodium & Swivel Walker
- Posterior Walker
- Forearm Crutches
- Reciprocating Gait Orthosis



School and Learning Problems

- Faced by about **80%** of children

- Problems Include

- Poor eye-hand coordination

- Comprehension

- Attention deficit / Hyperactivity

- Memory

- Organization

- Mathematics

	742	742
	<u>- 136</u>	<u>- 136</u>
	614	616



School and Learning Problems

- Malaysian children
 - Normal class (62.5%)
 - Special class (20%)
 - Not schooling (17.55)

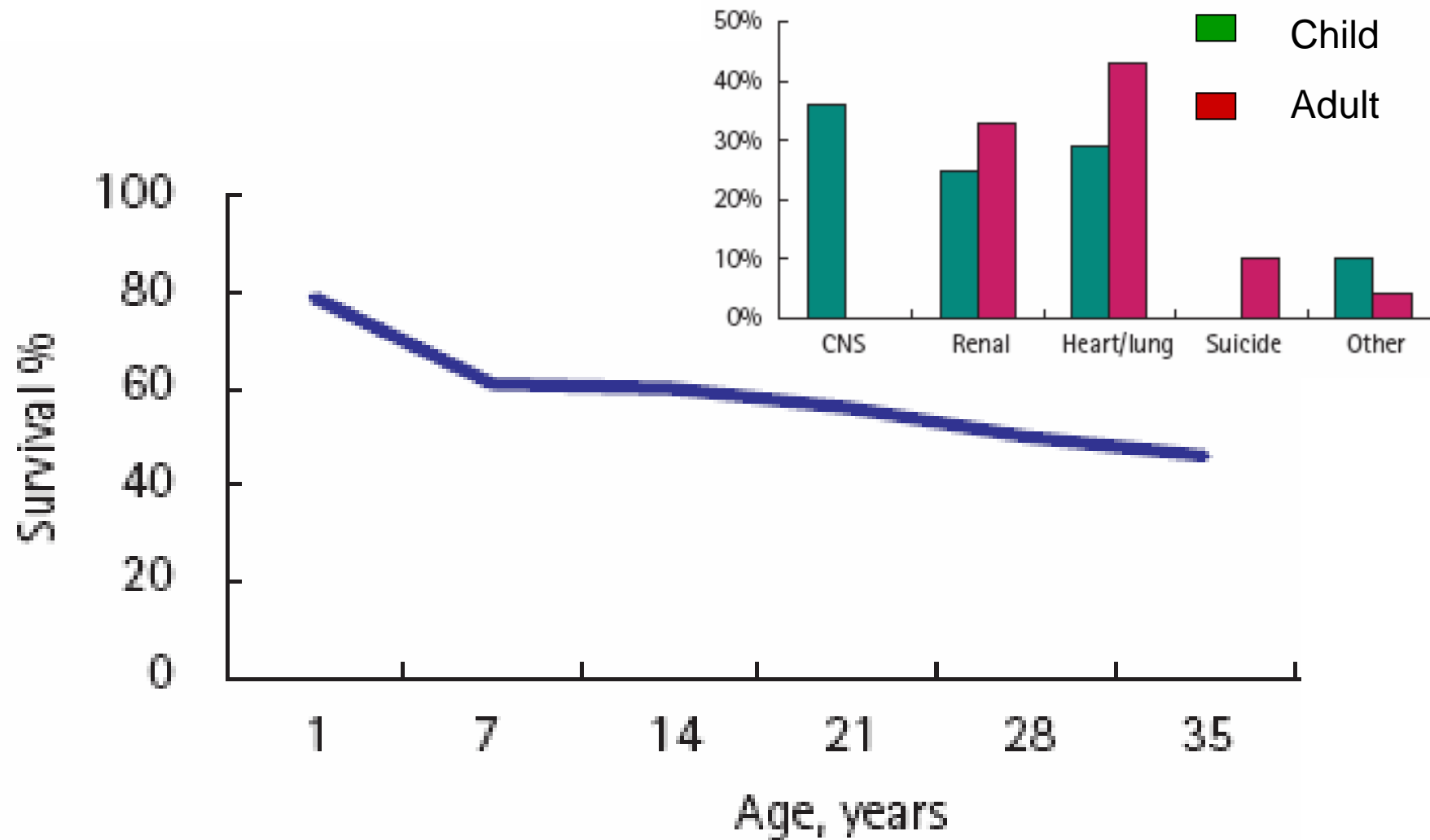
- Main Barrier
 - Incontinence (esp soiling)
 - Learning disability
 - Mobility and access still a main issue in Malaysia



Adolescent Issues

- Possible loss of ambulation
- Worsening of scoliosis & kyphosis
- Obesity
- Issues with body image and poor self esteem
- Rebelliousness
- Transition to adult-centred care

Survival & Cause of Death (UK)



Woodhouse CRJ, 2005



Maximizing Outcome in Adults

- Sexuality
 - Infertility
 - Common in males
 - Pregnancy

 - Independent living
 - Needs for assistance from caregiver
- Employment
 - Recreation
 - Access & community participation



Conclusions

- The prevalence of spina bifida is declining worldwide and ~ 50% of them is expected to live up to 35 years old
- Preservation of renal function could be achieved by more aggressive and early bladder management
- Functional independence and QoL issues need to be addressed and multisectorial involvement is important to achieve meaningful long term outcome.



Thank You