Unlocking potential:

Motor skills beyond ambulation in children with spina bifida and the relationship to lower limb muscle strength.

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Children's Health Queensland Hospital and Health Service pays respect to the Traditional Custodians of the lands on which we have the privilege to work on.

We acknowledge and pay our respects to Aboriginal and Torres Strait Islander Elders past, present and emerging.

Background



- Spina Bifida (Myelomeningocoele
 - MMC)
 - Congenital abnormality of the spinal cord
 - Failed neural tube closure
 - Paralysis and paresthesia below level of lesion
 - Delayed attainment of gross motor skills ^{1,2}

Ambulation in spina bifida

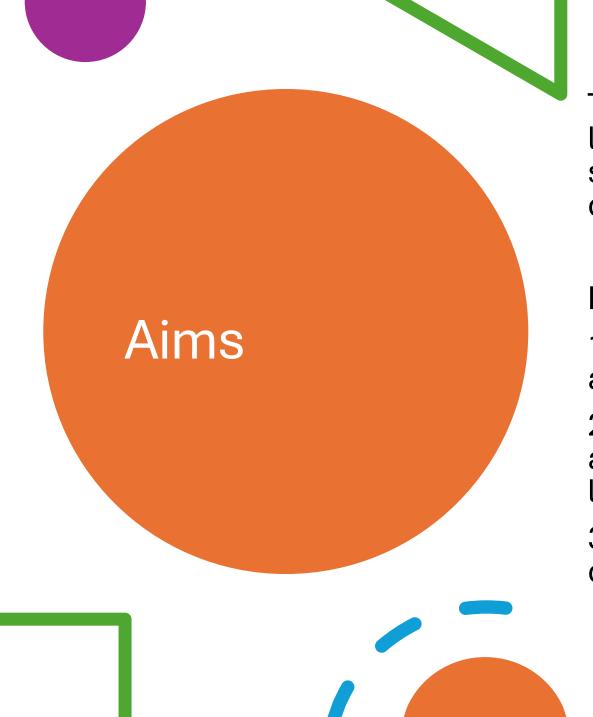
- Antigravity quadriceps ^{3,4}
- Pelvic stabilising muscles (hamstrings and hip abductors) ⁵
- Ankle stabilising muscles (ankle dorsiflexors and plantar flexors) ^{6,7}
- Thoracic and high lumbar non ambulant or with significant bracing
- Mid lumbar levels (L3-4) variable outcomes
- Sacral level community ambulation^{4,8}

What about other gross motor skills such as sitting, crawling, running and jumping??









To examine relationships between level of lesion, lower limb muscle strength and gross motor skills in children with myelomeningocoele.

Research questions:

- 1. What muscle strength is needed to achieve specific motor skills?
- 2. What motor skills can children achieve based on their level of lesion?
- 3. Can we predict motor skill in childhood based on MMT in infancy?

Clinical Measures

- Demographics & medical/surgical history
- Muscle Strength Assessment
 - Manual Muscle Testing (MMT) 9,10
 - Results for stronger leg used for analysis
- Gross motor skill assessment ¹¹
 - GMFM items and scoring criteria
- Neurological level of lesion
 - Broughton's classification ^{12,13}
- Functional mobility
 - Modified Hoffer's classification ¹⁴



Manual Muscle Testing

LO	WE	R LIMB	- MANUA	L N	MUSCLE TEST
U.R. Number :	F				PATIENT AGE: DATE OF MMT: REASON FOR MMT: PHYSIOTHERAPIST:
MUSCLES	÷	LEFT	RIGHT	÷	COMMENTS
QUADRATUS LUMBORUM T12-L1					
ABDOMINALS T8-T12					
LIOPSOAS L1-2					
SARTORIUS L1-3					
HIP ADDUCTORS L2-4					
QUADRICEPS L2-4					
MEDIAL HAMSTRINGS L4-S2					
ATERAL HAMSTRINGS L4-S1					
JLUTEUS MEDIUS L4-S1					
ELUTEUS MAXIMUS L5-S1					
TBIALIS ANTERIOR L4-L5					
TBIALIS POSTERIOR L4-L5					
PERONEUS LONG/BREVIS L5-S1					
PERONEUS TERTIUS L5-S1					
EXT HALLUCIS LONGUS L5-S1					
TOE EXTENSORS L5-S1					
LEX HALLUCIS LONGUS S1-S2					
OE FLEXORS S1-S2					
GASTROC SOLEUS \$1-\$2					

MUSCLE EXAMINATION QUALITY	UPPER LIMB STRENGTH
1. patient co-operative	
2 to a comment of full and discretional	

patient co-operative
 too young to follow directions/

not co-operative in some positions
3. patient unco-operative/crying results not necessarily accurate

MUCSLE STRENGTH TEST (MRC)
0-no palpable contraction
1-flicker or trace contraction
2-full ROM gravity eliminated
3-full ROM against gravity
4-full ROM against some resistance 5=full ROM against maximal resistance

GRIP STRENGTH

L1	L2	L3	L4	L5	<u>S</u> 1	52	53
4	SARTORIUS PECTINE			1	1	1	1
i	GRA	LONGUS	-	1	1	1	1
j	ADC	D BREWS CUCTORINA QUADRICE		1	I.	L	1
1			TRIAN	_		1	1
- 1	1	1	7	B POST N FAS LATA GLUT MEL	SAMIN .		l
- 1	ļ	- 1	3		DINOSUS	5	1
j	I	I		EXT HAL EXT D	KGE BAT	L	l
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- 1	1	1		1 4	ASTROCK SOLEUS A	MOPLANT	4
- 1	ļ	- 1		1	_	EMORIS IS MAX	7
1	1	- 1		l	FLEX H	FOOT IN	



Gross motor function

- **GMFM** scoring criteria
 - 5 static positions (sit, 4 point kneel, stand 3 or 20 sec, single leg stand)
 - 5 transition movements (4-point reciprocal crawl, pull to stand, cruise, walk hands held and walk unassisted)
 - 5 complex motor skills (walk with narrow base, walk along a line, run, jump, hop)
 - Additional motor skills (4-point non-reciprocal crawl, attain sit on small bench, attain sit on large bench)



Level of lesion



Non-Ambulant (NA)

• Uses a wheelchair for all mobility

Therapeutic Ambulant (TA)

- Walks in therapy often requiring bracing above the knee
- Wheelchair for most mobility inside and outside

Household Ambulant (HA)

- Walks indoors
- Wheelchair for all outdoors mobility, may use wheelchair for some indoor mobility

Community
Ambulant &
Wheelchair (CAW)

- Walks indoors and outdoors
- Uses wheelchair for distances

Community Ambulant (CA)

- Walks for all mobility indoor and outdoors without any aids
- Manages all distances

Ambulation (Modified Hoffer's Classification)

Procedure and Analysis

- Both assessments completed on day of clinic attendance
- Spearman's rank-order correlation co-efficient used to observe relationships between (p=<0.05)
 - Level of lesion and gross motor skill performance
 - Level of lesion and ambulation
 - MMT grade and gross motor skill performance
 - MMT grade and ambulation
- Non-parametric analysis required due to the ordinal nature of MMT and GMFM scores

Spearman's Correlation co-efficient (r_s) between

- 0-0.3 represents negligible correlation
- 0.3-0.5 is a low positive
- 0.5-0.7 is a moderate positive
- 0.7-0.9 is a high positive
- 0.9-1 is a very high positive ¹⁵

Results

Participants

- 42 children with MMC
- 4 16 years (Median age 8 yrs 1 month)
- Attended QCH Spinal Disabilities Clinic 2018 2023
- Ethics approval and parent consent

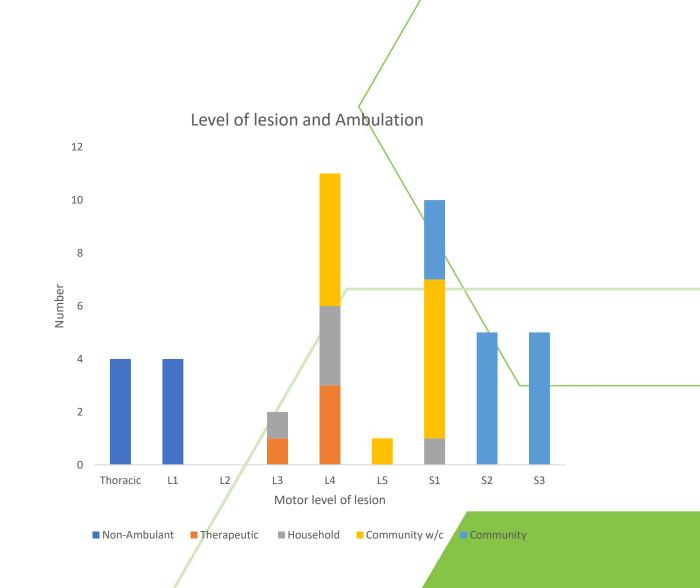
Excluded If:

- Unable to follow instructions
- History of orthopaedic surgery if preceding
 6/12
- Skin breakdown that precluded necessary assessment positions



Demographics

- 90% Arnold Chiari Malformation
- 76% VP shunt
- 5 % Antenatal closure
- 64% foot deformity
- 33% hip dislocation/subluxation
- 21% spinal deformity



Muscle strength, gross motor skills and ambulation

	4-point	Tran	nsitions	Independen	t Stand	Stepping/Walking			More complex gross motor skills Ambulation						
GMFM Item number	45	3	7 52		56	65			9 73	7.		7 5		0 / 82	2
	4 POINT: CRAWLS RECIPRO-CALLY FORWARD 1.8m	ON THE FLOOR: ATTAIN SIT ON LARGE BENCH	ON THE FLOOR: PULLS TO STD AT LARGE BENCH	STD: ARMS FREE 3 ST SEC 20	D: ARMS FREE SEC	STD 2 HANDS ON LARGE BENCH: CRUISE 5 STEPS	STD 2 HANDS HELD: WALKS FORWARD 10 STEPS	STD: WALKS FORWARD 10 STEPS	STD: WALKS FORWARD 10 STEPS BETWEEN PARALLEL LINES 20cm APART	STD: WALKS FORWARD 10 STEPS ON A STRAIGHT LINE	STD: RUNS 4.5m (15') STOPS & RETURN	STD: LIFTS FOOT ARMS FREE 10 SEC	STD: JUMPS 30cm (12') HIGH BOTH FEET	STD ON R FOOT: HOPS ON R FOOT 10 TIMES	MODIFIED HOFFER'S CLASSI- FICATION (NA, TA, HA, CAW, CA)
Iliopsoas L1-2	.795**	.784*	* .735**	.786**	.803**	.823**	.767**	* .737*	* .752**	.699*	.755*	* .727**	* .682*	* .618**	.723**
Quadriceps L2-4	.878**	.857**	* .809**	.733**	.677**	.846**	.739**	.620*	* .640**	.616**	.673*	* .624*	* .582*	* .505**	.683**
Medial Hamstrings L4- S2	.735**	.713**	* .649**	.828**	.878**	.754**	.840**	* .833*	* .864**	.805**	.847*	* .788**	* .811*	* .718**	.828**
Lateral Hamstrings L4- S1	.700**	.678**	* .612**	.832**	.903**	.695**	.811**	* .825*	* .905**	.841**	* .860**	* .824**	* .863*	* .828**	.851**
Gluteus Medius L4-S1	.623**	.578*	* .534**	.816**	.869**	.601**	.752**	* .813*	* .918**	.867**	* .865*	* .848*	.916*	* .827**	.858**
Gluteus Maximus L5-S1	.581**	.539*	* .498**	.846**	.883**	.537**	.777**	* .839*	* .914**	.872**	.883*	* .873**	* .903* ⁻	* .824**	.869**
Tibialis Anterior L4-L5	.545**	.506*	* .467**	.746**	.861**	.583**	.686**	* .767*	* .893**	.883**	* .834*	* .848*	.932*	* .872**	.858**
Tibialis Posterior L4-L5	.448**	.415**	* .384*	.710**	.858**	.479**	.599**	* .676*	* .869* [*]	.854**	.759*	* .829**	* .905*	* .826**	.788**
Peroneus Longus/Brevis L5-S1	.426**	.395*	.365*	.683**	.816**	.456**	.571**	* .646*	* .791**	.821**	* .723*	* .797**	* .851*	* .820**	.804**
Peroneus Tertius L5-S1	.466**	.432*	* .399*	.712**	.862**	.499**	.625**	* .707*	* .872**	.832**	.798*	* .823**	* .881*	* .858**	.831**
Toe Extensors L5-S1	.447**	.415**	* .383*	.668**	.842**	.478**	.598**	* .675*	* .866**	.819**	* .795**	* .812**	* .877*	* .911**	.802**
Toe Flexors S1- S2	.395**	.366*	.338*	.617**	.770**	.422**	.528**	* .596*	* .759**	.781*	* .732*	* .774*	* .818*	* .872**	.745**
Gastrocnemius S1-S2	.358*	.332*	.307*	.596**	.734**	.383*	.479**	* .541*	* .718**	.726**	.664*	* .774**	* .793*	* .818**	.773**

Question 1. What muscle strength is needed to achieve specific motor skills?



Pull to stand at large bench
hip flexors
adductors and
quadriceps



Reciprocal 4 pnt crawl – hip flexors, quads and hamstrings



Independent
walking
hip and knee flexors,
gluteals and ankle
dorsiflexors





More advanced gross
motor skills
pelvic stability muscles
and foot and ankle
muscles

Key gross motor skills and related muscle strength

Question 2. What motor skills can children achieve based on their level of lesion?

Expected motor skills based on level of lesion

Thoracic

Likely to achieve at least momentary sitting balance. May rely on upper limbs for support. May achieve transition from floor to small bench

Wheelchair for mobility

Mid lumbar (L3)

Likely to be able to reciprocal crawl, pull to stand and transition from floor to large bench. Needs support for standing and stepping

Ambulant with aids in therapy or home Wheelchair for community mobility

High sacral (S1)

Independent steps, can narrow base of support, runs. May have difficulty with static standing. Unlikely to be able to walk along a line, stand on one leg, jump or hop.

Ambulant in community. May use wheelchair for distances

High lumbar (L1-2)

Likely to attain sitting on a small bench and partially attain sitting on a large bench. Most can attain 4-point position and modified crawl for floor mobility Wheelchair for mobility

Low lumbar (L4-5)

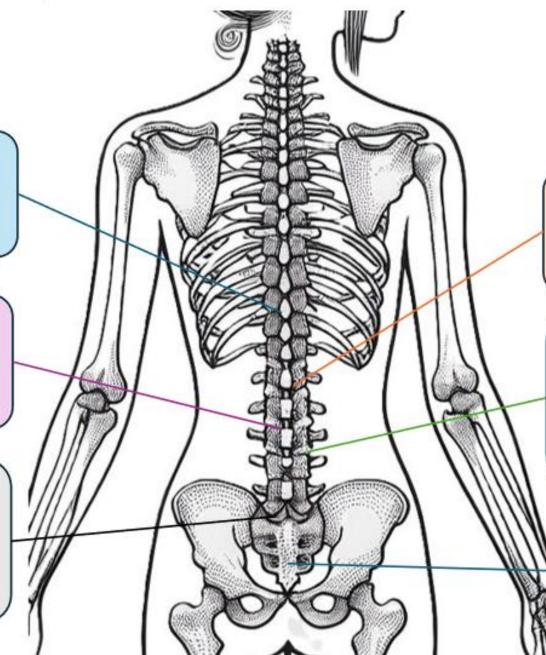
Can pull to stand and cruise, can manage a few steps with support. Difficulty standing still for >3 seconds. May take independent steps.

Ambulant with or without aids in therapy or home or community. Likely to use wheelchair for community mobility

Low sacral (S2-3)

Can walk, stand still, run. May be able to walk along a straight line, stand on one leg, jump and hop.

Ambulant in community. Does not need wheelchair for distances



Thoracic & high lumbar level lesions



Thoracic (weak abdominals and absent lower limb strength)

- May need upper limbs to prop in sitting
- Difficulty with independent floor mobility

High lumbar (abdominals, hip flexors +/- adductors)

Achieve 4 pnt kneel and modified crawl

Considerations for therapy:

- Will not achieve weight bearing with upper limb support need for standing frames
- Impact on fine motor and bimanual play need for supported seating
- Delayed floor mobility and impact on cognition, language and social development as well as spatial and topographical memory -Early introduction of wheeled mobility

Midlumbar levels (L3)

(Hip flexors, adductors, strong quads)

- Reciprocal crawl, pull to stand and transfer onto high bench
- Need support for standing and stepping
- Ambulate at home, require aids, orthotics and w/c mobility



Considerations for therapy:

Use weight bearing to achieve independence in pressure relieving activities and transfers
Upright activities may be for function, therapy or experiential

Low Lumbar (L4-5)



(Hamstrings +/- gluteus medius)

- Pull to stand, cruise, take steps
- Difficulty with static stand
- Walk with or without aids (w/c for community)

Considerations for therapy:

- AFOs improve gait pattern,
- Walking aids may reduce trunk sway
- Wheelchairs assist in functional mobility and fatigue management

Sacral Levels



High sacral (Glut max >grade 2, weak gastroc and foot muscles)

- Walk independently
- Difficulty walking along a line, single leg stand, jump and hop

Low sacral (Strong gastroc, may or may not have strong foot intrinsics)

- Can walk, stand still, run
- May walk along a line, jump, hop and single leg stand
- Only 1 child with S3 level lesion was able to achieve all motor skills

Considerations for therapy:

 The role of CNS and sensory impairments (tactile, visual, prioprioceptive and vestibular) have on motor control and gross motor skills Question 3.

Can we predict motor skill in childhood based on MMT in infancy?

Muscle strength in infancy and gross motor skills

- Subgroup n=19 (median age 6 yrs)
- MMT first 6 months of life
- Graded 0-3

		Ambulation		<	Key Gross Motor Items			
	Ambulant	Ambulant	Modified	ON THE	STD:	STD, 2	STD:	
	(therapeutic,	(household	Hoffer's	PULLS TO	MAINTAIN	HANDS	WALKS	
	household or	or	Classificatio	STD AT	S ARMS	HELD:	FORWARD	
	community)	community)	n	LARGE	FREE 20	WALKS	10 STEPS	
	Vs Non-	vs Non or		BENCH	SEC	FORWARD		
	ambulant	therapeutic				10 STEPS		
		ambulant						
Quadriceps L2-4	.998**	.573*	.563*	.726**	.401	.609**	.504*	
Medial Hamstrings L4-S2	.583*	.588*	.474*	.504*	.524*	.794**	.758**	
Lateral Hamstrings L4-S1	.530*	.754**	.586*	.475*	.637**	.898**	.900**	
Gluteus Medius L4-S1	.449	.783**	.821**	.309	.826**	.874**	.890**	
Gluteus Maximus L5-S1	.434	.757**	.620**	.299	.536*	.745**	.739**	
Tibialis Anterior L4-L5	.391	.862**	.742**		.767**	.987**	1.00**	



Need to look beyond strength of quadriceps when determining potential for ambulation in infants with MMC

Take home points

- Several positive relationships have been identified between strength of lower limb muscles and gross motor skills in children with spina bifida
- While quadriceps strength is important for achieving supported weight bearing, pelvic and ankle stability muscles appear to be necessary for achieving independent walking and more complex gross motor skills
- Understanding the relationship between muscle strength or level of lesion and gross motor skills may improve goal setting and planning for adaptive equipment in children with spina bifida



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References

- 1. Williams EN, Broughton NS, Menelaus MB. Age-related walking in children with spina bifida. Developmental Medicine And Child Neurology. 1999;41(7):446-9.
- 2. Bartonek Å. Motor development toward ambulation in preschool children with myelomeningocele---a prospective study. Pediatric Physical Therapy. 2010;22(1):52-60.
- 3. McDonald CM, Jaffe KM, Mosca VS, Shurtleff DB. Ambulatory outcome of children with myelomeningocele: effect of lower-extremity muscle strength. Dev Med Child Neurol. 1991;33(6):482-90.
- 4. Seitzberg A, Lind M, Biering-Sørensen F. Ambulation in adults with myelomeningocele. Is it possible to predict the level of ambulation in early life? Child's Nervous System: Chns: Official Journal Of The International Society For Pediatric Neurosurgery. 2008;24(2):231-7.
- 5. Asher M, Olson J. Factors affecting the ambulatory status of patients with spina bifida cystica. The Journal Of Bone And Joint Surgery American Volume. 1983;65(3):350-6.
- 6. Schoenmakers MAGC, Gulmans VAM, Gooskens RHJM, Helders PJM. Spina bifida at the sacral level: more than minor gait disturbances. Clinical Rehabilitation. 2004;18(2):178-85.
- 7. Bartonek A, Saraste H, Samuelsson L, Skoog M. Ambulation in patients with myelomeningocele: a 12-year follow-up. Journal Of Pediatric Orthopedics. 1999;19(2):202-6.
- 8. Bendt M, Bartonek Å. Walking in adults with spina bifida with respect to muscle function. European Journal of Physiotherapy. 2016;18(3):154-60.
- 9. Tan JL, Thomas NM, Johnston LM. Reproducibility of Muscle Strength Testing for Children with Spina Bifida. Physical & Occupational Therapy in Pediatrics. 2017;37(4):362-73.
- 10. Hislop HJ, Avers D, Brown M, Daniels L. Daniels and Worthingham's muscle testing: techniques of manual examination and performance testing. 9th ed. St. Louis, Mo.: Elsevier; 2014.
- 11. Russell D RP, Wright M, Avery LM. . GMFM (GMFM-66 & GMFM-88) User's Manual. 3 ed. London: Mac Keith Press; 2021.
- 12. Broughton N, Menelaus M, Cole W, Shurtleff D. The natural history of hip deformity in myelomeningocele. The Journal of Bone & Joint Surgery British Volume. 1993;75-B(5):760-3.
- 13. Bartonek A, Saraste H, Knutson LM. Comparison of different systems to classify the neurological level of lesion in patients with myelomeningocele. Developmental Medicine And Child Neurology. 1999;41(12):796-805.
- 14. Bartonek A, Saraste H. Factors influencing ambulation in myelomeningocele: a cross-sectional study. Developmental Medicine And Child Neurology. 2001;43(4):253-60.
- 15. Hinkle DE, Wiersma W, Jurs SG. Applied statistics for the behavioral sciences : hbk. 5th ed: Houghton Mifflin; 2003.