

# Malaysian Children with Spina Bifida: Relationship Between Functional Outcome and Level of Lesion

L C Ong, Y N Lim, A Sofiah

## ABSTRACT

**Purpose:** To determine the relationship between spinal lesion level and functional outcome in children with spina bifida.

**Methods:** Prospective observational study of 66 children who attended the Spina Bifida Clinic from 1994-1997. Data were obtained from serial physical examination and parent interview to determine mobility status, neurosensory deficits, continence and school placement.

**Results:** Eighteen (27.3%) had high-level (thoracic and thoracolumbar), 27 (40.9%) intermediate (low lumbar) and 21 (31.8%) low-level (sacral) lesions. Children with high-level lesions experienced more mobility problems (independent ambulation, balance and use of appliances), than those with intermediate or low-level lesions ( $p < 0.001$ ). 58.8% of children with low-level lesions used diapers, compared with those with high (90%) or intermediate (100%) level lesions ( $p = 0.005$ ), but there were no significant differences in the incidence of soiling or urinary incontinence among all three groups. There were no significant differences among the three groups in terms of school placement, skin breakdown, epilepsy or visual defect.

**Conclusion:** Although there is good correlation between the level of the lesion and mobility in children with spina bifida, other measures of functional outcome like continence and school placement are more difficult to predict. These data are important for realistic counselling of families with newborns with spina and planning long term rehabilitation resources.

**Keywords:** spina bifida, outcome, children

*Singapore Med J 2002 Vol 43(1):012-017*

## INTRODUCTION

The surgical treatment of spina bifida and associated hydrocephalus has ensured long-term survival in patients selected for treatment. Continued care of

these patients, however, is a long-term process with the attendant problems of motor deficits, incontinence, cognitive impairment and psychosocial adjustment. A multidisciplinary approach from health care professionals and other allied professions is required to help these patients and their families cope with these various problems from infancy, childhood, adolescence right up to adulthood.

Previous research in developed countries reported a large disparity in functional outcome<sup>(1-4)</sup>, although mobility was shown to have a good correlation with the neurosegmental level of the lesion<sup>(4-7)</sup>. In Malaysia, the functional outcome of children with spina bifida is largely unknown, and may differ from those reported because of differences in access to health care and educational policies.

The objective of this study was to obtain data on the functional outcome (in the areas of mobility, continence and education) of a group of Malaysian children with spina bifida, and to relate these with their level of lesion. This information will not only help in realistic counselling of parents with newborns with spina bifida, but also help plan for appropriate resources for long-term rehabilitation.

## METHODS

This is a prospective observational study of children who attended the Spina Bifida Clinic in Hospital Kuala Lumpur from 1994 to 1997. The clinic was set up in 1994 to facilitate the medical needs of these children. The clinic served as referral centre for children in the Klang Valley although there were referrals from the suburbs and other states as well. Due to space and time constraint, it had only a paediatric neurologist, nephrologist and urologist in attendance. Separate sessions were held with the neurosurgeon, orthopaedic surgeon but decisions in treatment (including liaison with the school and social authorities), were coordinated by the Spina Bifida Clinic. Only patients who had attended at least three sessions at the clinic (with complete neurological, urological and social assessment) and who had treatment plans initiated by the family were included in the analysis.

Department of  
Paediatrics  
Universiti  
Kebangsaan  
Malaysia  
Jalan Raja Muda  
Abdul Aziz  
50300 Kuala Lumpur  
Malaysia

L C Ong, MRCP (UK)  
Associate Professor

Y N Lim, MRCP (UK)  
Consultant Paediatric  
Nephrologist

A Sofiah, MMed  
Paeds (UKM)  
Consultant Paediatric  
Neurologist

Correspondence to:  
Professor Ong Lai Choo  
Department of  
Paediatrics  
Hospital Universiti  
Kebangsaan Malaysia  
Jalan Yaakob Latif  
56000 Kuala Lumpur  
Malaysia  
Tel: (603) 9170 2186  
Fax: (603) 9173 7827  
Email: onglc@  
mail.hukm.ukm.my

**Table I. Relationship between level of lesion and functional outcome in children with spina bifida.**

Outcome	High level (above L3) n=18	Intermediate level (L3-L5) n=27	Low level (Sacral) n=21	P* value
Ambulatory status				
Community	0	15 (65.6)	21 (100.0)	<0.001
Home	18 (100.0)	12 (34.4)	0	
Appliances required				<0.001
Orthoses only	2 (11.1)	10 (37.0)	0	
Wheelchair	16 (88.9)	16 (59.3)	0	
Balance problems				<0.001**
None	0 (0.0)	22 (81.5)	21 (100.0)	
Mild	8 (44.4)	5 (18.5)	0	
Moderate	7 (38.9)	0	0	
Severe	3 (16.7)	0	0	
Skin breakdown	3 (16.7)	6 (23.1)	1 (4.8)	0.220
Hydrocephalus	10 (55.6)	16 (59.3)	2 (9.5)	0.001
Shunted	9	14	2	
Shunt complications	3	6	1	
Epilepsy	1 (5.6)	1 (3.7)	1 (4.8)	0.957
Visual problems	3 (16.7)	2 (7.4)	1 (4.8)	0.401
Urinary problems				
Infections	14 (82.4)	21 (77.8)	18 (85.7)	0.777
Good stream	3 (16.7)	3 (11.1)	4 (19.0)	0.448
Clean intermittent	8 (44.4)	13 (48.1)	14 (66.6)	0.248
Catheterisation				
Dry day & night	1	2	3	
Dry by day	0	2	3	
Wet all the time	7	9	8	
Bowel problems				
Constipation	12 (66.7)	23 (85.1)	12 (57.1)	0.09
Laxatives/enema	10 (56.6)	20 (74.1)	10 (47.6)	0.155

Figures in parentheses indicate percentages

\* Chi-square analysis with two degrees of freedom

\*\* None and mild vs moderate and severe

The level of the lesion was determined by the level of motor involvement, and categorised as high (thoracic and thoracolumbar down to level of L2), intermediate (L3 to L5) and low (sacral segments only). Outcome was assessed according to neurosensory deficits (hydrocephalus, epilepsy, shunt placement, visual problems), mobility (ambulation, balance, use of appliances), urinary problems (urinary infections, use of intermittent catheterisation, continence), bowel problems (constipation, soiling, regular use of laxatives/enemas and diapers) and educational placement (normal school, special schools for the handicapped, dropped out from school). A child was considered to be a community ambulator if he or she managed to walk independently with or without the use of braces, crutches or walkers. Passive use of a stroller or wheelchair, walking only in the home environment or during therapy sessions was not considered community ambulation. These, and children who crawled about in the home

without attaining an upright position, were considered home ambulators. Balance problems were assessed with the child performing fine motor tasks in an upright position in a wheelchair; these were categorised as none, mild (able to use both hands simultaneously even with attendant scoliosis or kyphosis), moderate (needed to stabilise trunk with one hand on wheelchair) and severe (unable to maintain upright position or required both hands for stabilisation). Appliances referred to any physical aids required for ambulation (braces, orthoses, rollators, wheelchairs).

For urinary incontinence, a child was considered reliably dry if accidental wetting (severe enough to warrant changing of diapers, pads or underpants) occurred less than once a week. Soiling was said to occur on a regular basis if it occurred more than once weekly. Chi-square test (or Fisher exact test for cell values less than five) was used for analysis and a p value of less than 0.05 was considered statistically significant.

**Table II. Relationship between level of lesion and continence in children with spina bifida above four years of age.**

	High level n=10	Intermediate level n=18	Low level n=17	P* value
Urinary control				
Dry day & night	1 (10.0)	4 (22.7)	6 (35.3)	
Dry by day	1 (10.0)	2 (11.1)	3 (17.6)	0.206**
Wet all the time	8 (80.0)	12 (66.7)	8 (47.1)	
Soiling	6 (60.0)	9 (50.0)	8 (47.1)	0.804
Use of diapers	9 (90.0)	18 (100.0)	10 (58.8)	0.005

Figures in parentheses indicate percentages

\* Chi-square analysis with two degrees of freedom

\*\* None and mild vs moderate and severe

## RESULTS

There were 66 children who attended the clinic on a regular basis during that period, with ages ranging from two to 17 years (median six, inter quartile range three to nine years). Thirty-two (48.5%) were males; 29 (43.9%) were Malays, 19 (28.8%) Chinese and 18 (27.2%) Indians. Eighteen (27.3%) had high, 27 (40.9%) intermediate and 21 (31.8%) low motor level lesions.

Table I shows the relationship between the level of lesion and the functional status of the patients. Thirty-six (54.6%) were community ambulators, of whom 31 (86.1%) walked independently without the use of any aids. Of the 30 children whose ambulatory activities were limited to their home, 13 (43.3%) had no functional use of their lower limbs and mobilised by crawling, creeping and required assistance with transfer from wheelchair to bed. There was a significant difference in the ambulatory status between the three groups – all children with low level lesions and about half of those with intermediate level lesions were community ambulators, whereas all children with high level lesions were home ambulators. Of the 12 children with intermediate level lesions who were home ambulators, 5 (41.7%) had severely limited mobility, requiring help in and out of their wheelchairs. All 10 children who experienced moderate or severe problems with balance had high level lesions whereas none of the children with low or intermediate level lesions had balance problems severe enough to interfere with hand function.

Forty-four (66.7%) children in the study group required an appliance for mobility, of which 32 (72.7%) used wheelchairs on a regular basis. None of the children with low level lesions used any appliances; in contrast all children with high level lesions required one, the majority of whom (88.9%) used wheelchairs. Of the 26 children with intermediate level lesions who required an appliance, 16 (61.5%) used a wheelchair. Ten (15.2%) children developed skin breakdown at some point during the study period. Although the incidence of skin breakdown among the children

with low level lesions was lower than those with intermediate or high level lesions, this did not reach statistical significance.

Forty-nine (74.2%) patients suffered from constipation, and 43 (65.1%) required laxatives, enemas or practised manual evacuation on a regular basis. These measures were not mutually exclusive, as many children incorporated two or more such measures in their toileting regime. Although children with intermediate level lesions had the highest incidence of constipation (with consequent need for laxatives, enemas or manual evacuation), these differences were not statistically significant.

Fifty-three (80%) children had a documented urinary tract infection necessitating treatment at some point during the study period, with similar rates of infection among the three levels of lesions. Only 10 (15.2%) children reported a good stream of micturition; again there was no significance difference among the three groups in terms of dribbling or poor stream of micturition. Thirty-five (53%) children practised regular clean intermittent catheterisation (CIC); of whom 5 (14.3%) were reliably dry at all times, 6 (17.1%) were dry only during the day and 24 (68.6%) remained wet at all times. There was no significant difference in the rate of CIC use among the three groups. Among the children who practised CIC, those low-level lesions were most likely to remain dry at least during the day (42.8%), compared to those with intermediate (30.8%) and high (12.5%) level lesions. However, the number of children who achieved continence was small and these differences were not statistically significant.

Twenty-eight (42.4%) children had hydrocephalus, of whom 25 (89.3%) underwent a shunting procedure in early infancy. Nine (36%) of those shunted had a shunt malfunction (blockage with or without concomitant infection) at some point in life. Children with low-level lesions were less likely to develop hydrocephalus than those with intermediate or high-level lesions, but the rates of shunting and shunt

**Table III. Relationship between school placement and level of lesion in children with spina bifida.**

School placement	High level n=10	Intermediate level n=15	Low level n=15
Normal school	6 (60.0)	7 (46.7)	12 (80.0)
Integrated class for slow learners	0	3 (20.0)	2 (13.3)
School for handicapped	1 (10.0)	2 (13.3)	0
Dropped out	3 (30.0)	3 (20.0)	1 (6.7)

Figures in parentheses indicate percentages

complications were similar for children with hydrocephalus in all three groups. The incidence of epilepsy (4.5%) and visual defects (9.1%) was low in this study, with no significant difference among the three groups.

Assuming that normal Malaysian children would have achieved social continence (dry at least during the day, no soiling) by four years of age, a sub-analysis of bowel and continence among those aged above four years was carried out (Table II). Out of these 45 children, 11 (24.5%) were dry at all times, 6 (13.3%) were dry by day only, and 28 (62.2%) were wet all the time. Children with low-level lesions were more likely to achieve urinary continence (52.9%), compared to those with intermediate-level (33.2%) and high-level lesions (20%). These differences, however, were not statistically significant. Twenty-three (51.1%) of children aged over four years continued to soil on a regular basis, with no significant differences among the three groups. Thirty-seven (82.2%) children aged above four years wore diapers daily; with a significantly lower rate of diaper usage among children with low-level lesions as compared to those with intermediate or high-level lesions.

Forty (60.6%) children in the study were of school-going age (Table III). Of these, 25 (62.5%) were attending normal class, 8 (20%) attended either special classes in integrated schools or schools for the handicapped while 7 (17.5%) did not attend school at all. Seven (70%) of the 10 children with high-level lesions attended school, compared to 12 (80%) of the 15 children with intermediate-level lesions and 14 (93.3%) of the 15 children with low-level lesions. These differences were not statistically significant ( $p = 0.306$ ). More children with low-level lesions attended mainstream school (normal or integrated classes) compared to those with intermediate or high-level lesions (93.3% vs 64%), but this did not reach statistical significance ( $p = 0.059$ ). The children with high or intermediate-level lesions cited multiple problems (transport problems, inaccessible classrooms and toilets, social stigma of wetting and soiling) as the reasons for not

attending school. The sole child with low-level lesion who was not schooling was an adolescent who cited poor academic performance and difficulty with proper toilet access to perform CIC as the reasons for dropping out of school.

## DISCUSSION

The relationship between independent ambulation and the neurosegmental level of the lesion in spina bifida is well documented. Like other studies, all our patients with sacral level lesions walked independently without aids<sup>(1,4-8)</sup>. Long-term studies indicate that this pattern remained stable right into early adulthood<sup>(1,5)</sup>. None of our patients with high-level lesions and two thirds of those with intermediate-level lesions achieved community ambulation. This is in contrast to other studies which reported ambulation rates of 8 - 11% and 80% among those with high and intermediate-level lesions respectively<sup>(1,4-6,8,9)</sup>. The discrepancy may be due to the contrasting management philosophies and resources available at different centres. Improved orthopaedic procedures and high quality orthoses enabled many children with high and intermediate-level lesions in developed countries to achieve walking<sup>(9)</sup>. These facilities are not readily available in Malaysia and might explain the much higher rate of wheelchair use in this study. A study of South African children with myelomeningocele<sup>(7)</sup> noted that ambulation was more likely in children from urban areas, citing better access to orthopaedic surgery and maintenance of appliances as possible factors. It should be noted, however, that cessation of walking eventually occurred in all patients with high-level lesions. This usually occurs in the second decade<sup>(4,6)</sup> but has been reported to occur as early as seven to nine years of age<sup>(5)</sup>. It appears that, irrespective of the extent of orthopaedic and orthotic intervention, the walking ability in adulthood is ultimately dependent on the underlying neurologic deficit. In this aspect, the philosophy that mobility in a wheelchair outweighs the importance of upright "walking", provides a more pragmatic goal in the Malaysian setting. The ambulatory status of those

with intermediate-level lesions are much more difficult to predict, due to the complex interplay of factors such as availability of orthoses, spinal cord tethering, weight gain and motivation in an individual patient.

The incidence of hydrocephalus, epilepsy and visual problems was much lower in this study compared to others<sup>(1,3,7,10)</sup>. This may reflect different selection procedures for early aggressive intervention in the newborn with spina bifida who develops hydrocephalus. Selection criteria which precluded shunting of those with massive hydrocephalus would leave a cohort of less disabled survivors. The other reason for the relatively high rate of disability in other series could be that these patient cohorts were from an earlier era (1960s), when surgery was less sophisticated<sup>(1)</sup>. Like others<sup>(1,4,7)</sup>, the incidence of hydrocephalus was higher among children with thoracic or lumbar lesions than those with sacral lesions. However, the long-term impact on intellectual ability in these children is determined by the occurrence of shunt dysfunction rather than the presence of hydrocephalus per se<sup>(1,4,7)</sup>.

The overall incidence of skin breakdown in this study was lower than others<sup>(1,11)</sup>. As the cumulative incidence of skin breakdown is known to increase with age<sup>(11)</sup>, the relatively younger age of the patients in our study might explain the discrepancy found. This study did not demonstrate a higher skin breakdown rate among patients with high-level lesions. Okamoto et al<sup>(11)</sup> also found the rate of skin breakdown was similar irrespective of the level of the spinal lesion, although the location (perineum/gibbus as opposed to ankle/foot) depended on the level of paraplegia.

Half of our patients practised regular CIC, a rate comparable to some studies on children<sup>(2,12-14)</sup>. This is higher than that of older series<sup>(1,10)</sup>, in whom many patients had urinary diversion procedures before CIC became the standard mode of therapy. Of interest is that the rate of continence never exceeded 50%, even among those practising CIC. Other studies<sup>(2,9,12,15)</sup> have also noted the dismal rate of achievement of continence. Uehling et al<sup>(14)</sup> reported a much higher success rate (81%), but this was among patients who managed to continue CIC for more than five years. Like other studies<sup>(15)</sup>, we found that the incidence of urinary problems, including continence and the need for CIC, did not differ among patients with different levels of lesion.

Sensory loss<sup>(1)</sup>, urodynamic pattern of detrusor-sphincter dysfunction<sup>(13)</sup>, social class and urban living<sup>(7)</sup> appear to determine continence more than the level of the lesion. Although proper urodynamic evaluation

allows for more rational use of CIC, pharmacological agents and urosurgery, other social factors (family compliance, mobility and access to toilet facilities, age and cognitive ability) are equally important in helping these patients achieve social continence. As with other studies<sup>(10,12,16,17)</sup>, half of our patients soiled regularly. The level of the spinal lesion did not appear to influence the incidence of soiling. Studies have shown that, barring physical problems (spastic extremities, mental retardation), regular toilet training with adjuvant therapy (laxatives, enemas and manual evacuation) from an early age can produce good results and prevent the development of secondary megacolon/rectum<sup>(3,4,17,18)</sup>. Although patients in this study with low level lesions had a lower rate of diaper use, more than half of this group still used diapers on a regular basis. This highlights the need for improved measures to achieve both urine and bowel continence, irrespective of the level of the lesion.

Two-thirds of our patients managed to attend normal school, a rate comparable to others<sup>(2,3,12)</sup>. Incontinence (especially soiling) and learning disability, rather than physical disability, has been cited as the main obstacle to mainstream schooling<sup>(1,2,19)</sup>. In this study, more children with low-level lesions attended mainstream school than their counterparts with higher lesions. Although this did not reach statistical significance, the small numbers involved could have resulted in a beta error. Feedback from the patients indicated that mobility and access are still major issues for Malaysian school children with spina bifida. There is a need for better commitment from the social and education authorities if the policy of integration is to succeed, especially for those who are physically but not cognitively disabled.

In conclusion, ambulation could only be accurately predicted for children with low and high-level lesions. Those with intermediate-level lesions had variable outcome and mobility appeared to be influenced by factors other than the neurological deficit per se. There did not appear to be a clear association between urinary or bowel continence and the level of the lesion. The low rate of achievement of continence warrants improved medical and social care, irrespective of the level of the lesion. Although the majority of children (including those with high-level lesions) were attending mainstream school, much still needs to be done to improve access and facilities for the physically disabled. As improved medical care results in more children with spina bifida surviving beyond infancy, the results of this study may help towards realistic counselling of their families and improved rehabilitation resources to improve the functional outcome of these children.

## REFERENCES

1. Hunt GM Poulton A. Open spina bifida: a complete cohort reviewed 25 years after closure. *Dev Med Child Neurol* 1995; 37 (1):19-29.
2. Redaelli T, Cassinis A, Cosignani F, Conti B, Onofri MP, Dall'Acqua A. Interdisciplinary treatment of spina bifida children. *Paraplegia* 1992; 30:683-9.
3. Steinbok P, Irvine B, Cochrane DD, Irwin BJ. Long-term outcome and complications of children born with myelomeningocele. *Childs Nerv Syst* 1992; 8 (2):92-6.
4. Shurtleff DB, editor. *Myelodysplasias and Extrophies: Significance, Prevention and Treatment*. Orlando: Grune and Stratton, 1986.
5. Williams EN, Broughton NS, Menelaus MB. Age related walking in children with spina bifida. *Dev Med Child Neurol* 1999; 41:446-9.
6. McDonald CM, Mosca VS, Shurtleff DB. Ambulatory outcome of children with myelomeningocele: effect of lower limb strength. *Dev Med Child Neurol* 1991; 33:483-90.
7. Buccimazza S, Molteno C, Dunne T. Pre-school follow-up of a cohort of children with myelomeningocele in Cape Town, South Africa. *Annals Trop Paeds* 1999; 19 (3):245-52.
8. Stillwell A, Menelaus MB. Walking ability in mature patients with spina bifida. *J Pediatr Orthop* 1983; 3 (2):184-90.
9. Charney EB, Melchionni RN, Smith OR. Community ambulation by children with myelomeningocele and high level paralysis. *J Pediatr Orthop* 1991; 11:579-82.
10. Malone PS, Wheeler RA, Williams JE. Continence in patients with spina bifida: long term results. *Arch Dis Child* 1994; 70:107-10.
11. Okamoto GA, Lamers JV, Shurtleff DB. Skin breakdown in patients with myelomeningocele. *Arch Phys Med Rehabil* 1983; 64 (1):20-3.
12. Kalucy M, Boer C, Stanley F. School-aged children with spina bifida in Western Australia parental perspectives on functional outcome. *Dev Med Child Neurol* 1996; 38:325-34.
13. Knoll M, Madersbacher H. The chances of a spina bifida patient becoming continent/socially dry conservative therapy. *Paraplegia* 1993; 31 (1):22-7.
14. Uehling DT, Smith J, Meyer J, Bruskewitz R. Impact of an intermittent catheterisation program on children with myelomeningocele. *Pediatrics* 1985; 76 (6):892-5.
15. Merovach RA, Bogaert GA, Baskin LS, Lazzaretti CC, Edwards MS, Kogan BA. Lower urinary tract function in ambulatory children with spina bifida. *Br J Urol* 1996; 77: 593-6.
16. Purcell MH, Gregory JG. Intermittent catheterisation: evaluation of complete dryness and independence in children with myelomeningocele. *J Urol* 1984; 132 (3):518-20.
17. King JC, Currie DM, Wright E. Bowel training in spina bifida: importance of education, patient compliance, age and anal reflexes. *Arch Phys Med Rehabil* 1994; 75:243-7.
18. Okamoto GA, Souza J, Telzrow RW, Holm RA, McCartin R, Shurtleff DB. Toileting skills in children with myelomeningocele: rates of learning. *Arch Phys Med Rehabil* 1984; 65 (4):182-5.
19. Wasson CM, Bannister CM, Ward GS. Factors affecting the school placement of children with spina bifida. *Eur J Pediatr Surg* 1992; 2 (Suppl 1):29-34.