Low-lying spinal cord and tethered cord syndrome in children with anorectal malformations

Andrew Teck Kwee Teo[1], MBBS, MRCSE, *Bin Kee Gan*[1], MMed, MRCP, *Janice Su Zhen Tung*[1], MBBS, *Yee Low*[2], FRCSE, FAMS, *Wan Tew Seow*[1], FRACS, FAMS

INTRODUCTION Anorectal malformations (ARMs) and low-lying spinal cord (LLC) are commonly associated owing to their common embryonic origin. LLC may lead to tethered cord syndromes (TCS), requiring surgery. This study aimed to review the incidence of LLC in children with ARMs using ultrasonography (US) and magnetic resonance (MR) imaging, the incidence of TCS and the surgical outcomes of these patients after detethering.

METHODS We conducted a retrospective study of children who underwent surgery for ARMs in 2002–2009 at KK Women's and Children's Hospital, Singapore.

RESULTS Out of 101 (16.8%) ARM patients, 17 had LLC, of which 12 (70.6%) were high ARMs. 12 of the 17 (70.6%) patients had abnormal US and MR imaging findings. Five (29.4%) had normal US but abnormal MR imaging results; in these five patients, MR imaging was performed due to new symptoms and equivocal US findings. These 17 patients subsequently underwent surgical detethering. Three out of seven patients with TCS improved after surgery. None of the 17 patients had any complications.

CONCLUSION LLC appeared to be associated with high ARMs, although this was not statistically significant. LLC should be investigated for whenever ARM is diagnosed, regardless of its type. Lumbar US is useful for first-line screening for LLC. Abnormal US or onset of new symptoms should subsequently be investigated with MR imaging. Equivocal US findings are also likely to benefit from further MR imaging. Surgery to detether LLC can improve outcome in TCS, while prophylactic detethering for asymptomatic patients with lipoma of the filum terminale has very low surgical risk.

Keywords: anorectal malformation, low-lying spinal cord, surgical outcome, tethered cord, tethered cord syndrome
Singapore Med J 2012; 53(9): 570–576

INTRODUCTION

A low-lying spinal cord (LLC) is defined as the conus medullaris ending below the L2 vertebrae.[1,2] An LLC is usually abnormally fixed to a caudal structure such as a lipoma or scar, which limits caudal-cranial movement.[3] This may be attributed to tethering of the spinal cord. The association between anorectal malformations (ARMs) and LLC is well described.[3] It arises due to their common embryonic origin, the caudal cell mass in the gastrula,[2] which is involved in secondary neurulation. The association of LLC with the types of ARMs is, however, less elaborated on[4] and also controversial.

The two main diagnostic imaging modalities for LLC are lumbar ultrasonography (US) and magnetic resonance (MR) imaging of the lumbar spine. US is a good screening tool, and although MR imaging is probably the diagnostic imaging of choice,[4] it is not feasible to perform it for every child with ARM to detect LLC. The extent to which US and MR imaging are used to detect LLC thus varies in different centres. The choice of diagnostic imaging is important because LLC may lead to tethered spinal cord syndrome (TCS), resulting in neurological, musculoskeletal, urological or gastrointestinal abnormalities.[1] There is thus a need to recognise and institute early management, which includes surgical detethering for patients with symptomatic TCS. Management of asymptomatic patients is more controversial, as some surgeons advocate prophylactic detethering even before the onset of TCS, while others advocate conservative management.[3]

This study reviewed the incidence of LLC in children with ARMs who were diagnosed using US and MR imaging, and evaluated whether LLC is associated with certain types of ARMs. We also studied the role of US and MR imaging in order to determine the best diagnostic imaging strategy in the diagnosis of LLC in ARMs. Finally, we reviewed the surgical outcomes to evaluate the effectiveness of surgical detethering to improve the functional outcomes of TCS, and thus the role of prophylactic detethering.

METHODS

We conducted a retrospective study of children with ARMs who were investigated for LLC at the KK Women's and Children's Hospital (KKH), Singapore. Approval for this study was obtained from and granted by the Singapore Health Services Institutional Review Board. The inclusion criteria for our retrospective study were all children who had a lumbar US performed and who were diagnosed with and underwent surgery for ARMs in 2002–2009 at KKH. The case records, lumbar US and MR imaging lumbar spine reports, urodynamic study (UDS) reports and operative records were reviewed. Children who did not undergo

[1]Department of Paediatric Neurosurgery, [2]Department of Paediatric Surgery, KK Women's and Children's Hospital, Singapore
Correspondence: Dr Seow Wan Tew, Senior Consultant and Head, Department of Paediatric Neurosurgery, KK Women's and Children's Hospital, 100 Bukit Timah Road, Singapore 229899. seow.wan.tew@kkh.com.sg

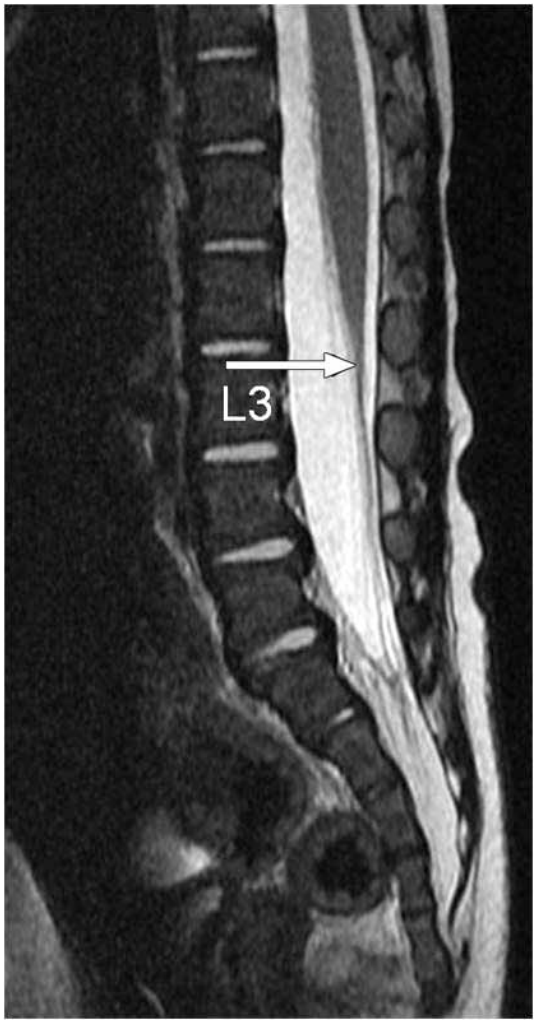

Fig. 1 Sagittal-T2 MR image shows a low-lying conus (arrow) that ends at L3.

a documented lumbar US were excluded. The assessment and diagnosis of ARM were made by a paediatric general surgeon. ARMs were classified into four main groups (low, intermediate, high and cloacal or rare anomalies) according to the Wingspread classification (WC).[5] For our study, the patients were subsequently divided into three ARM groups so that comparison of our data could be made with other similar studies. The groups were high-type ARMs (which included high or intermediate ARMs in WC), low-type ARMs (which were low in WC) and those with cloacal anomalies.

The presence of LLC on US and MR imaging was evaluated and reported by a radiologist. The MR images included axial and sagittal T1- and T2-weighted images. MR images of LLC and lipoma of the filum are shown in Figs. 1 and 2, respectively. The patients were reviewed by the paediatric neurosurgeon, who confirmed the radiologic diagnosis of LLC. The diagnosis of TCS was made based on the presence of any urodynamic dysfunction on UDS performed by a paediatric general surgeon or the presence of neurological signs, including urinary incontinence on clinical examination performed by the paediatric neurosurgeon. Subsequent detethering of the spinal cord, both for symptomatic TCS as well as prophylactic for asymptomatic non-TCS, was performed by the same paediatric

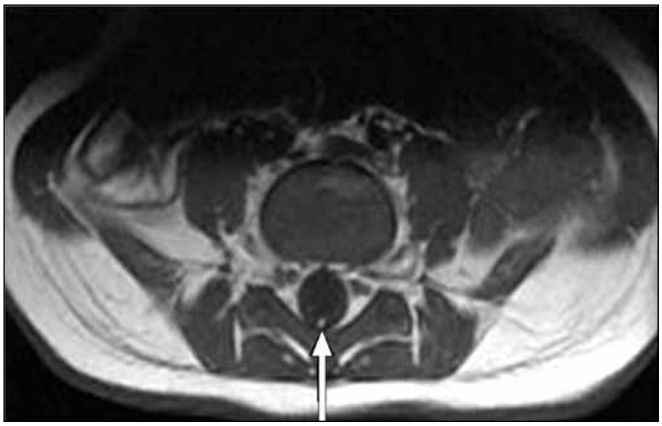

Fig. 2 Axial-T1 MR image shows lipoma of the filum terminale (arrow) as a high-signal intensity round structure in the thecal sac.

neurosurgeon. The outcome measures used to evaluate surgical detethering were any improvements in postoperative urodynamics and previous symptoms such as neurological deficits, as well as any postoperative complications.

Bivariate relations between LLC, non-LLC, TCS and non-TCS with each of the low-type, high-type and cloacal ARMs were examined using chi-square tests. The tests were evaluated at $p = 0.05$ level of significance. Statistical analyses were performed using the Statistical Package for the Social Sciences version 16.0.2 (IBM Corporation, Armonk, NY, USA). The sensitivity and specificity of US were evaluated based on the assumption that for patients who underwent further MR imaging, the findings were diagnostic of whether LLC was present or absent. Patients who did not undergo MR imaging were assumed to be non-LLC on initial US, and those who were not referred back to the hospital for neurological deficits or who did not have further abnormal imaging were definitively non-LLC cases. Five children had LLC on US but did not undergo further MR imaging. Two of these children had congenital cardiac anomalies and subsequently succumbed to the condition. The other three defaulted follow-up. These five patients were not included in the US analysis.

RESULTS

There were 137 children who underwent surgery for ARMs during the period, of which 101 patients were included in the study (Fig. 3). 36 children were excluded, as they did not have a lumbar US. 17 out of the 101 (16.8%) children evaluated had LLC, and of these, seven had TCS while ten did not (non-TCS) (Table I and Fig. 3). Nine were male and eight were female. Among these 17 children, four (23.5%) had low-type and 12 (70.6%) had high-type (seven intermediate and five high) ARMs, while one (5.9%) had cloacal malformation. The higher numbers of high-type ARMs were not statistically significant ($p = 0.171$).

Out of the 17 children with LLC, 12 (70.6%) had concordant abnormal US and MR imaging findings, indicating LLC. Four of these had TCS. The other five (29.4%) children had normal US, i.e. no LLC seen on US, but had subsequent abnormal MR imaging findings of definitive LLC. Three of these five children

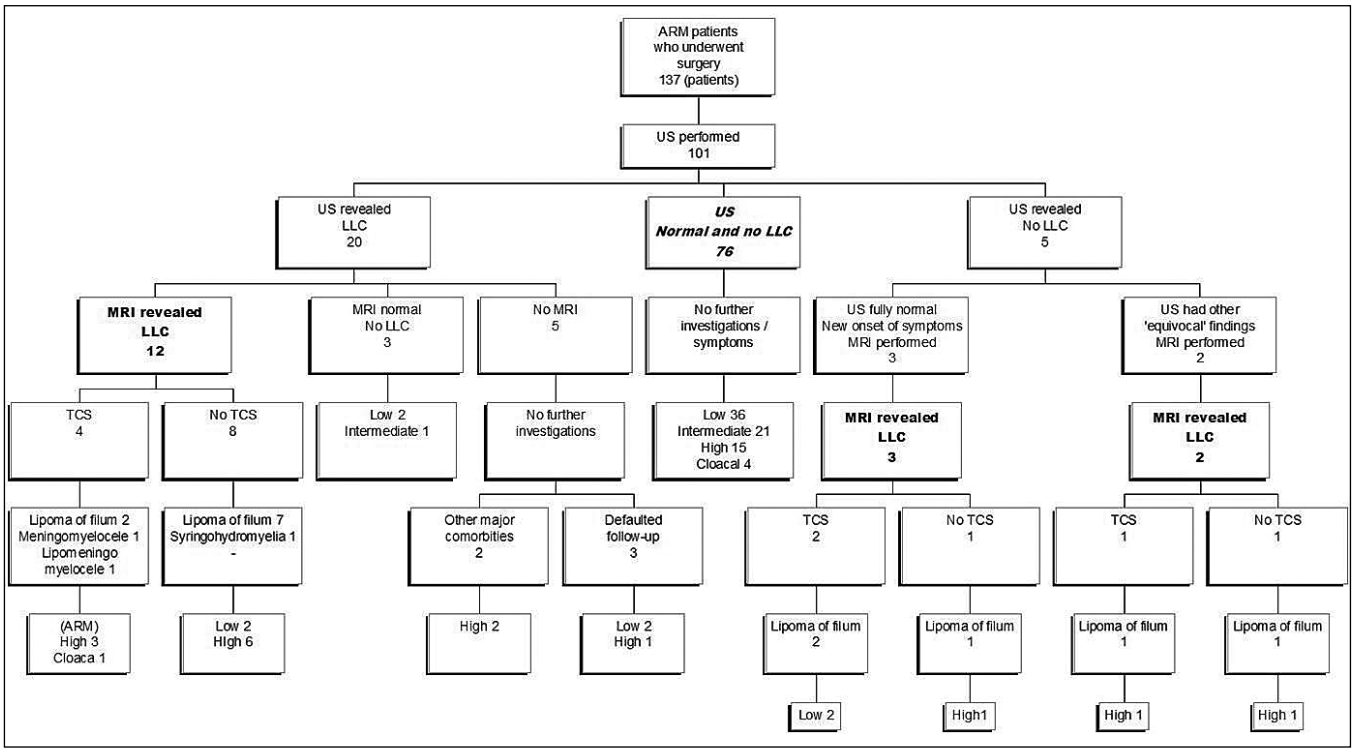

Fig. 3 Flowchart summary of the 101 patients included in the study.

were referred to the paediatric neurosurgery service, and MR imaging was performed at one year, six-and-a-half years and five years due to new onset of symptoms. Two of these three children had TCS. The other two children with normal US (reported) but abnormal MR imaging underwent imaging again, as these US had other 'equivocal' findings on review. One showed that the conus medullaris was within the lower limits of normal at L2. The other child had a US suggesting a filar cyst that measured 2 mm in diameter but which, on subsequent MR imaging, revealed LLC and lipoma of the filum terminale. The mean and median ages of the 17 children when US was performed were 3.65 days and 2.00 days, respectively, while the mean and median ages when MR imaging was performed were 14.59 months and 5.00 months, respectively.

14 of the 17 patients with LLC (82.4%) had lipoma of the filum terminale, while there was one patient each who had syringohydromyelia, lipoma of the conus medullaris and meningomyelocele. The children with syringohydromyelia and lipomyelomeningocele had high-type ARMs (intermediate ARMs of rectovestibular and rectourethral fistula, respectively), while the child with meningomyelocele had a cloacal anomaly. Both the children with meningomyelocele and lipoma of the conus medullaris had abnormal urodynamics, while the child with syringohydromyelia had normal urodynamics. The child with meningomyelocele also had a foot drop. In the 12 children with concordant US and MR imaging findings, there were nine cases of lipoma of the filum, one syringohydromyelia, one lipoma of the conus medullaris and one meningomyelocele. All five patients with discordant US and MR imaging findings had lipoma of the filum terminale. Four of the 17 children (23.53%) had VACTERL syndrome and two had trisomy 21. Two of the four VACTERL syndrome children had TCS, three had lipoma of the filum terminale and one had meningomyelocele. The clinical summary of these 17 children is illustrated in Table I.

The remaining 79 out of 101 children included in the study were not diagnosed with LLC. Of these, 38 (48.1%) were low-type ARMs, 37 (46.8%) were high-type ARMs (22 intermediate and 15 high) and four were cloacal (5.1%) anomalies. The approximately equal numbers of low-type versus high-type ARMs were not statistically significant ($p = 0.171$). Of these 79 children, 76 had normal US with no evidence of LLC. They also did not have any abnormal clinical signs and were not referred for new clinical changes. Three children who had LLC on US had subsequent normal follow-up MR imaging within three months. An analysis of the US findings for LLC and non-LLC is shown in Table II. Based on the assumptions previously mentioned, the sensitivity and specificity of US were 70.6% and 96.2%, respectively, while the negative and positive predictive values of US in our study were 93.8% and 80.0%, respectively.

In the seven children with TCS, five had lipoma of the filum terminale, one had meningomyelocele and one had lipoma of the conus medullaris. There were two low-type ARMS, four high-type (two intermediate and two high) ARMs and one cloacal anomaly. Of the ten children without TCS, nine had lipoma of the filum terminale and one had syringohydromyelia. There were two low-type and eight high-type (five intermediate, three high) ARMs. No statistical significance ($p = 0.566$) regarding the types of ARMs was observed in both the TCS and non-TCS groups. There were two cases of VACTERL syndrome in each of the TCS and non-TCS group. 16 of the 17 children with LLC underwent UDS. Seven children had abnormal UDS that revealed neurogenic bladder, thus indicating the presence of TCS. The one child without

Table I. Clinical summary of the 17 children with LLC.

Patient	Gender	Type of ARM	MR imaging findings	Pre-op UDS	Post-op UDS	Other anomalies	Follow-up period
1	F	High	Syringohydromyelia	Normal	None		8 yrs
2	F	High	Fibrolipoma and tethered cord (Referred for constipation)	Normal	Normal	VACTERL syndrome	5 yrs
3	F	High	LLC and fibrolipoma	Normal	None		6.5 yrs
4	F	Cloacal	Lipoma of the conus medullaris and tethered cord	Neurogenic bladder	Neurogenic bladder		1.5 yrs
5	M	Low	LLC and lipoma of the filum terminale (Referred for urinary overflow continence)	Voids by overflow and dribbling; high urinary retention	Normal	Trisomy 21	4 mths
6	F	High	LLC, lipoma of the filum terminale; partial sacral agenesis	Normal	Normal	Goldenhar syndrome	5.5 yrs
7	F	Low	Lipoma of the filum terminale (Referred for repeated urinary tract infection and abnormal UDS)	Overactive bladder with unstable detrusor contractions	Normal	VACTERL syndrome	6 mths
8	M	High	Tethered cord with lipoma of the filum terminale	Normal	None		3 yrs
9	M	High	Tethered cord, LLC, meningomyelocele	Neurogenic bladder	Neurogenic bladder	VACTERL syndrome; left foot weakness pre- and postoperatively	4 yrs
10	M	Low	LLC, mild tethering	Normal	None	Trisomy 21	3.5 yrs
11	F	Low	LLC, lipoma of the filum terminale	Normal	None		4 mths
12	M	High	LLC, lipoma of the filum terminale	Neurogenic bladder	Neurogenic bladder		3 yrs
13	M	High	Tethered cord, small lipoma of the filum terminale	None	None	VACTERL syndrome	3 yrs
14	F	High	Tethered cord; lipoma of the filum terminale	Normal	None		3 yrs
15	M	High	LLC, lipoma of the filum terminale	Neurogenic bladder	Neurogenic bladder		1.5 yrs
16	M	High	LLC, lipoma of the filum terminale	Overactive bladder with unstable detrusor contractions	Normal		1 yrs
17	M	High	LLC, lipoma of the filum terminale	Normal	Normal		1 mth

LLC: low-lying spinal cord; ARM: anorectal malformation; US: ultrasonography; MR: magnetic resonance; UDS: urodynamic study; M: male; F: female

Table II. Analysis of the US findings of LLC in anorectal malformation patients.

US finding	Confirmed diagnosis		
	LLC	No LLC	Total
LLC	12	3	15
No LLC	5	76*	81
Total	17	79	96

Note: 5 children were not included in the analysis, as they did not undergo further MR imaging.
*Assumed value. LLC: low-lying spinal cord; US: ultrasonography

UDS did not have neurological motor signs or urinary/bowel incontinence to indicate the presence of TCS. The child with myelomeningocele had both a neurogenic bladder and left foot weakness. All 17 children subsequently underwent surgical detethering. The mean and median durations from MR imaging to surgery were 5.35 and 4.00 months (range 0–16 months), respectively. The mean and median ages of the children who underwent surgery were 19.88 and 10.00 months (range 1 month to 5 years), respectively. Intra-operative histological findings of the lesions causing LLC in the 17 patients were consistent with the pre-operative radiological findings.

Postoperatively, three of the seven patients with TCS improved (Table I) and had normal urodynamics ($p = 0.051$); the patients were aged $\geq$ 15 months. Four children did not improve; one of them had myelomeningocele with both neurogenic bladder and left foot weakness, one had lipoma of the conus medullaris and two had lipoma of the filum terminale. All the children (except the child with myelomeningocele) had no neurological motor signs pre- and postoperatively. None of the children had any postoperative complications. The ten non-TCS children who underwent prophylactic detethering did not develop new symptoms on follow-up. The mean and median duration of neurosurgical postoperative follow-up were 2.92 and 3.00 years (range 1 month to 8 years), respectively.

DISCUSSION

There is no previously published literature on ARM and LLC in our local Singapore paediatric population. Our study examined the role of US and MR imaging in the diagnosis of LLC in ARM patients. We also compared the clinical differences between the TCS and the non-TCS groups, evaluated the surgical management and outcome of these children, and analysed the patterns of ARM types in LLC, non-LLC, TCS and non-TCS.

In our study, 16.8% of the evaluated children with ARM had LLC. This incidence is consistent with that of other studies, in which the percentage varies from 10% to 64%.[3,4,6-8] The

incidence of LLC is affected by the referral patterns and inclusion of the study populations, the choice of radiological diagnosis for LLC in ARM and practice differences in screening.[4,7] Our study population consisted mostly of ARM patients who had postnatal US studies to screen for LLC, and being population-based, the study highly reflects the incidence of LLC in ARM in our population. There have been mixed findings regarding the relationship of LLC with the types of ARM. LLC is known to be associated with high and complex ARM as well as cloacal and sacral anomalies.[3,6-8] However, it has also been reported that LLC was associated with low-type ARMs or was independent of the types of ARMs and the severity of sacral anomalies.[8] Other studies[9,10] have indicated that the incidence of LLC in VACTERL syndrome patients was higher (range 39%–86%) compared to the incidence of 23.52% in our study. The most common lesions in LLC were reported to be syringomyelia, altered sac morphology,[8] lipoma of the conus medullaris, intramedullary lipomas and lipoma of the filum terminale.[11]

LLC in our study appeared to be associated with high-type ARMs. There also appeared to be an equal proportion of low- and high-type ARMs in non-LLC patients. These findings were, however, not statistically significant. There was also no significant relationship between the types of ARM and the presence of TCS. LLC also did not appear to be strongly associated with VACTERL syndrome based on the incidence. Lipoma of the filum terminale was the most common spinal lesion but did not seem to be related to TCS. This was probably because the tethering effects of lipoma of the filum terminale may not occur until during the period of growth spurts later in childhood, during which the spinal cord becomes stretched. This stretching leads to impairment of mitochondrial oxidative metabolism and subsequent neuronal dysfunction.[12] Our findings suggest that LLC is common in our population of ARM and hence should be investigated whenever ARM is diagnosed, regardless of the types of ARM and other associated anomalies.

Diagnostic imaging modalities, such as US and MR imaging of the lumbar spine, aim to demonstrate the presence of LLC accurately and provide details for surgery in symptomatic TCS and prophylactic detethering in asymptomatic non-TCS. They also show the lesions causing tethering of the spinal cord such as lipomatous filum, lipoma of the conus medullaris, myelomeningocele and meningocele.[1,8] One study[7] reported a 500% increase in the detection rate of LLC in ARM after the introduction of screening US or MR imaging. The extent to which US and/or MR imaging is used, however, varies in different centres. US is safe, inexpensive, does not require sedation and is recommended for patients below three months of age before posterior ossification of the spine.[4,13,14] US is thus an excellent initial screening tool,[4] but it may not accurately detect an LLC[1,3,4,15] or a small lipomatous filum.[4] MR imaging is the gold standard for evaluation of the spine[4] and TCS,[1] as it assesses the conus medullaris, the thickness of the filum terminale, traction lesions and associated bony dysraphisms,[11] as well as clearly demonstrates intraspinal pathology.[8] The discordant rates between US and MR imaging in diagnosing spinal dysraphism have been reported to be 7%–30%,[4] although one study[13] reported that US exactly correlated with MR imaging in 32 out of 38 spinal dysraphism cases. It has been suggested that MR imaging be performed in cases of concurrent spinal anomalies and ARM,[16] as well as in cases of VACTERL syndrome.

Our study evaluated US and MR imaging for diagnosis of LLC in ARM. The majority of the ARM children with LLC had both US and MR imaging results that were concordant. The five cases with discordant findings were cases of lipoma of the filum terminale. US in our study did not miss LLC associated with the more complicated anomalies of spinal dysraphism such as lipoma of the conus medullaris, syringohydromyelia and meningomyelocele. Failure of US to diagnose LLC also did not appear to be associated with TCS. Our results thus suggest that US is an appropriate first-line investigation and screening tool for LLC in our population of ARM. MR imaging is not necessary in all cases of ARM but rather in cases where the US findings are abnormal or equivocal, as well as in children who have new onset of symptoms. Abnormal or equivocal US findings demonstrating lipoma of the filum terminale may lower the threshold for proceeding with an MR imaging, as all our cases of discordancy were lipoma of the filum terminale. VACTERL syndrome patients in our population may not necessarily lower the threshold for performing MR imaging based on the incidence of VACTERL syndrome in LLC.

It was reported that the sacral ratio was a strong predictor of LLC in ARM and a that low sacral ratio predicted a poor prognosis.[6] This is not unexpected, since the development of the sacrum is also from the same embryonic origin, the caudal cell mass. It was also recently recommended that MR imaging be performed for ARM patients with both sacral hypodevelopment and sacral ratio < 0.6.[4] Future investigations could evaluate whether sacral ratio could influence the consideration for MR imaging in our population. Future studies could also evaluate possible aetiology for discordancy between US and MR imaging in the different spinal lesions co-existing with ARM.

LLC can lead to TCS, which may in turn result in neurological, musculoskeletal, urological or gastrointestinal abnormalities.[1] It was reported that about 18%–21% of ARM patients with LLC had symptoms,[4,7] while another study[15] revealed that all patients had bladder dysfunction and 60% had orthopaedic deformities. In one study,[7] the proportions of TCS and non-TCS in LLC were 18% and 82%, respectively, and the proportions of LLC patients who underwent surgery for symptoms and asymptomatic detethering were 18% and 14%, respectively. Most centres perform detethering for TCS[17] but the results are varied. While detethering may arrest the progression of symptoms of TCS,[3] it may not improve the prognosis.[6] There were also reports that while orthopaedic and motor-sensory symptoms in TCS improved with detethering, urinary and bowel symptoms did not.[4,6-8,15] Results for detethering based on UDS results were also mixed.[18]

The poor results for bowel and urinary function were thought to be further contributed by the anorectal defect and abnormal sacrum in addition to the tethered cord, with a low sacral ratio also predicting a worse functional prognosis.[6] Previous animal experiments had also indicated that neurologic pathways controlling urinary and faecal continence may be established during early embryogenesis,[4] and detethering after these establishments thus may not improve function.

Opinion also differs regarding the timing of prophylactic surgery for patients without TCS.[3,6] There are recommendations for early prophylactic detethering because neurological symptoms would worsen with age[8] and future permanent neurological deterioration could be prevented.[3,17] This is because LLC, being fixed caudally, results in stretching of the spinal cord as the child grows, which, as mentioned above, leads to neuronal dysfunction.[12] One study recommended prophylactic surgery, particularly for lipoma of the filum terminale due to the short and long-term benefits.[19] Prophylactic surgery was, however, not recommended for lipoma of the conus medullaris due to its questionable long-term benefits,[19] although a more recent study revealed that long-term surgical outcome on spinal cord lipomas is further influenced by the extent of resection and age at surgery.[20] On the other hand, there are recommendations for a conservative approach to LLC altogether, as there is no clear evidence for the benefits.[3]

In our study, 41.2% of the ARM children with LLC had TCS and underwent detethering, while 58.8% without TCS underwent prophylactic detethering. Detethering for symptomatic TCS led to improvement in urodynamics in 42.9% of the children, although this was not statistically significant. One of the four patients who did not improve was a child with meningomyelocele, whose motor deficits also did not improve. Meningomyelocele has a different embryology from LLC, and so the outcome of this patient was not unexpected. There were no new complications or clinical signs after detethering in any of the 17 children. Our study illustrates that surgical detethering can lead to improvement in urinary function, as manifested by the improved UDS in almost half of the patients, although this was not statistically significant. The improvement in our study did not correlate with a younger age at surgery and thus did not appear to support the postulation of early establishments of neurological pathways for urinary function. Future studies could investigate other possible aetiologies for the favourable surgical outcome in urinary function in our local study population.

At our institution, all patients with LLC are recommended to undergo prophylactic detethering, and despite our high rates of prophylactic detethering, there were no postoperative complications in our study group and these patients remained asymptomatic on follow-up. 90% of our patients who underwent prophylactic detethering were cases of lipoma of the filum terminale, and the outcome for the latter is consistent with the favourable outcomes in various series.[19,21] Prophylactic detethering of the spinal cord in cases of lipoma of the filum terminale is generally straightforward with minimal operative risk[19,22] and is thus recommended. Based on the relatively short duration of follow-up, our results do not provide an insight to the long-term outcomes. Future studies on our current population over a longer follow-up period would provide further evaluation of the long-term outcomes for both the TCS group and the asymptomatic group in our population.

Our study had a few limitations. Firstly, the ARM population was obtained via the surgical records of ARM patients who had undergone surgery. This may not capture the small proportion of patients with ARM who did not undergo surgery at KKH. A prospective study looking at ARM patients with LLC would provide a more accurate evaluation of the relations of LLC and TCS in ARM as well as of the surgical outcomes. Another limitation was the small sample size of the LLC, TCS and non-TCS groups, which, although illustrated the trends and patterns, limited us from drawing a statistically significant conclusion. It also limited us from performing a regression analysis to take into account potential confounders like surgeon experience and technical expertise, which may have resulted in the favourable outcomes in our study. The small study sample size was, however, unavoidable due to the natural incidence of LLC in our ARM population.

In conclusion, 16.8% of children with ARM in our study had LLC. LLC appeared to be associated with high ARM, although it was not statistically significant. There was also no significant relationship between the type of ARM and the presence of TCS or the type of sacral spinal anomalies. LLC should be investigated for whenever ARM is diagnosed, regardless of the type. Lumbar US is useful as a first-line screening for LLC. Abnormal US or the onset of new symptoms should subsequently be investigated with MR imaging, while equivocal US finding is also likely to benefit from further MR imaging. Surgery to detether LLC can improve the outcomes in TCS, while surgery performed prophylactically in asymptomatic patients with lipoma of the filum terminale has very low surgical risk.

REFERENCES

1. Hertzler DA 2nd, DePowell JJ, Stevenson CB, Mangano FT. Tethered cord syndrome: a review of the literature from embryology to adult presentation. Neurosurg Focus 2010; 29:E1.
2. Patterson S. Sonographic assessment of the neonatal spine and the potential for new technologies to aid in diagnosis. J Diagn Med Sonography 2009; 25:4-22.
3. Suppiej A, Dal Zotto L, Cappellari A, et al. Tethered cord in patients with anorectal malformation: preliminary results. Pediatr Surg Int 2009; 25:851-5.
4. Kim SM, Chang HK, Lee MJ, et al. Spinal dysraphism with anorectal malformation: lumbosacral magnetic resonance imaging evaluation of 120 patients. J Pediatr Surg 2010; 45:769-76.
5. Wakhlu AK. Management of congenital anorectal malformations. Indian Pediatr 1995; 32:1239-42.
6. Levitt MA, Patel M, Rodriguez G, Gaylin DS, Pena A. The tethered spinal cord in patients with anorectal malformations. J Pediatr Surg 1997; 32:462-8.
7. Tuuha SE, Aziz D, Drake J, Wales P, Kim PC. Is surgery necessary for asymptomatic tethered cord in anorectal malformation patients? J Pediatr Surg 2004; 39:773-7.
8. Uchida K, Inoue M, Matsubara T, et al. Evaluation and treatment for spinal

cord tethering in patients with anorectal malformation. Eur J Pediatr Surg 2007; 17:408-11.
9. Kuo MF, Tsai Y, Hsu WM, et al. Tethered spinal cord and VACTERL association. J Neurosurg 2007; 106(3 suppl):201-4.
10. O'Neill BR, Yu AK, Tyler-Kabara EC. Prevalence of tethered spinal cord in infants with VACTERL. J Neurosurg Pediatr 2010; 6:177-82.
11. Raghavan N, Barkovich AJ, Edwards M, Norman D. MR imaging in the tethered spinal cord syndrome. AJR Am J Roentgenol 1989; 152:843-52.
12. Yamada S, Zinke DE, Sanders D. Pathophysiology of "tethered cord syndrome". J Neurosurg 1981; 54:494-503.
13. Dick EA, Patel K, Owens CM, De Bruyn R. Spinal ultrasound in infants. Br J Radiol 2002; 75:384-92.
14. Mosiello G, Capitanucci ML, Gatti C, et al. How to investigate neurovesical dysfunction in children with anorectal malformations. J Urol 2003; 170:1610-3.
15. Morimoto K, Takemoto O, Wakayama A. Tethered cord associated with anorectal malformation. Pediatr Neurosurg 2003; 38:79-82.
16. Heij HA, Nievelstein RA, de Zwart I, et al. Abnormal anatomy of the lumbosacral region imaged by magnetic resonance in children with anorectal malformations. Arch Dis Child 1996; 74:441-4.
17. Bui CJ, Tubbs RS, Oakes WJ. Tethered cord syndrome in children: a review. Neurosurg Focus 2007; 23:E2.
18. Guerra LA, Pike J, Milks J, Barrowman N, Leonard M. Outcome in patients who underwent tethered cord release for occult spinal dysraphism. J Urol 2006; 176:1729-32.
19. Pierre-Kahn A, Zerah M, Renier D, et al. Congenital lumbosacral lipomas. Childs Nerv Syst 1997; 13:298-334.
20. Pang D, Zovickian J, Oviedo A. Long-term outcome of total and near-total resection of spinal cord lipomas and radical reconstruction of the neural placode, Part II: Outcome Analysis and Preoperative Profiling. Neurosurgery 2010; 66:253-73.
21. Kasliwal MK, Mahapatra AK. Surgery for spinal cord lipomas. Indian J Pediatr 2007; 74:357-62.
22. Ng SYP, Gan BK, Seow WT. Screening and management of tethered spinal cord in children. J Paediatr Obstet Gynaecol 2010; 36:105-114.