The horizontal sacrum as an indicator of the tethered spinal cord in spina bifida aperta and occulta

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Object. The authors report on symptomatic patients with myelomeningocele (MMC) and lipomyelomeningocele (LMMC) who were found to have changes in their lumbosacral angle (LSA) corresponding to the onset of symptoms indicative of a tethered spinal cord.

Methods. The authors review data obtained in these two cohorts of patients and compare the LSAs measured in the perinatal period with those seen when the patients presented with symptoms of a tethered spinal cord.

Results. Children with LMMC, roughly one third of studied cases, were symptomatic due to a tethered spinal cord at their most recent follow-up. In children in whom the MMC was the closed form at birth, 20 of 30 had symptoms that could be indicative of a tethered spinal cord at their most recent follow-up. The LSA was altered in both groups with symptoms.

Conclusions. Signs and symptoms indicative of a tethered spinal cord appear to correspond to increases in the LSA. (DOI: 10.3171/FOC-07/08/E11)

KEY WORDS • myelodysplasia • myelomeningocele • spinal dysraphism • spine

In humans the LSA is approximately 20° at birth and increases to about 70° by 5 years of age, after which it remains constant.1 The LSA appears to be attributable to the ontogeny of bipedal position rather than obstetrical requirements. Indeed, in humans who acquire bipedalism early in life there is evidence of precocious formation of the LSA, and those who do not walk, walk late, or have impaired gait due to disease (for example, poliomyelitis) develop only a very minimal LSA.13

The horizontal sacrum, which is an exaggeration of the normal LSA, has been identified by our group in cases of symptomatic tethered spinal cord in populations of patients with MMCs and LMMCs.11,13 This entity is also found in other disease processes such as hypochondroplasia, atelosteogenesis Type II, Weismann-Netter-Stuhl syndrome, and spondylolisthesis.4,6,10 In the present study we review the findings obtained in these two groups of patients, one with an open and one with a closed form of spinal dysraphism.

Clinical Material and Methods

In a combined retrospective analysis of 50 consecutive patients who underwent LMMC repair and 30 patients who underwent MMC repair, the LSA was measured on radiography (recumbent) or MR imaging in the perinatal period and when these patients presented with signs and symptoms of a tethered spinal cord (for example, progressive lower-extremity dysfunction). This angle was determined by the intersection of two straight lines drawn on radiographs (recumbent) of the lumbosacral spine. The first line was drawn perpendicular to a line tangential to the anterior surface of the L-3 VB and the second line was drawn perpendicular to the sacral line, which is drawn by joining the middle of the anterior border of the S-1 VB of the first sacral vertebra with that of S-2 (Fig. 1). Angles were determined using a standard goniometer. The mean age for the group with an LMMC was 12.5 years (range 4–13 years); there were 28 girls and 22 boys. For this

Abbreviations used in this paper: LMMC = lipomyelomeningocele; LSA = lumbosacral angle; MMC = myelomeningocele; MR = magnetic resonance; VB = vertebral body.

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group, data obtained in 30 age-matched (25 male and 25 female) controls were also analyzed after they underwent lateral recumbent radiographs that also included the lumbosacral junction. These studies were most often acquired for observation of the gastrointestinal tract and thoracic viscera, and no patient had or was found to have diseases that involved the nervous system or spine. None of these patients (controls) had a diagnosis of spina bifida aperta or occult spinal dysraphism. Measurements were made of the LSA over time in all patients with an LMMC.

Additionally, we also analyzed LSA-related data obtained in 30 children with a closed-type MMC (treated at our institution between 1993 and 2001) who did not have a significant kyphotic, scoliotic, or lordotic deformity. These were 30 patients of a larger population of 71 cases involving total MMC closure during this period. These 30 patients were chosen because appropriate radiographs of the lumbosacral spine had been obtained perinatally and because they later presented with symptoms or signs of a tethered cord. All radiographs analyzed were those of patients in the recumbent position. In these patients the mean age was 4.9 years at presentation of tethered cord symptoms after MMC closure. None of the patients had a high thoracic or cervical MMC. Twenty controls were observed and were chosen from our hospital database because they were 8 years of age or younger (mean 3.5 years) and at least two lateral lumbosacral radiographs had been acquired over time. These controls were also without the diagnoses of spina bifida aperta, occult spinal dysraphism, or diseases of the lumbosacral vertebrae. In the majority of these control individuals, x-ray films had been obtained to observe the cavities of the thorax and abdomen. Statistical analysis was performed with SPSS 8.0 for Windows (SPSS Inc.).

**Results**

Appropriate imaging studies were available for 25 LMMC cases (lateral imaging studies that included the entire lumbar spine and sacrum, both perinatally and at presentation for sign/symptoms of a tethered spinal cord). Roughly one third of these patients exhibited symptoms due to a tethered spinal cord at their most recent follow-up (Fig. 2). Nine patients (36%) were found to have corresponding LSA measurements of greater than 70° with seven (28%) of these patients presenting with signs of a tethered spinal cord (for example, decreased lower-extremity function and bladder incontinence). The LSA measurements were statistically (p < 0.05) greater in the symptomatic patient population than in controls. Changes in the LSA were noted in only two asymptomatic patients (p ≥ 0.05). The perinatal LSA measurement for our study group ranged from 13 to 55° (mean 40°), and at presentation for signs or symptoms of tethered spinal cord this angle ranged from 45 to 90° (mean 75°). No single sign or symptom was found to correlate with an increase in the LSA in these patients. The LSA measured in controls during the perinatal period ranged from 5 to 42° (mean 25°) and at the second LSA measurement interval (age-matched controls) it ranged from 25 to 65° (mean 58°). None of our patients was found to have significant exaggeration of lumbar lordosis, although this curvature varies significantly in the general population and is difficult to define. Analysis of data obtained in symptomatic and non-symptomatic patients after surgical repair of their LMMC demonstrated a mean change in the LSA of 13 and 5°, respectively. The standard deviation for the symptomatic group was 11.546°, and for the nonsymptomatic group it was 8.174°. In symptomatic patients, the mean difference in the LSA (Measurement 2 – Measurement 1) was larger than that in the asymptomatic patients. This was statistically significant (p = 0.0202). The types of LMMCs seen in these patients were: caudal in 13 cases, dorsal in eight, and transitional in four. Of the seven patients with an increased LSA and signs/symptoms of a tethered spinal cord, three lesions were of the dorsal type, two were the caudal type, and two were the transitional type. The most common cutaneous stigmata in this group were lumbosacral subcutaneous masses and flat capillary hemangiomas. No single sign or symptom of a tethered spinal cord appeared to correlate with any degree of increase in the LSA. Of the excluded 25 patients in whom perinatal radiographic imaging was not performed, none was symp-
tomatic at their most recent follow-up and none had a grossly exaggerated LSA.

The LSA in children with an MMC closed at birth and for all ages ranged from 5 to 110° for the first measurement interval, with a mean of 65°, and from 25 to 120° for the second measurement interval, with a mean of 79°. Twenty of the 30 patients exhibited symptoms that could be indicative of a tethered spinal cord in clinic notes that were made at or near the time of their second LSA measurement (Fig. 3). These symptoms were increased hip subluxation, increased incidence of cystitis, increased external deviation of the foot, increased ureteral reflux, increased scoliosis, increased spasticity in the lower extremities, increased back pain, syringomyelia, decreased lower-extremity function, increased abnormality of urodynamics, and increased incontinence of the bladder or bowel. Five of these 20 patients went on to undergo spinal cord detethering soon after their second LSA measurement. The LSA values were increased at the second measurement in 26 of the total 30 patients and in all but one of the symptomatic patients. In addition, in two patients with a symptomatic tethered cord, there was no change in the LSA. The average change between the first and second LSA measurement ranged from 0 to 60°, with a mean change of 24°. The range for the time between measurements was 6 months to 7 years, with a mean of 3.4 years. Of these 30 patients, currently 11 ambulate with ankle-foot orthoses only, eight ambulate with ankle-foot orthoses and a walker or cane, and the remaining patients are not community ambulators.

In these 30 patients, the level of the MMC was as follows: three were thoracic, nine were thoracolumbar, 15 were lumbosacral, and three were sacral. The mean increase in the LSA for each of the aforementioned levels was as follows: thoracic 15°, thoracolumbar 20°, lumbosacral 25°, and sacral 10°. Children with more superior defects tended to have the greatest inclination of their LSA. Of the patients who did not exhibit an increase in their LSA between measurements, two had a decrease in their angle by 2 and 5°, respectively, and two had no change in the LSA. The range for the initial LSA measurement in our 20 controls was 5 to 37°, with a mean of 29°, and for the second LSA measurement the range was 28 to 82°, with a mean of 67°.

Statistical analysis of symptomatic compared with nonsymptomatic patients with myelodysplasia demonstrated a mean change in the LSA of 18.85 and 6.69°, respectively. The standard deviation for the symptomatic group was 15.621°, and for the nonsymptomatic group it was 10.159°. In symptomatic patients, the mean difference in the LSA (Measurement 2 – Measurement 1) was greater than that in the asymptomatic patients. This was statistically significant (p = 0.0371).

Discussion

The sacrum articulates with the ilia and transmits the weight of the body to the lower extremities through its upper segments. This bone, which is most differentiated in primates, is tilted anteriorly the greatest in humans. This rotation contributes to the formation of the LSA, which is generally not found in apes.1 In ancient man, the LSA is increased more than that in apes and reached its maximum in modern man, in whom it is slightly greater in females.2

Abitbol1 has reported that the LSA varies from 20 to 45° in children by the end of their 1st year of life and this corresponds to our measurements in controls. We found that mean perinatal measurement of this angle in our group of patients born, for example, with an LMMC, was 40°, which may suggest an earlier alteration in the angulation of the sacrum in these patients preoperatively. Reigel et al.7 found that release of a tethered spinal cord alters the course of lordosis in upper lumbar lesions but had little effect on the normal progression of lordosis in patients with lower lumbar or sacral lesions. Indeed, we did not observe significant changes in the LSA following detethering procedures in many of our patients with LMMCs or MMcs, but radiographs or MR images are not routinely obtained postoperatively in patients with LMMCs. Perhaps, reversal of this osseous deformity could be identified at long-term follow-up into adulthood, although the propensity for spinal cord retethering in this population might cloud this observation. Interestingly, in a perusal of other publications regarding patients with an LMMC, lateral radiographs of the lumbosacral spine obtained in symptomatic patients have demonstrated the “horizontal sacrum,” although no comments were made regarding this finding.

The mechanism underlying the greater propensity to develop a more horizontally positioned sacrum in forms of a tethered spinal cord is speculative, but it is known, for example, that spinal deformities (kyphosis, for example) in children with MMcs who have little function below L-1 develop as a result of the majority of the spinal extensors being rotated ventral to the equator of the spine.13 Interestingly, a tethered cord in children with MMcs may influence their growth rate1 and contribute to scoliosis in this group and others.12 Chernukha et al.3 have commented indirectly on this, stating that an increase in a patient’s lordosis may indicate neurological symptoms. Of note, of the total of 25 patients with a LMMC we examined, eight lesions (32%) were of the dorsal type, 13 (52%) were of the caudal type, and four (16%) were of the transitional type. However, of the seven patients with an increased LSA and signs/symptoms of a tethered spinal cord, three (43%) of the lesions were of the dorsal type, two (29%) were of the caudal type, and two (29%) were of the transitional type. These differences may reflect the difference in difficulty between the initial repair of these types and their proclivity to again become tethered in the long term.
Conclusions

We found that in children with an LMMC or MMC the LSA is often increased for their age and often increases at the time when symptoms of a tethered spinal cord manifest clinically. Although we do not advocate the use of the LSA as the sole indicator of the tethered spinal cord, this finding may aid the clinician in making clinical decisions regarding these patients when they present with symptoms of this pathological entity.

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References


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