The natural history of cord tethering in transitional LMMC remains unclear, but not all children suffer deterioration, and, in a significant proportion, function is not normal at birth. Surgery, as it is currently practiced, is generally safe but does not confer long-term immunity from deterioration.

The risk of deterioration, its pattern, and its timing are related in part to the morphology of the malformation. Patients with asymmetrical malformations may exhibit unilateral functional neurological or orthopedic abnormalities, which conspire with normal neurodevelopment to render these abnormalities apparent at an early age. Symmetrical malformations present later in childhood in association with bilateral and/or urinary dysfunction.

The rate of functional deterioration in patients following surgery appears to be equal to or is slower than the rate of deterioration in patients who do not undergo surgery. Many patients will require more than one untethering procedure to address evolving functional impairment. Structural abnormalities require end organ-specific orthopedic or urological interventions. A formal structured multidisciplinary monitoring team is required to provide clinical and functional surveillance. Such monitoring is required following operative untethering and debulking for the life of the patient.

Perhaps the most challenging cause of tethering in patients with spina bifida occulta is LMMC (lumbosacral lipoma, conus medullaris lipoma), in particular the transitional variety. Lipoma of the conus medullaris and its clinical variants were recognized in the early 1900s as a result of spontaneous deterioration in patients in whom a lumbosacral mass was noted at birth. The loss of function led surgeons to recommend surgery prior to the development of symptoms. Because the pattern of deterioration seemed unpredictable and symptoms in many patients were not reversed by surgery, many surgeons recommended that surgery be undertaken in infancy prior to the development of symptoms. The underlying presumptions were that cord tethering and the intramedullary lipomatous mass were the principal causes of deterioration, that untethering and lipoma debulking could be achieved without significant acute or long-term functional impairment, and that retethering and subsequent deterioration could be prevented. The authors of many series have shown good short-term results, with acceptable operative morbidity, following untethering. In the few long-term follow-up studies of surgically treated patients with transitional LMMCs, investigators have demonstrated a significant risk of symptom progression and deterioration despite surgically adequate untethering.

The purpose of this paper was to review the evidence regarding the timing and pattern of deterioration prior to and following initial cord untethering in patients with transitional LMMCs, to describe the differences in the presentation of transitional lesions based on the morphol-
ogy of the patient’s malformation, and to discuss the operative burden that these children bear so that they have optimized function.

**Risk of Deterioration With Nonoperative Management**

There is little published on the natural history of patients with LMMCs who do not undergo surgical intervention. A randomized clinical trial would add to our knowledge of the natural history of this condition, but the diversity of expert opinion and the lack of equipoise makes such a study unlikely. In the absence of definitive data, an estimate of the risk of deterioration before surgical intervention has been derived from the published clinical series in which authors described the age and frequency of presenting symptoms in patients prior to surgery. Cochrane et al. have applied survival analysis techniques to these data using the patient age when the “abnormal examination” was documented as the end point. The number of patients at specific ages and their condition (intact, asymptomatic, or abnormal status) were taken directly from Fig. 1 in the study published by Kaney and Bierbrauer and Figs. 22 and 30 in the study published by Pierre-Kahn. The assumptions made and the details of the analysis have been reported previously. These data are reproduced in Fig. 1 in the present report, the graph of which shows a comparison of deterioration rates for patients treated nonsurgically and those following successful untethering.

Kulkarni et al. subsequently published the data from the neurosurgery service of the NEM by using a similar analytic approach. The two estimates of deterioration in nonsurgically treated patients are compared in Table 1. The data derived from the historical literature show a more rapid deterioration over time than that documented in the NEM series. Other authors have postulated that there are differing patterns of presentation and deterioration that depend on the patient’s age, patient’s sex, and the nature of the malformation.

**Incidence and Timing of Deterioration Following Technically Satisfactory Untethering**

The determination of deterioration following cord untethering and lipoma debulking was derived from an outcome analysis of data obtained in a consecutive cohort of patients with transitional LMMC treated at British Columbia’s Children’s Hospital and from long-term outcomes reported in the literature. The pattern of postoperative deterioration compared with preoperative symptoms from the British Columbia Children’s Hospital series is shown in Table 2. Of those patients presenting with normal neurological function, without orthopedic or sphincteric dysfunction, 59% continued to experience clinically normal function throughout the last follow-up. Nine other patients, with preoperative normal function, suffered neurological, urological, or orthopedic deterioration. All patients with neurological dys-

---

**TABLE 1**

*Time to deterioration in patients with LMMCs treated conservatively*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Follow-Up (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>12</td>
</tr>
<tr>
<td>Cochrane et al., 2000</td>
<td>78</td>
</tr>
<tr>
<td>Kulkarni et al., 2004</td>
<td>95</td>
</tr>
</tbody>
</table>

* Values reflect the percentages of patients in whom functional deterioration occurred.

**TABLE 2**

*Summary of pre- and postoperative symptoms in patients with LMMCs*

<table>
<thead>
<tr>
<th>Postop Clinical Symptom</th>
<th>No. of Patients</th>
<th>Preoperative Clinical Symptom Status</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Normal</td>
<td>Neuro</td>
</tr>
<tr>
<td>no change from preop</td>
<td>22</td>
<td>13 (59)</td>
</tr>
<tr>
<td>functional change</td>
<td></td>
<td></td>
</tr>
<tr>
<td>neurological</td>
<td>13</td>
<td>5 (23)</td>
</tr>
<tr>
<td>orthopedic</td>
<td>9</td>
<td>3 (14)</td>
</tr>
<tr>
<td>urological</td>
<td>6</td>
<td>1 (5)</td>
</tr>
<tr>
<td>total</td>
<td>50</td>
<td>22</td>
</tr>
</tbody>
</table>

* Data derived from the study published by Cochrane et al. Abbreviations: Ortho = orthopedic; Neuro = neurological; Uro = urological.
function and orthopedic deformity evident preoperatively experienced further neurological deterioration, and the majority of patients with sphincteric dysfunction continued to suffer urinary deterioration. Time to the development of additional symptoms, either new or worsening of preoperative symptoms, following untethering/debulking is shown in Table 3 and varied with the symptom/sign. The postoperative symptom-specific intervals when 50% of cases were estimated to have shown deterioration were as follows: neurological function, 19 months; orthopedic function, 24 months; and urological function, 95 months.

Figure 1 provides an approximation of the time to deterioration for nonsurgically treated LMMCs; the curves indicate the deterioration following surgical untethering. The British Columbia’s Children’s Hospital, Paris, and Pittsburgh series included all patients and the Chicago series included only those with symptoms prior to untethering. The findings in the more recently published NEM series were not substantially different from reports published earlier. In looking at the graph in Fig. 1, the surgical series are above and parallel to the surgically treated curve. The slopes of the postoperative curves are not obviously different from the surgically untreated curve. With the exception of the NEM series, all postoperative curves lie above the literature derived natural history curve, suggesting that untethering does confer a benefit by stabilizing functions but that it does not protect against subsequent deterioration.

**Table 3**

<table>
<thead>
<tr>
<th>Primary Symptoms at Deterioration</th>
<th>Time to Deterioration (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>normal preuntethering</td>
<td>100</td>
</tr>
<tr>
<td>abnormal preuntethering</td>
<td>36</td>
</tr>
<tr>
<td>w/ functional change</td>
<td></td>
</tr>
<tr>
<td>neurological</td>
<td>19</td>
</tr>
<tr>
<td>orthopedic</td>
<td>24</td>
</tr>
<tr>
<td>urological</td>
<td>95</td>
</tr>
</tbody>
</table>

* Data derived from study published by Cochrane et al.  † Time to deterioration in 50% of the patient population.

**Relationship of Malformation’s Morphology to Pattern of Deterioration Before or After Untethering**

The anatomical variation of transitional LMMCs has been described by many authors. The constituents of the lipoma vary from case to case and with the age of the patient, as do the relationships of fat, fibrous tissue, muscle, and bone to neural elements. In the extensive review by Pierre-Kahn and colleagues, the authors described malformations of both cord and nerve roots in 66% of patients with dorsal or transitional lipomas of the conus medullaris. The lipoma–cord interface is also variable, lying symmetrically in the spinal canal or being rotated, often to the left. The clinical marker of an asymmetrical lipoma–neural interface is the presence of the subcutaneous lipoma centered off the midline. These observations prompted an analysis of the relationship of the anatomy of the lipoma–neural interface and a patient’s neurological dysfunction.

The malformations were classified as symmetrical (symmetrical LMMCs) if the epicenter of the lipoma was in the midline, if the lipoma arose symmetrically from the dorsal and caudal aspect of the neural placode, and if the amount of cerebrospinal fluid in both right and left lateral gutters was similar. Asymmetrical malformations (asymmetrical LMMCs) were characterized by the neural placode’s presence asymmetrically (rotated to the right or left) within the subarachnoid space (Figs. 2–6).

The relationship of the clinical symptomatology at diagnosis to the symmetry of the placode is shown in Table 4. Symmetrical malformations, when associated with symptomatology, exhibited bilateral deficits, and asymmetrical lesions were associated with unilateral deficits usually on the side to which the neural placode was rotated—that is, the side of the short roots.

**Table 4**

<table>
<thead>
<tr>
<th>Orientation</th>
<th>Lt</th>
<th>Rt</th>
<th>Bilat</th>
<th>Normal</th>
</tr>
</thead>
<tbody>
<tr>
<td>symmetrical</td>
<td>2</td>
<td>2</td>
<td>7</td>
<td>13</td>
</tr>
<tr>
<td>asymmetrical</td>
<td>6</td>
<td>0</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>lt</td>
<td>0</td>
<td>2</td>
<td>1</td>
<td>7</td>
</tr>
</tbody>
</table>

* Values reflect the number of patients.
Among patients with normal preoperative function who underwent successful untethering and subsequently suffered functional deterioration, the pattern was similar. Asymmetrical malformations were associated with ipsilateral deterioration in nine of 11 patients in whom deterioration occurred, and patients harboring symmetrical malformations exhibited bilateral signs or bladder deterioration in four of four cases. Proportionally more patients with symmetrical malformations retained normal function following untethering, and when deterioration did occur, it did so later in the postoperative period (range 50–65 months), whereas in those with asymmetrical malformations, whose function was normal preoperatively, the patient exhibited signs of deterioration earlier in the postoperative period (mean 22 months) (Table 2).

From the technical perspective, the morphology of the malformation predicts the technical ease and likelihood of successful untethering as well as the risks of untethering and lipoma debulking. The features of most relevance are: 1) access to the subarachnoid space lateral to the dorsal roots at the dural–lipoma interface and 2) the nature of the lipoma–placode interface (Figs. 4–6).

Injury to nerve roots is governed by the ease with which the subarachnoid space lateral to the dorsal roots can be accessed. This is a “safe space” and marks the location where the lipoma and placode can be untethered from the dura mater. In symmetrical lesions, this dural-lipoma-placode interface is readily followed, and untethering is achieved by following the subarachnoid space from rostral to caudal on each side of the placode. In the rotated transitional LMMC, the dura-lipoma-placode interface on the side rotated ventrally is more difficult to follow and requires the definition of anatomy concurrently from above and below. This is accomplished by first defining the “easy” side of the placode, untethering along the dural-lipoma-placode interface to the caudal end of the placode and then using this access to follow the interface rostrally on the rotated side, whereas on the contralateral side, progressing caudally and ventrally. Dissection on the rotated side is facilitated by judicious debulking of the intraspinal extradural lipoma to create the access corridor to the ventrally rotated dural–lipoma interface. This can be achieved safely if the surgeon works from the subarachnoid space “out” through the lipoma while protecting the lipoma–placode and roots. If one attempts to create this access corridor by entering through the lipoma, it is more likely that one will misinterpret the pathological anatomy and injure the roots and placode. The latter occurs because the surgical trajectory is too medial and the surgeon inadvertently enters the placode.

The lipoma–placode is variable in its content. Neural and mesenchymal elements are commonly present (Figs. 4–6). During dissection, the placode–lipoma interface is the usual site of injury to conus medullaris function, commonly resulting in incontinence. Regardless of the tools used to define and separate the lipoma–placode interface (carbon dioxide or yttrium aluminium garnet laser, traditional sharp microdissection), the decision regarding the extent of resection of the nonlipomatous elements becomes important from the perspective of injury risk and successful untethering. Electrophysiological monitoring and stimulation may help in defining tissues to be preserved. When in doubt, the surgeon’s only option is preservation with capacious dural graft placement.

Dural repair of the untethered and debulked rotated transitional LMMC is usually straightforward because the placode can be gently rotated and retracted to provide access for repair.
Operative Burden of Patients With LMMC During the 1st 18 Years of Life

There has been little written on the total operative burden borne by patients with spina bifida. We have reviewed the operations performed in a group of “graduates” of our multidisciplinary spinal cord clinic who had LMMCs and follow-up of at least 18 years. The operations were categorized based on the functions that the operation was intended to optimize. Of the 21 graduates, nine patients had remained clinically stable following their first untethering procedure and 12 patients underwent additional untethering operations to treat subsequent clinical deterioration. Function in seven of these patients stabilized following cord untethering; in five patients additional operations were required for orthopedic management of foot deformity (three–six interventions per patient) and/or urological procedures (one–seven procedures per patient).

Those patients who only had one cord untethering also fell into two groups. Five of nine patients required no other surgeries for orthopedic or sphincteric dysfunction whereas four patients required multiple procedures to treat orthopedic deformity.

Discussion

Although the original description of LMMC was reported in 1857 by Johnson (cited by Hoffman et al.) and subsequent authors have reported large series of LMMCs, the natural history of the lesion remains unclear. In the 1950s Bassett described the tendency for patients with LMMCs to experience spontaneous deterioration and observed that older children were likely to have neurological, orthopedic, or urological impairment, a clinical state seen less frequently in infants. The presumption was that patients were born functionally normal and that they subsequently and inevitably suffered deterioration of function. The presumed origin of the functional deterioration in patients with LMMCs was cord tethering and it was thought that this process was correctable by operative intervention. Myelodysplasia has not been thought to play an important role; indeed, as one group put it, “despite the grossly deranged anatomical configuration of the spinal cord in these patients, normal neurological function can be preserved.”

With further clinical experience, it has become clear that not all infants have normal function at birth or during infancy. The proportion of surgically untreated patients who exhibit abnormal neurological, urological, or orthopedic function at or close to birth is on the order of 40%. Both estimates of the natural history show progressive functional loss in the study populations, confirming that not all of these children deteriorate and that the time to functional loss can be spread over many years (Table 1).

Significantly, the natural history of LMMCs does not
necessary follow a progressive course of stability or deterioration. Rapid increases\(^\text{17}\) and, less commonly, decreases\(^\text{14}\) in lipoma mass have been reported (Fig. 7).

Postuntethering symptomatic deterioration unrelated to the operative intervention has been reported in most surgical series, although the frequency and time to deterioration vary.\(^\text{3,7,18,20}\) The time to development of specific symptoms differs depending on the symptom being assessed and the duration of follow-up.\(^\text{6,8}\) Neurological and orthopedic deterioration is detected sooner than deterioration in urinary control. The likelihood of deterioration was higher in patients with symptoms at the time of the original surgery.\(^\text{15}\)

The neuroanatomical features of LMMCs vary significantly. Both Choux and associates\(^\text{5}\) and Pierre-Kahn and colleagues\(^\text{19}\) have described additional neural abnormalities in these patients. Small syrinx cavities rostral to the lipoma, split cord malformations, and intralipomatous and short nerve roots have been described. Short roots were seen in 26% of conus medullaris lipomas in the Paris series and were significant in that “they may prevent the surgeon from adequately untethering the malformation.”\(^\text{18}\)

The relationship of the anatomy of the malformation to the presenting or subsequent symptoms and signs is likely important in determining the expectations for outcomes after surgery. The presence of an asymmetrical malformation would appear to predict the development of an asymmetrical neurological/orthopedic deficit on the side to which the placode is rotated. The etiological roles of medullosystem, failure to untether the cord, and retethering remain unclear. In the setting of a normal child with asymmetrical (rotated) LMMC, it would seem reasonable to advise patients that dysfunction on the side to which the placode is rotated may occur and may not be reliably prevented by untethering. There is no evidence in the current literature to support or oppose the deferring of operative untethering on the basis that a lateralized deficit will develop in a patient with an asymmetrical malformation or that untethering will prevent it.

Patients with symmetrical malformations appear to exhibit a different pattern. They are more likely to retain normal function at follow-up than those with asymmetrical malformations. They are less likely to sustain a neurological deficit as a result of untethering and lipoma debulking, and if they do experience deterioration, they do so more commonly with bilateral/sphincter signs such that deterioration is only apparent at an older age.\(^\text{6,8}\)

Cord untethering and lipoma debulking can be effective in stabilizing function in infants and children with LMMCs, but the operative burden can be significant. Cord untethering on one or more occasions can address functional deficits but structural abnormalities (orthopedic or urinary), either preexisting or evolving despite untethering, will require end-organ–specific interventions.

Transitional LMMC is a complex malformation that differs significantly from other forms of cord tethering and other less complex lipomas of the conus medullaris or terminal filum. Whereas natural history remains unclear, there is increasing evidence that not all children inevitably suffer functional deterioration, at least not within the published follow-up intervals, and function in a significant proportion is not normal at birth. Operative intervention, as it is currently practiced, is generally safe and may stabilize function; it does not, however, confer long-term immunity from deterioration.

The morphology of the patient’s transitional LMMC may be important in suggesting the pattern and timing of neurological dysfunction and, therefore, the monitoring and follow-up requirements for these patients. Untethering in patients with asymmetrical malformations, either prior to or after the declaration of symptoms and signs, does not predictably preserve or restore normal neurological function. The nature of the neurological and/or orthopedic dysfunction these patients tend to show and normal

---

**Fig. 6.** Sagittal (left) and axial (right) MR images revealing an asymmetrical transitional LMMC rotated to the left. Arrows indicate the dural-placode-lipoma interface. In additional to the rotation of the malformation to the left, the lipoma is composed of multiple elements, many of which appear to respond to stimulation during intraoperative monitoring.

**Fig. 7.** Sagittal MR images showing a substantial increase in the extradural (arrowhead) and intradural (arrow) intramedullary lipoma between 2 and 4 months. Initial imaging had been done when symptoms appeared. At 4 months of age, the infant presented with paraplegia with only partial recovery following debulking. Untethering was not achieved.

D. D. Cochrane

Neurosurg. Focus / Volume 23 / August, 2007
neurodevelopment conspire to render these functional abnormalities apparent at an early age. The malformation remains a surgical challenge. Symmetrical malformations are technically easier to address without operative morbidity and, when symptomatic, present in childhood with unilateral, bilateral and/or urinary dysfunction.

**Conclusions**

The rate of deterioration in patients following surgery appears to be equal or less than that in patients who do not undergo cord untethering. Regardless of whether untethering is proposed in asymptomatic infants or intervention is deferred until symptoms are recognized, a formal structured multidisciplinary monitoring team is required to provide clinical and functional surveillance. Such monitoring is required following operative untethering and debulking for the patient's lifetime.

**References**


**Acknowledgment**

I express my sincere appreciation to R. Kariyattil, Amrita Institute of Medical Sciences, Cochin, India, for providing Fig. 7.