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The medical records of patients at Children's Hospital and Medical Center treated for lipomyelomeningocele repair were reviewed to assess the long-term outcome of surgery. In total, 108 children were cared for between 1952 and 1987, and long-term data are available for 80 of 96 children who underwent surgery. Cutaneous manifestations and associated malformations were common. Surgical repair was performed at ages 1 week to 17 years; there was no operative mortality. Thirty-five of 38 children with normal preoperative examination were without deficits on long-term follow-up review. Bowel and bladder paralysis was present in 42 children and did not recover after release of cord tethering. Eleven children had return of symptoms and were reexplored 3 to 8 years after initial surgery. Surgical repair with release of cord tethering at the time of diagnosis is advocated, regardless of patient age.

Key Words • lipomyelomeningocele • lipoma • spinal dysraphism • tethered spinal cord • timing of surgery • children

The spectrum of spinal dysraphism includes multiple skin-covered lesions associated with spina bifida occulta. Lipomyelomeningocele is a subcutaneous lipoma occurring in the lumbosacral region connected to an intradural fatty component by a fibrous stalk, resulting in a tethered spinal cord. Children with lipomyelomeningocele most frequently have intact neurological function at birth but can progress to profound bowel, bladder, and lower-extremity dysfunction. Controversy exists regarding the correct timing of surgery for lipoma debulking and release of cord tethering. Many investigators recommend early surgery to prevent injury to the roots of the cauda equina from traction due to conus tethering. Others consider that surgery in the young child carries excessive risk of nerve injury.

At Children’s Hospital and Medical Center in Seattle, Washington, the management of children with lipomyelomeningocele is coordinated through a multidisciplinary group of pediatricians, neurosurgeons, orthopedic and urological surgeons, nurses, therapists, and rehabilitation specialists. This study consists of a retrospective review of the medical records of children with lipomyelomeningocele cared for between 1952 and 1987. The purpose of our study was to assess the timing, complications, and long-term neurological outcome following surgery.

Clinical Material and Methods

Patients from Washington, Alaska, Montana, and Idaho were referred to the Congenital Defects Clinic for comprehensive evaluation. A neurological assessment was performed by neurosurgeons, pediatricians, and physical therapists, and associated anomalies were evaluated by multiple specialists in congenital defects. Bladder function was assessed by renal and bladder ultrasound, dynamic voiding studies, intravenous pyelography, and cystometric studies. Electromyography was performed in many children with focal motor and sensory neurological deficits. All patients with suspected lipomyelomeningocele underwent plain spine films and myelography. Early studies in this series utilized injections of Pantopaque, which was replaced by metrizamide in 1976. High-resolution computerized tomography scans following intrathecal administration of contrast material began in 1980. Magnetic resonance imaging was added in 1986.

Patients were followed postoperatively in the Birth Defects Clinic with comprehensive evaluations on a
Long-term outcome of lipomyelomeningocele surgery

### TABLE 1

<table>
<thead>
<tr>
<th>Chief Complaint</th>
<th>No. of Cases</th>
</tr>
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<tbody>
<tr>
<td>lumbar sacral fatty mass</td>
<td>80</td>
</tr>
<tr>
<td>loss of bowel or bladder function</td>
<td>42</td>
</tr>
<tr>
<td>low-back or leg pain</td>
<td>12</td>
</tr>
<tr>
<td>foot ulceration</td>
<td>9</td>
</tr>
<tr>
<td>gait difficulties</td>
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<tr>
<td>orthopedic foot deformity</td>
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### TABLE 2

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<thead>
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<th>Lesions</th>
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<tr>
<td>midline lumbar sacral fatty masses</td>
<td>80</td>
</tr>
<tr>
<td>skin dimples</td>
<td>14</td>
</tr>
<tr>
<td>hair patches</td>
<td>11</td>
</tr>
<tr>
<td>superficial hemangiomas</td>
<td>9</td>
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<tr>
<td>skin tags</td>
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<td>depigmented regions</td>
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</table>

### TABLE 3

<table>
<thead>
<tr>
<th>Anomalies</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>scoliosis</td>
<td>7</td>
</tr>
<tr>
<td>amniotic band extremity deformity</td>
<td>6</td>
</tr>
<tr>
<td>sacral dysgenesis</td>
<td>4</td>
</tr>
<tr>
<td>anterior anal displacement with stenosis</td>
<td>2</td>
</tr>
<tr>
<td>hydromyelia</td>
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</table>

**Patient Presentation**

The records of 108 children treated for lipomyelomeningocele between 1952 and 1987 were evaluated. The incidence of these cases increased during the 35 years of the study. Permission for surgery was not given by the families of 12 children. Of the 96 children who underwent surgical repair, long-term data were available for 80, and these form the subject of this study. The remaining 16 children moved away from the referral region and information regarding their status is unknown.

The 80 patients in this series included 29 males and 51 females. From 1952 to 1967, the average age of patients on initial examination was 5½ years as compared with an average age of 4 months during the last 5 years of the study. Infants and children less than 3 years of age were most commonly brought for evaluation of a midline or eccentric lumbar sacral fatty mass, whereas symptoms of tethered spinal cord were present in older children as well (Table 1). Associated cutaneous manifestations (Table 2) and other anomalies were common (Table 3).

Seven children exhibited scoliosis on initial evaluation; of these, four had normal neurological examination and the other three patients had S1-2 motor levels. Seventeen patients had hydronephrosis, and six had renal dysgenesis upon initial ultrasound evaluation. No child had hydrocephalus. Magnetic resonance imaging, performed in 17 children, did not demonstrate Arnold-Chiari malformation or associated dysraphic conditions. Twelve children had a family history of spinal dysraphism, including myelomeningocele and diastematomyelia.

Thirty-eight children (47.5%) were normal on initial neurological examination (Fig. 1). In the remaining 42 children, the functional level ranged from S3-4 to L-4 and paralysis of bowel and bladder was confirmed in each of these children by ultrasound or dynamic voiding studies. Fixed neurological deficits and bladder paralysis were more common with increasing age (Fig. 2). In infants aged less than 1 year, normal examination

![Fig. 1. Preoperative neurological functional level in 80 children with lipomyelomeningocele.](image1)

![Fig. 2. Preoperative neurological findings in 80 children with lipomyelomeningocele as a function of age.](image2)
was twice as likely as functional loss, while in older children, fixed deficits were predominant.

Previous publications describing diagnosis,\textsuperscript{10} histopathology,\textsuperscript{2} and classification\textsuperscript{3} of skin-covered sacrococcygeal lesions have reported many of the patients included in the present series.

Results

Surgery and Postoperative Complications

Surgical repair was performed in children at ages 1 week to 17 years. All but three of 17 cases treated surgically during 1986 and 1987 were in infants less than 6 months of age, and 44 of all 80 operative repairs were in children less than 1 year of age. The most common level of lipoma attachment to the conus was lumbosacral (52%); lumbar attachment was seen in only 12 cases.

Postoperative complications included two subcutaneous cerebrospinal fluid leaks, which resolved spontaneously. There were 14 superficial and seven deep wound breakdowns; these were managed conservatively except for two deep draining sinuses that required excision and closure. One case was complicated by epidural abscess and meningitis requiring multiple drainage procedures. Children with normal preoperative neurological examination had fixed postoperative deficits limited to a unilateral S-1 root injury. No surgical mortality or complications due to anesthesia were reported. The overall complication rate of the series was 33%; from 1983 to 1987, it was less than 10% (three superficial wound breakdowns).

Long-Term Outcome

Thirty-five of 38 children with normal preoperative examination had no neurological deficits or bladder paralysis upon long-term follow-up examination. The three children with unilateral postoperative S-1 root injury retained normal bladder and bowel function. Ten children were followed until 21 years of age with an average follow-up interval of 6.2 years (Fig. 3).

None of the 42 children with preoperative bowel and bladder paralysis recovered normal function. The average follow-up period for this group was 5.2 years and 10 children were followed until 21 years of age (Fig. 3). In contrast, improvement in motor and sensory deficits was observed following repair and release of cord tethering. All but one of the children with preoperative L-4 or L-5 function improved to an S-1 or S-2 level 12 to 18 months following surgery. In children with preoperative functional levels of S-2 or lower, improvement in perineal sensation and gait was detected within 12 months of repair (Fig. 4). Of seven children with preoperative scoliosis (< 45°), three with S1–2 function were stabilized after surgery and the other four had improved 1 to 4 years postoperatively.

Three to eight years after initial surgery, 11 children developed new symptoms of cord tethering including foot ulceration, radicular pain, progressive weakness, and changed bladder function. Scoliosis did not accompany onset of recurrent symptoms. These patients were re-explored without neurological morbidity and adhesions of the lipoma-conus interface with the dura were present. Ten of these children had return of function to their initial level within 1 year and bladder dysfunction reversed in one of two children with new incontinence.

The greatest morbidity in this series of lipomyelomeningoceles patients was bladder and bowel paralysis. Children over 3 years of age required intermittent catheterization, antibiotic prophylaxis, and urinary acidification; bowel programs included digital stimulation and enemas. Children with bladder paralysis had at least one urinary tract infection each year requiring symptomatic oral antibiotic therapy. Sixteen children developed pyelonephritis necessitating intravenous antibiotics and hospitalizations averaging 10 days in duration. One child with bladder paralysis since 4 years of age developed renal vascular hypertension secondary to chronic infection; when she reached 20 years of age, a renal transplant was necessary.

In our series, all children over 3 years of age were ambulatory and only four required functional bracing. Intelligence was normal in all children of school age or

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{fig3.png}
\caption{Long-term postoperative follow-up period in 80 children with lipomyelomeningocele after initial repair and release of tethered spinal cord.}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{fig4.png}
\caption{Neurological examination in 80 children with lipomyelomeningocele 1 year following repair and release of tethered spinal cord: 47.5% were normal and functional levels were at S3-4 in 20%, S2-3 in 23.75%, S1–2 in 6.25%, L-5 in 1.25%, and L-4 in 1.25%.}
\end{figure}
Long-term outcome of lipomyelomeningocele surgery

older. Scoliosis developed 2 to 6 years after surgery in seven children with normal preoperative spinal alignment. Three of these children, two with normal neurological examination and one with an SI–2 functional level, required correction by instrumentation. Scoliosis was not a criterion for reexamination in the absence of other neurological changes.

Discussion

Series Overview

Lipomyelomeningocele is the most common closed skin-covered lesion of spinal dysraphism requiring neurosurgical management. At our medical center, the incidence was 8% of myelomeningoceles. The lesion was more common in females by a ratio of nearly 2:1, and associated anomalies were frequent. No child exhibited hydrocephalus or Arnold-Chiari malformation, and all had normal intelligence.

Clinical Presentation

Most infants with lipomyelomeningocele had normal neurological examination at birth and presented for evaluation of a superficial lipoma. Insidious loss of function increased with age due to progressive conus tethering and potential injury to the neural elements. Fixed deficits were three times more common in children under 1 year of age, and bowel and bladder paralysis was complete by the time motor or sensation loss was apparent on examination. Detection of bladder paralysis in children less than 2 years of age was difficult and unreliable except by dynamic contrast-enhanced bladder-emptying studies.

Surgical Procedures

We consider that repair of lipomyelomeningocele and release of cord tethering can be accomplished with very small neurosurgical risk, regardless of patient age. No complications relating to anesthesia occurred in the series. The goals of each operation were debulking of the intradural lipoma, preservation of all neural elements, release of conus tethering, and watertight reconstitution of the dura. Resection of the lipoma was aided by the use of a CO₂ operating laser and an ultrasonic aspirator. The resection techniques of Hoffman, et al., Schut, et al., and McLone and coworkers were utilized. There was no correlation between the location or size of the lipoma and neurological function, confirming the findings of Lassman and James and Rogers, et al. It was not possible to define the attachment anatomy using the classification system of Chapman, et al., or the morphology subtypes described by Hukuba, et al.

Other large reported series confirm the safety of surgery in patients of all ages. During the first 25 years of this study, radical debulking of the subcutaneous lipoma mass was a surgical goal; dehiscence of skin margins and subcutaneous closure layers was common in that era. We conclude that extensive resection of the subcutaneous fatty mass is unnecessary and leads to devascularization of the overlying skin which compromises wound healing.

Analysis of Results

Neurologically normal children who underwent surgery at an early age had normal function upon long-term follow-up examination and were the only patients who retained normal bladder function. Therefore, it is necessary to treat children with lipomyelomeningocele early, before critical neurological function is irretrievably lost.

The major cause of morbidity in patients with lipomyelomeningocele is bladder paralysis. Intermittent catheterization and aggressive urinary/renal sepsis surveillance is critical to minimizing kidney injury. Eleven children in our series were reexplored and removal of adhesions and neurolysis were performed without added neurological morbidity, which is similar to the rate described by Chapman and Beyerl. Scoliosis was a common sign in children with tethering but was not apparent in the group who had progressive neurological symptoms after initial repair. Reigel and Tamaki, et al., advocated release of adhesions and cord tethering for scoliosis and reported improvement in spinal curvature. We now reexplore children who develop postoperative scoliosis.

Conclusions

Lipomyelomeningocele is a serious lesion which, with growth and cord tethering, produces neurological loss and bladder paralysis. Children operated on in advance of deterioration have retained normal neurological function on long-term follow-up examination. Since there is little risk of nerve injury regardless of the patient’s age, we now routinely operate on children with lipomyelomeningocele at 1 to 3 months of age, as normal bladder function can only be achieved by early surgery.

Acknowledgment

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References

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