



LIPOMENINGOMYELOCELE: CLASSIFICATION, MANAGEMENT AND CONTROVERSIES

Definition :

Lipomyelomeningocele is a form of Open spinal dysraphism in which a subcutaneous fibrofatty mass traverses the lumbodorsal fascia, causes a spinal laminar defect, displaces the dura, and infiltrates and tethers the spinal cord.

- 1857 – Johnson
- 1971- Rogers and colleagues- used Term “LMM”

Oakes W: Management of spinal cord lipomas and lipomyelomeningoceles, in Wilkins RH, Rengachary SS (eds): Neurosurgery Update II. New York: McGraw-Hill, 1991, Vol 3, pp 3497–3504.

Classification

1) **Modified Chapman classification :**

- 1982
- according to relationship of lipoma-cord interface
- Dorsal
- Caudal or Terminal
- Transitional (Dorso caudal or Dorsolateral)

2) Recently- Chaotic lipoma

1) Chapman PH: Congenital intraspinal lipomas:Anatomic considerations and surgical treatment. *Childs Brain* 9:37–47, 1982

2) 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 I: anatomy, embryology, and surgical technique. *Neurosurgery*. 2009;65(3):511-528.

Dorsal Lipoma

- Fibrolipomatous stalk tethering cord proximal to conus
- Usually at middle lumbar to lumbosacral level
- Dorsal spinal cord dysraphic at site of attachment of lipoma
- Site of attachment medial to the dorsal root entry
- Normal spinal cord distal to myeloschisis.
- Roots lie within the subarachnoid space

Caudal or terminal lipoma

- Directly from conus medullaris or filum terminale
- Largely or wholly intradural
- Nerve roots entangled in the lipoma
- Lipoma-cord interface caudal to the dorsal root entry zone.
- Filum may be fatty, thickened and sometimes attached to subcutaneous tissue (sacral dimple).


Transitional lipoma

- Characteristics of both Type 1 and type 2.
- No normal spinal cord distal to lipoma attachment
- Initially dorsal roots may be separate but caudally become enmeshed into the lipoma.
- Frequently asymmetric attachment to cord

Chaotic lipoma

- Does not follow the rules of other lipoma
- Begin dorsally, caudal portion is ventral to placode.
- Does engulf neural tissue and nerve roots
- The fusion line - distinct rostrally and less predictable.

Pang et al. LONG-TERM OUTCOME OF TOTAL AND NEAR-TOTAL RESECTION OF SPINAL CORD LIPOMAS AND RADICAL RECONSTRUCTION OF THE NEURAL PLACODE: PART I—SURGICAL TECHNIQUE. NEUROSURGERY. VOLUME 65 | NUMBER 3 | SEPTEMBER 2009 | 511-529.

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- Sometimes confusing blend of the ventral fat and neural placode
 - Impossible task of separating fat from neural tissue at surgery
 - Associated with sacral agenesis.

Clinical Presentation

Infants	Toddler	Older children	Young adults
Decreased spontaneous leg movements	Delayed walking	Asymmetric motor/ sensory development	Back pain
Absent reflexes	Abnormal gait	Back/leg pain	Leg cramping/pain
Leg atrophy (32%)		UMN signs	Spasticity
Foot asymmetry		Painless ulceration (7%)	Hyperreflexia
Decreased urinary stream			Bowel/bladder incontinence.

Cutaneous Manifestations

SERIES	HOFFMAN ET AL., 1985 (N = 97)	KANEV ET AL., 1990 (N = 80)
Soft tissue mass	97	80
Skin dimple	26	14
Hemangioma	24	9
Hypertrichosis	1	11
Skin tag/tail-like appendage	5	6
Atretic or denuded skin patch	1	
Dermal sinus hypopigmentation	3	3

Sphincter disorders

- Most common trouble - with sacral agenesis (69%).
- **Micturation difficulties**-Incontinence
 - Dysuria,
 - Urge incontinence
 - Dribbling
- Hyperactive bladder, vesicosphincter dyssynergia
- Bladder infection or pyelonephritis

- **Bowel dysfunction** - Rare
 - Severe constipation
 - Faecal retention than to
 - Ass. Urinary disorders

➤ **Orthopaedic syndrome :**

- Club foot (30%)
 - - Limb length discrepancy,
 - High pedal arches,
 - Hammer toes,
 - Calcaneo-varus/ valgus deformity.
-
- Intractable pain in the legs, back, pelvis or perineum(33%).
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- **Sexual Dysfunction**
 - Rarely reported, probably not rare
 - 25%

The Caudal Syndrome

- Currarino – 1981
- Associated with
 - Sacral agenesis,
 - Presacral Mass (more commonly anterior meningocele, teratoma, enteric cyst or lipoma)
 - Perineal malformation (anorectal stenosis in urinary or sexual malformations)
- Incidence - 1.8 to 5.1%.
- Diagnosis - at birth, due to perineal malformations.
- *HLXB9* gene on chromosome 7

Necker Functional Score

Score	Motor	Sensory	Bladder	Bowel
1	Wheelchair <i>Major deficit*</i>	Skin ulceration Amputation	Day and night incontinence <i>Incontinence*</i>	Incontinence
2	Major orthosis Two crutches	Pain	Night incontinence <i>Retention*</i>	Painful constipation <i>Digital maneuvers*</i>
3	Distal orthosis	Painless deficit	Intermittent catheterization	Constipation
4	Fatigue on walking	Normal	Dysuria, stress incontinence	Normal
5	Normal		Normal	

* for children under the age of 3

Asymptomatic patients have a score of 18 and a normal life is possible with a score above 15.

M. Memet Özek • Giuseppe Cinalli • Wirginia J. Maixner. The Spina Bifida Management and Outcome. Springer-Verlag Italia 2008.445-465

Associated anomalies

Cord and Roots Anomalies

- Terminal syringomyelia - 11-27% (terminal)
- Dorsal, cervical and bulbar hydromyelia in 3.3%, 0.6% and 0.3% of cases, respectively.

Associated with diastematomyelia - 9% .

- Dorsal arachnoid cyst above the lipoma
- Enteric cyst.
- Dural arteriovenous malformations,
- Intramedullary mature teratoma have also been reported.

Spinal Malformations

- - Spina bifida- 69%- 89%
 - L5 and sacral –common
- Sacral agenesis rare (27%)
- Scoliosis -malformations of the vertebral bodies or
 - shortening of one lower limb.
- Type I diastematomyelia(8%)
- Hemivertebrae (7 %)
- Vertebral fusion (6%)

Brain Malformations(3.6%)

- Chiari malformations (0.8%)
- Hemispheric cysts
- Dandy-Walker malformation

Other Malformations

- 16 % to 23%
- Urogenital and Anorectal malformations (18%).
- Cardiac, ear, eye, limb, rib malformations

Pre operative Evaluation

Prenatal Diagnosis

- Screening antenatal

Ultrasonography (US)

- From 17 wk
- Differentiate among fat, spinal fluid, and spinal cord
- Skeleton immature with poorly calcified bone,
- Defining the attachment of lipomas
- Relative inexpensiveness, ease of use, and lack of need for patient sedation.
- Disadvantage: operator dependent and possibly miss subtle lesions.

Plain Radiographs

- Dorsal fusion defect in the lamina (bony spina bifida)
- Widening of the spinal canal.
- Varying degrees of agenesis
- Deformity of the sacrum
- Absence or incomplete calcification-limits the utility in children <18 months

Magnetic Resonance Imaging

- Study of choice

- Detail of the spinal cord and filum
- Level of the conus
- Absence or presence and location of fat
- Insertion of the lipoma on the cord
- Rotation of the spinal cord
- Relationship of roots with the lipoma
- Syringomyelia and other malfunction
- Fat sat and Gadolinium- Dermal sinus
- Dynamic MRI-mobility of cord

Computed Tomography

- For Bony abnormalities (e.g., septum in a split cord malformation).
- CT Myelography:
 - some authors recommended
 - Invasive and requires a lumbar puncture,
 - Risk because the conus is low-lying and the bone may be incompletely ossified.

Urodynamic study

- To determine the extent of urologic involvement:
 - Use of Cystometry-
 - Routine
 - Preop and Post op
 - Voiding cystourethrography :
 - To evaluate structural abnormalities or non neurogenic functional problems
- Comparison of postoperative with baselines is useful

Indication of surgery

- Presence of orthopaedic, pain or urologic syndrome
- Neurological symptoms
- Prior to corrective spinal surgery.
- Asymptomatic infants > 2months *

CONSERVATIVE vs SURGERY

“At 9 years, the actual risks of deterioration were 33% for the conservatively treated patients and 46% for the surgically treated patients. The incidences and patterns of neurological deterioration seemed to be very similar, regardless of whether early surgery was performed. These results suggest that conservative treatment of asymptomatic patients is a reasonable option”.

CONSERVATIVE vs SURGERY

“Lipoma of the conus, associated with more severe deficits, and for which surgery cannot be considered to be free of risk and is of questionable long-term efficacy.”

Pierre-Kahn A, Zerah M, Renier D, Cinalli G, Sainte-Rose C, Lellouch-Tubiana A, Brunelle F, Le Merrer M, Giudicelli Y, Pichon J, Kleinknecht B, Nataf F: Congenital lumbosacral lipomas. **Child Nerv Syst** 13:298–335, 1997

CONSERVATIVE vs SURGERY

“Total and near-total resection of lipomas and complete reconstruction of the neural placode produced a much better long-term progression-free probability than partial resection and nonsurgical treatment. There are strong indications that partial resection in many cases produces worse scarring on the neural placode and worse prognosis than no surgery.”

CONSERVATIVE vs SURGERY

“...Author recommended prophylactic surgery which is safe and effective in preventing neurological deficits irrespective of type of lipoma. Most of patients benefit only to some extent even after surgery once deficits developed...”

Kasliwal M, Mahapatra A K., Surgery for Spinal cord lipomas. Indian Journal of Pediatrics.2007,74(4): 357-362.

Principal goals of surgery

- Detach the spinal cord from all tethering structures
- Decompress the intramedullary mass.
- Reconstruct the spinal cord and dural sac
- With Minimizing the risk for neurological deficits and preservation of the functional tissue

SURGICAL TECHNIQUE

Step 1: Exposure:

- Incision
- Laminectomy
- Entry point of stalk to identify

Step 2: Detachment of the Lipoma from the Dura

Step 3: Lipoma Resection

Step 4: Neurulation of the Neural Placode

Step 5: Expansile Duraplasty

Intraoperative Neurophysiological Monitoring

- (1) Posterior tibial and peroneal somatosensory evoked potentials:
 - Detect excessive traction or lateral pressure on the conus
- (2) Pudendal sensory evoked potentials:
 - Detect injury to the S2-4 segments
 - Vulnerable to injury during untethering procedures.
- (3) Bladder and external anal sphincter manometry and lower limb EMG :
 - Difference between nerve roots and tethering bands and scar tissue

Complication

- **Wound complication: 10-25 %**
 - CSF leakage - 2% to 27%
 - over sewn as a first step.
 - If leak persists, re-exploration
 - placement of an external spinal drainage
 - Wound infection & wound breakdown -
 - Necrosis of the overlying skin-
- Development of a pseudomeningocele
- Aseptic meningitis
- Meningitis, Intradural abscess.

➤ **Neurological and Urological deficits:**

- - Transitional lipomas, chaotic lipoma
- 1 to 2% surgery-related

- Transient : - Pain - most constant problem.
 - Disappeared usually in 3 or 4 days.
 - Urinary and motor deficits: 7.5%
 - Regressed in maximum of 6 wks
 - EMG and urodynamic findings - returned to preoperative status.

1) Jeffrey P. Blount, M.D., And Scott Elton, M.D. Spinal lipomas. Neurosurg Focus 10 (1):Article 3, 2001.

2) Satar N, Bauer SB, Scott RM, et al: Late effects of early surgery on lipoma and lipomeningocele in children less than 1 year old. J Urol 157:1434–1437, 1997.

- Permanent neurological complication ,(0 - 4%)
 - Sphincter-related, occurred in 3.4%
 - Urinary retention more frequent than incontinence.

- Delayed-onset deterioration :
 - 3.3 to 7% over 6 months to 20 years

1) Jeffrey P. Blount, M.D., And Scott Elton, M.D. Spinal lipomas. Neurosurg Focus 10 (1):Article 3, 2001.

2) Satar N, Bauer SB, Scott RM, et al: Late effects of early surgery on lipoma and lipomeningocele in children less than 1 year old. J Urol 157:1434–1437, 1997.

Postoperative Outcome of Preoperative Deficits

- - Regardless of the type:
 - Complete Improvement : 27%
patients
 - Partial improvement in 31%.
 - Pain – responds best than any other deficit.
 - Clubfoot and Scoliosis- never improved

Long-Term Postoperative Outcome in Symptomatic Patients

- Surgery beneficial as, 70% of patients were improved or stabilized.
- Better the immediate results, the better the long-term neurological outcome.

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Long-Term Postoperative Outcome in Asymptomatic Patients

- Risk of deterioration 40- 50% over long term .
- Due to
 - Postoperative scarring
 - Adherences
 - Re-tethering.

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Reoperation

- Indicated in
 - Postoperative recurrence or
 - Deficits
- Rate of reoperation- 5 - 10%
- Duration : 5-6 yr(B/w 1st and 2nd surgery)
- Results:
 - Improvement- 25-30 %
 - Stabilization -35-45 %
 - Worsen/continue to deteriorate- 15-25%

McLone DG, Naidich TP (1986) Laser resection of fifty spinal lipomas. Neurosurgery 18(5):611-615

Prognostic factors:

- Type of lipoma
- Quality of the surgery (adequate freeing of the cord)
- Age at surgery
- Malformation complex.
- Pre- operative Neurological deficits(Bladder involvement)

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Conclusion

- Safe and effective treatment :
technologic advances in operative techniques, surgical instrumentation, electrophysiological monitoring, imaging techniques, and our understanding of the disease process.
- The prognosis - If care is provided early.
- Early diagnosis , and surgery should be performed within weeks or months regardless of neuroulogical symptoms.
- Preventing the development or progression of neurological, orthopedic, and urologic deficits should be aim.