**RATIONALE OF TREATMENT OF LIPOMENINGIOMYELOCELE: EXPERIENCE AT BENHA UNIVERSITY HOSPITAL**

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***ABSTRACT***

***Background:*** Lipomyelomeningocele is a spectrum of closed neural tube defects that represents a challenge to neurosurgeons all over the world especially when associated with inherent tethered cord causing neurological deficit

***Objectives:*** we try to define the rationale of managing lipomeningomyelocele according to our experience at the Neurosurgery Department of Benha University Hospital between January 2013 and January 2022,

***Patients and methods:*** retrospective study was carried out on 56 patients with lipomeningiomyeloceledivided into 3 groups

* *Group A:* Patients without neurological or urological deterioration who were treated conservatively include 20 patients
* Group B: Patients without neurological or urological deterioration who underwent prophylactic early surgery include 16 patients
* Group C: Patients who were treated surgically after the onset of neurological or urological deterioration include 20 patients

***Results:*** we noticed that 9 of the patients managed conservatively (45 %) continue to to deteriorate while 7 of the patients managed surgically earlier than the appearance of deterioration (43.8%) continue to deteriorate with no statistically significant difference between the two groups in comparable follow up periods

***Conclusions:*** we support the option of expectant treatment for asymptomatic patients as it carries nearly the same probability of developing deterioration in either nervous or urinary conditionand surgery when deterioration started could stop this deterioration although could not reverse it.

***Key words*:** lipomeningomyelocele, tethered cord

 Lipomeningiomyelocele is a general term that encompses all lumbosacral lipomas.1,2 Lipomyelomeningocele is a spectrum of closed neural tube defects that represents a challenge to neurosurgeons all over the world especially when associated with inherent tethered cord causing neurological deficit.3,4

 The prevalence of lipomyelomeningocele has been found to range between 0.3 and 0.6 per 10,000 live births.5

 A fatty mass positioned in the midline or just off the midline in the lumbosacral region represents the commonest symptom together with other skin lesions, including a hairy nevus, skin dimples, and cutaneous hemangiomas.6,7 Because the fatty mass is clinically apparent at birth, those affected are generally diagnosed before neurological symptoms present, and as many as 48% have been found to be neurologically intact on initial diagnosis.8,9

 The progressive neurological and urinary deficits occur due to the tethered cord syndrome caused by restriction of upward movement of the conus medullaris during axial growth.10,11,12,13

 Urinary dysfunction diagnosed by urodynamic testing can be due to detrusor paresis, external sphincter dysfunction, or most commonly, detrusorsphincter dyssynergy.14

 Postnatal MRI imaging is the invstigation of choice for lipomyelomeningocele and reveals:15

1. expansion of the spinal canal and subarachnoid space.
2. The cord and the dura extend dorsally through the spinal dysraphism.
3. Most cases present with a deformed and stretched neural placode that is rotated toward the lipoma on one side.
4. The meninges herniate on the opposite side.
5. Spinal roots on the side of the lipoma emerge nearer to the neural foramina. These roots are shorter than the roots that emerge from the side where the meninges herniate, and these short roots serve to tether the spinal cord.
6. The neural placode is frequently segmental.
7. A lipomatous dura mater can result if the lipoma surrounds the spinal cord or infiltrates the extradural space.

 Much controversy exists on the treatment of these lesions. Some physicians have advocated surgical treatment for all patients either symptomatic or not, while others have proposed that surgery should be postponed until symptoms develop.16,17

 In this study we try to define the rationale of managing lipomeningomyelocele according to our experience at the Neurosurgery Department of Benha University Hospital between January 2013 and January 2022, this means when, why and how to manage such cases.

**PATIENTS AND METHODS:**

***Study:***A retrospective study was carried out on 66 patients with lipomeningiomyelocele who were treated between January 2013 and January 2022, at the Neurosurgery Department of Benha University Hospital. Radiological examinations, patient records, surgical descriptions were reviewed and statistically analyzed.

***Patient grouping:***

* *Group A:* Patients without neurological or urological deterioration who were treated conservatively include 20 patients
* Group B: Patients without neurological or urological deterioration who underwent prophylactic early surgery include 16 patients
* Group C: Patients who were treated surgically after the onset of neurological or urological deterioration include (9 patients). This group includes also the patients already born with a deficit that recognized later in life (14 patients) and patients that deteriorate after prophylactic surgery (7 patients).

 ***Surgery:***

* General anesthesia in ventral decubitus.
* A rectilinear midline incision starting one vertebra above and extending to one vertebra below the subcutaneous fat mass, and proceeding around the cutaneous stigma.
* Circumferential dissection around the lipoma and/or the lipofibromatous talus.
* The penetration of the lipoma into the thoracolumbar fascia was viewed. The fascia was opened along the midline with a scalpel and the paravertebral muscles were carefully disinserted and laterally retracted using a periosteal elevator.
* The lamina and the spinous processes of the upper and lower vertebrae were viewed and the penetration of the lipoma or lipomatous talus into the osseous failure was observed.
* Laminectomy of the upper vertebra was usually performed. The normal condition of the dura mater and penetration of the lipofibromatoustalus were identified.
* The remaining surgical procedures were performed under magnification. The dura mater was sectioned along the midline, while bypassing the lipoma or lipofibromatous talus. The adhesion of the lipofibromatous talus to the medullary conus, and the relationship with the nerve radicles was identified.
* Using microsurgery scissors, the lipoma was resected as close as possible to the medullary conus, and as much adipose tissue as possible was removed. However, no attempt was made to resect the intramedullary portion of the lipoma, thus leaving the small remainder of the lipoma freely gliding within the canal sac. The arachnoid bands were also sectioned, but care was taken to lyse this adhesion without damaging functional nerve roots. Intraoperative nerve monitoring helps much in that.
* The filum terminale was identified and sectioned.
* Duraplasty with fascia was commonly performed.
* The paravertebral musculature and of the lumbar fascia were then closed in planes. The skin was sutured in a tension free manner.

***Follow up***

Patients were followed up initially monthly for 3 months, then every three months for the rest of follow up period. Patients were examined both neurologically and urologically using Urodynamics

**RESULTS:**

***Preoperatively:*** The 56 patients included 20 males and 36 females women, of ages ranging from 2months to 15 years (average of 8 years).

The following table describes the patient data in details

|  |  |  |  |
| --- | --- | --- | --- |
|  | Group A | Group B | Group C |
| Number  | 20 | 16 | 30 |
| Patient criteriaAge Sex* Male
* female
 | 2m- 4 years (2 y on average)614 | 6y- 11 y (8,5 y on average)610 | 7y- 15y (11 y on average) 1416 |
| Pretreatment dataClinical* lipoma
* G/U malformations
* Skin stigmata

Radiological * Spina bifida
* Low conus

Urodynamics  | 1501218100 | 128121378 | 2461616146 |
| Posttreatment dataImprovedStabilizedDeteriorated  | -119 | -97 | 0300 |
| Follow up period | 18- 60 month | 24 – 65 month | 20 – 72 month |

***Follow up:*** we followed up the patients for a pereiod ranged from1.5 to 6 years (3 years on average)

***Early postoperative (during first month) complications***

* No deaths occurred among our patients
* Transient altered bladder and bowel function after operative procedure in the form of frequent urinary dribbling and tendency to constipate occurred in 6 patients (16.7%).3 patients in group B and the other in group C. This recovered in 2 weeks.
* Reduced lower limb power in the immediate postoperative period which became normal by 5th postoperative day in three patients in group B (15%). These patients were given methylprednisolone in pulse therapy for 3 days after operative procedure.
* One patient in group B had surgical site infection which required local debridement and secondary suturing under antibiotic cover.
* Twelve patients, 8 in group B (40%) and 4 in group C (25%) had persistent seroma formation after removal of suction drain. Seroma was aspirated under aseptic conditions on outpatient department basis, and oral antibiotics were given.
* No CSF leak occurred in our series

***Late postoperative (after first month) complications:*** all in group B

* Four (25%) patients developed decreased lower limb power after surgical procedure. These patients were managed with regular physiotherapy. Improvement occurred after 24 month followup.
* One (6.2%) patient had decreased sensations over plantar aspect of foot and one patient suffered from altered sensations over dorsum and lateral side of foot. Parents of both patients have been taught foot care.
* Two (12.5%) patients had developed urinary incontinence. Clean intermittent catheterization in the daytime with continuous overnight drainage could achieve dry interval period of 3–4 h in both patients. One patient had developed constipation which responded to diet modifications (high residue, low carbohydrate diet) and oral laxatives. Two Patients with fecal pseudoincontinence were managed with rectal washouts

**DISCUSSION:**

 Surgical objectives in a lipomyelomeningocele repair include removal of the adipose mass, identification of the defect in the lumbosacral fascia for release of the tether, possible release of the filum terminale, preservation of neural elements, and prevention of retethering of the spinal cord.6,18

 Timing of surgical intervention has remained a rather controversial topic, with some advocating for intervention prior to presentation of neurological dysfunction, and some advocating for waiting for dysfunction to occur. In the absence of good natural history data on the rate of neurological deterioration secondary to lipomyelomeningocele, it is unclear how many individuals would remain asymptomatic without intervention.7

 ***Rendeli et al., 2007*** concluded that better results could be achieved when surgery was done before the age of one year.19 ***Pang et al***. raise this limit to two years.20 Both argued that early surgery could prevent the deterioration.

 In their study, ***Lavanya and Varsha, 2018*** agreed with ***Pang et al.*** andconcluded thatearly excision of lipomeningocele helped to prevent the development of neurological and urological deficits.21

 In the current study we noticed that 9 of the patients managed conservatively (45 %) continue to to deteriorate while 7 of the patients managed surgically earlier than the appearance of deterioration (43.8%) continue to deteriorate with no statistically significant difference between the two groups in comparable follow up periods. This supports the option of expectant treatment for asymptomatic patients.

 ***Kulkarni et al.*** found that no difference in the incidence of neurological deterioration when early prophylactic surgery was tried or when surgery was postponed till the occurrence of deterioration and concluded that expectant treatment is a reasonable option.17

 For patients of lipomeningocele with established neurological deficits surgery is associated with major risks and is of questionable long-term efficacy.10 A series of eighty patients by Kanev et al. showed that 92.1% of children with a normal preoperative examination had no neurological deficits or bladder dysfunction at long-term follow-up, and all had normal bladder function. However, children with preoperative bowel and bladder paralysis did not show improvement after lipomeningocele repair though patients from this subgroup showed improvement in sensory and motor function of lower limbs.22

For patients of lipomeningocele with established neurological deficitsin our study, surgery could stop the deterioration in all patients although did not reverse the deterioration that had already occurred.

**CONCLUSIONS:** we support the option of expectant treatment for asymptomatic patients as it carries nearly the same probability of developing deterioration in either nervous or urinary conditionand surgery when deterioration started could stop this deterioration although could not reverse it.

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**الملخص العربي**

**استراتيجية علاج التكيس النخاع شوكي الدهني:خبرة مستشفي بنها الجامعي**

**الخلفية العلمية:** التكيس النخاع شوكي الدهني هو احد نطاقات تشوهات الانبوبة العصبية المنغلقة والتي تمثل تحديا لجراحي الاعصاب حول العالم خصوصا بارتباطها بمتلازمة الخبل العصبي المربوط والتي تسبب علل عصبية

**الهدف من الدراسة:**محاولة تعريف استراتيجية علاج التكيس النخاع شوكي الدهني طبقا لحبرة مستشفي بنها الجامعي في الفترة من يناير 2013 حتي يناير 2022

**المرضي ووسائل البحث:** دراسة مرجعية ل 56 مريض ب التكيس النخاع شوكي الدهني والذين قسموا الي ثلاث مجموعات:

1. 20 مرض بلا اي اعراض تم علاجهم تحفظيا
2. 16 مريض بلا اي اعراض خضعوا لجراحة وقائية مبكرة
3. 20 مريض خضعوا لجراحة استئصال ال التكيس النخاع شوكي الدهني بعد بداية الاعراض

**النتائج:**45% من مرضي المجموعة الاولي استمروا بالتدهور بينما 43% من مرضي المجموعة الثانية استمروا بالتدهور دون اي فروق احصائية مهمة

**الخلاصة:**نحن ندعم العلاج التحفظي للمرضي بلا اي علامات وتاجيل الجراحة لما بعد ظهور اي تدهور