

Reoperation in Spinal Dysraphism: Does it Help in Reversing the Neurological Deficits?

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ABSTRACT

Aims: After initial primary repair by inexperienced hands for the spectrum of pathological conditions in spinal dysraphism (SD), a few percentage of patients present with recurrent symptoms and worsening neurological status especially when primarily pathology is not identified and dealt properly. When the primary intradural tethering element is left untouched, worsening of symptoms is common. In this retrospective study, we tried to analyze the symptomatology, functional outcome at 1–2 months after the second surgery and associated complications.

Subjects and Methods: All patients underwent second surgery at author's institution. Pre and post-operative data were evaluated using Necker –Enfants Malades (NEM) neurological and modified Hoffer ambulatory scale. **Results:** The main presenting complaints were bladder incontinence and limb weakness. Preoperative mean scores for motor and bladder were 3.56 and 2.78 out of 5, 2.67 out of 4, and 2.11 out of 3 for bowel and sensory function, respectively. Postoperative mean score for motor, sensory, bladder, and bowel function revealed good neurological improvement. Statistically neurological improvement in bladder and bowel function was significant. More than 60% of patients had normal ambulation at follow-up.

Conclusions: Patients presenting with recurrent symptoms in an operated case of SD need to be investigated, cause of recurrence has to be identified, and if needed repeat surgery is recommended at the earliest. Long-standing neurological deficits can potentially improve, especially bladder and bowel function which gives a good quality of life to the patients. Furthermore, we want to stress the fact that since it is an intradural pathology, these cases should be operated by experienced neurosurgeons, and this fact should be made aware among referring doctors.

KEYWORDS: *Reoperation, spinal dysraphism, tethered cord*

INTRODUCTION

After initial surgery in spinal dysraphism (SD), patient can again present with neurological deterioration which is often termed as recurrent tethered cord (RTC) syndrome. RTC can be due to untouched intradural pathology, inadequate detethering or retethering due to a variety of factors such as adhesions, large residual placode, small size of the canal, and nonrelease of the filum. The recurrence rate of 50% after 5 years of initially release and rate increases up to 57% by 2 years after the second release.^[1] RTC usually follows after improper first surgery when local intradural pathology is not addressed and is common in patients who are

operated by surgeons not trained in handling of neural tissue as well as improper technique. Literature from previous studies states patients presenting with RTC can improve after the second surgery and can lead a better functional life, even in long-standing neglected RTC cases. The main objectives of the study were to evaluate

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clinical symptoms and signs, functional outcome after second surgery, postoperative complications as well as highlighting the anatomical and technical aspects of tethered cord surgery.

SUBJECTS AND METHODS

A retrospective analysis of patients presenting with symptoms and signs of RTC to the Department of Neurosurgery, J N Medical College and Dr. Prabhakar Kore Hospital and MRC, Belagavi, from January 2008 to December 2015, was done. The study included patients who were previously operated for SD and presented with features of RTC. A total of nine patients were included in the study. Patient’s clinical features and indication for the first surgery was noted. All patients underwent magnetic resonance imaging (MRI) of spine and brain to evaluate the cause of RTC and rule out any other associated pathology such as hydrocephalus and Arnold-Chiari malformation. Patient’s pre- and post-operative neurological status were assessed used (NEM) Necker –Enfants Malades neurological scale^[2] [Table 1] and postoperative ambulation using modified Hoffer ambulatory scale [Table 2].^[3,4] The test used for this study was independent sample *t*-test. The decision to perform surgery was based on clinical symptoms such as pain, lower limb weakness, sensory disturbances, bladder and bowel disturbance, neurological deterioration, and radiographic findings including low lying conus, tethering of the spinal cord to the subcutaneous scar or to the inner surface of the spinal canal and presence of intradural lipoma etc. All postoperative outcomes were assessed between 1 and 2 months.

RESULTS

In our series, the youngest patient was aged 1½ years, and oldest was 16 years old. Out of nine patients, six were male and three were female patients. The pathology included patients who were previously operated for meningomyelocele (MMC), lipomenigocele (LMM), tethered cord syndrome (TCS) dermal sinus tract with TCS [Table 3]. All of them had persistence of previous

symptoms which were not resolved after first surgery associated with subsequent neurological deterioration.

Our patients presented with multiple complaints. However, the main presenting complaint was bladder incontinence (eight patients) followed by bowel incontinence, lower limb weakness, trophic ulcers, and foot deformities and backache [Figure 1].

Age at first surgery and diagnosis, age at second surgery, and presenting complaints are (detailed) in Table 3. Preoperative neurological status of patients is shown in Table 4. The surgical procedure was optimized, individualized according to the etiology and specific pathology. The second surgery was aimed at dealing with the intradural pathology with detethering of cord and dysraphic elements with compulsory release of filum. Majority of cases were first operated by pediatric surgeons or in untrained hands who were not experts in dealing with pathology and possibly without the use of operating microscope. Imaging findings and operative procedure are detailed in Table 5.

Following surgery patients were assessed for neurological improvement [Table 6]. At follow-up of 8–10 weeks, all patients had clinical improvement and significant neurological improvement with six patients being

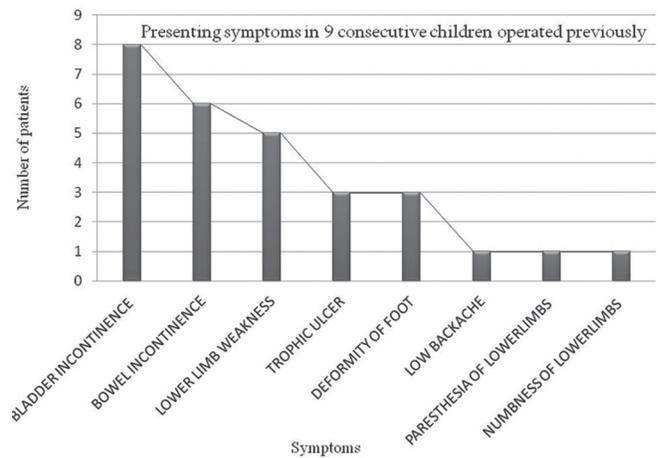


Figure 1: Clinical features in nine consecutive children operated previously

Table 1: The NEM neurological scale*

Score and description					
Area	1	2	3	4	5
Motor	Wheel chair bound	Major orthosis or 2 crutches	Minor or distal orthosis	Fatigue on walking	Normal
Sensory	Skin ulceration or amputation	Pain	Painless sensory deficit	Normal	
Bladder	Incontinence day and night	Nocturnal incontinence	Intermittent catheterization	Dysuria, infection, stress incontinence	Normal
Bowel	Incontinence	Painful constipation	Normal		

*Adapted from Pierre-Kahn et al.^[2] NEM: Necker–Enfants Malades

ambulatory (score of 5) on modified Hoffer ambulatory scale [Table 6]. One patient had cerebrospinal fluid leak which needed re-exploration and repair while the second patient had wound infection which was treated with antibiotics [Table 7].

Analysis shows a operative mean score for motor, sensory, bladder, and bowel function was 3.56, 2.67, 2.78, and 2.11, respectively. Postoperative mean score for motor, sensory, bladder, and bowel function was 4.33, 3.56, 4.78, and 2.89, which indicates good neurological improvement [Table 8].

Statistically significant neurological improvement in bladder ($P = 0.005$) and bowel ($P = 0.032$) function was seen, but improvement in motor and sensory function was statistically insignificant because motor function and sensation was relatively less affected in comparison with bowel and bladder function at second presentation in our study. Follow-up imaging was not done in our study as the patient had no neurological deterioration during their follow-up, but imaging will help in monitoring patient's neurological status on a long-term basis. The limitation of our study was intraoperative neurophysiological monitoring, preoperative urodynamics, and small sample size but IONM and urodynamics evaluation may not be possible in very young children (<3yrs).

Table 2: Modified Hoffer ambulatory scale*

Score	Symptom
1	Nonambulatory
2	Exercise ambulatory (only in therapeutic situations)
3	Household ambulatory (using crutch or brace indoors, wheel chair outdoors)
4	Community ambulatory (ambulate outdoors with or without brace, uses wheelchair for longer distance)
5	Normal ambulatory

*Adapted from Schoenmakers *et al.* and Hoffer *et al.*^[3,4]

DISCUSSION

RTC is a controversial matter surrounded by debate in terms of management and if left untreated can lead to progressive neurological disability. Repair of MMC, LMM, etc., usually follows RTC resulting from the adhesions of the placode within a too narrow spinal canal. Ten percent of those with spinal lipoma and one-third of MMC develop symptomatic RTC, mainly caused by the ischemic-metabolic injuries due to the cord stretching and consequence of arachnoid scarring.^[5-9] Patients with RTC can present with sensorimotor, sphincter-related issues-bowel, and bladder problems which sometimes compels the child to quit schooling due to incontinence, social embarrassment, etc., and also there is a high risk of recurrent urinary tract infection secondary to incomplete emptying, hydronephrosis with renal involvement secondary to reflux. In addition, the patient can develop limb length discrepancy, orthopedics issues such as foot deformities and gait disturbances.

Phuong *et al.*^[10] showed that majority of patients require end organ treatments who do not undergo untethering. Without surgery, patients will often experience a progressive neurological decline.^[11-13] It is important to know symptom characteristics, natural course of disease, and postoperative events to prevent complication and recurrence. Surgery is mandatory to prevent neurological worsening of symptoms. On a technical note, a good neuroanatomical knowledge, preoperative imaging, and renal workup along urodynamics are mandatory. The goal of surgical intervention is to deal with local pathology, debulking and disconnect the fibrous tissue, reconstruction of neural placode with aims to release the conus from the abnormal filum terminale as low as possible. Exposure should be good with caudal and cranial laminectomy; midline durotomy to expose the neural elements below the conus medullaris must be

Table 3: Clinical profile of our patients

Serial number	Age at surgery	First diagnosis	Age at second presentation (years)	Presenting complaints
1	3 years	Lipomeningocele	13	Weakness of lower limbs, bowel and bladder incontinence, trophic ulcer, deformity of foot
2	1 month	Lipomeningocele	5	Weakness of lower limbs, bowl and bladder incontinence
3	3 years	Tethered cord syndrome	16	Paresthesia of lower limb, numbness of lower limbs
4	1 year	Meningomyelocele	16	Bladder incontinence, trophic ulcer, deformity of foot
5	1 month	Meningomyelocele	4	Weakness of lower limbs, bowel and bladder incontinence, trophic ulcer
6	13 days	Meningomyelocele	16	Low backache, bladder incontinence
7	1 year	Dermal sinus tract with tethered cord syndrome	1 and 1/2	Weakness of lower limbs, bowel and bladder incontinence
8	1 year	Meningocele	15	Weakness of lower limbs, bowel and bladder incontinence
9	1 year	Meningocele	16	Bowel and bladder incontinence

Table 4: Preoperative neurological status patient

Patient number	Preoperative NEM score			
	Motor (out of 5)	Sensory (out of 4)	Bladder (out of 5)	Bowel (out of 3)
1	3	1	4	2
2	4	4	2	1
3	5	1	5	3
4	3	1	3	3
5	3	3	1	1
6	5	4	1	3
7	1	2	1	2
8	3	4	4	2
9	5	4	4	2

NEM: Necker-Enfants Malades

Table 5: Radiological diagnosis and surgical details

Patient number	Radiological diagnosis	Surgery performed
1	Conus at L4 with TCS, syringohydromyelia D7-L4, residual lipoma L4-S2	Lipoma excision, detethering of cord and release of filum
2	Conus at S4 with TCS	Detethering of cord, release of filum, arachnoid bands, and adhesiolysis
3	Conus at L5 with TCS, diastematomyelia L2-L5, syringomyelia D12-L2	Detethering of cord, release of filum and adhesion
4	Conus at L4 with TCS	Release of adhesions, detethering of cord, and release of filum
5	Conus at L5 with TCS, Spina bifida-L5, Syringohydromyelia - C5-L5	Detethering of cord, release of filum, reconstruction and closure using G patch
6	Conus at L4 with TCS	Release of adhesion, detethering of cord and release of filum
7	TCS, dermal sinus tract, intramedullary collection L3-S1, syrinx in lumbar region	Drainage of intramedullary abscess, excision of dermal sinus tract, detethering and release of filum
8	Conus at S2 with TCS	Detethering of cord and release of filum
9	Conus at L5 with TCS	Detethering of cord and release of filum

TCS: Tethered cord syndrome

Table 6: Postoperative neurological outcome and ambulatory level

Patient number	Postoperative NEM score				Modified Hoffer ambulatory scale (maximum score-5)	Outcome
	Motor (out of 5)	Sensory (out of 4)	Bladder (out of 5)	Bowel (out of 3)		
1	4	4	5	3	5	Improved
2	4	4	4	2	5	Improved
3	5	4	5	3	5	Improved
4	5	2	5	3	5	Improved
5	4	4	5	3	4	Improved
6	5	4	5	3	5	Improved
7	3	2	4	2	3	Improved
8	4	4	5	3	4	Improved
9	5	4	5	3	5	Improved

NEM: Necker-Enfants Malades

done without any traction on cord. Meticulous dissection using an operative microscope is mandatory due to the presence of extensive arachnoidal adhesion to ensure complete release of the spinal cord in a majority of the cases.

Following the duratomy, entire length of exposure should be checked for arachnoid bands, adhesion, and to identify rootlets. Filum can be identified with its typical dorsal midline location, slightly bluish color with its anteriorly located vessels, and the fat that

often infiltrates it. The absence of filum anteriorly, one may have to search laterally and/or rostrally. Rootlets and arachnoid bands are difficult to differentiate at times. The rootlets at the sacral levels are directed to both sides and may be identified by their size and situation. The arachnoid bands attached to the dura are slightly transparent, flimsy and thinner in diameter when compared to rootlets. The use of IONM may be useful for safe surgery, but in the absence of this, good neuroanatomical knowledge is required to preserve the

Table 7: Complication in nine operated patients for recurrent tethered cord

Patient number	Complication	Treatment
1	No complications	None
2	No complications	None
3	No complications	None
4	No complications	None
5	CSF leak	Exploration/repair
6	No complication	None
7	Wound infection	Intravenous antibiotics and observation
8	No complication	None
9	No complication	None

CSF: Cerebrospinal fluid

Table 8: Statistical score – pre- and post-operative period

Area	Mean		Postoperative (P)
	Preoperative	Postoperative	
Motor	3.56	4.33	0.148
Sensory	2.67	3.56	0.133
Bladder	2.78	4.78	0.005*
Bowel	2.11	2.89	0.032*

neural structures. Filum is sectioned after identification if not already sectioned during the previous operation which was the case in all of the patients in our series; it had not been cut during the previous operation. Detethering is confirmed intraoperatively by cranial ascent of filum. If not search for other cause such as bony spur, assessing the canal size, and the size of the remaining neural placode. Cutting placode dorsal to posterior rootlet line, over sewing of neural placode to accommodate it into the spinal canal with complete circumferential untethering of the spinal cord, finally cutting of ligamentum denticulatum and reconstruction of dural sac must be part of the surgery.

Herman *et al.*^[14] reported on 153 patients with re-TCS (100 patients with MMC and 53 with spinal lipoma) who underwent untethering operations. These authors reported that motor complaints improved in 63% of patients, pain improved in 90% of patients, and bladder function improved in 35% of patients.^[14] In thirty re-TCS surgeries, postoperative improvement was noted most often for pain (81%), and less often for urinary symptoms (53%), and weakness (48%).^[15] The preoperative use urodynamics and Intraoperative Neuro Monitoring (IONM) need a mention here. The use of IONM is feasible in all TCS patients. The identification of functional nervous structures and continuous guarding of the integrity of sacral motor roots by IONM may contribute to the safety of surgical detethering.^[16] Intraoperative use of microscope release of adhesion,

detethering, identification of filum, and its release and water-tight closure of dural closure with layered wound closure are the prerequisites for a favorable postoperative outcome.

CONCLUSIONS

Retethering of spinal cord is a known phenomenon after primary surgery for tethered cord release caused due to various conditions like MMC, LMM etc. Interpretation of MRI is of paramount importance which is missed sometimes leading to non-recognition of pathology in the course of disease. Good release of tethered cord at first instance prevents recurrence in many patients and it has to be done by trained and experienced personnel, who have trained in this field. The challenge lies not only in the release of tethered cord but also in identifying patients with RTC. Second surgery has good outcome in the majority of patients especially in those with early signs and symptoms of retethering and also requires mandatory follow-up after second surgery with multi-disciplinary team. Awareness should be made among referral population, general practitioners, pediatricians, and obstetricians that treatment of RTC should be carried out by trained neurosurgeons, especially in developing countries where there are no specific referral guidelines. Finally to conclude, “an experience in handling such cases would give better results” and requires long term followup, rehabilitation and importance of at least an annual checkup for preventing and managing kidney damage in all patients of tethered cord release.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Archibeck MJ, Smith JT, Carroll KL, Davitt JS, Stevens PM. Surgical release of tethered spinal cord: Survivorship analysis and orthopedic outcome. *J Pediatr Orthop* 1997;17:773-6.
2. Pierre-Kahn A, Zerah M, Renier D, Cinalli G, Sainte-Rose C, Lellouch-Tubiana A, *et al.* Congenital lumbosacral lipomas. *Childs Nerv Syst* 1997;13:298-334.
3. Schoenmakers MA, Gooskens RH, Gulmans VA, Hanlo PW, Vandertop WP, Uiterwaal CS, *et al.* Long-term outcome of neurosurgical untethering on neurosegmental motor and ambulation levels. *Dev Med Child Neurol* 2003;45:551-5.
4. Hoffer MM, Feiwell E, Perry R, Perry J, Bonnett C. Functional ambulation in patients with myelomeningocele. *J Bone Joint Surg Am* 1973;55:137-48.
5. Caldarelli M, Boscarelli A, Massimi L. Recurrent tethered cord: Radiological investigation and management. *Childs Nerv Syst* 2013;29:1601-9.
6. Kanev PM, Lemire RJ, Loeser JD, Berger MS. Management and long-term follow-up review of children with

- lipomyelomeningocele, 1952-1987. *J Neurosurg* 1990;73:48-52.
7. Sakamoto H, Hakuba A, Fujitani K, Nishimura S. Surgical treatment of the retethered spinal cord after repair of lipomyelomeningocele. *J Neurosurg* 1991;74:709-14.
 8. Sutton LN. Lipomyelomeningocele. *Neurosurg Clin N Am* 1995;6:325-38.
 9. Yong RL, Habrock-Bach T, Vaughan M, Kestle JR, Steinbok P. Symptomatic retethering of the spinal cord after section of a tight filum terminale. *Neurosurgery* 2011;68:1594-601.
 10. Phuong LK, Schoeberl KA, Raffel C. Natural history of tethered cord in patients with meningomyelocele. *Neurosurgery* 2002;50:989-93.
 11. Harwood-Nash DC, McHugh K. Diastematomyelia in 172 children: The impact of modern neuroradiology. *Pediatr Neurosurg* 1990-1991;16:247-51.
 12. McLone DG, La Marca F. The tethered spinal cord: Diagnosis, significance, and management. *Semin Pediatr Neurol* 1997;4:192-208.
 13. Kang JK, Lee KS, Jeun SS, Lee IW, Kim MC. Role of surgery for maintaining urological function and prevention of retethering in the treatment of lipomeningomyelocele: Experience recorded in 75 lipomeningomyelocele patients. *Childs Nerv Syst* 2003;19:23-9.
 14. Herman JM, McLone DG, Storrs BB, Dauser RC. Analysis of 153 patients with myelomeningocele or spinal lipoma reoperated upon for a tethered cord. Presentation, management and outcome. *Pediatr Neurosurg* 1993;19:243-9.
 15. Maher CO, Goumnerova L, Madsen JR, Proctor M, Scott RM. Outcome following multiple repeated spinal cord untethering operations. *J Neurosurg* 2007;106 6 Suppl: 434-8.
 16. Hoving EW, Haitzma E, Oude Ophuis CM, Journée HL. The value of intraoperative neurophysiological monitoring in tethered cord surgery. *Childs Nerv Syst* 2011;27:1445-52.