ORIGINAL ARTICLE

Surgical outcome of pediatric patients with symptomatic tethered cord syndrome at a tertiary care hospital

Muhammad Mujahid Sharif¹, Masood Mukhtar², Adil Aziz Khan³, Zalan Khan⁴, Shabana Abbas⁵, Sadaf Abbas⁶

¹ Assistant Professor Neurosurgery, Department of Surgery, Fauji Foundation Hospital, Rawalpindi, Pakistan

^{2, 3} Classified Specialist Neurosurgery, Department of Surgery, Fauji Foundation Hospital, Rawalpindi, Pakistan

^{4, 5} House Officer Neurosurgery, Department of Surgery, Fauji Foundation Hospital, Rawalpindi, Pakistan

⁶ Medical Officer Neurosurgery, Department of Surgery, Fauji Foundation Hospital, Rawalpindi, Pakistan

Author's Contribution

¹ Conceptualization of study and writing ² Data analysis

² Contributed data an

³ Contributed data and analysis tool ^{4, 5,6} Collection of data

Article Info.

Conflict of interest: Nil Funding Sources: Nil

Correspondence

Muhammad Mujahid Sharif doctormujahidns@gmail.com

Cite this article as: Sharif M M, Mukhtar M, Khan A, Khan Z, Abbas S, Abbas S. Surgical outcome of pediatric patients with symptomatic tethered cord syndrome at a tertiary care hospital. JSTMU. 2021; 5(1):23-27.

ABSTRACT

Introduction: Patients with tethered cord syndrome become symptomatic during the period of their growth spurt. Apart from progressive foot and spinal deformities, patients also develop motor deficits and bladder and bowel dysfunction. Early diagnosis and adequate release of the tethered cord are indispensable to successful management.

Objectives: To study the trends in the neurological outcome after surgical treatment of cord tethering in pediatric population.

Methodology: Study was carried out from January 2018 to December 2019 at neurosurgery department, Fauji Foundation Hospital Rawalpindi. Thirty-five patients were included in the study who fulfilled the inclusion criteria.

Results: The average age of the patients at the time of the intervention was 11 years. There were 21 females (60.0%) and 14 males (40.0%). The distribution of neurological status improvement tells improvement in back pain in 28 patients (80%), improvement in motor function in 14 patients (40%), and improvement in urinary incontinence in 7 patients (20%).

Conclusion: This study concludes that back ache improves significantly after surgery while urinary incontinence is the least benefited symptom. Untethering along with the rehabilitation also helps majority of the patients with motor deficit. The ratio of symptomatic relief was best for backache and motor function and then improvement in the urinary control.

Keywords: Backache, Motor deficit, Neurological outcome, Tethered cord syndrome

Introduction

Tethered cord is a disorder which refers to the hitching of the spinal cord to the structures within the spine such as dura, scar tissue, a bony spur or a lipoma of the cord.^{1, 2} If it is related to spinal cord dysplasia, it is classified as primary, and if it develops after surgery on the back, it is classified as secondary.³ Primary tethering is usually from an inelastic short, thick filum which is tethering the cord caudal end as a short & thickened filum terminal.⁴ The secondary tethering is induced by scarring after repair of a spinal meningocele.⁵ Adhesions cause traction on the spinal cord during truncal movements resulting into aggravation of symptoms of the tethered cord syndrome.⁶ Patients usually present with the symptoms of backache, gait difficulty, muscle atrophy, sensory deficit and bladder dysfunction.⁷ Cutaneous findings like hypertrichosis, subcutaneous lipoma, hemangiomatous skin, dermal sinus, are present in the spina bifida occulta.⁸ Orthopedic abnormalities are also present like foot arches, scoliosis and kyphosis.⁹ Radiographically, there is a profound conus medullaris and thick filum terminale.¹⁰ Preoperative cystometrogram is strongly recommended especially if the patient seems incontinent.¹¹ Electrophysiologic studies are also helpful in the early detection of subtle symptomatic cord tethering.¹²

JSTMU Journal of Shifa Tameer-e-Millat University

Aim of the surgery is to prevent the progression of the deficits and relief from the pain.¹³ Early is the surgery, better are the results.^{14, 15} The thickened filum terminal is identified and transected in primary tethered cord syndrome to relieve the symptoms.¹⁶ There are high chances of favorable outcome in primary tethered cord syndrome.^{17, 18} Goal of the surgery is to excise the sac and repair dura in myelomeningoceles the and lipomyelomeningoceles.¹⁹ There is a risk of re-tethering in these patients due to scar formation and adhesions.²⁰ Symptoms of re-tethering do occur during growth spurt.²¹ Multiple untethering surgeries are often required in such patients.²² Pain was the most benefited symptom after surgery.²³ Motor deficit also improved in significant number of patients after six months of vigorous physical therapy. ²⁴ Urinary symptoms were the least to recover.²⁵

Purpose of our study is to highlight the trends in the neurological outcome after surgical treatment of tethered cord syndrome patients. Even motor deficits have favorable outcome but urinary incontinence leaves its mark permanently. Lifelong indwelling urinary catheter is not without complications and such patients need to learn and act the technique of intermittent self-catheterization for the rest of their lives.

Methodology

This prospective study, that comprised of thirty-five patients, was conducted at Fauji Foundation Hospital in the department of Neurosurgery, Rawalpindi from January 2018 to December 2019. Only those patients were included in the study who were symptomatic and there was no previous surgical history for tethered cord syndrome. Asymptomatic patients were excluded from the study. Non probability purposive technique was used in sampling. Patients were enrolled from neurosurgery department of Fauji foundation hospital Rawalpindi. The detailed history was taken and thorough central nervous system examination was performed to assess the preoperative pain and neurological status of the patients. Medical research council (MRC) grading (Table 1) was used for motor functions of the patients and visual analogue scale (VAS) was used for pain assessment (Figure 1). After preoperative assessment, informed consent was taken for inclusion in study. Final outcome was assessed at 06 months of follow up though patients were appraised and

rehabilitated every month. Follow up was ensured through telephone contact. The data was collected by the researcher consultant himself on preformed proforma and result was compiled. All data was interpreted with SPSS version 14.0. Post stratification chi square test was applied taking p-value (≤0.05) as statistically significant.

Table 1: MRC grading of motor function

Muscle Grade	Observation
0	No contraction
1	Ficker or trace contraction
2	Active movement with gravity eliminated
3	Active movement against gravity
4	Active movement against gravity and resistance
5	Normal power

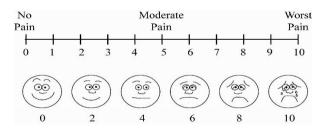


Figure 1: Visual analogue score:

Results

Mean age of the patients was 11 years. There were 10 (28.5%) patients in the age range of 07 -09 years, 15 (42.8%) patients in the age range of 10 -12 years, 10 (28.5%) patients in the age range of 13 -15 years (Table 2). In the distribution of patients by gender, there were 21 (60.0%) female and 14 (40.0%) male patients (Table 3).

At preoperative visit there were 28 (80%) patients who had severe back pain. The final outcome of pain control was assessed at 06 months of the follow up and it was only mild in nature in 28 (80%) patients. At preoperative visit there were 28 (24.0%) patients with moderate to severe motor deficits and after six months of surgery and vigorous physical therapy this percentage fell to 20% (Table 4).

https://j.stmu.edu.pk

JSTMU Journal of Shifa Tameer-e-Millat University

Table 2: Distribution of patients by age (n = 35)

Age (Years)	No. of Patients	Percentage		
07 – 09	10	28.5		
10 – 12	15	42.8		
13 – 15	10	28.5		

Sex	No. of Patients	Percentage		
Male	21	60.0		
Female	14	40.0		
Total	35	100.0		

Table 4: Comparison of pain

Back pain	Preoperative	Postoperative	Preoperative			Postoperative				
Mild	0	28(80%)	T test	Sig	95% CI		T Test	Sig	959	% CI
iiiid	0	20(0070)	1 1001	Olg	Lower	Upper	15.65	.000	Lower	Upper
Moderate	07 (20%)	07(20%)	33.00	.000	2.58	2.92	15.05	.000	1.02	1.32

Table 5: preoperative and postoperative motor function (n = 35).

Motor deficit	Preoperative	Postoperative (06 months)	Chi Square	P-Value
Severe (MRC 0-1)	07	02		
Moderate (MRC 2-3)	21	05	25.22	0.000
Mild (MRC 4-5)	07	28		

*0.05% level of significance.

Table 6: Distribution of patients

Urinary incontinence	Preoperative	Postoperative	Chi Square	P-Value
Yes	35 (100%)	28(80%)	7.778	0.005
No	0	07(20%)	1.110	0.005

*0.05 level of significance

Table 7: Results at 6 months of follow up (Chi square was applied on all parameters collectively)

Neurological status	Yes	No	Chi Square	P- Value
Improvement in pain	28(80%)	7(20%)	26.25	0.000
Improvement in motor	14(40%)	21(60%)		
Improvement in urinary incontinence	07(20%)	28(80%)		

*0.05 level of significance

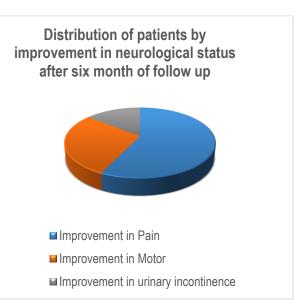


Figure 2: Distribution of patients by improvement in neurological status after six months of follow up

At preoperative visit there were 35 (100%) patients who had urinary incontinence. final outcome of the urinary control was assessed at 06 months of follow up (Table 5). Only 7 (20%) patients got relief from the incontinence but 28 (80%) patients had still urinary incontinence (Table 6). These patients were advised intermittent selfcatheterization. In the distribution of improvement in neurological status, there were 28 (80%) patients had improvement in back pain, 14 (40%) patients had improvement in motor function and 07 (20%) patients had improvement in urinary incontinence (Table 7; Figure 2).

Discussion

Tethered cord syndrome is a set of symptoms consisting of backache, weakness in legs, gait abnormality and inability to control urine.¹ It occurs because spinal cord is anchored with the surrounding structures like dura, lipoma, tumor, scar tissue and adhesions.² Tethering causes abnormal stretch on the distal cord resulting into aggravation of symptoms.³ These symptoms abruptly appear during growth spurt phase of the children.⁴ Tethering of the cord is always associated with spina bifida in children.⁵ Tethering occurs secondarily due to the scar of the myelomeningocele excision in spina bifida apperta.⁶ Spina bifida occulta is associated with diastematomyelia, lipoma of the cord, thickened filum terminale and dermal sinus tract.⁷

Children usually present with the obvious mass on the back, subcutaneous swelling, hyperpigmentation of the skin, hypertrichosis or any dimple on the back.⁸ common complaints are backache aggravated by movements and relieved by rest.⁹ Other symptoms are pain and numbness in the legs, gait abnormalities, scoliosis and poor bladder control.¹⁰ These symptoms increase in their intensity as child grows.¹¹ Magnetic resonance imaging is the gold standard investigation to evaluate the tethering of the cord.¹² Urodynamic studies help to assess the bladder control.¹³

Prompt untethering of the cord is carried out as the symptoms of tethering appear.¹⁴ Surgery varies from section of the tight filum terminal to the exploration of the previous surgical scar and untethering of the cord.¹⁵ Dura is closed while making sure that there are no adhesions of the cord around it.¹⁶ post-operative recoveries are favorable if preoperative motor deficit is mild.¹⁷ Vigorous physical therapy and rehabilitation help in improving the motor abilities and urinary control.¹⁸⁻²⁰ Though there are less chances of post-operative complications, still patient may develop worsening of the motor deficits and poor urinary control.²¹⁻²³

Khan MA conducted a study in which they studied the outcome in children as well adult patients with tethered cord syndrome. Their study showed improvement in backache in 70% of the patients while in our study 80% of the patients got relief from the backache. Their patients had a better outcome as far as urinary symptoms were

concerned.²¹ Their study exhibited improvement in urinary symptoms in 34% of the patients while it was 20% in our study.

Seki T studied the effectiveness of prophylactic surgery in asymptomatic tethered cord syndrome. Mean age at the time of surgery was 26 months. Follow up at 94 months and 177 months showed increasing neurological deficit in 14% of the patients.²² This study is the evidence that repeated release surgeries are needed to untether the cord in pediatric population. Sofuoglu OE were interested to find out the surgical outcome in adults with tethered cord syndrome. Their study showed 60.9% improvement in backache while it was 80% in our study. Urological symptoms improved in 34.8% of the patients while in our study this percentage was 20%.23 Garg K included 24 patients aged ≥16 years who underwent release surgery for tethered cord syndrome. They planned surgery as early as possible after the radiological evidence of tethering. Their results showed 83.3% improvement in backache comparable to our study.²⁴ Urological symptoms improved in 50% of their patients which is a very promising outcome. Patients presented late in our study and showed urological improvement only in 20% of the patients.²⁵

Above discussion emphasizes that backache improves significantly after the surgery. Early recognition of the symptoms of tethering of the cord is very important, especially, the urinary incontinence. If patients are presenting late with months and years of history of incontinence then even untethering the cord may not be fruitful. Most of the studies are showing improvement in backache with a good percentage of 70 to 80% but improvement in urological symptoms is from 20 to 35%. Motor deficits also improve with Aggressive physical therapy and rehabilitation. Multiple release surgeries are considered with recurrent tethered cord syndromes. Early recognition of the symptoms and prompt action carries the good outcome.

Conclusion

It is concluded that early recognition of the symptoms of tethered cord syndrome and prompt release of the cord is advised to achieve better results. Tethered cord syndrome patients who present with backache have good results post operatively than those who present with urinary incontinence.

References

- Khan MA, Ahmad M, Sajjad F, Ali S. Out Come of Surgical Management of Tethered Cord Syndrome. Pak J Neurol Surg. 2015; 19(4):276-80.
- Seki T, Hida K, Yano S, Houkin K. Surgical Outcomes of Pediatric Patients with Asymptomatic Tethered Cord Syndrome. Asian Spine J. 2018; 12(3):551.
 POL https://doi.org/10.4194/jaci.2040.40.2.554
 - DOI: https://doi.org/10.4184/asj.2018.12.3.551
- Tuite GF, Thompson DN, Austin PF, Bauer SB. Evaluation and management of tethered cord syndrome in occult spinal dysraphism: Recommendations from the international children's continence society. Neurourol. Urodyn. 2018; 37(3):890-903. DOI: https://doi.org/10.1002/nau.23382
- Roy RR, Haque SM, Al Mamun A, Rahman M. Tethered Cord Syndrome Causing Chronic Kidney Disease in a Child: A rare event Case Reports. J Pediatr. Nephrol . 2017; 5(2).
- Steinbok P, MacNeily AE, Hengel AR, Afshar K, Landgraf JM, Hader W, et al. Filum section for urinary incontinence in children with occult tethered cord syndrome: a randomized, controlled pilot study. J Urol. 2016; 195(4):1183-8. DOI: https://doi.org/10.1016/j.juro.2015.09.082
- Sofuoglu OE, Abdallah A, Emel E, Ofluoglu AE, Gunes M, Guler B. Management of tethered cord syndrome in adults: experience of 23 cases. Turk Neurosurg. 2017; 27(2):226-36. DOI: https://doi.org/10.5137/1019-5149.jtn.15892-15.1
- Starnoni D, Duff JM, Chittur Viswanathan G. Duplicated filum terminale in non-split cord malformations: An underrecognized cause for treatment failure in tethered cord syndrome. J Spinal Cord Med. 2017; 40(4):481-4.
 DOL https://doi.org/10.00000.0016.12027006
 - DOI: https://doi.org/10.1080/10790268.2016.1227896
- Abdallah A, Emel E, Abdallah BG, Asiltürk M, Sofuoğlu ÖE. Factors affecting the surgical outcomes of tethered cord syndrome in adults: a retrospective study. Neur Review. 2018; 41(1):229-39. DOI: https://doi.org/10.1007/s10143-017-0842-z
- Garg K, Tandon V, Kumar R, Sharma BS, Mahapatra AK. Management of adult tethered cord syndrome: our experience and review of literature. Neurol India. 2014; 62(2):137. DOI: https://doi.org/10.4103/0028-3886.132329
- Hou Y, Sun J, Shi J, Guo Y, Wang Y, Shi G, et al. Clinical evaluation of an innovative operative procedure in the treatment of the tethered cord syndrome. Spine J 2018; 18(6):998-1004. DOI: https://doi.org/10.1016/j.spinee.2017.10.009
- Thuy M, Chaseling R, Fowler A. Spinal cord detethering procedures in children: a 5 year retrospective cohort study of the early post-operative course. J Clin Neurosci. 2015; 22(5):838-42. DOI: https://doi.org/10.1016/j.jocn.2014.11.019
- Seki T, Hida K, Yano S, Sasamori T, Hamauch S, Koyanagi I, et al. Surgical outcome of children and adolescents with tethered cord syndrome. Asian Spine J. 2016; 10(5):940-4. DOI: https://doi.org/10.4184/asj.2016.10.5.940
- Broderick KM, Munoz O, Herndon CA, Joseph DB, Kitchens DM. Utility of urodynamics in the management of asymptomatic tethered cord in children. World J Urol. 2015 1;33(8):1139-42. DOI: https://doi.org/10.1007/s00345-014-1414-2
- Shweikeh F, Al-Khouja L, Nuño M, Johnson JP, Drazin D, Adamo MA. Disparities in clinical and economic outcomes in children and adolescents following surgery for tethered cord syndrome in the United States. J Neurosurg Pediatr. 2015; 15(4):427-33. DOI: https://doi.org/10.3171/2014.9.PEDS14241
- Shukla M, Sardhara J, Sahu RN, Sharma P, Behari S, Jaiswal AK, Srivastava AK, Mehrotra A, Das KK, Bhaisora KS. Adult versus pediatric tethered cord syndrome: Clinicoradiological differences and its management. Asian J Neurosurg. 2018; 13(2):264.

DOI: https://doi.org/10.4103/1793-5482.228566

 Glenn C, Cheema AA, Safavi–Abbasi S, Gross NL, Martin MD, Mapstone TB. Spinal cord detethering in children with tethered cord syndrome and Chiari type 1 malformations. J. Clin Neurosci. 2015; 22(11):1749-52.

DOI: https://doi.org/10.1016/j.jocn.2015.05.023

 Behaine J, Latif AM, Greenfield JP. Fecal incontinence as a predominant symptom in a case of multiply recurrent tethered cord: diagnosis and operative strategies. J Neurosurg Pediatr. 2015; 16(6):748-51.

DOI: https://doi.org/10.3171/2015.5.PEDS15124

- Ur Rehman Z, Khan MM, Ayub S. Surgical management of tethered cord syndrome in children and adolescents. Khyber J Med Sci 2017; 10(1):58.
- Iqbal N, Qadeer M, Sharif SY. Variation in Outcome in Tethered Cord Syndrome. Asian Spine J 2016; 10(4):711-8. DOI: https://doi.org/10.4184/asj.2016.10.4.711
- DOInchez T, John RM. Early identification of tethered cord syndrome: a clinical challenge. J Pediatr Health Care. 2014; 28(3):e23-33. DOI: https://doi.org/10.1016/j.pedhc.2013.06.007
- Yasar S, Dogan A, Kayhan S, Kirmizigoz S, Kaplan A, et al. Surgery for tethered cord syndrome: when and how?. J Turgut Ozal Med Cent. 2018; 25(3):0503-10.
- DOI: https://doi.org/10.5455/annalsmedres.2018.06.124
 22. Lee JY, Kim KH, Park K, Wang KC. Retethering : A Neurosurgical Viewpoint. J Korean Neurosurg Soc. 2020; 63(3):346-357. DOI: https://doi.org/10.3340/jkns.2020.0039
- Shah M, Nouman MA, Khan MM, Nabi A, Tariq M, Ahmed A.Clinical correlates and surgical management of occult spinal dysraphism with tethered cord syndrome. Khyber J Med Sci. 2015; 8(1).
- 24. Granada C, Loveless M, Justice T, Moriarty T, Mutchnick I, Dietrich JE, et al, Hertweck P. Tethered Cord Syndrome in the Pediatric-Adolescent Gynecologic Patient. J Pediatr Adolesc Gy. 2015; 28(5):309-12.

DOI: https://doi.org/10.1016/j.jpag.2014.09.005

 Jalai CM, Wang C, Marascalchi BJ, Horn SR, Poorman GW, Bono OJ, et al. Trends in the presentation, surgical treatment, and outcomes of tethered cord syndrome: A nationwide study from 2001 to 2010. J Clin Neurosci. 2017; 41:92-7 DOI: https://doi.org/10.1016/j.jocn.2017.03.034