

Untethering of the Tethered Cord with Spina Bifida: Experience in the Prince of Wales Hospital, Hong Kong

XL ZHU, HT WONG, WS POON, CK YEUNG

Abstract

The outcomes of 40 spina bifida cases undergone untethering surgery in the Prince of Wales Hospital with a mean follow-up of 4.5 years were reviewed. There were 11 cases with lipoma of filum (4 asymptomatic) and 29 cases with lipoma of conus (10 asymptomatic). The surgical complication rate was zero for the filum lipoma group, 17.2% local complication and 3.4% neurological complication for the conus lipoma group. For the long-term outcomes of the asymptomatic patients, all remained asymptomatic except one case. In symptomatic patients with filum lipoma and conus lipoma, the respective long-term outcomes were 29% and 37% improved, 57% and 53% stable and 14% and 11% deteriorated. The results are consistent with current international data. The results strengthen our policy to recommend untethering surgery for all young children with tethered cord and for patients of all ages who are deteriorating neurologically due to tethered cord. For older children who are asymptomatic or with stable neurological deficit, untethering surgery will be an option opened to the parents and patient.

Key words

Spina bifida; Spinal dysraphism; Spinal lipoma; Tethered cord

Introduction

Spina bifida is a common congenital and developmental disorder in Paediatrics. Due to the abnormal development in the early embryonic stage, the neurological, skeletal-muscular, urogenital and anorectal systems may all be involved, causing impairment and disability. On the other hand, under proper management, for the majority of the

patients, their cognitive function can be well preserved. They are continent and ambulatory socially. They are not only independent in activity of daily living but also working and enjoying normal social life. As a major step to achieve this, a Combined Spina Bifida Clinic has been set up in the Prince of Wales Hospital since 1995 and over a hundred of patients have been managed and followed up in the clinic. One of the major roles of Paediatric Neurosurgery in the management of spina bifida is untethering of the tethered spinal cord, a topic with controversies. In this paper, we review the outcome of our patients undergone untethering surgery from 1992 to 2002, and discussed our policy for the untethering surgery.

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Materials and Methods

All cases received untethering surgery in our hospital and followed up in the Combined Spina Bifida Clinic were recruited in this retrospective study. It has been our routine protocol to have all the patients assessed pre- and post-operatively by the Paediatrician, Paediatric Surgeon,

Orthopaedic Surgeon, Physiotherapist, Occupational Therapists and Spina Bifida Nurse Specialist since 1995. Assessed by Child Assessment Centre was requested as needed. Urodynamic study were performed pre- and post-operatively. Dissection under the microscope, intraoperative nerve root electrical stimulation with lower limb and sphincter electromyography monitoring were routinely employed to prevent damage to the functional nerve tissue. Laminotomy, whenever possible, was performed instead of laminectomy. The medical records, results of investigation and image films were reviewed. Special attention was paid to the surgical complication and functional outcome on follow-up. Urodynamic study results were not analysed in this paper.

Results

From 1992 to 2002, there were 40 cases received untethering operation in our hospital. 18 were female and 22 were male. At the time of operation, four cases were younger than 7 months old, sixteen cases were 7 to 12 months old, twelve cases were 12 to 24 months old, two cases were 3 to 4 years old, three cases were 5 to 12 years old, one cases were 12 to 18 years old and two were adult cases of 38 and 36 years old respectively. Eight cases were operated before 1995. Five cases had myelomeningocele repaired at neonate.

Pre-operatively, 14 cases were asymptomatic, 26 were symptomatic including 12 cases with stable neurological deficits and 14 cases with newly developed neurological symptoms or signs.

Among the 40 cases, 29 had lipoma of the conus, and 11 had lipoma of the filum (tight filum with or without lipoma of the filum). All the cases had a low-lying spinal cord with the conus at L3 or below.

Post-operative complications were summarised in Table 1. There were five cases developed local complication: all were

CSF leak from the wounds required repair or CSF diversion. All the cases with CSF leak were treated successfully without long-term sequel. There one case had neurological complication, in which a boy had both ankle motor powers decreased from 2/5 preoperatively to 0/5 postoperatively. All the surgical complications were occurred in 29 patients with conus lipoma. The complication rates for the conus lipoma group were local complication 17.2%, neurological complication 3.4%.

All the patients were followed up post-operatively for 1 to 11 years (mean 4.6 years, median 4 years). The long-term outcomes were summarised in Table 2. During the follow-up period, 27 cases were stable including 13 of the 14 asymptomatic cases and 14 symptomatic cases. There were 7 patients whose neurological function improved including one in bladder function and six in lower limb function. Two patients with pain all resolved but neurological function static. There four cases deteriorated including one originally asymptomatic case whose bladder function gradually deteriorated, one case with worsening of the clubfoot, and two cases with progressive scoliosis (one of them has VATER syndrome).

Discussion

Common neurosurgical problems associated with spina bifida are tethered cord, myelomeningocele (spina bifida aperta), hydrocephalus, Chiari malformation. Frequently, hydrocephalus and Arnold-Chiari malformation are in

Table 1 Post-operative complications

	Lipoma of filum	Lipoma of conus
	11	29
Local complication	0	5 (17.2%)
Neurological complication	0	1 (3.4%)

Table 2 Long-term outcomes of patients who were asymptomatic and asymptomatic pre-operatively

	Lipoma of filium		Lipoma of conus	
	Asymp Pts*	Symp Pts**	Asymp Pts*	Symp Pts**
	4	7	10	19
Asymtomatic	4 (100%)	0	9 (90%)	0
Improvement	0	2 (29%)	0	7 (37%)
Stable	0	4 (57%)	0	10 (53%)
Deterioration	0	1 (14%)	1 (10%)	2 (11%)

*Asymp Pts = Asymptomatic Patients

**Symp Pts = Symptomatic Patients

association with spina bifida aperta. With the routine folic acid supplement in early pregnancy and prenatal screening, the incidence of spina bifida aperta has been much decreased in recent years. Yet the incidence of spina bifida with lipoma and tethered cord seems no significant change. The management of this group of patient remains a challenge to health workers in the field of Paediatrics.

In newborns, the conus medullaris is at L1/L2 level.^{1,2} "Tethered cord" implies that not only the spinal cord is low-lying but also being fasten and under tension. It can be symptomatic or asymptomatic. Tethering lesions can be a conus lipoma with or without lipomeningocele or lipomyelomeningocele, a tight filum terminale with or without lipoma, or diastematomyelia. The main purpose of untethering surgery is to release the spinal cord from tension created by the tethering lesion in order to prevent neurological deterioration, or to stabilise or improve the existing neurological deficit. It has been shown that complications of untethering surgery for a tethered cord with lipoma of the conus is approaching zero, and the outcome is generally excellent. On the other hand, surgery for the conus lipoma has a much greater risk of inflicting a neurological insult as well as local complication mainly CSF leak and wound infection. The incidence of having local complication after surgery for conus lipoma is about 20%, and neurological complication is about 4%. Regarding symptomatic conus lipoma, after untethering surgery, chance of improvement is about 25%, remained stable about 50%, and further deterioration in about 25%. For neurogenic bladder, chance of returned to normal is less than 20%, and chance of reversal of motor deficit is less than 50%.³⁻⁶ Better outcome in terms of neurological function is associated with early surgery in the infant. There are controversies over prophylactic surgery for asymptomatic lipoma of the conus. Long-term incidence of postoperative deterioration reported 16.7% and 46.9% from big centers in Chicago and France respectively, explaining the different opinions of recommend and not recommend prophylactic surgery for asymptomatic conus lipoma from the two centers respectively.^{7,8} One major problem is that little knowledge of the natural history of lipoma of the conus is known. Retrospective studies showed that about 37-60% of the children who were normal at birth developed neuro-orthopaedic syndrome later with increasing age, especially during the first 2 years of life.^{6,9} With the advanced surgery and anesthesiology for pediatrics, the timing of untethering surgery has become much earlier than previous. The untethering surgery can be done safely nowadays before 6

months of age in an otherwise normal baby.

Review of our own series show that the surgical complication rate is consistent with the international data, also the long-term result in the filum lipoma subgroup and symptomatic conus lipoma subgroup. Our result of the asymptomatic conus lipoma has a lower long-term deterioration rate than the Paris group and close to the Chicago group. However, to interpret the results, several factors should be bared in mind: (1) This is a retrospective study. (2) The number of cases is small. (3) The mean follow-up time is 4.6 years only. It may not be long enough for the true incidence of long-term deterioration. (4) In children who inherited a primary insult in the spine cord and are still in the developmental stage, it is not easy to make judgment that whether their lower limb and sphinctor function are along their own "normal" curve. In other words, it is difficult to tell if a symptomatic child with delayed milestones yet in progress would have been progressed faster if surgery were done. In this regard, the setting of a combined clinic with timely and up-to-date communication with other specialties regarding the child's developmental progress is invaluable.

Currently, for spina bifida occulta patients with tethered cord, our approach is to provide the background knowledge to the parents/patients about the available information on the benefit and risk of the surgery, including the controversy about conus lipoma, and the limited data on its natural history. We recommend untethering surgery for all young children with tethered cord, especially infants; and for patients of all ages who are deteriorating neurologically due to tethered cord. For older children who are asymptomatic or with stable neurological deficit, untethering surgery will be an option. Although a randomised controlled trial is unlikely for the untethering surgery due to ethical reasons, a well-planned cohort study may shed light into this field regarding its long-term results with and without surgery.

Conclusion

The outcome of untethering surgery of our spina bifida patients with tethered cord was reviewed. It showed that the surgical complication rates and long-term outcome are consistent with current international data. The results strengthen our policy to recommend untethering surgery for all young children with tethered cord and for patients of all ages who are deteriorating neurologically due to

tethered cord; For older children who are asymptomatic or with stable neurological deficit, untethering surgery will be an option opened to the parents and patients.

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