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Analysis of different treatment modalities of tethered cord syndrome

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Abstract *Introduction:* Although MRI has improved the diagnosis of tethered cord, many controversies still exist in the treatment of tethered cord syndrome (TCS). Especially the indications for and timing of surgical release have remained topics of discussion. *Materials and methods:* We retrospectively analysed a group of 41 spina bifida occulta (SBO) patients with a tethered cord to evaluate the results of treatment. Patients were divided into four groups. Twelve asymptomatic tethered cord patients underwent prophylactic surgery (group 1). Ten patients were operated upon because of progressive symptoms (group 2). In the third group 9 patients were treated conservatively at first, but underwent surgery after further progression of symptoms. The 10 patients in group 4 were treated conservatively. *Results:* The course of symptoms was analysed in the separate groups. Patients in group 1 remained stable during the mean follow-up of 5.7 years. Neurological symptoms appeared to improve most after early surgery (group 2), but progression of symptoms continued in spite of surgical release.

Keywords Tethered cord syndrome · Spina bifida occulta · Detethering

Introduction

A tethered cord (TC) is a pathologic fixation of the spinal cord. The tethered cord syndrome (TCS) results from this tethering, and it may manifest with progressive neurological, urological and/or orthopaedic symptoms [1, 4, 12, 18]. Knowledge of the pathologic causes of TCS has expanded especially since the development of MRI. Despite these diagnostic improvements many controversies persist about the treatment of TCS. Especially the indications for and the timing of surgical release have remained topics of discussion. Some authors propose that repetitive surgical detethering guarantees a favourable long-term outcome [3, 7, 10, 11, 16, 18]. Others state that progression of symptoms will occur in spite of surgical release [14, 15, 19]. They wonder whether this progressive course may be due to the natural history of TCS, although in fact the natural course of TCS is unknown. In addition to this, the degree of surgical detethering is not easy to quantify. Therefore it remains difficult to evaluate the effectiveness of surgical release objectively. Finally, the diversity of TC lesions and their accompanying symptoms requires a subclassification based on morphology.

In order to contribute to this discussion we retrospectively analysed a group of patients with TC due to spina bifida occulta (SBO) treated by the Spina Bifida Team of the University Hospital Groningen. A total of 41 patients were included. These patients were divided into four groups based on their treatment. The first group consisted of patients who were operated upon in a prophylactic fashion. Patients in the second group were treated surgically because of progression of their symptoms [1, 9, 13, 16, 17, 19]. The third group consisted of patients who were treated conservatively at first, but who were finally operated upon after progression of their symptoms. Patients in the fourth group were treated conservatively. These different treatment modalities are analysed, and the results are discussed.

Patients and methods

A total of 41 patients were included in the analysis. During a 14-year period (1986–1999), 31 patients were operated on. All patients were followed up regularly by the Spina Bifida Team, and all patients had MRI investigations of the spine (Fig. 1).

The first group consists of 12 (6 female and 6 male) patients. Their mean age is 7.7 years (range 1.7–20 years). All patients had SBO. The type of tethering lesion is shown for each in Table 1.

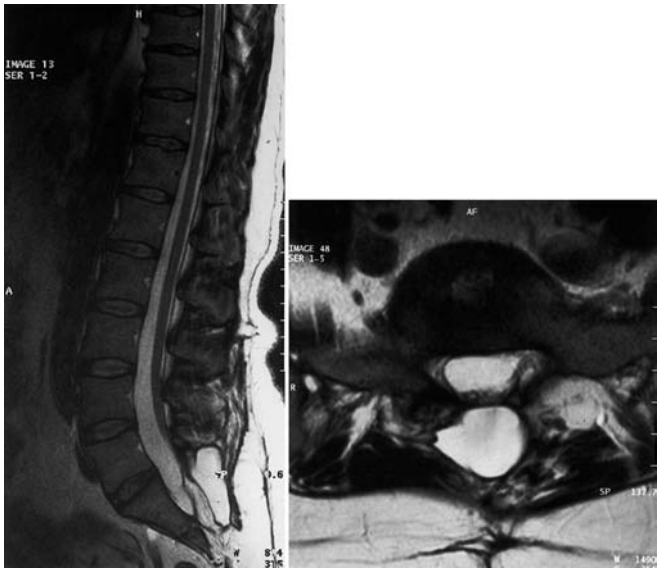

Fig. 1 An illustrative case of a patient with intra spinal lipoma and a thickened filum terminale. This patient had both progressive neurological symptoms and painful symptoms that improved postoperatively

Table 1 Summary of abnormal pathology found on MRI, reported per group (*SCM* spinal cord malformation)

	Group 1 (*n*=12)	Group 2 (*n*=10)	Group 3 (*n*=9)	Group 4 (*n*=10)
Lipoma	6	7	7	4
Filum terminale	6	6	4	3
SCM	1	1		4
Meningocele manqué	2	–	–	–
Dermal sinus	1	–	–	–
Arachnoidal strands	–	1	2	–
Dermoid cyst	–	–	–	1
Teratoma	–	1	–	–
Syrinx	–	1	–	2

The second group consists of 10 (7 female and 3 male) patients,. with a mean age of 27.5 years (range 5.2–54.6 years). The nature of their tethering lesions is shown in Table 1. In the second group 9 patients were operated upon because of progression of neurological symptoms. Neurological decline presented either as gait problems, or as sensory and/or motor deficits. Urological symptoms were present in 3 patients. Pain was reported in 1 patient. In contrast to neurological and urological symptoms, pain and orthopaedic symptoms were never the sole indication for surgery (Table 2).

The third group contains 9 patients, 3 male and 7 female. Their mean age is 28.3 years (range 8.9–49.7 years). All patients showed neurological deterioration. Additional orthopaedic symptoms were present in 3 patients. One patient had additional urological complaints. Pain was present in 3 patients (Table 2). All patients were treated conservatively for more than a year before having surgery. Eventually, progression of symptoms led to the decision that an operation was indicated.

The fourth group consists of 10 patients, 6 male and 4 female, with a mean age of 34.4 years (range 9.0–54.5 years). The nature of their tethering lesions is shown in Table 1.

The changes in symptoms were evaluated during the follow-up period. The outcome was analysed in relation to differences in the results of operation at the end of follow-up and patient age [8, 13].

Results

Group 1

All 12 patients remained stable during the follow-up period. The mean duration of follow-up was 5.7 years (range 0.9–11 years). The mean age at surgery was 1.9 years (range 0.3–9.1 years). Surgical complications developed in 4 patients. Three patients had CSF leakage from the wound, and in 1 of these re-exploration was necessary. Another patient had a temporary bladder retention. All these complications resolved during the early postoperative period.

Group 2

The mean age at operation was 19.3 years (range 1.9–49.7 years). Three patients developed postoperative complications. One patient had a CSF leakage requiring re-exploration. Another patient had urinary problems, and 1 patient had a partial caudal syndrome. All these complications resolved during the early postoperative period.

In the end (after a mean follow-up of 7.1 years), 4 patients showed improvement of their symptoms, 2 pa-

Table 2 Indication(s) for operation by group

[a] Total number of patients in whom this was the indication for operation
[b] In combination with another indication for operation
[c] Only indication for operation

Indication	Group 2			Group 3		
	Total[a]	Combination[b]	Solitary[c]	Total[a]	Combination[b]	Solitary[c]
Neurologic	9	3	6	9	6	3
Orthopaedic				3	3	
Urological	3	2	1	1	1	
Pain	1	1		3	3	

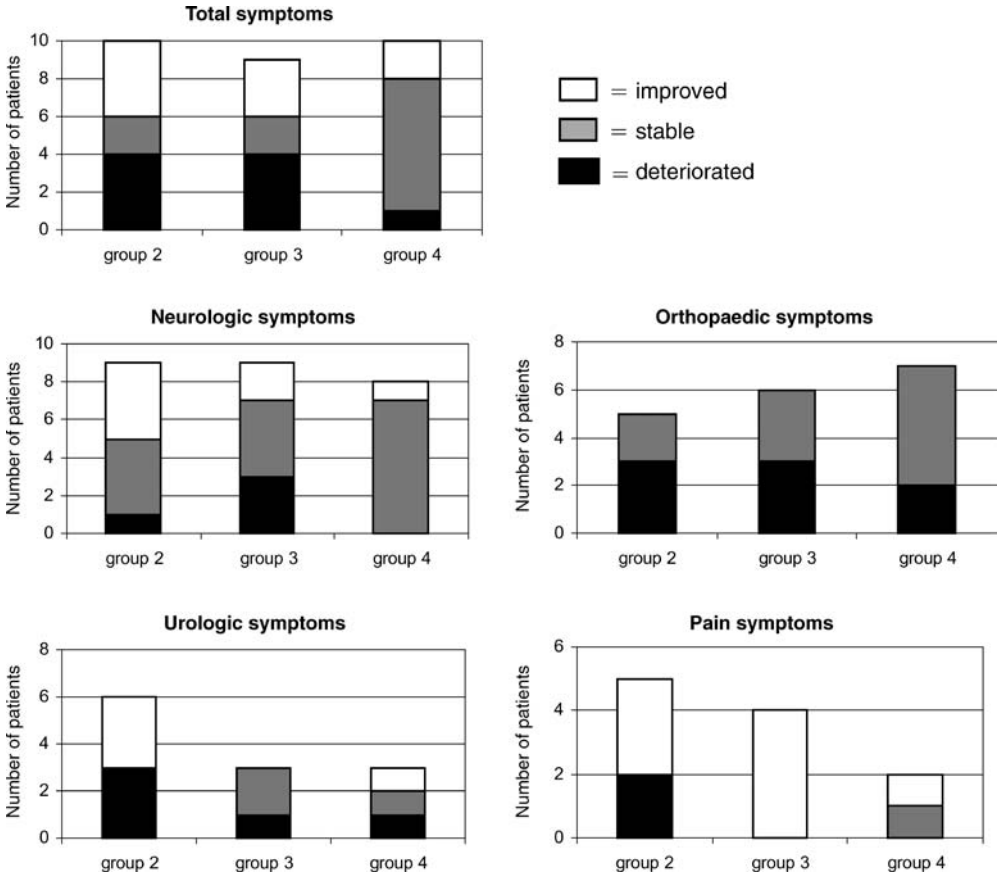

Fig. 2 Results of treatment. Numbers of patients with improvement, stabilisation and deterioration of symptoms

tients remained stable and 4 patients had deteriorated (Fig. 2).

Group 3

The average period of conservative treatment before surgery in this group was 7.8 years (range 1.1–20.9 years). The mean age of these patients at the time of their operations was 24 years (range 3.8–55.3 years). Postoperative complications were seen in 3 patients. One patient had CSF leakage, which resolved spontaneously. One patient had voiding problems, and another patient had liquor hypotension complaints. All complications were resolved during the early postoperative period.

The follow-up period after surgery was 5.1 years (range 0.9–10.2 years). Ultimately (mean total follow up of 12.9 years, ranging between 3.9–29.3 years), 3 patients experienced improvement of their symptoms, 2 patients remained stable, and 4 patients deteriorated (Fig. 2).

Group 4

The mean follow-up in this group was 5.1 years (range 6 months to 20 years). At first contact 9 patients were

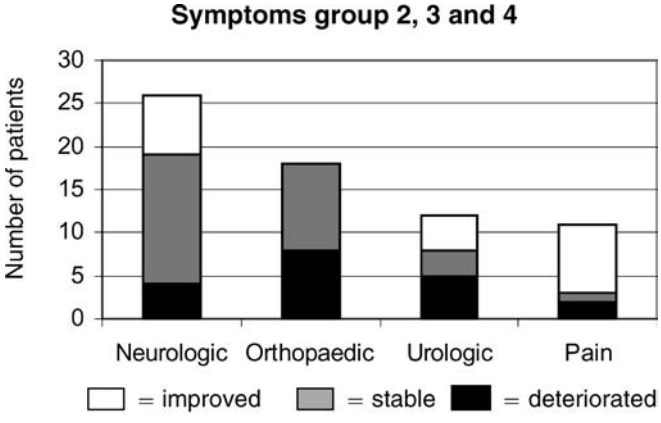

Fig. 3 Results of treatment. Groups 2, 3 and 4 together. Numbers of patients with improvement, stabilisation and deterioration of symptoms

symptomatic. In the end 2 patients experienced spontaneous improvement of symptoms, 7 patients remained stable and 1 patient showed deterioration (Fig. 2).

Concerning the age of onset of the TCS no differences in surgical outcome could be found in these series.

Because the numbers of patients with orthopaedic, urological and pain symptoms are low an analysis per

group is not possible. Differences between the symptoms were only analysed for patients from group 2, 3 and 4 together. There were 25 patients with neurological symptoms, 7 of whom improved. Sixteen patients had orthopaedic symptoms. None of these patients experienced any improvement. Urological symptoms were present in 9 patients, 4 of whom showed improvement. Ten patients had painful symptoms, and 8 of them had improved by the end of follow-up (Fig. 3).

Discussion and conclusions

A retrospective description of a historical group of patients requires a critical discussion. The decisions on how to manage these patients were taken on an individual basis. Therefore, the profiles of the four different groups must be strongly biased and statistical analysis is of limited value. However, the importance of this series must be sought in the recognition of some remarkable aspects like the use of MRI in all patients, the follow-up screening by a multidisciplinary Spina Bifida Team and the evaluation of a nonsurgical group (group 4).

The results observed in the four different groups will be discussed first.

The patients in the first group are characterised by their young age and by the absence of development of symptoms after surgery. Postoperative complications occurred in 33% (4/12) of the patients, but these problems were all resolved. Therefore prophylactic surgery can be carried out safely, and it may have a role in the prevention of symptoms among these young patients. Many authors have emphasized that early operation leads to a better outcome [2, 8, 10, 11, 12, 17], which would provide support for a policy of prophylactic detethering. Long-term follow up is required to evaluate prophylactic surgery.

In the second group, 90% (9/10) of the patients had neurological symptoms. These neurological symptoms appeared to be influenced most by surgery (improved in 40% [4/10], stabilised in 40% [4/10] and worse in 20% [2/10]). Neurological symptoms improved more markedly after operation in the patients in group 2 than in those in group 3 (improved in 22% [2/9], stabilised in 44% [4/9], and deteriorated in 33% [3/9]).

Analysis of the results of operation by the different symptoms shows that neurological and urological symptoms changed in a similar way (Fig. 3). There was a tendency for both to improve or stabilise, although urological symptoms show a more diverse pattern with more deterioration. Orthopaedic symptoms did not improve, and even showed deteriorated. The main orthopaedic symptoms were scoliosis, clubfoot and vertebral deformities, all of which are structural deformities. This may explain why these symptoms cannot improve and stabilisation is the best that can be achieved. Painful symptoms show a distinct pattern separate from that of the other symptoms. They were reported most often by adult patients (age >18 years) [8]. In groups 2 and 3, 62% (8/13) of the adult patients had painful symptoms, while 17% (1/6) of the younger patients reported pain. Many patients experience improvement of painful symptoms.

Group 4 may illustrate the natural course of a selected group of patients. Only 1 patient (10% (1/10)) deteriorated during follow-up.

Comparison of the follow-up periods of groups 2 and 3 (Fig. 4) reveals a difference between the beginning and the end of follow-up in the results of the operation. Patients in group 2 show 60% (6/10) improvement at the first postoperative examination. At the end of follow-up improvement is 40% (4/10). Patients in group 3 show less improvement immediately after operation (44% [4/9]), and at the end of follow-up the improvement rate is down to 33% (3/9). These figures show that improvement after operation is often temporary, patients falling back to the level recorded before the operation. Because patients in group 3 improve less after operation, further

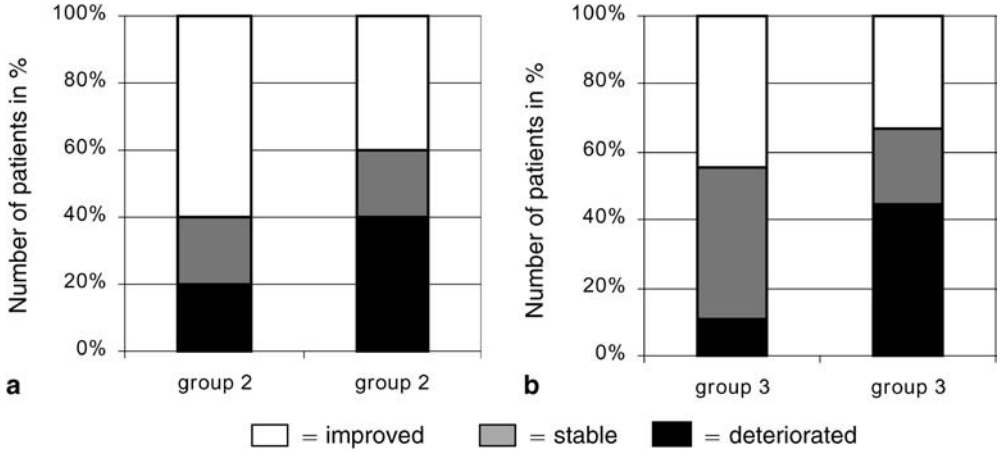

Fig. 4a,b Results **a** at the beginning and **b** at the end of follow-up in groups 2 and 3

deterioration during the follow-up period will take these patients down to a level worse than the level prior to surgery. This series of patients shows that in TCS patients with progression of symptoms the progression often continues despite surgery. For about 70% (20/29) of patients the best that can be achieved in the long term is stabilisation or arrested progression of symptoms.

The role of repetitive surgery cannot be evaluated in our series.

In conclusion, prophylactic detethering appears to be effective among asymptomatic young patients, and it can be done safely in such patients.

Neurological symptoms and pain can be influenced most by surgical detethering.

Early surgical detethering seems to be more effective in improving neurological symptoms.

Once symptomatic, TCS patients tend to show further progression of symptoms even after surgical detethering.

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