Primary Tethered Cord Syndrome

35

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Contents

35.1 Introduction

While tethered cord syndrome (TCS) has often been referred to as a single entity in the past, this view is beginning to change. Secondary forms like those associated with myelomeningocele, lipoma, or trauma may be entirely different from primary ones. Criteria that had been thought to be important like low conus medullaris, thick filum terminale, and permanent cord traction are also now being questioned with respect to their contribution to the pathophysiology and to the clinical picture. Primary TCS may be entirely different from secondary forms related to trauma, myelomeningocele repair, or dysraphism in general. The disorder remains to be a challenge, both for diagnostic and therapeutic measures. Today there is no question, however, that neurosurgery has an important role in its treatment, and indications for surgery recently have rather widened than narrowed. Early diagnosis to prevent neurological deterioration is an urgent requirement, and even prophylactic surgery is now being advocated [2, 20, 61, 69]. On the other hand, there is evidence that a low conus and/or a thick filum does not necessarily lead to clinical symptoms typical for TCS [49, 58].

35.2 History and Definitions

Although there was an early report by Jones in 1891 with the illustrative title “Spina bifida occulta: no paralytic symptoms until seventeen

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years of age; spine trephined to relieve pressure on the cauda equina; recovery" that describes a patient with neonatal history of spina bifida in whom a "fibrous band" crossing the spinal canal had been transected and removed [29], the specific term tethered cord syndrome itself was first applied by Garceau in 1953 [15]. The initial definition included a conus medullaris situated below the level of L1/2 and a thickened filum as "hard criteria." Recently, an entity of an occult tight filum terminale syndrome, characterized by clinical findings consistent with a tethered cord syndrome, but with the conus ending in a normal position, has been recognized [79].

It is because of this controversy that one is hesitating to come up with strong criteria to define the condition. In 1993, Warder and Oaks in a larger series of patients have demonstrated that a low conus is not a necessary condition for TCS. At that time they stated that the disorder is diagnosed by progressive symptoms and thickened filum terminale [77, 78]. In 2001, the thick filum is not a primary issue anymore, and Warder then defines TCS rather along clinical observations as being a progressive form of neurological deterioration that results from spinal cord tethering by various dysraphic spinal abnormalities [76]. Yet 2 years later, tethered spinal cord still is defined as "…a condition in which the conus medullaris ends at a level below the L1-2 intervertebral space" by other authors [69].

It has been indicated that TCS may rather be a functional disorder of the lumbosacral spinal cord, and the only morphological abnormality in the macrolevel is a firm attachment of the spinal cord to the spinal canal. Otherwise, gross anatomical information is not adequate to diagnose the disorder. In a significant number of patients who present with the typical clinical picture of TCS, the diameter of the filum terminale is found within normal limits and the caudal end of the spinal cord is located in the normal position [81]. The combination of an elongated cord and a thick filum terminale, demonstrated by MRI or at operation, is not an essential feature for the diagnosis of TCS [82].

Excessive tension in the lumbosacral cord appears to be the most crucial factor in the pathogenesis of the disorder. Tension in the lower cord is related to impairment of oxidative metabolism in this region [80].

Posterior displacement of the conus and filum on MRI, lack of viscoelasticity by the stretch test of the filum during surgery, and fibrous displacement of glial tissue within the filum by histological studies, according to Yamada et al., would be the prevailing criteria to establish the diagnosis [81]. This would mean, however, that the exact diagnosis is only established *after* surgery. However, the same author has suggested that for TCS diagnosis, emphasis should rather be on its characteristic clinical picture [82].

For this review, we may therefore elaborate on a definition of TCS given by Yamada as early as 1981 [86]. Keeping in mind that spinal cord adhesion is the basic relevant pathogenetic factor, tethered cord syndrome is a *clinical entity* manifested by back and leg pain, progressive motor and sensory changes in the legs, urinary and sphincter control deficiencies, and spinal deformities like kyphoscoliosis. Research has added that it represents a *functional disorder* caused by lumbosacral neuronal dysfunction *due to traction* to and *tension* in the lower spinal cord [32, 81].

This overview focuses on TCS without dysraphism. Clinical signs in this group of patients may develop later in childhood or even in adulthood. The symptoms and findings are often distinct and point to the diagnosis [61]. The tight filum terminale is one major pathogenetic factor in this group of patients.

On imaging, posterior displacement of the conus and filum that attach the posterior arachnoid membrane often is the only radiological criterion [85]. This type of primary TCS may be considered a role model for the pathophysiology of the disorder, and release of the filum is less controversial than in other forms of occult spinal dysraphism or tethering due to tumors. Secondary causes such as arachnoiditis, trauma, and retethering after previous operations will not be discussed here, although they often also deserve the attention of the neurosurgeon.

35.3 Epidemiology and Prevalence

Data regarding the prevalence of primary tethered cord syndrome are rare. A recent survey in 5,499 Turkish primary school children including all primary causes ranging from tight filum terminale over myelomeningocele to intraspinal tumors suggested a prevalence of 0.1 % overall and of 1.4 % in 422 enuretic children [2]. Although the first symptoms usually occur between the age of 5 and 15 years, adult onset of the syndrome is well documented in the literature [1, 16, 20, 33, 35, 41, 44, 51, 59, 85].

Associated developmental disorders, such as cavovarous foot, length differences between the lower limbs, scoliosis, and cutaneous alteration in the lumbar and sacral regions (lipomas, hypertrichosis, hemangiomas, and dimples), may occur with varying frequencies [54, 61].

35.4 Etiology

35.4.1 Normal Anatomy of the Filum and the Conus

The filum terminale is a fibrovascular band which is mainly composed of 5- to 20-μm-thick longitudinal bundles of type 1 collagen separated by 3- to 10-μm intervals. A delicate (0.05–1.5 μm) meshwork of predominantly type 3 collagen transversal fibers connects these bundles. Abundant longitudinally oriented elastic and elaunin fibers are found inside collagen bundles that along with vascular structures add up to a complex three-dimensional structure [12].

It is commonly believed that in healthy humans, the filum fuses with the dura of the lower spinal canal at the level of the second sacral segment. While this is true in the majority, there is a wide range of fusion sites. In a more recent study by Hansasuta et al., the fusion was found to be located from L5 to S3 levels [24]. Moreover, the dural sac itself mostly ends at S2 with a range from S1 to S3. Interestingly, in 4 of 27 cases, the fila fused above the S1 level, and off-the-midline fusion was found in three cases. At birth, the conus medullaris rests at L2/3, and because of the growth of the spine relative to the spinal cord within the first 2 months, it comes to rest at its adult location of L1/L2 [61].

Yundt et al. measured the diameters of the filum terminale in vivo in the operating room on 31 children at an age between 2 and 14 years who underwent selective dorsal rhizotomy for spastic cerebral palsy. None of the children had clinical evidence of TCS. The authors found the diameter of the filum at 10 and 15 mm caudal to the conus to be around 1.2 mm with a standard deviation of little over 0.2 mm [87].

In another study with adult fresh cadavers, the mean diameter at the origin of the filum was slightly greater (1.38 mm) and of higher variability with a range of 0.4–2.5 mm [57]. With a filum thickness of more than 2 mm and with two fila originating below the L2 level, 6 of 41 cadavers fulfilled one of the original anatomical diagnostic criteria for tethered cord syndrome although there was no medical history of respective symptoms.

In 104 TCS patients without spinal dysraphism, Yamada et al. found the conus at the L2–3 intervertebral space or above in 37 and below the L2–3 level in 67 patients. The diameter of the filum was <2 mm in 60 and >or =2 mm in 44 patients [83]. The authors concluded that combination of an elongated cord and a thick filum terminale, demonstrated by MRI or at operation, is no longer an essential feature for the diagnosis of TCS [84].

35.4.2 Pathophysiology

Genetically, TCS has been linked to modifications on chromosomes 21 and 22 [50]. The embryopathy underlying tethering of the filum terminale is not yet satisfactorily understood. George et al. have immunostained 34 fila from patients with tethered cord syndrome. Caudal neural tube developmental markers H4C4 (CD44) and NOT1 exhibited significant alterations in tethered fila compared to controls. The authors suggested that the change in expression

of these markers may be indicative of altered cell identity in the filum and constitute the predisposition to tethering [17].

Embryologically, the disorder apparently is different from overt myelomeningocele and associated Arnold-Chiari type II malformation. Clinically, however, the association of TCS with spinal dysraphic disorders is so striking and obvious [3, 21, 26, 41, 49, 54, 61, 88] that a clear separation into different entities has not become well established yet. It has been postulated that primary TCS is a manifestation of local dysmorphogenesis of all three germ layers at the lumbosacral area, possibly triggered by a hemorrhagic, inflammatory, or some other local lesion occurring in embryogenesis [66].

TCS is also frequently been diagnosed in association with spinal tumors, especially with lipomas [7, 8, 20, 51, 61]. Oi et al. reported that the presence of lipomyelocele may delay development of neurological deficits [49].

The basic mechanical cause of primary TCS without spinal dysraphism clearly is to be found in an anchoring of the caudal end of the spinal cord by an altered, less elastic filum terminale.

Selcuki et al. on histology found that "more connective tissue with dense collagen fibers, some hyalinization and dilated capillaries" were present in the fila from a group of patients presenting with clinical symptoms of TCS and normal MR appearance of the fila. They suggested that this may reflect a decreased elasticity within the filum terminale [69].

When the spinal column grows after birth, it usually does so faster than the spinal cord which is fastened to the base of the brain, the dentate ligaments, and the nerve roots along its course. Stretching of the spinal cord can also occur with flexion and extension movement [71]. During surgery, rostral-to-caudal decrease in diameter, pallor, and absence of pulsation of the spinal cords all indicate traction [61]. Upon release, there is noticeable retraction of the cone in rostral direction, and the formerly linearly oriented and small blood vessels become undulating and widen. Both the lumbosacral spinal cord and the cauda equina fibers have been shown to be most susceptible to stretch and elongation which may initiate metabolic, vascular, and conduction changes [6]. The tethering effect of the tight filum terminale is transmitted to the spinal cord up to the level of D12. Above this level, the dentate ligaments mostly prevent further propagation of the tethering effect. Consequently the tethering force is exerted mainly on the conus medullaris [51, 65].

Constant or intermittent stretching induces traction and tension in the cord, giving rise to functional changes that in turn are responsible for the development of symptoms. Electrophysiological activity is depressed [13], and shifts in metabolism such as those indicated by an altered reduction/oxidation ratio of cytochrome oxidase in the mitochondria of the lower cord ensue [80, 86]. The latter suggests that there is impairment of oxidative metabolism, and other experimentators have confirmed this view [69]. These putative functional changes in TCS occur mainly within the lumbosacral portion of the spinal cord under disproportionate tension [84]. It is not yet clear to what extent they are a direct consequence of micromechanical injury to neurovascular structures or secondary to ischemia due to decreased blood supply. With the application of laser Doppler flowmetry before and after surgical release, it was shown, however, that there is a threefold increase in blood flow 2–3 cm rostral to the site of tethering [67]. Prolonged or accentuated neuronal dysfunction may lead to structural damage to the neuronal perikarya and later of the axons [86]. In animal experimentation, axonal transport of cholinergic enzymes has been shown to be compromised in spinal cord ischemia [43]. Kocak et al. found hypoxanthine and lipid peroxidation levels significantly elevated in an guinea pig model of TCS [36], consistent with the picture in ischemic injury. Subsequently, the latencies of the somatosensory and motor evoked potentials significantly increased, and the amplitudes decreased, indicating a defective conduction in the motor and sensorial nerve fibers. Aside from reversible changes like edema, the group also reported irreversible changes like scarcity of neurofilaments and destruction in axons and damage in myelin sheaths [36].

Thus, it appears that the pathophysiological mechanisms that give rise to the clinical findings in TCS are both axonal and neuronal, both elements being adversely affected by traction and ischemia.

35.5 Clinical Findings

TCS is primarily diagnosed on the basis of its characteristic symptomatology, accentuated by postural changes, since it is a functional disorder of the lumbosacral spinal cord [82]. The most common presenting symptoms of primary TCS are lumbar pain radiating to the lower limbs and lower limb palsy. Patient may also exhibit urinary and anal sphincter control deficiencies and kyphoscoliosis due to disbalance of the muscles innervated by the deficient cauda [54, 55, 82].

Appearance and progression of these symptoms are highly variable and may depend upon age, growth, and physical activity and overlap with changes caused by associated lesions. While changes like neurogenic bladder facilitate early recognition, preexisting neurological deficits, slow development, and acceptance of minor deficiencies may mask the disease [61].

Skeletal abnormalities like foot or leg length discrepancies, pes cavus, and varus or valgus deformities are always suspicious of TCS [47] as are cutaneous abnormalities often pointing to the diagnosis of occult dysraphism [20, 22, 35, 78, 79]. Syringomyelia may be associated or even caused by TCS, and the symptoms can overlap with those of primary TCS [11]. In children, recurrent meningitis of unknown cause as well as recurrent urinary tract infection should raise prompt further clinical investigation.

Late presentation of TCS has been suggested to be related to the degree of tethering and the cumulative effect of repeated microtrauma during flexion and extension [20]. In order to establish an early diagnosis, patients with persistent back or leg pain, neurological deficits, or skeletal deformities should be investigated with MRI.

TCS can have far-reaching consequences for the patient potentially causing major disability if it is not diagnosed and treated properly [9]. Even growth rate of children harboring the disorder may be decreased [64]. For early diagnosis, it is important to notice minor changes. Back pain, enuresis, stress incontinence, and delayed toilet training in children should not be taken casually by the observer. Lower extremity fatigue requiring frequent rest periods, unsteady gait, and postural changes may be important signs in the early phase of spinal cord tethering. This constellation of symptoms, possibly combined with skeletal abnormality, is not confined to childhood and adolescence [30, 49, 51]. In adults, however, onset of clinical symptoms is often precipitated by traumatic events, presumably causing stretch to the conus and the cauda, and clinical changes usually begin more suddenly.

35.6 Imaging and Other Diagnostic Techniques

MRI generally is the method of choice for investigating primary TCS without spinal dysraphism [1, 7, 23, 49, 54, 61]. Radiological criteria for an abnormal filum terminale in the past have included a caliber of more than 2 mm, the conus situated below the L2 vertebral level, and presence of fatty tissue at the level of conus medullaris [45]. These criteria have been rightfully challenged on the basis of the evidence that a significant proportion of symptomatic TCS patients do not show these features on imaging. What can be seen on imaging in these patients, however, is posterior displacement of the conus and filum with attachment on the dura of the lower spinal canal [85]. Although generally, the filum may be surgically sectioned at any level over its entire length, identification and distinction of the structure from the nerve roots of the cauda equina under the surgical microscope may not always be trivial. Thus, precise information on the site of the tether is desirable in the planning of a neurosurgical intervention. On T2-weighted images, nerve roots may have imaging characteristics similar to the filum terminale, and often it is hard to tell both apart even on axial images. Constructive interference of steady-state sequences (CISS) may overcome this dilemma by suppressing the root signal to the advantage of the filum terminale which can then be clearly visualized in its entire length (Fig. 35.1) [63].

The cause of this effect is not clearly known yet. A reduced partial volume effect of the thinner slices in CISS images would seem unlikely, because often there is no significant difference in

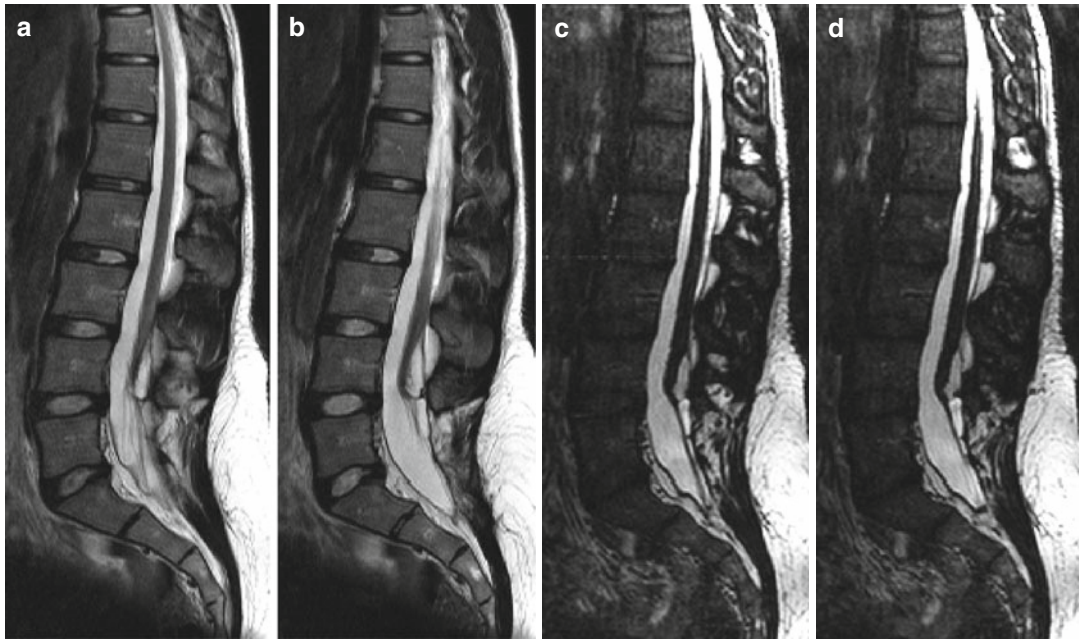

Fig. 35.1 (**a**, **b**) Sagittal MRI of a patient with TCS. An associated lipoma in the lumbar spine had been operated on several years ago. While the nerve roots of the cauda equina and the lipoma give a clear signal in T2-weighted images, the filum and exact site of tethering cannot be identified unequivocally. (**c**, **d**): Sagittal CISS sequences of the lumbar spine of the same patient. The nerve root signal is almost completely suppressed in the median sections of the spinal canal, and the filum terminale is easily recognized

the diameter of the filum and the surrounding nerve roots. It is well possible that the distinction may be caused by the motion of nerve roots as opposed to the relative stillness of the filum.

Scoliosis may present another problem especially in adults, because it will be hard to capture the entire width of the spinal canal in sagittal images [61]. Plain radiographs and computed tomography should always complement MR imaging in suspected spinal dysraphism [51, 54, 61, 83]. Apart from signs of spina bifida, kyphosis, and scoliosis, vertebral body fusion, fused lamina, increased interpeduncular distance, prominent or absent spinous processes, and midline hypercalcifications may be detected [61]. It has also been hypothesized that a horizontally angulated sacrum may predate the clinically appreciable symptoms of a tethered spinal cord after myelomeningocele repair [73]. Ultrasonography can be effective in screening for spinal cord location and tethering as well as for associated meningocele, diastematomyelia, teratoma, and lipoma [60].

SSEPs to tibial and peroneal nerve stimulation are valuable in correlation with static neurological deficit, in demonstrating deterioration on serial investigation before a permanent neurological deficit occurs, and during surgery for monitoring purposes [5, 12, 14, 16, 27, 36, 38, 39, 52, 53, 56, 65, 75, 80, 84, 86].

Since a fixed deficit in bladder function is irreversible by surgical untethering in most cases, early recognition of urinary dysfunction assumes greater importance. This may be accomplished by urocystometry and pelvic electromyography (EMG) [31].

35.7 Indications for and Timing of Surgery

In a series of 60 children from 3 to 18 years of age who met typical clinical criteria for TCS, Wehby et al. retrospectively analyzed the outcome over a mean follow-up period of 13.9 months after sectioning of the tight filum terminale [79]. All patients in this series had their

conus in a normal position on MRI. After surgery, urinary deficiency completely resolved in 52 % and markedly improved in 35 %. Fecal incontinence resolved in 56 % and improved in 41 %. Weakness, sensory abnormalities, and pain improved or resolved in all patients. Surgery was indicated only, when there were signs of spina bifida occulta, progressive neurogenic bladder instability unresponsive to conservative measures, and two or more of the following: fecal incontinence or chronic constipation, lower extremity weakness, gait changes, reflex/tone abnormalities, sensory disturbances, back/leg pain, limb length discrepancy, scoliosis/lordosis, recurrent urinary tract infections, abnormal voiding cystourethrogram/ultrasound, syringomyelia, or neurocutaneous stigmata.

This comprehensive list illustrates the complex clinical work-up involved in decision making for the surgical indication in TCS patients. The study also demonstrates that significant improvement of neurogenic bladder disturbances may be achieved even in patients in whom the disorder has already caused some urinary deficiency as long as primary symptoms are not ignored or accepted and diagnosis of TCS is established early by employing the whole range of diagnostic criteria.

Syringomyelia is associated with spinal dysraphism in a significant number of patients and may successfully be treated by untethering the spinal cord alone. Lee et al. [42] reported that 33 in a series of 135 patients with closed spinal dysraphism presented with a syrinx. In 31 of the 32 patients whose spinal cord was surgically untethered, follow-up imaging showed long-term stability or a decrease in the syrinx index. New urinary symptoms had developed in one patient whose syrinx increased after surgery and retethering could be proved.

When surgery is delayed over a longer period in established disease, results are not as favorable [35, 48, 49, 55, 57, 61, 82]. Since the disease leads to progressive neurological deterioration in most patients with neuroradiologically proven TCS including a low conus and spina bifida occulta, prophylactic surgery has been suggested and performed in children and adults with good results for several years follow-up in asymptomatic patients [62, 74].

Previous surgery (e.g., for myelomeningocele or spinal tumors) does not appear to compromise the results of surgical untethering [33] and therefore should not be a criterion to delay an operation in clinically progressive disease with clear radiological signs of TCS.

Having observed histological changes that reflected decreased elasticity of the filum terminale in urinary incontinent patients with normal results in radiological studies, Selcuki et al. even suggested sectioning of the filum terminale in all of these patients [67].

The indication of surgery in adult patients should be based on clinical symptoms, i.e., the presence and progression of a neurological deficit and pain syndromes that are clearly related to a tethered spinal cord. While radiological evidence in some severely symptomatic cases may not readily be obtained, there are other cases with an unequivocal radiological finding that has no clinical correlate.

From a series of 85 cases, Klekamp [34] has concluded that surgery in adult patients with a tethered cord syndrome should be reserved for those with symptoms. In the 43 patients who had been operated on with or without additional resection of a lipoma or dysraphic cyst, satisfying long-term pain relief was achieved in the majority, and long-term neurological stabilization tended to persist more often than it did in 25 conservatively treated patients.

While there is no consensus about the timing of surgery in primary TCS today, there is clearly a tendency toward early operation in established disease, especially when there are progressive clinical signs. Conservative wait-and-see management in patients with a clinical manifestation possibly related to primary tethering of the spinal cord and no radiological markers would at least necessitate a search for associated anomalies [48], urodynamic investigation, and close follow-up. On the other hand, asymptomatic patients with clear radiological signs of TCS may not become symptomatic for years [10, 28] but should be referred to surgery on appearance of the first symptoms [48, 62].

35.8 Operative Technique and Intraoperative Monitoring

Adherence to some general neurosurgical principles will help to improve operative results. With the patient in prone position, special efforts to ensure that the abdomen is free of pressure should be undertaken in order to reduce abdominal venous pressure and prevent unnecessary blood loss. In the presence of spinal curvature, this may be difficult. A midline incision will usually be carried out which avoids additional cutaneous manifestations and follows scoliotic deformities. In the presence of associated malformations and in recurrent surgery, as a rule the surgeon will work from normal to abnormal anatomy.

In primary TCS without associated lesions, hemilaminectomy will be sufficient to approach and transect the filum. Laminectomy and laminotomy will be necessary in cases of additional tumors or myelomeningocele; the latter should be carefully considered in the presence of multiple arch defects [35, 54, 61].

If primary dural closure cannot be achieved due to a small dural sac or a large surface after tumor resection that would promote retether, dural plasty with either thoracolumbar fascia or artificial material will be required [61], perhaps combined with retention sutures that may help to maintain a relatively normal position of the spinal cord within the thecal sac, thus decreasing the potential adherence of the dorsally scarred aspect of the dysmorphic cord to an overlying graft [70, 72].

Intraoperative monitoring with anal sphincter EMG has long been introduced in surgery for TCS [27]. A more elaborate setup with continuous EMG recording of leg muscles, continuous recording of tibial nerve SSEPs, recording of MEPs evoked by transcranial electrical stimulation, and recording of compound muscle action potentials or SSEP from the scalp upon electrical stimulation of the nerve roots (mapping) provides the surgeon with functional information on the state of the motor and sensory pathways and enables anatomical identification of nerve roots and their distinction from fibrous or neoplastic structures [38, 39, 52, 56].

Because electrical stimulation of the filum will also lead to motor activation due to activation of neighboring nerve roots, and because there is considerable inter-patient variability in electrical thresholds of nerve fibers, von Koch et al. have proposed a ratio, rather than an absolute number, for establishing the electrical criteria for the distinction of the filum and the roots [75]. In over 70 % of their 63 patients, muscle activation via the filum required 100 times the voltage needed to activate a motor root.

35.9 Results and Outcome

Because of the progressive clinical course of the disorder and the good results of untethering, there is general agreement that surgery is the method of choice for treatment of primary TCS. The condition is easily dealt with surgically with little risk of additional injury. It is also universally accepted that the likelihood of some improvement in neurological function and the elimination of pain is high [1, 3, 19, 20, 26, 35, 37, 41, 43, 48, 54, 59, 61, 68, 70, 75, 83]. There are, however, differences in outcome with respect to the extent of tethering and displacement of the conus, the presence of additional lesions like myelomeningocele and tumors, and the age of the patient at the onset of symptoms.

Neurogenic bladder may not improve after surgery at all [36, 46] or only in a small percentage of patients [4, 20, 60, 73], most probably depending on the duration and severity of the disease. The main urologic improvement is seen in bladder capacity [18], probably related to normalization of neurogenic detrusor overactivity [19]. Complete restoration of urinary function to a normal level was reported in all patients in a series of infants up to 3 years of age when surgery is performed shortly after occurrence of the first clinical signs, while untethering in children presenting at birth with upper motor neuron symptoms may result in poorer outcome [10]. Johnson and Levy [28] suggested that children with markedly decreased cord motion on phase MRI would not improve after surgery [28].

In adult patients with primary TCS, neurological findings and urinary deficits show a favorable long-term surgical outcome after tethered cord release, as most patients report improvement or stabilization of their symptoms. In addition, the overall postoperative complication rate is low [40]. A short duration from onset of symptoms to surgery again has also been associated with a good prognosis [25].

Albeit rare, possible complications of surgical untethering are deterioration of motor function [68], postoperative urinary tract infections [75], deterioration of preoperative normal bladder function [18, 19], erectile dysfunction [4], incomplete untethering [33], and cerebrospinal fluid leak [70]. Retethering may occur [33, 40, 68] and can usually be released surgically with good results.

35.10 Conclusive Remarks

In the literature, the term primary tethered cord syndrome apparently stands for at least three different entities – tight filum terminale with and without low conus medullaris as well as TCS associated with myelomeningocele and tumors. These entities are diagnosed at various different stages. It is this variety that mainly accounts for the different outcome reports and the ambiguity with respect to the indication for surgery. Nevertheless, there is common ground to stand on: neurological deterioration in a majority of patients with untreated primary TCS is natural and obvious. It is thus important that the diagnosis is established as early as possible. Surgical untethering ultimately remains the method of choice and should be offered to all patients who experience worsening of their condition. In some with clear evidence of TCS, even prophylactic surgery may be indicated.

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