



The Prevention of the Tethered Cord Syndrome

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Received date: July 02, 2021; Accepted date: July 16, 2021; Published date: July 23, 2021

Abstract

Considering that, in pediatric embryology, it is well known that the spinal cord tends to ascend within the spinal canal, namely soon after birth, till attaining its "adult level" (so rising from the level of the 4th lumbar vertebra till the level between the 2nd and the 3rd Lumbar vertebrae), we believe one is able to prevent the appearance of the Tethered Cord Syndrome by performing a proximal laminectomy just above the dysraphism, after having tubularized the "open" spinal medulla in the cases of a myelomeningocele.

After large dozens of neonatal operations for myelomeningocele not a single case of tethered cord syndrome appeared, leading to the conclusion that that type of complication is really preventable, what knowledge of developmental embryology and simple common sense justifies.

Keywords: Spinal dysraphism; Tethered cord syndrome; Myelomeningocele; Myelocele

Introduction

The severe problem of the Tethered Cord in cases of spinal dysraphism is a relatively infrequent complication, although generally with late appearance and leading to gait and sphincter difficulties that were not initially present. The objective of the technique we are about to describe is to obtain its prevention, by introducing the concept of primary proximal laminectomy (whether one deals with a simple lumbar lipoma or a severe form of myelomeningocele) [1].

Materials and Methods

Routinely performing a laminectomy (one or even two vertebrae), above the dysraphism, really prevents the appearance of a Tethered Cord Syndrome. Although used personally for more than 40 years, that technique has never increased the incidence of late spinal deformities, which could be a negative factor to be considered.

One aims to protect and preserve the exposed neural tissue. So Surgery, particularly in cases of an open wound, should be performed as soon as possible following birth, in order to prevent infection, but obviously not excluding a careful initial evaluation directed mainly to neurological or even urological alterations (namely through echography). The main purposes of surgery of Myelomeningocele are to preserve as much as possible the motor and sensory functions, to

restore the normal cord cerebrospinal fluid environment and to prevent central nervous system infection.

Surgical technique

Under general endotracheal anesthesia, with the patient in the prone position (with the head of the table moderately depressed to minimize eventual CSF loss at the time of incision of the sac, namely in meningoceles), after cleaning the surrounding skin with active antiseptics (hibitane or iodine, but caring that the neural plate is not touched by this solution), the exposed medulla is simply washed with saline. Surgery starts by developing two large cutaneous flaps based inferiorly and externally (associated with two small compensatory triangles), that will later be rotated to the midline (Figure1) [2].



Figure 1: Two rotation flaps, associated with two small compensatory triangles. Incision bordering the neural plate, placed at the interface between the skin and the arachnoid, whose excess will be removed.

It follows an incision bordering the neural plate, which one aims to fully preserve, the incision being made at the interface of the normal skin to the arachnoid, taking care not to introduce skin in the placode. The arachnoid is individualized (showing the terminal portion of the medulla and some nervous branches), its redundant portion is excised and then the dura is individualized.

The freeing of the dura is started at the upper end where one can see the vertebral lamina and is prolonged distally, excising any excess of fat or dura. Then one tries to tubularize the neural plate, through suturing the arachnoid and the dura. The tubularized neural plate will always be significantly larger than the proximal medulla, which some "so-called reputed neurosurgeons" seem not to realize, but is a real fact in our experience.

At this stage, one (or even two) proximal lamina is removed, so that the physiological movement upwards of the bulky medulla can take place during growth (Figure 2).

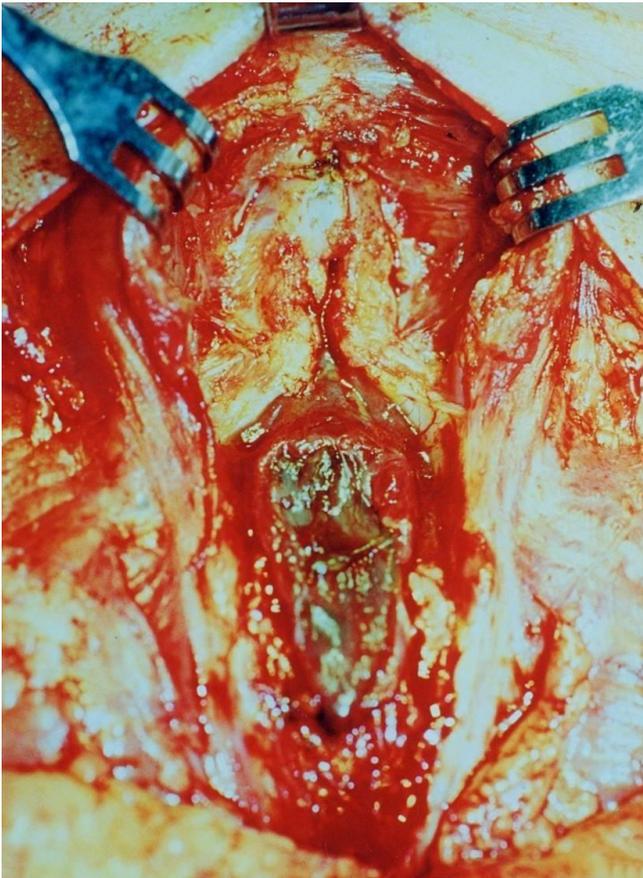


Figure 2: Proximal laminae to be removed.

After removing the laminae a normal looking medulla can be seen at the upper end of the surgical wound. Then, on either side, two longitudinal rectangular flaps are developed from the lumbar fascia and muscles, to be joined by a midline suture (If needed, freeing the flaps as far as the vertebral periosteum, to facilitate their mobilization). The suture of these flaps in the midline leaves a small opening at its proximal end allowing for the drainage of an eventually appearing hematoma [3,4]. In the thoracic and sacral areas it is not possible to provide this additional covering (Figure 3).

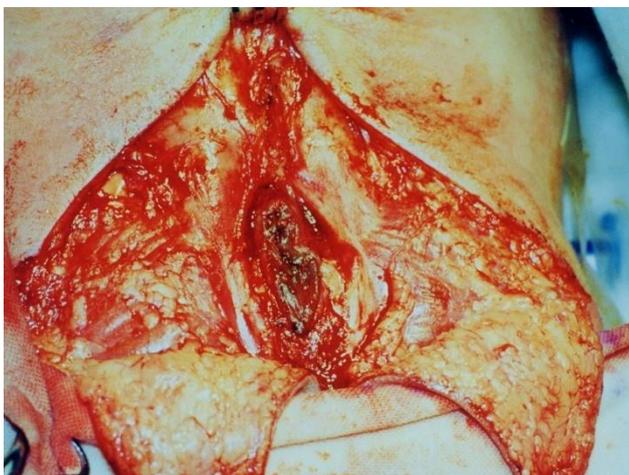


Figure 3: Dura having been tubularized, leaving a small proximal opening allowing for the drainage of an eventually possible hematoma. Two muscles/aponeuroses flap to cover the reconstructed dura.

The skin flaps initially raised are then rotated internally and sutured in the midline, excision being made of the two compensatory skin triangles initially referred to. For 24 to 48 hours a small aspiration drain is placed subcutaneously, through a separate hole. In the immediate post – operative period the patient is kept in a prone position.

Later, in the more frequent forms of dysraphism, localized at the lower lumbar region, the scars will remain concealed in “normal” pants (Figure 4).



Figure 4: Final scars, easily concealed when wearing “normal” pants.

Discussion

Spina bifida is one of the most crippling congenital malformations. Its incidence at world level is above 1 for a 1.000 live births (in Britain being more than double) [5]. The primary abnormality is incomplete fusion of the neural tube and overlying ectoderm, leading to a defect between the vertebral arches. The resulting nerve alterations may lead to paraplegia, incontinence and multiple orthopedic problems. The fusion of the neural folds is completed by the 4th week of embryonic development. The less severe anomalies involve failure of the vertebral arch fusion and protrusion of the meninges to form a meningocele. More severe anomalies involve the neuro-ectoderm with protrusion of the tube itself to form a myelomeningocele [6,7]. The localization of spinal dysraphism is mainly in the lumbar region. In meningoceles [8], if untreated, apart from the dangers of meningitis, epithelialization occurs slowly, leaving a puckered scarring mass covered with poor quality skin, liable to ulceration. A further argument for their early treatment.

The etiology of spinal dysraphism is unknown but there is an evident familial tendency. If a mother has had a child with myelomeningocele, the chances of having another child with the malformation increase significantly. On the other end a deficit in folates seems to be a very important factor for the appearance of these malformations as its incidence is markedly reduced if the mother takes Folic Acid during pregnancy (or even when projecting to get pregnant).

The conus medullaris ascends during the prenatal and postnatal life, mostly prenatal, from 8 to 25 weeks lying opposite L 2-3 disc space at birth, and ascending to its adult level, opposite or even cranial to the

1st/2nd disk space, by two months after birth [9-11]. Surprisingly, that fact is never valued in any of the well-known books on pediatric neurosurgery or even in articles on scientific journals, and surprisingly, quite wrongly, some even seem to doubt its existence.

Some more recent authors based in radiological studies of normal infants believe that, at birth, the conus medullaris is already at adult level. Nevertheless I believe we cannot rely on their studies performed in normal babies if one is dealing with spine malformed neonates [12-15].

Serial sections of embryos with myelomeningoceles [16-19] showed that the bulk of the neural plate tissue is much greater in the region of the spine defect than in the normal adjacent normal segments of the spinal cord. The neural tube defect is established prior to the spina bifida [20-22] and so the vertebral deformities would be a secondary phenomenon. It has been assumed that the ascent of the spinal cord within the osseous vertebral canal, taking place largely through differential growth, has a progressive tethering effect. More caudally located conus causes a suspicion of future spinal cord tethering.

The appearance of the tethered cord syndrome later in life, normally at adolescence, requiring difficult untethering of the cord because of spinal cord dysfunction, has been practically eliminated using the proposed primary laminectomy. After 40 years work (1960/2000) at the children's hospital de D Estefânia, in Lisboa, not a single case was detected at the Alcoitão rehabilitation center, where the patients with neurological sequelae (from the Lisbon area) are generally sent for rehabilitation. And identical results have so far happened with the pediatric neurosurgeon that follows the technique here advised. Nevertheless the same was not true for patients operated by other neurosurgeons, according to the "classical" method proposed in the literature (with no "preventive" laminectomy).

The argument used by a Belgian Urologist, when referring to another Surgeon that "claimed that he never had a fistula following hypospadias repair, could only be the result that he had not done many", certainly does not apply here (with my absence of a single known case of the Syndrome following the many dozens of patients I have treated "my way").

It is quite obvious that the tethered cord syndrome being rather infrequent, it is obviously impossible to have statistically significant figures. But a famous Nobel Prize winner said once that "the best way not to improve, is to be satisfied with what one is doing" Also there is an excellent English saying: "Guidelines are no god's lines". One must always think and try to understand if what he is doing is the best solution for his Patient. Prevention is certainly better (and even cheaper) than treatment!

According to Wolfe et al. no effective method to prevent re-tethering when a tethered cord syndrome is surgically treated, has been described. The fact that, when trying to correct a tethered cord syndrome, one may find that sticking is mainly between the neural placode and the dural scar and associated arachnoid adhesions, does not invalidate the placode's trial to ascend in the neonatal period [23,24].

In spite of repairing the spinal malformation, it remains essential the measurement of the cephalic perimeter and the performance of a transfontanelle areography for an early diagnosis of an eventual hydrocephalus, (that would impose a ventriculo-peritoneal shunt, at least after a short period of "armed expectancy").

Conclusion

In myelomeningoceles, surgery should be performed as early as possible, soon after birth, almost as an emergency. In the cases of a lipoma in a dysraphism, after clarification through MRI, surgery may be deferred to a later time, when the child seems fit (but not too late).

It is essential to minimize trauma to the neural plate, protecting it through good fascial muscle flaps, in the case a small dehiscence appears in the area in which the cutaneous flaps meet (fortunately an extremely rare event in our hands). Careful hemostasis is essential and the use of an electric stimulator is advisable, as one aims to preserve all the existing neural branches. The use of antibiotics is obviously indicated.

Proximal laminectomy, as a basic principle in the treatment of spinal dysraphism, and its role in the prevention of the tethered cord syndrome, is not quoted in any Article or Chapter of a Neurosurgery Book. But it is an easy, safe and not lengthy operation that should become an essential (and even routine practice), in all surgery for those malformations.

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