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Caudal Traction as a Pathogenetic Mechanism of Chiari Malformation Type I

Miguel Bautista Royo-Salvador, Marco Fiallos-Rivera and Horia Salca

Abstract

Despite the important achievements made with respect to our understanding of their clinical and image features, Chiari malformations are the result of etiopathogenetic mechanisms still sunk into mystery, while most of the efforts to dissipate it are isolated attempts that deal with rather late, secondary pathogenetic events, such as the reduction of the posterior fossa volume, the crowdedness of its contents or the disturbances of the cerebrospinal fluid flow at the level of the foramen magnum. Nevertheless, until new research will shed light onto many of these processes, the actual partial, fragmented knowledge can be structured in a much more reliable manner if one holds the theory of caudal traction as a guiding principle. We present a potential pathogenesis that could culminate into an abnormal axial tension throughout the spinal cord, as well as some image and therapeutic features found during our clinical practice, testifying in favor of this relentless caudal traction.

Keywords: Chiari malformation, tonsillar descent, hindbrain, spinal cord, caudal traction

1. Introduction

The commonest variant of Chiari malformations, the one that has been labeled "type I," including some recently derived variants (type 0, type 1.5), is unique among the central nervous system abnormalities by its capacity to elicit just as much apprehension within the community of patients, as bewilderment among the clinicians. Its ominous relationship with sudden death, as well as its resemblance with the tonsillar herniation seen in terminal stages of brain tumors, intracranial hemorrhage, and other space-occupying lesions, would very well serve to explain many of these feelings. Nevertheless, their deeper reason seems rather to be the apparent mystery that clouds its pathogenesis, hindering many attempts at agreement among the authors involved in its investigation.

Notwithstanding, an attentive eye can discover interesting pathogenetic clues issued from recent research that one only has to pin up at the right spots on an older scaffold initiated long ago by some intuitive theories that started to explore into these matters even from the discovery of the hindbrain malformation: while Hans Chiari favored hydrocephalus as the cause of tonsillar descent, Julius Arnold proposed the concept that cord tethering at the level of the associated

myelomeningocele determines a caudal traction along the spinal cord that ends in the tonsillar descent of Chiari malformation type II [1].

This is why every effort to unveil the origin and the mechanisms of formation of Chiari malformation type I should be greatly welcomed. It is very likely that the same can be extrapolated to the less common Chiari malformation type II, which could be just a more severe form of the same deformity, caused by more intense but qualitatively similar pathogenetic alterations. The unifying theory that follows is merely the result of attentive, scrupulous efforts to acknowledge valuable data in the middle of puzzling research results and connect them orderly in a logical explanation of the mechanisms likely to be involved in the production of Chiari malformation type I.

The concept of caudal traction as we use it through the following lines should not be understood merely from a physical point of view, as a purely mechanical force, as it refers to a biological system with certain viscoelastic properties and an intrinsic capacity to develop a reaction to any force acting upon it. The development of the human body is a continuous interplay of genetic, molecular, biochemical, and mechanical changes that result in a more or less dynamic structure and function. Absolutely all human beings, as well as other vertebrate species, are subjected to this phenomenon of caudal traction, which is a necessary part of the development of the spinal cord and brainstem, as they grow by lengthening, distinctly from the forebrain and cerebellum, which do it by expansion. In fact, the notion of caudal traction points to a group of deformities of the nervous system and its surrounding tissues, identifiable on diagnostic images and likely to result from this longitudinal growth of its caudal segments during development; they may be discovered at various stages during this process or even later, during adulthood, which is by no means a cease of it, but merely a continuation, as an involution—apparently a reversed process, but in fact an ongoing, caudal traction at a deep structural level of the involved neural organs. After all, the definition and understanding of this dynamic concept will certainly improve in parallel with the abilities of the diagnostic tools that we shall be able to use in these patients.

After an initial presentation of this new pathogenetic theory, we will follow with a second part where we shall bring into view some conditions quite likely to be produced by means of a mechanism of caudal traction and which are frequently associated with Chiari malformation type I. The third part of this chapter will deal with the clinical arguments of our demonstration, presenting a range of suggestive, but often neglected proofs of this pathogenesis, which we meet during the diagnosis and treatment of these patients.

2. Embryology of asynchronism

It is very likely that the events that eventually lead to Chiari malformations take place at a very early stage during embryogenesis; a plausible idea if one takes as an example the defects of neural tube closure, related in some way to our problem, as we know well enough that during their evolution, some of them can cause a Chiari malformation type II—and the future will probably show that the relationship between these conditions is not limited to this (and caudal traction could be the link). If some parallel, very early, processes, related but not identical to neurulation abnormalities, would finally result in a Chiari malformation type I, it means that actually all purported etiopathogenetic mechanisms of this condition are in fact late secondary features that simply result from the abnormal development of the cranio-cervical junction. Most importantly, both the small volume of the posterior fossa and the disturbances of cerebrospinal fluid circulation across the foramen magnum would be such *effects* wrongly converted in *causes* by the most prevalent theories that try to explain at present the genesis of Chiari malformations.

This concept of very early pathogenesis of Chiari malformation type I has also another important consequence in the way we should try to understand it: most, if not all of the morphological and mechanical changes involved in its generation take place in the diminute body of a human embryo, then fetus, and then child (probably of a comparatively decreasing magnitude throughout these stages), even though the diagnosis will eventually be secured only at an adult age. This invalidates many recent research results and actual misconceptions based on mature or adult human anatomy and physiology.

Perhaps Chiari malformation type I is the best example of the meaning of Lewis Wolpert's famous phrase "It is not birth, marriage, or death, but gastrulation which is truly the most important time in your life" [2], as indeed, the events that finally lead to its development seem to originate during gastrulation (third week postfertilization), that is, at a much earlier stage of embrionary development than that stated by all theories invoked nowadays.

Thus, the primordium of the central nervous system divides along its freshly defined anterior-posterior axis into four regions, corresponding to the future forebrain, midbrain, hindbrain, and spinal cord [3], well in advance of any significant differences in shape or length among them. Interestingly, while the first two limits are represented by discrete junctional areas that function as organizing centers for nearby neural territories—the so-called anterior neural ridge between the forebrain and midbrain and the isthmic organizer between the midbrain and anterior hindbrain [4]—there is no specific anatomical hint as to the precise location of the posterior hindbrain-spinal cord transition [3]; moreover, its final position depends on quite sophisticated but also delicate mechanisms involving a negative feedback loop between retinoic acid signaling, Cdx4 transcription factor, and the Cyp26 enzyme involved in the degradation of retinoic acid [3, 5]. Despite its importance for all future development of the nervous system, this hindbrain-spinal cord transition is exposed to be moved cranially or caudally by various alterations in these complex, interconnected signaling pathways [3, 5]. For example, experimental loss of Cdx4 function in zebrafish led to caudal displacement of the transition as far as that corresponding to two somites inside the spinal cord territory. As a consequence, the hindbrain-spinal cord transition along the developing neural tube will be matched to a different mesodermal counterpart, belonging to the first pairs of somites, either occipital or cervical. In this way, it becomes easy to figure out how an alteration of the Cdx4 gene or an equivalent disturbance of retinoic acid signaling could displace the transition caudally and place the junction between the developing brainstem and the spinal cord at the level of the future atlas, while the cerebellum might be expected to expand until the same area well below the occipital foramen. In fact, maternal administration of exogenous retinoic acid has been used to produce Chiari malformations in an experimental model in hamsters [6].

By and large, the tonsillar descent seen in Chiari malformation type I would thus be the result of delicate molecular abnormalities that occur early in a critical area of the future body plan, representing the precise border separating the head, with a neural-driven expansile growth in three directions, from the spine, with a somatic-driven tensile growth in one predominant direction (**Figure 1**). This is why its developmental importance and pathological associations and consequences are so complex and puzzling in their diversity, far outweighing the apparent trivial significance that it still has in the eyes of many clinicians.

Of course, it is difficult to apply such an ultra-early pathogenesis involving molecular and genetic signaling pathways to what we actually think and know about Chiari malformation type I, but here we have again a point where an analogy with Chiari malformation type II is quite welcome. Since Julius Arnold's days, it was already supposed that a myelomeningocele would "tether" the growing child's
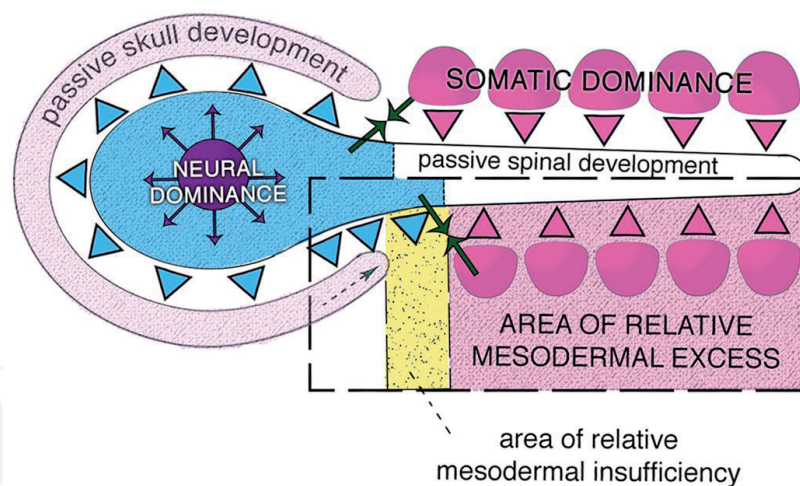
Figure 1.
The interplay of molecular and growth influences that grant a special importance to the cranio-cervical junction.

spinal cord and thus determine a progressive caudal traction on the cerebellum and the tonsillar descent through the occipital foramen. The concept of "tethering" involves both a pathological lesion that fixes the spinal cord to the vertebral column at some point and an uncompensated load, either in the form of continuous growth, repetitive forward flexion movements, or a fracture-dislocation with sudden cord traction. In fact, there is also a third, mandatory component, the lack of an adequate adaptive reaction of the body, which may be due to the overwhelming intensity, suddenness, or persistence of the pull. Now, all these features can yet be expressed in another way if we view tethering more generally, as a relative shortness of the spinal cord, underlying both Chiari malformation types I and II, with the mention that in the former, there is absolutely nothing of spinal cord tethering.

Therefore, patients with Chiari malformation type I would tend to have relatively shorter spinal cords because the neural territory assigned to the formation of the future spinal cord is reduced with respect to the nearby somitic mesoderm, a disproportion that will result in a continuous spinal cord tension during growth, as the neural tissue will always be "one step behind" its mesodermal counterpart (**Figure 1**).

Interestingly, similar arguments in favor of these pathogenetic mechanisms have come from the other side of the problem, that is, from attempts to explain a supposedly defective development of the mesodermal tissue composing the prospective vertebral column, resulting in idiopathic scoliosis: the so-called "Roth-Porter theory" invokes exactly the same "asynchronism" between the spinal cord and spine during growth [7–9], reflected in the tridimensional deformity of idiopathic scoliosis in a much more visible manner [10] than in the case of Chiari malformation type I, but we shall develop more of these aspects later.

Just as a collateral observation, here we should mention that retinoic acid was also suspected as a pathogenetic factor in adolescent idiopathic scoliosis [11].

By the way, given its role of mechanical support, the development of osseous tissue has always been regarded as being associated to the creation and maintenance of tensile or compressive forces in the neighboring tissues. Thus, the development of the cranial vault by intramembranous ossification seems to proceed by means of tensile forces created in the sutures by the growth of the underlying brain [12]. Just in the same way, it should not be surprising that the growing vertebral column could exert a barely perceptible but relentless, tensile force that by some yet unknown mechanism stimulates the growth of the contained spinal cord accordingly.

Well, this would be exactly the Achilles' heel in individuals with Chiari malformation type I, as they are exposed more than normal people to a deficiency of the homeostatic mechanisms that maintain coupled the growth of the two structures. In selected cases, this uncoupling can occur also in the absence of a tonsillar descent or with a minimal one, so that its pathological consequences do not require the 5 mm of descent that most authors use to define Chiari malformations.

3. Illustrative associations

Without pretending to be exhaustive, Chiari malformation type I is associated with a few pathological conditions that could be explained by similar mechanisms involving genetic and molecular abnormalities followed by an axial traction throughout the spinal cord and the brainstem. But before all, in order to have a crystal clear vision of these associations, we have to rule out any tonsillar descent that is obviously secondary to compressive forces from above, as in benign intracranial hypertension, hydrocephalus of any etiology, craniosynostosis, or Paget disease of the bone and other conditions with calvarial thickening, as these are not real instances of Chiari malformation [1] and only compound the problem unnecessarily: one should better consider them as merely secondary tonsillar descents in specific clinical contexts that require only the treatment of the primary pathology and nothing more, just as is always done in the posterior fossa tumors, the deadliest cause of downward displacement of the cerebellar tonsils, where nobody disputes the foremost therapeutic objective. Nevertheless, if really and honestly open-minded, one has to acknowledge that perhaps every tonsillar descent is secondary to a pathological process, even though in most cases its nature is still unknown. But in the actual state of knowledge, we should better consider as "Chiari malformation type I" only the apparently *primary* and *congenital* cases of tonsillar displacement, just keeping in mind that both features can still be open to debate in any particular case.

Malformations of the occipito-cervical junction, representing a diverse and complex group of pathological conditions and related deformities, are often multiple in the same patient and many times occur in conjunction with Chiari malformation type I—with as many as 38–40% of hindbrain herniations in cases of atlas assimilation combined with Klippel-Feil syndrome [13]. In these patients, the abnormal fusions involving the occiput, atlas, and other cervical vertebrae would most probably be generated by defects in the functions of Hox and Pax-1 genes at different levels in the occipital and cervical somites [14] at a more delayed stage than those mentioned above. A possible explanation could be that the genetic and molecular alterations are more severe and thus extend their effects over segmentation and resegmentation of the somites and specification of the sclerotomes, not only affecting the hindbrain-spinal cord boundary as we have mentioned. A second possibility might be that the anomalous establishment of this boundary creates the conditions for a defective feedback from the neural counterpart to the mesoderm, disturbing these molecular pathways and secondarily the formation of cervical vertebrae. And we could add to these qualitative alterations the obvious quantitative one: if too much mesodermal tissue has been wrongly assigned to build the prospective spine, it goes without saying that the amount of tissue left for building the skull will be insufficient (**Figure 1**). The consequences of this relative lack of occipito-cervical mesodermal tissue will be distinct from those of the lack of spinal cord progenitor tissue, as the prospective growth of this segment of somitic mesoderm will be governed by the underlying hindbrain which, as far as we know, is very strictly divided in rhombomeres with distinct features, as opposed to the

monotony of the spinal cord organization in these early stages. Their feedback over their corresponding (and quantitatively defective) mesodermal counterpart will put quite stressful limits on the availability of compensatory mechanisms and thus determine an abnormal formation of the osseous and ligamentous elements of the occipito-cervical junction.

The same could happen also at more cranial levels, corresponding to the first occipital rhombomeres, where the same disproportion between the neural tissue contained within and the nearby mesoderm that receives its developmental induction would produce the deformities of basilar impression, platybasia, brainstem kinking, and retroflexed odontoid, found in 7.7% of our patients with Chiari malformation type I (Royo-Salvador et al., unpublished data). All these osseous anomalies could probably be explained by the interplay of discrete but persistent compressive and tensile forces developed among occipito-vertebral mesodermal segments during their development, secondarily to the mentioned genetic and molecular defects, recording somehow to the tenets of the Hueter-Volkmann law as applied to the spine [15].

At the same time, the disproportion between the contained, apparently hypertrophic hindbrain and the corresponding scarcely available mesodermal tissue will create the conditions for what Roth described in 1986 with such a brilliant intuition as "cranio-cervical growth collision" [16]: the impaction of the developing hindbrain against the growing vertebral column, which surpasses and deforms the insufficient occipito-cervical junction mesodermal primordium (the former from the inside, the latter from below (**Figure 1**)), accentuating the tonsillar descent, enlarging the occipital foramen, and leaving too little room for the formation of the occipital bone. It is amazing how the actual general opinion is able to conceive only this last developmental step [1, 17], but yes, finally, there is a para-axial mesodermal insufficiency associated with the Chiari malformations, but it is an associated phenomenon, somehow delayed and of secondary importance.

Among cranio-cervical junction malformations, a special mention deserves odontoid retroflexion, as it is a bony deformity that although it is less known and more imprecisely defined, it was found to be more marked and more common in children and adults with Chiari malformation type I than in normal controls [18, 19]. Moreover, in children with Chiari malformation type I, a study found it was correlated with the presence of syringomyelia and with a lower position of the *obex* [18]—that is, with a more intense cranio-caudal distortion of the brainstem. The pathogenesis of odontoid retroflexion seems more clearly related to an abnormal caudal traction exerted by the growing cervical spine than that of other occipito-cervical junction malformations, its mechanism of action being also more prolonged, as the dental central synchondrosis that connects the odontoid to the body of the axis can persist until the age of 8 years [13], being thus exposed to this axial strain, transmitted through the occipito-cervical dura mater and neighboring ligaments and membranes. But the most important detail that these studies on odontoid retroflexion provide is that they prove indirectly that the cerebellar tonsils were *pulled* and not *pushed* into the cervical funnel—in other words, that traction overrides compression at least in these cases—because if the opposite were true, the odontoid had been displaced anteriorly in patients with Chiari malformation type I, as a result of the "overcrowding" of the posterior fossa [18].

Since long ago, observations were published on the frequent association between Chiari malformation type I and idiopathic scoliosis [20], even though no coherent explanation of this fact has ever been provided. As a specific point, we have to insist that the presence of syringomyelia is not really necessary, as many have thought so far. Among our patients, Chiari malformation type I was associated with idiopathic scoliosis in 78.8% of cases, out of which only 52.1% also had idiopathic

syringomyelia (Royo-Salvador et al., unpublished data). Instead, a common pathogenesis, based on an abnormal caudal traction, seems more likely to be involved: in fact, as we mentioned before, the concept of "neuro-vertebral growth asynchrony" was coined in the realm of idiopathic scoliosis and constitutes the mainstay of the Roth-Porter pathogenetic theory [7–9], which uses various mechanical experimental models to demonstrate that an uncoupling of the growth velocity between the spine and spinal cord makes the latter to lag behind, putting tension on the posterior elements which will grow at a slower pace (here we come once again in close contact to the Hueter-Volkmann law), so that the anterior elements will grow too much and the vertebral bodies, "tethered" posteriorly, will start to rotate around an axis represented by the spinal cord itself and will deviate to one side as they grow restrained in this way, thus creating the scoliotic curve [21]. It is not difficult to imagine how a similar mechanism of caudal traction would produce both a Chiari malformation type I and an idiopathic scoliosis if this intrinsic "tether" acted continuously over the vertebral column and spinal cord throughout their development and associated longitudinal growth, a fact especially conceivable if, following the mentioned alterations in the definition of the hindbrain-spinal cord boundary, there is a relative excess of mesenchymal tissue composing the sclerotomes of the future thoracic spine, even though it would be much later that this unbalanced tissue distribution would become manifested, during the growth spurt of the adolescence.

Last but not the least, among enlightening pathological associations of Chiari malformation type I is the tethered cord syndrome, maybe the most interesting of all, the most difficult to explain, and nevertheless, the most important, as it forms a bridge between Chiari malformation types I and II. In fact, this association should be better regarded as a separate third category of Chiari malformations, taking into account the different mechanism of relative spinal cord "shortening": if in Chiari malformation type I this originated in a caudal displacement of the hindbrain-spinal cord boundary and in Chiari malformation type II, in the traction exerted by a caudal myelomeningocele on the growing spinal cord, here there is an abnormal *filum terminale*, short, thickened, and/or lipomatous that hampers the spinal cord longitudinal growth and that alters the coupling between vertebral and neural growth. Among our patients, the level of the *conus medullaris* was below the L1 L2 disk in as many as 20.9%, and most interestingly, it was statistically correlated with the degree of tonsillar descent (Royo-Salvador et al., unpublished data).

Now of course, if one accepts that the pathogenesis of Chiari malformations includes a common pathway of relative shortening of the spinal cord with respect to the vertebral column, of various etiologies that can be grouped into these three large groups, an important question comes about: why not any patient with this relative spinal cord shortening has a tonsillar descent? Well, the answer is quite simple, because, as we have already pointed out, there is another decisive factor that will eventually determine the occurrence or not of a Chiari malformation: the adequacy of the neural tissue reaction to the tensile forces developed as a consequence of the growth asynchrony. In other words, the tonsils will descend only if this homeostatic mechanism doesn't function properly for one reason or another; moreover, any degree of tonsillar descent and of brainstem and fourth ventricle distortion should be possible in every one of the three main etiopathogenetic groups mentioned, so it should be time that we stop associating Chiari malformation type II only to myelomeningocele and instead, consider, for example, three degrees of tonsillar descent, perhaps labeled as Chiari malformation types 1, 1.5, and 2 (even four if a Chiari malformation type 0 were added) and defined with clear-cut morphological criteria, including measures of brainstem elongation and fourth ventricle distortion [22].

4. Clinical arguments

Magnetic resonance imaging, if scrutinized really carefully, can provide much more information than just detect Chiari malformation type I. Early on, we mentioned the special meaning that a retroflexed odontoid can get as a proof of caudal traction applied on the occipito-cervical junction (**Figure 2**).

In many Chiari malformation type I patients, we can ascertain a descent not only of the cerebellar tonsils but seemingly of the whole cerebellum, as there is a readily identifiable difference of width of subarachnoid spaces above and behind the cerebellum, a feature that others have labeled "obliteration of retrocerebellar cerebrospinal fluid spaces" [17] following a different interpretation; of course, if a diminished posterior fossa volume were the cause of the tonsillar descent, there would be no free subarachnoid space visible underneath the tentorium as we see in many patients (**Figure 3**).

But maybe the most spectacular image testimony of the mechanisms mentioned above is the feature that we called "tense spinal cord," which has also been described in relation to idiopathic scoliosis [9] but that we could identify in many patients with Chiari malformation type I with or without scoliosis: in sagittal cuts, the spinal cord does not follow closely to the curves of the spinal canal, but instead, it takes the shortest route within the canal and is thus more or less straightened, in some cases even stuck on the concave side of the lordotic or kyphotic curve of the spinal canal (**Figure 4a**), corresponding in axial cuts to an eccentric position of the spinal cord in the canal, closer to the concave side (**Figure 4b**).

We interpret in a similar way another associated image feature, denominated "lateralized spinal cord," visible in coronal or axial cuts (**Figure 5**) and that can be understood as a marker of tension through the spinal cord if one keeps in mind Porter's experimental model [9], this time conditioned by the presence of at least a minimal degree of scoliosis. All this is even easier to figure out by neurosurgeons, because here the spine recalls the principle of functioning of the Leyla retractor system introduced by Gazi Yasargil and so often used to hold brain spatulas. In other words, a central cable in a hollow curved construct will deviate towards the concavity if subjected to axial tension, and in the vertebral column, this can happen either in the sagittal plane, in the coronal plane, or in both.
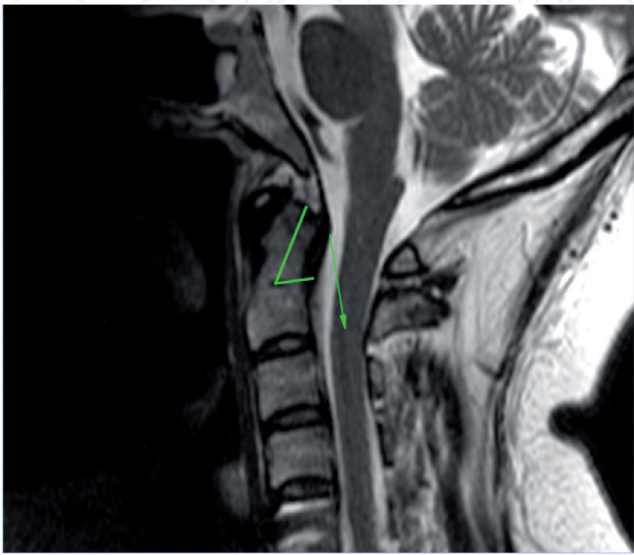

Figure 2.
The causative vector of a retroflexed odontoid is likely parallel to the caudal traction (arrow).
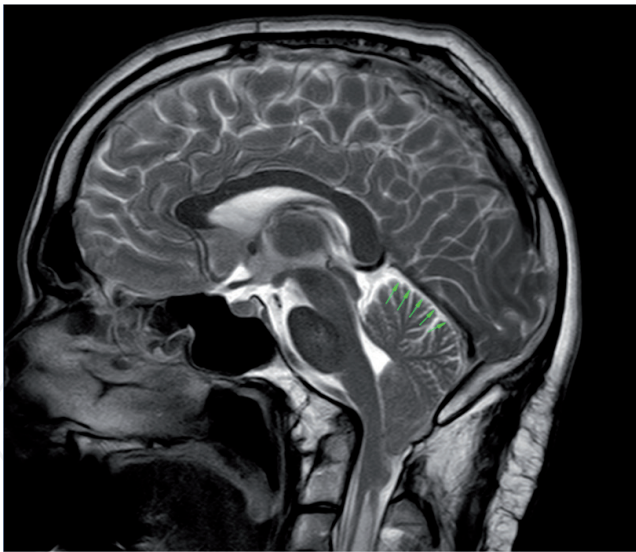
Figure 3.
Increased subarachnoid spaces between the tentorium and the cerebellum reflect the global displacement of the latter towards the foramen magnum.
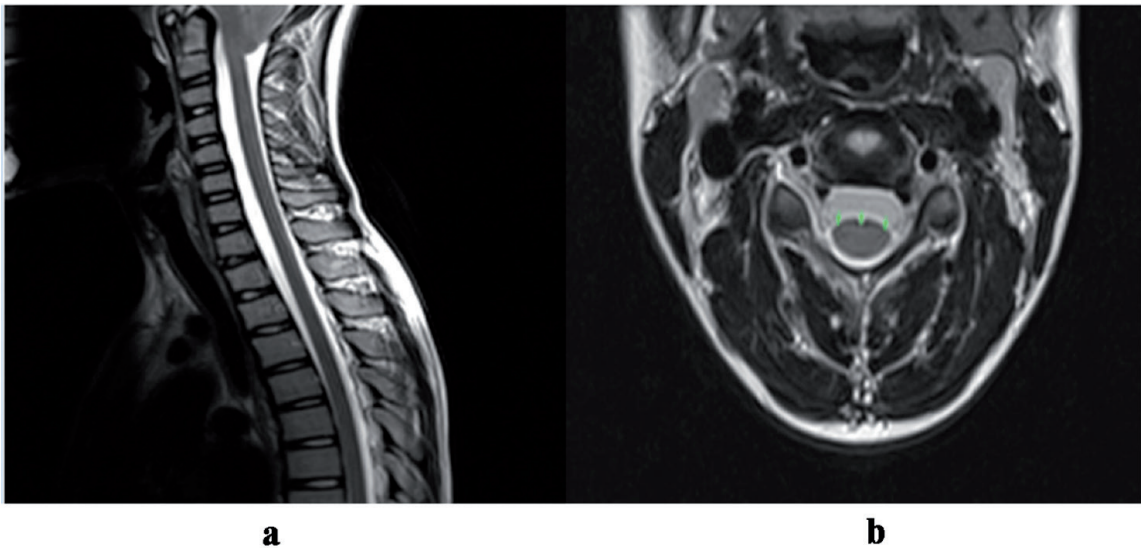

a   **b**

Figure 4.
Tense spinal cord in a sagittal cut (a) and an axial one (b). The spinal cord travels closer to the concavity of the curves and occupies an eccentric downward position in axial images (arrows).

As an expected consequence of an incomplete understanding of the etiopathogenesis of Chiari malformation type I, its surgical treatment seems the unhappy heir of a mysterious real estate, haunted by dreadful ghosts such as sleep apnea and sudden death. If in some cases it is indeed elementary caution and justified to do no treatment at all, as the tonsillar descent is merely an asymptomatic deformity discovered incidentally, in many other instances, the patients are left to struggle with their own despair as the obvious symptoms and signs they present are not recognized as such by the neurosurgeons in charge. And the reverse is also true: when an active treatment is chosen, it consists usually of suboccipital craniectomy, C1 laminectomy, and duraplasty, which is equivalent to performing an en bloc resection with healthy borders followed by radiotherapy and chemotherapy for a tumor of unknown behavior (not to mention the tonsillar resection added at times). Well, some minimalizing technical advances have been proposed, like leaving the dura mater or the atlas intact, but their problem rests in not getting to the heart of the matter—so, they might lack the desired efficacy. Recent efforts
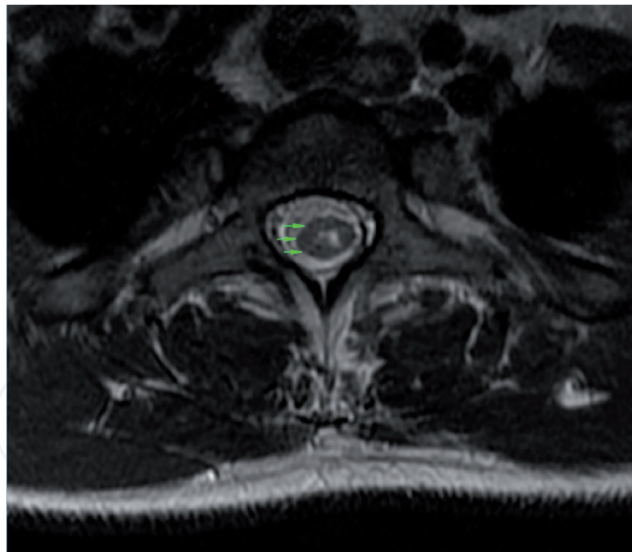
Figure 5.
Lateralized spinal cord deviated towards the left inside the spinal canal.

complicating more this subject have tried to define instances of cranio-cervical or atlantoaxial instability that supposedly would require complicated and risky procedures applied without firstly securing more confidently a diagnosis of genuine instability—one that is perfectly plausible in selected cases of traumatic spine injury.

Many of the delusions and mishaps issued from the actual therapeutic strategy applied to Chiari malformation type I could be avoided if, taking into account patiently all the facts presented above, one should switch his or her vision from the actual obsession to perform a *circumferential* decompression of the tonsils squeezed against the elongated brainstem to an objective of a rather *longitudinal* or *axial* release of the deformity that affects not only the brainstem-spinal cord junction but the whole of the brainstem *and* the spinal cord starting at the level of the *dorsum sellae*—upper end of the notochordal-influenced growth and somitic division of the mesoderm—until the very tailbone that at earlier stages was the advancing front of axial somatic growth and a possible regulator and intermediate of the coupling between the vertebral and spinal cord growth.

The most logical initial step for interfering with this pathogenesis, considering caudal traction as a final common pathway of multiple etiologies, would be to interrupt this unique route of producing damage to the brain, spinal cord, and spine itself. Technically, this is straightforward if done at the caudal end of the tense spinal cord instead of a frontal attack upon the delicate, impacted cranio-cervical junction. This should consist of a *filum terminale* release by means of the best available technique. A bonus of this approach is that it eliminates the concerns of a possible worsening of a hidden cranio-cervical instability. Its guarantee of success in releasing the conflict between the tonsils, brainstem-spinal cord junction, and occipital foramen stands in the continuous process of spinal growth which produced the progressive lengthening of the cord throughout the intrauterine life and childhood, by adding up collagen and elastin fibers to the complex tridimensional network that conforms the *pia mater* and holds the spinal cord connected mechanically with the vertebral column during its growth and movements. During adulthood, as we have already mentioned, although growth eventually stops, the axial tension is maintained by ongoing processes of degeneration and atrophy which paradoxically, instead of reverting it, will convert the mentioned *neuro-vertebral asynchrony* into a lifelong feature of the human body.
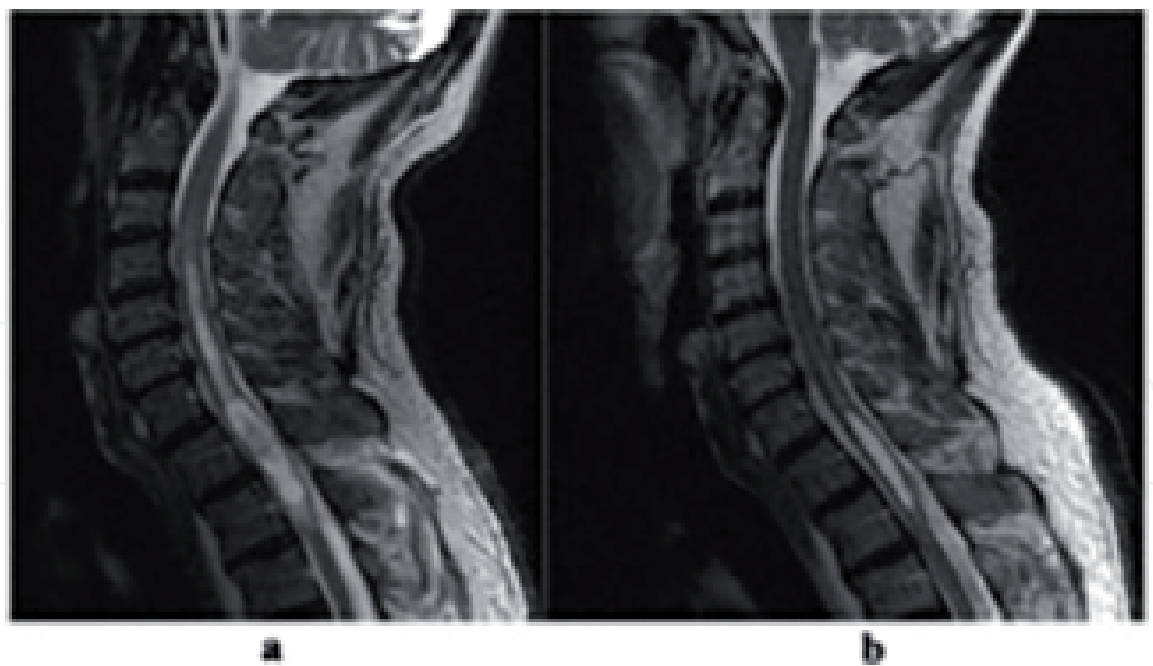
Figure 6.
Preoperative (a) and postoperative (b, at 30 months) magnetic resonance images of a 56-year-old male patient with Chiari malformation type 0, operated of filum terminale sectioning; the improvement of his idiopathic syringomyelia, visible also in a previous control (not shown), is now quite obvious, pointing to caudal traction as a possible mechanism.
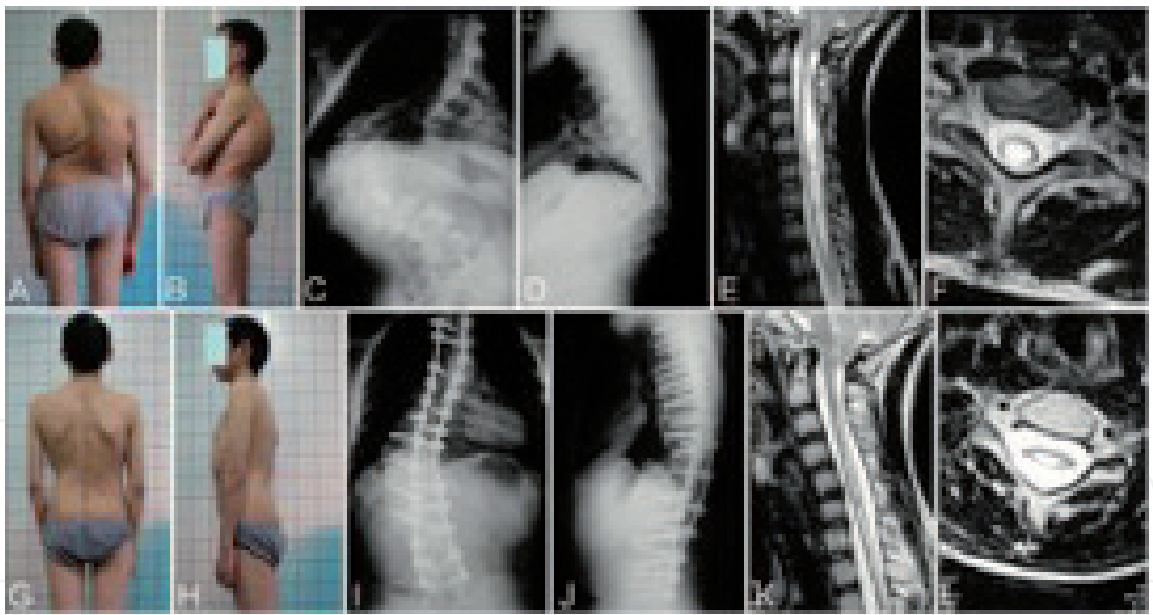

Figure 7.
Preoperative (A–F) and postoperative (G–L, at 3 months) photographs, radiographs, and magnetic resonance images of a 17-year-old male patient with Chiari malformation type I, idiopathic syringomyelia, and severe scoliosis, operated of spinal resection and instrumentation, with marked improvement of his syringomyelia (with permission from Wang et al. [25]).

Of course, the actual surgical approach of suboccipital craniectomy *does* lead to the completion of a similar release of longitudinal spinal tension but at a much higher cost and with much more risk of potential complications; moreover, it might be less efficient, because in the cervical spine, the stronger and more numerous dentate ligaments limit more the stress release than in the lumbar region.

Interestingly, against all odds, some subtle developments occurred in recent years in the surgical treatment of Chiari malformation type I, proving that more
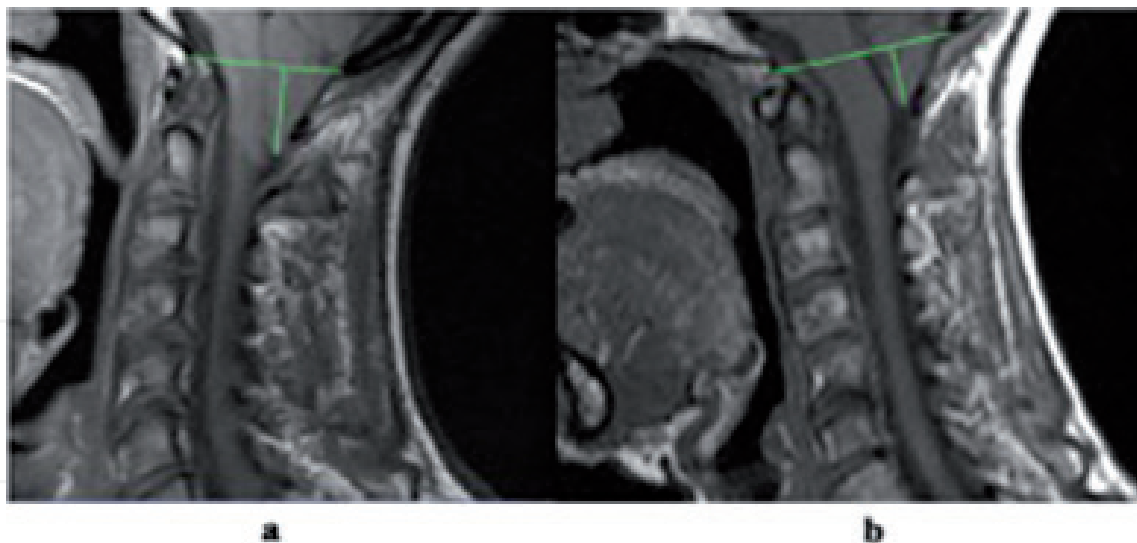
Figure 8.
Preoperative (a) and postoperative (b, at 7 years) magnetic resonance images of a 49-year-old female patient with Chiari malformation type I, operated of filum terminale *sectioning, with impressive improvement of the tonsillar descent from 17 to 13 mm with respect to McRae's line (measured in the cuts with maximal descent).*

and more clinicians are starting to accept that maybe suboccipital craniectomy is not the only surgical solution to this condition. For this reason, we can present here three "surgical" testimonies in favor of the theory of caudal traction, as follows:

1. In fact, it is suboccipital craniectomy itself that opened these new perspectives when in the hands of some fearless teams [23, 24]; it started to be used for treating patients with syringomyelia without tonsillar descent, with encouraging results, but they did not realize their meaning not even when they discovered that in these children, there were image features suggesting a caudal elongation of the brainstem with displacement of the obex and increased diameter of the *foramen magnum*, as occurs also in Chiari malformation type II [24]. In **Figure 6** we show a similar example from our series of patients, where sectioning the *filum terminale* determined a marked improvement of a cervicothoracic syringomyelia in a case of Chiari malformation type 0.

2. Another ingenious team discovered that if they performed idiopathic scoliosis correction by a technique of posterior vertebral column resection with spine shortening and instrumentation after applying compressive forces, the cerebrospinal fluid flow at the level of the *foramen magnum* improved in patients with Chiari malformation type I and syringomyelia, and in many of them, even the latter diminished in size [25]. Obviously, scoliosis surgery by no means could have accomplished any *circumferential decompression* of the occipital foramen but a release of the tension in the brainstem and tonsils (or, as we stated above, a welcome *longitudinal decompression*) (**Figure 7**).

3. Yet the most important testimony came from the hands of a group which operated 318 patients presenting both tethered cord syndromes defined according to very exigent criteria and Chiari malformation type I or low-lying cerebellar tonsils of 0–4 mm descent (that we consider as being also Chiari malformation type I, together with more and more authors [1]), but the technique used was not suboccipital craniectomy, but sectioning of the *filum terminale* by means of an L4 laminectomy. Their results were excellent, both concerning the clinical picture and various morphometric criteria of the posterior fossa contents,

demonstrating that the hindbrains of these patients were abnormally descended preoperatively and improved their position after the indirect surgery and applied to the other end of the spinal cord [22]. In **Figure 8** we present pre- and postoperative images of one of our cases, with a significant ascent of the tonsils after the filum terminale release.

5. Final remarks and future directions

In the actual state of knowledge, it is imperative to recognize that the development of the hindbrain and the spinal cord is a complex process regulated by genetic, molecular, mechanical, endocrine, and nervous homeostatic mechanisms that compensate one for another—within certain limits—in case of imbalances and disturbances. Nevertheless, it is exactly this complexity, coupled with the elevated functional requirements that the cranio-cervical junction has to meet, that makes their union so sensitive to various pathogenetic factors and determines malformations among which the one known as Chiari malformation type I is the most common. According to all the arguments presented in this chapter, the final common pathway of these etiopathogenetic aggressions seems to be caudal traction, a complex biological phenomenon that by no means should be reduced to a simple mechanical force of axial pull. There is still much left to discover about the physiologic mechanisms that govern the coupling between the growth of the vertebral column and that of the spinal cord during somatic development, where maybe future research will define the roles played by the pineal gland, the subcommissural organ, and the *filum terminale*, just to cite a few of the possible actors eligible for this casting.

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