The MRI in Arnold-Chiari Syndrome I and Idiopathic Syringomyelia

Author: Miguel B. Royo Salvador, MD, PhD

Introduction

Magnetic resonance imaging (MRI) is a crucial diagnostic tool for men, women, and children with a diagnosis of Syringomyelia, Syringobulbia, Chiari Malformation, Scoliosis, and additional related pathology. Radiology is a necessary part of the diagnostic considerations for the ongoing care of the patient.

There are key areas of reporting pathology that need improvement so that radiologists can positively contribute in the process of proper early identification of the onset of disease and accompanying pathology. This is crucial to prevent progression and irreversible damage. One current problem is that pathology of these diseases are frequently dismissed on MRI or termed "of no clinical significance" when it is clinically significant! This can lead to years of misdiagnoses, complications, and incorrect treatment leading to progression of the disease and irreversible damage. Ethically we have a responsibility to change this disturbing trend through better practices, updated education, and increased awareness among all providers across multiple disciplines.

Some ways that radiologists can positively support the Syringomyelia and Chiari community are by identifying all of the pathology which will aid in proper diagnosis. Measurement of the size of the cysts also termed syrinxes as well as tonsillar herniations is helpful for the neurosurgeon. All pathology must be reported to the ordering physician who will then consult with neurosurgery as well as other appropriate specialists to determine the treatment course based on the patient's history, presenting symptoms, and physical assessment. Syrinxes large and small as well as tonsillar herniations large and small should be measured, documented, and reported so that this vital information can be included with a thorough patient history and assessment to determine the appropriate treatment and specialists needed in the ongoing care of the patient.

Dr. Miguel B. Royo Salvador MD, PhD currently serves on the medical advisory board of Worldwide Syringomyelia & Chiari Task Force. He is Director of the Institut Chiari & Siringomielia & Escoliosis de Barcelona. His use of MRI to manage patients with Arnold-Chiari Syndrome I is provided below. All figures were provided by the author.

"Please Note: The Filum System ® is used by Dr. Miguel B. Royo Salvador in Europe but is not used in the United States. If clinicians are interested in learning more about the Filum System ®, please visit https://institutchiaribcn.com/en/presentation-filum-academy-barcelona/ "

Quantifying the descent of the cerebellar tonsils is not indispensable for the diagnosis of Arnold-

Chiari Syndrome

In medical bibliography, there is a certain controversy regarding the Arnold-Chiari I Syndrome* (ACHS.I) as to determining an abnormal position of the cerebellar tonsils (CT) and the possible clinical correlation.

Diagnosis of the Arnold-Chiari Syndrome I and the position of the cerebellar tonsils.

There are different publications by Nuclear Magnetic Resonance (MRI) specialists that link the CT position to the diagnosis of the Arnold-Chiari Syndrome I. For some, the distance from the end of the cerebellar tonsil to the foramen magnum (FM) is of 5 mm, for others when it is of 3 mm. In both cases without insisting on clinical correlation, the diagnosis is essentially drawn from the imaging (1).

In the early Nineties, when I was working on a protocol for the measurement of the descent of the cerebellar tonsils (DCT) for my doctoral thesis (2), I found that measuring this descent on the MR image from the tip of the descended CT to the FM had an important inconvenience: according to the size of the radiological image, the DCT could vary and it was difficult to scale it properly. DCT measurement in this manner was prone to error.

I therefore adopted a measurement scale for DCT in relation to the neighbouring anatomical vertebral structures, possible in most cases. I considered the position of the CT at different levels, aligned with: the foramen magnum, the first/second/third third of the distance FM-C1, the upper edge of the posterior arch of the first cervical vertebra (C1), the body of the posterior arch of C1, the lower edge of C1, the upper edge of the posterior arch of C2, the body of the posterior arch of C2, the lower edge of C2…

If there are no existing posterior vertebral references, which is quite infrequent, the measurement could equally be taken with anterior vertebral references.
One of the conclusions of the thesis was that the caudal traction force that causes the DCT in ACHS.I can be conditioned by different factors that modulate the intensity of the descent of the cerebellar tonsils, such as the size of the foramen magnum, the existence of other cranio-vertebral deformations, bioelasticity, age, …(figure 1).

Thus, a smaller DCT can correspond to a greater force, if the foramen magnum, or other causes, make the movement of the cerebellar tonsils difficult. What matters is therefore not quantifying the descent, but determining whether there is a caudal force that is traumatic to the whole nervous and cranio-vertebral system.

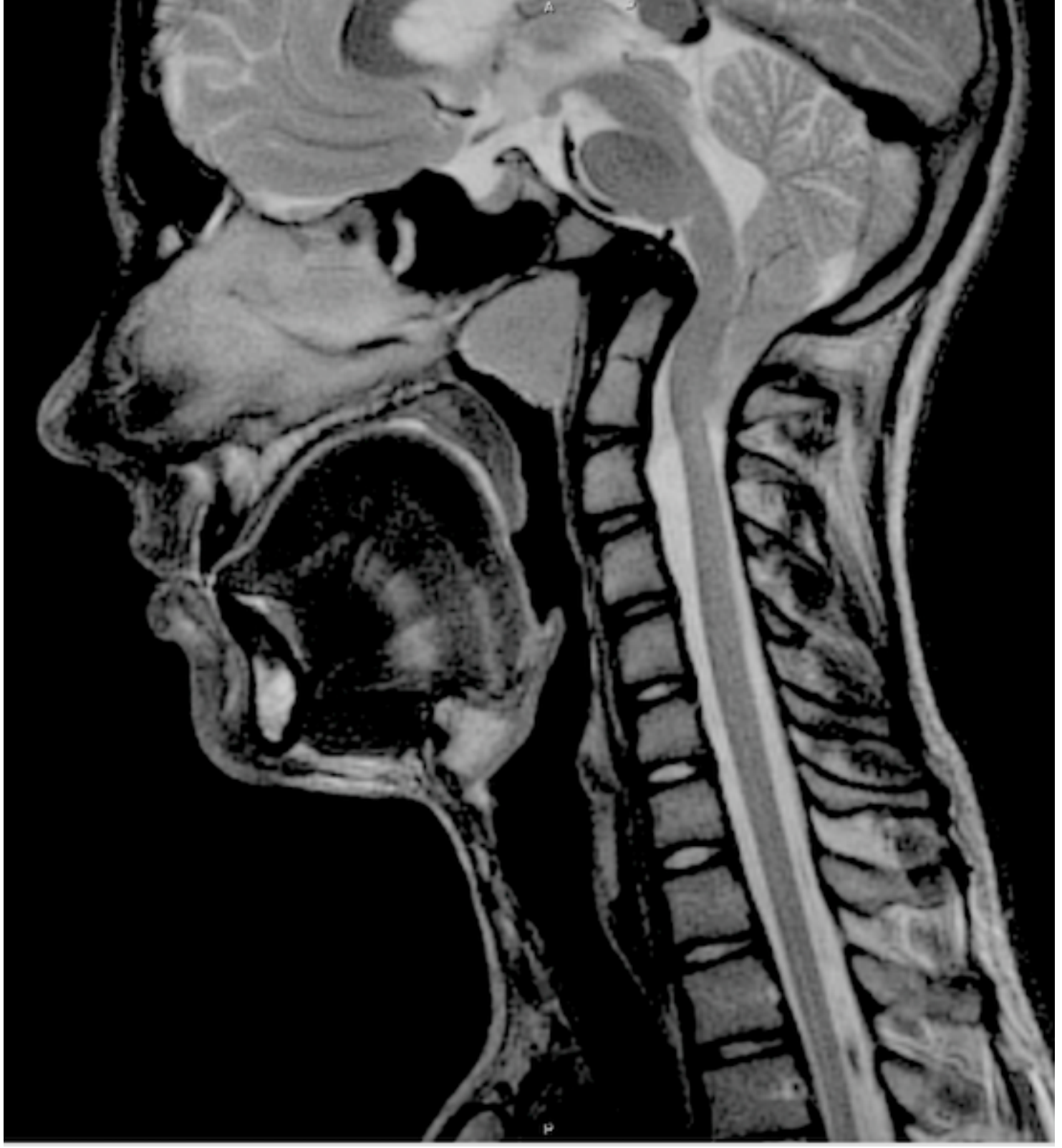


Figure 1.- Case 18044HC. 18 yr. old male from Germany with DCT, basilar invagination, platybasia, retroflexed odontoid, cervicothoracic I.SM, atlas assimilation. A significant share of cranio-vertebral malformations are generated by the caudal traction, and the malformations in turn hinder the descent of the cerebellar tonsils. We can also observe the increase in the supracerebellar space and the tension of the cervical spinal cord with a straightened cervical lordosis.

Not long ago, the Arnold-Chiari Syndrome type 0 was described: a patient with a symptomatology compatible with the Arnold-Chiari Syndrome but the position of the cerebellar tonsils is at 0 mm or close to the foramen magnum without surpassing it. With this, imaging as exclusive means for the diagnosis of the Arnold-Chiari Syndrome is no longer applicable.

We described the clinical Filum Disease in our method Filum System® (3): it is similar to ACHS.0 and it presents a compatible clinical picture without the conventional imaging expressions (MRI, x-ray).

The descent of the cerebellum in the ACHS.I

In our new description of the Filum Disease (FD), which includes Arnold-Chiari Syndrome I, idiopathic Syringomyelia and idiopathic Scoliosis amongst others, we observe that in cases where the cerebellar tonsils were close to or surpassed the foramen magnum, there was an increase in the supracerebellar space. Both observations indicate the existence of a descent of the entire cerebellum within the posterior fossa (figure 2).

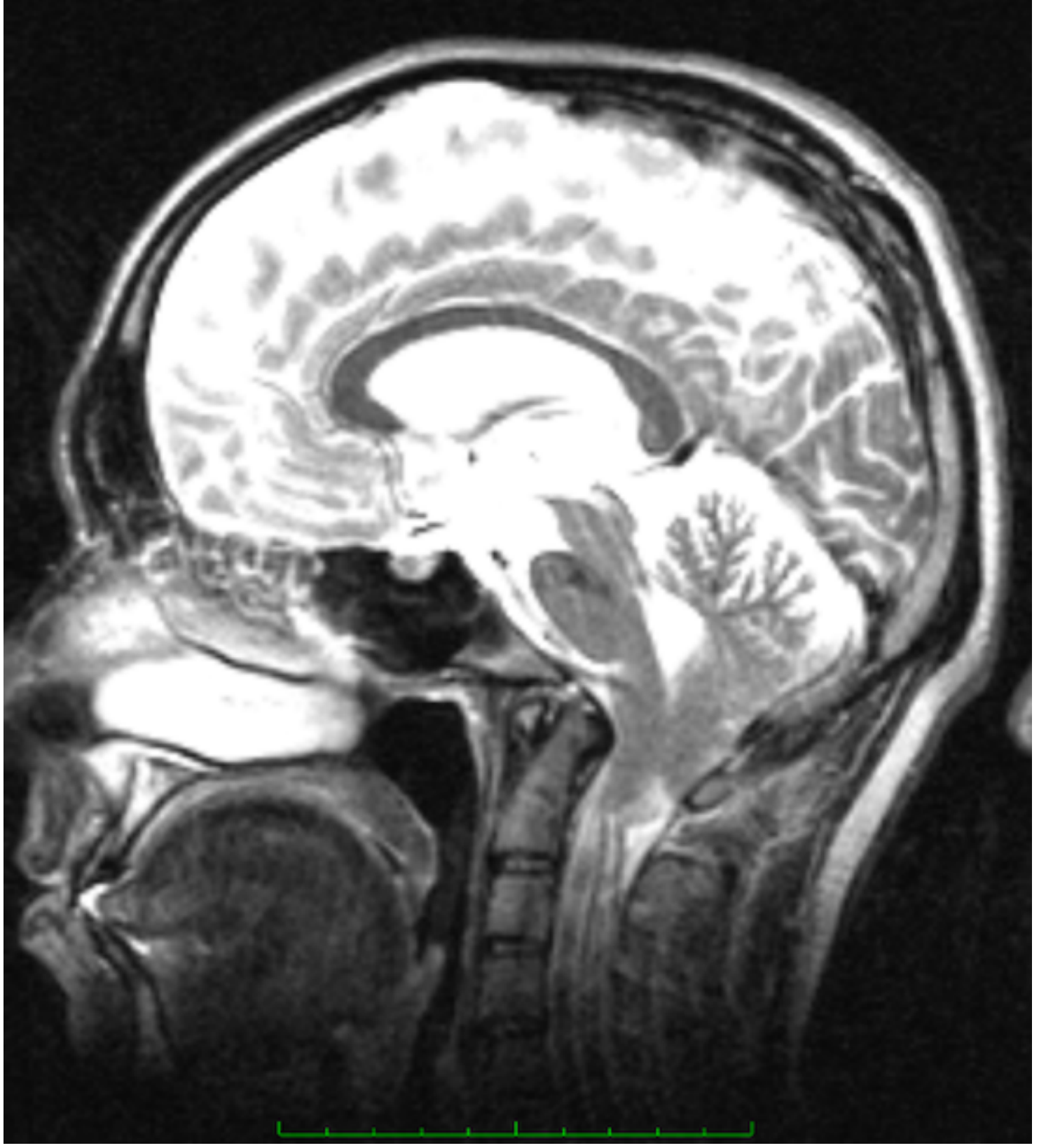


Figure 2.- Case 11705HC, 26 yr. old male with cervical and thoracic Syringomyelia, levoconvex thoracic kyphosis and scoliosis and a marked increase of the supracerebellar space.

In other words, we consider that not only the cerebellar tonsils descend, but also the cerebellum as a whole does so, therefrom the high frequency with which an increased cerebellar space can be observed alongside a descent of the cerebellar tonsils. This argument is of great significance as it contradicts the current stenosis of the posterior fossa mechanism, given that in the observation of thousands of typical ACHS.I cases, not one case has been found with a superior cerebellar herniation (transtentorial, rostral, pontine or mesencephalic), or simply a collapse of the supracerebellar space. This should occur with an equal frequency as the DCT, if the cerebellum is subjected to a space restriction by a posterior fossa stenosis. Hence, the suboccipital craniectomy, which is usually applied in ACHS.I, is unnecessary, as the procedure has the aim to increase the posterior fossa volume.

We therefore perceive the existence of an ACHS.I when there are signs that the cerebellum is displaced caudally towards the foramen magnum and that the important issue with the Arnold-Chiari Syndrome I is to identify the existence of a caudal force that causes functional and anatomical injuries to the entire Central Nervous System. This is of key importance to help our patients: as soon as the caudal force is found, it is convenient to eliminate it, due to present and progressive damage it can cause in the future.

In the radiology reports concerning MRIs with signs of a caudal displacement of nervous structures, using terms such as “not clinically relevant” should be avoided. Given that there is a significant proportion of cases where inspite of the displaced cerebellum, the displaced cerebellar tonsils or changes in the supra- or infracerebellar fossas being visible, the patient and/or the physician are not able to find the existing symptoms or signs that are correlated to the imaging findings.

Sometimes this can be due to a lack of appreciation by the patient, other times to inadequate patient histories and physical examinations, or insufficient clinical histories and physical examinations, or, as is frequently the case, because the MRI specialist does not have the clinical data or due to the unawareness of the physician, or Medicine in general, of the compatible clinical pictures as occurs with the Filum Disease, which is unknown to the majority of health care professionals.

Idiopathic Syringomyelia can be detected on MR imaging before the intramedullary cyst is formed.

Another controversy is what happens with the Magnetic Resonance Images of the idiopathic syringomyelia cavity and the neuroimaging specialists’ tendency to consider it as a benign dilation of the ependimary central canal of the spinal cord.

Pathological MR imaging in idiopathic syringomyelia that is being ignored or considered normal.

I described the different phases of the evolution of the syringomyelia cavity in my publications (4), from its beginnings with the ischemic phase going through the fusiform, dilated, fistulised, collapsed and atrophic phases.

Each of the syringomyelic cavity’s phases expresses itself in a specific way on the magnetic resonance as to their description and pathological meaning. Nevertheless, there are images that some MR specialists do not recognize or interpret with an ambiguous meaning, for example:

- The phase of central canal ischemia and edema in the Filum Disease indicates the start of the formation of a syringomyelic cavity, hydromyelia or ependimary dilation and results in an image of two paracentral rails (figure 3) or one centromedullary hypointense rail in the sagittal planes of the spinal cord. The latter image is described in Magnetic Resonance Imaging atlases as a sign of spinal cord edema and tends not to be described in MRI reports (figure 4).

Fig. 5.97a, b. Proton-density- and T2-weighted sagittal images. Multisegmental manifestations of disseminated encephalomyelitis in the thoracic cord and prominent central canal

Figure 3.- Figure 5.97a and b, and in detail, taken from ***"MRI Atlas Orthopedics and Neurosurgery The Spine. Martin Weyreuther and cols. Springer 2006.".***

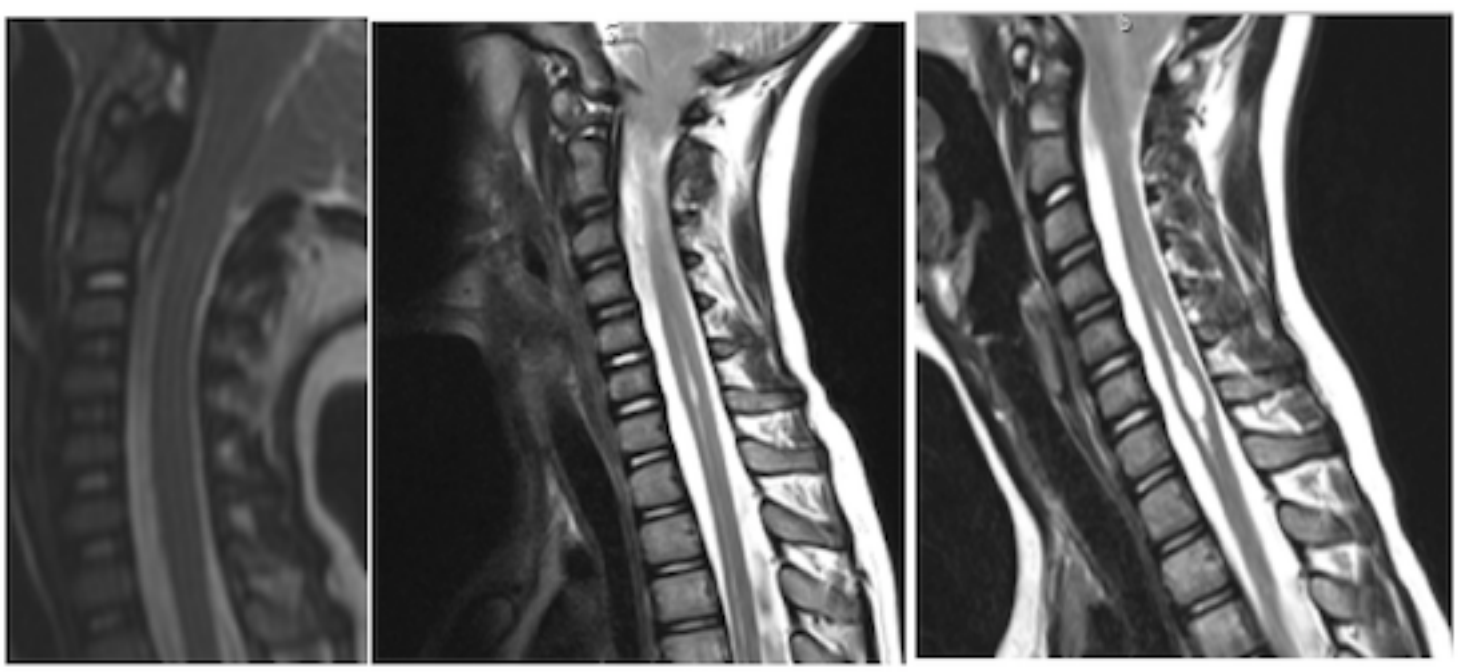


Figure 4 .- Case 1404P, patient affected by cervical and thoracic. Here the evolution of the centromedullary injury can be observed from its beginning to the formation of the syringomyelic cavity. On the MR image of the cranio-cervical region on the left hand side, taken in 2010, patient two years old, two parallel lines that follow the centre of the spinal cord can be seen, medullary ischemia-edema phase. On the image in the centre, at the age of five, we see the beginning syringomyelic cavity where the tissue necrosis due to ischemia appears (compatible with the conventional concept of the benign dilation of the ependimary canal). On the right hand side, now six years old, with the syringomyelia cavity at tension. We can also observe the progression of the descent of the cerebellar tonsils and of the tension of the cervical spinal cord.

- Many consider a dilation of the ependimary conduct to be benign or a variant of normality. According to the hydrodynamic theory, the dilation would be an expression of hyperpressure of the CSF when passing through the obex, located in the lower part of the fourth ventricle, towards the ependimary canal. According to the traction theory, the basis for the Filum Disease, it is most likely that any idiopathic image of ependimary canal dilation is pathological and due to peri-ependimary tissue necrosis in view of the ischemia that is produced by the spinal cord

traction transmitted by the filum terminale (figure 5).

Figura 5 .- Spontaneous evolution of idiopathic SM without treatment , case 10005HC. Left: MRI from 2010 with a presumed dilation of the ependimary conduct catalogued as "benign". Right: the same injury that has progressed to a syringomyelic cavity on a MR image from 2014.

- In cases of Arnold-Chiari Syndrome I and idiopathic Syringomyelia we can observe on the axial planes that the spinal cord is displaced to either of the sides of the vertebral canal. This is an obvious sign for the existence of a caudal force in the curvilinear vertebral canal, as is common in scoliosis. On the coronal plane, we can also see that the spinal cord goes from one internal convexity to another of the vertebral canal, placing itself as to follow the shortest path. This very important and pathognomonic sign for idiopathic scoliosis is currently not being observed or described in MRI reports (figure 6).

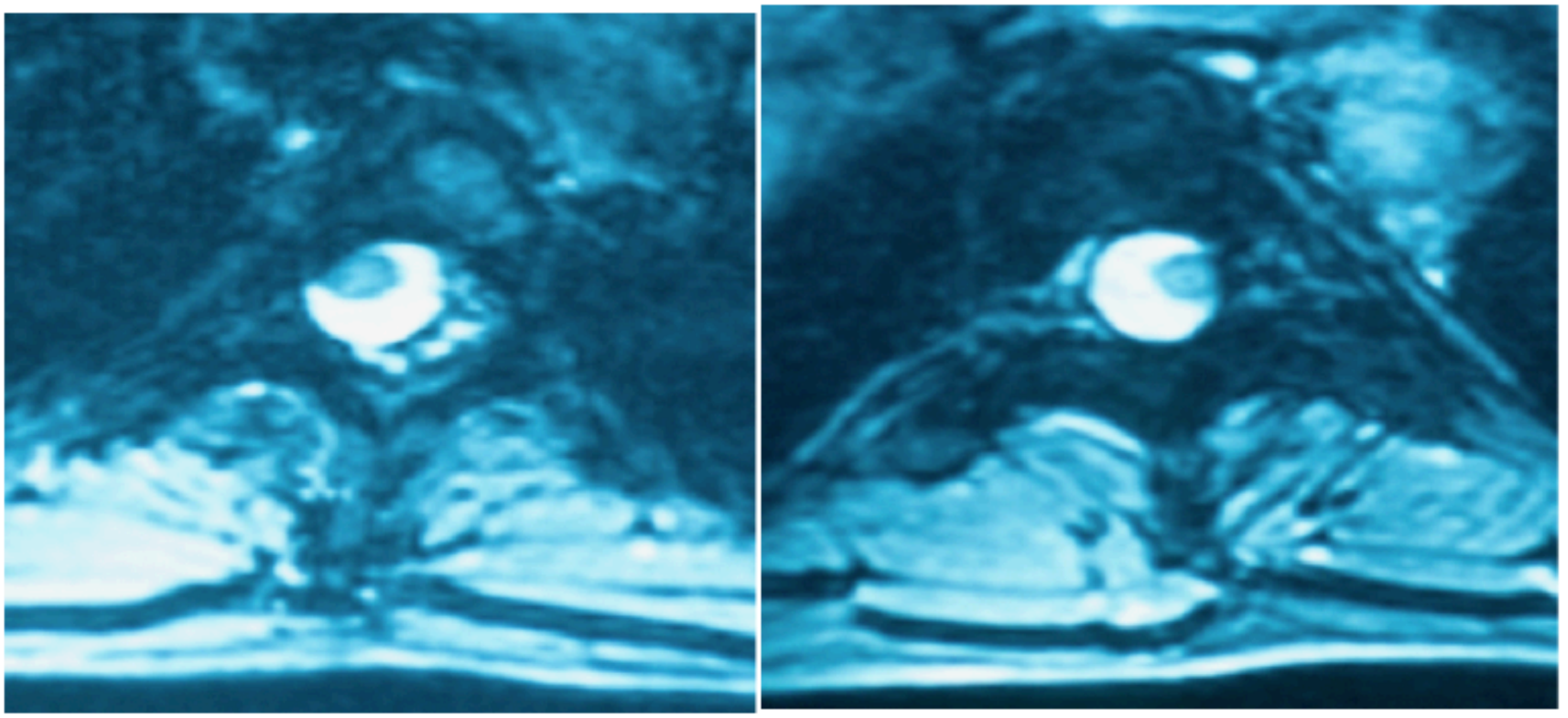
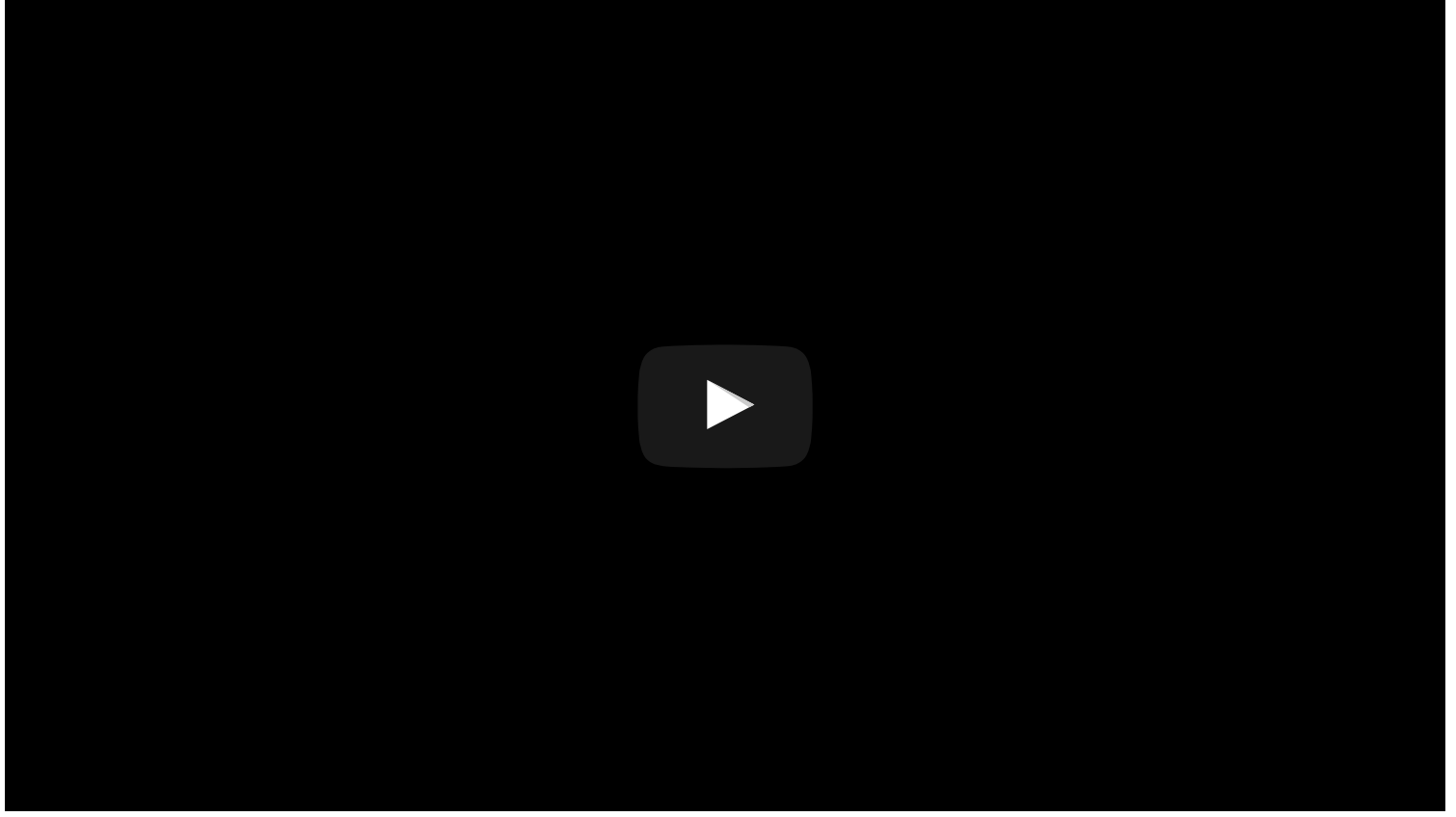


Figure 6.- Thoracic scoliosis with axial images on the high and low thoracic MRI of the same patient. Left: upper thoracic vertebra where the spinal cord leans into the right-concave side of the vertebral canal. Right: lower thoracic vertebra, the spinal cord leans towards levoconcave side of the spinal canal. This indicates the existence of a lateral force on both images that displaces and squashes the spinal cord towards the lateral wall of the spinal canal and eliminates the force of the denticulate ligaments that maintains the spinal cord in the centre of the vertebral canal when there is not a cranio-caudal or axial force.

How to diagnose the Arnold-Chiari Syndrome I, idiopathic Syringomyelia and idiopathic Scoliosis or the disease that includes all of them: the Filum Disease?

In order to diagnose these conditions in many cases it is necessary and indispensable to have the findings from the clinical history and physical examination available, given that the majority of Filum Disease cases show minimal expression or no expression on the imaging (MRI and x-ray). Thus imaging experts should limit themselves to the description of anomalies with possible diagnostic orientations and refrain from clinical commentary; these should be made by the specialist who gathers all the information, clinical as well regarding to imaging.



“Diagnosis of the NCV.S and the FD”, the first of the twelve Filum System® protocols, includes thirty-two clinical history parameters and significant imaging signs for the diagnosis of the Filum Disease. They facilitate an automatized diagnosis in the form of two tables: one concerning the Neuro-Cranio-Vertebral Stress (75 signs) and the other concerning clinical measurements (57 clinical and physical examination parameters) with regards to the Filum Disease. With the sum of the scores of both tables we obtain a value indicating with high probability the existence or non-existence of the Filum Disease.

When is a Filum Disease diagnosis convenient?

The Filum Disease causes two types of injuries: reversible and irreversible ones. When a diagnosis for Arnold-Chiari Syndrome I, idiopathic Syringomyelia and idiopathic Scoliosis is established, in the majority of cases many of the injuries are irreversible.

A diagnosis of these conditions in reversible phase is usually detected in the phase when the cerebellar tonsils are impacted (Arnold-Chiari Syndrome I); in the phase of ischemia-edema in idiopathic Syringomyelia and in idiopathic Scoliosis when the curvatures add up to less than 15-20º.

Applying the Filum System® health method in the reversible phase of the conditions allows to restore the patient near to normality in the majority of cases.

How can we reach an early diagnosis of the Filum Disease?

First, health care professionals need to be aware of the existence and know the details of the Filum Disease, given that currently this is not the case. Once the existence of the condition has been accepted, diagnostic out-patient protocols can be established with the help of the Filum System®.

Secondly, the public needs to gain knowledge of the Filum Disease to notice the existence of certain symptoms and to then approach professionals that have training concerning the condition.

When should the treatment for the Filum Disease be applied?

There is an indication for the application of the Filum System® treatment method in the majority of cases as soon as the

Filum Disease diagnosis has been established.

How many patients with the Filum Disease are there?

The sum of the incidences of the diseases that find primary and secondary expression in the Filum Disease can be of more the 20% of the world population, which would make the Filum Disease the most frequent and unknown disease of mankind.

Author: Miguel B. Royo Salvador, MD, PhD

* Author's note: the current International Statistical Classification of Diseases and Related Health Problems (10th Revision, 2016) of the World Health Organization lists the Arnold Chiari Syndrome (Q07.0), which some authors refer to as the Chiari malformation, Chiari or the Chiari syndrome. Leaving out the "Arnold" could give way to possible misunderstandings that the nomenclature of the WHO intents to avoid, as there are several other entities with similar names, the Chiari-Fromel syndrome, the Budd-Chiari Syndrome, Chiari Osteotomy.

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