

The Filum Disease and the Neuro-Cranio-vertebral Syndrome: definition, clinical picture and imaging features

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INTRODUCTION

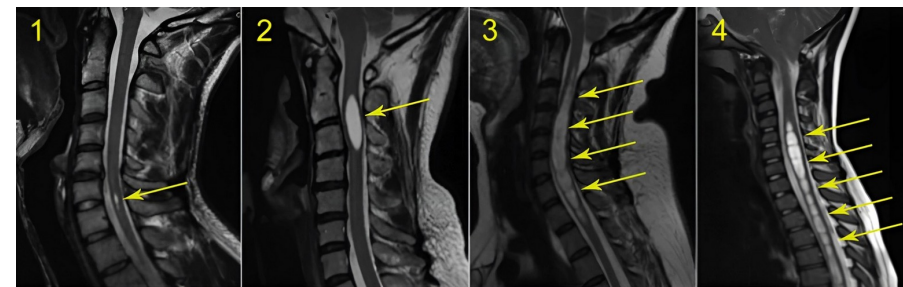
We propose the concept of Neuro-Cranio-vertebral Syndrome (NCVS) and Filum Disease (FD) to define a set of clinical and imaging manifestations that affect the nervous system, the skull and the spine in the form of known diseases like Arnold-Chiari Syndrome Type I (ACSI), Idiopathic Syringomyelia (ISM), Idiopathic Scoliosis (IS) and other anomalies like Platybasia, Basilar Impression, Retroflexed Odontoid and Brainstem Kinking. Based on the arguments outlined in the doctoral thesis “Contribution to the etiology of syringomyelia”[1], the traction of the spinal cord and brain is proposed as the main mechanism involved in the etiopathogenesis of this conditions.

METHODS

We selected a sample of 373 patients with diagnoses including ACSI, ISM, IS, Platybasia, Basilar Impression, Brainstem Kinking, low-lying conus medullaris, and related pathologies were evaluated at the Institut Chiari & Siringomielia & Escoliosis de Barcelona, disposing of complete clinical and imaging data, excluding those with prior neurosurgical interventions or neurological diseases that could alter their presentation. A descriptive analysis of demographic and clinical data and imaging features was followed by the analysis of associations using Pearson’s chi-square test, Kendall’s test for ordinal variables, and the Mantel-Haenszel chi-square test for stratified data ($p < 0.05$). Symptoms and signs were grouped by topography to create continuous variables, which were analyzed alongside imaging features using Student’s t-test and Pearson’s correlation coefficient with scatter plots.

REFERENCES

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Scheme 1. Classification of the extension of syringomyelic cavities Grade 1 (less than one vertebral segment); grade 2 (between 1 and 5 vertebral segments); grade 3 (between 6 and 10 vertebral segments); grade 4 (more than 10 vertebral segments).

RESULTS

Epidemiology

FD predominantly affects females (72%). The mean age at diagnosis is 33.6 years (SD ± 15.9), and in 48% of cases, the time from symptom onset to diagnosis exceeds 10 years.

Most Frequent Symptoms

- Headache (84%)
- Lumbosacral and cervical pain (72%)
- Balance disturbances (72%)
- Paresthesias (70%)
- Thoracic back pain (65%)
- Visual disturbances (57%)
- Lower limb pain (56%)
- Other common symptoms include nervousness, fatigue, upper limb weakness, nausea/vomiting, and sphincter dysfunction (~49–53%).

Predominant Clinical Signs

- Altered deep tendon reflexes in upper (86%) and lower limbs (82%)
- Abnormal plantar reflexes (73%)
- Reduced grip strength (70%)
- Sensory alterations (temperature, touch) (~65–69%)
- Spontaneous nystagmus (55%), positive Romberg test (50%)
- Positive Mingazzini’s test (66%)

- kinking of the brainstem: a common etiology with idiopathic syringomyelia, scoliosis and Chiari Malformations] Revista de Neurología 24:1241-1250.
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Relevant Imaging Findings

- Abnormal position of cerebellar tonsils (93%): 73% descent, 20% impaction (**Fig 2**)
- Low conus medullaris below T12–L1 (88%) (**Fig 3**)
- Vertebral column deviation (76%) (**Fig 4**)
- Multiple disc disease (72%) (**Fig 1**)
- Syringomyelic cavity with different extensions (52%) (**Scheme 1**)

Statistically Significant Correlations

- The level of the conus medullaris correlates with cerebellar tonsil descent ($p = 0.008$) and vertebral deviation ($p = 0.045$).
- Tonsillar descent is associated with cervical syringomyelia ($p = 0.003$), and inversely with non-cervical (thoracic/thoracolumbar) syringomyelia ($p = 0.001$).
- Descent of cerebellar tonsils and Deviation of the vertebral column ($p = 0.014$).
- Positive straight leg raise test ($p = 0.048$) and reduced grip strength ($p = 0.019$) are associated with the level of conus medullaris
- Altered tendon reflexes associated with cervical syringomyelia ($p = 0.005$) and vertebral deviation ($p < 0.001$)

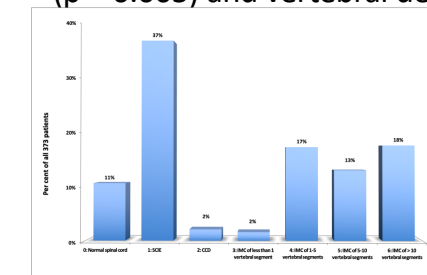


Fig 1. Distribution of the various degrees of medullary abnormalities in the selected 373 patients. SCIE = SpinalCordIschemia-Edema; CCD = Central Canal Dilatation; IMC = Intramedullary Cyst

