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Chiari I malformation: should we operate pictures or children? Proposal of a diagnostic and therapeutic flow chart based on the retrospective analysis of 450 monoinstitutional cases

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Introduction: There are still many discussions about treatme for Chiari I Malformation (CM1) and Syringomyelia in children, both on indications and on surgical technique; Complex Chiari are reported to need Craniovertebral Stabilization in as much ad 50% of the cases.

Objective: The aims of the present review were:

- to evaluate the results of the Craniovertebral Decompression with/without duroplasty and/or Tonsillar resection in a large series of operated Children (150), especially focusing on the controversial points of the association with tethered cord (real or so occult) and craniovertebral instability;
- to define the correct timing for surgery by the experience of follow-up in a series of 300 asymptomatic children about the natural history.

Methods: 150 children were **operated** for CM at the National Neurological Institute of Milan between 1986 and 2013. Age at time of surgery ranged from 1 up to 17 years (mean 11). Preoperative MR was performed in every case and extended on the whole nevraxis to role out associated malformations. All the patients had symptoms before surgery, in the 70% due to the associated syringomyelia.

300 **asymptomatic children** were followed clinically and by annual MRI for a mean time of 4 years: just 10 deserved surgery, despite high extent of downward tonsilar migration present in many cases, while 7 had an upward migration of the tonsils.

In the **surgical series**, there were no major surgical morbidity and no mortality. The preoperative symptoms improved when related to CM1. The associated Syringomyelia reduced in more than 80% of children and disappeared in a significant number, but often a reoperation with tonsilar resection was needed. The association with tethered cord remained rare (2%) despite it was searched in all the cases; when present and symptomatic it needed a double treatment, detethering by itself being insufficient to obtain also tonsilar ascent.

A high percentage of associated Craniovertebral Junction Malformations (CVJM) was documented at MRI; these cases were defined as Complex CM and submitted to dynamic MRI: mild instability was documented in few patients and none deserved fixation up to now.

Conclusions: Children may present peculiar picture of CM, as syncopes and acute paraparesis. The clinical symptoms are often more serious than in the adult population, but the results of surgery, especially on the syrinx, may be better if aggressively treated. In our experience, despite the high incidence of CVJM in CM1, true clinical and MRI instability deserving CV fixation is very rare. Asymptomatic children had a low risk to develop symptoms at mean time follow-up, suggesting that CM1 occasionally diagnosed by MRI performed for other symptoms may be safely followed by annual MRI along growing.