

# Chiari I Malformation: Should We Operate Pictures or Children?Proposal of a Diagnostic and Therapeutic Flow Chart Based on the Review of 450 Monoinstitutional Cases

Laura Grazia Valentini MD

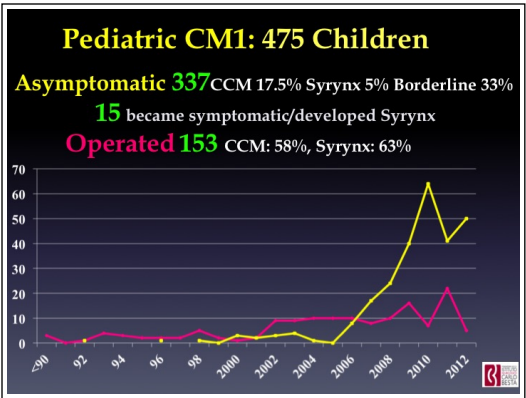
Fondazione Istituto Neurologico "C. Besta", Milano, Italy

## Introduction

There are still many discussions about treatment for Chiari I Malformation (CM1) and Syringomyelia, both on indications and on surgical technique; Complex Chiari (CCM) are reported to need Craniovertebral Stabilization in as much as 50%.

## Methods

The aims are to evaluate the results of CVD with/without duroplasty and/or Tonsillar resection in a large series of operated Children (150), focusing on the controversial points (association with tethered cord and craniovertebral instability) and to define the correct surgical timing by the follow-up in the series (300 asymptomatic children) about the natural history. 150 children were operated for CM at Institution. the asymptomatic children were followed by annual MRI for a mean time of 4,5 years.



## Results-Asymptomatic

377 pauci or a-symptomatic children were followed for a mean time of 4,5 years: 311 (92.2%) remained stable, 11 (3,3%) displayed cranial tonsils migration and 15 (4.5%) worsened and deserved surgery for symptoms or syrinx occurrence.

**Pediatric CM1:**  
Cause leading to Diagnosis

Headache  
Mental Retardation  
Growing Delay  
Epilepsy  
NF  
Autism  
Precocious Puberty  
Other

>30%  
**Associated Genetic Disorder**

Diagnosis

- Costello syndrome
- Idiopathic growth hormone deficiency
- PTEN syndrome
- Cutis marmorata telangiectatica congenita
- Charge syndrome
- Alport syndrome
- Angelman syndrome
- Opitz syndrome
- Neurofibromatosis type 1
- Crouzon syndrome
- Muenke syndrome
- Polysyndactyly
- Sagittal synostosis
- MEF2C haploinsufficiency
- 2p15p16 haploinsufficiency
- TAR syndrome
- Cloacal ectropia

CI, 5y,Cutis Marmorata

## Results - Operated Children

In the surgical series there was no major surgical morbidity nor mortality. Preoperative symptoms improved when related to CM1. Associated Syringomyelia reduced in >80% and disappeared in a significant number, but 30 pts needed CBL tonsils resection for failure of simple CV with duroplasty. An high percentage of associated Craniovertebral Junction Malformations (CVJM) was documented and defined as Complex CM (CCM). All were submitted to dynamic MRI or CT and none deserved fixation except one adolescent, that had true instability needing fixation (0,6%)

**Pediatric CM1 :  
CV Decompression  
145 Expansive Duroplasty**

**COMPLICATIONS:**

- Frequent venous bleeding by anomalous sinus, controlled by suturing
- 18 (12%) CSF leaks
- 6 (4%) needed revision (2 SS + CM1)

**Pediatric CM1**  
A couple of tricks to prevent CSF Leakage

1. Insertion of Dural Substitute between Arachnoid and Duroplasty

2. Suturing the Plasty by an unresorbable stitch (Prolene)

35 cases, no CSF collections !

**43 (28%) CVD + CBL tonsils/Resection**

4 second choice for failure  
39 for relevant tonsillar migration/ severe crowding

**Pediatric CM1: Results on syrinx  
24 Complete Shrinkage**

D.P.E., 13 years, Female:  
Syrinx collapse 8 months after surgery

**Pediatric CM1: Open Problems**  
Postoperative CV Instability

S.A, 8 y, Male: Postoperative CVJ Hypermobility

flexed

extended

B.T, 4 y, Male: Postoperative CVJ Instability?

## Conclusions

The surgical technique applied led to good results on CM1 related symptoms. This aggressive approach in symptomatic children was aimed to syrinx shrinkage: CVD + duroplasty as first choice, followed by tonsils coagulation/resection in case of failure of the first year. Despite the high incidence of CVJM observed, true clinical and MRI instability was quite rare (<1%). Surgery was spared in a huge population of pauci or asymptomatic CM1 children. They mainly remained stable; a few changed and had the same chance (<5%) to heal and to progress.

Symptoms, first of all headache, may be wrongly attributed to CM1, leading to a surgery burdened by a clinical failure despite the anatomic success. To avoid this, we propose the following Flow Chart for diagnosis and treatment of Pediatric CM1.

