Gestione del paziente asintomatico o con diagnosi incidentale

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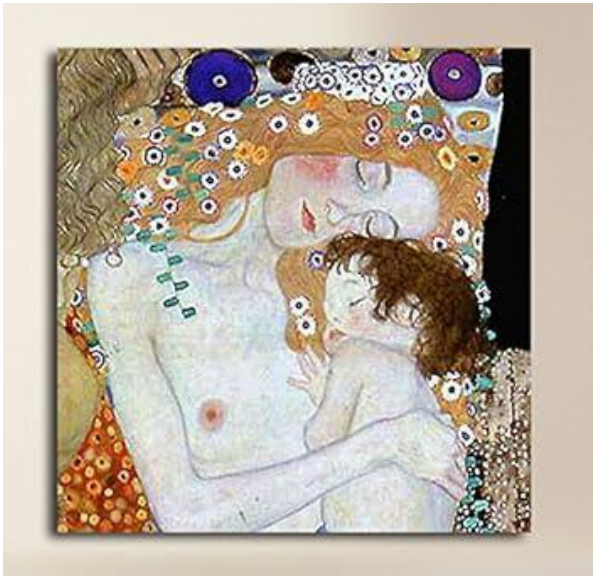
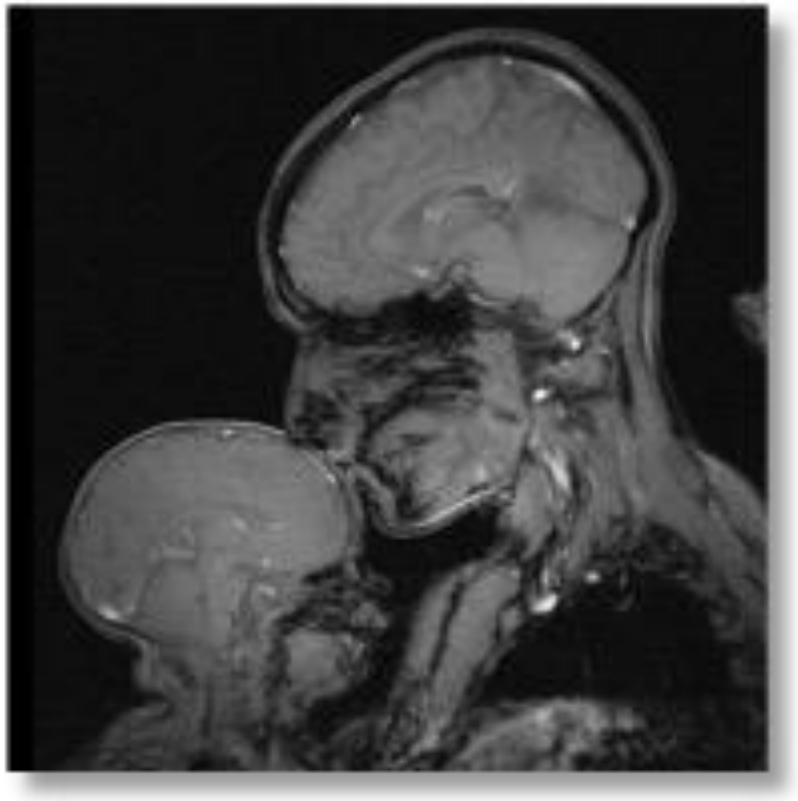
L’evoluzione dell’ «imaging»…

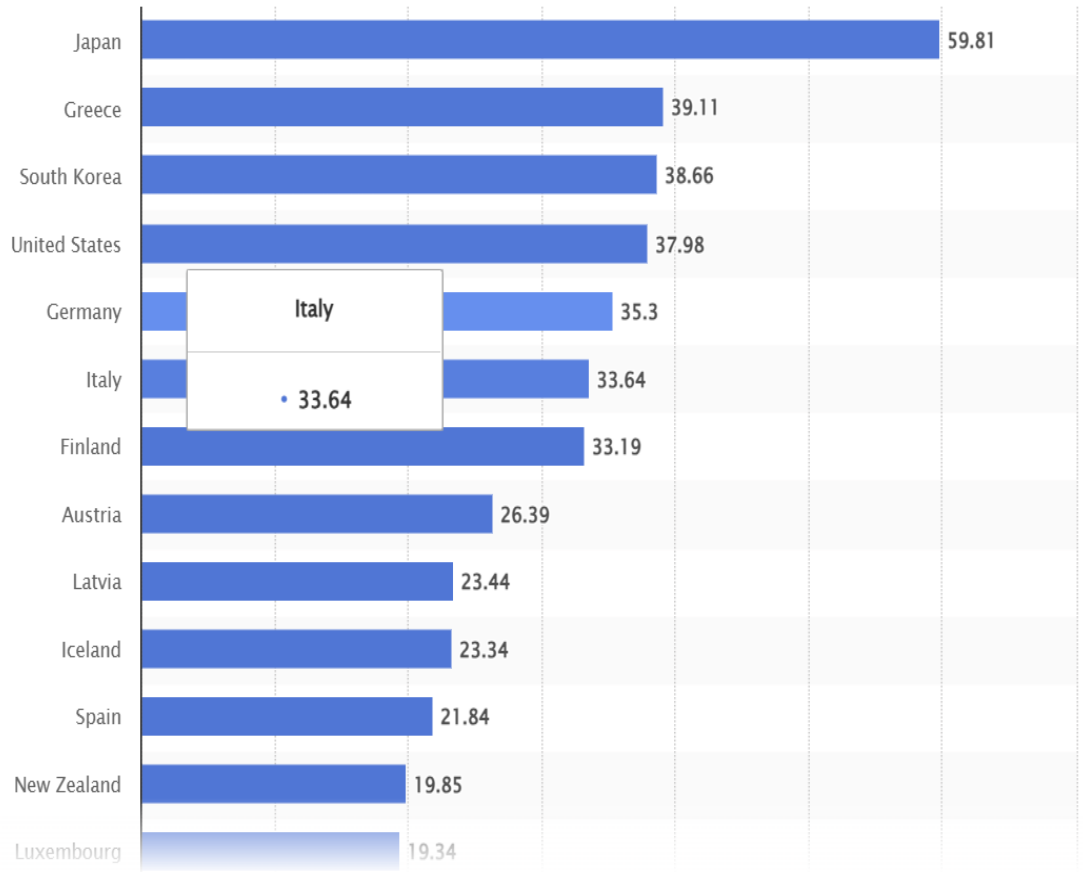




Diffusione della RM: effetti negativi e positivi

Number of magnetic resonance imaging (MRI) units in selected countries as of 2024

(per million population)



DOWNLOAD

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Source

→ Show sources information
→ Show publisher information
→ Use Ask Statista Research Service

Release date

2025

Region

OECD

Survey time period

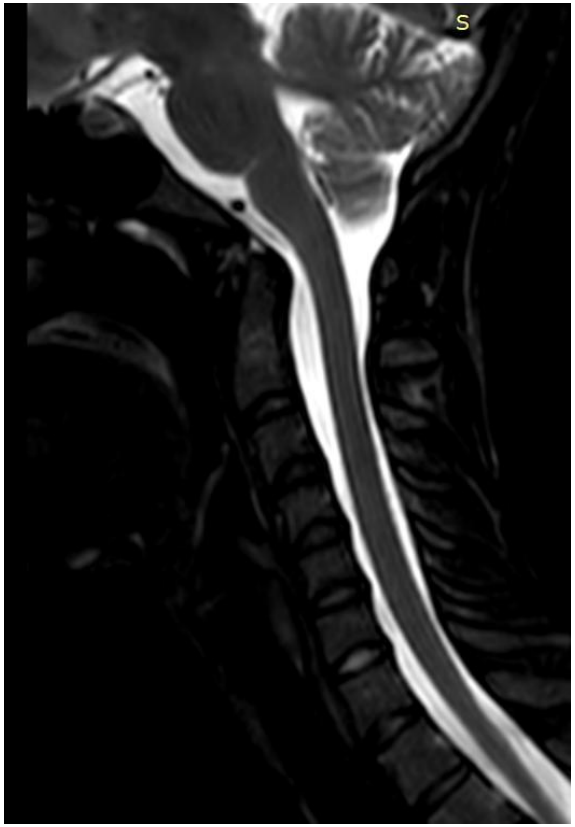
2024

Supplementary notes

Data are for 2024 or latest year available.

VOMIT

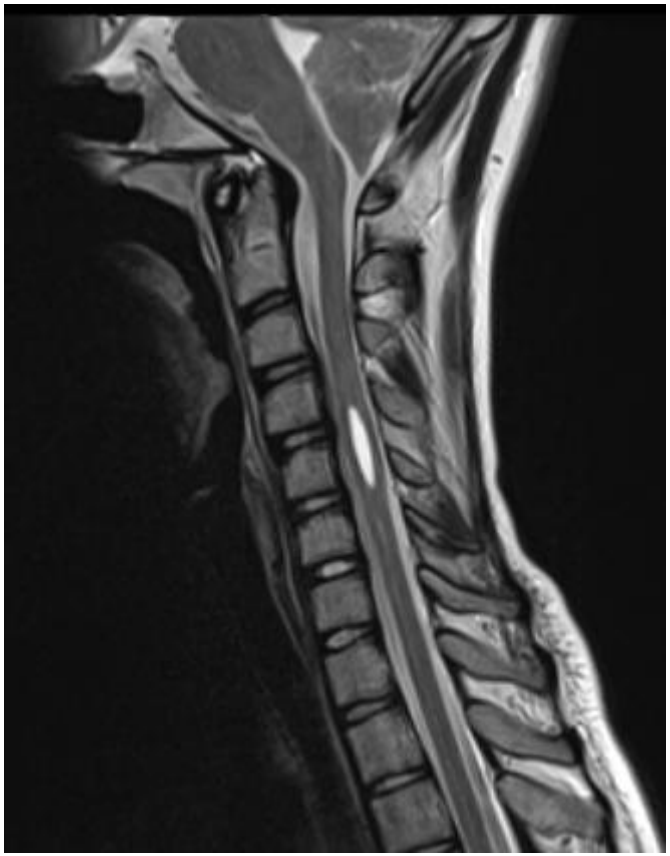
Victim
Of
Modern
Imaging
Technology



F, 19 anni
Diagnosi dopo esame di screening

LUCKI

Lucky
Unintended
Clinical
Knowledge from
Imaging



F, 12 anni
Diagnosi incidentale dopo trauma

Gestione della m. di Chiari incidentale (asintomatica)

Primo passaggio

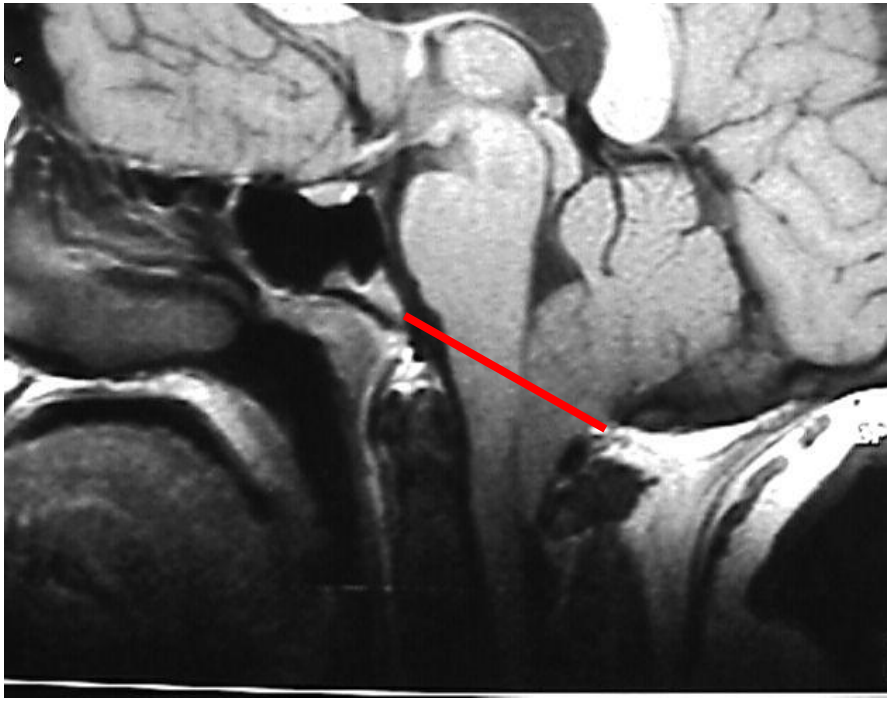
Diagnosi adeguata

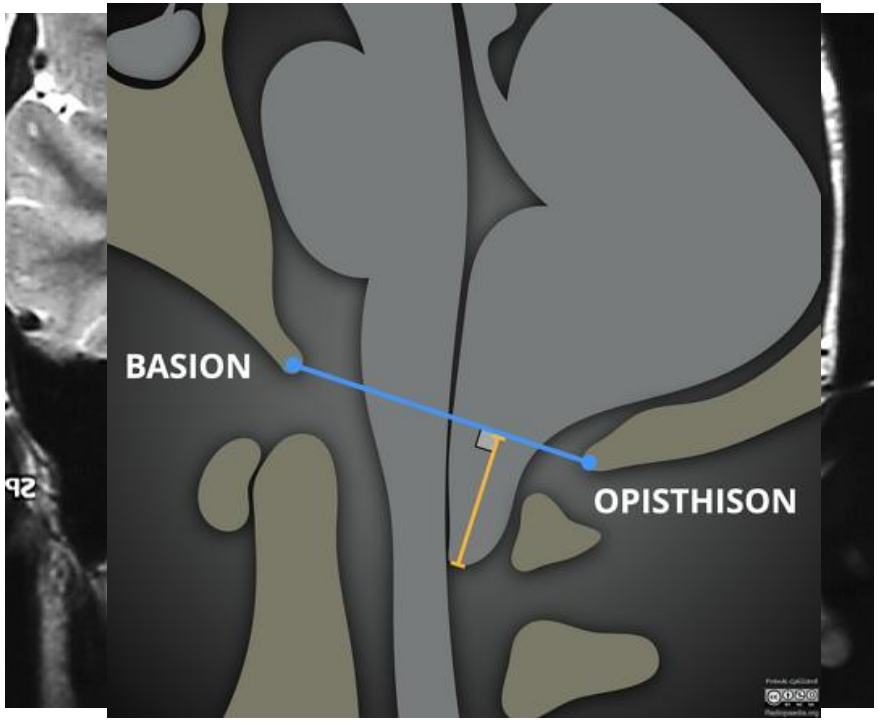
E’ una m. di Chiari?

E’ incidentale?

Definizione più accettata

"Prolapse of the cerebellar tonsils 5 mm or more below the foramen magnum (basion-opisthion line or McRae line)"





Elster AD and Chen MY, Radiology, 1992

Misurazioni

RESEARCH Open Access

Cerebellar tonsil ectopia measurement in type I Chiari malformation patients show poor inter-operator reliability

Braden J. Lawrence[1,2†], Aintzane Urbizu[3†], Philip A. Allen[4], Francis Loth[5], R. Shane Tubbs[6], Alexander C. Bunck[7], Jan-Robert Kröger[7], Brandon G. Rocque[8], Casey Madura[9], Jason A. Chen[10], Mark G. Luciano[11], Richard G. Ellenbogen[12], John N. Oshinski[13], Bermans J. Iskandar[14] and Bryn A. Martin[1*]

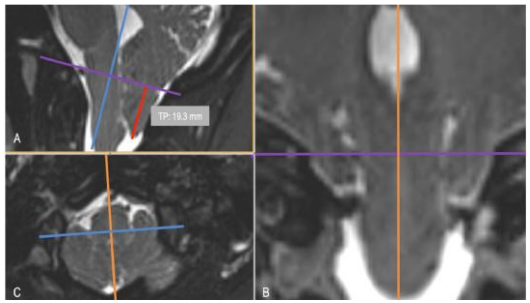


Table 2 Descriptive statistics calculated for seven expert operators by MRI subject group across 33 CM-I patients

Measure	Subject group	Operator 1	Operator 2	Operator 3	Operator 4	Operator 5	Operator 6	Operator 7
Range (mm)	All	13.00	28.00	28.00	22.30	23.00	17.00	30.10
	CM-Ia	8.50	10.00	16.20	13.40	10.00	12.00	15.30
	CM-Is	11.80	16.00	14.90	16.70	15.00	13.00	19.90
	Control	2.50	9.00	10.20	6.70	7.00	7.00	13.40
Mean (mm)	All	3.43	4.91	5.87	6.18	8.73	6.55	3.90
	CM-Ia	3.49	6.00	7.55	6.73	8.50	7.67	4.85
	CM-Is	6.10	9.09	10.65	10.37	12.73	8.73	9.34
	Control	0.42	− 0.90	− 1.23	0.93	4.60	2.80	− 3.23
SD (mm)	All	3.65	5.33	6.23	5.48	4.99	4.27	6.90
	CM-Ia	2.96	2.95	4.23	4.15	3.28	3.82	4.43
	CM-Is	3.78	4.40	4.08	4.99	5.28	4.09	5.74
	Control	0.86	2.43	2.78	2.00	2.01	1.89	3.63
Median (mm)	All	2.50	5.50	6.70	6.10	8.00	6.00	3.20
	CM-Ia	3.50	6.00	7.70	6.65	10.00	7.50	4.20
	CM-Is	5.00	8.00	10.50	8.30	10.00	7.00	7.90
	Control	0.00	0.00	− 0.70	0.00	5.50	3.00	− 2.70
IQR (mm)	All	5.30	8.00	10.50	9.15	5.50	7.50	10.55
	CM-Ia	5.83	5.00	6.40	6.95	5.75	6.75	6.90
	CM-Is	5.80	4.00	6.70	7.50	9.00	7.00	8.80
	Control	0.43	2.50	3.18	1.15	3.00	3.00	5.20

CM-Ia type I Chiari malformation asymptomatic, *CM-Is* type 1 Chiari malformation symptomatic, *IQR* interquartile range, *SD* standard deviation

7 Radiologi esperti su 33 RM

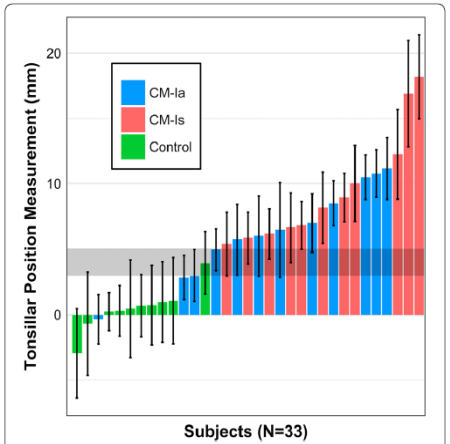
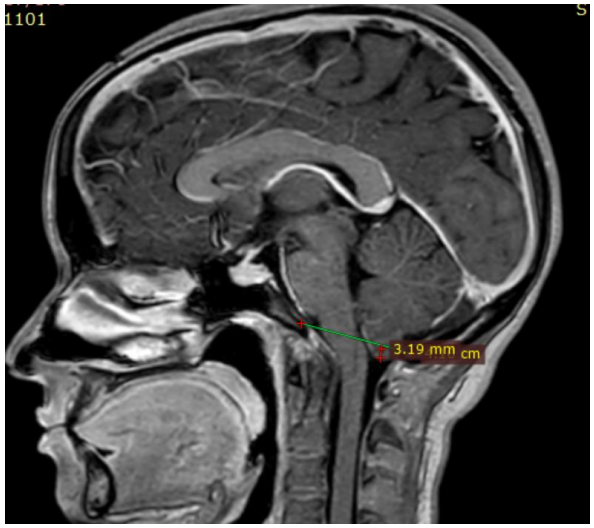
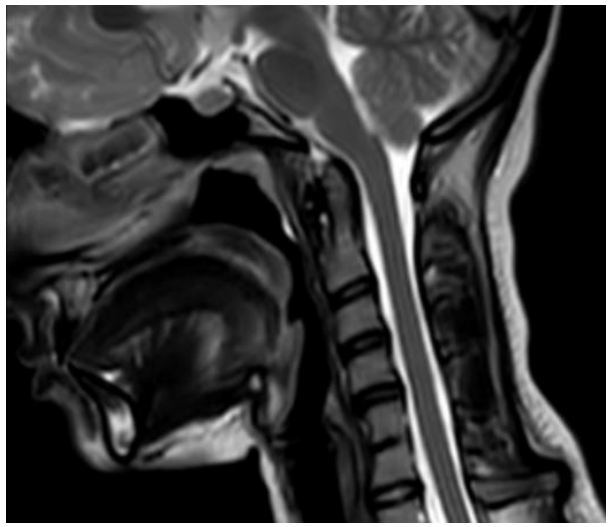


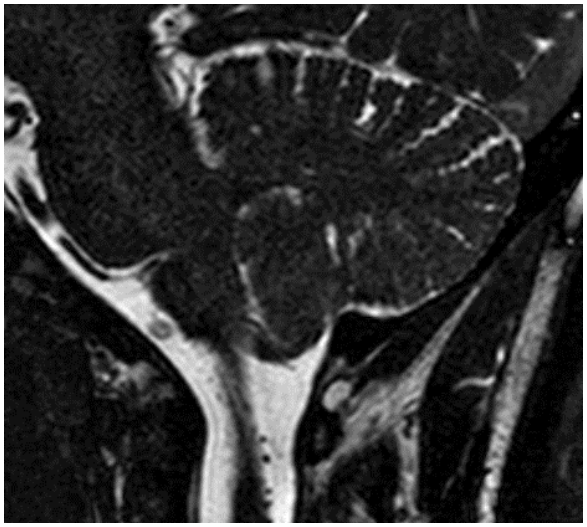
Fig. 2 Mean ± 1SD of N = 33 tonsillar position measurements for the seven operators (green = control, red = asymptomatic CM-I patient, blue = symptomatic CM-I patient). Shaded horizontal rectangle indicates TP 3–5 mm

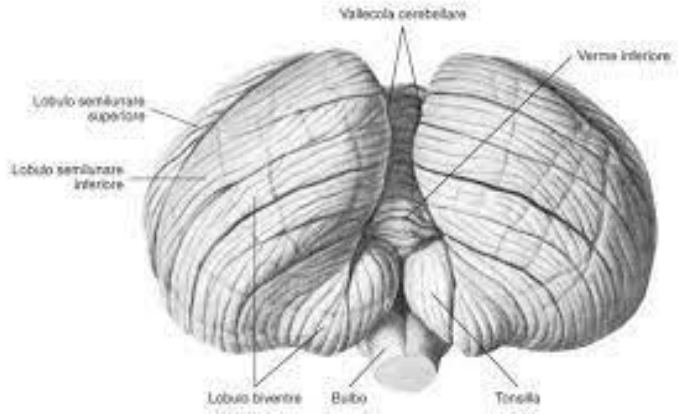
Chiari *borderline?*

Erniazione tonsillare < 5 mm (3-4 mm)









Misurazione sulla linea mediana!

Definizione più accettata?

"Prolapse of the cerebellar tonsils 5 mm or more below the foramen magnum (basion-opisthion line or McRae line)"

Emendamenti…

"Discesa caudale delle tonsille cerebellari ≥ 3 mm più almento uno dei seguenti indici di affollamento del forame magno/compressione degli spazi subaracnoidei:

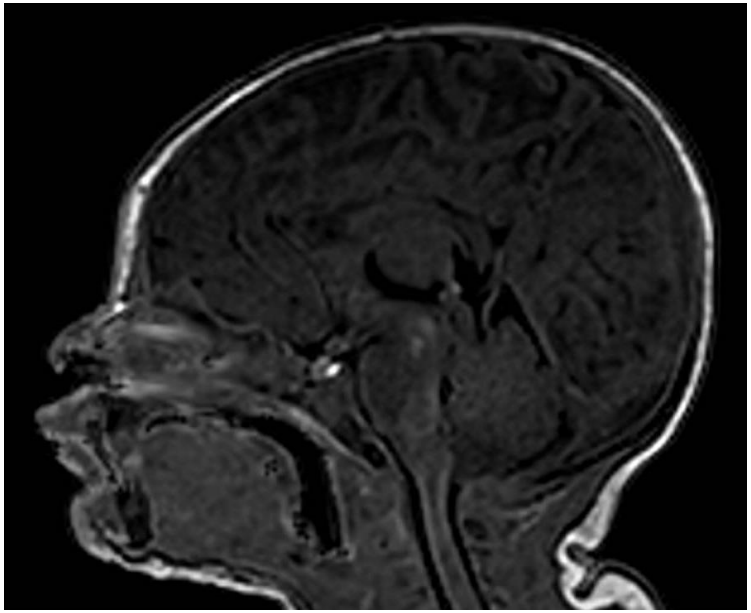
Compressione degli spazi liquorali posteriori e/o anteriori

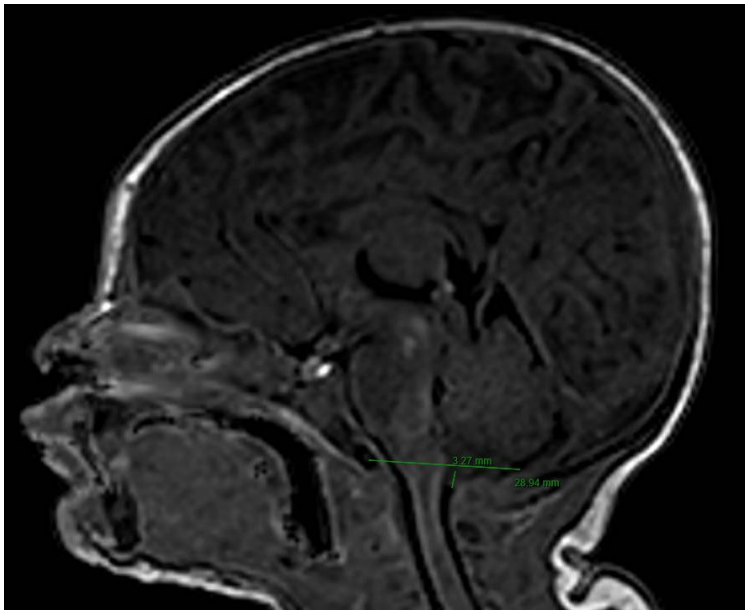
Ridotta altezza del sopraoccipite/aumentata inclinazione del tentorio

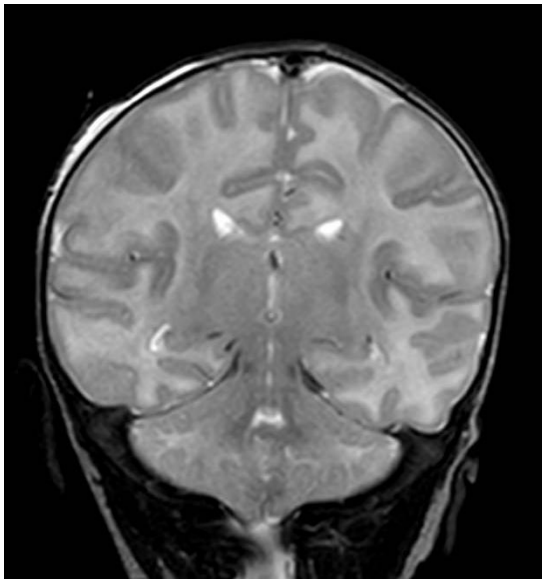
Aspetto appuntito delle tonsille cerebellari

Inginocchiamento del troncoencefalo (bulbo)"

Definizione

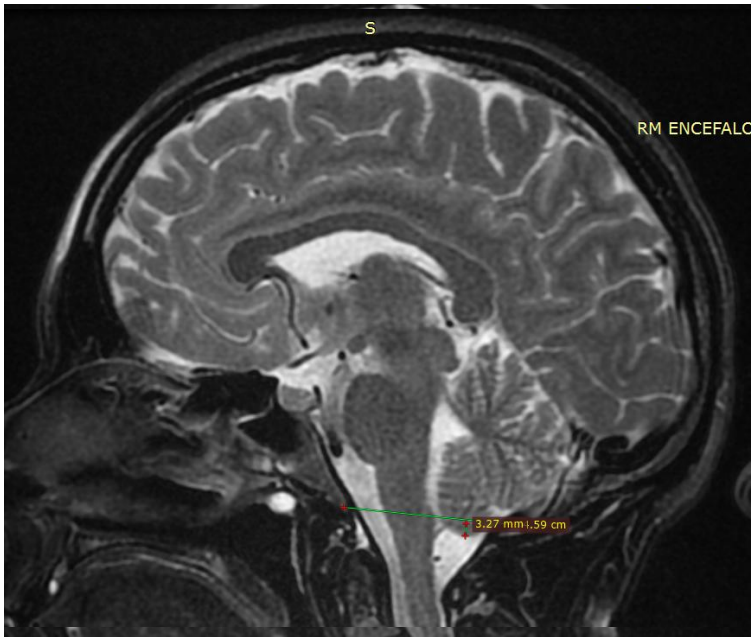


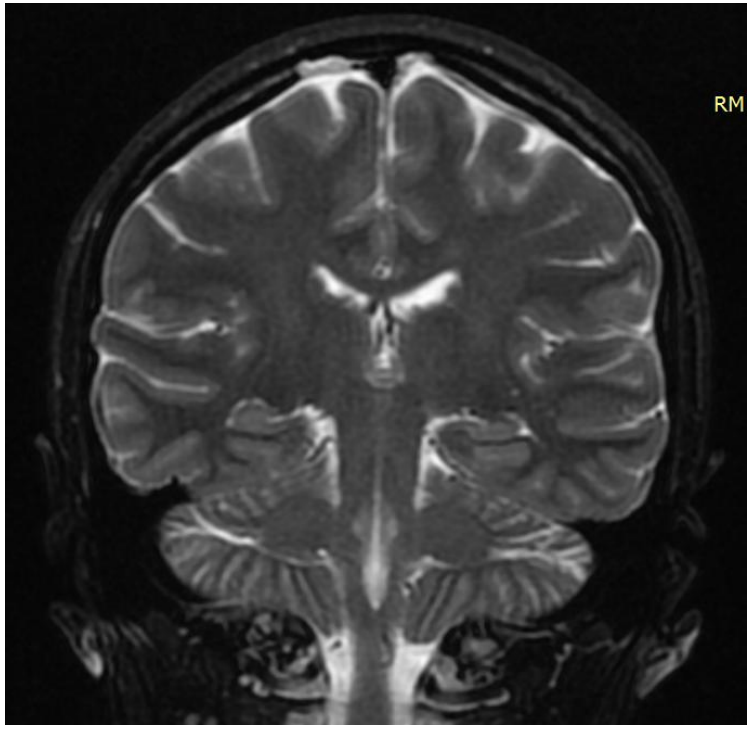




Discesa tonsillare lieve (> 3 mm) + tonsille appuntite + gossa posterior piccola (angolo tentoriale ridotto) + cisterna magna ridotta = m. di Chiari

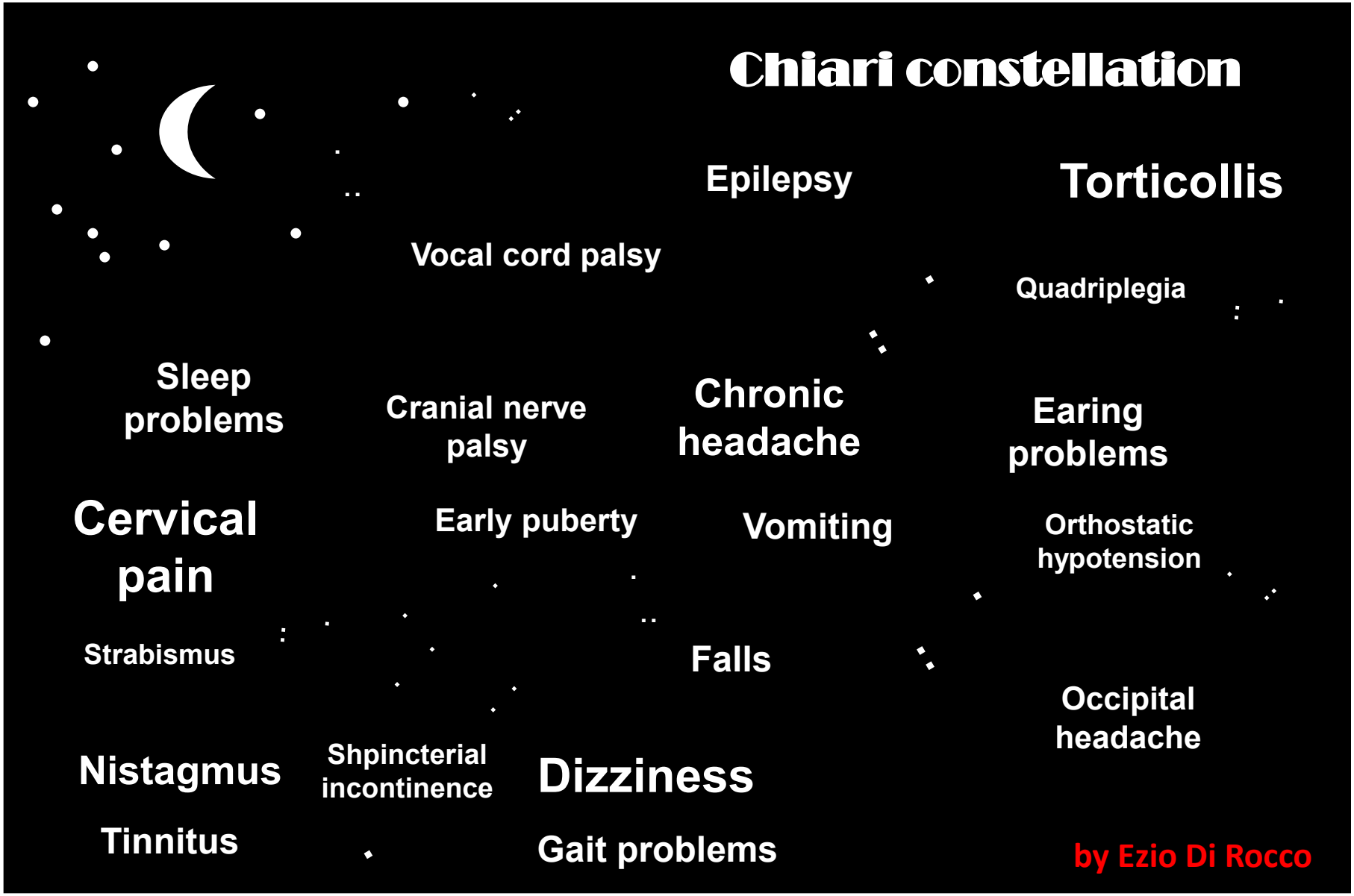
Definizione



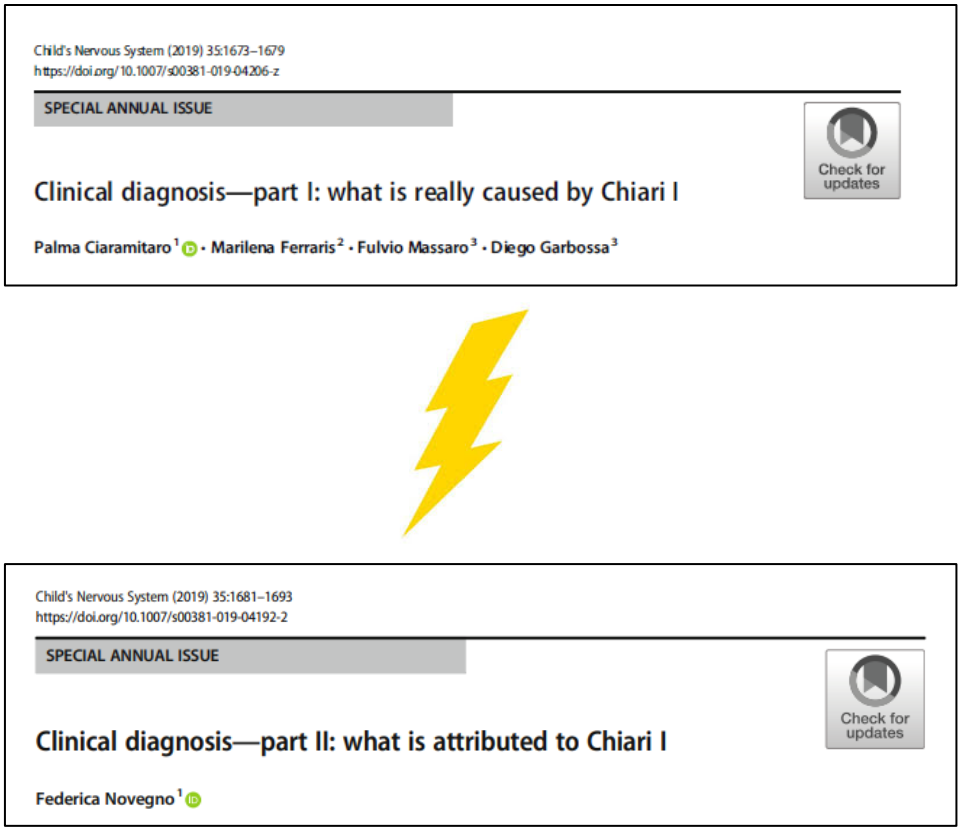


Discesa tonsillare lieve (> 3 mm) + tonsille arrotondate + normale volume della fossa posteriore + normali spazi subaracnoidei ≠ m. di Chiari ma semplice discesa tonsillare

Quadro clinico



Quadro clinico…



Child's Nervous System (2019) 35:1673–1679
https://doi.org/10.1007/s00381-019-04206-z

SPECIAL ANNUAL ISSUE

Clinical diagnosis—part I: what is really caused by Chiari I

Palma Ciaramitaro[1] · Marilena Ferraris[2] · Fulvio Massaro[3] · Diego Garbossa[3]

Child's Nervous System (2019) 35:1681–1693
https://doi.org/10.1007/s00381-019-04192-2

SPECIAL ANNUAL ISSUE

Clinical diagnosis—part II: what is attributed to Chiari I

Federica Novegno[1]

La diagnosi di m. di Chiari è incidentale nel 60-70% dei casi!
Oltre il 70% dei pazienti con diagnosi incidentale rimane asintomatico

Clinical picture: symptoms

Pain symptoms

Headache (suboccipital > generalized)
Cough headache
Neck pain
Retroorbital pain
Limbs and back nonradicular pain
Facial pain
Throat pain
Migraine

Non-pain symptoms

Neck, back and limbs dysesthesia
Dissociated/suspended sensory loss
Hypoesthesia/hyperesthesia
Paresthesia
Analgesia
Poor position sense
Dysphagia
Palpitations
Diplopia
Blurred vision
Photophobia
Dizziness
Disequilibrium
Vertigo
Nausea
Oscillopsia
Hearing loss
Hyperacusis

Massimi L et al, Adv Tech Standard Neurosurg, 2011

Clinical picture: signs

Brainstem

Syncope, drop attack
Sudden death
Sleep apnea
Respiratory imbalance
Recurrent aspiration
Palatal weakness
Tongue atrophy
Cricopharyngeal achalasia
Dysarthria
Dysphagia
Hiccups
Facial sensory loss/palsy
Vocal cord paralysis
Reduced gag reflex
Nystagmus
Hearing loss
Oculomotor nerves palsy
Papilledema
Motor/sensory loss
Hypertension
Sinus bradycardia

Cerebellum

Truncal/appendicular ataxia
Limb clumsiness
Dysmetria
Dysarthria
Nystagmus
Poor coordination
Tremors

Spinal cord

Hyper/hyporeflexia
Babinski sign
Clonus
Motor/sensory loss
Muscular weakness/atrophy
Spasticity
Loss of proprioception
Numbness
Urinary incontinence
Fecal incontinence
Impotence
Trophic abnormalities

Quadro clinico: certo vs possibile…

1. **Cefalea da sforzo**: tipicamente posteriore, di breve durata, esacerbata da tosse, sforzi, manovra di Valsalva o movimenti del collo; isolate o associate a vertigini, atassia, incertezza nella marcia. **>75%**

2. **Nistagmo**: sia verticale che orrizzontale. **23-70%**

3. **Segni midollari + scoliosi**: soprattutto in caso di siringomielia. **Fino a 80%**

4. **Atassia cerebellare: 40%**

CNS GUIDELINES FOR PATIENTS WITH CHIARI MALFORMATION

Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for Patients With Chiari Malformation: Symptoms

Eric M. Jackson, MD*, Sarah Jernigan, MD, MPH†, Jeffrey S. Raskin, MS MD§, Laurie L. Ackerman, MD‖, Libby Kosnik Infinger, MD, MPH¶, Cormac O. Maher, MD#, Toba Niazi, MD**, Jogi V. Pattisapu, MD††, Rabia Qaiser, MD‡‡, Carolyn Quinsey, MD‖‖, Brandon G. Rocque, MD, MS¶¶, Howard Silberstein, MD##, Shobhan Vachhrajani, MD, PhD, FRCSC***, David F. Bauer, MD, MPH†††§§§

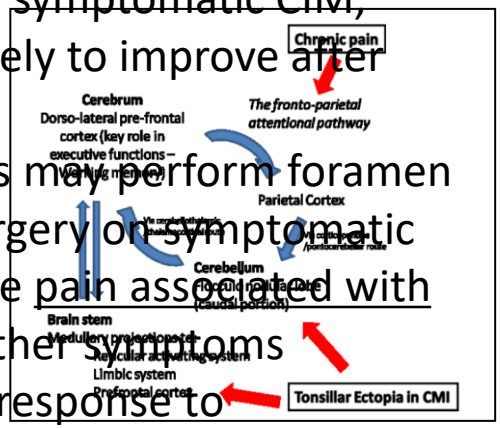
RECOMMENDATIONS

2-1. In patients operated for symptomatic CIM, what symptoms are most likely to improve after surger

Recommendation: Clinicians may perform foramen magnum decompression surgery on symptomatic patients with CIM to improve pain associated with strain-related headache. Other symptoms demonstrate more variable response to decompression.

Strength of recommendation: Grade C

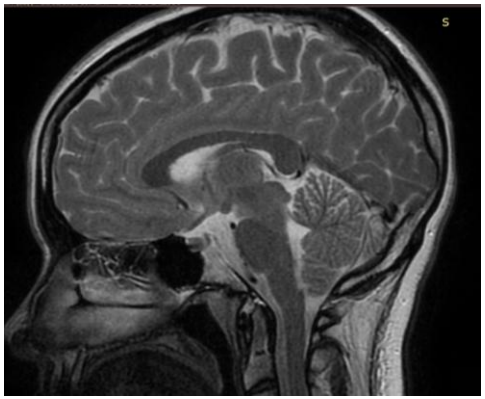
Level III evidence

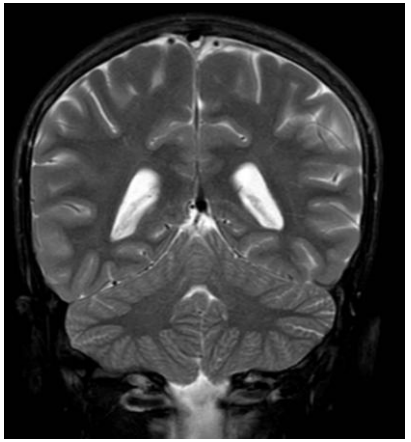


In sintesi: diagnosi «incidentale»

Primo scenario
No Chiari, no sintomi

M, 13 anni
Diagnosi incidentale di tonsille basse dopo trauma cranico (TC)





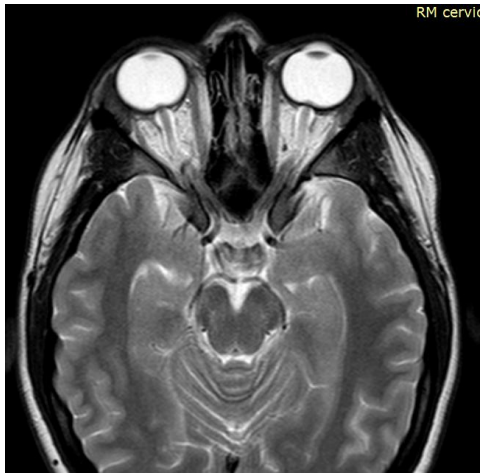
Restare in contatto con il medico
Nuova RM? Solo durante lo sviluppo?

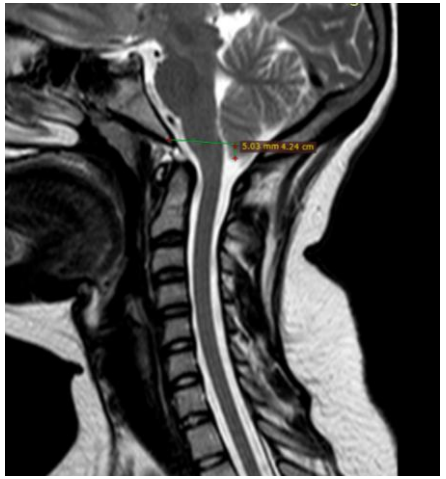
In sintesi: diagnosi «incidentale»

Primo scenario
No Chiari, no sintomi

Secondo scenario
No Chiari, sintomi

F, 15 anni
Diagnosi «incidentale» dopo RM eseguita per cefalea, papilledema e calo del visus (pseudotumor cerebri)





Trattammento della patologia primaria

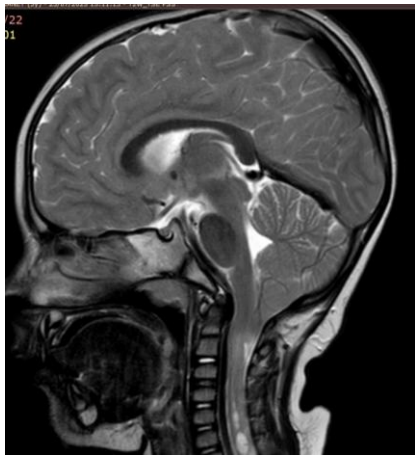
In sintesi: diagnosi «incidentale»

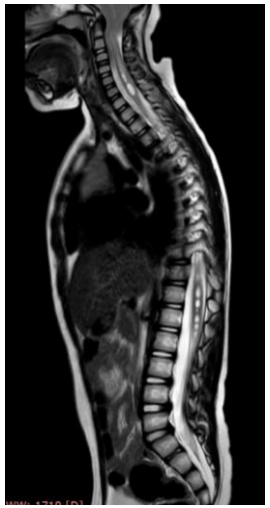
Primo scenario
No Chiari, no sintomi

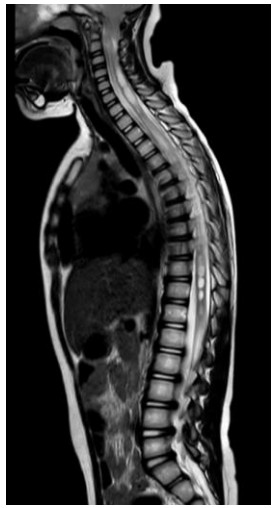
Secondo scenario
No Chiari, sintomi

Terzo scenario
Chiari, apparentemente no sintomi

F, 4 anni
Diagnosi «incidentale» dopo RM eseguita per scoliosi precoce e progressiva







Trattamento della m. di Chiari (e siringomielia)

In sintesi: diagnosi «incidentale»

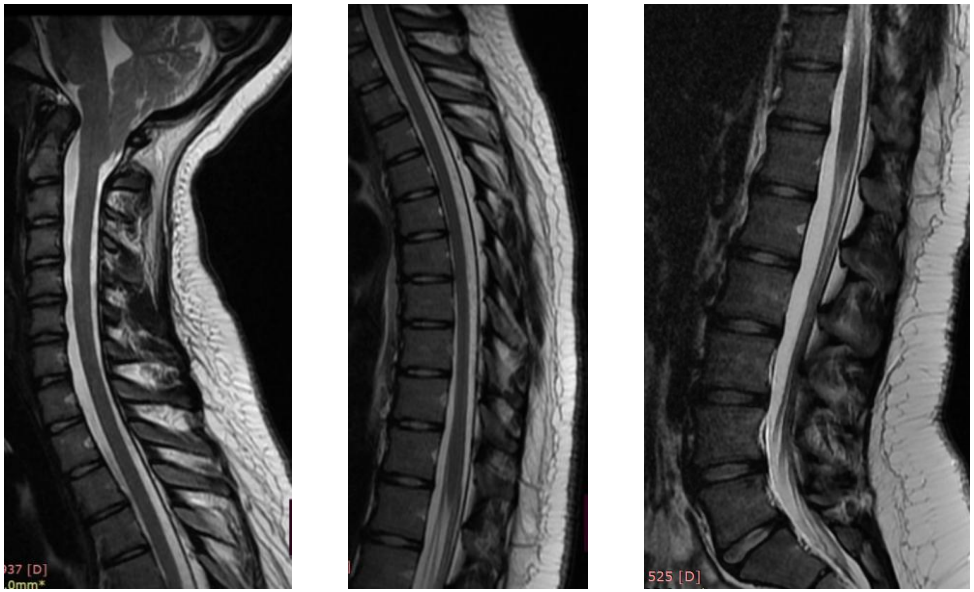
Primo scenario
No Chiari, no sintomi

Secondo scenario
No Chiari, sintomi

Terzo scenario
Chiari, apparentemente no sintomi

Quarto scenario
Chiari, no sintomi

F, 16 anni
Diagnosi incidentale dopo trauma sportivo (canottaggio), asintomatica



Follow-up

Gestione della m. di Chiari incidentale (asintomatica)

Secondo passaggio

Chiari incidentale

(m. di Chiari vera + paziente realmente asintomatico)

Chirurgia preventiva?

No

Follow-up?

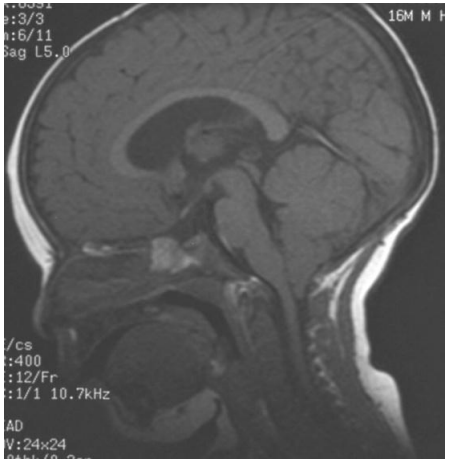
Sì

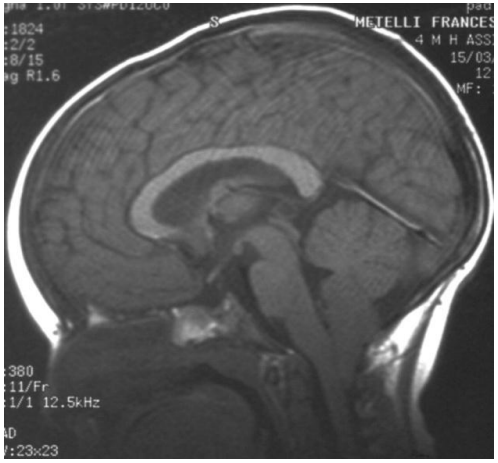
Per quanto tempo?
come?

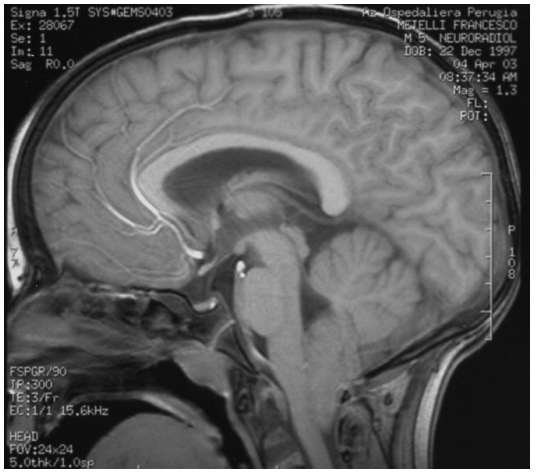
Restrizioni?

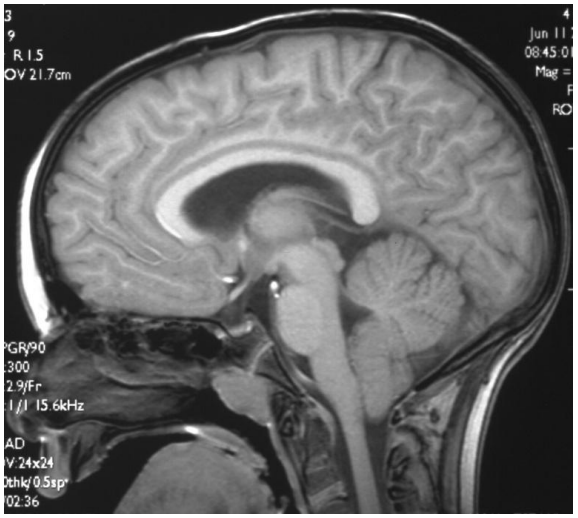
No (di solito)

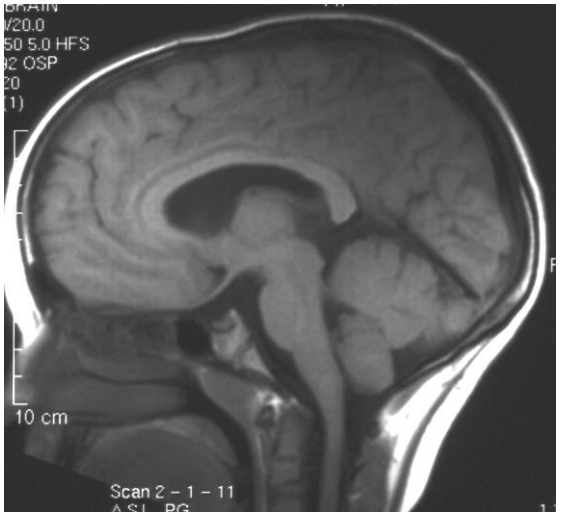
No chirurgia preventiva











Risoluzione spontanea in 10 anni (paziente asintomatico)

Storia naturale: serie personale

See the corresponding editorial in this issue, pp 177–178.

J Neurosurg Pediatrics 2:179–187, 2008

The natural history of the Chiari Type I anomaly

FEDERICA NOVEGNO, M.D., MASSIMO CALDARELLI, M.D., ANTONIO MASSA, M.D., DANIELA CHIEFFO, PH.D., LUCA MASSIMI, M.D., BENEDETTA PETTORINI, M.D., GIANPIERO TAMBURRINI, M.D., AND CONCEZIO DI ROCCO, M.D.

Department of Pediatric Neurosurgery, Catholic University Medical School, Rome, Italy

Neurol Sci (2011) 32 (Suppl 3):S275–S277
DOI 10.1007/s10072-011-0684-3

GENERAL

Natural history of Chiari type I malformation in children

Luca Massimi · Massimo Caldarelli · Paolo Frassanito · Concezio Di Rocco

- 36 bambini asintomatici
- Discesa tonsillare: > 10 mm in 54% dei casi
- Follow-up medio: 9,1 anni
- Peggioramento clinico/radiologico che ha richiesto chirurgia in 3 casi (8%), uno dei quali con idrocefalo

Natural history: literature

J Neurosurg Pediatrics 7:375–379, 2011

Outcomes in pediatric patients with Chiari malformation Type I followed up without surgery

Clinical article

David Benglis Jr., M.D., Derek Covington, B.A., Ritwik Bhatia, Sanjiv Bhatia, M.D., Mohamed Samy Elhammady, M.D., John Ragheb, M.D., Glenn Morrison, M.D., and David I. Sandberg, M.D.

Department of Neurosurgery, University of Miami/Miller School of Medicine and Miami Children's Hospital, Miami, Florida

- The great majority of cases (both poorly symptomatic and asymptomatic) did not need surgery (119/124: 96%)
- Syringomyelia unchanged in all patients and no new cases during follow-up
- Follow-up: 2.8 years

J Neurosurg Pediatrics 8:214–221, 2011

Natural history of Chiari malformation Type I following decision for conservative treatment

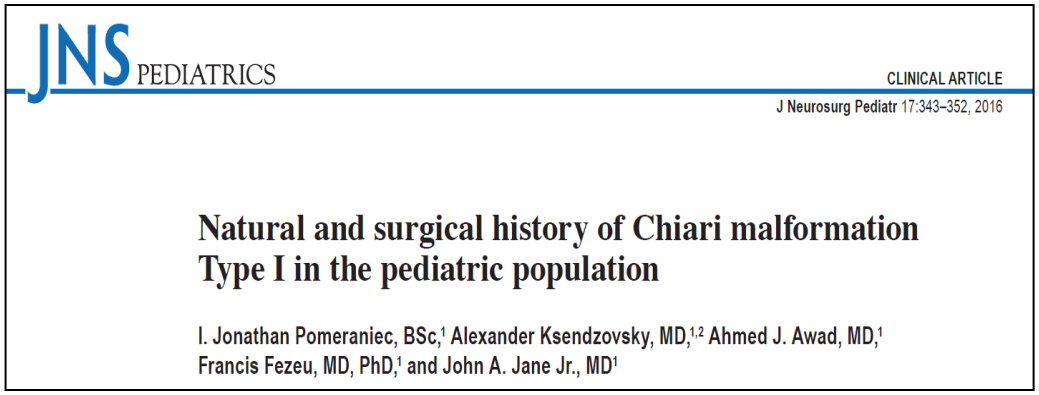
Clinical article

Jennifer Strahle, M.D.,[1] Karin M. Muraszko, M.D.,[1] Joseph Kapurch, B.S.,[1] J. Rajiv Bapuraj, M.D.,[2] Hugh J. L. Garton, M.D.,[1] and Cormac O. Maher, M.D.[1]

Departments of [1]Neurosurgery and [2]Radiology, University of Michigan, Ann Arbor, Michigan

- No patients (147 cases) needed surgery
- Only 9 children developed Chiari symptoms (6%)
- 8 cases developed syringomyelia (5.5%); in 5 of them, hydromyelia was already present; finally, syringomyelia regressed in 3 cases
- Follow-up: 4.6 years

Natural history: literature



JNS PEDIATRICS

CLINICAL ARTICLE

J Neurosurg Pediatr 17:343–352, 2016

Natural and surgical history of Chiari malformation Type I in the pediatric population

I. Jonathan Pomeraniec, BSc,[1] Alexander Ksendzovsky, MD,[1,2] Ahmed J. Awad, MD,[1] Francis Fezeu, MD, PhD,[1] and John A. Jane Jr., MD[1]

- 427 children
- 15 cases required surgery (3.5%)
- Median time to surgery: 21 months
- Median follow-up median: 25.5 months (range: 12-120 months)

Child's Nervous System (2019) 35:1793–1799
https://doi.org/10.1007/s00381-019-04310-0

SPECIAL ANNUAL ISSUE

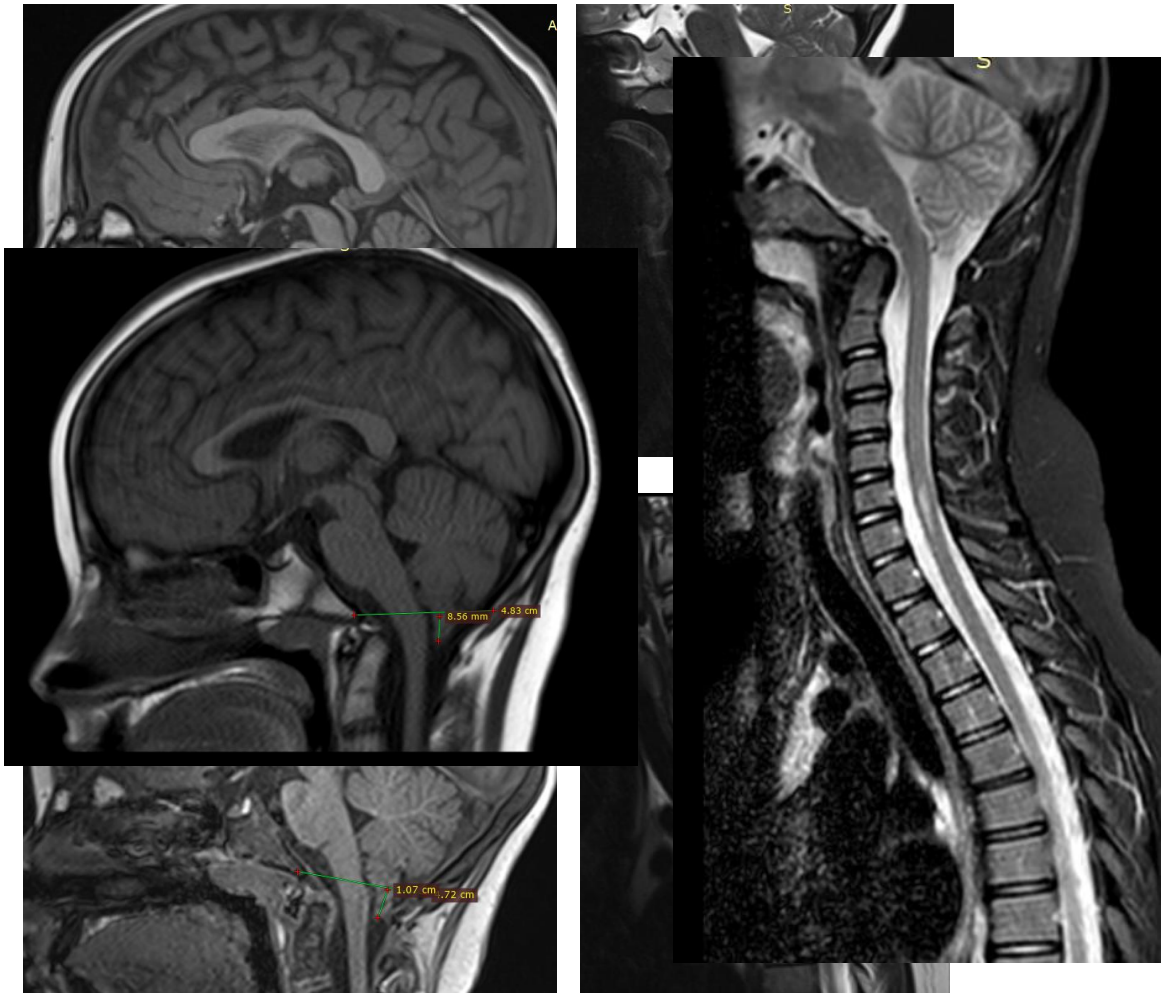
Check for updates

Chiari I malformation in children—the natural history

Ajay Chatrath[1] · Alexandria Marino[1] · Davis Taylor[1] · Mazin Elsarrag[1] · Sauson Soldozy[1] · John A. Jane Jr[1]

- 700 asymptomatic children with no syringomyelia: only 5% developed symptoms and only 2% developed syringomyelia
- 100 symptomatic children without syringomyelia: 7% worsened, 48% spontaneously improved, 2% developed syringomyelia

No chirurgia preventiva



2020

2025

2022

Syringomyelia: natural history

Neurosurg Focus. 2011 Dec;31(6):E13. doi: 10.3171/2011.9.FOCUS11208.

Natural history of untreated syringomyelia in pediatric patients.

Singhal A[1], Bowen-Roberts T, Steinbok P, Cochrane D, Byrne AT, Kerr JM.

Author information

Abstract

OBJECT: The natural history of syringomyelia in pediatric patients remains uncertain. Although symptomatic and operative cases of syringomyelia are well studied, there are fewer articles in the literature on the nonoperative syrinx and its clinical and radiological course. The purpose of this research was to analyze the natural history of untreated syringomyelia in pediatric patients presenting with minimal neurological symptoms.

METHODS: A review of the neurosurgery database at British Columbia's Children's Hospital identified all pediatric patients (< 18 years of age) with syringes identified on MR imaging. Patients were included in this study if they had at least 2 MR images of the spine, at least 1 year apart, while receiving nonoperative treatment. Magnetic resonance imaging was used to determine changes in the size of the syrinx over time. Clinic notes were analyzed to establish demographic and clinical features and to determine any clinical changes over time.

RESULTS: A total of 17 patients were included in the study. Symptoms at presentation were often mild and included limb numbness (3 cases), headaches (2 cases), mild sensory deficits (2 cases), mild motor deficits (3 cases), and intermittent incontinence (7 cases). The consultant neurosurgeon believed that the syrinx was not contributing to the symptoms in these 17 patients. The syrinx either remained unchanged (7 cases) or diminished in size (8 cases) in a total of 15 patients (88%). In the remaining 2 patients the authors noted an increase in syrinx size, in 1 of whom the clinical course also worsened. Both of these patients had a Chiari malformation and subsequently underwent craniocervical decompression. Overall, the mean change was -0.7 mm of maximal axial diameter (range -2.6 to +2.7 mm). Sixteen patients (94%) exhibited no worsening of symptoms over time.

CONCLUSIONS: Syringomyelia often remains stable in patients receiving nonoperative treatment. However, given that 2 (12%) of 17 syringes in this series enlarged, it is likely appropriate to include periodic imaging in the follow-up of these cases.

Syringomyelia: natural history

Child's Nervous System
https://doi.org/10.1007/s00381-023-06273-9

RESEARCH

Natural history of Chiari I malformation with syrinx and dilatation of the central canal in the pediatric population: the CHEO experience

Maria Fernanda Dien Esquivel[1] · Neetika Gupta[1] · Christian Alfred O'Brien[2] · Vid Bijelić[3] · Nick Barrowman[3] · Nagwa Wilson[1] · Albert Tu[4]

Received: 24 August 2023 / Accepted: 27 December 2023

Abstract

Purpose Given that syrinx is often considered an indication of surgery in children with Chiari I malformation (CM1), understanding of the natural history of these patients is very challenging. In this study, we investigate the natural history of children with CM1 that have syrinx and/or prominence of the central canal on presentation.

Methods All pediatric Chiari I patients who had syrinx and/or prominence of the central canal who underwent MR imaging of the head and spine from 2007 to 2020 were reviewed. Patients were divided into 3 groups (early surgery, delayed surgery, and conservative management). We focused on those patients who did not initially undergo surgery and had at least 1 year of clinical follow-up. We assessed if there were any radiological features that would correlate with need for delayed surgical intervention.

Results Thirty-seven patients met the inclusion criteria. Twenty-one patients were female and 16 were male. The mean age at presentation was 8.7 (5.8 SD). Fourteen (38%) patients had early surgical intervention, with a mean of 2.5 months after initial presentation, 8 (16%) had delayed surgery due to new or progressive neurological symptoms and 46% of patients did not require intervention during follow-up. The length of tonsillar herniation and the position of the obex were associated with the need of surgery in patients who were initially treated conservatively.

Conclusion In pediatric patients with CM1 with syringomyelia and prominence of the central canal, conservative treatment is initially appropriate when symptoms are absent or mild. Close follow-up of patients with CM1 and dilatation of the central canal who have an obex position below the foramen magnum and greater tonsillar herniation is suggested, as these patients show a trend towards clinical deterioration over time and may require earlier surgical intervention.

Keywords Chiari I malformation · Syrinx · Natural history · Neuroimaging · Neurosurgery · MRI · Pediatrics

35 International Experts: Consensus statement

Neurological Sciences (2022) 43:1311–1326
https://doi.org/10.1007/s10072-021-05317-9

ORIGINAL ARTICLE

Diagnosis and treatment of Chiari malformation type 1 in children: the International Consensus Document

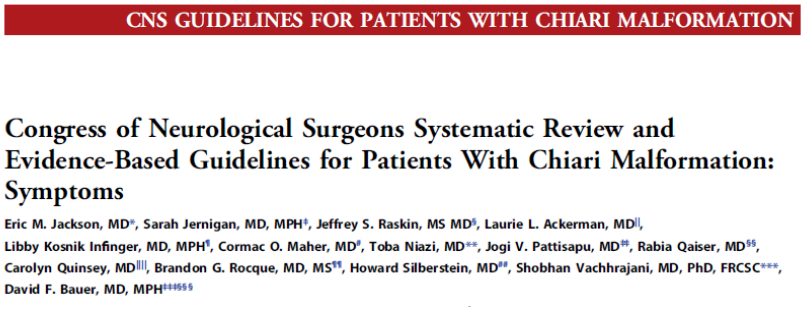
Luca Massimi[1] · Paola Peretta[2] · Alessandra Erbetta[3] · Alessandra Solari[4] · Mariangela Farinotti[4] · Palma Ciaramitaro[5] · Veronica Saletti[6] · Massimo Caldarelli[1] · Alexandre Casagrande Canheu[7] · Carlo Celada[8] · Luisa Chiapparini[3] · Daniela Chieffo[9] · Giuseppe Cinalli[10] · Federico Di Rocco[11] · Marika Furlanetto[12] · Flavio Giordano[13] · George Jallo[14] · Syril James[15] · Paola Lanteri[16] · Christian Lemarchand[17] · Martina Messing-Jünger[18] · Cecilia Parazzini[19] · Giovanna Paternoster[15] · Gianluca Piatelli[20] · Maria. A. Poca[21] · Prab Prabahkar[22] · Federica Ricci[23] · Andrea Righini[19] · Francesco Sala[24] · Juan Sahuquillo[21] · Marcus Stoodley[25] · Giuseppe Talamonti[26] · Dominic Thompson[27] · Fabio Triulzi[28] · Mino Zucchelli[29] · Laura Valentini[12] · International Experts - Jury of the Chiari & Syringomyelia Consensus Conference, "Milan, November 11–13, 2019"

Table 1 Planning for CM1 in children: indications to surgery

1	In *asymptomatic* children with incidentally discovered, isolated CM1 and *no syringomyelia*, surgery is not indicated	Agreement: 94.1%
2	In asymptomatic children with incidentally discovered CM1 and syringomyelia, surgery is indicated in cases of *syrinx larger than 5–8 mm*, and smaller syrinx increasing in size	Agreement: 82.4%
3	In children with *epilepsy* and CM1, surgical treatment of CM1 does not improve the seizure disorder	Agreement: 94.1%
4	In children with CM1 and *cognitive* and/or *behavioral* disorders (such as autism) not able to correctly report their symptoms because of communication impairment, careful clinical and instrumental assessments are mandatory to detect symptoms and signs of cerebellar tonsils ectopia	Agreement: 97.1%
5	In children with CM1 and cognitive and/or behavioral disorders (such as autism) and no clear CM1 symptoms, surgery is not indicated to improve the clinical picture	Agreement: 91.2%
6	Surgical treatment is not decided on *MRI pattern* in asymptomatic children with tonsillar ectopia ≥ 20 mm without syringomyelia	Agreement: 88.2%

27,000 patients (18,200 adults and 8,800 children), more than 40% of them operated on patients.

Guidelines


CNS GUIDELINES FOR PATIENTS WITH CHIARI MALFORMATION

Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for Patients With Chiari Malformation: Symptoms

Eric M. Jackson, MD*, Sarah Jernigan, MD, MPH‡, Jeffrey S. Raskin, MS MD§, Laurie L. Ackerman, MD‖, Libby Kosnik Infinger, MD, MPH¶, Cormac O. Maher, MD#, Toba Niazi, MD**, Jogi V. Pattisapu, MD††, Rabia Qaiser, MD§§, Carolyn Quinsey, MD‖‖, Brandon G. Rocque, MD, MS¶¶, Howard Silberstein, MD##, Shobhan Vachhrajani, MD, PhD, FRCSC***, David F. Bauer, MD, MPH‡‡§§§

RECOMMENDATIONS

2-2. In patients with asymptomatic CIM without syrinx, is prophylactic surgery indicated to prevent future need for surgery? What is the chance of developing symptoms in the future?

Recommendation: Clinicians should not perform prophylactic surgery on patients with asymptomatic CIM without syrinx. There is a small percentage of patients who develop new or worsening symptoms in the future.

Strength of recommendation: Grade C

Level III evidence

Follow-up

No linee guida né raccomandaizoni

➢ Key Points

- ❑ Classification and diagnosis: CM is primarily classified into types 1 and 2, with type 1 being most prevalent in adults. Diagnosis heavily relies on MRI, the gold standard for detecting tonsillar herniation and associated structural abnormalities.
- ❑ Clinical symptoms: CM1 is associated with neurological symptoms such as occipital headaches and dysfunctions of the brainstem and spinal cord. These symptoms are critical for diagnosing CS, the clinical manifestation of CM.
- ❑ Conservative management: Conservative management is recommended for asymptomatic or mildly symptomatic CM1 patients, with regular clinical and radiologic monitoring. Surgical intervention is reserved for patients with severe symptoms.
- ❑ Consensus recommendations: The WFNS Spine Committee developed standardized recommendations for CM diagnosis and management, emphasizing accurate clinical assessment and the use of advanced imaging techniques to guide treatment.
- ❑ Need for further research: The study highlights the need for more high-quality research to strengthen current guidelines and improve the long-term management of CM. The CCOS is suggested for evaluating postoperative outcomes and long-term effectiveness.

LITERATURE REVIEW

Chiari Malformation

Diagnosis, Classifications, Natural History, and Conservative Management. World Federation of Neurosurgical Societies Spine Committee Recommendations

Francesco Costa, MD,[a] Said Ait Benali, MD,[b] Fernando Dantas, MD,[c,d] Francesco Restelli, MD,[a] Elio Mazzapicchi, MD,[a] Saleh Baeesa, MD,[e] Onur Yaman, MD,[f] Salman Sharif, MD,[g] Oscar L. Alves, MD,[h,i] Mehmet Zileli, MD,[j] and Ricardo Botelho, MD[k]

Non c’è menzione su come realizzare la gestione conservativa

Proposta di follow-up

No linee guida né raccomandazioni: considerare la storia naturale

- Per quanto tempo: fino al termine dello sviluppo fisico (20-25 anni…)
- Controlli medici programmati (annualmente)
- RM encefalo-midollo programmate (ogni 2 anni)
- Cine-RM?
- RX dinamica cervicale nelle forme complesse?
- PEM? PESS? ABR? Polisonnografia?

Neurological Sciences (2022) 43:1311–1326
https://doi.org/10.1007/s10072-021-05317-9

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Table 2 Planning for CM1 in children: symptoms and follow-up

1	In *asymptomatic* children with incidentally discovered CM1 and *no syringomyelia*, neurological follow-up and whole neuraxis MRI should be performed until the end of growth, with a schedule based on clinical picture and degree of tonsils herniation.	Agreement: 91.2%
2	*Neuro-pediatric evaluation* is mandatory in all children with CM1 and *headache*	Agreement: 94.1%
3	Neuro-pediatric evaluation is mandatory in all children with CM1 and *symptoms* and/or signs of the brainstem, cerebellar, and/or cervical cord dysfunction to identify co-pathologies	Agreement: 91.2%
4	Neuro-pediatric evaluation is mandatory in all children with CM1 and *cognitive* and/or *behavioral disorders*	Agreement: 91.2%
5	Somatosensory evoked potentials (*SEPs*) may be used in CM1 children only in association with other tools (both clinical and instrumental)	Agreement: 90.9%
6	Auditory evoked potentials (*BAEPs*) may be used in CM1 children only in association with other tools (both clinical and instrumental)	Agreement: 87.9%
7	Motor evoked potentials (*MEPs*) may be used in CM1 children only in association with other tools (both clinical and instrumental)	Agreement: 84.8%
8	Polysomnography is indicated in very young children (< 6 years), in case of suspected apneas, and in case of severe cerebellar tonsils descent	Agreement: 88.2%

Guidelines

Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for Patients With Chiari Malformation: Diagnosis

David F. Bauer, MD, MPH[*§], Toba Niazi, MD[§], Rabia Qaiser, MD[‖], Libby Kosnik Infinger, MD, MPH[¶], Shobhan Vachhrajani, MD, PhD, FRCSC[#], Laurie L. Ackerman, MD[**], Eric M. Jackson, MD[‡‡], Sarah Jernigan, MD, MPH[§§], Cormac O. Maher, MD[‖‖], Jogi V. Pattisapu, MD[¶¶], Carolyn Quinsey, MD[##], Jeffrey S. Raskin, MS MD[***], Brandon G. Rocque, MD, MS[†††], Howard Silberstein, MD[§§§]

RECOMMENDATIONS

1-2. In patients with CIM, are advanced imaging modalities such as Cine MRI helpful to predict benefit from surgical decompression?

Recommendation: In patients with CIM, advanced imaging modalities such as Cine MRI may or may not predict benefit from surgical decompression.

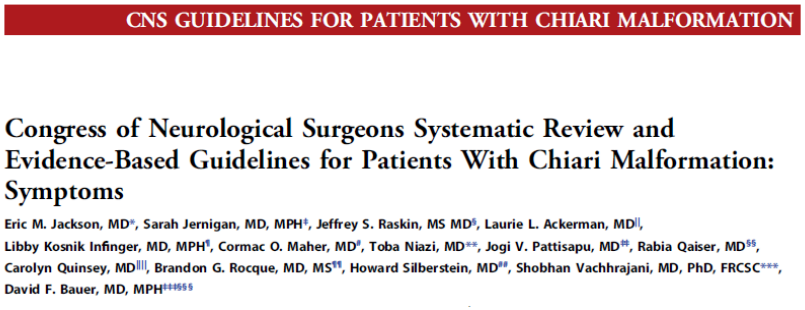
1-3. In patients with CIM, should flexion and extension radiographs be routinely performed to evaluate for cervical instability? Should preoperative evaluation include any specific evaluation for cranial cervical instability or ventral compression (clivoaxial angle, pB-C2, etc)? 3

Recommendation: In patients with CIM, measurement of the clivoaxial angle, pB-C2, or C- C2 sagittal vertebral alignment (C-C2SVA) may predict future craniocervical instability and the need for surgical stabilization.

Strength of recommendation: Grade C

Level III evidence

Guidelines



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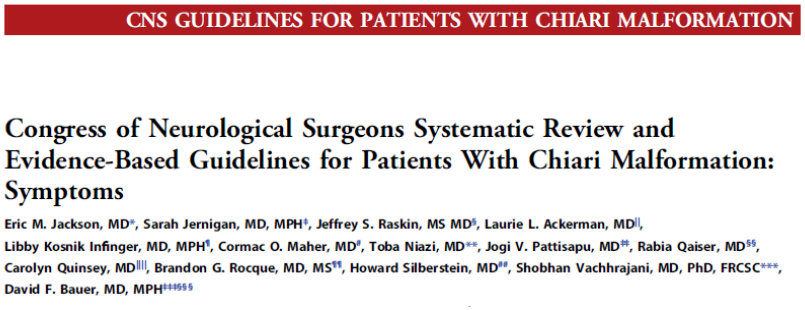
RECOMMENDATIONS

2-4. In patients with CIM, should sleep or swallow studies be routinely performed to evaluate for sleep apnea or dysphagia?

Recommendation: There is insufficient evidence to support routine sleep and swallow studies in patients with CIM without sleep or swallow symptoms.

Strength of recommendation: Grade insufficient…

Guidelines



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RECOMMENDATIONS

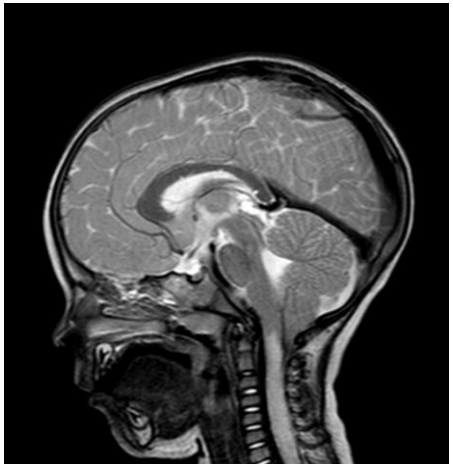
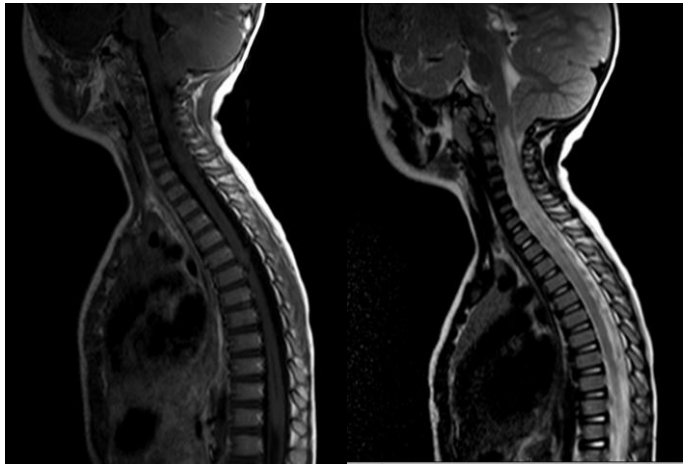
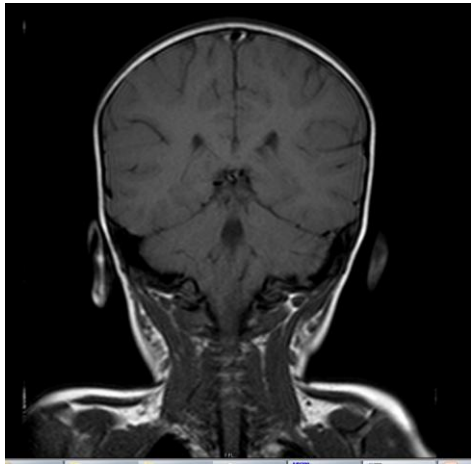
2-5. In patients with CIM, should siblings or first-degree relatives be screened for CIM?

Recommendation: Clinicians should not routinely screen asymptomatic siblings or first-degree relatives of patients with CIM.

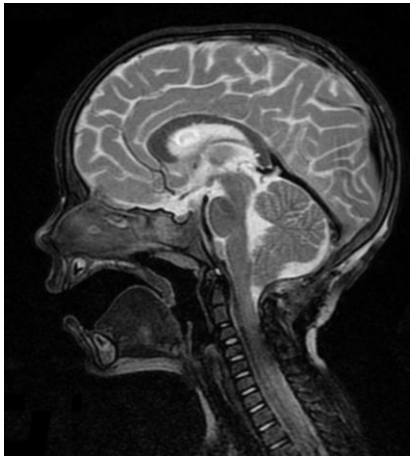
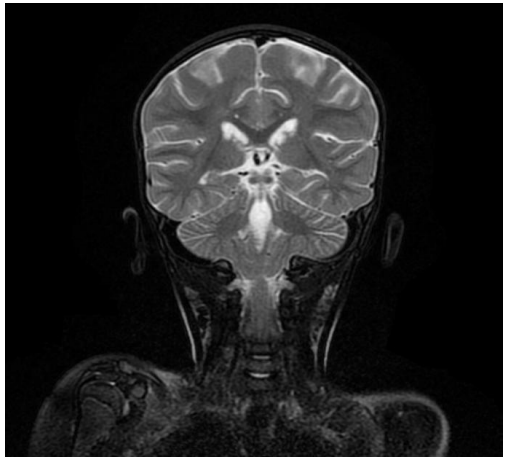
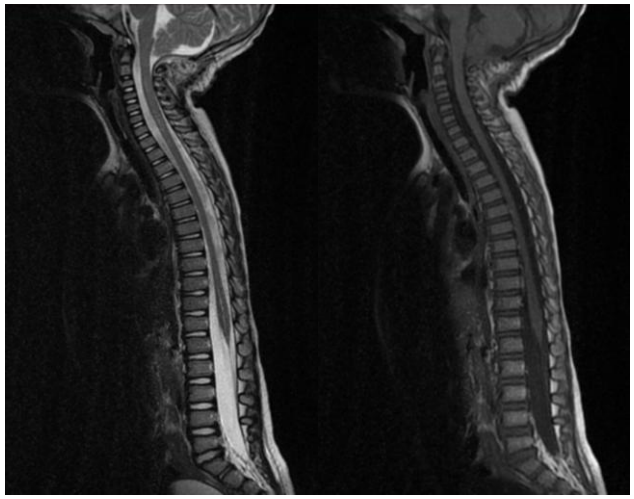
Strength of recommendation: Grade C

Level III evidence

Restrizioni

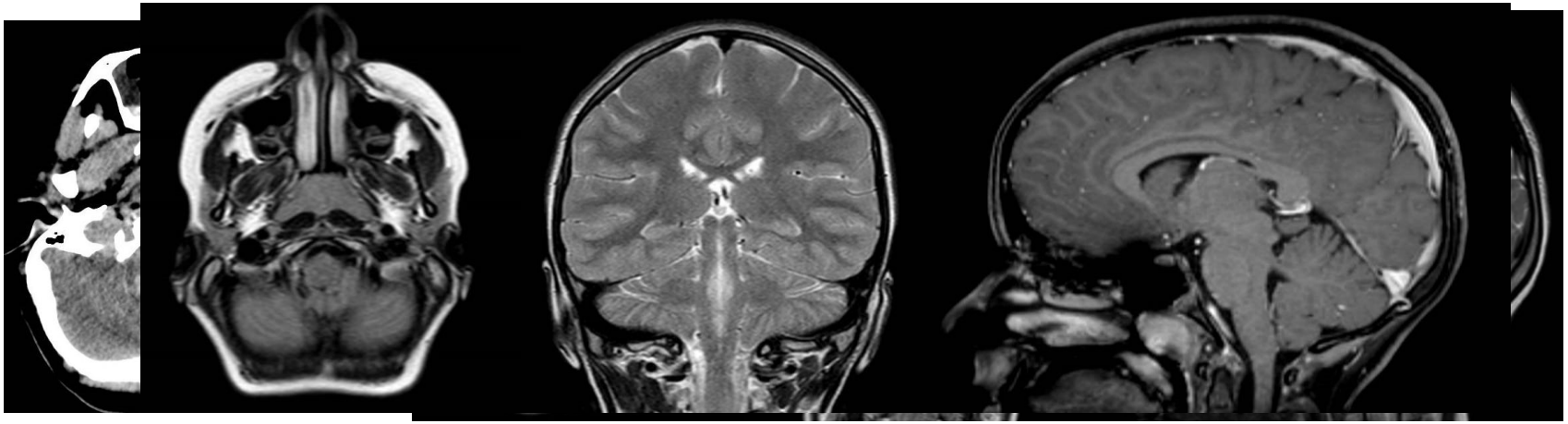
- M, 4 anni: tetraparesi improvvisa e arresto respiratorio dopo trauma cervicale lieve. Decompressione neurochirurgica in urgenza. Recupero completo dopo 3 mesi

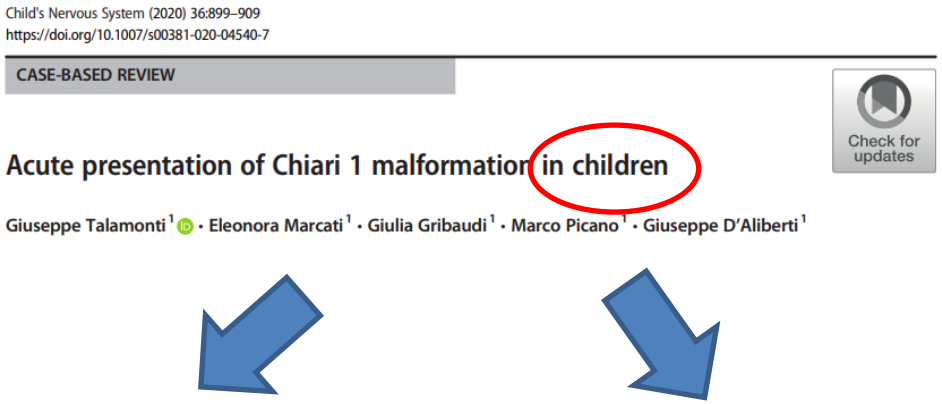
Esordio acuto

- 41 pazienti in 30 anni (~ 1% casistiche ampie), rischio stimato nella popolazione: 0,02%
- Età media: 19,1 anni (range: 1-71 anni)
- Trauma noto nel 41,5% dei casi
- Deficit motorio (36,5%), disturbi respiratori (29%), paralisi dei nervi cranici (17%), arresto cardiaco (14,5%), decesso (12%)
- Recupero completo: 73%

Massimi L et al, Neurosurgical Review, 2012



Esordio acuto



Child's Nervous System (2020) 36:899–909
https://doi.org/10.1007/s00381-020-04540-7

CASE-BASED REVIEW

Check for updates

Acute presentation of Chiari 1 malformation in children

Giuseppe Talamonti[1] · Eleonora Marcati[1] · Giulia Gribaudi[1] · Marco Picano[1] · Giuseppe D'Aliberti[1]

Symptoms other than cortico-spinal dysfunction

- 32 cases
- Age range: 11 mts-14 yrs
- Trauma: 11/32 (34%)
- Syringomyelia*: 16/32 (50%)
- Surgery: 25/32 (78%)
- Death: 2/32 (6%)
- Complete recovery: 21/30 (70%)

Symptoms of cortico-spinal dysfunction

- 17 cases
- Age range: 4 mts-17 yrs
- Trauma: 8/17 (47%)
- Syringomyelia*: 9/17 (53%)
- Surgery: 15/17 (88%)
- Death: 0/17
- Complete recovery: 10/17 (58%)

* Syringomyelia or hydromyelia or pre-syrinx state

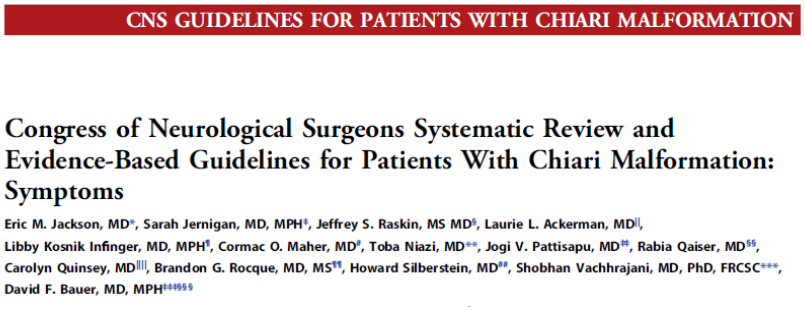
«Evidenze»

TABLE 1. Cohort of 328 sports participants with CM-I dichotomized by sport, level of competition, and number of concussions

Sport	Male†	Female†	Sports Participation*		Concussions	Level of Participation		
			No. of Months	No. of Seasons		Community/Recreation League	Middle School	High School & Above
Archery	0	1	24	8.0	0	1	0	0
Baseball	68	3	1058.5	352.8	0	42	8	10
Basketball	43	42	1190.5	396.8	3	85	24	29
Bowling	6	7	322	107.3	0	4	8	1
Cheerleading	0	21	595	198.3	0	5	3	11
Cross country	4	7	117	39.0	0	0	4	4
Dance	3	40	1841	613.7	0	23	8	7
Dodgeball	2	0	18	6.0	1	1	0	0
Equestrian	1	12	587	195.7	0	6	1	2
Figure skating	1	1	154	51.3	1	1	1	0
Flag football	16	1	77.5	25.8	0	14	1	0
Football	41	1	645	215.0	8	10	6	19
Golf	7	10	240	80.0	0	2	2	11
Gymnastics	3	26	495.5	165.2	0	23	1	2
Hockey	17	10	894.5	298.2	8	9	2	11
Lacrosse	7	1	56	18.7	0	0	3	2
Martial arts	12	5	476	158.7	0	7	1	2
Motocross	3	0	90	30.0	0	2	0	0
Rugby	0	0	0	0.0	0	0	0	0
Skiing	2	3	62.5	20.8	1	59	9	27
Soccer	52	58	2330.4	776.8	9	22	8	12
Softball	4	45	673.5	224.5	0	4	1	0
Swimming	15	24	565	188.3	0	21	6	6
Tee ball	13	7	72	24.0	0	18	0	0
Tennis	9	7	251	83.7	0	4	3	8
Track	9	12	195	65.0	0	2	6	8
Ultimate Frisbee	2	0	1	0.3	1	1	0	0
Volleyball	3	48	641.5	213.8	0	9	18	22
Weight lifting	1	1	42	14.0	0	0	0	0
Wrestling	11	1	207	69.0	1	3	2	6
Totals	355	394	13,922.4	4640.7	33	378	126	200

* Values represent cumulative participation.
† Some participants played multiple sports.

Guidelines



CNS GUIDELINES FOR PATIENTS WITH CHIARI MALFORMATION

Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for Patients With Chiari Malformation: Symptoms

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RECOMMENDATIONS

2-3. In patients with asymptomatic CIM without syrinx, should the patient have any activity restrictions to prevent future harm?

Recommendation: Clinicians should not recommend activity restrictions for patients with asymptomatic CIM without syrinx, as there is no evidence of future harm prevention.

Strength of recommendation: Grade C

Level III evidence

Grazie!

«Er grosso Colosseo tutto de marmo, cor bianco che risplenne ar sol de Roma.
'Na scalinata che te toje er fiato, pé la fatica e pure pé l'incanto.
Con tutti l'archi in ordine preciso, che te sembra d'arrivà in paradiso.
Le gallerie che corrono d'intorno te mostrano il cielo lì ner fonno.
Immensi tutti e due i bianchi cavalli, co' l'omini ar fianco tosti e belli…»

F. Ricci