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Postural control in Chiari I malformation: protocol for a paediatric prospective, observational cohort – potential role of posturography for surgical indication

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ABSTRACT

Introduction Chiari I malformation (CM1) is an anatomical abnormality characterised by the cerebellar tonsils descending at least 5 mm below the foramen magnum. CM1 causes obstruction of cerebrospinal fluid (CSF) circulation as well as direct compression on the brainstem, thus causing typical consequences (syringomyelia), and typical clinical features (characteristic headaches and neurological impairment). Surgery is the only available treatment, indicated when symptomatology is present. However, sometimes patients have atypical complaints, which are often suggestive of otolaryngological (ears, nose and throat, ENT) involvement. This may be difficult for a neurosurgeon to explain. Our study aims to investigate the relationship between one of these atypical symptoms, for example, postural instability, in a paediatric population using a Computerised Dynamic Posturography (Equitest, NeuroCom, Clackamas, OR). To our knowledge, there are no previously published studies carried out on children with CM1, using dynamic posturography.

Methods and analysis Forty-five children aged 6–18 years old presenting with radiologically confirmed CM1 and presenting ENT clinical complaints will be included in the study for a duration of 3 years. As primary endpoint, posturographic results will be described in the population study. Second, posturographic results will be compared between patients with and without indication for surgery. Finally, preoperative and postoperative posturographic results, as well as CSF circulation quality at foramen magnum level, syringomyelia, sleep apnoea syndrome, scoliosis and behaviour will be compared in the operated patient group.

Ethics and dissemination This protocol received ethical approval from the Clinical Research Delegation of Nancy University Hospital, in accordance with the National Commission on Informatics and Liberties (Commission Nationale de l'Informatique et des Libertés) (protocol number 2019PI256-107). Our data treatment was in accordance with the Methodology of reference Methodology Reference-004 specification for data policy. The study findings will be disseminated via peer-reviewed publications and conference presentations, especially to the Neurosphynx's rare disease healthcare network.

STRENGTHS AND LIMITATIONS OF THIS STUDY

⇒ This is the first trial which evaluates postural control strategies in a paediatric population affected by Chiari I malformation using computerised dynamic posturography.
⇒ Quantitative and qualitative data inherent to postural ability will be observed and collected, in order to ensure reproducibility of the research.
⇒ This protocol could represent the starting point for a wider, multicentre study.
⇒ This trial is a multidisciplinary approach to Chiari I malformation, combining neurosurgical, ears, nose and throat and biomechanical expertise.
⇒ The main limitation is that posturography can be unreliable in children under 6 year.

Trial registration number NCT04679792; Pre-results.

INTRODUCTION

Chiari I malformation (CM1) is a structural abnormality characterised by cerebellar tonsillar descent of at least 5 mm below the level of the foramen magnum,[1] into the vertebral canal. Once considered to be rare, it is now quite often diagnosed incidentally due to the increasing utilisation and sensitivity of neuroimaging, especially MRI. Although the true prevalence in the general population is difficult to establish, the imaging incidence in children younger than 18 years has been reported from 0.4% to 3.6%.[2 3]

Initially reported in the XII century by Nicholas Tulp in its 'Observationes Medicae',[4] its formal description has been attributed to Hans Chiari in 1896.[5 6] Despite CM1 is actually one of the most studied neurosurgical topics, some aspects concerning this malformation (ie, pathogenesis, evolution and treatment) remain controversial,[2] so that different

studies are in progress with the aim of clarifying the natural history and best management of CM1.[7]

While 14%–21% of people remain asymptomatic,[8] most patients present with a constellation of clinical features which are diverse and which can impair their quality of life. The most typical symptom attributed to CM1 is pain; this paroxysmal symptom is usually localised to the occipital-cervical region and it is associated with a Valsalva manoeuvre such as coughing, laughing or sneezing. Other neurological symptoms or signs are related to brainstem or cranial nerve compression or distortion, and are represented by long tract sensitive-motor deficit, hyperreflexia, Babinski sign, sleep apnoea syndrome and, less commonly, vocal cord paralysis, palatal weakness and dysarthria.[9] If syringomyelia is associated, as in 45%–75% of patients,[10 11] one can also see a typical clinical expression such as distal-to-proximal weakness of the upper limbs with a cape-like suspended sensory loss. Less commonly, cerebellar syndrome with nystagmus, dysarthric speech and ataxia may be present.[9]

Given the fact that the clinical presentation of CM1 is highly variable, and often accompanied by less than clear subjective somatic complaints,[12] in everyday practice it is sometimes difficult to state whether CM1 is really symptomatic or not. Some neuro-otological manifestations (such as nystagmus, dizziness and imbalance) are reported in CM1, but these features have been described as 'atypical',[13] thus creating confusion in interpretation.

Whether the malformation is symptomatic or not is of fundamental importance to determine if the patient is candidate for surgical intervention; surgery is the only possible treatment for this abnormality.

When analysing CM1 from an anatomical point of view, however, it is not unreasonable to think that patients may present with symptoms and signs suggestive of otolaryngological (ears, nose and throat, ENT) involvement.[14] As a matter of fact, it is important to keep in mind that many of the pathways and structures responsible for balance and gait control (ie, medial longitudinal fasciculus, spinocerebellar tracts, vestibulospinal fasciculus, reticulospinal fasciculus, vestibular nuclei and nerves) are located in the brainstem in the craniocervical region, feeding both afferent and efferent collateral fibres to the cerebellum.

Moreover, ataxia is rarely found on neurological standard clinical examination in CM1 patients and its formal characterisation is difficult in the paediatric population.

For these reasons, we feel that detailed clinical evaluation, including objective testing might be advantageous in this population. Literature about instrumental imbalance evaluation in CM1 patients is scarce, although ataxia and dizziness are frequently described.[9 15–18] We decided to evaluate one of these uncommon aspects in children, that is, imbalance, using Computerised Dynamic Posturography (CDP) (Equitest, Neurocom, Clackamas, OR) at our institution (LAPEM laboratory, Brabois University Hospital in Nancy, France). To our knowledge, this is the only study using CDP in children with CM1.

Preliminary search and pilot study selection

Initial research was conducted following four main axes:

- Typical symptoms of CM1 reported in neurosurgical reviews.
- ENT involvement.
- Developmental assessment of equilibrium control in children.
- Dynamic posturography recordings in paediatric population.

Systematic review of the literature at the time of study design was performed using an Index Medicus and PubMed electronic database, looking for typical symptoms of CM1 reported in neurosurgical articles. ENT studies reporting the interest of neuro-otological evaluation in Chiari I patients were checked, and the following clinical features were individualised: nystagmus, dizziness, hearing loss and gait imbalance.[19–34] Seven studies were selected to express the normal development of postural control in children,[35–41] two of which underline the feasibility and effectiveness of Equitest.[39 42]

Posturographic evaluation in CM1 patients has been used in a study published in 2019 by Palamar *et al*.[43] Static posturography (Tetrax Interactive Postural Balance System (Sunlight Medical, IT) was used in 36 adult subjects, trying to find a correlation between the risk of fall (Fall Index) and degree of tonsillar ectopia, as well as presence of syringomyelia. The authors did not report significant results, but found a more elevated Fall Index in patients with more than 1 cm tonsillar ptosis.

Study objectives

The aim of our research is to analyse the characteristics of balance control in a paediatric population presenting with CM1. This evaluation will be performed utilising CDP (Equitest). To our knowledge, no similar previous studies have been published.

We also wished to compare the CDP sensory organisation test (SOT) patterns between patients with surgical cases of CM1 and non-surgical ones. Surgical indication, determined on the basis of actual neurosurgical criteria, will not be influenced by the results of this study.

Our study will also compare preoperative vs 3 months postoperative assessments with regard to (1) posturographic results, (2) imaging results, (3) symptomatology, (4) sleep quality, (5) spinal balance (eg, scoliosis) and (6) behaviour in the group of operated patients.

METHODS AND ANALYSIS

Protocol design

This is a non-blinded, non-interventional, monocentric, multidisciplinary and prospective, observational, longitudinal clinical study. The ongoing results of the study will not influence surgical decisions.

Patients and public involvement

No patient was involved in the development of this study.

Population study

A total of 45 patients will be enrolled in the study for a duration of 3 years.

Inclusion criteria are:

- Children aged from 6 to 18 years with radiologically confirmed CM1.
- Children presenting clinical features suggesting ENT involvement (dizziness, nystagmus, gait impairment, motion sickness, malaises and atypical migraines which cannot be directly attributed to the CM1).
- Children whose parents/guardians agreed to participation in the study, after being supplied detailed oral and written information.

Diagnosis of CM1 was made by the presence of a caudal displacement of cerebellar tonsils of at least 5 mm under the foramen magnum (more precisely, under McRae's line, a radiographic line drawn on a midsagittal section of MRI that connects the anterior and posterior margins of the foramen magnum). This is traditionally used as a reference to determine foramen magnum level[1] and is associated with problems of cerebrospinal fluid circulation at the foramen magnum level, and a possible brainstem compression.

Non-inclusion criteria are:

- Chiari malformation secondary to other complex pathology (eg, craniostenosis, severe craniocervical malformation, intracranial hypertension, posterior fossa tumour).
- Inability to stand on the CDP platform, due to cerebral palsy, severe behavioural troubles, severe visual impairment or associated orthopaedic pathologies.
- Pre-existing vestibular pathology.
- Refusal of parent to allow use of the indexed personal data.

Study setting

Newly diagnosed CM1 patients recruited at Nancy University Regional Hospital (France) initially undergo neurosurgical evaluation to perform a complete clinical evaluation and precise anamnesis that will be recorded in a survey, ensuring that no valuable information is missed.

Afterward, patients are systematically adressed to perform these two fundamental examinations:

- Medullary MRI, to check for syringomyelia.
- Polysomnographic recording, to look for sleep apnoea syndrome.

Patients who present clinical pictures such as dizziness, nystagmus, gait impairment, disabling motion sickness, malaise and atypical migraines which cannot be attributed to their CM1 will be referred for ENT assessment, including neuro-otological examination and CDP.

Group definition

Patients will be allocated into two groups:

- Patients who will benefit from surgical intervention (followed up for several years after the intervention).
- Patients in whom there is no indication for surgery (followed until adulthood to check for CM1 modification over time).

Choice of comparators

Partition of the patients into the two groups will be made following standard neurosurgical selection criteria for surgery. The decision for surgical intervention is supported by the following examinations: (1) anamnestic and clinical elements, (2) cerebral and medullary MRI and (3) polysomnography. A decision making flow chart is illustrated in figure 1.

The criteria which lead the neurosurgeon to decide on surgical intervention are represented by the presence of at least one of the following aspects:

- Characteristic symptomatology (most of all, exertional headaches, usually occipitocervical, but also of frontal location; presence of symptoms of brainstem compression).
- Syringomyelia.
- Central sleep apnoea.

Investigations

ENT assessment will be carried out in both surgical and non-surgical groups presenting with symptoms requiring specialist examination, as mentioned above. It will consist of a neuro-otological examination and clinical vestibular assessment, if deemed necessary.

The aims of the neuro-otological examination are to detect and differentiate cerebellar and vestibular signs, identify segmental or axial deviations, and rule out confounding associated factors.

A cerebellar syndrome is recognised by the finger to nose test (dysmetria), alternating hand movements (dysdiadochokinesis) and by the increase of the polygon in the Fukuda stepping test (cerebellar ataxia).

With regard to a vestibular syndrome, the consequences of a vestibular lesion can be appreciated by evaluating the vestibulo-ocular pathway (evidenced by the presence of nystagmus), the vestibulo-spinal pathway (which can produce instability), the vegetative pathway (linked to nausea and vomiting) and the perceptual (vestibulocortical) pathway (which can produce true vertigo). Videonystagmography, along with normal caloric vestibular assessment, is very helpful at suggesting brainstem pathology and ruling out peripheral vestibular involvement.[21]

Patients are also evaluated for the presence of nystagmus, which is usually due to peripheral or central vestibular impairment.

Other evaluated factors included vergence insufficiency, refraction disorders, or other visual correction. It is sometimes necessary to ask for a complementary ophthalmological or orthoptic assessment.

Vertigo or dizziness can have multiple causes in the same patient and will be accurately assessed. The diagnosis of the type of vertigo relies mainly on the history taken from the patient and the clinical examination. Any

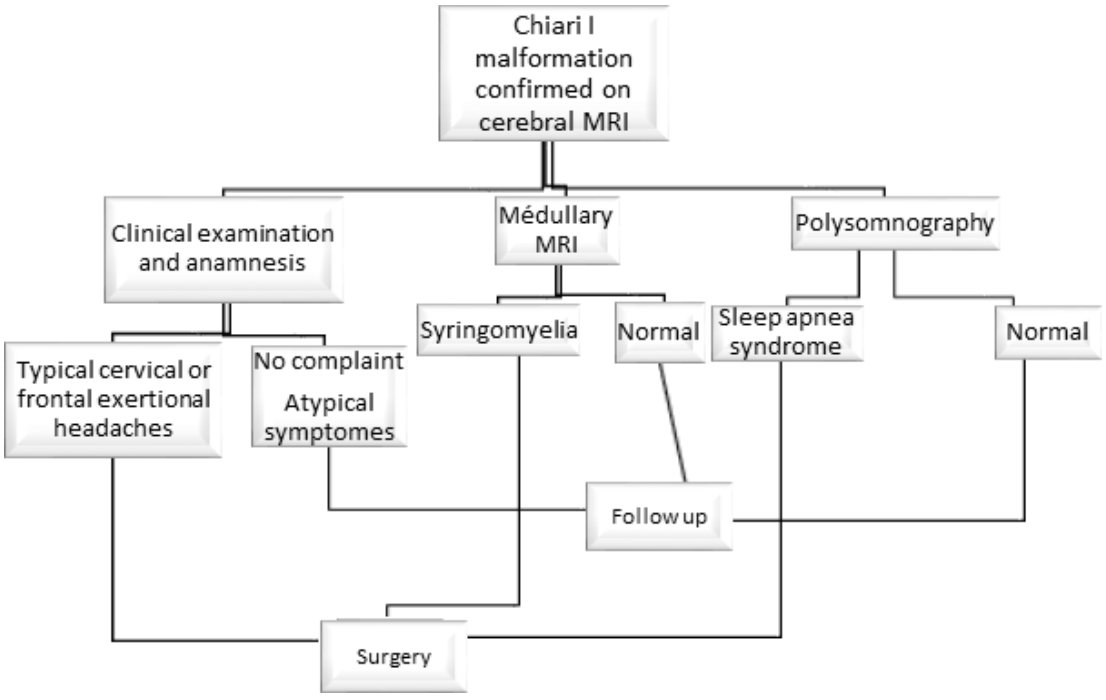

Figure 1 Decision making flow chart for management of CM1 patients. CM1, Chiari I malformation.

history of head trauma and falls, headache, drug intake and motion sickness susceptibility is noted. If spells of true vertigo are occurring, they need to be precisely described (starting date, number, frequency, duration, intensity) in order to determine an eventual evolutionary pattern. The following triggers are sought: head versus trunk movement, movement of the head in space, quick standing, stressing situations (cardiovascular origin). So-called 'false vertigo' should be eliminated.[44] Vertigo or unsteadiness can be associated with sensorineural and conductive hearing loss. Otitis media and previous ear surgery are noted. Complaints of tinnitus should also be addressed.

Otoscopic examination will also be carried out, with tympanometry and acoustic reflex test recordings (Interacoustics, Middelfart, Denmark) and determining hearing thresholds (pure-tone air and bone-conduction thresholds) in tone audiometry (from 250 Hz to 8000 Hz) and intelligibility in speech tests (Interacoustics).

After that, CDP will be carried out to determine balance control performances.

CDP (Equitest, NeuroCom, Clackamas, OR) assesses global balance performance and relative weight of sensory information (visual, vestibular and somatosensory) involved in balance control. The Equitest balance system consists of a dual platform with two footplates connected by a pin join. The footoplates are supported by five force transducers. The computer calculates the centre of foot pressure and the vertical component of the centre of gravity (CoG), using the subject's height entered by the operator. When a subject stands with ankles centred over the stripe on the dual platform, with feet an equal distance laterally from the centre line, he is in the a position called 'electrical zero position', which serves as a reference point for the calculation of sway angles.

The SOT consists of three 20 s trials under six different sensory conditions in which the surface and/or visual surround (ie, sensory inputs) are systematically manipulated (so-called 'sway referencing') (table 1).[43–46]

Examination in eyes closed situations (conditions 2 and 5) may be made more complex by 30° rhythmic flexoextension movements of the head, to better evaluate the somatosensory component of vestibular function and cervical muscles (see below). This might be of particular

Table 1 Computerised dynamic posturography

Postural control test		
Name	Situation	Sensory consequences
Condition 1	Fixed support, eyes open	–
Condition 2	Fixed support, eyes closed	Vision absent
Condition 3	Fixed support, SR surround	Altered vision
Condition 4	SR support, eyes open	Altered proprioception
Condition 5	SR support, eyes closed	Vision absent, altered proprioception
Condition 6	SR support SR surround	Altered vision and proprioception

Sensory organisation test (Equitest, NeuroCom, Clackamas, OR). Determination of the six conditions[42 46 57]
SR, sway-referenced.

Table 2 Computerised dynamic posturography

Name	Equation	Significance
Composite score	(C1+C2+3 (C3+C4+C5+C6))/14	Evaluate global balance performance. A low score represents poor postural control
Somatosensory ratio	C2/C1	Ability to use somatosensory input to maintain balance (even when visual cues are removed). A low score suggests poor use of somatosensory references
Visual ratio	C4/C1	Ability to use visual input to maintain balance (even when somatosensory cues are altered). A low score suggests poor use of visual references
Vestibular ratio	C5/C1	Ability to use vestibular input system to maintain balance (even when visual cues are removed and somatosensory cues are altered). a low score suggests poor use of vestibular cues or that vestibular information is unavailable
Visual preference ratio	C3+C6/C2+C5	Degree to which patient relies on visual information to maintain balance (correct/incorrect information). A low score suggests reliance on visual cues even when they are inaccurate

Sensory organisation test (Equitest, NeuroCom, Clackamas, OR). Significance of composite score and sensory ratios.

interest in patients with Chiari malformation. To protect against falls, patients wear a safety harness connected to the ceiling and an operator stands within reaching distance. An equilibrium score (ES) is calculated by comparing the subject's anterior–posterior sway during each 20 s SOT trial to the maximal theoretical sway limits of stability, which is based on the individual's height and size of the base of support. It represents an angle (8.0 anteriorly and 4.5 posteriorly) at which the subject can lean in any direction before the CoG would move beyond a point that allows him/her to remain upright (ie, point of falling). The following formula is used to calculate the ES:

$$\text{Equilibrium} = 12.5^\circ - (\theta\text{max} - \theta\text{min}) \,/\, 12.5^\circ \times 100$$

where θmax indicates the greatest anteroposterior CoG sway angle, θmin indicates the lowest antero-posterior CoG sway angle. Lower sways lead to a higher ES, indicating a better balance control performance (a score of 100 represents no sway, while 0 indicates sway that exceeds the limit of stability, resulting in a fall). Table 2 shows Composite Equilibrium Score (CES) and sensory ratios calculation's method.

Posturography is also useful in evaluation of the patient with multiple pathologies affecting several components of the sensorimotor chain (inner ear, vision and somesthetic).[47]

An example of CDP preoperative and postoperative results is illustrated in the figure 2.

Association between Chiari malformation and scoliosis is well known; scoliosis can be a factor in balance impairment[48] and altered oculomotor functions.[49] Accordingly, these factors will also be taken into account in the assessment.

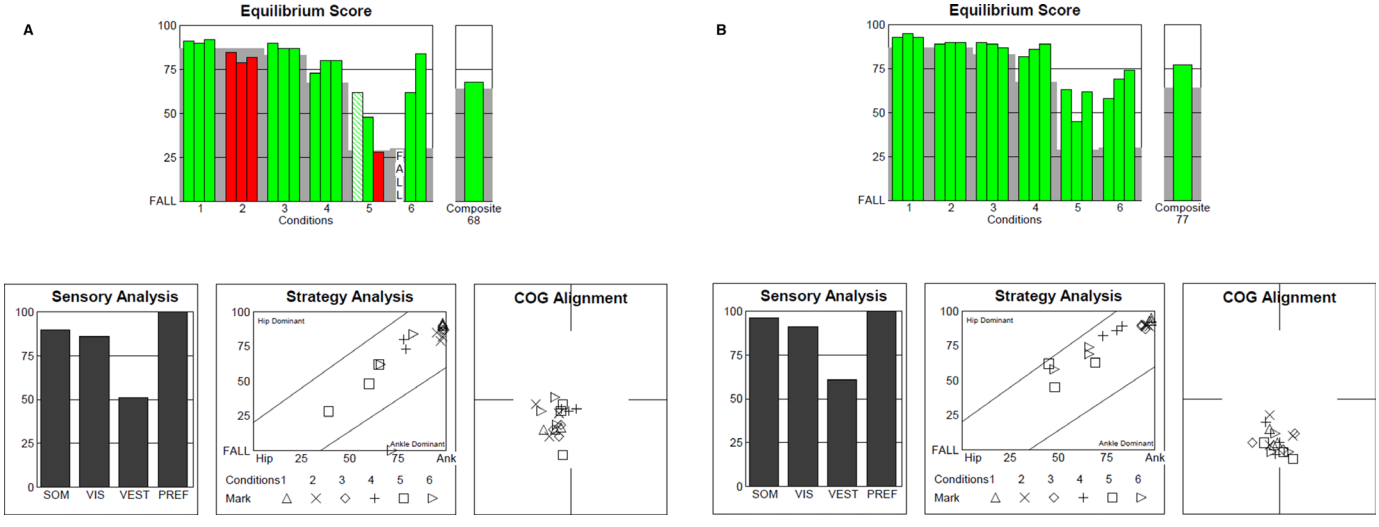

Figure 2 Posturographic results (Equitest, NeuroCom, Clackamas, or) in one of operated patients. Sensory Organisation Test: the composite equilibrium score increased from 68 before surgery (figure 2A) to 77 after surgery (figure 2B). Somatosensory (SOM), visual (VIS) and vestibular (VEST) ratios increased, respectively, from 90 to 96, 85 to 91 and 51 to 60 before to after the surgery.

Outcomes

Primary outcome

The CES will be our primary endpoint, along with the reported visual, somesthetic and vestibular components.

We also will carry out the following evaluations:

- A Head Shake Sensory Organisation Test (details of this assessment are outlined in the literature[50–53] consisting during 20 s of repetitive forward and backward flexions of the head, these head tilts provocative condition of 30° in the pitch plane (cervical flexion/extension) at 0.33 Hz stimulating the cervical proprioception and muscles, as well as the two inner ears. This sensory stimulation was tested in eyes closed condition, both stable (during condition 2) and sway-referenced platform (during condition 5). We feel it important to assess these parameters as these patients may present with cervical pain and muscle impairment.
- Lateral displacements of the CoG, used to quantify the postural sway in the mediallaterally plane.

We also wish to evaluate postural strategies that will be adopted. As outlined in the literature, there are two strategies; a bottom-up regulation model, where the body is oscillating like an inverted pendulum ('ankle strategy'), and a top down strategy (a pattern favouring visual preference involved ('hip strategy'). According to this model, postural control in the sagittal plane is by default exerted around the ankle joint and then (if the postural challenge increases) by the hip joint[54–56]

Secondary outcome

The secondary outcomes are the following.

- CDP results comparison between surgical and not surgical patients.
- Efficacy of surgical intervention to restore cerebrospinal fluid circulation at foramen magnum level.

A single observer (IS) will be in charge of assessing the radiological assessments by comparing preoperative and 3 months postoperative MRI, using a three-point scale: 0=any modification appreciated; 1=improved tonsillar ptosis; 2=resolved tonsillar ptosis with reappearance of a cisterna magna.

- Effectiveness of surgical intervention on a pre-existing syringomyelia.

Improvement will be considered when the cavity's size (maximum anteroposterior diameter) is reduced by at least 30% between preoperative and 3 months postoperative medullary MRI. Stability will be considered in case of cavity's size reduction from 0% to less than 30%, while any degree of increase in size will be considered as aggravation.

- Efficacy of surgical intervention to improve sleep quality.

In case of pre-existing apnoea syndrome before surgery, evolution in the ratio between total and partial respiratory interruption (ie, apnoea/hypopnoea index) between preoperative and 3 months postoperative polysomnography will be considered. A reduction of at least 50% in this ratio will be considered as improvement.

- Effectiveness of surgical intervention to arrest vertebral imbalance (scoliosis).

In case of pre-existing vertebral imbalance, corresponding to a Cobb's angle over 10°, none or minimal Cobb's angle change (<5°) of the pre-existing scoliosis will be considered as a favourable outcome.

- Influence of surgery on patient's behaviour (yes/no).

Based on parents' feelings at least 3 months after surgical intervention, change in patient's behaviour (yes/no) will be assessed at a routine follow-up.

Statistical methods

Primary outcome

Data will be analysed with counts and percentages for qualitative variables and means SD or median and extreme values for quantitative variables (depending on the nature of the distribution).

Secondary outcomes

Intergroup comparison of posturographic results will be performed using Student's t-test or Mann-Whitney U test depending on the nature of the distribution. Comparison between 'before surgery' and 'after surgery results' will be performed using Mc Nemar test or symmetry test categorical variables and Student's test or Wilcoxon's test on paired series for continuous variables.

Data collection method

The data are collected by the principal investigator in an electronic and anonymous database which is password protected and stored on the principal investigator's professional computer.

Data collection will be carried out in accordance with the National Commission on Informatics and Liberties (Commission Nationale de l'Informatique et des Libertés, CNIL) policy.

During the study, the collection of data will be interrupted and the data previously collected will not be used if the person participating in the research objects to the use of their data.

Data monitoring: formal committee and interim analysis

No data monitoring committee has been appointed, due to the absence of recruitment problem and inclusion criteria based on neurosurgical arguments.

An interim analysis of outcomes has not been planned, due to the prompt interpretation of investigation's results.

Timeline

Patients' data are collecting from September 2020 to March 2023.

Adverse effects

No adverse effect of the proposed investigation is conceivable.

ETHICS AND DISSEMINATION

Approvals

This protocol received ethical approval from the Delegation for Clinical Research and Innovation (Délégation à la Recherche Clinique et à l'Innovation—DRCI) of the Nancy University Hospital, in accordance with the National Commission for Computing and Liberties (CNIL) (protocol number 2019PI256-107). The CNIL is an independent administrative authority responsible for ensuring the protection of personal data contained in computer or paper files and processing. It ensures that data processing is at the service of the citizen and that it does not infringe on human identity, human rights, privacy, individual or public freedoms. All procedures performed in this study involving human participants were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

DRCI manages ethical issues and research quality; patients' data are checked and elaborated by Methodology, Data Management and Statistic Unit (Unité de Data Management et Statistique), in accordance with Chapter IX of Law No. 78-17 of 6 January 1978 relating to data processing, files and freedoms. The DRCI, as study Promoter (which is not Investigator of the study), has signed a commitment to comply with the methodology reference–004 specification for data policy. This article was supported by a grant from the French Ministry of Health.

Protocol amendment

If a modification of the analysis plan provided for in this version of the protocol were to be made, this would be justified and validated by the DRCI.

Data management and confidentiality

The first author (IS) is the only person who has knowledge of the identity of the recruited patients and the results of patients' investigations. Data are collected in a protected computerised database only accessible to first author. In accordance with the third paragraph of Article 56 of the Data Protection Act, the presentation of the data processing results may in no case allow the direct or indirect identification of the persons involved in the research.

Data collection will be carried out in accordance with the National Commission on Informatics and Liberties (CNIL) policy.

Patient's families give their oral consent to anonymous treatment of data to the first author (IS). Oral information as well as written details has been supplied to the family in order to provide the principles of the study (aims, data treatment modality). Family were also given the opportunity to refuse by written non consent to data use.

Protocol promotion and funding

This study has been selected as the award winner of 'Neurosphynx 2021 call for project'. NeuroSphinx rare diseases healthcare network coordinates the investigators concerned with rare pelvic and medullary malformations with sphincter damage. The diseases and malformations concerned are those which affect the marrow and the caudal pole. NeuroSphinx links together three Rare Disease Reference Centres, of which C-MAVEM is the specific one who organises the caregiving of syringomyelia, Chiari and vertebral-medullary malformations. It promotes patients-health givers communication and research advances.

Thanks to this award, we have received financial support from Neurosphynx in order to contribute to the publication, and also to maintenance of the posturography.

Sponsor

Direction for Research of the Nancy University Hospital as study Promoter. Vandoeuvre-lès-Nancy, France. Email: d.deoliveira@chru-nancy.fr

NeuroSphinx rare diseases healthcare network call for project as award winner 2021 (IS).

NeuroSphinx-Hôpital Necker, Paris, France. Email: contact@neurosphinx.fr

Dissemination plan

The study's results will be submitted for publication in a peer-reviewed journal, conforming to the definition of the outcome presented in this protocol and will be discussed in specialised congress (European and International Society for Paediatric Neurosurgery, French Society for Pediatric Neurosurgery and European Society for Clinical Evaluation of Balance Disorders meetings). Results will be also presented in Neurosphynx's meetings, as award winner, thus allowing Chiari patients to be informed about.

The DRCI of the Nancy University Hospital is the owner of the data. Nevertheless, the main author and the principal investigator may dispose of data and, in the respect of DRCI policy, for the purposes described above.

The observations done in this study could represent a starting point for other researches, maybe multicentric and could give useful information about how to better manage Chiari's patients presenting with ENT symptoms.

Steering committee

It is composed by the main authors of this report, headed by PP, principal investigator of the study.

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Correction notice This article has been corrected since it was first published. Affiliation number 5 is now correct.

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Collaborators Dr. Art Mallinson (Vancouver, British Columbia, CA) for his helpful advice in the final read-through of the manuscript.

Contributors IS, OK and PP have conceived and designed the study; IS, OK, PP and AJ manage medical aspects of the patients; IS has written the manuscript while OK, PP and TR have participated in writing it; AP realises the posturographic recordings; TR cares about statistical analysis.

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Competing interests None declared.

Patient and public involvement Patients and/or the public were not involved in the design, or conduct, or reporting, or dissemination plans of this research.

Patient consent for publication Not applicable.

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REFERENCES

1 Raybaud C, Jallo GI. Chiari 1 deformity in children: etiopathogenesis and radiologic diagnosis. *Handb Clin Neurol* 2018;155:25–48.
2 Meadows J, Kraut M, Guarnieri M, *et al*. Asymptomatic Chiari type I malformations identified on magnetic resonance imaging. *J Neurosurg* 2000;92:920–6.
3 Chatrath A, Marino A, Taylor D, *et al*. Chiari I malformation in children-the natural history. *Childs Nerv Syst* 2019;35:1793–9.
4 Tulpius N. Observationes medicae. Lugduni Batavorum [Leiden]. Joh. du Vivie 1716; RACS Catalogue No.: C 474.
5 Chiari H. Concerning alterations in the cerebellum resulting from cerebral hydrocephalus. 1891. *Pediatr Neurosci* 1987;13:3–8.
6 Chiari H. Über Veränderungen des Kleinhirns, des pons und Der medulla oblongata in Folge von congenitaler Hydrocephalie des Grosshirns. *Denkschr Kais Akad Wiss* 1896;63:71–116.
7 Piper RJ, Afshari FT, Soon WC, *et al*. Uk Chiari 1 study: protocol for a prospective, observational, multicentre study. *BMJ Open* 2021;11:e043712.
8 Piper RJ, Pike M, Harrington R, *et al*. Chiari malformations: principles of diagnosis and management. *BMJ* 2019;365:l1159.
9 Steinbok P. Clinical features of Chiari I malformations. *Childs Nerv Syst* 2004;20:329–31.
10 Fernández AA, Guerrero AI, Martínez MI, *et al*. Malformations of the craniocervical junction (Chiari type I and syringomyelia: classification, diagnosis and treatment). *BMC Musculoskelet Disord* 2009;10 Suppl 1:S1.
11 Kahn EN, Muraszko KM, Maher CO. Prevalence of Chiari I malformation and syringomyelia. *Neurosurg Clin N Am* 2015;26:501–7.
12 Alexander H, Tsering D, Myseros JS, *et al*. Management of Chiari I malformations: a paradigm in evolution. *Child's Nervous System* 2019;35:1809–26.
13 Novegno F. Clinical diagnosis-part II: what is attributed to Chiari I. *Childs Nerv Syst* 2019;35:1681–93.
14 Saez RJ, Onofrio BM, Yanagihara T. Experience with Arnold-Chiari malformation, 1960 to 1970. *J Neurosurg* 1976;45:416–22.
15 Milhorat TH, Chou MW, Trinidad EM, *et al*. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery* 1999;44:1005–17.
16 Langridge B, Phillips E, Choi D. Chiari malformation type 1: a systematic review of natural history and conservative management. *World Neurosurg* 2017;104:213–9.
17 Jarski P, Zimny M, Linart M, *et al*. Results of the surgical treatment in children with Chiari malformation type I. *Child's Nervous System* 2019;35:1911–4.
18 Hersh DS, Groves ML, Boop FA. Management of Chiari malformations: opinions from different centers-a review. *Childs Nerv Syst* 2019;35:1869–73.
19 Rydell RE, Pulec JL. Arnold-Chiari malformation. neuro-otologic symptoms. *Arch Otolaryngol* 1971;94:8–12.
20 Spooner JW, Baloh RW. Arnold-Chiari malformation: improvement in eye movements after surgical treatment. *Brain* 1981;104:51–60.
21 Longridge NS, Mallinson AI. Arnold-Chiari malformation and the otolaryngologist: place of magnetic resonance imaging and electronystagmography. *Laryngoscope* 1985;95:335–9.
22 Zimmerman CF, Roach ES, Troost BT. See-Saw nystagmus associated with Chiari malformation. *Arch Neurol* 1986;43:299–300.
23 Mossman SS, Bronstein AM, Gresty MA, *et al*. Convergence nystagmus associated with Arnold-Chiari malformation. *Arch Neurol* 1990;47:357–9.
24 Albers FW, Ingels KJ. Otoneurological manifestations in Chiari-I malformation. *J Laryngol Otol* 1993;107:441–3.
25 Ahmmed AU, Mackenzie I, Das VK, *et al*. Audio-vestibular manifestations of Chiari malformation and outcome of surgical decompression: a case report. *J Laryngol Otol* 1996;110:1060–4.
26 Sperling NM, Franco RA, Milhorat TH. Otologic manifestations of Chiari I malformation. *Otol Neurotol* 2001;22:678–81.
27 Naya Gálvez MJ, Fraile Rodrigo JJ, Liesa RF, *et al*. Otorhinolaryngologic manifestations in Chiari malformation. *Am J Otolaryngol* 2002;23:99–104.
28 Simons JP, Ruscetta MN, Chi DH. Sensorineural hearing impairment in children with Chiari I malformation. *Ann Otol Rhinol Laryngol* 2008;117:443–7.
29 Korres S, Balatsouras DG, Zournas C, *et al*. Periodic alternating nystagmus associated with Arnold-Chiari malformation. *J Laryngol Otol* 2001;115:1001–4.
30 Kumar A, Patni AH, Charbel F. The Chiari I malformation and the neurotologist. *Otol Neurotol* 2002;23:727–35.
31 Al-Awami A, Flanders ME, Andermann F, *et al*. Resolution of periodic alternating nystagmus after decompression for Chiari malformation. *Can J Ophthalmol* 2005;40:778–80.
32 Liebenberg WA, Georges H, Demetriades AK, *et al*. Does posterior fossa decompression improve oculomotor and vestibulo-ocular manifestations in Chiari 1 malformation? *Acta Neurochir* 2005;147:1239–40.
33 Unal M, Bagdatoglu C. Arnold-Chiari type I malformation presenting as benign paroxysmal positional vertigo in an adult patient. *J Laryngol Otol* 2007;121:296–8.
34 Guerra Jiménez G, Mazón Gutiérrez Ángel, Marco de Lucas E, *et al*. Audio-vestibular signs and symptoms in Chiari malformation type I. Case series and literature review. *Acta Otorrinolaringol Esp* 2015;66:28–35.
35 Nashner LM, Black FO, Wall C. Adaptation to altered support and visual conditions during stance: patients with vestibular deficits. *J Neurosci* 1982;2:536–44.
36 Forssberg H, Nashner LM. Ontogenetic development of postural control in man: adaptation to altered support and visual conditions during stance. *J Neurosci* 1982;2:545–52.
37 Shumway-Cook A, Woollacott MH. The growth of stability: postural control from a development perspective. *J Mot Behav* 1985;17:131–47.
38 Hsu Y-S, Kuan C-C, Young Y-H. Assessing the development of balance function in children using stabilometry. *Int J Pediatr Otorhinolaryngol* 2009;73:737–40.
39 Hirabayashi S, Iwasaki Y. Developmental perspective of sensory organization on postural control. *Brain Dev* 1995;17:111–3.
40 Barozzi S, Socci M, Soi D, *et al*. Reliability of postural control measures in children and young adolescents. *Eur Arch Otorhinolaryngol* 2014;271:2069–77.
41 Rine RM, Rubish K, Feeney C. Measurement of sensory system effectiveness and maturational changes in postural control in young children. *Pediatr Phys Ther* 1998;10:16???22–22.
42 Sinno S, Dumas G, Mallinson A, *et al*. Changes in the sensory weighting strategies in balance control throughout maturation in children. *J Am Acad Audiol* 2021;32:122–36.
43 Palamar D, Güler H, Hancı M, *et al*. Posturographic examination of body balance in patients with Chiari type I malformation and correlation with the presence of syringomyelia and degree of cerebellar ectopia. *Turk J Phys Med Rehabil* 2019;65:74–9.
44 Perrin P, Mallinson A, Van Nechel C, *et al*. Defining Clinical-Posturographic and Intra-Posturographic Discordances: what do these two concepts mean? *J Int Adv Otol* 2018;14:127–9.

45 Black FO, Paloski WH, Doxey-Gasway DD, *et al*. Vestibular plasticity following orbital spaceflight: recovery from postflight postural instability. *Acta Otolaryngol Suppl* 1995;520 Pt 2:450–4.
46 Nashner LM. Computerized dynamic posturography. In: Jacobson GP, Newman CW, Kartush JM, eds. *Handbook of balance function testing*. London: Thompson Learning, 1997.
47 Kingma H, Gauchard GC, de Waele C, *et al*. Stocktaking on the development of posturography for clinical use. *J Vestib Res* 2011;21:117–25.
48 Haumont T, Gauchard GC, Lascombes P, *et al*. Postural instability in early-stage idiopathic scoliosis in adolescent girls. *Spine* 2011;36:E847–54.
49 Lion A, Haumont T, Gauchard GC, *et al*. Visuo-oculomotor deficiency at early-stage idiopathic scoliosis in adolescent girls. *Spine* 2013;38:238–44.
50 Paloski WH, Wood SJ, Feiveson AH, *et al*. Destabilization of human balance control by static and dynamic head tilts. *Gait Posture* 2006;23:315–23.
51 Honaker JA, Converse CM, Shepard NT. Modified head shake computerized dynamic posturography. *Am J Audiol* 2009;18:108–13.
52 Mishra A, Davis S, Speers R, *et al*. Head shake computerized dynamic posturography in peripheral vestibular lesions. *Am J Audiol* 2009;18:53–9.
53 Honaker JA, Janky KL, Patterson JN, *et al*. Modified head shake sensory organization test: sensitivity and specificity. *Gait Posture* 2016;49:67–72.
54 Nashner LM, McCollum G. The organization of human postural movements: a formal basis and experimental synthesis. *Behav Brain Sci* 1985;8:135–50.
55 Perrin P, Deviterne D, Hugel F, *et al*. Judo, better than dance, develops sensorimotor adaptabilities involved in balance control. *Gait Posture* 2002;15:187–94.
56 Duchene Y, Mornieux G, Petel A, *et al*. The trunk's contribution to postural control under challenging balance conditions. *Gait Posture* 2021;84:102–7.
57 Lion A, Bosser G, Gauchard GC, *et al*. Exercise and dehydration: a possible role of inner ear in balance control disorder. *J Electromyogr Kinesiol* 2010;20:1196–202.