Pulsatile cerebrospinal fluid dynamics in Chiari I malformation syringomyelia: Predictive value in posterior fossa decompression and insights into the syringogenesis

ABSTRACT

Background: Pathophysiological mechanisms underlying the syringomyelia associated with Chiari I malformation (CM-1) are still not completely understood, and reliable predictors of the outcome of posterior fossa decompression (PFD) are lacking accordingly. The reported prospective case-series study aimed to prove the existence of a pulsatile, biphasic systolic–diastolic cerebrospinal fluid (CSF) dynamics inside the syrinx associated with CM-1 and to assess its predictive value of patients' outcome after PFD. Insights into the syringogenesis are also reported.

Methods: Fourteen patients with symptomatic CM-1 syringomyelia underwent a preoperative neuroimaging study protocol involving conventional T1/T2 and cardiac-gated cine phase-contrast magnetic resonance imaging sequences. Peak systolic and diastolic velocities were acquired at four regions of interest (ROIs): syrinx, ventral, and dorsal cervical subarachnoid space and foramen magnum region. Data were reported as mean ± standard deviation. After PFD, the patients underwent a scheduled follow-up lasting 3 years. One-way analysis of variance with Bonferroni Post hoc test of multiple comparisons was performed. P was <0.001.

Results: All symptoms but atrophy and spasticity improved. PFD caused a significant velocity changing of each ROI. Syrinx and premedullary cistern velocities were found to be decreased within the 1st month after PFD (<0.001). A caudad and cephalad CSF jet flow was found inside the syrinx during systole and diastole, respectively.

Conclusion: Syrinx and premedullary cistern velocities are related to an early improvement of symptoms in patients with CM-1 syringomyelia who underwent PFD. The existence of a biphasic pulsatile systolic–diastolic CSF pattern inside the syrinx validates the "transmedullary" theory about the syringogenesis.

Keywords: Cerebrospinal fluid circulation, Chiari I malformation, cine magnetic resonance imaging, posterior fossa decompression, syringomyelia

INTRODUCTION

The prevalence of syringomyelia ranges from 40% to 70% in patients with Chiari I malformation (CM-1), and the coexistence of both these conditions makes the decision-making process about the indication for surgery even more difficult.1,2 The main reasons for this lie in the still debated pathogenesis about the syrinx formation, heterogeneous symptomatology,
multiplicity of treatment options, and especially dramatically lack of reliable predictors of outcome on the basis of which to select those patients who will most likely benefit from surgery[3,4]. In fact, most of the previous reported predictors include anamnestic factors focused on the type and duration of symptoms, neurologic or quality of life scales, or also static magnetic resonance imaging (MRI) measurements.[5,6] Nevertheless, Chiari I-related syringomyelia is a very complex pathology where most of the theories about its formation, persistence, and progression involve dynamic factors of the cerebrospinal fluid (CSF) circulation, volume changes of the spinal canal and its contents, cardiac-related pulsations, abdominal pressure, along with a reduced volume of the posterior fossa.[7,8] On the other hand, the constant evolution of the MRI techniques and the implementation of CSF velocity-coded sequences have led to the comprehension of the fundamental features of the motion of the CSF along the neuraxis. They also provided a noninvasive qualitative–quantitative evaluation of the morphofunctional characteristics of the syrinx during the cardiac cycle.[9,10] Cine phase-contrast has been among the first motion-sensitive MRI sequence proved to have a reliable role in the prediction of patients’ outcomes for several pathologies.[11,12,13,14,15] Nevertheless, its predictive value in the evaluation of patients affected by syringomyelia has been largely underestimated.

The aim of this prospective case-series study is first to test the hypothesis of the existence of a biphasic, systolic–diastolic CSF pulsatile pattern inside the syrinx, as a result of a communication between the syrinx and the subarachnoid space and second to assess its predictive value of patients’ outcome after posterior fossa decompression (PFD). Some insights into the pathophysiology underlying the syrinx formation and the theories about the hindbrain-related syringomyelia are also reported.

METHODS

The present study, approved by the Internal Institutional Review Board, started in January 2015 and finished in December 2019. Eligible criteria were the following: age ranging between 18 and 70 years old, evidence for Chiari I malformation and associated syringomyelia, and scheduled PFD surgery. Patients with previous moderate-to-severe brain injuries, CSF pathologies, previous neurosurgical procedures, chronic obstructive pneumopathy, obstructive sleep apnea syndrome, or thoraco-abdominal masses were excluded from the study. In all the patients, an exhaustive neurological assessment aimed to detect symptoms and signs related to the syrinx and CM-1 was evaluated preoperatively. All the patients underwent T1-T2-weighted spin-echo images of the brain and spine. The preoperative imaging workup was also involved by default retrospective cardiac-gated cine phase-contrast MRI (Magnetom 1.5T MRI Scanner, Siemens Healthineers, Erlangen, Germany, in all of them). Acquisition datasets were as follows: repetition time: 50 ms, echo time: 10 ms, flip angle: 15°, two signals acquisition, 256 × 192 matrix, thickness: 3 mm, velocity encoding: 5 cm/s, and increased to 10 cm/s in case of aliasing. A retrospective cardiac gating technique was used. Cardiac triggering was achieved with finger photoplethysmography. Radio-frequency pulses ranged between 12 and 24 depending on the heart rate, which, in turn, ranged between 50 and 100 bpm. CSF flow data were acquired in mid-sagittal and axial planes.

Cardiac cycle was fractioned in 14 different phases and the highest velocity (peak) was calculated both in systole and diastole for each voxel within each region of interest (ROI) at each of these phases. Acquired images were reviewed in a video loop mode studying the relationships of the cardiac cycle on the motion of the CSF within the subarachnoid space.

Four ROIs were studied in detail, namely the syrinx, cervical ventral, and dorsal subarachnoid space and foramen magnum. The measurements were executed with a Dicom imaging workstation (Osirix DICOM Viewer®, Pixmeo, Bernex, Switzerland).

Craniocaudal length and maximum anterior–posterior (AP) diameter of the syrinx were calculated on the sagittal and axial T2-weighted MRI, respectively. Maximum AP diameter of the ventral and dorsal cervical subarachnoid space (C-SAS) was measured on sagittal and axial T2 MRI at the level of the C4–C5 intervertebral disk. AP diameter of premedullary cistern and cisterna magna was measured at the level of the McRae line. Peak systolic and diastolic velocities were calculated at each ROI and reported as mean ± standard deviation.

All the patients underwent a PFD. Surgeries were executed by the same surgeon (RG) and involved in all cases a wide suboccipital craniectomy, C1 laminectomy, or C1 and C2 depending on cases, dura opening, microneurolysis of the arachnoid bends at the level of foramen of Magendie, and duraplasty with a xenogeneic bovine pericardium dural substitute (Tutopatch, SIAD Healthcare S.p.a., Assago, Milano, Italy). The scheduled follow-up involved a postoperative – within 72 h – neurological evaluation, along with the same neuroimaging protocol performed preoperatively. The same clinicoradiological evaluations were executed at the 1st, 2nd, 3rd, and 6th month and 1st, 2nd, and 3rd year after the PFD. Statistical analysis was carried out by means of a commercially available software (SPSS Statistics for Windows, version x. 0, SPSS Inc., Chicago, IL, USA).
One-way analysis of variance (ANOVA) was performed for each parameter. Significant results in the ANOVA model were further investigated with Bonferroni Post hoc test of multiple comparisons. $P$ was set as $\leq 0.001$. The results were reported based on the Strengthening the Reporting of Observational Study in Epidemiology statement.\footnote{40}

RESULTS

Demographic data
Fourteen patients were enrolled. Patients’ ages ranged between 17 and 51 years, with an average age of 30.5 years. The male/female ratio was 0.7.

Symptoms and spinal cord metameric involvement
Data about preoperative symptoms are reported in Graph 1. Headache, dissociated sensory loss, lower cranial nerve dysfunction, and weakness were most frequent. C2–C5 metameres were most frequently involved [Graph 2]. Headache immediately disappeared after surgery, whereas a full recovery from dyesthetic pain, weakness, and dissociated sensory loss was seen in all the treated patients within 6 months. Conversely, atrophy and spasticity were almost unaffected by surgery. The overall trend of symptoms and clinical signs is reported in Graph 3.

Magnetic resonance imaging and cine phase-contrast magnetic resonance imaging data
Overall data about the ROI measurements are reported in Table 1.

Syrinx
All the syrinx parameters decreased after surgery ($<0.001$) [Graphs 4-6]. Bonferroni multiple comparisons test showed that the length and AP diameter decreased at 1st year and 1st month, respectively, whereas systolic and diastolic peak velocity declined at postoperative and 2nd-month follow-up, respectively [Table 2].

Ventral (cervical subarachnoid space)
AP diameter increases immediately after surgery ($<0.001$) [Graph 7]. Peak systolic and diastolic velocities significantly decreased at 3rd-and 6th-month, respectively [Graph 8 and Table 3].

Dorsal (cervical subarachnoid space)
AP diameter increased starting from the 3rd month after surgery [Graph 9]. Here, the systolic velocity diminished very earlier than the diastolic one [Graph 10 and Table 4].

Foramen magnum region
Premedullary cistern showed a significant enlargement 3 months after surgery [Graph 11], whereas cisterna magna appeared effectively increased by surgery [Graph 12].

Illustrative cases
Case no. 1
A 45-year-old female with occasional history of headache exacerbated by cough, underwent a laparoscopic cholecystectomy for a gallstone disease. Two weeks later, she started to suffer from a severe ataxia, dysphagia, and dyesthetic
Case no. 1
A 37-year-old male with a severe spinal deformity was diagnosed with a CM-I and a concomitant C2–C7 syringomyelia [Figure 3a]. The patient reported severe headach and a slow but progressive dissociated sensory loss in the arms. A CM-I was found, with a C2-T1 syrinx [Figure 2a and b]. Preoperative cine MRI clearly showed a caudal hyperintense signal inside the syrinx during systole [Figure 2c]. During diastole, a tenuous hypointense signal was visible inside the syrinx as a consequence of the cephalad CSF flow [Figure 2d]. The patient was operated on by means of a PFD. The postoperative cine MRI was performed 48 h after surgery revealed a significant attenuation of the systolic signal inside the syrinx [Figure 2e]. The same signal was found to be almost disappeared at the 1st-month evaluation [Figure 2f and Video 2].

Case no. 3
A 37-year-old male with a severe spinal deformity was diagnosed with a CM-I and a concomitant C2–C7 syringomyelia [Figure 3a]. The patient reported severe headach and a slow but progressive dissociated sensory loss in the arms, especially during the Valsalva maneuver and cough. Cine MRI revealed a caudal CSF jet flow inside the syrinx during systole [Figure 3b and Video 3]. After PFD, the sagittal extension and the AP diameter of the syrinx collapsed already in the early postoperative evaluation [Figure 3c]. Cine MRI performed at the 2nd month showed a significant reduction of the jet flow inside the syrinx during systole [Figure 3d and Video 4].
showed the disappearance of the systolic signal inside the syrinx [Figure 3d and Video 4]. The patient recovered almost completely from his symptoms within 3 months.

**DISCUSSION**

The natural history of the hindbrain-related syringomyelia is characterized by a stepwise and severe myelopathy causing a progressive neurological deterioration mainly characterized by the occurrence of paralysis, sensory loss, and drug-resistant chronic pain. The already controversial landscape related to the management of syringomyelia associated with Chiari I malformation is furtherly complicated by the lack of reliable predictors of outcome based on which to better select those patients who could benefit from the PFD. In the past two decades, the advent and implementation of the velocity-coded cine phase-contrast MRI sequences in the routine clinical practice have allowed to shift from a static to a dynamic conception of the CSF pathologies at large, dramatically changing the approach to these illnesses. Cine MRI has also led to new insights into the pathophysiology of the CSF circulation. A further valuable strength of this technique lies undoubtedly in its noninvasiveness, which furthermore justifies its wide employment. The employment of the cine-phase contrast MRI in CM-1 patients with syringomyelia is not a novelty. Nevertheless, almost the totality of the reported studies focused on parameters different from the direct measurement of the velocities inside the syrinx. The results of the present study allow to confirm the existence of a systolic–diastolic pulsatile pattern inside the syrinx, which can be measured preoperatively by means of cine phase-contrast MRI, thus achieving quantitative and qualitative paramount information about the syrinx itself. According to the reported data, the preoperative average syrinx velocity was 1.23 ± 0.34 and 0.72 ± 0.35 cm/s for the systolic and diastolic one, respectively. Similar results have been reported by Honey et al. After the PFD, we found a significant decrease of the peak systolic velocity already in the immediate postoperative evaluation. The reduction of diastolic velocity resulted significant starting from the 2nd-month follow-up, these data suggesting a later normalization of this parameter compared with the systolic velocity. The disappearance of the systolic–diastolic pattern inside the syrinx follows mainly the trend of the recovery from ataxia, lower cranial nerve dysfunction, dysesthetic pain, and headache. At the level of the foramen magnum region, average peak systolic velocity of the premedullary cistern also decreased immediately after surgery, allowing to identify this parameter as a positive predictor of outcome. Furthermore, the increase of the AP diameter of the premedullary cistern also decreased immediately after surgery, allowing to identify this parameter as a positive predictor of outcome. Furthermore, the increase of the AP diameter of the premedullary cistern became significant starting from 3 months after surgery, namely much more belatedly than the normalization of the systolic CSF velocity. In contrast, the normalization of the velocities in correspondence of the ventral C-SAS occurred at the 3rd- and 6th-month follow-up indicated a poor correlation between the C-SAS velocities and the recovery from symptoms. Similar data regraded the dorsal C-SAS.

The findings of the present study also lead to important considerations about the main theories explaining the formation and progression of the syringomyelia in patients with CM-1.
Briefly, the seminal “pressure dissociation” or “slosh” theory of syrinx progression by Williams,[15,56-58] later revised by Oldfield et al. and Heiss et al.,[12,13] involved that the presence of a CSF flow obstruction at the craniovertebral junction causes a pressure dissociation between the cranial and spinal subarachnoid spaces by means of a “ball valve mechanism.” CSF would pass the area of obstruction during coughing, sneezing, or Valsalva but would be unable to flow back once the spinal pressure had normalized. In the Williams theory, the periphery of the syrinx tends to be more ischemic than the center.

Table 1: Overall Data about the ROIs Measurements

| ROI and Parameter | Pre-op. | Post-op. | 1st mo | 2nd mo | 3rd mo | 6th mo | 1st Y | 2nd Y | 3rd Y |
|-------------------|---------|----------|--------|--------|--------|--------|-------|-------|-------|
| SYRINX            |         |          |        |        |        |        |       |       |       |
| Length (cm)       |         |          |        |        |        |        |       |       |       |
| Average           | 14.01   | 13.06    | 10.11  | 8.32   | 9.41   | 8.19   | 8.12  | 8.21  | 5.27  |
| SD                | 6.22    | 7.39     | 6.29   | 4.94   | 4.98   | 3.13   | 2.86  | 2.61  | 1.83  |
| AP diameter (mm)  |         |          |        |        |        |        |       |       |       |
| Average           | 12.85   | 11.60    | 7.98   | 6.80   | 6.53   | 5.92   | 5.18  | 4.76  | 4.62  |
| SD                | 2.05    | 2.70     | 1.75   | 1.79   | 2.90   | 1.41   | 2.05  | 1.74  | 1.87  |
| Sys Vel. (cm/sec.)|         |          |        |        |        |        |       |       |       |
| Average           | 1.23    | 0.67     | 0.30   | 0.32   | 0.23   | 0.18   | ND    | ND    | ND    |
| SD                | 0.34    | 0.28     | 0.18   | 0.09   | 0.12   | 0.04   | ND    | ND    | ND    |
| Dias Vel. (cm/sec.)|       |          |        |        |        |        |       |       |       |
| Average           | 0.72    | 0.58     | 0.45   | 0.36   | 0.32   | 0.18   | ND    | ND    | ND    |
| SD                | 0.35    | 0.15     | 0.10   | 0.09   | 0.11   | 0.04   | ND    | ND    | ND    |
| VENTRAL C-SAS     |         |          |        |        |        |        |       |       |       |
| AP diameter (mm)  |         |          |        |        |        |        |       |       |       |
| Average           | 1.23    | 2.78     | 2.94   | 3.02   | 3.62   | 4.24   | 4.23  | 4.52  | 4.48  |
| SD                | 0.72    | 0.74     | 1.04   | 0.87   | 0.63   | 0.43   | 0.42  | 0.67  | 0.46  |
| Sys Vel. (cm/sec.)|         |          |        |        |        |        |       |       |       |
| Average           | 5.63    | 5.47     | 5.28   | 4.48   | 3.38   | 3.73   | 2.68  | 2.16  | 1.94  |
| SD                | 1.20    | 1.45     | 2.32   | 1.50   | 1.81   | 2.09   | 1.27  | 0.57  | 0.78  |
| Dias Vel. (cm/sec.)|       |          |        |        |        |        |       |       |       |
| Average           | 3.48    | 3.45     | 3.35   | 3.26   | 2.24   | 1.98   | 1.67  | 1.65  | 1.28  |
| SD                | 1.18    | 1.34     | 1.46   | 1.47   | 1.08   | 0.84   | 0.83  | 0.93  | 0.78  |
| DORSAL C-SAS      |         |          |        |        |        |        |       |       |       |
| AP diameter (mm)  |         |          |        |        |        |        |       |       |       |
| Average           | 1.36    | 1.52     | 1.62   | 1.96   | 2.72   | 4.38   | 4.67  | 4.56  | 4.27  |
| SD                | 0.60    | 0.77     | 0.97   | 0.82   | 1.21   | 1.41   | 0.65  | 0.51  | 0.67  |
| Sys Vel. (cm/sec.)|         |          |        |        |        |        |       |       |       |
| Average           | 4.84    | 4.68     | 3.27   | 3.16   | 2.98   | 2.32   | 2.24  | 2.18  | 2.16  |
| SD                | 0.77    | 1.76     | 1.90   | 1.06   | 1.50   | 1.35   | 1.50  | 1.16  | 0.91  |
| Dias Vel. (cm/sec.)|       |          |        |        |        |        |       |       |       |
| Average           | 2.98    | 2.87     | 2.25   | 2.23   | 2.14   | 1.98   | 1.76  | 1.37  | 1.28  |
| SD                | 1.22    | 1.34     | 1.20   | 1.15   | 0.88   | 1.05   | 0.73  | 0.85  | 0.64  |
| FORAMEN MAGNUM REGION |       |          |        |        |        |        |       |       |       |
| Premedullary Cistern AP Diameter (mm) |         |          |        |        |        |        |       |       |       |
| Average           | 2.35    | 3.07     | 3.21   | 3.30   | 3.57   | 4.14   | 4.28  | 4.35  | 4.35  |
| SD                | 0.81    | 0.72     | 0.78   | 0.81   | 0.53   | 0.48   | 0.49  | 0.50  | 0.50  |
| Cisterna Magna AP Diameter (mm) |         |          |        |        |        |        |       |       |       |
| Average           | 2.36    | 4.42     | 5.64   | 6.07   | 7.21   | 10.57  | 11.29 | 11.50 | 11.93 |
| SD                | 0.44    | 0.68     | 0.82   | 0.66   | 0.87   | 1.38   | 1.72  | 1.36  | 2.03  |
| Sys Vel. (cm/sec.)|         |          |        |        |        |        |       |       |       |
| Average           | 5.39    | 4.62     | 3.21   | 1.22   | 1.08   | 0.75   | 0.70  | 0.68  | 0.62  |
| SD                | 0.78    | 1.43     | 0.92   | 0.18   | 0.30   | 0.32   | 0.30  | 0.29  | 0.28  |
| Dias Vel. (cm/sec.)|       |          |        |        |        |        |       |       |       |
| Average           | 2.71    | 2.46     | 1.16   | 0.87   | 0.83   | 0.82   | 0.81  | 0.81  | 0.66  |
| SD                | 1.52    | 1.00     | 0.20   | 0.15   | 0.17   | 0.19   | 0.22  | 0.23  | 0.23  |

AP: anterior-posterior; C-SAS: cervical subarachnoid space; Dias: diastolic velocity; mo: month; ND: not detectable; ROI: Region of Interest; SD: standard deviation; Sys: systolic velocity; Y: year.
Table 2: Summary of ANOVA and Bonferroni multiple comparisons tests of the Syrinx Parameters

| Parameter       | ANOVA Overall Follow-Up | Bonferroni Multiple Comparisons |
|-----------------|-------------------------|---------------------------------|
|                 | Pre-op vs. Post-op      | Pre-op vs. 1st mo               | Pre-op vs. 2nd mo | Pre-op vs. 3rd mo | Pre-op vs. 6th mo | Pre-op vs. 1st Y |
| Length          | <0.001                  | 0.717                           | 0.265             | 0.012             | 0.040             | 0.015             | <0.001           |
| AP diameter     | <0.001                  | 0.179                           | <0.001            | nc                | nc                | nc                | nc               |
| Systolic Velocity| <0.001                  | <0.001                          | nc                | <0.001            | <0.001            | <0.001            | nc               |
| Diastolic Velocity| <0.001                  | 0.187                           | 0.009             | <0.001            | nc                | nc                | nc               |

AP: anterior-posterior; mo: month; Y: year; nc: not calculated

Table 3: Summary of ANOVA and Bonferroni multiple comparisons tests of the Ventral C-SAS

| Parameter       | ANOVA Overall Follow-Up | Bonferroni Multiple Comparisons |
|-----------------|-------------------------|---------------------------------|
|                 | Pre-op vs. Post-op      | Pre-op vs. 1st mo               | Pre-op vs. 2nd mo | Pre-op vs. 3rd mo | Pre-op vs. 6th mo | Pre-op vs. 1st Y |
| AP diameter     | <0.001                  | 0.001                           | nc                | nc                | nc                | nc               |
| Systolic Velocity| <0.001                  | 0.754                           | 0.801             | 0.035             | <0.001            | nc                |
| Diastolic Velocity| <0.001                  | 0.948                           | 0.790             | 0.653             | 0.007             | <0.001            |

AP: anterior-posterior; C-SAS: cervical subarachnoid space; mo: month; nc: not calculated; Y: year.

Table 4: Summary of ANOVA and Bonferroni multiple comparisons tests of the Dorsal C-SAS

| Parameter       | ANOVA Overall Follow-Up | Bonferroni Multiple Comparisons |
|-----------------|-------------------------|---------------------------------|
|                 | Pre-op vs. Post-op      | Pre-op vs. 1st mo               | Pre-op vs. 2nd mo | Pre-op vs. 3rd mo | Pre-op vs. 6th mo | Pre-op vs. 1st Y |
| AP diameter     | <0.001                  | 0.830                           | 0.123             | 0.104             | 0.046             | 0.027             | <0.001           |
| Systolic Velocity| <0.001                  | 0.757                           | 0.008             | <0.001            | nc                | nc                | nc               |
| Diastolic Velocity| <0.001                  | 0.830                           | 0.123             | 0.104             | 0.046             | 0.027             | <0.001           |

AP: anterior-posterior; C-SAS: cervical subarachnoid space; mo: month; nc: not calculated; Y: year.

Table 5: Summary of ANOVA and Bonferroni multiple comparisons tests of the Foramen Magnum Region

| Parameter       | ANOVA Overall Follow-Up | Bonferroni Multiple Comparisons |
|-----------------|-------------------------|---------------------------------|
|                 | Pre-op vs. Post-op      | Pre-op vs. 1st mo               | Pre-op vs. 2nd mo | Pre-op vs. 3rd mo |
| Premedullary Cistern | AP diameter          | <0.001                          | 0.019             | 0.008             | 0.008             | <0.001           |
| Cisterna Magna   | AP diameter            | <0.001                          | <0.001            | nc                | nc                | nc               |
| Foramen Magnum   | Systolic Velocity      | <0.001                          | 0.090             | <0.001            | nc                | nc               |
|                  | Diastolic Velocity     | <0.001                          | <0.001            | nc                | nc                | Nc               |

AP: anterior-posterior; mo: month; nc: not calculated; Y: year.

Figure 1: Sagittal T1- (a) and T2-weighted magnetic resonance imaging (b) of the brain and spine, and axial T2-weighted magnetic resonance imaging (c) (C4–C5 intervertebral disk) showing a Chiari I malformation with a holocord syringomyelia. (d) Preoperative cine phase-contrast magnetic resonance imaging during systole revealing the cerebellar tonsils moving downward, the dilation of the syrinx synchronous with the systole, and a cerebrospinal fluid jet flow inside the syrinx. (e) Cine magnetic resonance imaging performed 1 month after posterior fossa decompression documenting a significant systolic reduction of flow inside the syrinx.
the intracranial pressure would then be normalized by the CSF flowing through the obex into the central canal, ultimately leading to the syrinx formation. Increasing abdominal pressure is transmitted to the epidural spinal veins producing an ascending venous pressure wave from the spinal subarachnoid space through the foramen magnum. Due to the obstruction at the level of craniocervical junction, the pressure wave ascends to the spinal axis, expanding the syrinx. Oldfield et al. and Heiss et al. proved that in the presence of an obstruction of the foramen magnum and a “piston effect” by the cerebellar tonsils on the spinal subarachnoid space, increasing of abdominal pressure is transmitted to the spinal cord through the Virchow–Robin perivascular spaces. They also demonstrated that the propelling force at the base of this “transmedullary” CSF flow is the heartbeat, with systole and diastole leading to a self-maintaining mechanism. Indeed, a CSF flow through the Virchow–Robin perivascular spaces associated with the genesis of syringomyelia was described several years before by Ball and Dayan.

Rai and Rai reported in 2015 their valuable alternative hypothesis about the Chiari malformation-associated syringogenesis, namely the “volume change” theory. This theory is based on the CSF flow dynamics at the level of the posterior fossa and the subsequent changes in volume of the spinal canal and its content during flexion and extension of the head, leading to the syrinx formation as a compensatory mechanism.

The "volume change" theory constitutes the rationale for Goel's technique, namely stand-alone C1–C2 fixation. This because the instability of the atlantoaxial complex has been reported to be the cause of Chiari 1 malformation, syringomyelia, and basilar invagination. Based on this evidence, syringomyelia secondary to Chiari malformation
ought to be treated with stabilization of the atlantoaxial joint, with no need for PFD with or without intradural manipulation.[63‑68]

In our study, the finding of a clear systolic pattern of the injection of the syrinx by a CSF jet flow which is synchronous with the heartbeat, as reported in the illustrative case, supports the Oldfield and Heiss “transmedullary” theory.

This concept is furtherly reinforced by the fact that, according to these observations, CSF systolic velocity inside the syrinx decreases immediately after surgery.

From a technical standpoint, the adjustment of the velocity encoding in case of aliasing is the key to avoid artifacts during acquisition of the dynamic CSF study.

Limitations of the study
Our study has some important limitations which ought to be mentioned. First, the number of the patients is limited and series by far wider are necessary to draw a definitive conclusion and to validate the proposed neuroimaging study algorithm as the approach of choice in these patients. Second, the search for a systolic–diastolic CSF flow pattern is often difficult to achieve during cine phase-contrast MRI, and the technique is affected by a not-negligible risk of false positives and negatives. Artifacts coming from a limited velocity encoding or aspects as prospective versus retrospective cardiac gating are further technical aspects making difference. Third, the reported study did not involve a control group, being basically an observational case series study. Randomized trials are necessary to achieve definitive conclusions.

CONCLUSION
A biphasic systolic–diastolic CSF pulsatile pattern, detected with cine phase-contrast MRI, can be documented in a large part of the patients diagnosed with syringomyelia associated with Chiari I malformation.

PFD causes an early decrease of the velocities inside the syrinx and at the level of the foramen magnum. These velocities are predictors of positive outcome for an early recovery from headache, dysesthetic pain, weakness, lower cranial nerve dysfunction, and dissociated sensory loss.

The evidence of a pulsatile CSF flow pattern inside the syrinx supports the Oldfield and Heiss “transmedullary” theory about the formation and progression of the syrinx.

Further studies are necessary the definitively validate these data.

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Conflicts of interest
There are no conflicts of interest.

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