Original Article

Is foramen magnum decompression for acquired Chiari I malformation like putting a finger in the dyke? - A simplistic overview of artificial intelligence in assessing critical upstream and downstream etiologies

ABSTRACT

Background: Missed diagnosis of evolving or coexisting idiopathic (IIH) and spontaneous intracranial hypotension (SIH) is often the reason for persistent or worsening symptoms after foramen magnum decompression for Chiari malformation (CM) I. We explore the role of artificial intelligence (AI)/convolutional neural networks (CNN) in Chiari I malformation in a combinatorial role for the first time in literature, exploring both upstream and downstream magnetic resonance findings as initial screening profilers in CM-1. We have also put together a review of all existing subtypes of CM and discuss the role of upright (gravity-aided) magnetic resonance imaging (MRI) in evaluating equivocal tonsillar descent on a lying-down MRI. We have formulated a workflow algorithm MaChiP 1.0 (Manjila Chiari Protocol 1.0) using upstream and downstream profilers, that cause de novo or worsening Chiari I malformation, which we plan to implement using AI.

Materials and Methods: The PRISMA guidelines were used for "CM and machine learning and CNN" on PubMed database articles, and four articles specific to the topic were encountered. The radiologic criteria for IIH and SIH were applied from neurosurgical literature, and they were applied between primary and secondary (acquired) Chiari I malformations. An upstream etiology such as IIH or SIH and an isolated downstream etiology in the spine were characterized using the existing body of literature. We propose the utility of using four selected criteria for IIH and SIH each, over MRI T2 images of the brain and spine, predominantly sagittal sequences in upstream etiology in the brain and multiplanar MRI in spinal lesions.

Results: Using MaChiP 1.0 (patent/ copyright pending) concepts, we have proposed the upstream and downstream profilers implicated in progressive Chiari I malformation. The upstream profilers included findings of brain sagging, slope of the third ventricular floor, pontomesencephalic angle, mamillopontine distance, lateral ventricular angle, internal cerebral vein–vein of Galen angle, and displacement of iter, clivus length, tonsillar descent, etc., suggestive of SIH. The IIH features noted in upstream pathologies were posterior flattening of globe of the eye, partial empty sella, optic nerve sheath distortion, and optic nerve tortuosity in MRI. The downstream etiologies involved spinal cerebrospinal fluid (CSF) leak from dural tear, meningeal diverticula, CSF-venous fistulae, etc.

Conclusion: AI would help offer predictive analysis along the spectrum of upstream and downstream etiologies, ensuring safety and efficacy in treating secondary (acquired) Chiari I malformation, especially with coexisting IIH and SIH. The MaChiP 1.0 algorithm can help document worsening of a previously diagnosed CM-1 and

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find the exact etiology of a secondary CM-I. However, the role of posterior fossa morphometry and cine-flow MRI data for intracranial CSF flow dynamics, along with advanced spinal CSF studies using dynamic myelo-CT scanning in the formation of secondary CM-I is still being evaluated.

Keywords: Acquired Chiari, artificial intelligence, Chiari I malformation, convolutional neural networks, idiopathic intracranial hypertension, Manjila Chiari Protocol 1.0 (MaChiP 1.0), spontaneous intracranial hypotension

INTRODUCTION

Chiari malformations (CMs) are a group of disorders which were traditionally described as entities, in which there is either herniation of posterior cranial fossa content below the foramen magnum (FM) or hypoplasia of the posterior cranial fossa and the cerebellum.[1] CMs I–III were proposed based on the degree of hindbrain or tonsillar herniation through the FM according to the original Chiari classification system. Chiari type IV corresponds to cerebellar aplasia or hypoplasia. More recently, Chiari type 0 and 1.5 have also been suggested, with type 0 CM representing a symptomatic patient with a syrinx and "0" mm of tonsillar descent and type 1.5 describing tonsillar herniation with additional caudal descent of the brainstem [Table 1].[2]

CM-I has been a difficult condition to treat, and we tried to explore the differentiation of various subtypes less than Chiari II [colored pink in Table 1]. Chiari 0 malformation is described as syringohydromyelia that resolves after FM decompression, along with low-lying cerebellar tonsils that block but do not or minimally descend through FM, beyond the basion-opisthion line. Chiari 0.5 malformation is characterized by ventral tonsillar herniation beyond a line that bisects the medulla to the anterior and posterior parts at the level of FM. Tonsillar herniation through FM, if present, will always be <5 mm.[3] CM-I is defined as herniation of one or both cerebellar tonsils >5 mm through FM. Importantly, no herniation of brainstem is present.[4,5] Chiari 1.5 malformation can be described as advanced form of CM-I. It is defined by herniation of cerebellar tonsils as well as the elongated brainstem, or part of it, through the FM.[6] As for Chiari II malformation, it is characterized by herniation of cerebellar tonsils and vermis and caudal displacement of brainstem and fourth ventricle through an enlarged FM. Chiari II is almost always associated with myelomeningocele.[7]

Hans Chiari in 1891 defined Chiari III malformation as herniation of cerebellum into a high cervical hydroencephalocele along with the other findings of Chiari II malformation. However, the term Chiari III had expanded later on to describe the herniation of posterior fossa contents and cerebral tissue (e.g., occipital lobe and lateral ventricles) into a high cervical or a low occipital encephalocele. In contrast, Chiari IV malformation refers to cerebellar hypoplasia along with occipital encephalocele that contains supratentorial structures only (e.g., occipital lobe and choroid plexus of lateral ventricles).[8] The term Chiari 3.5 malformation was proposed by Christian Fisahn in 2016 to describe a case that was reported earlier by Giuseppe Muscatello in 1846, in a premature girl with occipitocervical encephalocele along with missing cerebellum, tentorium cerebelli, and transverse sinuses. Finally, Chiari V malformation was introduced by Shane Tubbs to describe a patient with occipital lobe herniation through FM and absent cerebellum.[9]

Magnetic resonance imaging (MRI) of the brain and cervical spine remains the imaging modality of choice for the initial assessment of CM-1, but if there is suspicion of spontaneous intracranial hypotension (SIH), a pan-spine MRI is indicated.[10] Conventionally, the defining radiological feature of the CM type I is tonsillar descent of 5 mm or more beyond the FM. The descent is measured by calculating the perpendicular distance between the tip of herniated tonsil and the McRae's line (a radiographic line drawn on a lateral midsagittal section of computed tomography [CT] or MRI, joining the basion and opisthion representing the level of FM).[11] In addition to clinical history and examination, neuroimaging is essential in the diagnosis of CM-I to assess the anatomical structures and cerebrospinal fluid (CSF) dynamics that are associated with a CM-I.

ACQUIRED CHIARI I MALFORMATION

Acquired or secondary CM is a rare occurrence, and the literature describing its exact pathophysiology and approach to treatment remains sparse.[12,13] The underlying cause in all cases is an imbalance between calvarial volume and intracranial contents or excess or diminished movement of CSF out of the cranial cavity.[14] Decreased cranial vault capacity is seen in Paget's disease, rickets, craniosynostosis, and erythroid hyperplasia, while tumors, acute hydrocephalus, and idiopathic intracranial hypertension (IIH) cause an increase in intracranial content. Excess CSF drainage due to spontaneous CSF leaks, lumbar drainage, or secondary to a lumboperitoneal shunt also leads to acquired tonsillar descent. The posterior fossa volume and morphometry as well as intracranial dentate ligament and craniovertebral junction ligaments are implicated as causative factors in worsening tonsillar descent.[15-19]

Table 1: Different types of Chiari malformations reported in neurosurgical literature

	Chiari 0	Chiari 0.5	Chiari 1	Chiari 1.5	Chiari 2	Chiari 3	Chiari 3.5	Chiari IV	Chiari V
Anatomy	Cerebellar tonsils are blocking but not descending through the FM + syrinx (Chiari 0 classified under syringomyelia)	Ventral cerebellar tonsil herniation across a line that bisects the medulla (to anterior and posterior parts) at the level of the FM Cerebellar tonsils descend <5 mm through FM	Caudal descent of one or both cerebellar tonsils through FM >5 mm	Caudal descent of the cerebellar tonsils+all/part of brainstem through FM	Elongated small cerebellum and brainstem with caudal displacement of cerebellar tonsils and vermis, fourth ventricle and brainstem through an enlarged FM. Almost always associated with myelomeningocele	Herniation of dysplastic posterior fossa contents into an associated occipitocervical encephalocele (rhombencephalocele)	Occipitocervical encephalocele connecting with the stomach. Anomalous anatomy of posterior fossa	Occipital encephalocele with supratentorial contents (cortical and ventricular tissue), absent or severely hypoplastic cerebellum, large posterior fossa CSF spaces, and small brainstem	Herniation of occipital lobe through FM Absent cerebellum
Radiological finding	Syrinx No herniation but low-lying tonsils pressing against FM	Cerebellar tonsillar position <5 mm Downward tonsillar herniation and ventral tonsillar position relative to a novel line bisecting the caudal medulla at the level of the FM	Pointed "peg-like tonsils" Vertically oriented sulci "sergeant stripes" Cerebellar tonsillar position shows >5 mm + no descent of brainstem	At least one herniated tonsil and a downward herniation of all/part of brainstem and no radiological findings of Chiari II Possibly Posterior angulation of odontoid process	Antenatal US Lemon sign Banana cerebellum sign Ventriculomegaly Fetal myeloschisis Posterior fossa Herniation of cerebellar tonsils and vermis through FM Caudal displacement of medulla±pons "Beaked" tectum of midbrain due to fusion of colliculi Aqueductal stenosis Kinking of brainstem Rostral herniation of superior cerebellum through the tentorium Cerebral hemispheres Dysgenesis of corpus callosum Large massa intermedia Cortical heterotopias Polymicrogyria Skull Lückenschädel of the skull Low-lying tentorium cerebelli with large incisura Scalloping of the petrous bone Short clivus Enlargement of FM Spine Myelomeningocele Tethered cord Filum lipoma	MRI: Occipitocervical encephalocele Herniation of brain (cerebellum and occipital lobe), ventricles (fourth or lateral), Cisterns, medulla, and pons through Chiari II malformation findings	N/A	MRI Occipital encephalocele with supratentorial contents Cerebellar hypoplasia Small posterior fossa	Herniation of occipital lobe through FM Absent cerebellum

Contd...

Table 1: Contd...

	Chiari 0	Chiari 0.5	Chiari 1	Chiari 1.5	Chiari 2	Chiari 3	Chiari 3.5	Chiari IV	Chiari V
Etiology/ mechanism	Arachnoid adhesions at 4th ventricular outlet block CSF flow	Ventrolateral wrapping of the cerebellar tonsils and compression of the lateral medulla and the exiting lower cranial nerves	Congenital Abnormal skull base Cervical segmentation anomalies Small cranial vault Excessive brain tissue Acquired Intracranial hypertension Intraspinal hypotension	Similar to Chiari 1 malformation plus Smaller posterior fossa Posterior angulation of odontoid process disrupting CSF flow and increasing syringomyelia size	Defective closure of neural tube and incomplete spinal occlusion results in leakage of CSF and collapse of cranial vesicles whom distention is necessary for brain development	Similar to Chiari II with added failure of induction of endochondral bone by incomplete closure neural tube or failure of ossification centers to fuse completely	Neurenteric fistula/canal. Axial canal of notochord (connects amniotic and yolk sac to equalize their pressure) fails to close	Undeveloped/ underdeveloped cerebellum	-
Reference		Peter F. Morgenstern	Julius Arnold (1835–1915) and Hans Chiari (1851–1916)		Julius Arnold (1835–1915) and Hans Chiari (1851–1916)	Julius Arnold (1835–1915) and Hans Chiari (1851–1916)	Described by Giuseppe Muscatello, 1894 Named by Christian Fisahn, 2016	Julius Arnold (1835–1915) and Hans Chiari (1851–1916)	R. Shane Tubbs

CSF - Cerebrospinal fluid; FM - Foramen magnum; MRI - Magnetic resonance imaging; US - Ultrasound; N/A - Not available

Acquired CM was first reported in 1976 by Hoffman and Tucker as a delayed complication in an extrathecal shunt drainage. Clinical presentation of acquired Chiari is like congenital CMs, with suboccipital headaches, neck pain, and neck stiffness. Motor sensory deficits are seen in cases that also have associated syringomyelia, but the overall incidence of syrinx is low in acquired Chiari as compared to congenital CM. CT or MRI will demonstrate downward descent of the cerebellar tonsils and may point to the diagnosis in surgical pathologies such as tumors, arachnoid cysts, craniosynostosis, and achondroplasia causing a secondary CM-I malformation. Phase contrast cine flow MRI is also useful in assessing CSF flow across the FM.[20-24]

Treatment of acquired Chiari is the surgical or interventional treatment of its etiology, independent of whether the clinical presentation is acute or chronic. The overarching principle of management is to restore the CSF dynamics at the craniovertebral junction and relieve the headaches. The interventional treatment ranges from blood patch to endovascular embolization. The bony extent of the cranial decompression, the need for duroplasty, the choice for the dural substitute material and/or dural sealant used, and the need for an expansile cranioplasty, all remain controversial. It is important not to overtreat asymptomatic patients with only radiological abnormalities; however, it is imperative to follow these patients closely to observe any change in symptoms over a period. Space occupying lesions are treated surgically, either by tumor excision or cystoperitoneal shunting for arachnoid cysts. In cases that are secondary to overdrainage of CSF after shunt, changing the valve apparatus such as a programmable and gravitational valve, likely with a siphon guard to prevent overdrainage, is the first step. When these measures fail, or there is established craniocephalic disproportion, surgery to increase posterior fossa volume is indicated.[25]

MATERIALS AND METHODS

Downstream and upstream etiologies of acquired chiari malformation

The upstream pathologies causing acquired CM can be either IIH or SIH.[26,27] It is often difficult to apply strict clinical and radiological criteria to clearly diagnose them. The downstream etiologies such as spontaneous spinal CSF leaks might require additional dedicated tests like dynamic myelographic studies.

Downstream etiologies

The most common cause of acquired Chiari is low CSF pressure such as SIH.[28] This can be secondary to drainage of CSF by lumbar shunting, CSF drainage by lumbar taps, or

spontaneous leaks.[29-34] The spontaneous spinal leak of CSF is usually located in the cervicothoracic region. These patients may also have some structural predisposing conditions such as congenital weakness of the dural sac, meningeal diverticula (like Marfan syndrome), connective tissue disorders, spondylotic spurs, or disk herniation, which make them more prone to develop this condition. Occasionally, they may report a trivial trauma before the onset of symptoms.

SIH patients present with postural or orthostatic headaches.[35] This differs from that of congenital Chiari, being shorter in duration.[36] It is also rare for syrinx to be associated with an acquired Chiari secondary to a spinal leak. A history like a trivial trauma, or connective tissue disorders, can provide a valuable clue to the diagnosis of acquired CM.

The CSF drainage causes sagging of the brain and subsequent intracranial hypotension distorts the pain-sensitive structures and neural structures, often leading to cranial nerve palsies.[37] It is difficult to locate the leak, especially in low-flow leaks. The best initial treatment is bed rest, hydration, and caffeine. Those who fail conservative management undergo an epidural blood patch followed by surgical repair of the leak if the blood patch fails.

CSF-venous fistulas represent a recently recognized entity that potentially precipitate excess CSF drainage. The term describes direct communications between the spinal subarachnoid space and the adjacent paraspinal veins.[38] They can be spontaneous or iatrogenic and occur from T7 to T12 most commonly. Spinal veins play a significant role in the normal absorption of CSF and contribute to 1/5th of CSF absorption. Spinal arachnoid granulations give rise to a fistula which can drain excess CSF, leading to intracranial hypotension.[39] Despite advanced imaging modalities such asdigital subtraction myelography or intrathecal gadolinium magnetic resonance (MR) myelography, these dynamic lesions are difficult to be diagnosed.

In patients where the leak can be defined with imaging, surgery is the treatment of choice.[40] Spinal intracranial hypotension can be treated surgically with direct CSF-venous fistula ligation via hemilaminectomy, nerve root ligations for a CSF-venous fistula, or a direct surgical repair of the spontaneous ventral dural tears causing CSF leaks.[41] The usual site is the lower thoracic nerve roots, in which case the nerve root is ligated to seal the fistulous connection. In rare instances when functional cervical or lumbar roots are involved, transvenous embolization with Onyx has been successfully reported in a few cases.[42]

Upstream etiologies

Pseudotumor cerebri or IIH is increased intracranial pressure (ICP) in the absence of a mass lesion or obstruction to CSF absorption. The pathophysiology is a complex interplay between the vascular bed, the CSF space, and the brain parenchyma, within the closed system of the skull.[43-52] The incidence of CM-1 in patients diagnosed with IIH is 6%. While some researchers attribute abnormal CSF absorption in IIH to venous congestions, others suggest that increased ICP is the main causative factor.[53]

An association between IIH and CM has been hypothesized and a common pathophysiologic mechanism has been proposed.[54-56] A study compared pulsatile and static ICP in patients with IIH and CM; it found that pulsatile ICP was elevated and comparable in both IIH and CM patients, whereas static ICP was higher in the IIH cohort. The comparable and increased pulsatile ICP pointed toward impaired intracranial compliance as a common pathophysiologic mechanism. Despite multiple studies, the exact relationship between CM and IIH is still poorly understood. Raised ICP and brain swelling in IIH may cause downward herniation of the intracranial contents through the FM corresponding to a radiological CM. Alternatively, a subset of Chiari I patients may have abnormal CSF flow at the FM causing raised ICP.

The treatment is aimed at the underlying cause. In the case of IIH with mild or asymptomatic CM-1, acetazolamide is the first-line treatment. Patients with visual symptoms benefit from optic nerve sheath fenestration without significantly altering the underlying disease process. It is difficult to distinguish Chiari 1 with papilledema from IIH, and there may be a significant overlap in these two conditions. Reports of early deterioration after surgery for presumed acquired CM secondary to pseudotumor cerebri point to our sparse understanding of these etiologies. Good clinical success with CSF diversion shunts or transvenous stent placement has been noted in patients with IIH.

UPRIGHT MAGNETIC RESONANCE IMAGING AND GRAVITY-PRONE CHIARI MALFORMATION I

Recently, upright MRI systems including MR Open scanner from Paramed Medical system, G-Scan Brio from Esaote, and the Stand-Up MRI system from the FONAR corporation, have been contemplated as a dynamic gravity-aided non- invasive investigational study.[57] This is because tonsils may be lower when the patient is upright owing to the gravitational forces and thus increasing the sensitivity of MRI. CM-1 linked to hypermobility syndromes such as Ehlers-Danlos and Marfan syndrome is increasingly diagnosed using flexion and extension features in an upright MRI.

We propose that there is an intricate relation between CM-induced tonsillar herniation and gravity. There have been reports of intracranial hypertension in some space travelers due to zero gravity.[58] This could exacerbate an existing CM or cause an acquired CM. This correlation of CM and gravitational forces can be extrapolated to include upright and dynamic MRI systems in the screening of CMs. We feel that the patients with equivocal tonsillar descent and classic Chiari symptoms should be subjected to an upright MRI to check if the tonsillar descent worsens.

RESULTS

ARTIFICIAL INTELLIGENCE IN CHIARI MALFORMATION I

The application of artificial intelligence (AI) principles in the disease progression leads to a diagnosis of coexisting IIH or SIH with CM-1 [Figure 1]. This detection of SIH can be of clinical use in the decision-making between surgical treatment and conservative line of management. MRI measured values including measures of brain sagging TE, slope of the third ventricular floor, pontomesencephalic angle, mamillopontine distance, lateral ventricular angle, internal cerebral vein-vein of Galen angle, and displacement of iter, clivus length, and tonsillar descent are recognized by Houk *et al*. as features of SIH. as features of SIH. Based on these parameters compared with the historical MRI studies, the progression of the disease can be clearly defined, and appropriate treatment modality could be opted.

On the other hand, IIH has MRI and magnetic resonance venography (MRV) findings with classic features of empty sella turcica, optic nerve protrusion, distension of the optic nerve sheath, optic nerve tortuosity, posterior globe flattening, and transverse sinus occlusion, although absence of these findings does not rule out the diagnosis of IIH.

MRI brain regular protocol with / without contrast and T2 sequences along with whole spine T2 sequences for screening is incorporated into the baseline image acquisitions, for applying MaChiP 1.0 (Manjila Chiari Protocol 1.0) algorithm (patent and copyright pending). The compiled information in the form of interpreted output data would be a useful tool in the prediction of disease progression.

Optimal output of the progression of the abovementioned individual parameters could be charted for deciding the optimum treatment protocol. Image data are required to be uniformly formatted to avoid the errors as the different scanners are using different imaging software.

The basic protocols of regular data feeding, deep learning, developing neural networks, and final output could be followed in studying CM-I. The optimized data based on the described algorithm would get appropriate output as it is better classified in terms of accuracy. Once these refined imaging data are added for machine learning, we can get an appropriate logical output for the desired disease progression.

The dimensionality feature reduction and selection of the features of decisive nature are the crucial parts of data analysis. Once feature selection based on algorithms is accomplished, the pattern recognition of different subjects could be classified. Once the classified data are incorporated, the machine learning will be gaining the intended accuracy. Based on these parameters, the basic structure of the analytic model can be created. Once the analytic model is ready, the sieve analysis of the collected subject MR images can determine the desired output.

The implementation of deep learning convolutional neural network (CNN) models in neuroimaging is still an emerging area of research.[59,60] Tanaka *et al*. was able to train CNN models like ResNet50 and VGG19 to automatically detect clinically significant CM 1 on MRI with a high sensitivity and specificity. These models have the potential to be upgraded by incorporating clinical outcome data, postoperative imaging, and postoperative outcomes to create a multifaceted clinical support tool.

DISCUSSION

MAGNETIC RESONANCE IMAGING-BASED CHIARI I PREDICTION USING DEEP LEARNING MODEL

Resnet50/XceptionNet are now being used to train deep learning models for the classification of CM-I on MRI to assist clinicians in diagnosis and decision-making. Fast region-based CNN (R-CNN) provides accurate cerebellar herniation estimation using CNNs. They build on previous work to efficiently classify object proposals using deep convolutional networks. Fast R-CNN models depend on the selective search algorithm for generating region proposals. Each proposal is fed to a pretrained CNN for classification. Images are annotated using VOTT for Cerebellar Tonsil Bounding Box and FM Bounding Box and train the model. Alternatively, a faster R-CNN can be employed, which offers a better performance.

ResNet-50 model is a CNN that is 50 layers deep and can be trained well using the deep learning library called Keras. Keras contains pretrained learning models which are

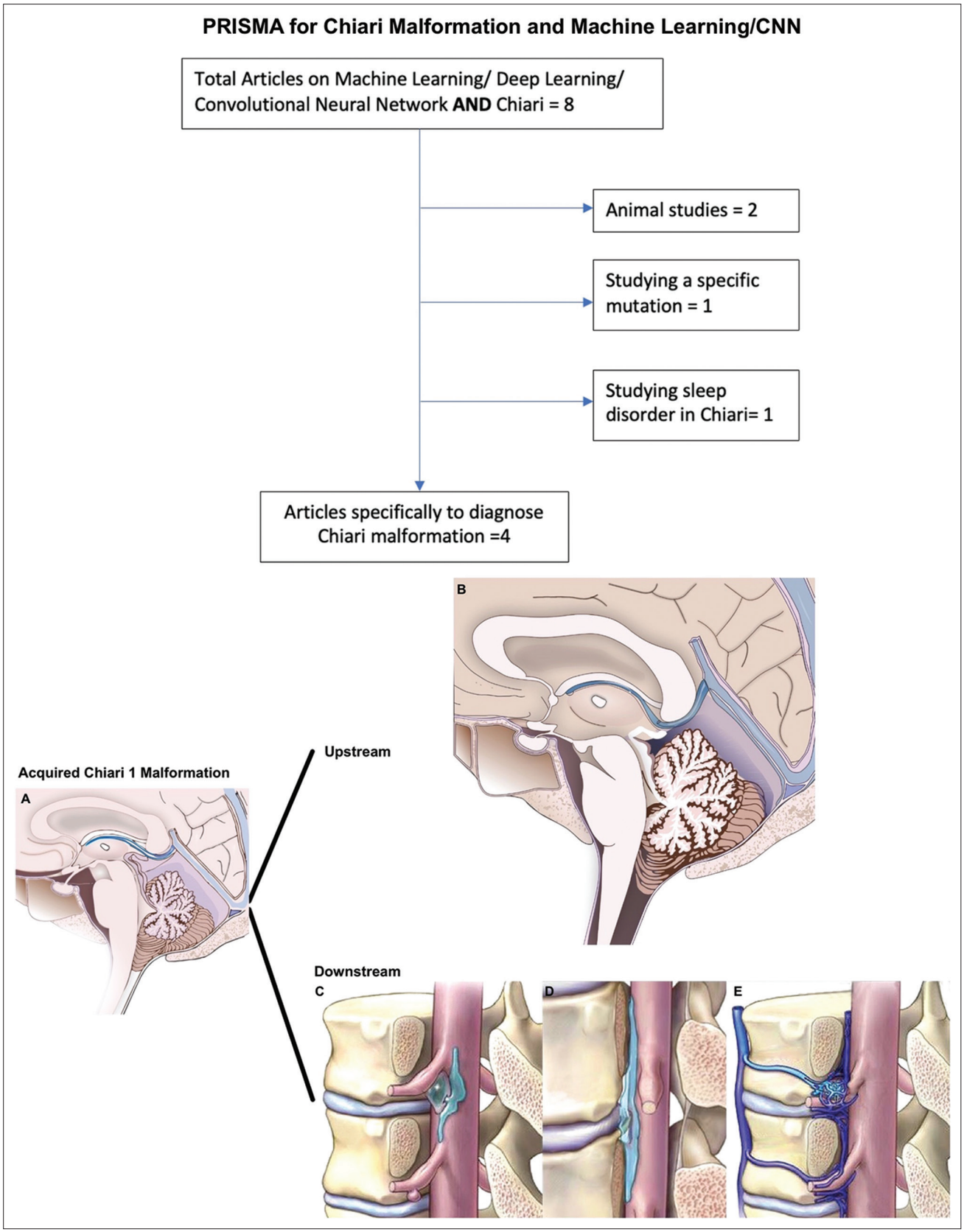

Figure 1:The artistic rendition of acquired or worsening Chiari I malformation in A, upstream intracranial findings in B and spinal CSF leak from various lesions causing SIH shown in C-E; CNN= Convolutional Neural Network

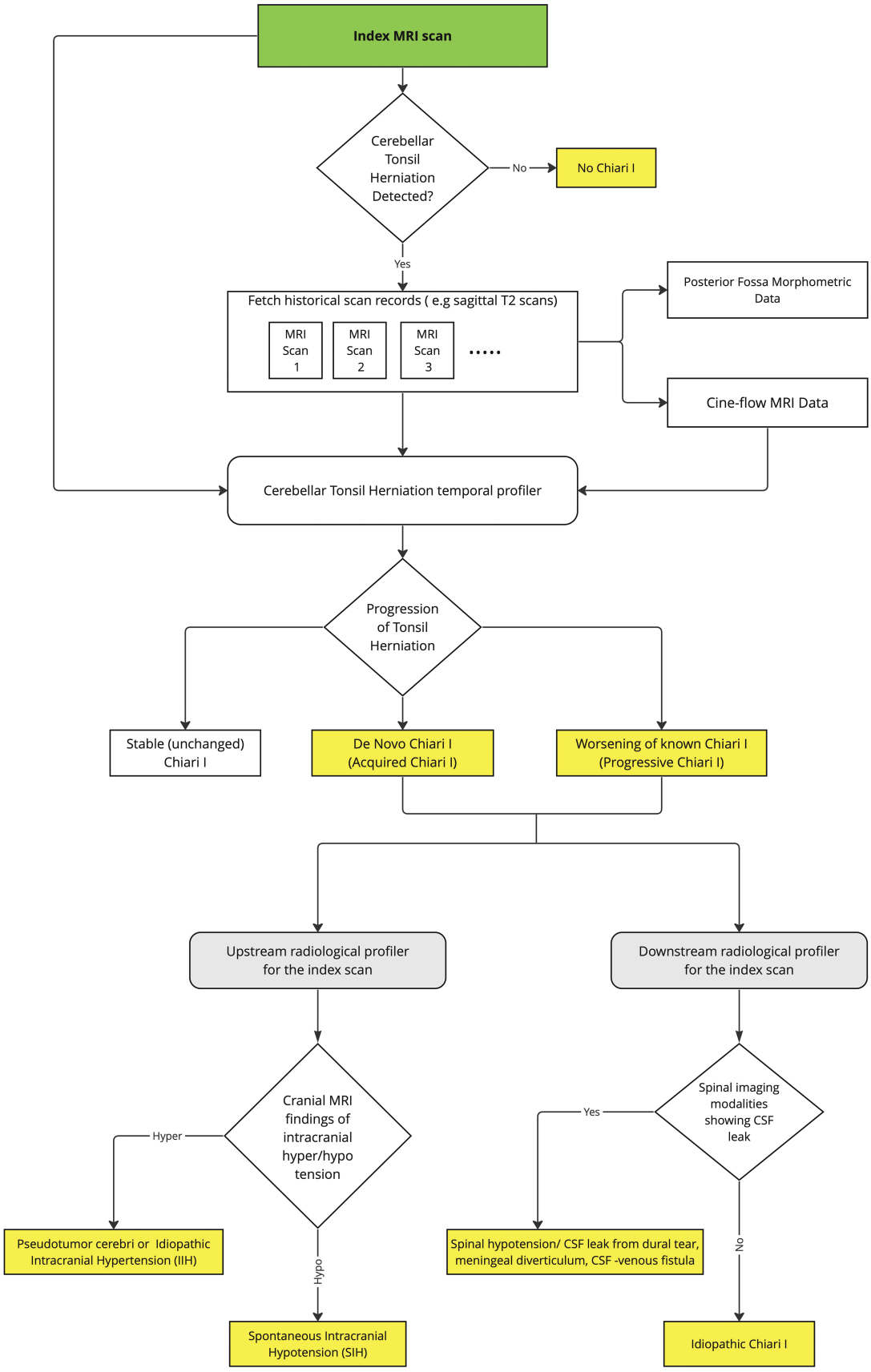

Figure 2: A novel artificial intelligence-based algorithm for Chiari detection and characterization. MRI: Magnetic resonance imaging, CSF: Cerebrospinal fluid, SIH: Spontaneous intracranial hypotension, IIH: Idiopathic intracranial hypertension

actually open-source networks. We feel that ResNet would offer a stable platform for analyzing both the upstream and downstream radiologic profilers once a CM-I is identified by the radiologist.

The proposed MaChiP solution requires automated software systems capable of fetching the historical MRI scans from Picture archiving and communication system (PACS) and running the proposed CM-I detection algorithm on these scans [Figure 2]. The software system must also have the machine intelligence required to recognize and detect various radiological findings and observations. Studies such as Lin *et al*. and Tanaka *et al.* have explored using CNN models on MRI to aid in CM-I diagnosis and decision-making.[61] Tanaka *et al*. have used ResNet model pretrained on ImageNet dataset as the substrate of their neural network model and employed transfer learning to train the ResNet to defect CM-I malformation on a small dataset (135 images).[62] They employ techniques like skull stripping and image augmentations (rotation, scaling, and offset parameters), as well as K-fold cross-validation to make the training more effective.

We propose a similar CNN classifier to detect CM-I [Figure 3]. However, to implement our detection algorithm, we not only need our model to be able to classify scans based on the presence or absence of CM-I malformation but also be able to measure the tonsillar herniation in millimeters. In essence, we need to turn the CM-I classification model to a CM-I herniation measurement regression model. To achieve this, we would build a faster R-CNN model over CNN to draw bounding boxes over cerebellar tonsil and FM.[63] A study by Ma *et al*. used faster R-CNN over MRI images to similar effect.[64] With bounding boxes over cerebellar tonsil and FM, we can easily compute the tonsillar herniation depth as shown in Figure 3.

Following herniation measurement over all available MRI scans of the patient, our algorithm proceeds to investigate and infer causes of CM I. To do so, we employ two different software modules: upstream radiological profiler and downstream radiological profiler. Upstream radiological profiler aims to classify an input sagittal brain MRI into intracranial hypertension and intracranial hypotension [Figure 4]. Of note, only these four highly specific/sensitive MR diagnostic features in sagittal planes are selected although absence of these findings does not rule out the diagnosis of IIH. Furthermore, an MRV which offers a significant clue in the diagnosis of IIH can also be included later in the AI study.

Downstream radiological profiler is a classifier that detects presence or absence of spinal hypotension on sagittal,

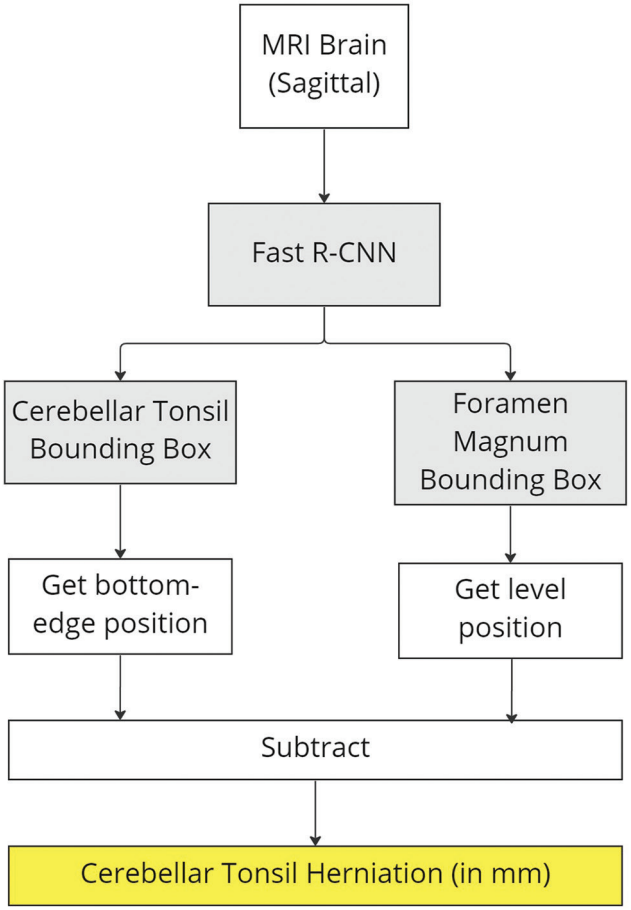

Figure 3: Artificial intelligence algorithm for calculating cerebellar tonsil herniation as the first step. MRI: Magnetic resonance imaging, R-CNN: Region-based convolutional neural network

coronal, and axial spine MR images. It does so based on the detection of meningeal diverticula, dural tear, and CSF venous fistula as shown in Figure 5.

Computationally, these profilers are a set of ResNet classifiers used to detect each of radiological findings in MRI. Though it is possible to build a single CNN for upstream and downstream radiological profilers to perform classification, having a set of specialized classifiers for each radiological finding aids in achieving better clinical interpretability of the classification. Better preprocessing steps and image treatments could be applied to the MR images which are specific to the observation that model must make. This will also aid in improving the general accuracy of the models.

Challenges and limitations

The algorithm expects that the historical MRI scan records of the patient are accessible digitally and instantly, which is often a cumbersome task [Figure 6]. Uniformity of digital data is also a technical difficulty as the follow-up imaging is done every few years and the imaging technology is evolving

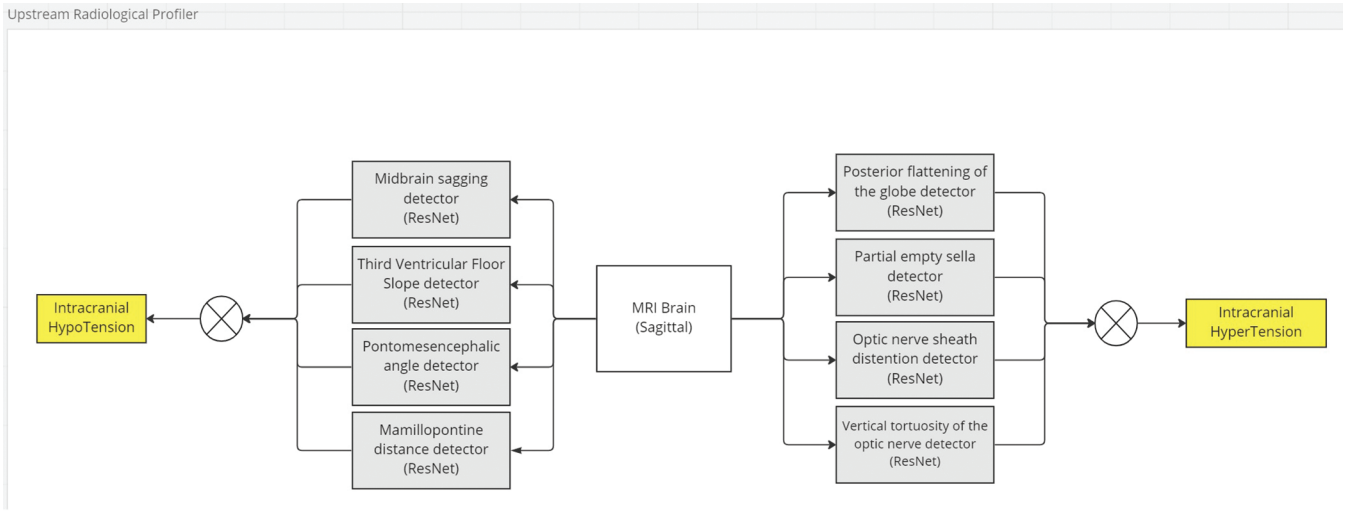

Figure 4: Artificial intelligence algorithm for studying the upstream radiological profilers from the magnetic resonance findings (4 parameters each selected for idiopathic intracranial hypertension and spontaneous intracranial hypotension from sagittal T2 sequences for proof-of-concept studies). MRI: Magnetic resonance imaging

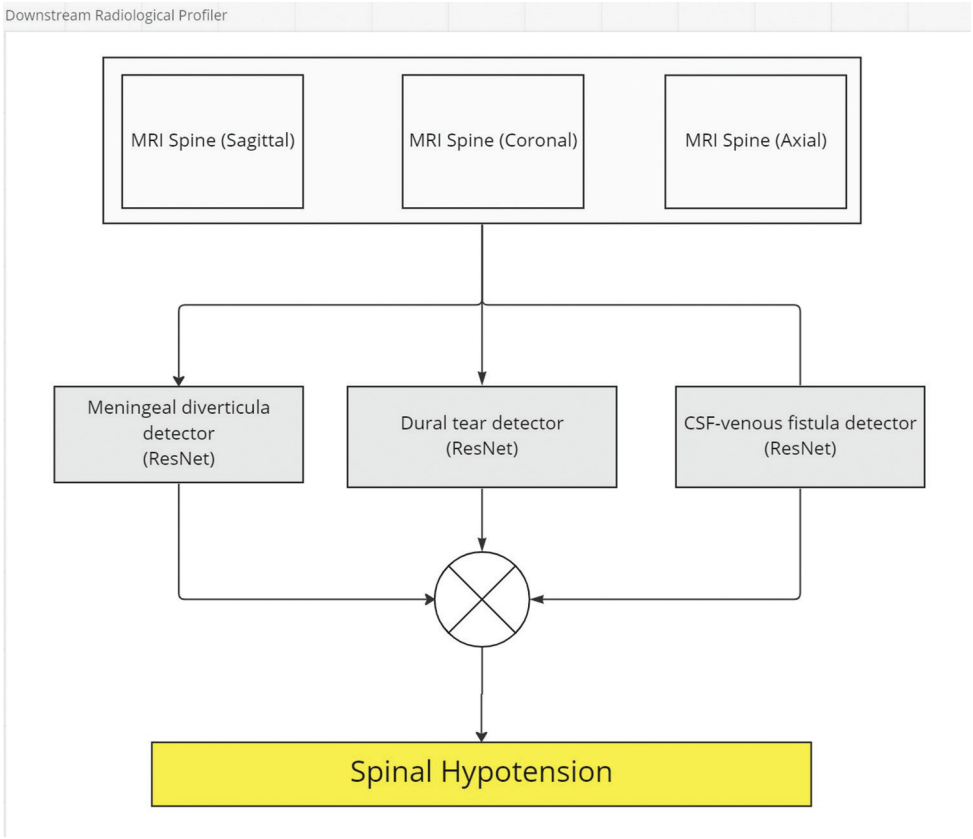

Figure 5: Artificial intelligence algorithm for downstream radiological profilers on magnetic resonance imaging findings (multiplanar studies on magnetic resonance axial, coronal, and sagittal sequences) for spontaneous intracranial hypotension diagnosing meningeal diverticula, spinal dural tears (cerebrospinal fluid [CSF] leaks), and CSF-venous fistula detectors using ResNet. MRI: Magnetic resonance imaging, CSF: Cerebrospinal fluid

every day. The comparison of an obsolete format with a newer format would be arduous and can pose a tremendous technical workload.

Though data science techniques -like transfer learning and data augmentation- help in building models with large enough data, building models that are robust enough to be used in clinical practice requires huge amounts of data. Building a huge cohort of MR scans and training neural networks on them would be a prohibitively expensive and time-intensive exercise.

The posterior fossa morphometry also has a crucial role in the formation of a secondary CM-I malformation, which poses some

unique challenges when building the cohort of scans for training the model [Figure 7]. Likewise the effect of gravity aided MRI and cine flow MRI findings at the craniovertebral junction are required to be incorporated to the MaChiP algorithm. Special attention needs to be taken to ensure that no racial/ethnic group be underrepresented in the cohort; otherwise, we would risk building unintended biases into the AI.

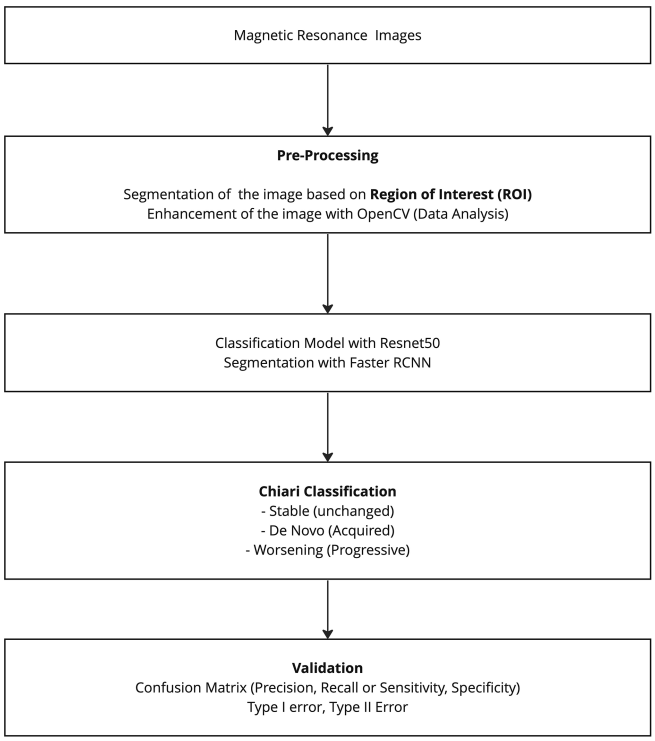

Figure 6: Pipeline for preprocessing, classification, probability scoring, and validating radiologic findings of an acquired Chiari case. MRI: Magnetic resonance imaging

CONCLUSION

CM-I malformation, whether primary or acquired (secondary) in the setting of IIH or SIH, can often be extremely difficult to treat, and we feel that the AI can help reduce the gap in the clinical workflow in these conditions. This article provides a comprehensive update of all the subtypes of CMs, with a special attention to gravity-aided (upright) MR scans if there is a questionable <5 mm tonsillar descent with classic symptoms. When a patient is diagnosed with <5 mm Chiari for the first time, it is prudent to look for upstream and downstream etiologies such as IIH or SIH, evaluated as radiologic profilers. The future of CM diagnosis lies in upright MRI, computation of CSF space at the craniocervical junction via cine-flow MRI, and morphometric/volumetric measurements, all of which can be performed in association with machine-learning models. The novel AI algorithm proposed here can help identifying secondary or acquired CM I and the underlying etiology. Therefore, it helps the radiologist and neurosurgeon to fix the problem rather than put a finger on the dyke by performing a FM decompression.

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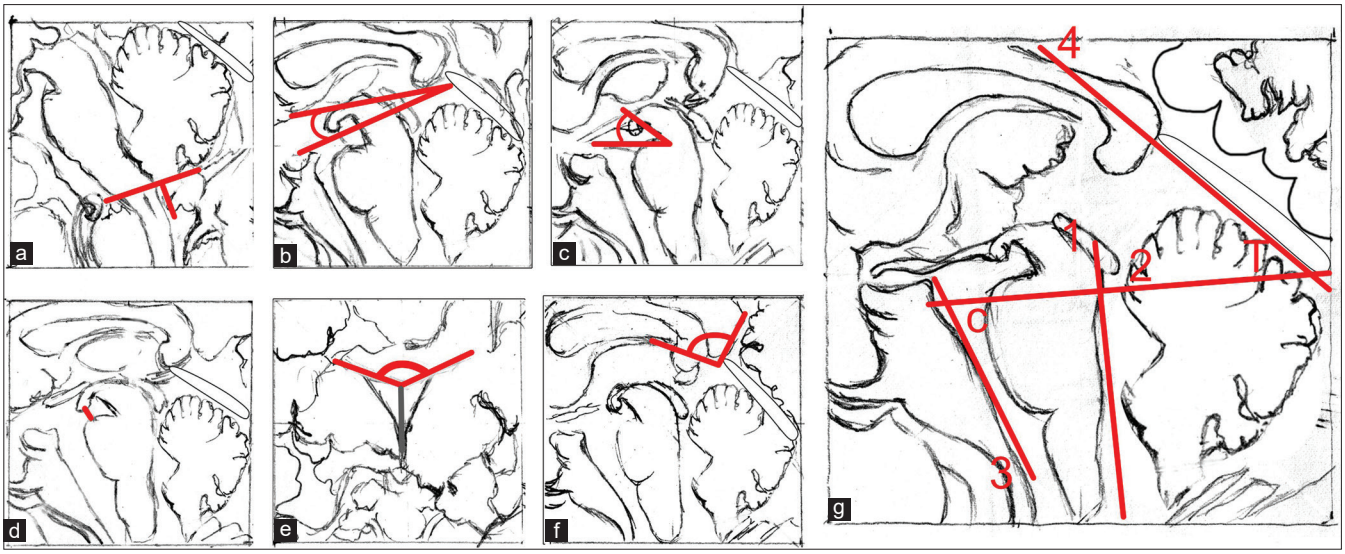

Figure 7: Magnetic resonance imaging showing intracranial anatomical measurements - brain sagging TE, slope of the third ventricular floor, pontomesencephalic angle, mamillopontine distance, lateral ventricular angle, internal cerebral vein-vein of Galen angle, and tonsillar descent (a-f) and posterior fossa morphometric measurements (note lines 1-4 and annotated T for tentorium cerebelli and C for clivus in g) that can assess causative factors that lead to acquired Chiari I malformation

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Conflicts of interest

There are no conflicts of interest.

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