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MR Imaging of Arnold Chiari Malformations

*Dr. Parveen Chandna[1], Dr. Anju Dhiman[2] , Dr. Dalip Singh Dhiman[3] , Dr. Nazir Ahmad Pandit [4]
Assistant Professor [1], Assistant Professor [2] , Professor and HOD[3], Medical Superintendent [4]
Department of Radio-Diagnosis,
N.C. MEDICAL COLLEGE AND HOSPITAL, ISRANA, PANIPAT, HARYANA , INDIA
Corresponding Author: Dr. Parveen Chandna*

Abstract
MR imaging especially complements USG due to its exceptionally high resolution, multiplanar capabilities, superior soft tissue contrast and enlarged field view. The earliest prenatal diagnosis has been achieved with the help of ultrasounds at 18 weeks followed by MRI at 23 weeks. MRI (pre and postnatal) depicts the scalloping of the clivus, small posterior fossa with hypoplastic cerebellum , large encephalocele , the low lying tentorium, hydrocephalus and the dysgenesis of the corpus callosum. Fetal MRI has been found to be complementary to the ultrasonography which further reaffirms additional concrete findings in its vicinity such as migrational abnormalities or callosal anomalies. However, MRI can further establish the extent of cerebellar herniation into the spinal canal. MRI of the head and cervical spine establishes and confirms the diagnosis, the same has been the modality of choice in evaluating Chiari I as the same depicts cerebellar tonsillar descent bigger than 5 mm below the foramen magnum (McRae line). Furthermore , shrunken posterior fossa and syrinx can be well visualised. Based on the size of the syrinx dire need has been felt to encompass whole spine MRI modality .

Key words: *Chiari malformation, posterior fossa, cerebellum, inferior herniation and cerebellar tonsillar herniation.*

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I. INTRODUCTION

Arnold-Chiari malformation is the significant term assigned to a special category of deformities related to the hindbrain and posterior fossa resulting in cerebellar tonsillar herniation through the foramen magnum. Atrophy of the cerebellum may be associated with other intracranial or extracranial deformities like encephalocele, spinal dysraphism , syrinx and hydrocephalus [1,4].

TYPES OF ARNOLD CHIARI MALFORMATION -

CHIARI 0 MALFORMATION : Inferior herniation of cerebellar tonsils 3mm or less below the foramen magnum, Syrinx and presence or absence of Crowding at foramen magnum:

CHIARI I MALFORMATION : Most common type affects both children and adults. However , the vermis, medulla and fourth ventricle have been detected as quite normal or only with minimal deformity occasionally. Inferior herniation of cerebellar tonsils through the foramen magnum into the upper cervical spinal canal (5mm or more tonsillar descent in patients with age>15 years) and (6mm or more tonsillar descent in patients with age<15 years). The cerebellar tonsils obstruct the flow of CSF and may cause increase of fluid in the spinal cord(Syringomyelia) and osseous anomalies have been well detected in 25 percent patients particularly in the form of basilar invaginations , atlanto-occipital assimilation and klippel-feil anomaly.[2,5]

CHIARI 1.5 : Inferior herniation of cerebellar tonsils (12mm or more tonsillar descent, elongation and displacement of 4th ventricle and brainstem especially medulla and small posterior fossa.

CHIARI II MALFORMATION: present at birth and affects infants .It can be revealed in the form of herniation in the inferior portions of the vermis, fourth ventricle and medulla into the spinal canal, kink in the medulla and displacing normal structures. The same is associated with small posterior fossa, petrous ridges, fenestrated falx, concave clivus and low lying transverse sutures. It is visualized following birth defect myelomeningocele ,a form of spina bifida.

However, other malformations include syringohydromyelia , both the tonsils and brainstem which have been herniated inferiorly into the spinal canal to occlude CSF flow in the brain and thus responsible for hydrocephalus and segmentation anomalies .[3,6,8] **THIS TYPE IS CALLED ARNOLD CHIARI MALFORMATION.**

Chiari III Malformation: severely affects infants, Inferior herniation of the hindbrain below the foramen magnum in association with the birth defect occipital encephalocele , further in association with innumerable distinguished features of the Chiari II malformation and spina bifida . MR has been regarded as the best modality to depict all sorts of ACM.[7,9,10]

CHIARI IV: affects infants. Hypoplasia/aplasia of the cerebellum has been particularly seen with spina bifida.[11,12]

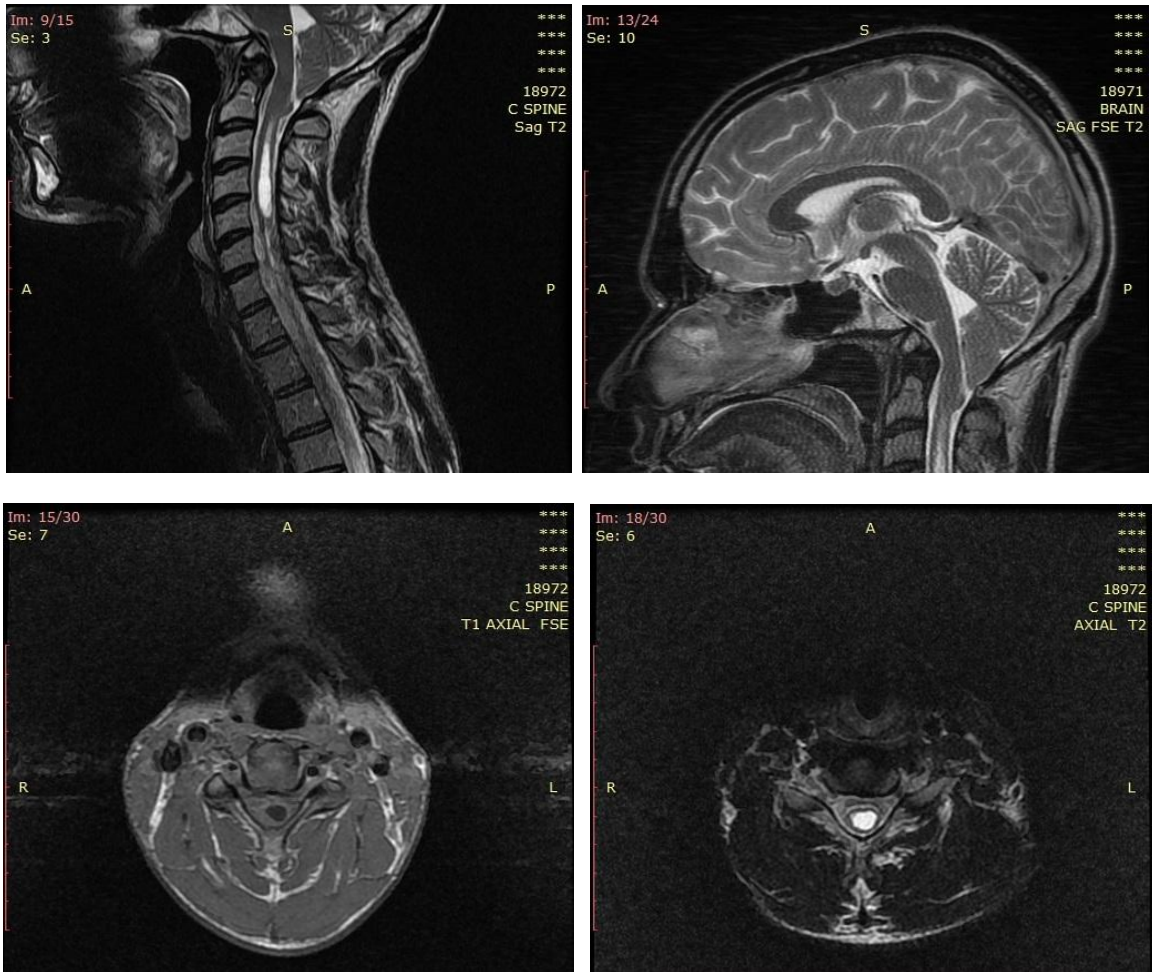
II. Material And Methods :

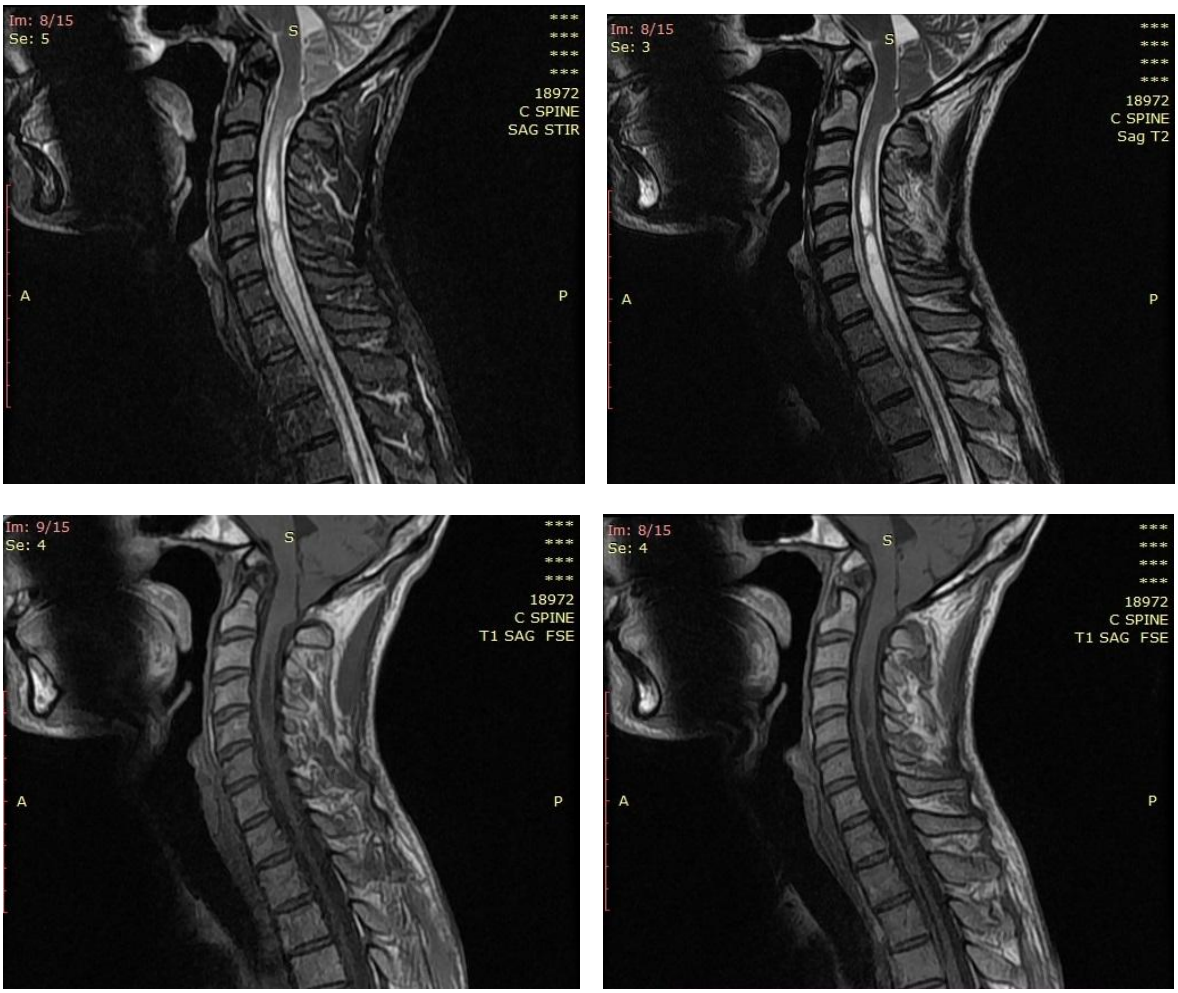
The study had been carried out in the Department of Radiodiagnosis, N .C. MEDICAL COLLEGE AND HOSPITAL, ISRANA, PANIPAT, HARYANA , for a period of 6 months from APRIL 2022 to October 2022. After taking a brief note of properly informed written consent and complete history, thorough clinical examination was done and these patients were subjected to MRI scan. To explicitly explain the utility of **GE SIGNA EXCITE 1.5T** in early detection and diagnosis.

III. Results

Case report

A 30 year old male patient visited medicine OPD with chief complaints of mild posterior headache, dizziness, nausea and vomiting for the last 1 month. Patient also complained related to neck pain, deteriorating vision and problems of imbalance. Numbness and weakness in left upper limb and both lower limbs for 1.5 years. The weakness was progressive in nature. Poor hand coordination with difficulty in swallowing since 15 days. On examination , both sensory as well as motor involvement had been noted.





CHIARI MALFORMATION TYPE 1.5-

There is caudal herniation of b/l cerebellar tonsils and medulla below foramen magnum. The cerebellar tonsils seen extending approx. 16mm (CC) below the foramen magnum with kinking and flattening of medulla. Extensive syringohydromyelia involving cervical and thoracic spine extending from C2 to upper dorsal vertebral level especially marked at the level of C3-C7 vertebrae where cord is expanded with thinning of cord parenchyma. Few septations are noted within the syrinx.

TABLE : ARNOLD CHIARI MALFORMATIONS (TYPES)

ARNOLD CHIARI MALFORMATIONS -TYPES	MRI FINDINGS
CHIARI 0	1. Inferior herniation of cerebellar tonsils 3mm or less below the foramen magnum 2. Syrinx 3. Presence or absence of Crowding at foramen magnum
CHIARI 1	1. Inferior herniation of cerebellar tonsils (5mm or more tonsillar descent in patients with age>15 years) and (6mm or more tonsillar descent in patients with age<15 years) 2. Syringomyelia 3. 4th ventricle, vermis, medulla remains in normal position or only with minimal deformity
CHIARI1.5	1. Inferior herniation of cerebellar tonsils (12mm or more tonsillar descent) 2. Elongation and displacement of 4th ventricle and brainstem especially medulla. 3. small posterior fossa
CHIARI 2 **ARNOLD CHIARI MALFORMATION**	1. Inferior Herniation of the cerebellar tonsils 2. syringohydromyelia 3. Hydrocephalus 4. Kink in the medulla 5. Myelomeningocele. 6. small posterior fossa

	7. and segmentation anomalies .
CHIARI 3	1. Inferior herniation of the hindbrain below the foramen magnum with occipital encephalocele . 2. spina bifida
CHIARI 4	1. Hypoplasia/aplasia of the cerebellum 2. Spina bifida.

IV. DISCUSSION AND CONCLUSION

Magnetic resonance imaging has been considered as one of the noninvasive diagnostic modality especially employed to evaluate the spinal cord, CSF and brain. MRI can establish the extent of cerebellar herniation into the spinal canal. The herniated cerebellar tonsils through the foramen magnum into the spinal canal occlude the normal flow of CSF. Hence, this obstruction is largely responsible for increased fluid in the brain (HYDROCEPHALUS) or in the spinal cord (SYRINGOMYELIA). Chiari I and 1.5 has been duly evaluated and further confirmed through magnetic resonance imaging (MRI) especially in a child or adult. Whereas, Chiari II to IV have been evaluated initially by ultrasound in utero followed by fetal MRI for detailed evaluation and confirmation.[8,13,14]

Fetal MR imaging is generally carried out following identification of an anomaly on prenatal US but confirmation can be made out only when familial genetic syndrome exists. Both sides of the fetal brain and posterior fossa anatomy have been clearly observed without artifact from calvarial ossification. MR imaging has least influence generated through amniotic fluid scarcity or maternal obesity. MR imaging facilitates to establish additional diagnostic features in half of the cases and altered prognostic information in about one fifth cases, and clinical management in about 35 percent of cases.[11,15,18]

US has been regarded as the first modality of choice in pregnancy and fetal imaging, being depicted as noninvasive, devoid of ionizing radiation, availability accessible to a larger extent and real-time associated with color Doppler capabilities. It is quite safe, envisaging standard guidelines for obstetric scanning .US can be performed throughout the second and third trimester, as early as 12 weeks, but most examinations have been carried out amid gestation in abundance at about 20 weeks.[16,17,20]

Cine MRI: has to be carried out to evaluate CSF flow. CSF is forced out of the ventricles of the brain with succesive heart beat into the cisterna magna(CM) and further descending into spinal canal. CSF flow takes a reverse route when the heart relaxes. This cine MRI captures the fluid movement and can evaluate blocking of the back-and-forth flow of CSF between the brain and spine through Arnold chiari malformation.[19,21,24]
Fetal MR imaging has established our inculcated understanding related to this complex multilevel malformation, which needs to be reclassified as a fetal disruption rather than a unique malformation. Further investigations are quite important due to the unified theory which may explicitly depict the combination of the spinal dysraphia and the Chiari 2 malformation, but certainly not relevant, being unable to reveal the additional cerebral findings such as migrational abnormalities or callosal malformations. Advanced fetal MR imaging including diffusion tensor imaging with tractography can create a usual specific impact related to this complex fetal abnormality.[22,23]

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