NEUROLOGY

NEUROIMAGING

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Abstract: *Availability of neuroimaging facilities has made the evaluation of neurological problems easier in the last few decades. Computed tomography scan of brain is the initial choice in very sick children because of its wider availability, faster turnaround time and lower cost. Cranial ultrasonography is an important modality in the follow up of infants in the postnatal period, particulary in the evaluation of hypoxic ischemic encephalopathy, subependymal- periventricular- intraventricular hemorrhage and hydrocephalus. It is used as a point of care investigation by neonatologists. Absence of radiation exposure and precision makes magnetic resonance imaging the modality of choice in emergency situations. But the disadvantages are the need for sedation or brief anaesthesia, longer procedural time and cost. But benefits outweigh the disadvantages and additional tools like Magnetic resonance angiography, Magnetic resonance venography and Magnetic resonance spectroscopy add precious information for further evaluation.*

Keywords: *Neuroimaging, Cranial ultrasonography, CT, MRI, MRV, MRA, MRS.*

Neuroimaging plays an important and growing role in the diagnosis and management of neurological diseases in children. Neuroimaging falls into two broad categories (a) structural imaging deals with the structure of the brain and (b) functional imaging is used to measure the brain function; it is useful for diagnosing metabolic diseases and as a research tool in cognitive neurosciences including neuropsychology.

Modalities of neuro imaging

The major imaging modalities for structural and functional evaluation of the central nervous system (CNS) are ultrasonography (USG), computed tomography (CT), magnetic resonance imaging (MRI) and nuclear medicine techniques. USG and CT provide rapid screening for gross macro structural abnormalities. MRI often provides the most definitive macro structural, micro structural and functional imaging information. Nuclear medicine techniques may offer additional functional data.

X-ray skull

Plain films of the skull can show fractures of the skull, erosions, hyperostosis, calcifications, overriding of sutures (craniostenosis), widening of sutures (raised intra cranial pressure) and inflammation of the sinuses and mastoids. Skull X-rays are rarely used nowadays because of their low yield and have been replaced by computed tomography.

Ultrasonogram cranium

Cranial ultrasonography is the most frequently used neuroimaging modality in prenatal and perinatal period. Developmental anomalies of central nervous system such as holoprosencephaly, lissencephaly, encephalocele, Dandy walker malformation, spina bifida, hydrocephalus etc., can be detected by sonography. In newborns, open anterior fontanel provides an excellent window to visualize the infant brain. It is helpful in the evaluation of hypoxic ischemic encephalopathy, subependymal- periventricular-intraventricular hemorrhage, hydrocephalus and major migrational anomalies such as agyria- pachygyria complex.

Computed tomography

Computed tomography has been available since 1970s for clinical use and is widely available for emergencies and medically unstable patients. Lesions appearing as low density (appearing dark) include edema, infarct, inflammation, necrosis and cysts. High density (appearing bright) lesions include calcifications and hemorrhage. Fat containing lesions appear less dense and air appears as the lowest density. Modern CT devices allow appropriate visualization of midline structures and the ventricular system, providing sufficient diagnostic yield for herniation and hydrocephalus. However, CT is generally suboptimal for imaging of structures in the posterior fossa and brain stem. The greatest practical advantage of CT, particularly over MRI, is its greater availability, much faster imaging time, lower cost and in the context of known or suspected metallic foreign bodies, when MRI is contraindicated.[1]

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Magnetic resonance imaging

Magnetic resonance imaging is the most powerful neuro imaging modality for adults and children. Painless and noninvasive, MRI uses powerful magnets and radio waves to produce excellent detailed images of the brain. It provides multiplanar imaging with high resolution without repositioning the patient. However it has less tolerance for patient movement and hence sedation is required for most of the children.[2]

Cerebrospinal fluid, muscle, deoxyhemoglobin, hemosiderin and substances with long T1 relaxation times appear dark on T1W (T1 weighted) images. Tissues with short T1 relaxation time such as fat, methemoglobin appear bright on T1W images. On T2W images, structures with long T2 relaxation times such as CSF, edema, infarct, tumours, demyelination appear bright whereas muscle, deoxyhemoglobin, and hemosiderin appear dark because of short T2 relaxation times.

Patients with paramagnetic metallic implants such as aneurysm clips, brain stimulators (deep brain stimulator or vagus nerve stimulator) or pacemakers are ineligible for MRI. These metallic foreign bodies could move or heat up, resulting in injuries to surrounding tissues. In a closed type MRI system, patients with claustrophobia may be unable to remain still during the procedure. Similarly, children or patients with cognitive or behavioral impairments may not be able to undergo the test. This obstacle can sometimes be overcome with the use of sedation or with use of newer open MRIs.[3]

The various neuroimaging findings in different neurological conditions are discussed below:

Neuro infections

Viral infections: Viral infections of central nervous system can take the form of meningitis, encephalitis, encephalomyelitis or encephalomyeloradiculitis. Neurotropic viruses have special predilection for certain regions of the neuraxis producing characteristic imaging features that may aid in prioritizing diagnostic considerations in the differential diagnosis (Table I).[4,5]

Bacterial meningitis: Neuroimaging is neither sensitive nor specific for the diagnosis of meningitis. Abnormal enhancement of the pia and arachnoid (leptomeninges) caused by inflammatory breakdown of the blood-brain barrier is seen in only 50% of patients.[6] Neuroimaging is most useful for excluding herniation before lumbar puncture and for detecting complications such as subdural effusion / empyema (Fig.5), ventriculitis (Fig.6&7), hydrocephalus (Fig.8), sinus venous thrombosis, infarct and cerebral abscess. Similar to parenchymal abscess, pus in the ventricles or epidural or subdural space shows restricted diffusion, a finding that is useful for distinguishing simple effusion from empyema in the context of acute bacterial meningitis.[7]

Table I. Characteristic imaging features in certain viral infections

Viral infection	Characteristic imaging features
Herpes simplex virus (Fig.1)	Inferomedial temporal lobe, insula, limbic system
Japanese encephalitis virus (Fig.2)	Thalamus (90%), basal ganglia, substantia nigra
West Nile virus	Thalamus, basal ganglia, cranial nerves, spinal cord, cauda equina
Dengue virus	Globus pallidi, thalami, hippocampus, temporal lobe, pons, spinal cord
Chikungunya virus	Cerebral white matter, cauda equina
Rabies virus	Brainstem, hippocampus, limbic system, hypothalamus
Human immunodeficiency virus	Symmetric periventricular white matter involvement, cerebral atrophy
Entero virus (Fig.3)	Medulla, pons, midbrain, splenium of corpus callosum
Epstein-Barr virus	Cerebellum, pons, cerebral peduncles, splenium of corpus callosum
Measles virus	Bilateral striatal necrosis, ADEM pattern
Varicella encephalitis	Cortex, gray- white matter junction, vasculopathy
Influenza virus (Fig.4)	Acute necrotising encephalopathy - diffuse brain edema, symmetric thalami, brainstem, cerebellum

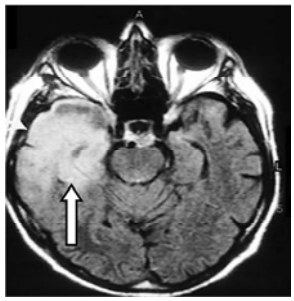

Fig.1. Axial MR FLAIR showing right temporal lobe involvement suggestive of HSV encephalitis

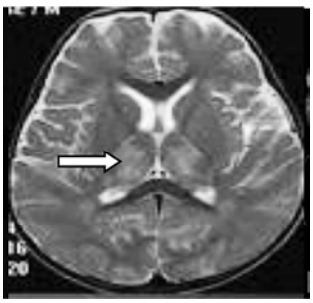

Fig.2. Axial T2 MR showing bilateral asymmetrical thalamic involvement in a child with JE encephalitis

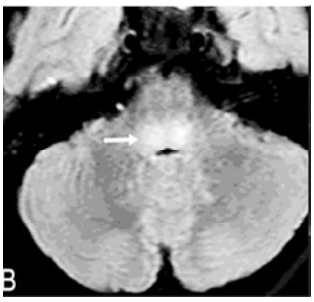

Fig.3. Axial FLAIR sequence showing posterior medulla involved in a child with entero virus encephalitis

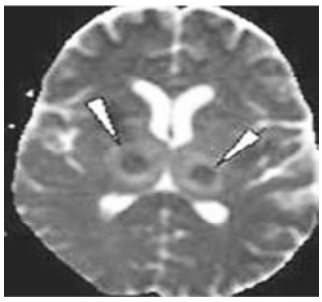

Fig.4.Axial T2MR showing bilateral symmetrical thalamic involvement with central areas of hemorrhage suggestive of acute necrotising encephalitis

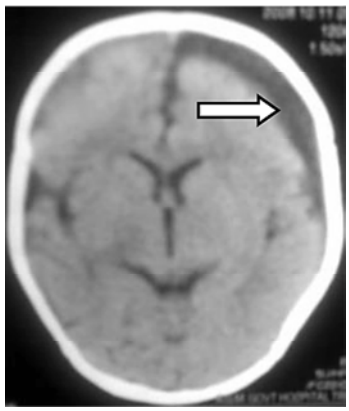

Fig.5. Plain CT brain - left frontal subdural effusion in bacterial meningitis

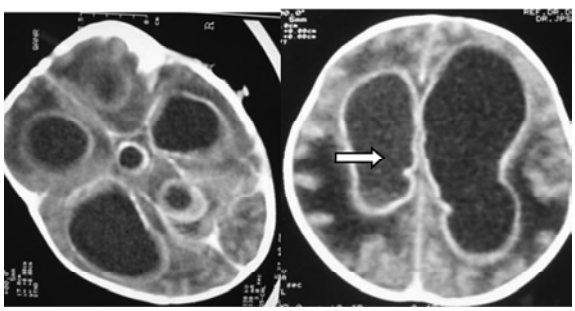

Fig.6&7. Contrast CT brain showing enhancement of ependymal lining of ventricles suggestive of ventriculitis

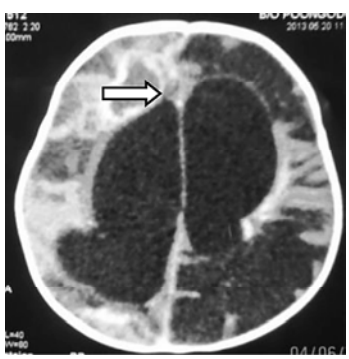

Fig.8. Contrast CT brain - Right frontal abscess with hydrocephalus

Tuberculous meningitis: Neuroimaging manifestations of tuberculous meningitis (TBM) include characteristic thick or nodular enhancement in the basal cisterns, although this finding may also be noted in meningitis due to other granulomatous (fungus or sarcoid) and neoplastic (carcinoma or lymphoma) disorders. Other possible sequelae of tuberculous meningitis such as infarcts, hydrocephalus, tuberculomas may be visualized.[8] The triad characterized by basal meningeal enhancement, hydrocephalus (Fig.9) and deep infarcts is highly suggestive of tuberculous meningitis.[9]

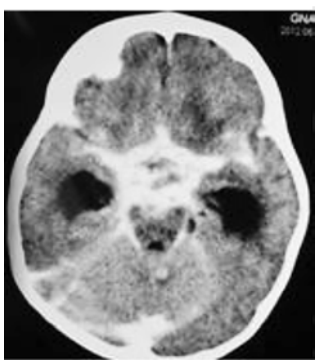

Fig.9. Contrast CT brain showing thick basal exudates and hydrocephalus in TBM

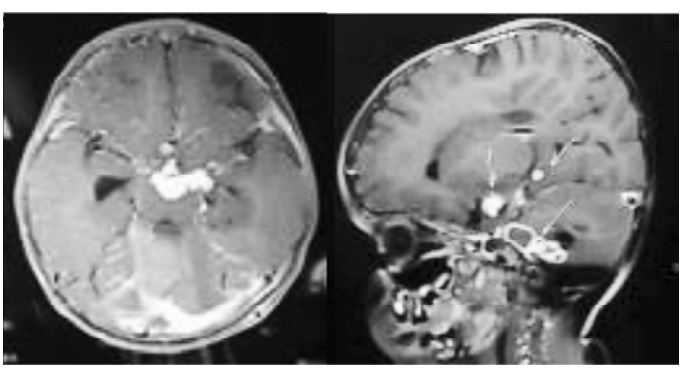

Fig.10. Post Gado contrast MR axial and sagittal views showing thick basal exudates, tuberculomas in interpeduncular region, pons and hydrocephalus

The characteristics of tuberculomas in MRI studies depend on whether they are caseous and the caseous matter has a solid or liquified center (Fig.10).[10,11] Non-caseous tuberculomas usually have a hypointense signal on T1 and a hyperintense signal on T2, with homogenous enhancement after gadolinium administration. Solid caseous granulomas have iso or hyposignal on both T1- and T2- weighted sequences.[12] Rajshekar, et al considered the differences between focal granulomatous lesions caused by cysticercosis and tuberculomas in individuals with seizures who have come from endemic regions and inferred that it would to be a difficult task without biopsy, but the occurrence of increased intracranial pressure and focal neurologic deficit, size greater than 2 cm, irregular outline and association with deviation of midline cerebral structures favor the possibility of tuberculoma.[13]

Tuberculomas are reported to demonstrate lipid peaks at 0.9 ppm, 1.3 ppm, 2.0 ppm and 2.8 ppm at MR spectroscopy and these peaks are believed to correlate with the presence of the high lipid content of the mycolic acid in the mycobacterial cell wall.

Neurocysticercosis

Radiographic appearance of neurocysticercosis mirrors the pathophysiologic stage of cyst degeneration. In vesicular stage, viable cysts show no surrounding edema or rim enhancement, however scolex may be seen (Fig.11). Visualisation of scolex in computed tomography or MR imaging is a strong evidence for neurocysticercosis. Colloid vesicular stage hydrocephalus shows ring like pattern of contrast enhancement with surrounding edema (Fig.12). The granular nodular stage begins with cyst retraction and formation of a granulomatous nodule and calcified nodular stage (Fig.13) is the last stage with gliosis and calcification.[14] Table II shows distinguishing features between tuberculoma and neuro cysticerosis.

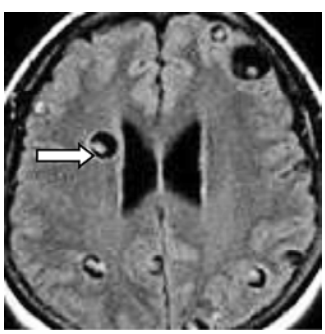

Fig.11.Axial MR FLAIR image shows multiple cysticerci in vesicular stage

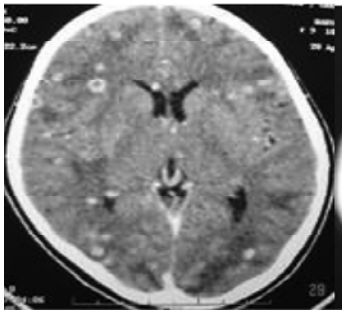

Fig.12. Contrast CT showing ring enhancing and disc enhancing lesions with surrounding edema

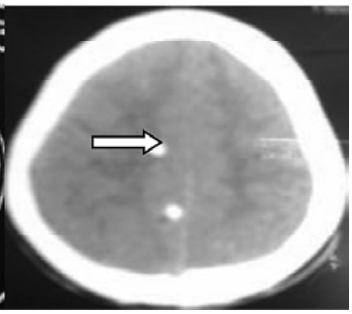

Fig.13. Plain CT showing calcified granulomas

Table II. Tuberculoma and neurocysticercosis - Distinguishing features in MRI

Tuberculoma	Neurocysticercosis
Size >20 mm	< 20mm
Irregular thick outline	Rounded thin outline
Marked perilesional edema	Less surrounding edema
Infratentorial or supratentorial	Predominantly supratentorial
Midline shift more likely	Midline shift less likely
Hypointense core in T2W images	Hyperintense core, eccentric dot best seen in fluid attenuated inversion recovery (FLAIR) or diffusion weighted imaging (DWI)
Lipid peak present	Lipid peak absent

Neurocutaneous syndromes

Phakomatoses or neurocutaneous syndromes are a heterogeneous group of congenital disorders with variable degree of penetration, primarily involving structures derived from the embryological neuroectoderm (central nervous system and peripheral nerves). In addition, surface ectodermal structures like skin, eye and other systems may also be involved.[15] Various neurocutaneous syndromes and their imaging findings are depicted in Table III.

Developmental anomalies

Central nervous system anomalies represent one of the most frequently involved structures with an estimated incidence of 1 per 100 births.[16] Cortical malformations must

Table III. Neurocutaneous syndromes and their imaging findings

Neurocutaneous syndrome	MR imaging findings
Sturge -Weber syndrome (Fig.14)	CT - subcortical calcification usually in parieto occipital region - parenchymal volume loss MRI T1contrast - prominent leptomeningeal enhancement in affected area - due to congested internal cerebral veins called 'pial angiomatosis', resulting in venous congestive ischemia with infarction and obliteration of cerebral parenchyma - enlarged ipsilateral choroid plexus
Tuberous sclerosis (Fig.15,16)	Subependymal hamartomas-(88% calcified), high in T1 and iso to high in T2, cortical/subcortical tubers- high in T2 and low in T1, subependymal giant cell astrocytoma - heterogeneous intensity on T1, T2 with contrast enhancement near the foramen of Monro leading to obstruction and hydrocephalus
Neurofibromatosis type I (Fig.17)	Focal areas of signal intensity (FASI) or unidentified bright objects (UBO) in deep white matter and basal ganglia or corpus callosum, i.e. areas of T2/FLAIR hyperintensity with no contrast enhancement, optic nerve glioma or optic pathway glioma, sphenoid wing dysplasia
Neurofibromatosis type II (Fig.18)	Intracranial vestibular schwannoma, intracranial and spinal meningioma, intraspinal-intramedullary ependymoma

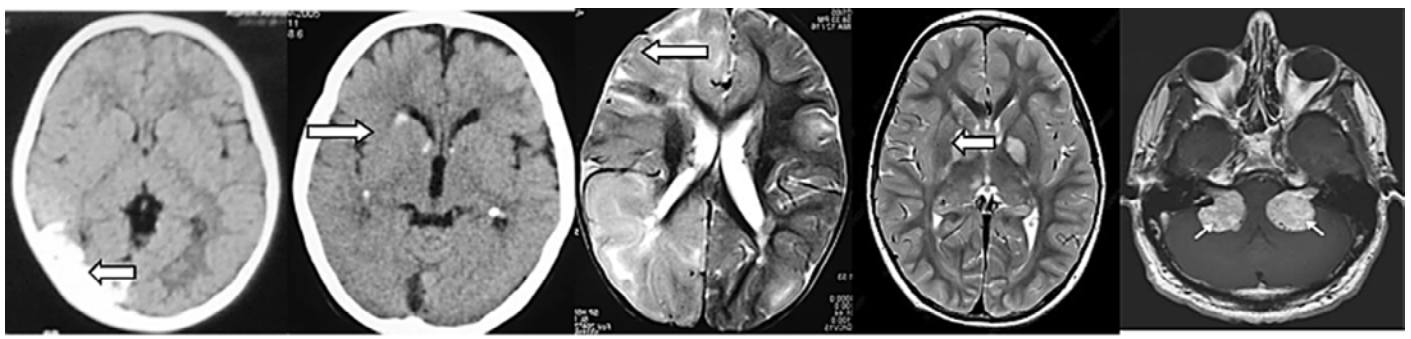

Fig.14. Plain CT shows calcified pial angioma in right parieto occipital region suggestive of Sturge Weber Syndrome

Fig.15. Plain CT brain shows subependymal calcified tubers in a child with tuberous sclerosis

Fig.16. Axial T2MR showing subependymal tubers and cortical tubers

Fig.17.Axial T2MR showing FASI in the basal ganglia in Neurofibromatosis type I

Fig.18.Axial MR FLAIR sequence showing bilateral acoustic neuromas in neurofibromatosis type II

be ruled out in essentially every pediatric patient with developmental delay or epilepsy. It is very difficult to make a diagnosis of congenital brain malformation based on clinical findings and use of CT or MRI is essential in these cases. Imaging findings in different developmental anomalies of central nervous system are described in Table IV.[17,18]

Inborn errors of metabolism

Neuroimaging is a particularly useful adjunct in the diagnosis of inborn errors of metabolism and should not be delayed until laboratory results are known, as it may provide early crucial clues to the diagnosis (Table V).[19,20]

Degenerative disorders - leucodystrophies

Magnetic resonance imaging has become the primary imaging modality in children with leukodystrophy and plays an important role in the identification, localization, and characterization of underlying white matter abnormalities in the affected patients. Systematic analysis of the finer details of involvement may help to narrow down the diagnosis (Table VI).[20]

Gray matter degenerative disorders

Generally imaging findings in gray matter degenerative disorders are nonspecific, usually diffuse

Table IV. Developmental anomalies of central nervous system - Imaging findings

Congenital malformation	Imaging findings
Dandy Walker malformation	Complete or partial vermian agenesis, cystic dilatation of fourth ventricle and enlargement of the posterior fossa with elevation of the transverse sinus, tentorium, and torcula
Arnold Chiari malformation (Fig.19-22)	Type 1 - Herniation of cerebellar tonsils into cervical canal - descent of >6 mm is abnormal Type 2 - Herniation of vermis, tonsils and medulla Myelomeningocele (nearly 100%) Type 3 - Type 2 + occipital encephalocele
Arachnoid cyst (Fig.23)	Extracerebral mass containing cerebrospinal fluid encircled by walls composed of arachnoid membrane
Callosal agenesis (Fig.24)	Complete callosal agenesis shows high riding third ventricle with spoke-like orientation of gyri around it, lateral ventricles widely separated, parallel and non-converging.
Joubert syndrome (Fig.25)	Small dysplastic or aplastic cerebellar vermis, prominent thickened elongated superior cerebellar peduncles giving characteristic molar tooth sign, key hole fourth ventricle
Lissencephaly (Fig.26)	No or few cerebral gyri and sulci Type I lissencephaly-typical figure eight configuration of brain with thickened cortex, flat broad gyri and shallow sylvian fissures Type II lissencephaly - thickened cortex having polymicrogyria
Schizencephaly (Fig.27-29)	Grey matter-lined cleft extending from the ependymal surface to pia mater, closed lip - cleft walls are in apposition, open lip - cleft walls are separated and filled with CSF
Heterotopia (Fig.30)	Presence of normal neurons at abnormal sites – nodular (common), band or laminar- a layer of neurons interposed between ventricle and cortex
Holoprosencephaly (Fig.31-34)	Alobar - single midline ventricle, absent interhemispheric fissure, falx cerebri, fused thalami and basal ganglia Semilobar - rudimentary occipital and temporal horns interhemispheric fissure - absent anteriorly thalami and basal ganglia partially separated Lobar - ventral portions of frontal lobes fused rudimentary frontal horns of lateral ventricles
Hemimegalencephaly (Fig.35)	Hamartomatous overgrowth of a part or all of one cerebral hemisphere

cortical atrophy. Specific findings may help us to narrow down the diagnosis in certain disorders (Table VII).

Demyelinating disorders

Magnetic resonance imaging plays an important role in the diagnosis, delineating the type, extent of demyelination and in the follow up of these disorders. Typical MRI findings in different types of demyelinating disorders are depicted in Table VIII.[21,22,23]

Stroke

Neuroimaging is essential for diagnosis and differentiation of stroke from stroke mimics such as hypoglycemia, demyelinating disorders, tumors, posterior reversible leukoencephalopathy syndrome and hemiplegic migraine. In general, the presence of diffusion restriction in the distribution of an arterial territory confirm stroke, although other entities such as brain tumors, abscesses, white matter diseases and seizures can exhibit reduced

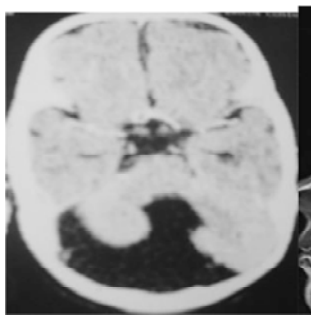

Fig.19. Plain CT brain showing Dandy Walker malformation

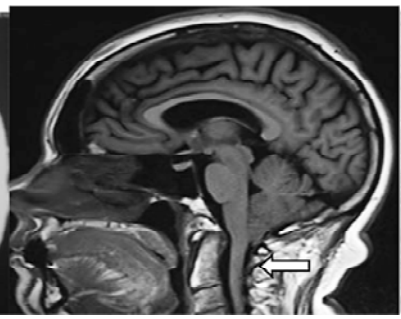

Fig.20. Sagittal T1 MR shows descent of cerebellar tonsils into cervical canal - Chiari I malformation

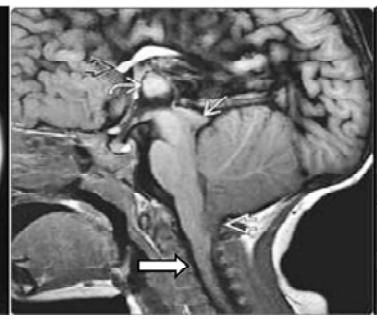

Fig.21. SagittalT1 MR showing descent of tonsils and medulla into cervical canal - Chiari II malformation

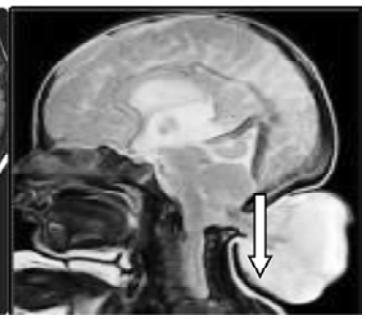

Fig.22.Sagittal T2 MR showing descent of medulla, tonsils with occipital encephalocoele - Chiari III malformation

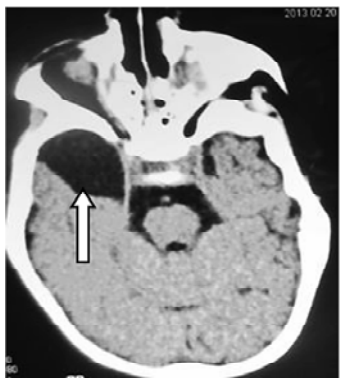

Fig.23. Plain CT brain showing arachnoid cyst in right temporal region

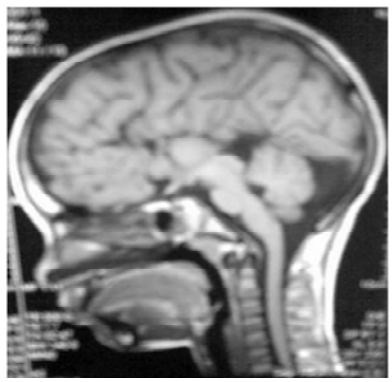

Fig.24. Sagittal T1 MR showing absence of corpus callosum

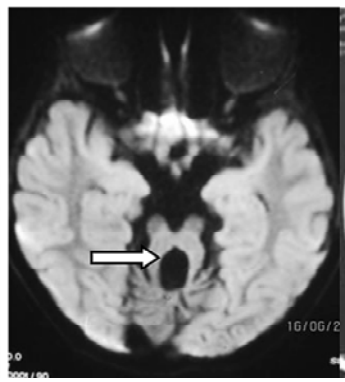

Fig.25. Axial MR FLAIR showing characteristic molar tooth sign suggestive of Joubert syndrome

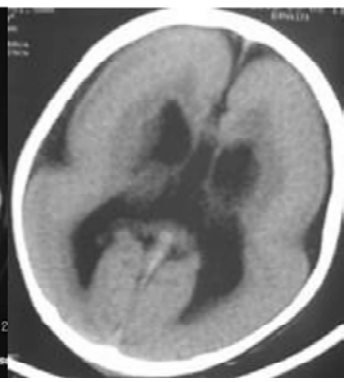

Fig.26. Plain CT brain showing smooth surface of brain with shallow sylvian fissure - figure of 8 appearance in Lissencephaly

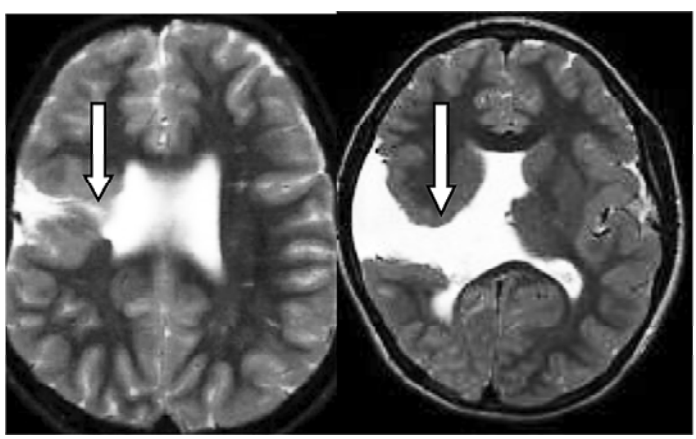

Fig.27. Axial T2 MR showing closed lip schizencephaly

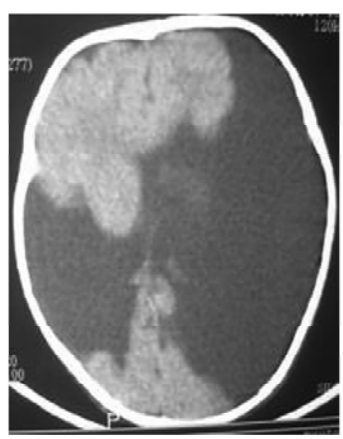

Fig.28. Axial T2 MR showing unilateral open lip schizencephaly

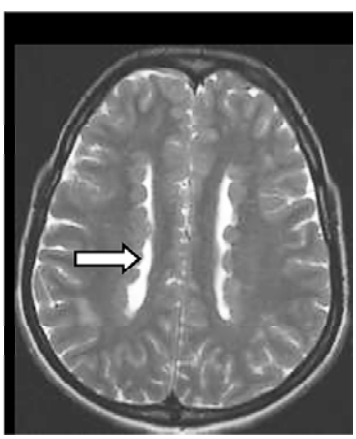

Fig.29. Plain CT brain showing bilateral schizencephaly

Fig.30. Axial T2MR showing nodular heterotopia

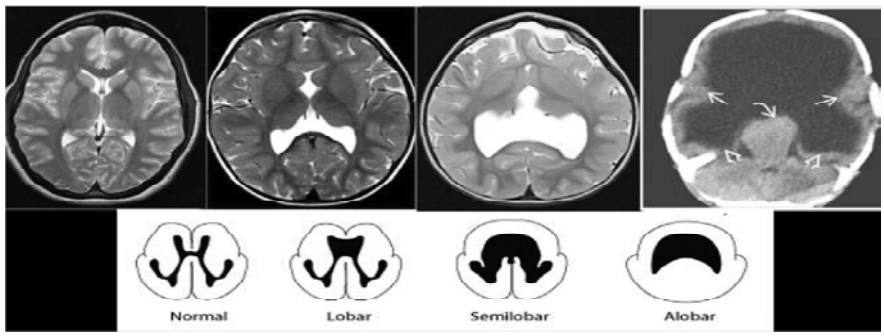

Fig. 31-34. Types of Holoprosencephaly

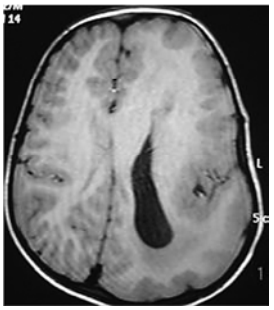

Fig.35. Axial T1 MR showing left hemimegalencephaly

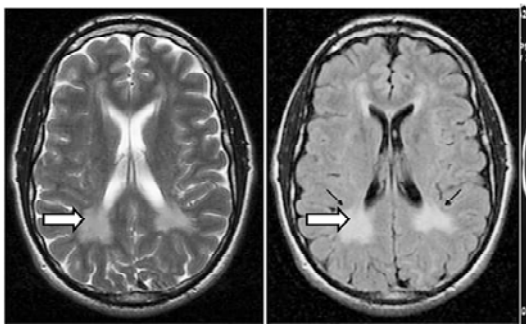

Fig.36(a & b).Phenyl ketonuria Axial T2 and FLAIR MR image shows periventricular white matter changes predominantly in parieto occipital region

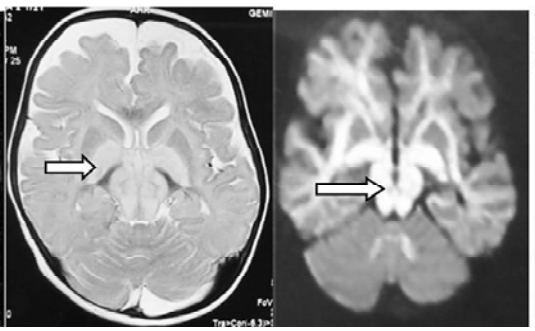

Fig.37(a & b).Maple syrup urine disease Axial T2 and DWI MR image showing high signal intensities in dorsal midbrain, cerebral peduncles and globi pallidi with diffusion restriction

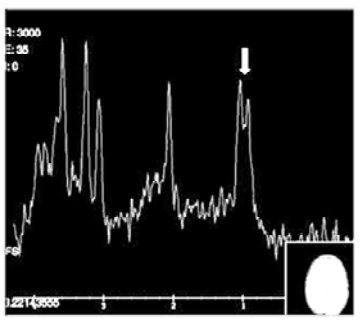

Fig.38.MR Spectroscopy abnormal wide peak at 0.9 ppm representing branched chain amino acids and keto acids

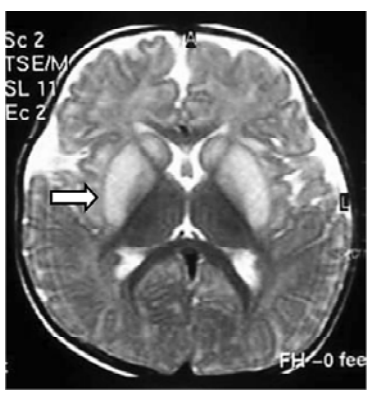

Fig.39. Methyl malonic acidemia Axial T2MR image shows bilateral symmetrical involvement of caudate and putamen

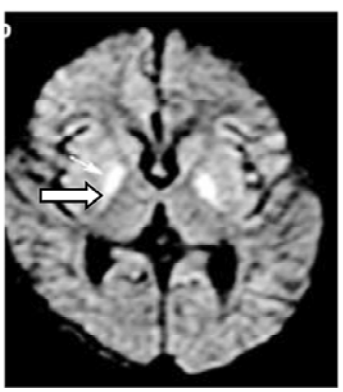

Fig.40.Propionic acidemia DWI showing diffusion restriction in both globus pallidi

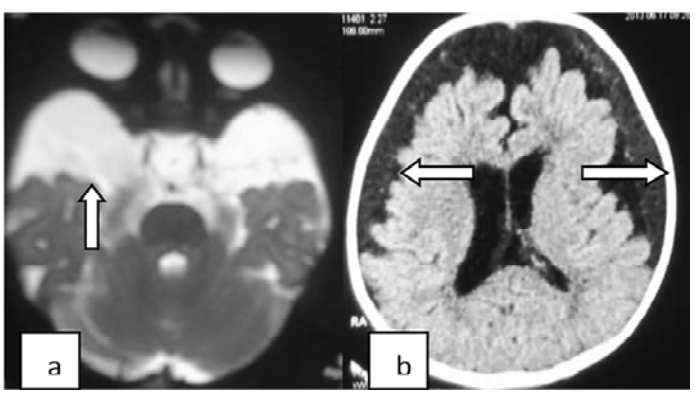

Fig.41a.Glutaric acidemia type I Axial T2MR image shows bats wing appearance

b. Axial FLAIR image showing bilateral chronic subdural hemorrhage in the same child

Table V. Important clues to diagnosis from neuroimaging in inborn errors of metabolism

Inborn error of metabolism	MRI findings	MRS findings
Phenyl ketonuria (Fig.36 a & b)	High signal intensities in periventricular/ parieto-occipital white matter	Phenylalanine peak at 7.37 ppm
Maple syrup urine disease (Fig.37 a & b / Fig.38)	Increased signal of cerebellar and perirolandic white matter, dorsal brainstem, cerebral peduncles, posterior limb of internal capsule,thalami and globi pallidi with diffusion restriction	Branched chain amino and ketoacids at 0.9 ppm
Methymalonic academia (Fig.39) Propionic academia (Fig.40)	Bilateral caudate and putamen,bilateral globi pallidi early - diffuse edemalate - hypomyelination, volume loss	
Glutaric aciduria type I (Fig.41a&b)	Bilateral fronto temporal atrophy, wide sylvian fissure - 'Bat wing' appearance, hilateral putamen involvement	
Nonketotic hyperglycinemia (Fig.42)	T2-hyperintense lesions of themyelinated white matter tracts, corpus callosal agenesis, vermian hypoplasia	Glycine peak at 3.5ppm
Sulfite oxidase deficiency (Fig.43)	Extensive cystic encephalomalacia	Sulfocysteine -3.6ppm Taurine - 3.4ppm Cysteine - 2.9 ppm
Creatine deficiency syndromes (Fig.44 a&b))	Normal	Absent or severely deficient creatine peak
Mitochondrial disorders (Fig.45 a, b & c)	T2-hyperintense lesions in dorsal midbrain, cerebral peduncles, pontine corticospinal tracts, dorsalmedulla, subcortical white matter with diffusion restriction	Lactate doublet at 1.33ppm

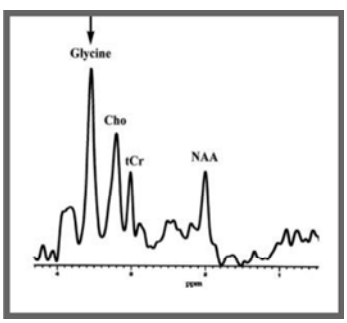

Fig.42. MR Spectroscopy showing elevated glycine peak in a neonate with non ketotic hyperglycinemia

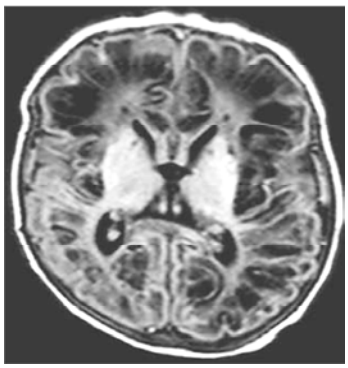

Fig.43. Sulfite oxidase deficiency Axial FLAIR MR image shows multicystic encephalomalacia

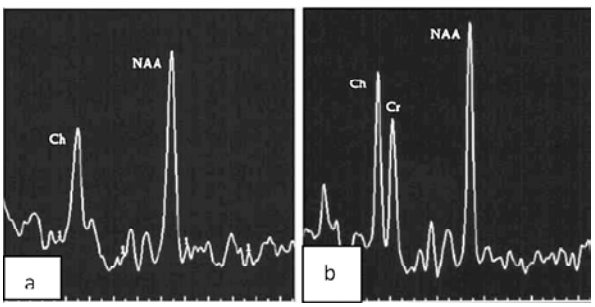

Fig.44a.Creatine deficiency disorder MR Spectroscopy showing absent creatine peak b. MR Spectroscopy in a normal child for comparison

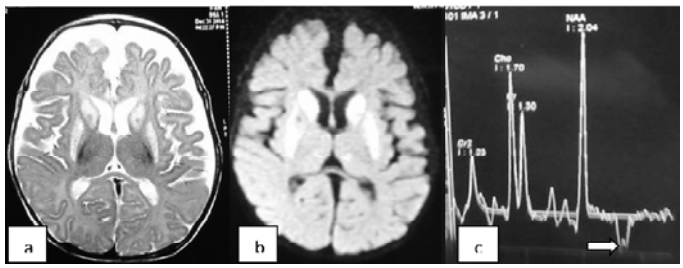

Fig.45a.Leighs encephalopathy Axial T2MR image showing bilateral symmetrical hyperintensities in basal ganglia

b.DWI showing diffusion restriction in the same areas and thalami

c.MR Spectroscopy showing inverted doublet lactate peak

Table VI. Leucodystrophies - Imaging findings

Disease	MRI findings	MRS findings
Metachromatic leucodystrophy (Fig.46 a & b)	Bilateral symmetric confluent areas of high signal intensity in the periventricular white matter with sparing of the subcortical U fibers. Tigroid or leopard skin pattern suggestive of sparing of the perivascular white matter seen. Corpus callosum, internal capsule, and corticospinal tracts, cerebellar white matter are frequently involved. No contrast enhancement.	
Krabbe disease (Fig.47 a & b)	Periventricular white matter in parieto occipital regions, thalami, corticospinal tracts, internal capsule, corona radiata, cerebellum may be involved. Optic nerve hypertrophy may be seen.	
Adrenoleucodystrophy (Fig.48 a & b)	Symmetric periventricular white matter in parieto occipital regions (peri-trigonal region), splenium of corpus callosum, cerebellar white matter. 3 zones- Inner zone - irreversible gliosis - markedly hyperintense at T2W. Intermediate zone- active inflammation- iso or hypointense in T2W and readily **enhances with contrast**. Outer zone - active demyelination moderately hyperintense at T2W.	
Alexander (Fig.49 a & b)	Extensive cerebral white matter involvement with frontal predominance, cavitations in white matter	
Canavan disease (Fig.50a & b)	Symmetric areas of homogeneous high signal intensity in T2W throughout the white matter including the subcortical U fibers, internal, external capsules, cerebellar white matter and globus pallidi.	marked elevation of NAA peak
Vanishing white matter disease (Fig.51)	Symmetric and diffuse white matter involvement with signal intensity which is close to, or similar to cerebrospinal fluid in every sequence.	Marked decrease or absence of NAA, creatine, choline peaks
Vander Knapp disease - Megalencephalic leukoencephalopathy with subcortical cysts (Fig.52)	Bilateral diffuse subcortical white matter involvement and subcortical cysts in anterior temporal lobes	

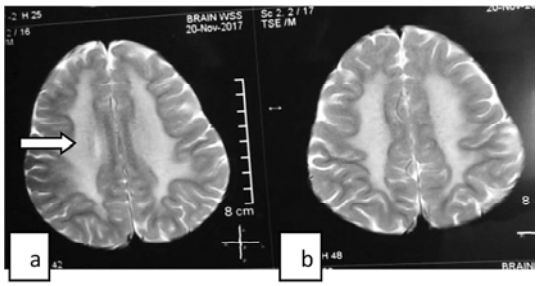

Fig.46a. Metachromatic leucodystrophy Axial T2 MR image showing bilateral symmetrical periventricular white matter hyperintensity. b. Tigroid pattern.

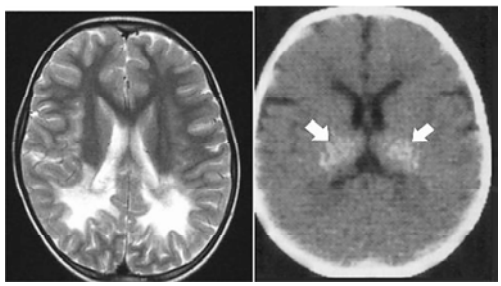

Fig.47a.Krabbe disease Axial T2 MR showing peri ventricular white matter hyperintensity

b. CT brain showing hyperdense thalami

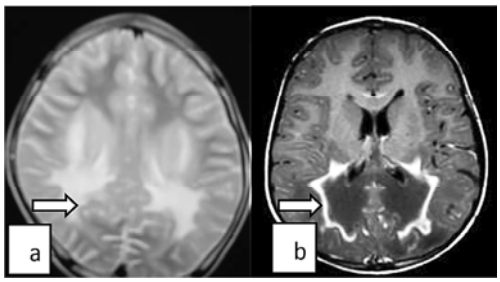

Fig.48a. Adreno leucodystrophy Axial T2 MR image shows symmetrical white matter hyperintensities in the parieto occipital region b. Axial T1 contrast showing contrast enhancement

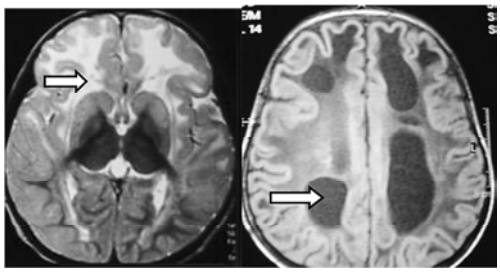

Fig.49a. Alexander disease Axial T2 MR showing frontal white matter involvement

b. Axial FLAIR showing cysts in the white matter

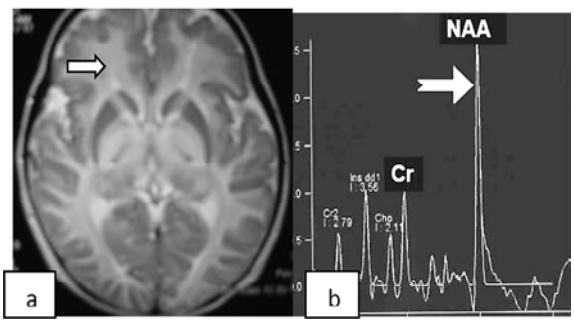

Fig.50a. Canavan disease Axial T2 MR image showing periventricular and subcortical white matter including U fibers and bilateral globus pallidi

b. MR Spectroscopy showing large NAA peak

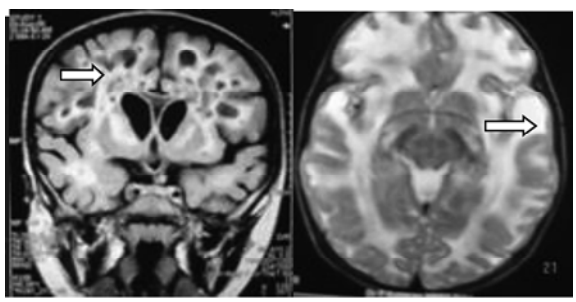

Fig.51.Vanishing White Matter disease Coronal FLAIR shows diffuse white matter involvement with cystic changes

Fig.52.Vander Knapp disease Axial T2 MR image showing diffuse white matter involvement with cyst in the anterior temporal pole

diffusivity (Fig.60 a & b). MRA of the head and neck is most often warranted to evaluate for transient cerebral arteriopathy (TCA), arterial dissection, moyamoya disease (Fig.61) and fibromuscular dysplasia.[24] In a child who is medically unstable, CT with CT angiogram (CTA) of the head and neck may be preferable. Repeat imaging may be performed to assess hemorrhagic transformation, infarct extension, mass effect, herniation, and stroke recurrence. Follow-up imaging is often performed between six weeks and three months to evaluate for progression or improvement of existing arteriopathies.[25]

Table VII. Diagnostic imaging findings in certain gray matter disorders

Gray matter disorder	MRI findings
Neuronal ceroid lipofuscinosis (Fig.53)	Diffuse cerebral and cerebellar atrophy
Mucopolysaccharidoses (Fig.54)	Prominent perivascular spaces, periventricular white matter involvement, cord compression in cranio cervical region
Fucosidosis (Fig.55 a&b)	Bilateral pallidal hyperintense signaling on T1W images and hypointense signaling on T2 W images
Panthothenate kinase associated neurodegeneration (PKAN) (Fig.56)	"Eye of the tiger"sign - central T2 hyperintense spot within the hypointense globi pallidi due to gliosis and vacuolisation in T2W images
Wilson disease (Fig.57)	Hyperintensity involving globus pallidi, putamen , caudate, thalami. “Face of the giant panda” sign in the midbrain with high signal in tegmentum and normal red nuclei

Table VIII. Neuro imaging findings in demyelinating disorders

Disease	MRI findings
Acute disseminated encephalo myelitis (ADEM) (Fig.58 a & b)	Bilateral, asymmetric, multiple poorly demarcated lesions in deep and subcortical white matter, (thalami and basal ganglia frequently affected) number of lesions varies with size ranging from <5 mm to 5 cm, in large confluent intramedullary lesions in spinal cord that extend over multiple segments are common. The degree of contrast enhancement is variable.
Multiple sclerosis (MS) (Fig.59 a & b)	Multiple well-demarcated lesions in the periventricular, juxtacortical, infratentorial, and spinal cord white matter. T1-weighted sequences may reveal "black holes" that represent complete tissue loss resulting from a previous inflammatory event
Neuromyelitis optica (NMO)	Bilateral optic nerve involvement and extension of T2 hyperintense signal posteriorly as far as chiasm, involvement of periaqueductal grey, hypothalamus, dorsal pons, medulla,corpus callosum, High T2 signal spanning at least three vertebral segments, often many more (known as a longitudinally extensive spinal cord lesion), cord swelling usually present in acute phase
Myelin oligodendrocyte glycoprotein (MOG) associated demyelination	Bilateral or recurrent optic neuritis sparing the optic chiasm, longitudinally extensive transverse myelitis involving lumbar segment and conus medullaris

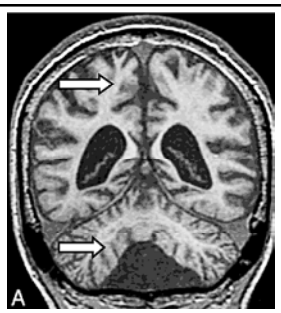

Fig.53. Neuronal ceroid lipofuscinosis Coronal T1 MR showing diffuse cerebral and cerebellar atrophy

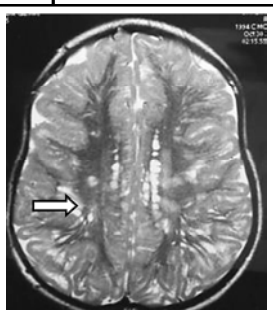

Fig.54.MPS Axial T2 MR showing prominent peri vascular spaces (Virchow Robin spaces)

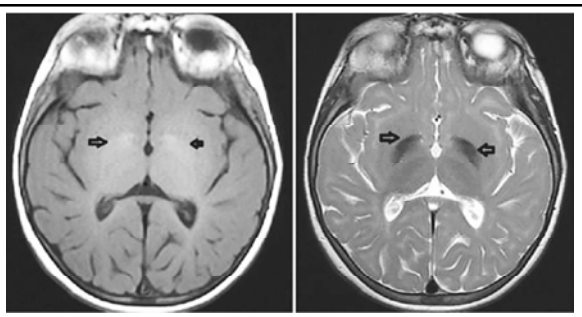

Fig.55a. Fucosidosis Axial T1 MR showing hyperintense globus pallidi

b. T2 MR showing hypointense globus pallidi characteristic

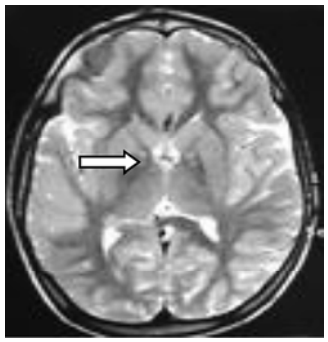

Fig.56. PKAN Axial T2 MR shows Eye of the tiger¨sign - central T2 hyperintense spot within the hypo intense globi pallidi

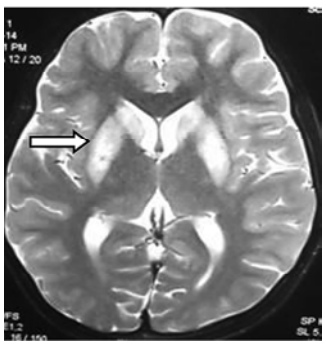

Fig.57. Wilson disease Axial T2 MR image shows hyperintense caudate and putamen

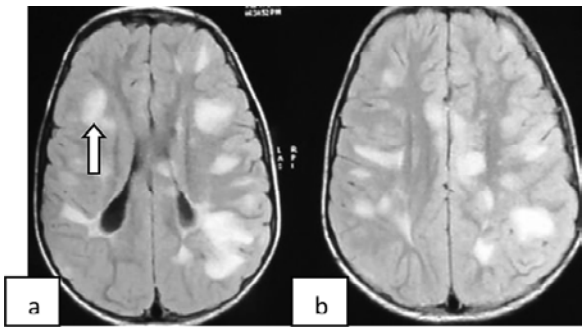

Fig.58 a & b. Acute disseminated encephalomyelitis Axial MR FLAIR sequences show bilateral, asymmetric, multiple hyperintense lesions in the deep and subcortical white matter

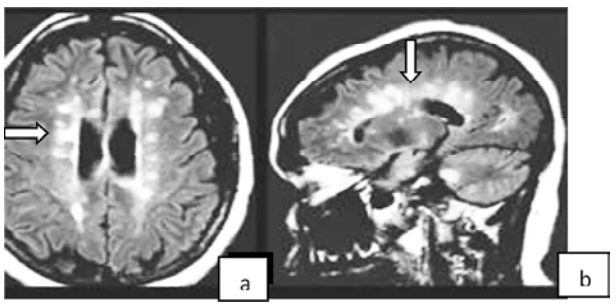

Fig.59 a & b. Axial and Coronal MR images showing multiple hyperintense lesions in periventricular white matter perpendicular to ventricular margins – Dawson's fingers. Involvement of corpus callosum seen in coronal picture

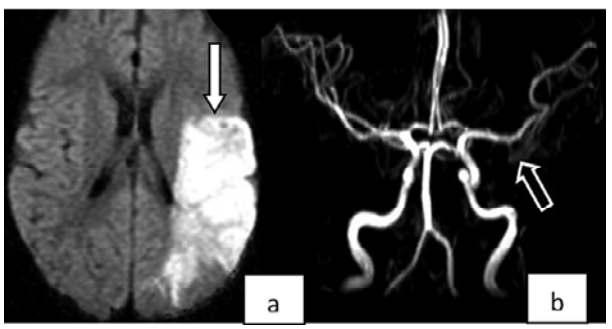

Fig.60 a & b. Axial DWI showing acute infarct in the left parietal region and MR Angiography showing focal irregularity of left middle cerebral artery

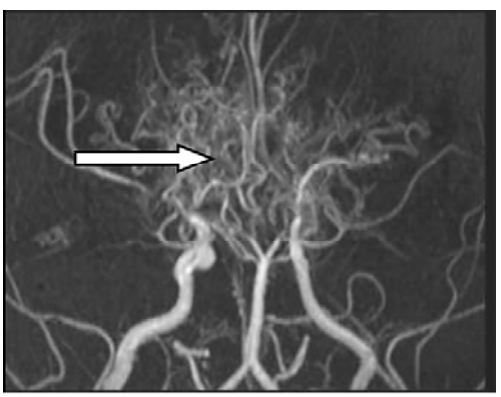

Fig.61.Moya Moya disease MR Angiography depicting occlusion of distal ICA and proximal MCAs bilaterally with collaterals with typical appearance of "puff of smoke"

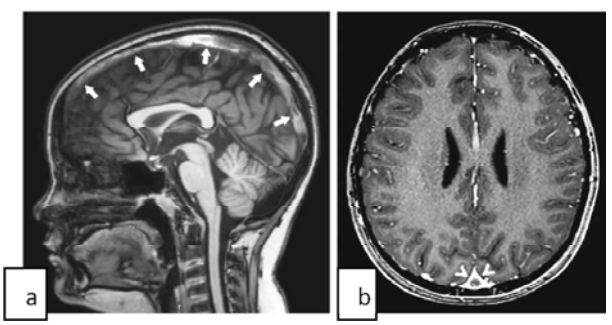

Fig 62.a.Superior sagittal sinus thrombosis. Sagittal T1 MR image showing abnormal hyperintense signal in the superior sagittal sinus

b. Axial contrast MR demonstrates the 'empty delta' sign of the superior sagittal sinus with contrast outlining a triangular thrombus

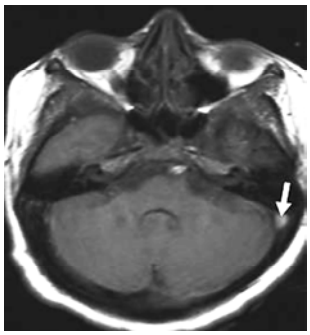

Fig.63. Sigmoid sinus thrombosis. Axial FLAIR MR image showing hyperintense signal at left sigmoid sinus

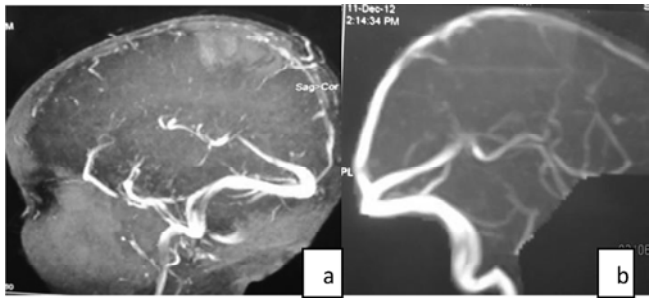

Fig.64a. MR Venogram shows thrombosis of superior sagittal sinus due to pyogenic meningitis. b. Partial recanalisation after three months

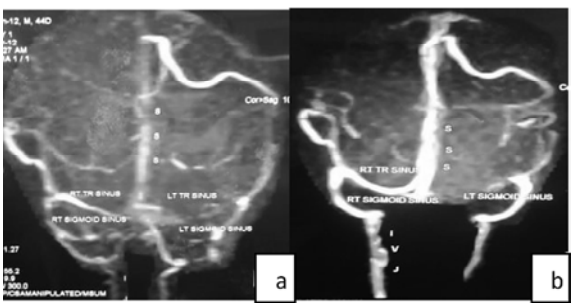

Fig.65a. MR Venogram shows thrombosis of superior sagittal, transverse, sigmoid sinuses in a child with protein C deficiency. b. Complete recanalisation of the sinuses after three months

Cerebral venous sinus thrombosis (CVST)

An ischemic infarction that crosses usual arterial boundaries particularly with a hemorrhagic component or in close proximity to a venous sinus is suggestive of CVST. Brain parenchymal changes in frontal, parietal and occipital lobes usually correspond to superior sagittal sinus thrombosis. Temporal lobe parenchymal changes correspond to transverse and sigmoid sinus thrombosis. Deep parenchymal abnormalities including thalamic hemorrhage, edema or intraventricular hemorrhage correspond to thrombosis of the vein of Galen or straight sinus.[26]

The primary sign of acute CVST on a noncontrast CT is hyperdensity of a cortical vein or dural sinus (Fig.62 a). Thrombosis of the posterior portion of the superior sagittal sinus may appear as a dense triangle, the dense or filled delta sign. Contrast-enhanced CT may show the classic "empty delta" sign, in which a central hypointensity due to very slow or absent flow within the sinus is surrounded by contrast enhancement in the surrounding triangular shape in the posterior aspect of the superior sagittal sinus (Fig.62 b). On CT, high hemoglobin concentration in the setting of dehydration or polycythemia can also be confused with clot, but in these individuals the entire vascular system is dense.[27]

MRI is more sensitive for the detection of CVST. MRI of the brain is suggestive of CVST by the absence of a fluid void signal in the sinus while T2 hypointensity is

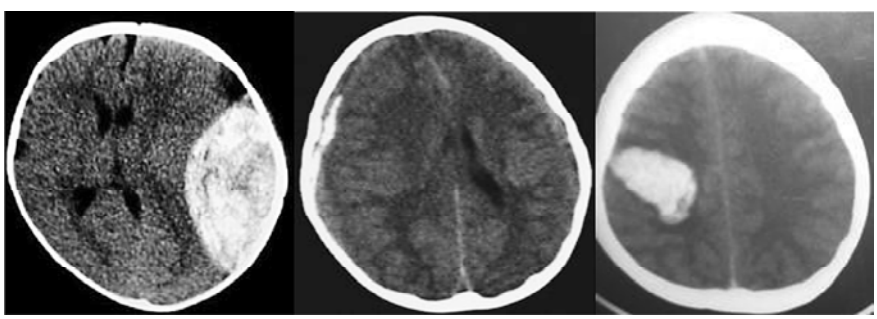

Fig.66a. Plain CT showing extradural hemorrhage (biconvex shaped) in the left parietal region with midline shift. b. Plain CT shows subdural hemorrhage (plano convex) in right parietal region with midline shift. c. Plain CT showing intra parenchymal bleed in the right parietal region

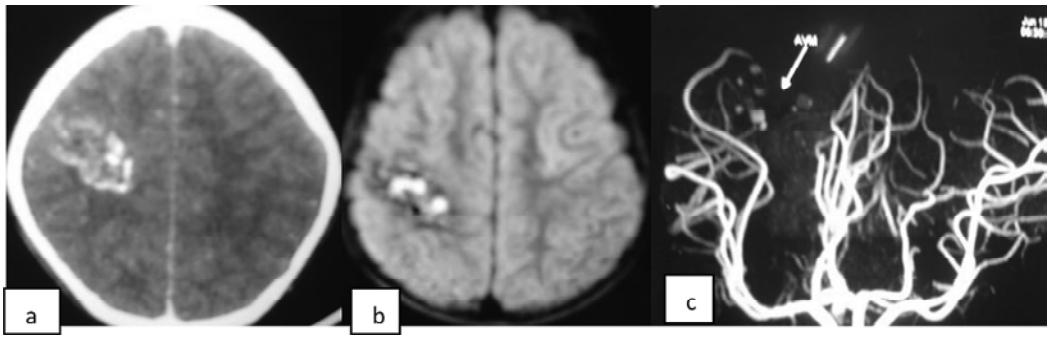

Fig.67a. Contrast CT showing enhancement of feeding vessels. b. Axial FLAIR image shows conglomeration of flow voids forming a "honeycomb" pattern. c. MR Angiography showing AVM

suggestive of a thrombus (Fig.63).[28] MRV shows non-visualization of occluded veins or sinuses due to absent signal and flow defect (Fig.64 a & b and 65a & b).[29]

A follow-up CT venography or MR venography at 3 to 6 months after diagnosis is reasonable to assess for recanalisation of the occluded cortical vein/sinuses.

CT is often the first neuroimaging modality performed because of its sensitivity for detecting hemorrhage, its short scan time and its availability in the emergency setting (Fig.66 a, b & c). If hematologic causes for hemorrhagic stroke are ruled out, CT angiography or MR angiography should be done to diagnose underlying arteriovenous malformations (AVM) or aneurysms. If no vascular malformation is noted during the acute period, repeat neuroimaging should be obtained after the hematoma has resolved because small vascular lesions can be compressed and concealed by the hematoma.

CT scan can demonstrate vascular calcifications associated with AVM. Contrast CT shows enhancement of the vascular channels (Fig.67a). MRI can demonstrate both dilated feeding arteries and enlarged draining veins. MRA can further delineate the anatomy and microarchitecture of an AVM (Fig.67 b, c).

Conclusion

Neuro imaging especially magnetic resonance imaging is a valuable tool in the hands of the clinician that helps in the diagnosis, management and follow up of neurological diseases in children.

Points to Remember

- ***Neuroimaging is an invaluable tool in the evaluation of neurological problems.***
- ***Cranial ultrasonography is the most frequently used neuroimaging modality in the perinatal period, particulary in the evaluation of hypoxic ischemic encephalopathy, subependymal- periventricular-intraventricular hemorrhage and hydrocephalus.***
- ***CT brain is the first imaging modality in unstable patients as it is widely available for emergencies, has shorter imaging time and lower cost. However, CT is generally suboptimal for imaging of structures in the posterior fossa and brain stem.***
- ***MRI is an indispensable tool in diverse CNS problems such as developmental anomalies, infections, neurocutaneous syndromes, demyelination and metabolic disorders.***
- ***Constraints with MRI are the need for sedation in young infants and its contraindication in the presence of metallic devices and implants.***
- ***MRA, MRV and MRS are additional facilities useful in identifying vascular and metabolic pathology.***

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CLIPPINGS

Corona viruses

Coronaviruses (CoVs) are a large family of enveloped, single stranded, zoonotic RNA viruses. They rapidly mutate and recombine leading to novel CoVs that can spread from animals to humans. The novel CoVs severe acute respiratory syndrome coronavirus (SARS-CoV) emerged in 2002 and Middle East respiratory syndrome coronavirus (MERS-CoV) in 2012. The 2019 novel coronavirus (SARS-CoV-2) is currently causing a severe outbreak of disease (termed COVID-19). This seem to less commonly affect children, cause fewer symptoms and less severe disease and are associated with much lower case-fatality rates in children.

Petra Zimmermann P., Nigel Curtis N. Coronavirus Infections in Children Including COVID-19 An Overview of the Epidemiology, Clinical Features, Diagnosis, Treatment and Prevention Options in Children.