# CONGENITAL ANOMALY OF HEAD AND NERVOUS SYSTEM COMMONLY FOUND IN THAILAND

**Vallop LOAPAIBUL[1] M.D.,**
**Kawee TUNGSUBUTRA[2] M.D., F.R.C.R. (ENGLAND)**

## ABSTRACT

Congenital ANORMALY of head and nervous system can be diagnosed accurately by CT and/or MRI. The early recognition of the deformed or the deprivation part or parts of the anatomical structures of the normal bones and/or soft tissue structures, early reconstruction of the IMPAIRMENT may help the innocent child to grow up and grow up to be a useful citizen and relief the burden of the family and the country. The conditions commonly found in Thailand and can be amendable are diagnosed by CT and/or MRI will be listed as followed.

1. **Fronto-Ethmoidal meningocoele. 2 cases.**
2. **Agenesis of Corpus Callosum**
3. **DANDY WALKER MALFORMATION (Cyst)**
4. **Schizencephaly: BRAIN CLEFT,OPEN LIP**
5. **Schizencephaly: BRAIN CLEFT, CLOSE LIP.**
6. **Lissencephaly. 6.1 Case 1**
   **6.2 Case 2**
7. **Holoprosencephaly**
8. **STURGE–WEBER Syndrome (S.W.S.), Neurocutaneous diseases.**

## INTRODUCTION

**CONGENITAL** anormally is a condition which is UNPREVENTABLE and UNPREDICTABLE. Early recognition and Accurate Diagnosis by CT and /or MRI, the impairment and absent parts can be repaired or reconstructed by modern surgery and artificial organ or the donated organ bank in future will be very beneficial.

## CONGENITAL ANOMALY;

### 1. FRONTO-ETHMOIDAL MENINGO -ENCEPHALOCOELE. Case I

**Clinical:** A boy, age 2 years, There is an abnormal mass between both eyes and between the middle part of nose bridge. The most prominent part is under the left eye obstructing the lower field of vision of medial part of both eyes. (black arrow) No nervous deficit.

Parents want their child to have plastic reconstruction. Fig. A. Non-contrast CT. showing bone defect at the white arrow.

[1] Department of Radiology, Faculty of Medicine, Khon Kaen University, Khon Kaen, THAILAND.

## FRONTO-ETHMOIDAL MENINGO-ENCEPHALOCOELE. Case I

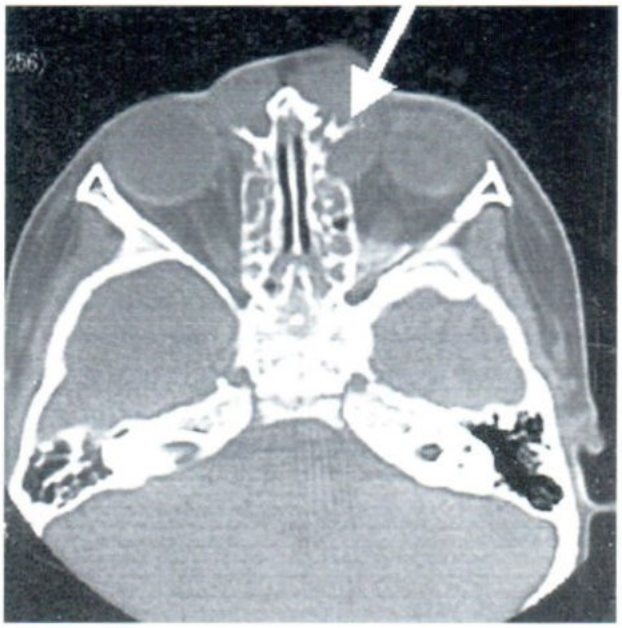


**Fig. A** NC.CT.

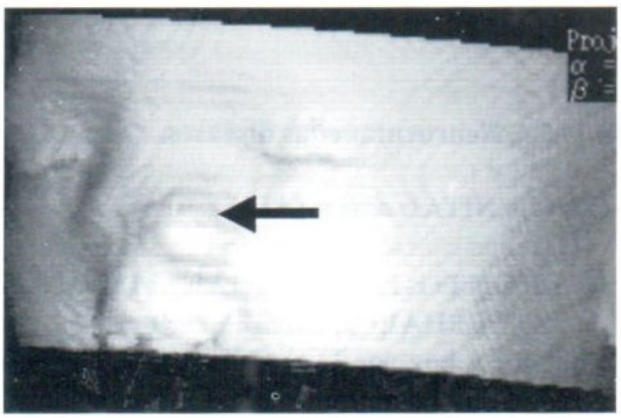


**Fig. B** 3D. Reconstruction of the CT., black arrow shows Fronto-Ethmoidal Meningocoele.

## 2. FRONTO-ETHMOIDAL MENINGOCOELE, Case II:

Clinical: A boy, age 8 months, having a bulging mass between the eyes and nose bridge. The bone defect showing as a bulging mass clinically known as "Meningocoele"

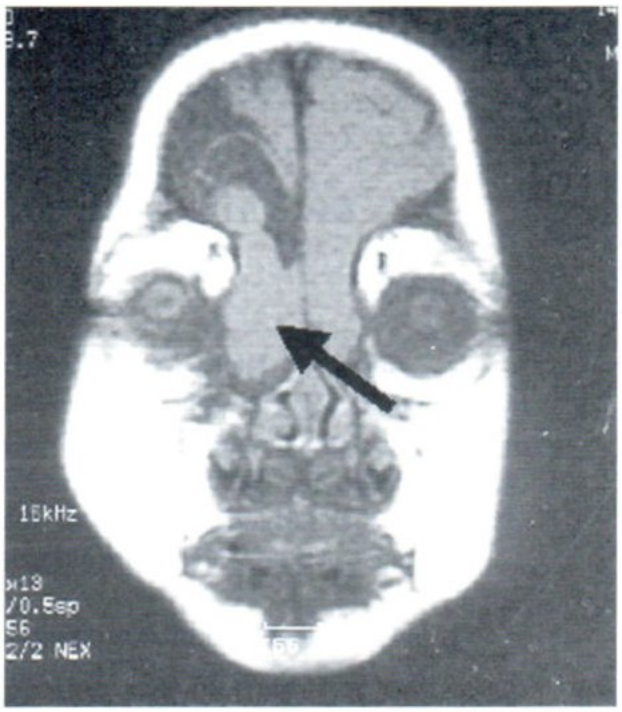



**Fig. A** MRI, CORONAL, $T_1W_1$

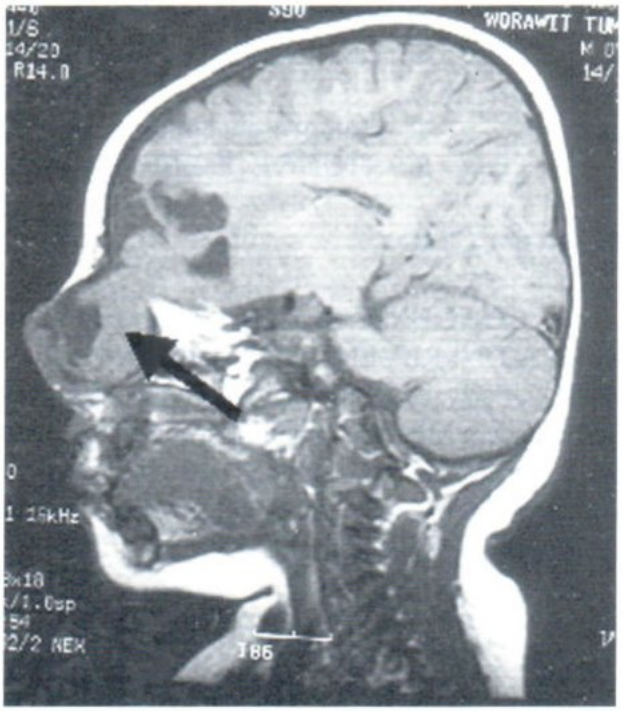



**Fig. B** MRI, SAGITTAL, $T_1W_1$

A and B, showing herniation of brain and meninges through the bone defect at the Fronto -Ethmoidal region. (black arrows)

## 3. AGENESIS OF CORPUS CALLOSAL.

**Clinical:** Male child, 1 year, head circumference 46 cm., retarded brain and body development. Out standing bigger head than normal child of the same age group.

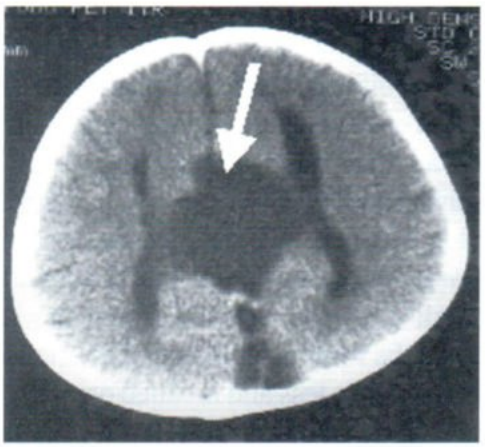


**Fig. A** NC.CT. shows absence of Corpus Callosum. Having central dorsal Interhemispheric cyst between lateral ventricles.

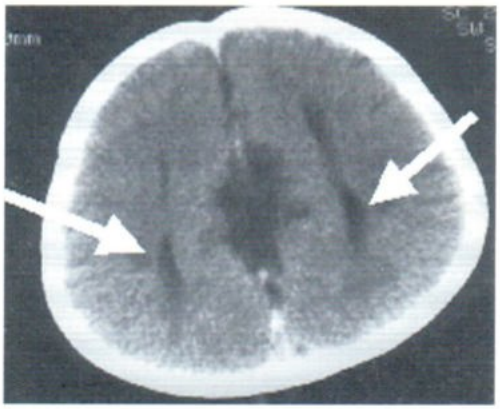


**Fig. B** NC.CT at different level of the head. CT. at upper level above the cyst, showing the lateral ventricles of both sides running parallelly between the left and right sides of the cyst seen in fig. A.

Corpus Callosum is the transverse nerve fibers which join the rt. and lt. hemispheres together. It is composed of mostly white matter situating at the deeper parts of longitudinal tissue consisted of 4 parts from anterior to posterior, Rostrum, Genu, Trunk and Splenium having a special name as Commissura Magna Cerebri.

## 4. DANDY WALKER MALFORMATION. (Cyst)

**Clinical:** Female, 1 month baby, born by Caesarian section: Having larger head than normal baby, vomiting occasionally, no convulsion, retarded development. CT. showed a large cyst at the posterior fossa with connection to the 4th ventricle. (black arrows).

Dandy Walker cyst is a congenital anomaly consisting of aplasia or hypoplasia of cerebellar vermis and cystic transformation of 4th ventricle. Congenital hydrocephalus, Dandy Walker cyst is caused by obstruction of foramen of Luschka and foramen of Magendie, which are the connections between the lateral ventricle and 4th ventricle (APERTURA MEDIANA VENTRICULI QUARTI.)

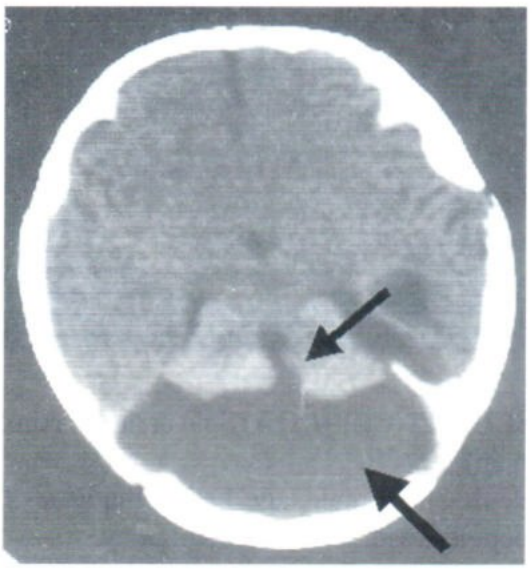
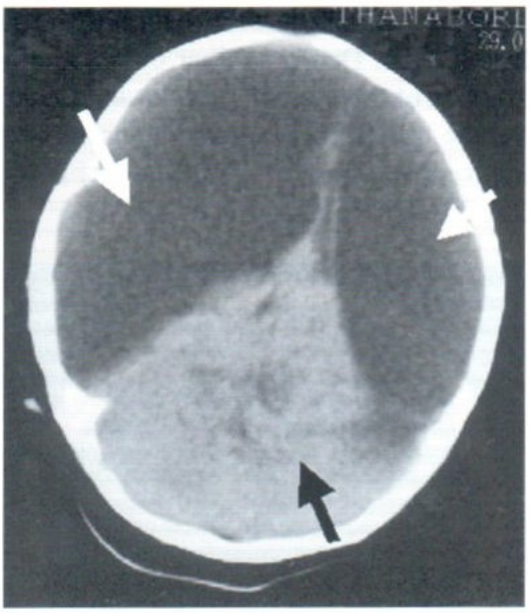


**Fig.4** Showed, "DANDY WALKER MALFORMATION", lower black arrow showed large cyst at the posterior fossa. Upper small black arrow showed connection with 4th ventricle.

**5. HYDRANENCEPHALY.**

**Clinical:** Child, male, age 3 months, no activities and bodily development. CT. showed cerebral hemispheres are replaced by water or CSF. The brain tissues seen in the picture are parts of cerebellum which is in the posterior fossa. Hydranencephaly is the combination of 2 words; HYDRO+ANENCEPHALY, i.e. Absence of cerebral hemispheres which are replaced by water. The baby is incompatible with life.



**6. SCHIZENCEPHALY;** Split brain or brain cleft (open lip)

**Clinical:** Child, male, age 1 year, big head, slow development with spastic cerebral palsy.

CT. showed empty spaces without brain tissues making the brain cortex, arachnoidal spaces and ventricular lumen have free connection. These findings are called open lips schizencephaly.

Schizencephaly is a condition which there is gray matter lined cleft, starting from ventricular ependyma to PIA mater which may consisted of;

1. There are cysts or cavities in the brain cortex connecting with arechnoidal spaces by pores or clefts.
2. There are cavities in the brain which may be found in fetus in utero or young infants and may or may not connecting with arachnoidal space. These cavities may occur from the result of destructive lesions or may occur congenitally. Schizencephaly may be found in 2 different types. The open lip is compatible with when one open his lips, looking into his mouth through a mirror, one may see a large cavity with a Uvula in the middle. The close lip is looking like one close his lips and only he can see a small cleft between the upper and lower lips. In this case, there is a small cleft connected between the cerebral cortex and the ventricular lumen. If the cleft is wide, the brain tissue is less and is called open lip. On the contrary, if the cleft is narrow, it is a close lip type. Close lip, schizencephaly, there is a small cleft connecting between brain cortex, arachnoidal space and ventricular lumen.

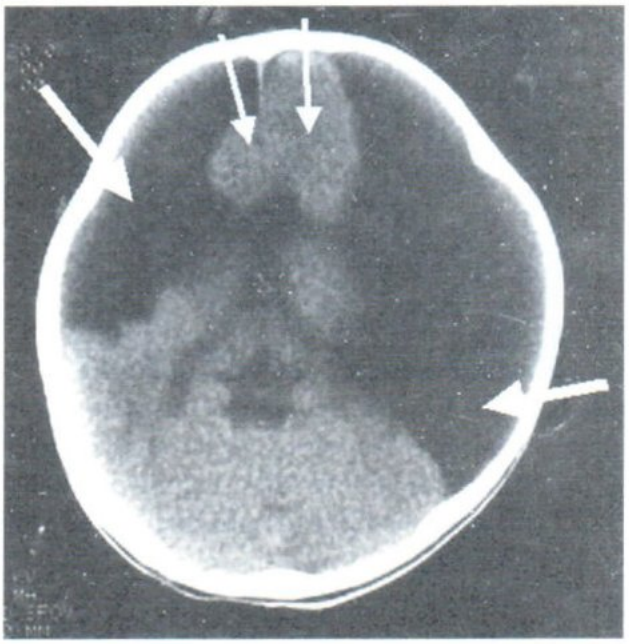
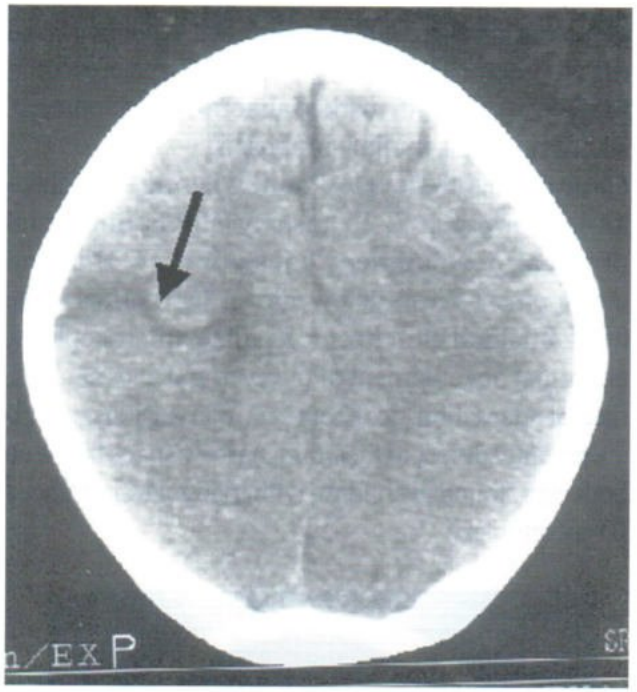


**Fig. 6** Open lip Schizencephaly: split brain or brain cleft (open lip)
[ A Uvula in the oral cavity]

**7. SCHIZENCEPHALY;** Brain cleft (close lip)
**Clinical:** Infant, male, age I year and 7 months. Lt. arm and leg weakness for 1 year. CT. show narrow brain cleft connecting from cerebral cortex to ventricles.

Black arrow showing small cleft connecting between cerebral cortex and ventricle. Schizencephaly: Brain cleft (Close lip)



**8. LISSENCEPHALY** (Smooth brain)
**Case I:**
**Clinical:** Female, age 16 years, delay development, both physically and mentally. CT, showed smooth brain, having small number of sulci and gyri. The sulci and gyri are broader than normal brain, looking smooth in outline. The cerebral cortex is thick, cerebral sulci is small in number, each one is broad. Gyri are flat. Lissencephaly having smooth brain, having small number of sulci and gyri. The sulci and gyri are broader than normal brain, looking smooth in outline. The cerebral cortex is thick, cerebral sulci are small in number, each one is broad. Gyri are flat. Lissencephaly having smooth brain, gyrus is shallow or no gyrus, in the worse cases, they are agyria (completely smooth) or pachygyria (incomplete). The convolution number are few. Smooth cerebral hemisphere or small numbers of convolution can be found in some species of mammals such as "bats" or in some rodents.

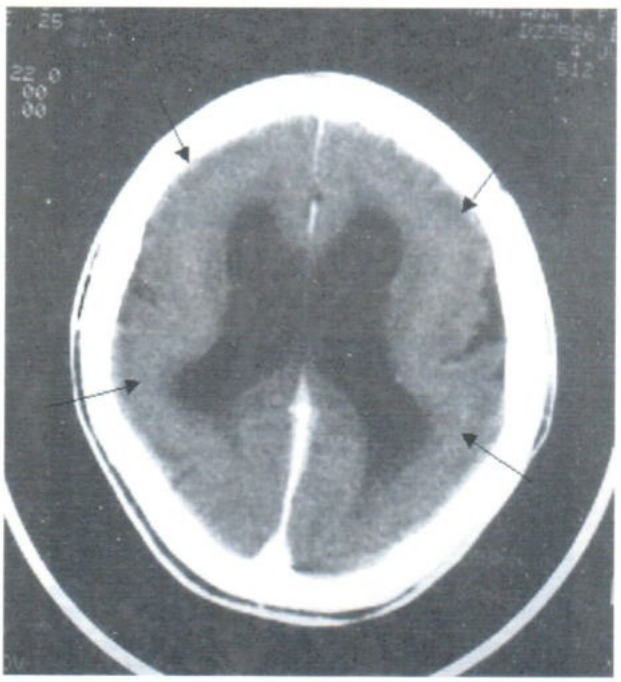


**Fig. 8** CT, of lissencephaly showing smooth brain. few numbers of sulci and gyri. The complete absence of gyrus is called "Agyria." The incomplete absence with shallow gyri are called "Pachygyria"

**9. LISSENCEPHALY** (smooth brain)

**Case II:**

**Clinical:** Male child age 5 months, slow development since birth.

**Fig. A.** is $T_1W_1$, MRI., **Fig. B.** is $T_2W_1$, MRI.

Cerebral cortex thick, syvian fissures shallow, the furrows are natural development to increase the area of brain tissues to impove the intelligence of human being. Gyrus is the tortuous elevations or convolution of brain surface.

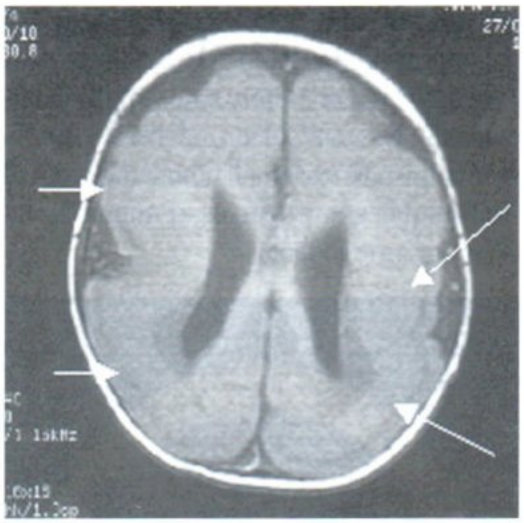


**Fig. A** $T_1W_1$, MRI.

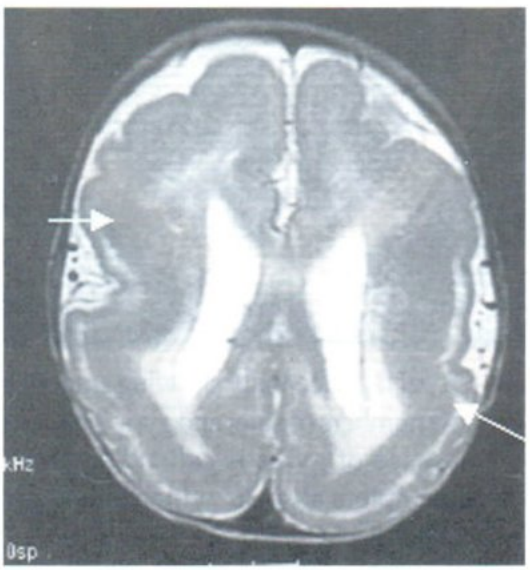


**Fig. B** $T_2W_1$, MRI.

**10. HOLOPROSENCEPHALY**

**Clinical:** Female, age 6 months retarded development since birth.

CT. shows semilobar holoprosencephaly:

**Absence** of **septum pellucidum.**

**Septum pellucidum** is double layers of thin membrane, triangular shape separated the anterior horn of lateral ventricles into left and right sides at the median plane.

**Holoprosencephaly:** Failure of normal development of prosencephalus together with deficit in midline facial development, There may be only one eye present in the face in case with marked abnormal development. such as cyclops, midline proboscis.

**Causes of Mal-Development:** Having extrachromosome in group 13-15, trisomy DI. **The abnormal physical development consisting of:** Bilateral development of ears at lower level than normal in the head, bilateral cleft lip, cleft palate, microcephaly, abnormal development of the eyes and eyes sockets, mental retardation, deaf both ears, convulsions, ventricular septal defect in the brain, narrow space between both eyes, found in children with triangular shape head (Hypotelorism)

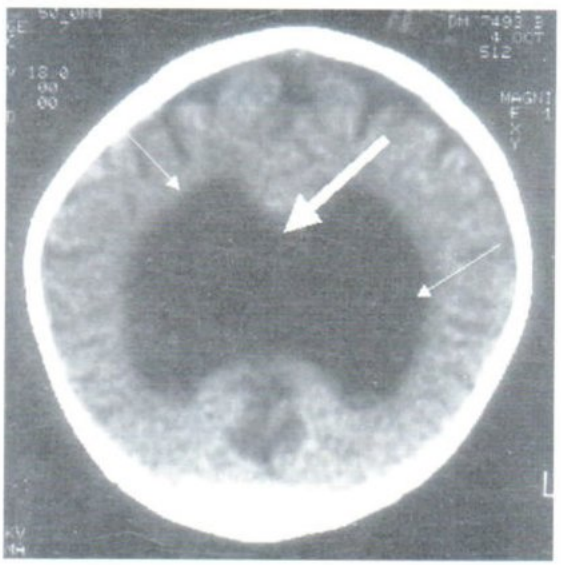


**Fig. 10** CT. shows abscence of septum pellucidum.

## 11. STURGE–WEBER SYNDROME (SWS); CONGENITAL NEUROCUTANEOUS DISEASE.

**Clinical:** Male, age 14 years.: Convulsion,on and off, controlled by anti-convulsive drugs. The convulsive attacks increase in frequency as the boy grow older.

**Fig.A.** NC.CT., **Fig.B.** CE.CT. Both figures show gyral enhancement at right cerebral cortex indicating atrophy of right cerebral hemisphere and thickening of calvarium right side.

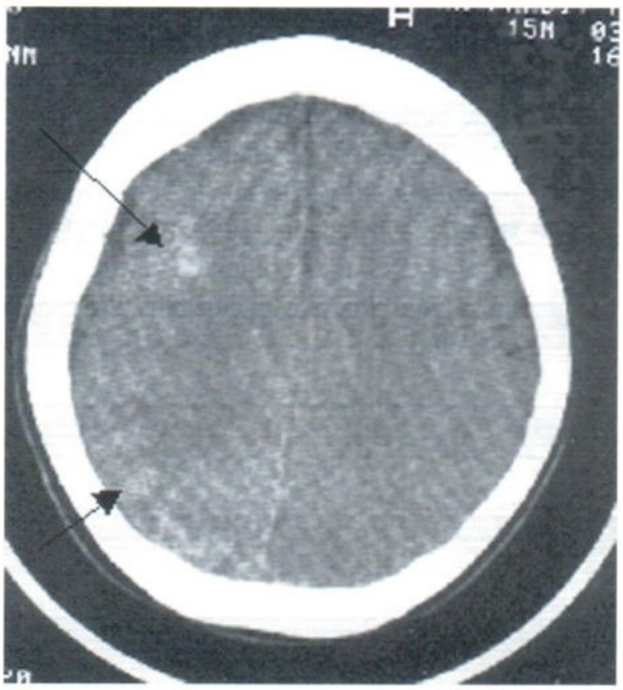


**Fig. A** NC.CT. shows cortical calcifications.

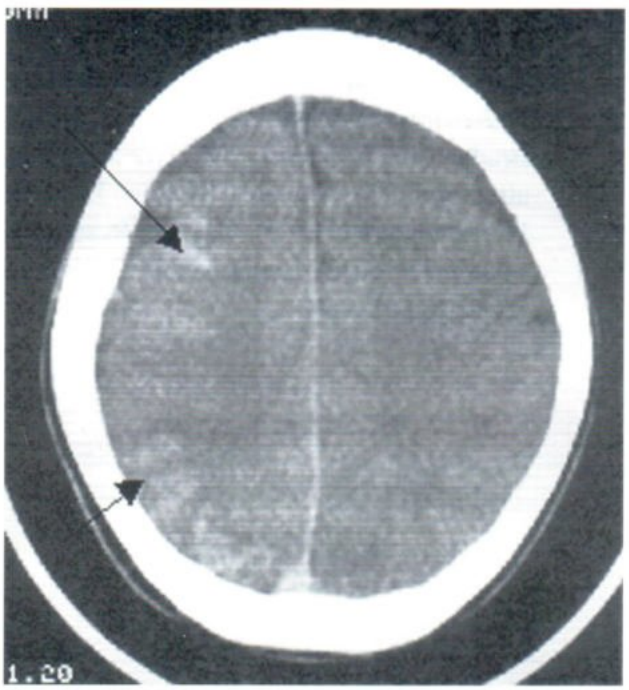


**Fig. B** CE.CT. shows enhancement of cortical calcifications.

***Sturge-Weber syndrome is a congenital abnormality characterized by:***

1. Capillaries haemangioma at the face presented at birth with varying areas. It present as a red skin on the face at one or both sides of the face.
2. Presence of angioma at the leptomeninges and choroid, late glaucoma, intra-cranial calcifications,
3. Mental retardation
4. Contralateral hemiplegia and epilepsy due to cerebral hemorrhages from intra-cranial angioma.
5. A port-wine stain half of the face in the area supplies by the first branch of trigeminal nerve due to the abnormalities of cerebral vessels, resulting in contra lateral hemiplegia.

## DISCUSSION

The development of CT. and MRI., parallel with the improvement of opaque media using with CT. and MRI making the diagnosis of the lesions in the brain more convenience , accurate and quick. The most important diagnosis are the number of the lesions, the position of the lesion or lesions and the nature of the lesion. The treatment can be pre-planned and choose the right and best method of treatment available.

## CONCLUSION

CT. and MRI. with or without contrast media

should be used for prompt and accurate diagnosis of the diseases of central nervous system. The treatment of the lesions in the CNS. should be a team approach. The team should consist of, radiodiagnostician, radiotherapist, surgeon, physician and pathologist.

## REFERENCES


1. Zimmerman RA, Bilaniuk LT, Gusuard DA. Pediatric cerebral anomalies. In: Stark DD,
2. Bradley WG Jr, Quencu RM, Hydrocephalus, atrophy, and intracranial CSF flow. In: Stark DD, Bradley WG Jr. eds. Magnetic Resonance Imaging 2nd ed. St. Louis: Mosby-Yearbook 1992; 28:913.967.
3. Gomori JM, Grossman RI. Goldberg HI. Occult cerebral vascular malformations: high field MR imaging. Radiology 1986; 158: 707-713.
4. Teresi LM, Davis SJ. Cerebrovascular maformations. In: Stark DD, Bradley WG Jr eds. Magnetic resonance imaging 2nd ed. St. Louis: Mosby Yearbook; 1991: Chpt 29.
5. Barkovich AJ. Pediatric neuroimmaging. New York: Raven press, 1995.
6. Chamberlain MC. Pediatric aids: a longitudinal comparative MRI and CT brain imaging study. J. Child Neul. 1993; 8: 175-181.
7. Faerber E. Cranial computed tomography in infants and children. Philadelphia, PA: Lippincot 1986.