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Outcome of 14 Patients with Dandy Walker Malformation in Al Nasiriyah Teaching Hospital (2014 – 2020)

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Abstract:

early workers. Although the Taggart and Walker theory has probably been the one most CSF path ways dilatation accompanied by a posterior compartment cavity& both cerebellar atrophy is known as Dandy Walker malformation, is a rare syndrome with facing a challenge in its presentation & management. The sign & symptom rely on congenital malformation in toddler. If the problem due to raised ICP the infant is presented with large size head& sign & symptom of CSF pathways dilatation. In older children, cerebellar atrophy principally imitate axial trunk instead of peripheral limb ,intellectual retardation , papilledema & extra ocular muscle extirpation are apparent .

Materials:

By this retrospective study 14 cases were collected , diagnosed and treated throw out our neurosurgical field in Nasiriyah teaching hospital from 2014 to 2020. The diagnosis depends on radiologic investigation. Presently, the most definitive diagnostic imaging method is magnetic resonance imaging.

Outcome:

Surgical treatment of 10 cases with douple catheter of ventricular system & posterior cranial fossa sac.Four cases treated with standard shunt system originating in the lateral ventricle with aid of endoscope for fenestration of membrane responsible for non communication to facilitate creation of one compartment. Patient outcome good and discharged well.

Conclusion:

DWS is a rare congenital brain abnormality, presenting usually in first year of life with enlargement of the head. In older children , it can be present with varied neurological symptom. It is diagnosed with advanced imaging & treated primarily with CP or VP shunting or both. Prognosis varies due to several factor , but an early diagnosis & appropriate treatment have a good prognostic value .

Key Words:
Dandy-walker malformation, dysgenesis, hydrocephalus, surgical treatment endoscope.

Definition and Pathogenesis:

The dandy walker syndrome although described initially at the end of 19th century, It doesn't until 1914 that the first detailabout disease approved by ((Dandy and Blackfan)).[1] The cause of the disease of darn in the foramina of (luschka & magendie) was apparent in this period & subsequently elaborated via((Taggart and Walker)),[2] who attributed the accompanied CSF pathways dilatation & bulging of the 4th ventricle to foraminal atresia so as to cause mal development of the cerebellar vermis& arrest of rear migration of swarming the cavity & awning. Subsequently, the term Dandy-Walker syndrome has become the accepted terminology, even though it does not acknowledge the roles of other commonly accepted, it must be recognized that there is evidence to indicate that it may be in correct. A second opinion was suggested through((Benda)), that suggested the disease clear up mal development in site of the 4th ventriclebut don't restricted to the aperture.[3] He guess that defeat of retroactive change in rear medullary velum associated with a hereditary loss of cerebellar vermis result in cavity apparent at tailpiece of 4th ventricle & isolation of cerebellar lobe. Advance research lend reinforce this opinion. It appear in research study in human being,[4] some problem are specialized for Dandy Walker Malformation happened prior to aperature would normally sincere & so spit suspicious function of foraminal darn. More ever, the appearance of numerous other anomalies accompanied with Dandy-Walker Malformation pointed that the way the disease develops is something primary & further sophisticated malprogress.

Data Collection in Al - Nasiriyah Neurosurgical field

All patients are collected in Al-Nasiriyah teaching hospital in department of Neurosurgery , ten cases was brought to the neurosurgery consultant out-patient directly & four cases refer from pediatric hospital. Age of ten patient during infancy & other was older children. Almost of the patient have bulging of all fontanels & sutures diastasis. Lambdoid suture is broad, elevation of the occipital bone in comparison of remaining of skull bone. Transillumination of skull , (EOP) unnaturally elevated in some cases. Four cases with cerebellar dysfunction , papilledema , nystagmus. Prosopopoeia depend on radiological Ix. X-rays of the skull is benefit but have limited importance. There is prove of macrocranium and suture diastasis associated with expansion of posterior cranial fossa. Old age child x-ray film appear elevated piece of torcular & both transverse sinuses. Ultrasonography has become a widely accepted and useful method to determine the gross intracranial anatomy of infants, particularly the size & ranking of CSF pathways . Scan made of an infant with a Dandy – Walker Deformity appears a huge cavity in posterior cranial fossa which communicate with enlargement 4th ventricle. The cerebellum is tinny and the diminutive cerebellar lobe are deviated antero-laterally. The lateral & third ventricle dilated and the occipital horn of lateral ventricle are characteristically divergent as a consumption of raised part of the tentorium & displacement by cyst as in the figure (1,2,3). Presently, the most definitive diagnostic imaging method is (MRI).[5,6] Multiplanar MRI is used to assess the cerebellar vermis and hemisphere for hypoplasia or atrophy. Mid sagittal MR images will demonstrate a rotational axis & pressure at the vermis & better accurate any caudal expansion of cavity to up cervical

vertebral column or rostral expansion across tentorial incisura. Through axial succession both cerebellar lobe is usually shrinkage for 2 tinny small knot of neural parenchyma deviated antero-laterally & did not attached via midline parenchyma. Both lateral & third ventricular system mild - moderate expansion. Dynamic MRI pulse sequences visualize flow through the aqueduct to help determine the presence or absence of stenosis, see figure (4,5).

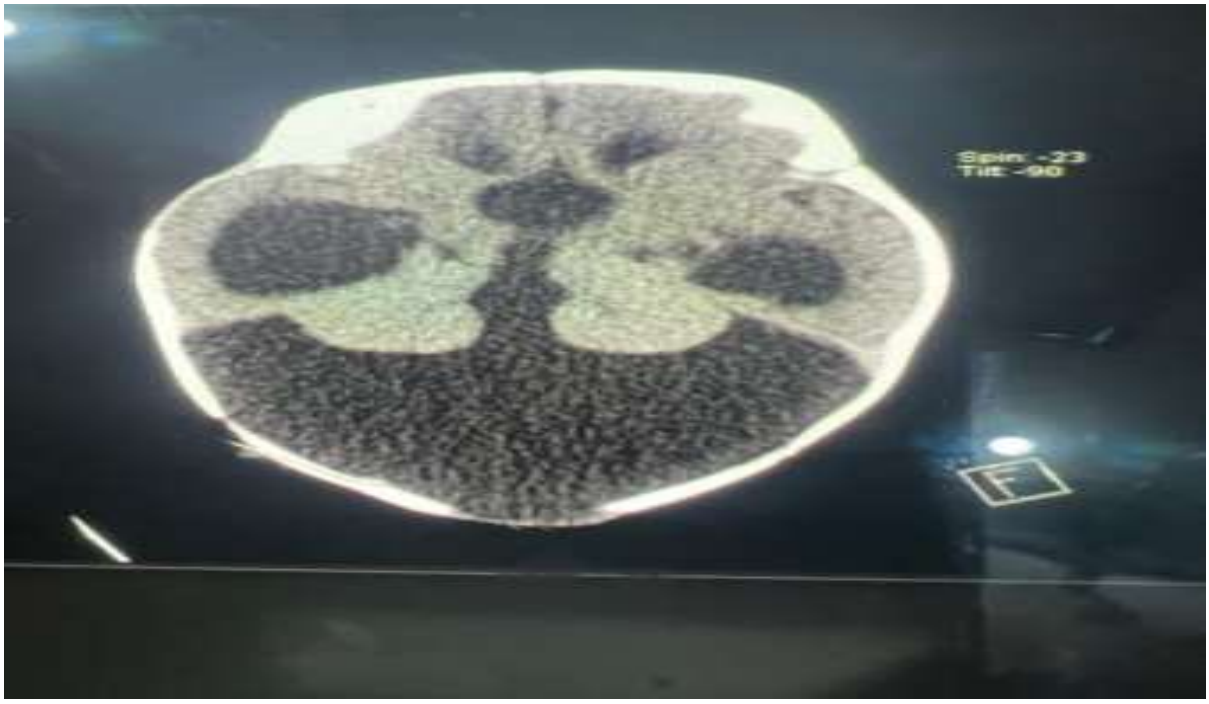

Figure (1):Axial Non Contrast C-T

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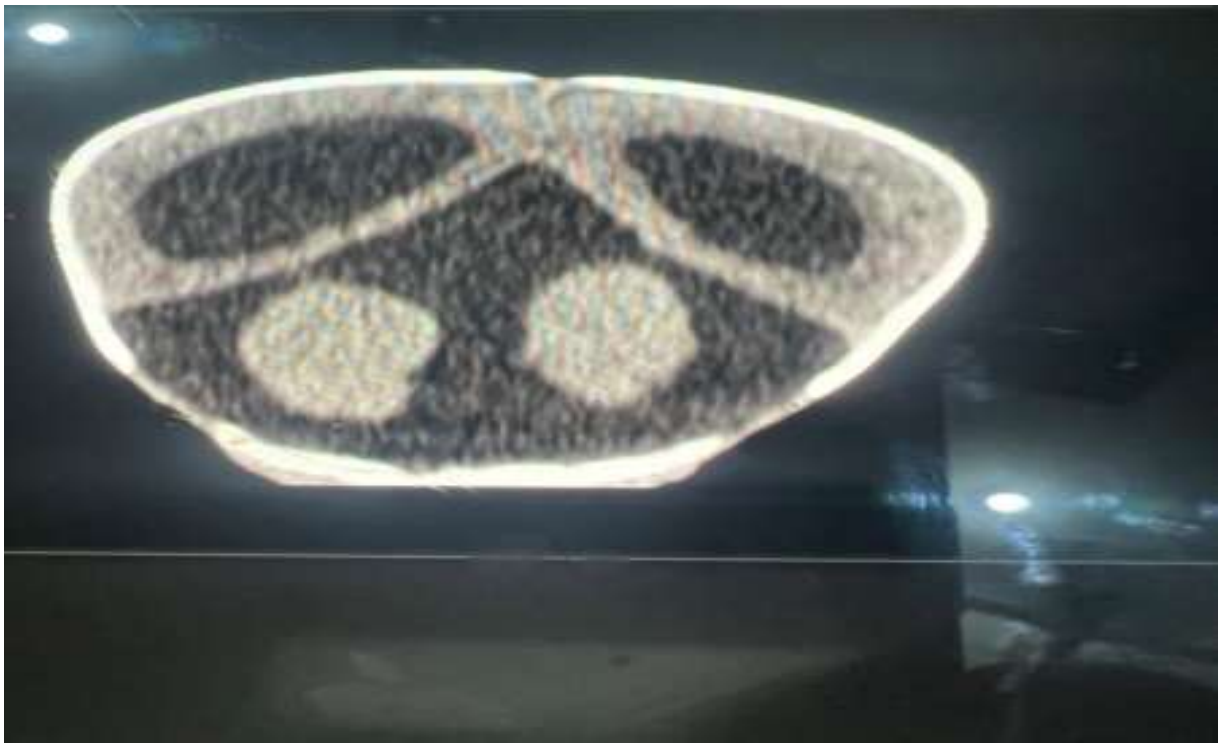


Figure (2): Coronal Non Contrast C-T Scan

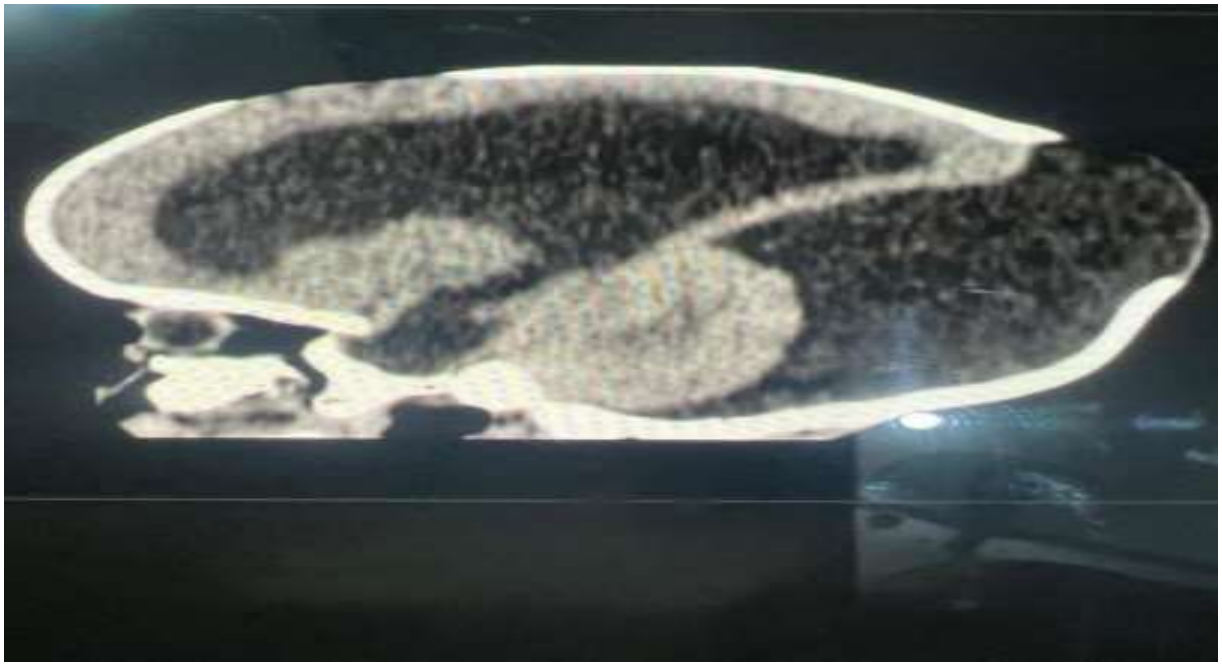


Figure (3): Sagital Non Contrast C-T Scan

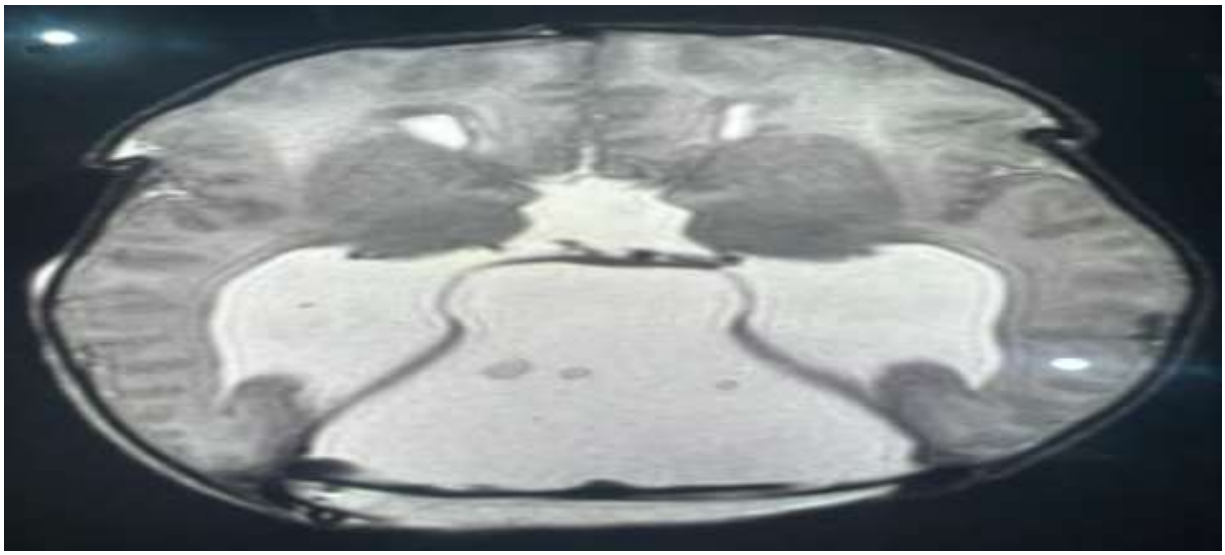


Figure (4): Axial MRI T2

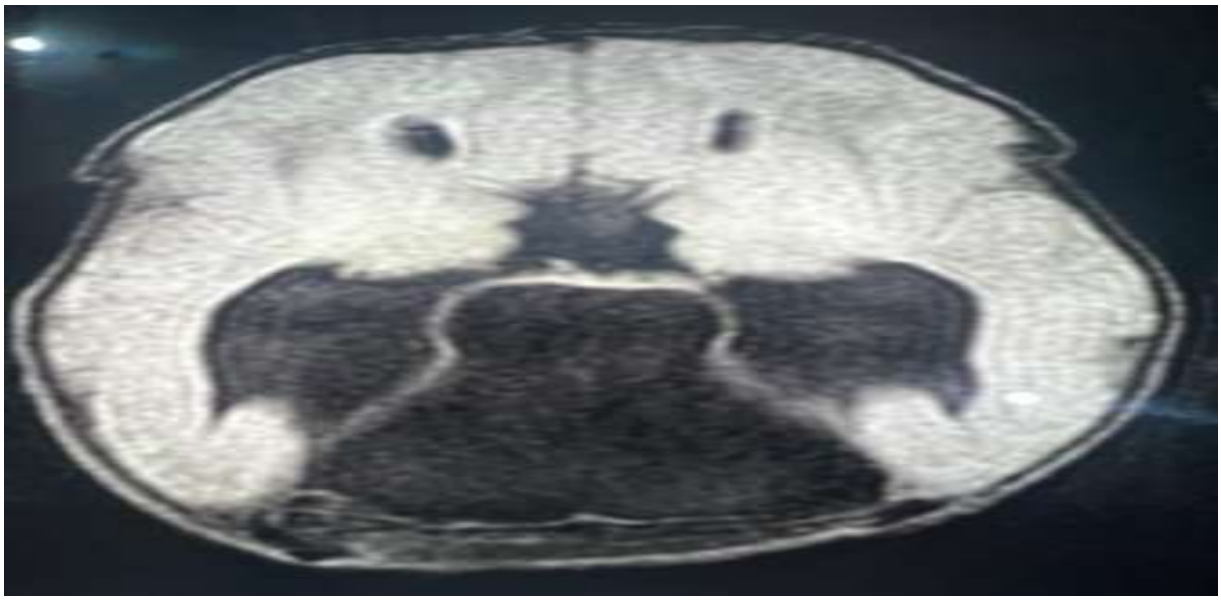
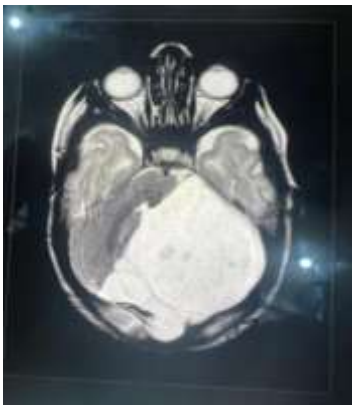


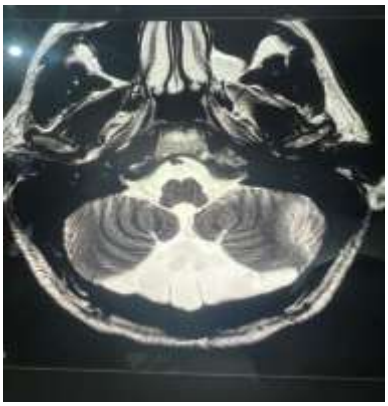
Figure (5): Axial Flair MRI

Differential Diagnosis:

Some cases are in x-ray, brain C-T scan &MRI similar to the Dandy –Walker syndrome like expansion of Cisterna Magna as in picture (A). & the appearance of arachnoid cyst picture (B){{axial T2 MRI}}, picture (C) {{ sagital T1 MRI}}, picture (D) {{coronal T2 MRI}}. Because the giant Cisterna Magna does not full almost the whole posterior fossa, isolated 4th ventricle is distinguished from it & cerebellar vermis is very clear.[5]Extra –axial arachnoid cyst is another challenge but it is not in center & deviate the normal 4th ventricle . If doubt is present , a positive contrast agent may be used to check out aqueduct of Sylvius , 4th ventricle availability & link with abnormal cavity.



Picture (A)



Picture (B)

Picture (C)

Picture (D)

Figure (6):

ASSOCIATED ABNORMALITY:

1.Dandy –Walker syndrome is associated with 65% of other CNS malformation & responsible for outcome and prognosis.

2.CNS malformation like neuronal migration disorder of cerebellum & cerebrum , brainstem malformation , hamartomas, abnormal development of brain before birth known as polymicrogyria, stenosis of aqueduct of Sylvius , myelomeningocele & meningocele.

3.Systemic anomaly include split lip & palate , supernumerary fingers and or toes, webbed or conjoined fingers or toes.

Treatment:

Dandy-Walker syndrome submitted to several changes of management from which it firstly apparent & later up on & till this time no worldwide acceptance of its management. The first one to discuss eradication of cavity membrane & posterior Dandy and Blackfan..[1] This procedure is a unique solution profitable for many years till roses the CSF shunting tools. CSF shunt tools apply if membrane cavity can't be eradicated completely with surgery& hold ventricular dilatation. Advance observation reveal the choice of prosperity of cavity membrane eradication surgery was a little that the basic method of management should be shunting. This the only solution agreement through last years, but controversy remain abundant regarding exact site of caudal tip of shunt tool. Several scriber supported benefit from basic shunt tools inserting in the lateral ventricle. Coworkers& Raimondi have supported use double CSF shunt of lateral ventricles & posterior fossa cavity in order to prevent obstruction of aqueduct of sylvius & shunting of only the lateral ventricles result in rising up transtentorial herniation due to loss of shrinkage of cavity.[7] Another seniors, however , including ourselves, suggested that stricture of aqueduct of sylvius is rare & simply discovered through introducing a contrast material to lateral ventricle at same onset of C-T scanning .If attachment between the lateral ventricle & cavity is found, it is better to put the caudal tip of shunt in the cavity . This barrage of CSF , not similar to lateral ventricle , does not shrinkage on pressure relieve , slightly due to huge posterior compartment & accompanied cerebellar atrophy. So shunt task if don’t work because caudal tip obstruction is seldom happened. In case there is no attachment among the lateral ventricle & cavity , Endoscope step inside through a lateral ventricle & the layer accountable for nonattachment perforated for easy acting single independent part .Catheter is guided via ventricle to the cavity of posterior compartment in order to prevent intricate shunt device.

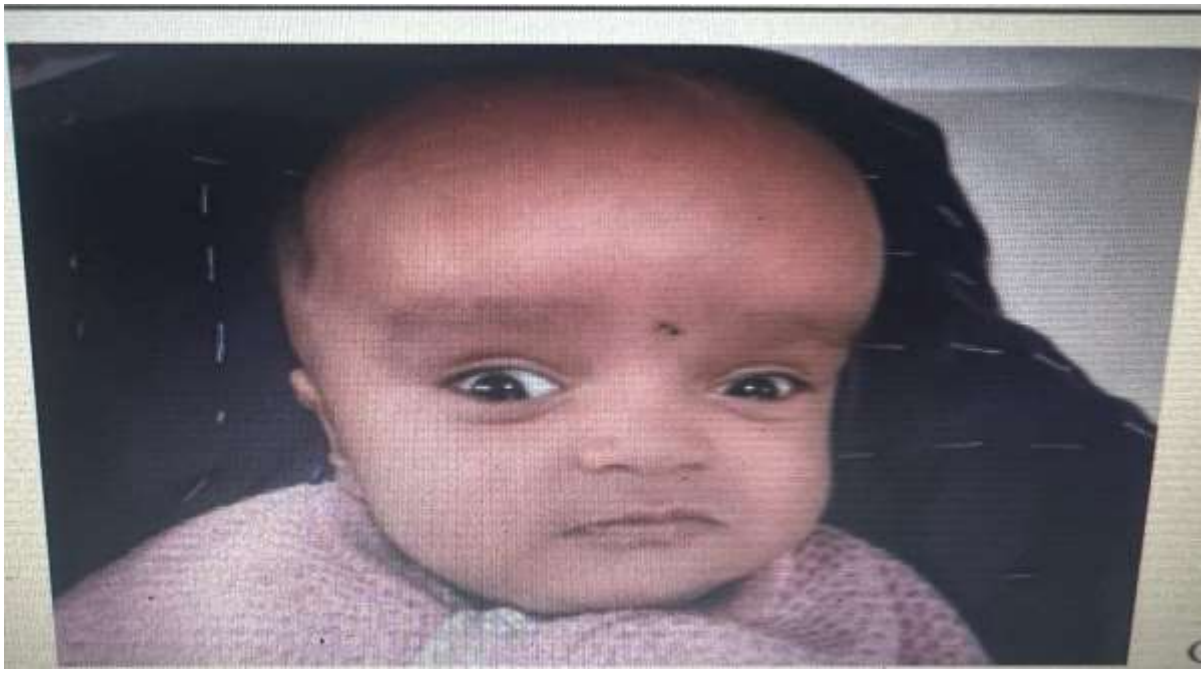


Figure (7):Clinical Picture Macrocephaly

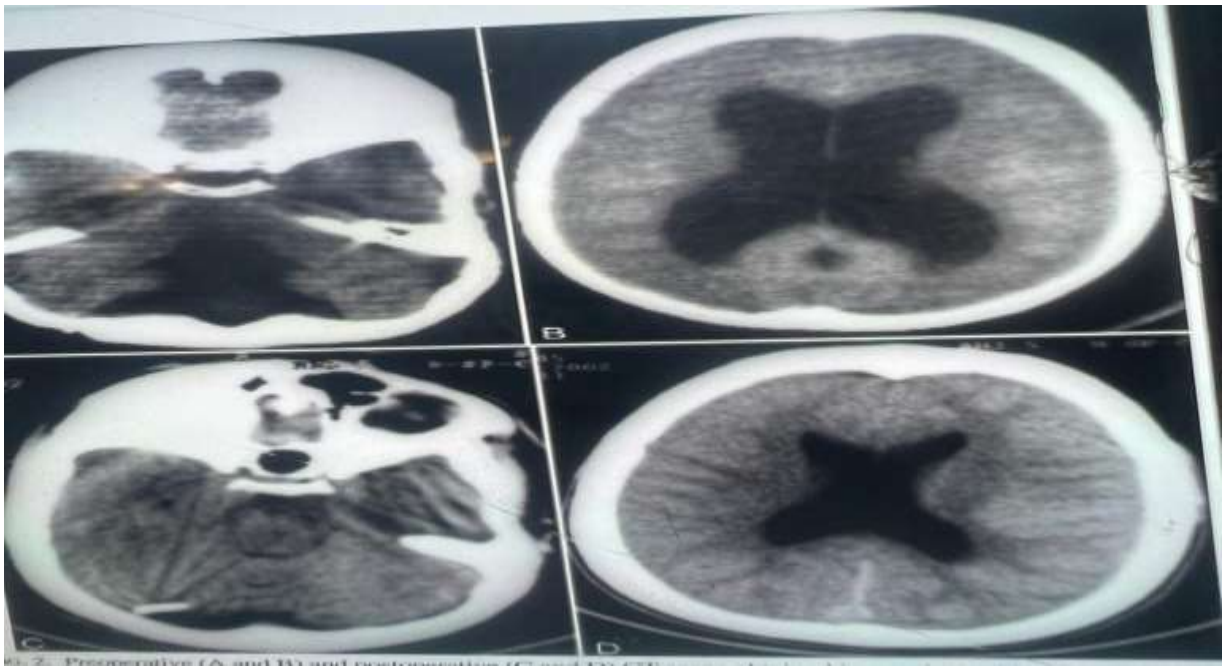


Figure (8): Pre operative (A,B) & post operative (C,D) C-T scan done in a child suffering from a Dandy-Walker varient made for him shunt device. observe marked decrease of cavity volume& average decrease in ventricle quantity.

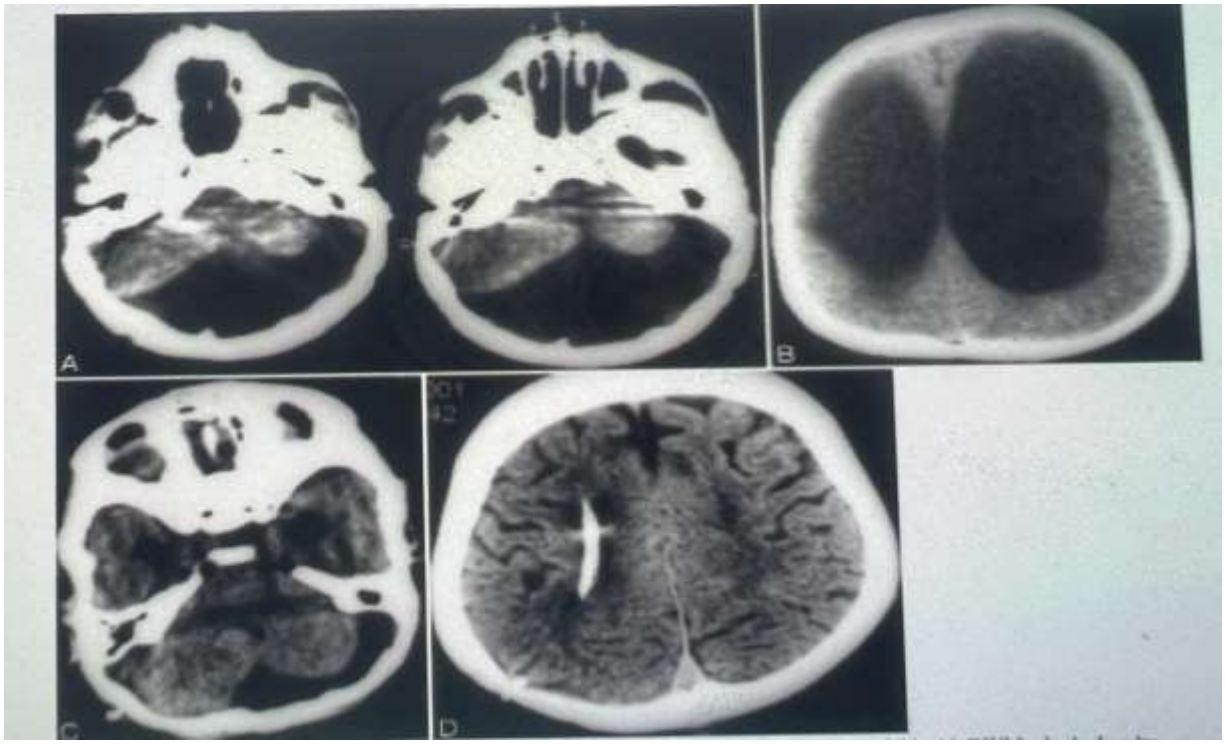


Figure (9): Pre operative(A,B) & post opetrative(C,D) scan done in infant suffer from DWM for whom a shunt device was done. observe marked decrease of ventricle volume with a leftovers posterior compartment cavity.

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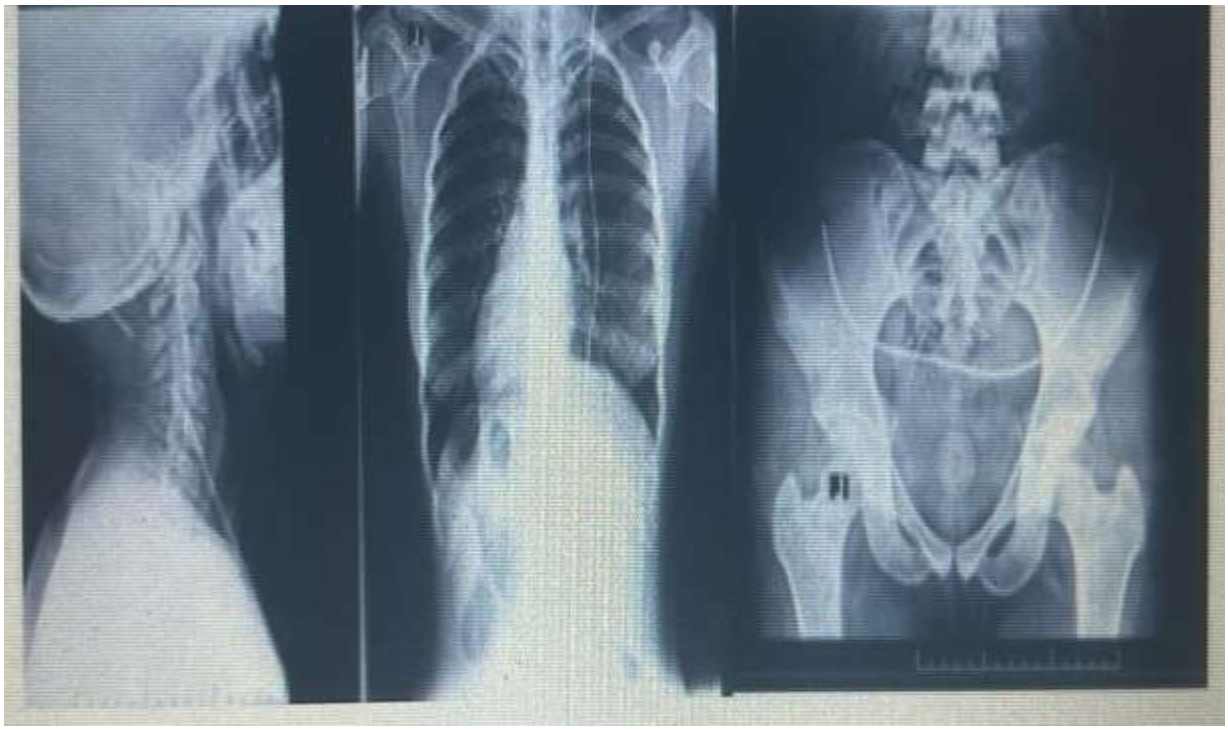

Figure (10): X-ray obtained after surgery show course of shunt device through the body.

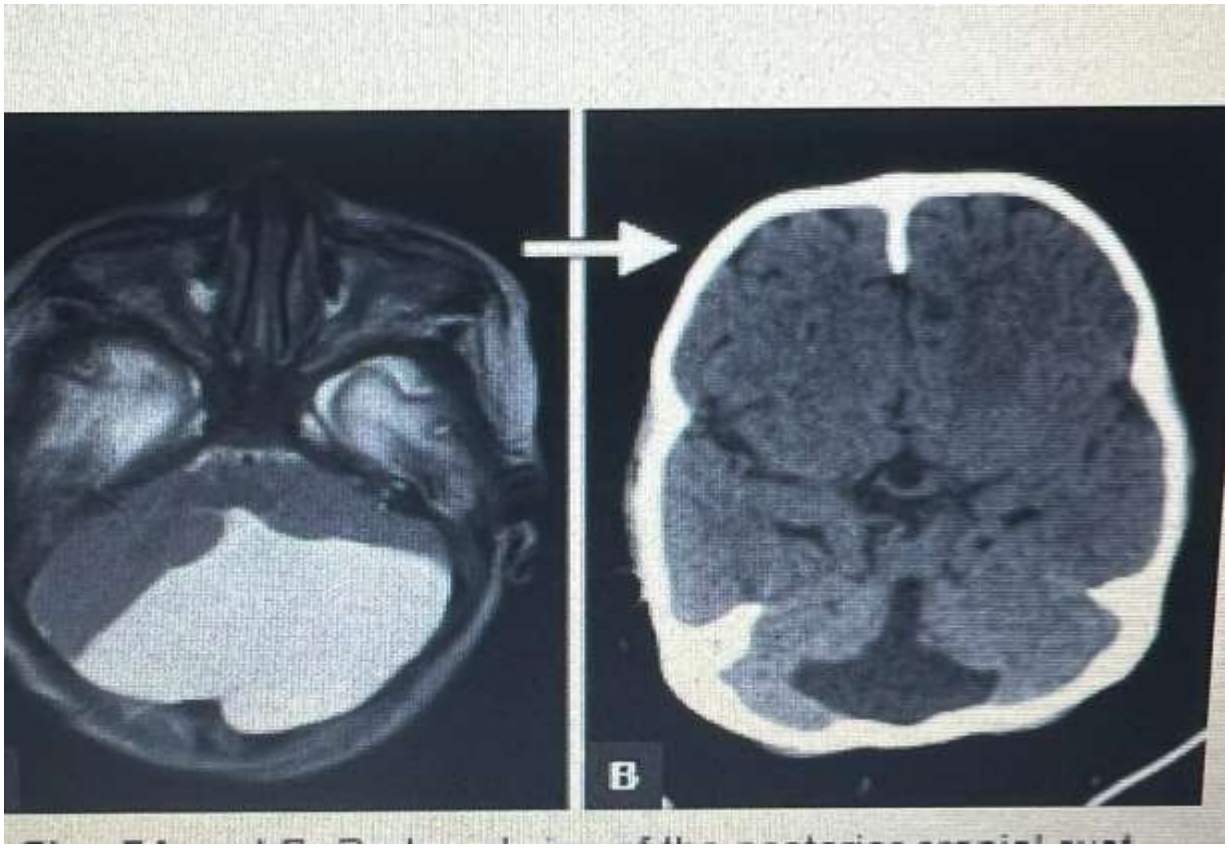

Figure (11): Reduced size of posterior cranial cyst post operatively

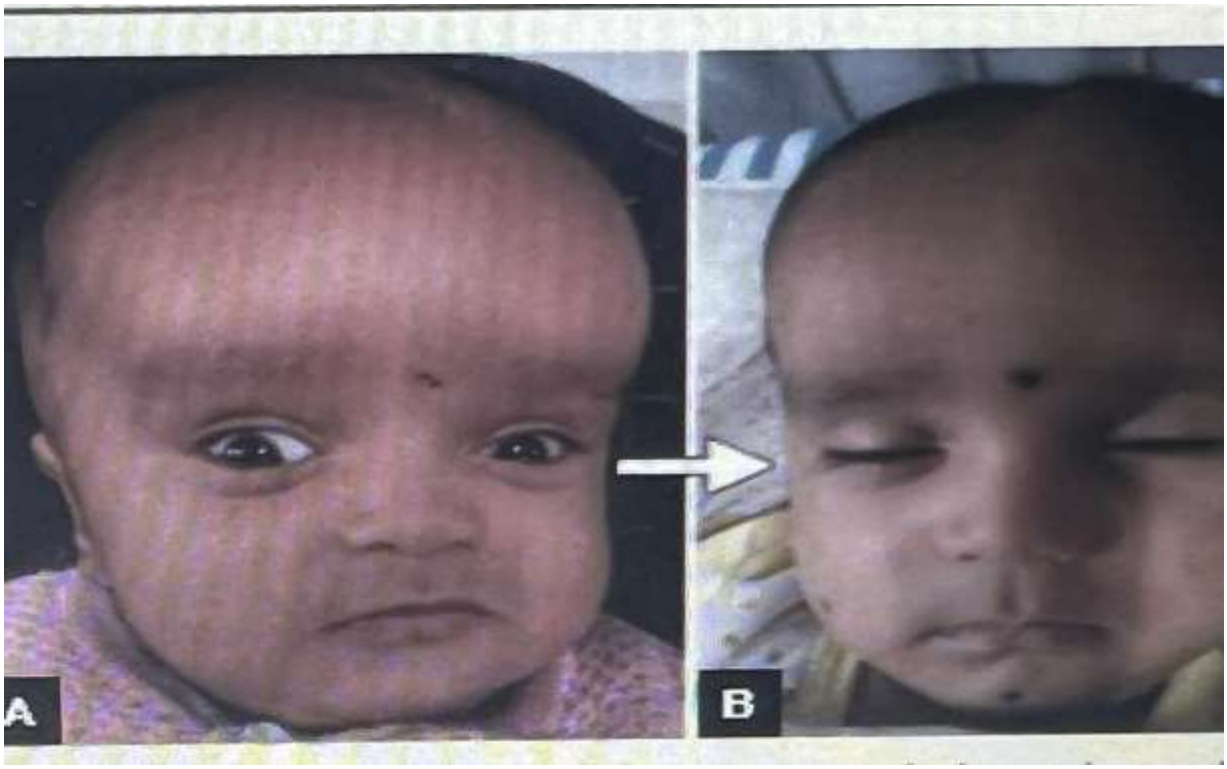

Figure (12): Improvement in macrocephaly post operatively

Results and Discussion:

Outcome of management of patient depend up on death rate, disease prevalence, or evolutional & professional efficiency . Death rate in Fisher's series of 27 cases was 41 percent.[8] And it was 26 percent in sequence of 23 patient documented via Sawaya and Mclaurin.[9]Mortality was 28 percent in sequence of 18 patients documented via Carmel et al.[10] Main reason of dissipate life was pollution of shunt device& cardio respiratory stand still. Second intricacy is a vulnerablein the Dandy – Walker syndrome & has been condole to the temporary task of a rudimentary brain stem. Disease prevalence concern with ventricular device originally concern to mechanical shunt mal function & pollution of shunt device .Ratio of such complication has reduced at this time. We believe malfunction of shunt device is less more by employing the cavity instead of ventricle, for CSF drainage. The maximum excruciating view of Dandy- Walker syndrome is prolong period result . In consequence documented via Carmel et al, only 6 / 13 surviving person were considered to have normal intelligence on follow up.[10] Sawaya and Mclaurin performed detailed psychometric testing on 14 survivors and found that 71 percent had subnormal intelligence (IQ < 83).[9] Although additional investigation[11,12,13] have demonstrated a higher degree of intellectual functioning, the prospective thorough of Dandy- Walker variants to the patient sequence may supportdeviation the evolutionary result of these patients. Availability of accompanied disorder never linked completely with minimum IQ scale unless for the coherently minimum scale of these person with atrophy of the corpus callosum. From these monitoring we can decidedthat the Dandy –Walker syndrome is don't restricted to a mechanical upset of CSF pathway nevertheless explained a additional universal disturbance of CNS growth

Conclusions:

The dandy- walker syndrome is a rare congenital brain abnormality, presenting usually in first year of life with enlargement of head . In older children ,it can present with varied neurological symptoms. It is diagnosed with advanced imaging &treated primarily with CP or VP shunting or both. Prognosis varies due to several factors, but an early diagnosis & appropriate treatment have a good prognostic value.

Recommendations:

Treatment is based on the symptoms resulting from Dandy –Walker syndrome. A careful evaluation by a healthcare provider is essential before initiating treatment. For example , healthcare providers may recommend:

1.VP shunt: surgeons place a VP shunt , a small device to drain excess fluid from the brain. The shunt can reduce pressure on the brain and improve symptom.

2.Medications: Your child provider may prescribe medications to control seizures.

3.Therapy: Physical and occupational therapy can help children maintain muscle strength . Therapists can also teach children new methods of doing their regular activities. Speech therapy can help with language and speech development.

4.Special education: The right learning environment can help children reach educational and social goals.

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