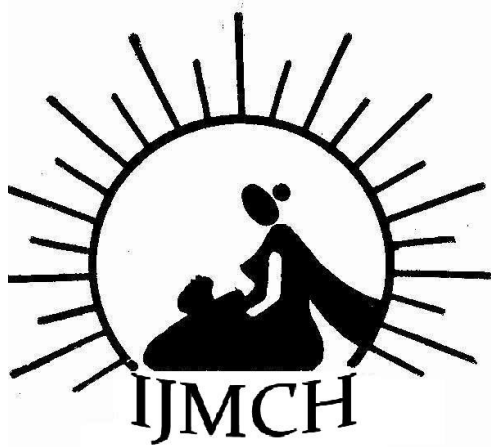


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CASE REPORT

DILEMMAS IN THE MANAGEMENT OF FOETAL HYDROCEPHALUS

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ABSTRACT:

A primigravida presented at 27 weeks of pregnancy with moderate hydrocephalus. She was in the dilemma whether to continue the pregnancy or opt for termination. 32 weeks ultrasonography showed moderate hydrocephalus due to aqueductal stenosis with a V : H ratio 46 %. The prognosis was consulted with pediatric neurosurgeon and was told to be good with respect to survival and newborn would need V.P shunt. Induction of labor was done at 37 weeks and emergency C-section was done for CPD. Head circumference of the newborn was 36 cm. C.T scan of the brain showed dilated lateral and third ventricles. The baby was discharged after 7 days. After 2 months head circumference increased and baby underwent ventriculoperitoneal shunt surgery and is doing fine.

Key words: *Hydrocephalus, Cephalocentesis, Caeserean section, V-P Shunt.*

INTRODUCTION:

Foetal hydrocephalus is a challenging obstetric scenario. A current prevalence of congenital and infantile hydrocephalus is between 0.48 to 0.81/1000 births (live and still)⁽¹⁾. Congenital hydrocephalus is a well known entity which is usually associated with other intracranial and extracranial malformations. The complications related to this condition range from intracranial (hypoplasia of corpus callosum, encephalocele and arachnoid cysts) in 37% cases to extracranial malformations (myelomeningocele, renal agenesis, Fallot's tetralogy, septal defects, cleft palate, Meckel-Gruber Syndrome etc.) in 63 % cases⁽²⁾. The ability to diagnose severe hydrocephalus antenatally by ultrasound means that some cases are prevented by termination⁽¹⁾. But the remaining cases of mild and moderate will always lead to a clinical dilemma as the prognosis is guarded.

CASE REPORT:

Mrs. XY, aged 25 years, primigravida with a married life of 1 year was detected of having moderate hydrocephalus of baby at 27 weeks of gestation and was referred to BLDE Hospital Bijapur. She was perceiving foetal movements well. She had no history of fever with rashes, no history suggestive of severe preeclampsia and urinary tract infection. She was not anemic and there was no edema. She was investigated for TORCH titres and thyroid profile. Toxoplasmosis , IgG and IgM was normal whereas she was IgG positive for Rubella, Cytomegalovirus and Herpes simplex virus. Patient was regularly followed and ultrasonography at 32 weeks showed moderate hydrocephalus due to aqueductal stenosis with a V:H ratio 46% with moderate thinning of parenchyma. The prognosis was consulted with pediatric neurosurgeon and was told to be good with respect to survival and would need post natal CT brain evaluation and mostly need ventriculoperitoneal shunt. Patient was admitted at 37 weeks of gestation for induction of labor.



Figure 1: Antenatal USG picture showing moderate dilatation of both lateral ventricles and 3rd ventricle suggestive of aqueductal stenosis. Moderate compression of cerebral cortex .

At admission: She had no pallor, no edema, no icterus, vital signs were normal, per abdominal examination- Uterus 37 weeks size , relaxed, cephalic presentation, 4/5th palpable with regular foetal heart rate. Per vaginal examination revealed – Cervix 2.5 cm length, closed internal os with Bishop score 5. Moderate CPD was diagnosed by Muller-Munrokkerr method.

She was induced with prostaglandin E₂ gel initially and later by 25 microgram misoprostol tablets 4 to 6th hourly interval for 6 doses and was given a trial of labour. She was taken up for emergency C-section in view of non progress of labor. Per vaginal findings before C- section were Cervix 3 cm dilated, 50% effaced, vertex at -1 station, caput ++. A male baby of 2.6 kg weight with normal apgar score was extracted. Post operative period was uneventful.

On examination of baby : At birth head circumference was 36 cm. Anterior fontanelle was normal 3 x 3 cm size without any bulging, forehead was broad.

Neurosonogram showed moderate dilatation of 3rd and lateral ventricles. 4th ventricle was normal with obstructive hydrocephalus due to aqueductal stenosis. CT brain showed asymmetrical dilatation at lateral and 3rd ventricles with normal 4th ventricle and selectively small posterior fossa, features of corpus callosal dysgenesis associated with periventricular nodular heterotopia.



Figure 2:Hydrocephalic baby at birth

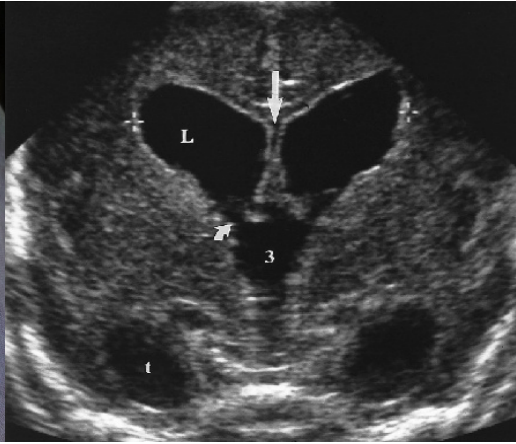


Figure 3: Ultrasonography showing Congenital Hydrocephalus

Figure 3: Ultrasonography showing Congenital Hydrocephalus–Aqueductal Stenosis. Coronal image shows marked symmetric dilatation of the lateral (L) and third (3) ventricles and the foramina of Monro (curved arrow). The temporal horns (t) are prominently dilated. The fourth ventricle was normal in size, indicating congenital aqueductal stenosis.



Figure 4 : CT scan of the brain showing dilated lateral and 3rd ventricles

Baby's head circumference was serially measured during the stay in hospital . It increased by only 0.25 cm. Mother and baby were discharged on 7th day. Later, on follow up of baby there was a 2 cm increase in size of head circumference at 2 months. Baby underwent a ventriculoperitoneal shunt surgery at NIMHANS Hospital Bangalore and is doing fine.

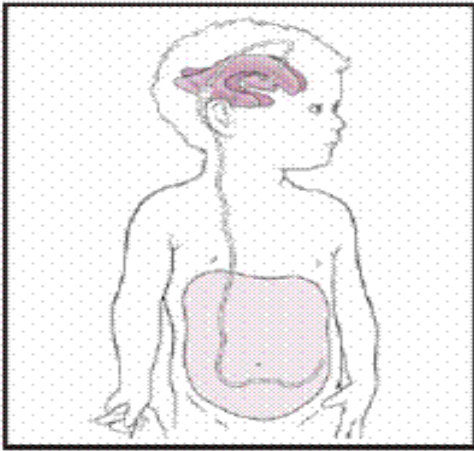


Figure 5: Ventriculoperitoneal Shunt



Figure 6: Ventricles after shunt was placed

DISCUSSION:

Hydrocephalus is not a specific disease, rather it represents a diverse group of conditions. It results from impaired circulation and absorption of CSF or, in the rare circumstances from increased production of CSF by choroid plexus papilloma⁽³⁾. It denotes an increase in size of CSF spaces like ventriculomegaly associated with an increase in intracranial pressure⁽⁴⁾. The development of high resolution 2D real time scanner coupled with increasing expertise has established the role of cranial sonography in the evaluation of neuroanatomy of the brain parenchyma and ventricular system with high precision⁽⁵⁾.

Ventriculomegaly is defined as bilateral enlargement of the ventricular system beyond the upper limit (2SD) of distribution for the normal population of equal age ; in practice the ventricle to hemisphere ratio is used to determine ventriculomegaly⁽⁶⁾. Ventriculomegaly is of greatest clinical significance when shown to be progressive or associated with recognised underlying abnormalities. The causes⁽⁶⁾ of pathological ventriculomegaly are :- 1) Holoprosencephaly results from disordered forebrain development and is characterized by the presence of a single ventricle (usually enlarged), absence of the corpus callosum, major forebrain dysgenesis, and facial abnormalities almost always involving the fetal eyes. The prognosis is extremely poor, perinatal death is common and severe mental retardation is uniform. 2) Ventriculomegaly due to aqueductal stenosis presents a major management dilemma. Perinatal outcome with ventriculomegaly due to aqueductal stenosis is usually, but not invariably, poor. 3) Ventriculomegaly due to major neural tube defect where again prognosis is bad. 4) Unilateral or occasionally bilateral cystic cerebellar degeneration (Dandy-Walker cysts). The prognosis is generally poor. Predisposing factors for the above causes are genetic, environmental, TORCH infections, folic acid deficiency.

The size of lateral ventricle LVW gradually increased as age advanced i.e., from 12.19 ± 1.42 mm at 32 weeks of gestational age to 18.54 ± 1.51 mm at 13 to 18 months of age⁽⁷⁾.

The CT scan or MRI along with USG in an infant are the most important tools to identify the specific cause and severity of hydrocephalus⁽¹⁾.

It has been suggested that the term mild ventriculomegaly should be limited only to cases with measurements of 10 to 12 mm, whereas values of 12.1 to 15 mm should be referred to as moderate ventriculomegaly because they tend to have in general a worse outcome⁽⁸⁾.

Fetuses with isolated mild ventriculomegaly usually have a good outcome, and in most instances the ventricles stabilize or return to normal size throughout gestation⁽⁹⁾. However, these fetuses run an increased risk of neurologic compromise and, in some cases, develop severe cerebral anomalies, including hydrocephalus white matter injury, and cortical plate abnormalities, in the last part of the gestation or after birth⁽¹⁰⁾. The risk is particularly increased when the width is greater than 12 mm, when the dilatation affects both lateral ventricles⁽⁸⁾.

When the diagnosis of severe ventriculomegaly is made before viability, many parents would probably request termination of pregnancy⁽¹¹⁾. In continuing pregnancies, no modifications of standard obstetric management are required. A C-section is recommended only in those cases with associated macrocrania. Cephalocentesis to reduce cranial size is associated with significant morbidity and is indicated only in cases with severe associated abnormalities which are incompatible with postnatal survival or those characterized by the virtual absence of cognitive function⁽¹²⁾. Cephalocentesis is followed by vaginal delivery.

The management of pregnant women with hydrocephalic fetuses presents a dilemma for the treating physician. One side of management scale is represented by termination of the pregnancy once the diagnosis is confirmed. On the other hand, pregnancy may be allowed to continue to term and vaginal delivery is achieved after performing destructive procedures to the baby's head, which may lead to significant maternal morbidity and definite fetal mortality⁽¹³⁾.

In this case there was moderate hydrocephalus with aqueductal stenosis without any CNS malformation. Therefore destructive operation was not done⁽¹⁴⁾ and labour was induced at 37 weeks in an attempt to have vaginal delivery and because of failure of trial of labor emergency caesarian section was performed and a live male baby of good apgar score was extracted. At 2 months after birth, because head circumference increased by 2 cm ventriculoperitoneal shunt surgery was done and baby is fine.

Therapy for hydrocephalus depends on cause. Long term results with medical management like Acetazolamide and Furosemide are disappointing. Most cases require extracranial shunts, particularly a VP shunt (occasionally a ventriculostomy suffices). The major complications of shunting are occlusion (characterised by headache, papilloedema, emesis and mental status change) and bacterial infection (fever, headache, meningismus) usually due to staphylococcus epidermidis. With meticulous preparation shunt infection rate can be reduced to < 5%⁽³⁾.

The prognosis depends on the cause of dilated ventricles. Hydrocephalic children are at increased risk for various developmental disabilities like memory function, visual problems. Most of them are pleasant and mild mannered, while some children show aggressive and delinquent behaviour. It is imperative that hydrocephalic children receive long term follow up in a multidisciplinary setting⁽¹⁵⁾.

CONCLUSION:

Ultrasonographic scanning coupled with good clinical judgment remains the mainstay of management of hydrocephalus. During this time the decision for continuing or terminating the pregnancy is taken after patient counseling. The various treatment options of intrauterine fetal therapy available in developed countries cannot be carried out in our context. In case of isolated hydrocephalus, due to aqueductal stenosis V.P shunts are carried out with good prognosis⁽¹⁰⁾. The use of folic acid 5 mg daily 3 months prior to conception and during the first trimester of pregnancy has been reported to be beneficial in the prevention of neural tube defects⁽¹⁶⁾.

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