

# Congenital Hydrocephalus with Aqueductal Stenosis: A Rare Condition Case Report

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## ABSTRACT

Congenital Hydrocephalus is due to buildup of excess cerebrospinal fluid in the brain at birth. Fluid buildup leads to neurological and psychological damages to the child. Aqueductal stenosis means a narrowing of the passageway between the third and fourth ventricles. In this case report, Ventriculoperitoneal shunt is placed to relieve the pressure, but was removed due to Meningitis. As per review of literature, incidence of shunt failure in infants below 2 years is 5-12% and with more than 60% mortality rate.

**Keywords:** Hydrocephalus, Cerebrospinal fluid, Shunt surgery, Aqueductal stenosis, Large communicating.

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## INTRODUCTION

Congenital hydrocephalus is characterized by abnormal buildup of cerebrospinal fluid (CSF) in the brain in newborns. This excess fluid causes an abnormal widening of ventricle spaces in the brain (ventriculomegalia) and can put harmful pressure on brain tissue. Hydrocephalus symptoms include a large head with a thin, transparent scalp, a bulging forehead with increased spaces between the bones of the skull (fontanelles), and a downward gaze.<sup>1</sup> Seizures, abnormal reflexes, slow heartbeat and respiratory rate, headaches, vomiting, irritability, weakness, and visual problems all are possible symptoms. The most standard treatment is to surgically insert a shunt system to transport the excess CSF and allow for re-absorption. Blindness and mental deterioration may occur if left untreated. Hydrocephalus is categorized based on type of brain defect and pressure of CSF viz., communicating hydrocephalus where there is no blockage in the ventricles but the fluid is not readily absorbed. Noncommunicating (obstructive) hydrocephalus occurs when the CSF is blocked, causing the pathways upstream of the block to widen (dilate), resulting in increased pressure inside the brain. The flow of CSF out of the skull may be obstructed in some cases. Arachnoid cysts are a type of brain growth that can obstruct fluid flow.<sup>2</sup> Other causes include mutations in many other genes, brain and/or spinal cord malformations, infections, bleeding inside the cavities of the brain (intraventricular haemorrhage), trauma, teratogen exposure, or a congenital brain tumor.

## CASE REPORT

A five months old male child, known case of congenital hydrocephalus with aqueductal stenosis with history of right VP shunt placement in March 2021 and removal of shunt due to meningitis with shunt failure in May 2021 and brought for review.

## Chief Complaints

Progressive increase in head size,

Occasional vomiting,

no fever, no seizures

## Past History

Known case of congenital hydrocephalus with aqueductal stenosis

VP shunt placement in March 2021

removal of shunt due to meningitis with shunt failure in May 2021

no other co-morbidities

## Physical Examination

Baby is active and afebrile

PR: 120/Min

BP: 80/50 mmHg

CVS: S1+S2

RS- BAE +

PA- soft

CNS- No FND

## Diagnostic Assessment

Parameters	Abnormal values
Hb%	7.1 g/dl
Hematocrit	22.9 %
MCV	69FL
MCHV	21.5pg
Total WBC count	13,100 cells/cu.mm
ESR	28mm/1 <sup>st</sup> hr
Total RBC	3.6 millions/cu.mm

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**Figure 1:** Real Time Neuro sonography.

### Other investigations

- Real time Neuro sonography were performed (Figure 1)- Grossly dilated bilateral and 3<sup>rd</sup> ventricle, with Impression of Bilateral gross hydrocephalus.
- CT scan of brain- Dilated bilateral lateral and 3<sup>rd</sup> ventricle. [Temporal horns 44mm, 3<sup>rd</sup> ventricle 20mm], Dilated 4<sup>th</sup> ventricle (30mm), white matter differentiation, no evidence of extra axial fluid collections, Bony calvarium is normal, with Impression of Large communicating hydrocephalus.

### Follow-up and Outcomes

According to all the investigations and imaging reports, mentioned above, physician advised ventriculoperitoneal shunt placement and prescribed below medications for symptomatic relief.

### Plan

- First line therapy- shunt placement and endoscopic third ventriculostomy
- Supplementary therapy- Antibiotics to avoid infections and relapse of meningitis
- Diuretics- Acetazolamide to drain excess CSF.

Sl. no	Category	Generic name	Dose	ROA
1	Syp.Ocef	Cephalexin	125mg	PO
2	Syp.Levipil	Levetiracetam	20mg	PO
3	Syp.Paracetamol	Paracetamol	120mg	SOS
4	Inj.Monocef	Cefpodoxmine	250mg	IV
5	Inj.Amikacin	Amikacin	25mg	IV
6	Syp.Bevon	Benfotiamine	15ml	PO

## DISCUSSION

Congenital hydrocephalus with aqueductal stenosis is a rare disease in infants which may alter the structure of brain.<sup>3</sup> Hydrocephalus leads to accumulation of CSF and causes accelerated head growth which is one of the major symptoms in this patient for readmission in hospital. Aqueductal stenosis means narrowing of ventricle of brain which blocks the flow of CSF and increases CSF pressure in infants. Shunt Placement is first line therapy, but Shunt failure is common in majority of cases due to shunt infections, in this case patient has suffered from meningitis and shunt was removed to reduce infection. Usually systemic or intraventricular antibiotic therapy will be prescribed.<sup>4</sup> A study on pediatric population, states that shunt removal in addition to antibiotic treatment and external ventricular drainage is the most effective approach to eradicate shunt infection.

Another treatment option is endoscopic third ventriculostomy (ETV) which is a surgical method that is somewhat helpful in avoiding certain shunt-related complications such as shunt obstruction, infection, or over drainage. Furthermore, physical therapy can help children to improve their growth. Task oriented skills need to be practiced daily such as sitting to standing and also skills should be trained repetitively.

CSF need to be obtained for estimation of CSF biomarkers for assessment of infection in CSF. Based on lab profile Hemoglobin levels are abnormal, treating anemia helps to reduce pressure on body.

Hydrocephalus delays the developmental and behavioral growth of a child. However, it is also important that emphasis is placed on family centered care and treatment in natural environments. The mutual goal is usually to increase functional activity, which in turn, decreases disability.<sup>5</sup> However, in this child, there is a possibility of reduction of symptoms which may delay milestones due to brain cortical growth and if the surgical procedures were delayed, a poor prognosis.

## CONCLUSION

Congenital Hydrocephalus with Aqueductal stenosis is potentially a fatal disease. The aim of this case report is to highlight the morbidity and mortality of the disease and how surgical management will alleviate the patient of the symptoms and complications of the disease. Left untreated Congenital Hydrocephalus with Aqueductal stenosis is a fatal disease with natural progression leading to inevitable death. However, medical intervention also have their attendant complications and a careful watch is necessary.

As a physician Assistant subtle Adjustments in the patient management and correction of minor problems like anemia and hypoproteinemia through proper pharmacological intervention may go a long way hand in hand with the physician to treat the disease with better outcomes.

**Ethical committee approval:** Obtained approval from the hospital ethical committee. We also obtained informed consent from the patient which we have attached with this case report for the journal's reference.

## CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

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