

**PRESENTATION AND MANAGEMENT OF PAEDIATRIC
HYDROCEPHALUS IN TWO ACADEMIC HOSPITALS IN
GAUTENG PROVINCE, SOUTH AFRICA**

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DECLARATION

I, Dr Cyril Agbor declare that this master's research report, which I hereby submit for the degree Master of Medicine in Neurosurgery at the University of Witwatersrand, is my work and has not been submitted previously by me for a degree at another university nor has it been submitted elsewhere for publication.

Sign: 

Date: 21/08/2020

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DEDICATION

I dedicate this work to:

My loving wife, Mrs Grace Cyril Agbor

For standing by me through thick and thin.

My Parents

Chief Napoleon Agbor Ntui & Mrs Francisca Agbor Ntui

For your unwavering love and kindness

ABSTRACT

Background

Hydrocephalus is a leading paediatric neurosurgical condition in the sub - Saharan African region. It is associated with preventable mortality and morbidity that may be related to socio-demographic, clinical and management characteristics. An audit of current outcome of hydrocephalus at selected hospitals in Gauteng may improve practice.

Aim

The aim of the study was to describe the clinical and radiological features of hydrocephalus and its management among children attending Charlotte Maxeke Johannesburg Academic Hospital (CMJAH) and Chris Hani Baragwanath Academic Hospital (CHBAH), Gauteng Province

Methodology

This was a prospective analytic cross-sectional study of newly diagnosed children (aged 0-16 years) with hydrocephalus conducted at the Neurosurgical units of CHBAH and CMJAH between September 2017 to August 2018. Socio-demographic and clinical characteristics were obtained from parents or caregivers using a self-administered questionnaire. Physical examination, assessment of the radiological image of the brain and management of each participant was recorded. Data were analysed using Stata statistical software to determine the pattern and determinants of the presentation and management of the condition.

Results

The median age of the children was 6 (2.5 - 16) months with 79% (n= 113) of them younger than 24 months. The male to female ratio among hydrocephalic children was 1.2:1. About three-fifths (3/5) of the parents only had at most primary level education and about 68.5% were unemployed.

The majority of children (85.8%) below the age of 2 years presented with enlarged head whereas, the majority of children (n=28, 83.3 %) above two years presented with features of raised ICP. Thirty seven percent (37%) previously had meningitis while 21.7% have had spinal bifida and 23% had an abnormal motor function.

During the antenatal period, 7.9% of the mothers had eclampsia while 38% of them took other drugs besides haematinics (such as antiretroviral drugs). Labour was prolonged in 31.5% of cases. Among the babies delivered, 33.6% were preterm and 28.7% having low birth weight. About 16.8% had a recurrent infection, while 0.49% had traumatic brain injury during the neonatal period.

The majority of the patient (32.2%) had communicating hydrocephalus and, aqueductal stenosis (21.7%) was the most prevalent cause of obstructive hydrocephalus. Majority (92.3%) of the patient have operative intervention, mostly ventriculoperitoneal shunt for cerebrospinal fluid (CSF) diversion

Discussion

Paediatric hydrocephalus is a common paediatric neurosurgery condition. It occurs more frequently among children below 24 months of age. Low socioeconomic status and exposure to predisposing factors during pregnancy, labour and postnatal periods lead to hydrocephalus. More than one-third were due to infective processes such as meningitis. The ventriculoperitoneal shunt remains the most commonly used method of diverting cerebrospinal fluid, even though it is fraught with high morbidity.

Conclusion

Reduction in the risk of perinatal and neonatal infections could drastically reduce the risk of meningitis and hydrocephalus in our environment. Public enlightenment campaigns on the risks and management of hydrocephalus among personnel and patients attending peripheral hospitals can help to reduce morbidity and mortality from hydrocephalus as the majority of our patients were delivered at the primary healthcare centres.

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1. CHAPTER ONE: INTRODUCTION

1.1. Background

Hydrocephalus is defined as an increased volume of cerebrospinal fluid that may result in active dilation of the ventricular system, with or without increased intracranial pressure (ICP), due to imbalance between the production and absorption of cerebrospinal fluid (CSF) (1, 2). Hydrocephalus is due to anomalies in the anatomy and/or deranged hydrodynamics of CSF that results in the build-up of CSF in the ventricles. Hydrocephalus can be acute, which occurs over hours or days. It may also be chronic that occurs over months or years. Hydrocephalus can occur as an isolated condition or in association with numerous other neurological conditions and diseases (1).

As early as the 5th century BC, Hippocrates described hydrocephalus as the accumulation of water within the head while Galen showed the association between the CSF and the brain and choroid plexus but failed to relate to the pathophysiology of the hydrocephalus. In the 17th century, Willis explained the production of CSF by choroid plexus and its absorption in the venous system. In 1701, Pacchioni proved that the arachnoid granulations were the site of production of CSF. By the 19th century, the current concept of hydrocephalus was established (3). Hydrocephalus is a condition that represents multiple causes with varying clinical presentation and radiological characteristics (4). Its spectrum might vary with locality. A knowledge of its spectrum of clinical manifestation coupled with timely intervention, could improve the outcome and impact on a better quality of life.

1.2. Literature review

1.2.1. Prevalence

The global prevalence rate of hydrocephalus is about 1.2/1000 children (5). Thus, public health burden of hydrocephalus among the paediatric population cannot be over-emphasised. The prevalence varies significantly across populations and regions, and the estimated annual new cases of infant hydrocephalus in sub-Saharan Africa (SSA) is 100,000 -375,000 (6). Also, the projected annual cost of diagnosis and management of hydrocephalus to the health systems in SSA was \$1.4 to \$56 billion (7).

Furthermore, Warf and Collaboration (7) estimated that between 1000 and 2000 new cases of infant hydrocephalus occur every year in Uganda. The same researchers also predicted the

occurrence of 2,900 to 4,800 new cases of neonatal hydrocephalus each year in Mozambique (7). Although hydrocephalus is not associated with ethnicity or gender in the high-income countries (8), it was found that more males than females were affected in the low income countries, as the male to female ratio was found to be 1.3:1 in Nigeria (9). The age of occurrence of hydrocephalus has been studied. The peak age of hydrocephalus was found to be between one to six months and more than half of the affected children were younger than two years (9). Furthermore, Jaiswal and Jaiswal (10) reported that 38% of cases were congenital, while 62% were acquired (8).

1.2.2. Types of hydrocephalus

Hydrocephalus may be classified into congenital or acquired. Congenital hydrocephalus occurs in utero and it is noted at time of delivery. It may be due to either events or environmental influences that occur during foetal growth, or genetic and/or anatomic abnormalities. Acquired hydrocephalus develops at the time of delivery or some point afterwards. This type of hydrocephalus can affect individuals of all ages and may be caused by injury or disease (11). Hydrocephalus can also be categorised into communicating or non-communicating based on the site of obstruction of the CSF. In the communicating type, there is blockage CSF flow beyond the ventricles. Non-communicating hydrocephalus, otherwise termed obstructive hydrocephalus, occurs when CSF flow is obstructed within the ventricular systems (11).

1.2.3. Aetiology and risk factors

The causes of hydrocephalus vary with the age of the child. In children aged 0 to 2 years, hydrocephalus is usually caused by a perinatal haemorrhage, meningitis, and developmental abnormalities, the most common being aqueductal stenosis and spinal bifida. In children aged 2 to 10 years, the most frequent causes of hydrocephalus are posterior fossa tumours and aqueductal stenosis (12).

Congenital Causes of hydrocephalus

About 55% of all childhood hydrocephalus are congenital (12). Congenital hydrocephalus can result from central nervous system (CNS) malformations, genetic defects, teratogens, intrauterine infection, intraventricular haemorrhage, and trauma (13). A rare aetiology of hydrocephalus is obstruction caused by a congenital CNS tumour, especially if located near the midline. Primary aqueductal stenosis accounts for about 5% of congenital hydrocephalus, whereas aqueductal stenosis secondary to neoplasm, infection, or haemorrhage accounts for another 5% (12). Primary aqueductal stenosis usually presents in infancy. Secondary aqueductal stenosis is caused by gliosis as a result of inflammation consequent to intrauterine infection or germinal matrix haemorrhage (12). Anatomic defects commonly observed with idiopathic congenital hydrocephalus are associated with abnormalities of hindbrain development and include Chiari malformations, Dandy-Walker malformation (DWM), and others. DWM is associated with atresia of the foramen of Luschka and Magendie and affects 2% to 4% of neonates with hydrocephalus. About 50% of all patients with DWM develop hydrocephalus (12).

Congenital hydrocephalus can be part of a major brain malformation, such as an encephalocele or holoprosencephaly, or can be associated with genetically transmitted metabolic diseases, such as achondroplasia and Hurler disease (12). Other causes of congenital hydrocephalus include agenesis of the foramen of Monro, congenital tumours, arachnoid cysts, vascular malformations (vein of Galen), and intrauterine toxoplasmosis (12).

Acquired Causes of hydrocephalus

Hydrocephalus may occur because of central nervous system (CNS) infections. These include meningitis, mostly bacterial, which can result in hydrocephalus by either inflammatory aqueductal stenosis or leptomeningeal fibrosis. In some parts of the world, parasitic disease, such as intraventricular cysticercosis, can cause hydrocephalus by mechanical obstruction (12).

Intracranial bleed into the subarachnoid space or, less commonly, into the ventricular system can predispose to hydrocephalus. Post-haemorrhagic hydrocephalus (PHH) usually occurs after IVH and can be related to prematurity, head injury, or rupture of a vascular malformation. The bleed incites an inflammation followed by fibrosis. The main mechanism for hydrocephalus is impaired absorption of CSF (communicating hydrocephalus), although some

obstruction to CSF flow also may occur. Approximately one-third of extremely low-birth-weight infants with an IVH develop PHH (12).

Intracranial abnormal masses account for 20% of all cases of hydrocephalus in children. Brain tumours, such as medulloblastoma, astrocytoma, and ependymoma, may predispose children to hydrocephalus. Similarly, cysts, abscesses, vascular malformations, or hematomas can also cause hydrocephalus (12).

Increased venous sinus pressure with subsequent impair CSF absorption can also lead to hydrocephalus. This can be related to venous sinus thrombosis, some craniosynostosis or achondroplasia hypertension (12). Iatrogenic causes of hydrocephalus include hypervitaminosis A, which can lead to hydrocephalus by increasing secretion of CSF or by increasing permeability of the blood-brain barrier. Hypervitaminosis A is a more common cause of idiopathic intracranial hypertension (12).

Child-related risk factors have been associated with the development of congenital hydrocephalus. These risk factors include male sex, preterm birth (< 28 weeks), birth weight below the 10th percentile or above the 90th percentile and being firstborn (14).

Currently, South Africa is undergoing health and demographic transitions and has a high prevalence of infectious diseases such as the Human Immunodeficiency Virus infection and tuberculosis which could increase the incidence of hydrocephalus in the country (15). A prospective study in East Africa showed that 57% of paediatric hydrocephalus was related to the infectious process (7). South Africa is a middle-income country with multi-racial demography. Scanty studies currently exist about the epidemiology of hydrocephalus in South Africa.

Poor attendance at prenatal care by women is strongly associated with congenital hydrocephalus among their children(16). Furthermore, maternal hypertension or preeclampsia, and maternal diabetes (pregestational and/or gestational) have been shown to have a strong association with congenital hydrocephalus (17). In addition, maternal exposure to several medications has been implicated in the evolution of congenital hydrocephalus. Such implicated drugs include vaginal metronidazole treatment during the 2nd and 3rd month of pregnancy and first-trimester maternal exposure to antidepressants (14). Maternal alcohol and illicit drug use can also predispose to the development of congenital hydrocephalus (17). A large population-based cohort study showed that there is a significantly increased risk of congenital hydrocephalus in infants with low socioeconomic status (8). Another study further observed

that babies of women who were homemakers were at increased risk of congenital hydrocephalus (18).

1.2.4. Clinical manifestation

The clinical manifestations of hydrocephalus vary and are dependent on factors such as the patient's age, aetiology, rate of progression of raised intracranial pressure (ICP) and any co-existing pathology. The main clinical presentations of hydrocephalus are related to increased ICP (1). For children below two years, head enlargement, headache, fever, altered sensorium, seizures, vertigo, neurological deficit, diminished vision and excessive crying are the typical clinical features. Sleepiness, irritability, sunset eyes, and failure to thrive are other common clinical features in this age group (1).

In children older than two years, the head may not be enlarged since the disease process is occurring after the closure of the fontanelles and sutures. Clinical manifestations usually include blurred vision, diplopia, papilledema or optic atrophy. Other features are abducens nerve paresis, hypothalamic dysfunction (abnormal stature, obesity and diabetes insipidus), poor coordination, gait disturbance, spastic lower extremities with hyperreflexia, low intelligence quotient (IQ) and learning difficulties (1, 19).

The differences in aetiology, pathologies and clinical features of hydrocephalus across populations could be related to socio-economic conditions. Thus, the socio-economic status of the parent of children with hydrocephalus may influence the condition through higher risk factor thereby impacting on the pattern aetiology, presentation pattern or outcome of patients managed at our centre.

1.2.5. Investigation

Radiological imaging is an essential tool in the confirmation of a diagnosis of hydrocephalus and its complications (12). In a new-born, ultrasonography is the preferred modality for the initial examination because it avoids ionising radiation, does not require sedation/anaesthesia, and is usually readily available and portable. Ultrasound is suitable for imaging the lateral ventricles but does not assess the posterior fossa thoroughly; the diagnostic accuracy of ultrasound also depends upon the expertise of the user. Infants found to have progressive ventriculomegaly on ultrasound should generally undergo additional imaging with magnetic resonance imaging (MRI) (12). In older infants and children with suspected hydrocephalus,

computed tomography (CT) or MRI should be performed. These imaging studies may also detect associated central nervous system malformations or tumours (12)

Hydrocephalus is characterised by ventriculomegaly and / or evidence of increased intracranial pressure (ICP). The determination of whether ICP is elevated is based on the clinical and radiographic findings. Radiographically, results that suggest increased pressure include include attenuation of the ventricles, basal cisterns and other CSF spaces, brain herniation, loss of grey-white matter differentiation, enlargement of the recesses of the third ventricle, dilation of the temporal horns of the lateral ventricle. Interstitial oedema of the periventricular tissues (seen on T2-weighted or FLAIR [fluid-attenuated inversion recovery] magnetic resonance imaging (MRI) sequences) and effacement of the cortical sulci (12).

The images in acute communicating (extra-ventricular obstructive) hydrocephalus, may show enlargement of all the ventricles, especially of the third and fourth ventricles with associated trans-ependymal oedema and upward bowing of the corpus callosum. On the other hand, chronic non-communicating (intraventricular obstructive) hydrocephalus may show marked ventriculomegaly, elevated and thinned corpus callosum, fenestration of the septum pellucidum and depression of the fornix. Other radiological features of chronic non-communicating hydrocephalus include inferior displacement of the floor of the third ventricle, ballooning of the recesses and erosion of the Sella turcica (20, 21). Also imaging finding depends of the site of obstruction in the ventricular system; The dilated part of the ventricle if usually proximal to the site of obstruction. (20).

1.2.6. Treatment

Most hydrocephalus are progressive, and neurologic deterioration will occur if the hydrocephalus is not treated. Thus, it is important to ensure timely management of the children with hydrocephalus. The goals of treatment are to restore the CSF dynamics and intracranial pressure (ICP) to levels that is as near to normal as possible and to encourage normal neurologic development. The treatment of hydrocephalus can be expectant, medical or surgical.

Watchful waiting

Asymptomatic patients who do not have elevated intracranial pressure (ICP), and are achieving expected developmental milestones, and do not have severe ventriculomegaly or obvious obstruction of the CSF pathway on neuroimaging can be managed with watchful waiting. During this period, young infants undergo serial head circumference measurements, monthly or bi-monthly, head ultrasounds or magnetic resonance imaging (MRI), and assessment of gross motor skills (22).

Temporising measures

A period of observation or medical management may be initiated, especially in preterm infants, if there is no obstruction or operable lesion and the accumulation of CSF is slight and slowly progressive (1, 20). The temporizing measure may also be indicated in cases with ongoing infection, a life-threatening presentation, and those not stable enough to undergo surgery. Measures include temporary external ventricular drain (EVD) placement, use of diuretics like furosemide and acetazolamide to decrease CSF production (23). Serial lumbar punctures (LP) in preterm infants with post-haemorrhagic hydrocephalus. However, the routine use of periodic lumbar punctures (LP) as a preventive measure in neonates with intraventricular haemorrhage does not appear to be effective is not advised (24). On rare occasions, hydrocephalus stops progressing, so-called "arrested hydrocephalus". The arrest could be due to the development of an alternate route of cerebrospinal fluid (CSF) absorption or because normal mechanisms for CSF management become re-established which in this case, shunting is unnecessary (25).

Definitive treatment

If clinical features of increased ICP evolve and/or there is a progression of ventriculomegaly on imaging, surgical intervention is generally warranted. In many cases, the most effective treatment is surgically diverting CSF, using a shunt or third ventriculostomy. Rare exceptions include cases of hydrocephalus caused by a vein of Galen malformation, embolisation of the malformation may be more appropriate than surgical drainage (22, 26). The timing of surgical intervention in patients with hydrocephalus is determined by the severity of symptoms and the neuroimaging findings. Acute rapidly progressive hydrocephalus requires urgent surgical intervention, typically with a CSF shunt or endoscopic third ventriculostomy (ETV) (23).

Removal of lesion

If the hydrocephalus is due to an anatomic cause such as a resectable brain tumour, the relief of CSF pathway could be achieved by removal of the mechanical obstruction by excising the tumour, often with intraoperative EVD placement (27, 28). If the tumour is unresectable, then a CSF shunt or ETV can be performed to address the hydrocephalus (29).

CSF diversion

However, CSF diversion is required to evacuate excess fluid, relieve the pressure build-up, and treat congenital hydrocephalus in most cases. In moderate to severe hydrocephalus, CSF diversion is performed as early as the child is 'medically fit' to undergo the procedure. The CSF diversion is indicated with hydrocephalus if any of the following are present: symptoms raised ICP, progression of ventriculomegaly and/or obvious obstruction of the CSF pathway evident on neuroimaging. This could be achieved using shunts, or endoscopic third ventriculostomy (30). The choice of diversion procedure is determined by the age of the patient and the underlying cause.

CSF shunt

CSF shunts have long been the standard treatment for hydrocephalus in children. CSF shunt procedure is preferred in patients with a history of IVH, meningitis, or previous shunting, CSF shunt is a mechanical device placed to prevent the excessive accumulation of CSF. The shunt enables CSF to flow from the ventricles into the systemic circulation or to the peritoneum where it is absorbed, bypassing the site of obstruction. The shunt consists of a ventricular catheter, a valve and a distal catheter. The distal end of the system most commonly placed in the peritoneal cavity (VP shunt). Less commonly, the distal catheter is inserted in the right atrium of the heart (ventriculoatrial [VA] shunt) or pleural space (ventriculopleural shunt). The CSF can also be diverted to the cisterns, galea of subdural space. Ventriculoperitoneal (VP) shunt, is mainstay of CSF shunt procedure, though it frequently complicated by infection and/or malfunction.

Endoscopic third ventriculostomy (ETV) is an alternative approach. It has many advantages over CSF shunting in that it is durable, and potentially avoids the long-term shunt complications. ETV is a procedure in which a small fenestration is made in the tuber cinereum on the floor of the third ventricle using an endoscope within the ventricular system

through a burr hole in the skull. This enables the CSF to flow directly to the basal cistern bypassing the obstruction.

Its success depends on the age of the patient, aetiology of hydrocephalus, presence of scar in the basal cistern and pre-intervention complications (31). Generally, patients with third or fourth ventricular outlet obstruction or with clear aqueductal stenosis and for those with pineal region tumours and tectal tumours, respond well to ETV. It is contraindicated in the treatment of obstructive hydrocephalus in infants <3 months old, because the likelihood of success is low (around 25 percent) in this age group. Other contraindications include abnormally narrow prepontine space, remarkably distorted ventricular anatomy, intraventricular haemorrhage, and sepsis involving the ventricles and potential causes of adhesive and fibrosis of cisterns like radiotherapy (20, 32). The ETV success rate is low in patients with a history of intraventricular haemorrhage (IVH), meningitis, or previous shunting. However, if patients with these disorders also have acquired aqueductal stenosis, ETV is attempted prior to pursuing shunting, because there is moderate success with this approach. For children in whom ETV is unsuccessful (i.e., hydrocephalus progresses following ETV), perform a shunting procedure, because repeating the ETV acutely is not likely to be successful in some cases (33). Imaging parameters used in assessing the effectiveness of surgical intervention include - reduction in ventricle size, amount of CSF over the cerebral hemispheres, presence of a flow void in the third ventriculostomy site and degree of periventricular oedema (29).

In spite of the fact that though the complication rate of ETV is low, serious events should always be kept in mind. Understanding of potential surgical complications at each step, careful intraoperative monitoring and procedures, and close postoperative monitoring are necessary to prevent these complications. Patients who had surgical intervention for hydrocephalus need long-term follow-up. Visits are scheduled as appropriate. Neuroimaging studies are typically obtained postoperatively. Postoperative imaging for children who underwent endoscopic third ventriculostomy (ETV) should include magnetic resonance cerebrospinal fluid (CSF) CSF flow study to demonstrate flow through the ventriculostomy.

1.2.7. Complications

There could however be local, systemic or technique and device -related complications of the surgical interventions (11). In general, complications of shunted hydrocephalus are due to malfunction of the shunt. Malfunction may be caused by infection or mechanical failure. Approximately 40% of standard shunts malfunction within the first year of installation, and 5% per year malfunction in subsequent years (34).

Shunt infection occurs in approximately 5 to 15% of procedures (35, 36). This may lead to ventriculitis and may contribute to poor cognitive outcome and death (37). The risk of shunt infections seems to be higher in new-borns compared with older infants and children (38). Perioperative period antibiotic prophylaxis reduced the risk of shunt infection by approximately 50%, and also, the use of antibiotic-impregnated catheters appears to lower the risk of infection (39, 40).

Mechanical failure is most common during the first year after shunt placement. The majority of this failures stem from obstruction at the ventricular catheter (41). Fractured tubing is responsible for shunt failure in approximately 15% of cases. Other causes include shunt migration (partial or complete) and excessive CSF drainage (over drainage). Mechanical failure requires prompt recognition and surgical intervention.

Over drainage can cause functional shunt failure, leading to subnormal ICP with subsequent development of neurologic symptoms such as postural headache and nausea (34). Over drainage greatly reduces the size of the ventricles causing the catheter to lie against the ependyma and choroid plexus, and these tissues block the holes in the catheter. Other common complications are related to the distal site of CSF drainage. include perforation of viscus and intestinal obstruction.

Although, endoscopic third ventriculostomy (ETV) is a safe procedure, a variety of complications have been reported, mostly related with the surgical procedure. This complication could occur intraoperative or postoperative complications of ETV include haemorrhage, injury to neural structures and late sudden deterioration (42). Infection, hematoma, and cerebrospinal fluid leaks may present in the direct postoperative period.

Bouras and Sgouros (42) reported an overall complication rate was 8.8 percent, including intraoperative haemorrhage (3.9 percent), infection (1.8 percent), CSF leak (1.7 percent), and other surgical complications (eg, thalamic infarct and subdural, intracerebral, and epidural

hematoma: [1.1 percent]). Permanent morbidity, including hemiparesis, gaze palsy, memory disorders, altered consciousness, and/or hypothalamic dysfunction, occurred in 2.1 percent. Postoperative mortality was 0.2 percent, and two died at 25 months and 60 months following ETV, due to acute hydrocephalus from stoma blockage.

1.2.8. Prognosis

The prognosis of hydrocephalus depends on the cause, associated disorder(s), the severity of symptoms, and time of diagnosis (34). In untreated hydrocephalus, approximately 50% of affected patients die before three years of age, and about 80% die before reaching adulthood (34). Treatment improves the outcome for hydrocephalus not associated with malignant tumour, with 89% and 95% survival in two reports (38, 43).

Seizures frequently occur in children with shunted hydrocephalus (43, 44). In one report of children treated with ventriculoperitoneal (VP) shunt and followed for a mean of eight years, 32% had epilepsy (44). Seizures may be present at the time the hydrocephalus is diagnosed. However, shunt placement and complications also predispose to epilepsy. The incidence of seizures varies according to the aetiology of hydrocephalus. In the study described above, the risks in patients with infection, intraventricular haemorrhage (IVH), and spina bifida were approximately 50%, 30%, and 7% respectively (44).

Functional outcome is dependent on many factors, including the degree of prematurity, other central nervous systems (CNS) malformations, other congenital abnormalities, and epilepsy, as well as sensory and motor impairments (34). A study reviewed outcomes of 129 children who underwent shunt placement before the age of two years and who were followed up for at least ten years, motor deficits, visual or auditory deficits, and epilepsy occurred in 60%, 25%, and 30% of patients, respectively (43). IQ was >90 in 32% and was <50 in 21%. Attendance at a regular school was possible for 60 %, of the remainder, 31% were in special classes or were institutionalised, and 9% were not considered educable (43). Casey, Kimmings (38) showed that children with hydrocephalus caused by infection or by IVH were more likely to need special education services than those with congenital hydrocephalus (52 and 60% versus 29 %).

A study, IQ was ≥ 85 in 33 %, 70 to 84 in 30%, 50 to 69 in 21 %, and <50 in 16 %. Median IQ was decreased among those who were born preterm compared with term (median IQ score 68 versus 76); among those with isolated hydrocephalus at birth compared with those with

hydrocephalus and myelomeningocele or with acquired hydrocephalus (median IQ score 60 versus 77); and among those with cerebral palsy and/or epilepsy compared with those without (median IQ score 66 versus 78) (45). There was a discrepancy between median verbal and performance IQ (90 and 76, respectively) (45). The discrepancy was also noticed other studies (46).

Socio-demographic characteristics of hydrocephalic patients may also influence the treatment and prognosis of hydrocephalus in our environment. The impact of hydrocephalus on the patient, the healthcare system and the society cannot be overemphasised. Its high prevalence among the paediatric age group in low- and middle-income countries calls for concerted efforts and research on its presentation and management strategies in our environment. Also, the knowledge of its socio-demographic characteristics; clinical and radiological features; treatment and the eventual outcome is critical to the preparation of the health care delivery system.

1.3. Justification of the study

There is a paucity of local research on the epidemiology of hydrocephalus in South Africa. (47), in spite of it substantial physical, economic and financing responsibility on the patients and his/her family, healthcare providers and society (48).

Thus, the public health burden of hydrocephalus among the paediatric population made it imperative to heighten awareness about its risk factors/aetiology, clinical sequelae and management. Hence, adequate allocation of health care resources for its prevention and management in sub-Saharan Africa including South Africa is necessary.

Describing the epidemiology and management of hydrocephalus in South Africa is an important step towards appreciating its enormous burden. Therefore, this study aims to audit paediatric hydrocephalus in South Africa by describing the pattern of presentation and management of children presenting with the condition at two academic centres in Gauteng to improve practice.

1.4. Research question

What are the clinical and radiological presentations of hydrocephalus among children attending Chris Hani Baragwanath and Charlotte Maxeke Johannesburg Academic Hospitals, and how are they managed?

1.5. Study aims and objectives

1.5.1. Aim

The study aims to describe the clinical and radiological presentations and management of hydrocephalus among children attending Charlotte Maxeke Johannesburg Academic Hospital (CMJAH) and Chris Hani Baragwanath Academic Hospital (CHBAH), Gauteng.

1.5.2. Objectives

The specific objectives of the study are

1. To describe the socio-demographic characteristics of children with hydrocephalus
2. To describe the clinical and radiological presentations of hydrocephalus
3. To evaluate the management options of children with hydrocephalus

2. CHAPTER TWO: RESEARCH METHODOLOGY

2.1. Study setting

This study was conducted in the neurosurgical units of CHBAH and CMJAH. These public hospitals manage patients with hydrocephalus in southern Gauteng.

2.2. Study design.

This was is a prospective analytic cross-sectional study of newly diagnosed children with hydrocephalus at the two hospitals from September 2017 to August 2018.

2.3. Study sampling.

All children newly diagnosed with hydrocephalus, were consecutively recruited into the study.

2.4. Sample size calculation

The sample size was calculated using the formula below (49);

$$n = \frac{Z^2 p q}{d^2}$$

where :

n = desired sample size

Z = standard of deviation (95% confidence level i.e. 1.96)

P = expected or prevalence rate of hydrocephalus from previous study

Q = 1-p

d=degree of accuracy desired (0.052)

Using a prevalence of hydrocephalus of 1.5% (50), the sample size will be

$$\begin{aligned} n &= \frac{(1.96)^2 (0.015) (1 - 0.015)}{(0.05)^2} \\ &= 23 \end{aligned}$$

2.5. Study population

Patients aged 0-16 years with newly diagnosed hydrocephalus (based on imaging) at Chris Hani Baragwanath and Charlotte Maxeke Johannesburg Academic Hospitals from September 2017 to August 2018.

2.5.1. Inclusion criteria

- Patients aged 0-16 years with newly diagnosed hydrocephalus using imaging
- Only newly diagnosed and untreated hydrocephalus

2.5.2. Exclusion criteria

- Patients who had been previously treated for hydrocephalus
- Patients outside the included age range

2.6. Recruitment of Participants

Parents or guardians were given the participant's information sheet to read and understand the essence of the study (Appendix I). They were then counselled about the study, after which informed consent obtained (Appendix II). Assent was also obtained from children who were older than seven years (Appendix III). A structured proforma (Appendix IV) was administered to the parent or caregiver of the child. Parents' age, marital status, occupation, ethnicity/race and parity were elicited and recorded. Also, the family history was elicited. Demographic data of the patients such as age and, the gender of the child; the relevant clinical history of the disease, duration of the illness, including developmental history, family history, gestational age at delivery of the patient and any records of imaging investigations were recorded.

Clinical examinations conducted on the participants included: anthropometric measurements using non-stretchable plastic tape to measure the head circumference and length of the baby to the nearest 0.5 cm. The paediatric weighing scale was used to weigh the participants. The weighing machines, (Detector Medic Scale and Salter thermoscale (England)), was validated daily before the commencement of the clinic with a known weight. A thorough general, central nervous system (CNS) and systemic examination were performed, to elicit any associated anomaly. The radiologic findings of the computed tomography and /or magnetic resonance

image scan of each participant were recorded. The treatment administered was documented from the patients' case note.

2.7. Statistical analysis

2.7.1. Data management

The data were transferred from the proforma to an Excel spreadsheet in a password-protected personal computer. The data was checked for consistency of values (data validation). If this is not possible such erroneous/unexpected value will be coded as 'missing'. In addition, double-entry was checked for and the data was cleaned before analysis was commenced.

2.7.2. Data analysis plan

All Information and radiological results obtained were analysed using the STATA 14, (Stata Corp. 2015) statistical software. Descriptive analysis was presented as a frequency percentage and charts. The mean \pm standard deviation or median (interquartile range) was used for presenting normally distributed, and non-normally distributed continuous variables respectively. Student's t-test was also used to compare differences between continuous variables such as age among the acquired and congenital hydrocephalus group. The distribution of data was performed using the Shapiro-Wilk test, continuous data with p-value < 0.05 was considered not normally distributed. Chi-square test was used to test for association between categorical variables such as the race and the type of hydrocephalus. Statistical significance was set at p-value < 0.05 .

2.8. Ethical considerations

The study was commenced after registration with South Africa National Health Research Database (NHRD) and obtaining approval from Human Research Ethics Committee (Medical) at the University of Witwatersrand) (Appendix V) and hospital's Chief Executive Officers (CEO) (Appendix VI and VII). Utmost confidentiality was guaranteed.

3. CHAPTER THREE: RESULTS

3.1. Socio-demographic characteristics of the participants

In all, 143 hydrocephalus babies were recruited with more males (n= 78/143, 54.6%) than female (n= 65/143, 45.5%) (Sex ratio was 1.2: 1). The majority of the children were younger than 2 years (n= 113/143, 79.0%). More than three-fifth of the parents (Mother: 90/143, 62.9%; Fathers:86/143, 60.1%) only had at most primary level education . The majority of the mothers were Blacks (n=136/143, 95.1%) and also unemployed (n= 98/143, 68.5%).

Table 3.1:1: Socio-demographic characteristics of children with Hydrocephalus

Characteristics (N=143)	
Hospital sites, n(%)	n (%)
Charlotte Maxeke Academic JHB Academic Hospital (CMJAH)	61 (42.66)
Chris Hani Baragwanath Academic Hospital (CHBAH)	82 (57.34)
Gender, n(%)	
Male	78 (54.55)
Female	65 (45.45)
Age in months, median (IQR)	
<24	113 (79.02)
≥24	30 (20.98)
Mother's Occupation, n (%)	
Employed	35 (24.48)
Unemployed	98 (68.53)
Student	10 (6.99)
Father's Occupation, n (%)	
Employed	105 (73.43)
Unemployed	32 (22.38)
Student	6 (4.19)
Mother's Educational qualification, n(%)	
None	30 (20.98)
Primary	60 (41.96)
High school	42 (29.37)
Tertiary	11 (7.69)
Father's Educational qualification, n(%)	
None	13 (9.09)
Primary	73 (51.05)
High school	49 (34.27)
Tertiary	8 (5.59)
Ethnicity, n(%)	
Black	136 (95.11)
Coloured	5 (3.50)
White	2 (1.39)

Indians/Asian	0 (0)
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3.2. Types of Hydrocephalus

Figure 3.1 showed that 69.2% (95% CI: 61.1% - 76.3%) of the patients had acquired hydrocephalus whereas , 30.8% (95% CI: 23.7% – 38.9%) had congenital hydrocephalus

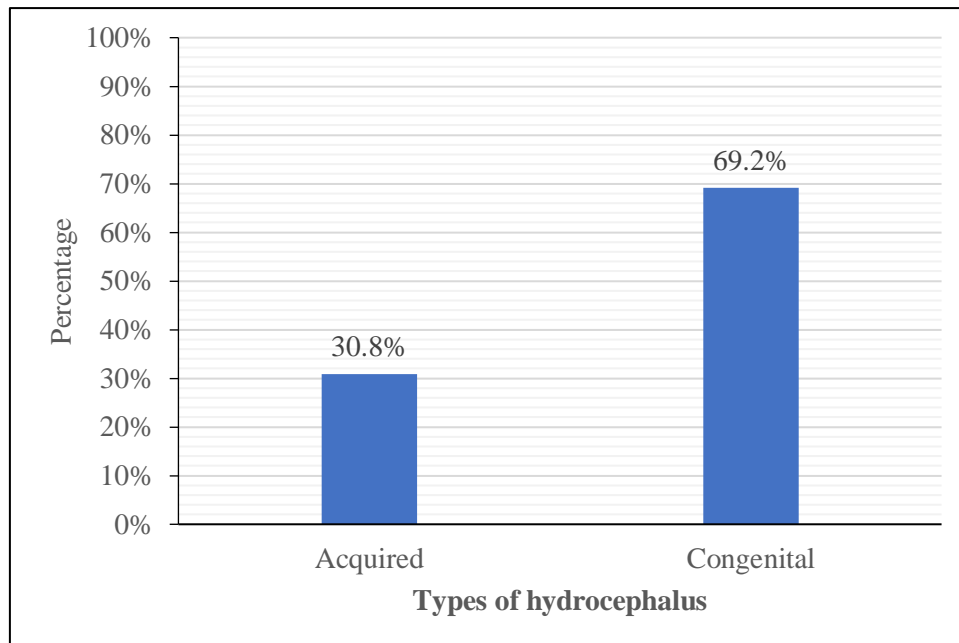


Figure 3.1:1: Types of hydrocephalus

3.3. Clinical and radiological presentations of hydrocephalus

Table 3: 2 and 3:3 below presents the clinical and radiological characteristics of the children.

Table 3.1:2: Clinical presentation of children under two years

Symptoms	Frequency (%)
Rapid increase in head circumference	
Yes	97 (85.84)
No	16 (14.16)
Duration (in days) of rapid increase of head circumference before presentation – if yes (Mean±SD)	60.56±72.93
Abnormal eye gaze	
Yes	59 (52.21)
No	54 (47.79)
Duration (in days) of abnormal eye gaze before presentation – if yes (Mean±SD)	61.68± 80.93
Seizures	
Yes	31 (27.43)
No	82 (72.57)
Duration of Seizures before presentation – if yes (Mean±SD)	18.18 ± 43.84
Vomiting	
Yes	57 (50.44)
No	56 (49.56)
Duration of Vomitting before presentation – if yes (Mean±SD)	8.28±9.48
Excessive sleepiness	
Yes	40 (35.4)
No	73 (64.6)
Duration of Excessive sleepiness before presentation – if yes (Mean±SD)	30.76±31.40
Poor feeding	
Yes	56 (49.56)
No	57 (50.44)
Duration of Poor feeding before presentation – if yes (Mean±SD)	9.95±9.02
Low muscle tone and strength	
Yes	20 (17.7)
No	93 (82.3)
Duration of Low muscle tone and stength before presentation – if yes (Mean±SD)	29.24± 69.88

Table 3.2 above showed that the symptoms of hydrocephalus in children younger than two years were increased head circumference (85.84%), poor feeding (56%) abnormal eye gaze (52.2%) and vomiting (50.4%). The clinical presentation of the children above two years is presented in Table 3.3.

Table 3.1:3: Clinical presentation of children older than two years

Symptoms	Frequency (%)
Headache	
Yes	28 (93.33)
No	2 (6.67)
Duration of headache - if yes (Mean±SD)	154.32± 143.76
Muscle spasm	
Yes	10 (33.33)
No	20 (66.67)
Duration of muscle spasm – if yes (Mean±SD)	147.6± 401.88
Delayed growth	
Yes	16 (53.33)
No	14 (46.67)
Duration of delayed growth – if yes (Mean±SD)	477.06± 432.6382
Trouble eating	
Yes	11 (36.67)
No	19 (63.33)
Duration of Trouble eating – if yes (Mean±SD)	132.91± 203.43
Excessive sleepiness	
Yes	17 (56.67)
No	13 (43.33)
Duration of excessive sleeping - if yes (Mean±SD)	205.76± 610.86
Irritability	
Yes	19 (63.33)
No	11 (36.67)
Duration of Irritability- if yes (Mean±SD)	64± 51.41
Loss of coordination	
Yes	19 (63.33)
No	11 (36.67)
Duration of Loss of coordination- if yes (Mean±SD)	450± 793.95
Loss of bladder control	
Yes	11 (36.67)
No	19 (63.33)
Duration of Loss of bladder control– if yes (Mean±SD)	619.42± 849.74
Larger than normal head	
Yes	9 (30)
No	21 (70)
Duration of larger than normal head – if yes (Mean±SD)	1637.44± 1249.39
Trouble staying awake or waking up	
Yes	4 (13.33)
No	26 (86.67)
Duration of trouble staying awake or waking up – if yes (Mean±SD)	56.5±90.12
Vomitting or Nausea	
Yes	13 (43.33)
No	17 (56.67)

Duration of vomiting or nausea – if yes (Mean±SD)	35± 70.26
Seizure	
Yes	15 (50.00)
No	15 (50.00)
Duration of seizure – if yes (Mean±SD)	211± 398.23
Problem concentrating	
Yes	10 (33.33)
No	20 (66.67)
Duration of problem concentrating – if yes (Mean±SD)	296.5± 323.14
Related Diseases and Conditions	
Meningitis	
Yes	53 (37.06)
No	90 (62.94)
Brain Tumour	
Yes	15 (10.49)
No	128 (89.51)
Epilepsy	
Yes	36 (25.17)
No	107 (74.83)
Spinal Bifida	
Yes	31 (21.68)
No	112 (78.32)
BG	
Yes	2 (1.4)
No	141 (98.6)
Other birth defects	
Yes	16 (11.19)
No	127 (89.44)

Table 3:3 shows the children above the age of two years, had the following symptoms - headache (93.33%), delayed growth (53.3%), excessive sleep (56.67%) irritability (63.33%), loss of coordination and seizure (63.33%) were the leading symptoms among children with hydrocephalus who were older than two years at presentation. The associated conditions that were commonly identified were meningitis (occurred in 37.06% of cases), brain tumour (occurred in 10.49% of cases), epilepsy (25.17%) and spinal bifida (21.68%).

3.4. Management

Table 3:4 outlines the obstetrics characteristics and the sources of referral of mothers of hydrocephalic babies

Table 3.1:4: Obstetrics characteristics and the sources of referral of mothers of hydrocephalic babies

Characteristics	Frequency, n (%)
Antenatal Booking	
Unbooked	4 (2.82)
Booked at Tertiary Health facility	20 (14.08)
Booked at Secondary Health facility	2 (1.41)
Booked at Primary Health facility	115 (80.99)
Booked at Health Centre	1 (0.7)
Diabetes Mellitus	
Yes	4 (2.82)
No	138 (97.18)
History of maternal pyrexia	
Yes	20 (14.08)
No	122 (85.92)
Skin Rash	
Yes	6 (4.2)
No	137 (95.8)
Exposure to radiation	
Yes	8 (5.71)
No	132 (94.29)
History of drug use in pregnancy	
Routine haematinics only	88 (61.97)
Routine haematinics and anti-retroviral drugs	47 (33.1)
Routine haematinics and anti-hypertensive drugs	6 (4.23)
Routine haematinics and Anti-tuberculosis drugs	1 (0.7)
Ante natal care was uneventful?	
Yes	121 (86.43)
No	6 (4.29)
Antepartum haemorrhage (APH)	1 (0.71)
Bled at 27weeks	1 (0.71)
Eclampsia	1 (0.71)
Pregnancy-induced hypertension (PIH)	10 (7.14)
Gestational age at delivery, Mean (SD)	36.3 ± 4.2
Preterm	48 (33.57)
Term	95 (66.43)
Place of Delivery	
Home	3 (2.1)
Tertiary	30 (20.98)
Secondary	2 (1.4)
Primary	104 (72.73)

Health centre	1 (0.7)
Private	3 (2.1)
Pre-labour Rupture of Membrane	
Yes	45 (31.47)
No	98 (68.53)
Mode of delivery	
Normal vaginal	96 (67.13)
Assisted vaginal	8 (5.59)
Caesarean section	39 (27.27)
Extended resuscitation	
Yes	71 (49.65)
No	72 (50.35)
Birth weight, Median (IQR)	2.9 (2.3 - 3.2)
<1.50	13 (9.09)
1.50 – 2.49	28 (19.58)
≥ 2.5	102 (71.33)
Maternal risk factors for sepsis	
Yes	95 (66.43)
No	48 (33.57)
Neonatal history	
Major illness	
Yes	31 (21.68)
No	112 (78.32)
Recurrent infections	
Yes	24 (16.78)
No	119 (83.22)
Exposure to HIV	
Yes	48 (33.57)
No	95 (66.43)
Exposure to Tuberculosis	
Yes	33 (23.08)
No	110 (76.92)
Operation surgical from other indication exception of hydrocephalus	
Yes	17 (11.89)
No	126 (88.11)
Traumatic Brain injury	
Yes	15 (10.49)
No	128 (89.51)
Immunisation	
Yes	120 (83.92)
No	23 (16.08)
Developmental Milestone	
Yes	62 (43.36)
No	81 (56.64)
Smoking Status	

Smokers	18 (12.59)
None Smokers	125 (87.41)
History of consanguinity	
Yes	1 (0.7)
No	142 (99.3)
Family history of hydrocephalus	
Yes	6 (4.26)
No	135 (95.74)

Table 3.4 shows that 80.99% of mothers to the patient with hydrocephalus had antenatal care at the primary health centre. During the pregnancy, 2.82% of the mothers had gestational diabetes mellitus, 7.85% had eclampsia, 7.85% had pregnancy-induced hypertension, 38% took other drugs besides haematinics. During antenatal care, the commonest drugs aside the haematinics were antiretroviral drugs. Labour, in most cases (n=104, 72.72% was at primary health care, with prolonged rupture of membrane occurred in 31.47% of cases. About 66.43% of the babies were delivered at term. Average birth weight was 2.9%, with 9.09% had very low birth weight, while 19.58% has a birth weight ranging between 1.50 and 2.49. During the neonatal period, 16.78% had the recurrent infection, 33.57% were exposed to HIV, and 10.49% had a traumatic brain injury. About 4.26% had it in the family history of hydrocephalus.

Table 3.1:5: Physical Examination findings

Head shape	N (%)
Normal	98 (87,72)
Dolichocephaly	3(2.65)
Brachycephaly	4 (3.54)
Plagiocephaly	8 (7.08)
Sutures	
Present	103 (72.03)
Absent	40 (27.97)
Overriding	
Not overriding	
Fontanelle	
Sunken	3 (2.63)
Flat	40 (35.09)
Bulging	71 (62.28)
Scalp Vein	
Present	73 (64.6)
Absent	40 (35.4)

Table 3.1:5: normal head shape was seen (n=98, 87.7205%), patent sutural line (n =103, (72.03%), enlarge scalp vein in (n=73,64. 60 %).

Table 3.1:6: Neurological Examination

Characteristics	Frequency	%
Motor Function		
Reflexes		
Decreased	31	21.68
Normal	55	38.46
Increased	57	39.86
Muscle strength		
Decreased	32	22.38
Normal	56	39.16
Increased	55	38.46
Muscle tone		
Decreased	31	21.99
Normal	55	39.01
Increased	55	39.01
Sensory Status		
Touch Sensation		
Decreased	3	2.1
Normal	140	97.9
Increased	-	-
Eye		
Vision		
Normal	101	70.63
Blur	29	20.28
Abnormal	13	9.09
Gaze		
Up	18	12.59
Down	62	43.36
Lateral	63	44.06
Hearing		
Decreased	6	4.2
Normal	136	95.1
Increased	1	0.7
Movement Status		
Co ordination		
Normal	70	60.34
Abnormal	46	39.66
Balance		
Normal	67	57.76
Abnormal	49	42.24

As outlined in Table 3.6, about 23% had an abnormal motor function, 29.38% had impaired vision and 4.02% had hearing loss

Table 3.1:7: Features of radiological investigation among children with hydrocephalus

Radiological features	Frequency (%)
Ventriculomegaly	
Yes	143 (100)
No	0 (0)
Transependymal oedema	
Yes	53 (37.06)
No	90 (62.94)
Thin and elevation of the corpus callosum	
Yes	102 (71.33)
No	41 (28.67)
Fenestration of septum pellucidum	
Yes	131 (91.61)
No	12 (8.39)
Ballooning of recess	
Yes	138 (96.5)
No	5 (3.5)
Sella turcica erosion	
Yes	63 (44.06)
No	80 (55.94)
Cortical thickness	
Yes	24 (16.78)
No	119 (83.22)
Calcification	
Yes	13 (9.09)
No	130 (90.91)
Mass	
Yes	5 (3.5)
No	138 (96.5)
Holoprosencephaly	
Yes	15 (10.49)
No	128 (89.51)
Dandy-Walker Malformation	
Yes	29 (20.28)
No	114 (79.72)
Arnold Chiari malformation	
Yes	11 (7.69)
No	132 (92.31)
Agenesis of the corpus callosum	
Yes	23 (16.08)
No	120 (83.92)
Blood in ventricle	
Yes	8 (5.59)
No	135 (94.41)

Table 3.1.7 all the patients in my study had ventriculomegaly on imaging, while 37.6% had trans ependymal oedema, 0.49% holoprosencephaly, 72.3% of the patients had thinning and elevation of the corpus callosum, (10.49%) had Dandy-Walker Malformation, and 7.69% Arnold Chiari malformation.

Table 3.1.8: Classification of Hydrocephalus Diagnosis

Diagnosis	Frequency (%)
Communicating hydrocephalus	46 (32.17)
Intraventricular haemorrhage (IVH)	3 (2.1)
Tumours	9 (6.29)
Aqueductal stenosis	31 (21.68)
Arnold chiari malformation (ARM)	9 (6.29)
Meningomyelocele (MMC)	16 (11.19)
Dandy walker syndrome (DWS)	22(15.38)
Infection	6 (4.2)
Cyst	1 (0.7)

Table 3.8 shows 32.17% of the patients had communicating hydrocephalus. Aqueductal stenosis accounted for 21.68%, Dandy walker syndrome for 15.38%, myelomeningocele (MMC) for 11.19%, intracranial tumours for 6.29%, Arnold Chiari malformation for 6.29%, infection for 4.20%, intraventricular haemorrhage (IVH) for 2.10%, and cysts for 0.70%. of diagnosis.

Table 3.9 below shows the treatment options, type of surgical intervention and postoperative complication.

Table 3.1:9 Treatment options, types of surgical interventions and complications

Method	n (%)
Non operative	11(7.7%)
Operative	132 (92.3%)
Cerebrospinal fluid ((CSF) diversion	
Ventriculoperitoneal shunt (V-P Shunt)	102 (92.3)
Ventriculo subgaleal shunts (VSG)	2 (1.4)
External ventricular drainage(EVD	14(9.79)
Endoscopic third ventriculostomy (ETV)only	1(0.70)
Endoscopic third ventriculostomy (ETV) in combination with other CSFdiversion procedures	13(9.09)
Endoscopic pellucidostomy	1(0.70)
Relief of obstruction	
Yes	46 (95.83)
No	2 (4.17)
Postoperative Complication	
None	13 (54.7)
Sepsis	9 (37.5)
Subdural haematoma (SDH)	1 (4.17)
Subdural empyema (SDE)	1 (4.17)

Table 3.9 shows that eleven patients (7.7%) were managed non-operatively while, 132 cases (92.3%) had surgical intervention. Among those who had surgery, 102 cases (77.27%) had ventriculoperitoneal shunt (VPS), two cases (2%) ventriculo sub-galea shunt (VGS, 14 cases (14.73%) underwent endoscopic third ventriculostomy (ETV) and one patient endoscopic pellucidostomy (0.07%). Also, 95.83% had relieved of obstruction. Following surgical intervention, 45.83% of cases developed complications. Of all patients that underwent surgery, 37.50% had sepsis, and 4.17 each had a subdural haematoma and subdural empyema.

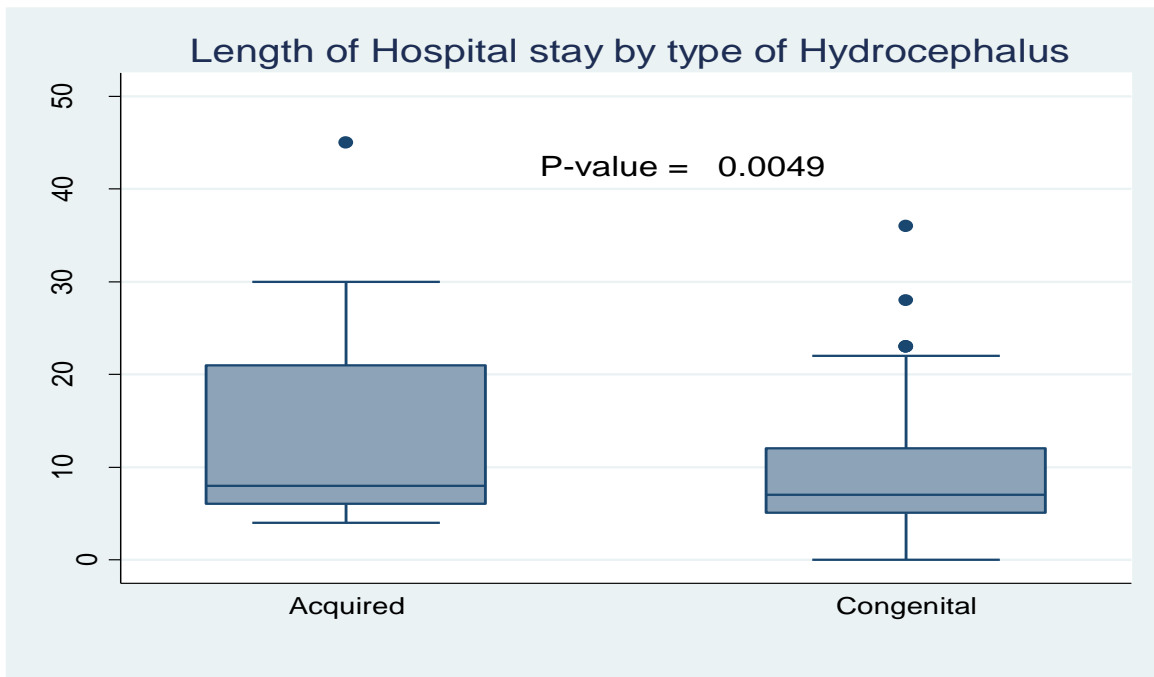


Figure 3.1:2: Length of hospital stay by type of hydrocephalus

The median length of hospital stay was lower among babies with congenital hydrocephalus (7, IQR = 5-12 days) compared to babies with acquired hydrocephalus (8, IQR= 6-21 days), p-value = 0.0049 see figure 3.4 above.

4. CHAPTER FOUR: DISCUSSION

4.1. Socio-demographic characteristics

In all, a total of 143 cases of paediatric hydrocephalus cases were reviewed during the period. 54.55% of our study population were males, giving a male to female ratio of 1.2: 1. Our result is comparable to a similar work conducted in Nigeria, where 54.55% of the children were males. The average age of the children presenting with hydrocephalus in this study was six months; however, Yusuf, Omokanye (9) reported the average age of three months which was lower as compared to our research, possibly because the target population were children aged one and below (9). Our study recruited participants aged one month to sixteen years. The majority (79%) of our participants were aged two years and below. This is not surprising because hydrocephalus in paediatric is predominantly a disease of infancy with symptoms presents at a younger age (51-53).

The majority of the parent in our study attained at best primary level education. The majority of the mothers were Blacks (n=136/143, 95.1%) and about (n= 98/143, 68.5%) were unemployed. The poor educational level and unemployment account for the low socioeconomic status of the parent to the children in my study. Low socioeconomic status with deprivation in health information, poor sanitation and malnutrition increases susceptibility to intrauterine infection before and during pregnancy and other risk factors thus predisposing to hydrocephalus. This collaborates with the finding in the study by Stein, Feldman (54) who examined socioeconomic status as a predictor for congenital hydrocephalus. Although women with low socioeconomic status are at higher risk of having an infant with low birth weight (55), the association between socioeconomic status and congenital hydrocephalus persisted after adjusting for birth weight. The extent to which low socioeconomic status is associated with an increased risk of congenital hydrocephalus is still unknown. It is s necessary to ascertain the degree to which this socioeconomic variation may be associate with other individual base factor variables such as infant's race/ethnicity, maternal age to cause paediatric hydrocephalus.

The majority of the parent are have lower or no income tend to use these patronize facilities our study centres health services are subsidised, and the bills are cheap. This account for the high percentage of the parent in this study been in the low socio-economic class.

Our study shows that hydrocephalus is a frequent disorder in this Gauteng province affecting mostly children below 24 months age mostly male. It also showed most of the parent were of

low socioeconomic status. Therefore, in order to detect the hydrocephalus early, surveillance of paediatric patient delivered by parents of low socioeconomic background should be routine during medical visits.

4.2. Clinical presentations of hydrocephalus

The presentation of hydrocephalus differs in children age two and below compared with the older children. The key reason for the difference in presenting symptoms by age group is because hydrocephalus predates the closure of the cranial sutures and obliteration of the fontanelle in children younger than two years. Thus, the tendency to present commonly with abnormal head enlargement, which was noted in 97% of our cases. Other clinical symptoms are often subtle included general irritability (51%), poor feeding (56%) and slow attainment of milestones. Clinical signs include bulging of the fontanelle, wide separation of the cranial sutures, prominence of scalp veins, and “setting sun” of the eyes. The finding in our study is in line with the works by Kumar and later Anand who showed that enlargement of the head, was the most common feature in infants and young children. In older children and adults, the closure of sutures and fontanelles precede the onset of hydrocephalus, leading to symptoms raised ICP, and headache (10, 56).

Antenatal and perinatal maternal characteristics can cause congenital hydrocephalus. Thus, in a 10-year retrospective study, Van Landingham, Nguyen (17), found the risk factors of hydrocephalus included lack of prenatal care, multiparous gestation, maternal diabetes, maternal chronic hypertension, maternal hypertension during gestation and alcohol use during pregnancy. Nonetheless, our study showed that almost all (97.89%) mothers of our patients with hydrocephalus booked for and had antenatal care at a primary health centre. However, some mothers in our study population had diabetes mellitus (2.8%), eclampsia and pregnancy-induced hypertension (7.85%), which had been identified as risk factors for hydrocephalus (17). Also, we found that about 38% of the mothers had other chronic medications (such as anti-retroviral drugs, anti-tuberculous and anti-hypertensive) during antenatal care. Since South Africa has one of the highest prevalence of HIV, it is expected that a higher percentage of women attending antenatal care may be on anti-retroviral drugs as it occurred in this study. Association between antenatal ingestion of ARVs and hydrocephalus should be clarified in future studies.

The majority of the children 66.43% were delivered at term. However, the 72.72% were delivered at the primary health centre. The primary health care staff lack the skills and facilities to identify a high-risk patient or diagnose hydrocephalus early. This may result in a delay in early diagnosis and referral for treatment, with associated morbidity.

Perinatal and postnatal infections are important pathological factors for the evolution of hydrocephalus. Hence, we found that prolonged rupture of membrane (PROM) occurred in 31.47% of hydrocephalus in our study. PROM predisposes to neonatal sepsis that may result in hydrocephalus. Average birth weight was 2.9%. About 9.09% had very low birth weight, while 19.58% has a birth weight ranging between 1.50 and 2.49

Our study showed that 16.78% of the babies with hydrocephalus had a recurrent infection during the neonatal period. The neonatal infection rate from our report is however lower than the rate of 25.8% that was reported El Awad and Al-Barki (57), in Saudi Arabia. There was a retrospective study with a very small sample size of 46 patients, about a third our sample size. Gauteng an HIV endemic environment, about 33.57% of the babies were exposed to HIV. Nevertheless, the direct role of HIV in the evolution of hydrocephalus still requires further clarifications.

In this study, only 4.26% had previous family history hydrocephalus. Our finding contrast with the report by Ali and Abdelaal (58), in Egypt, who report a family history of 20.8% in their hydrocephalic cases. The very high incidence of family history in Egypt may be related to the reported higher rate of consanguinity in that environment. Shawky, Elsayed (59) showed that the offspring of consanguineous unions might be at increased risk of genetic disorders, higher in mental retardation and hydrocephalus.

Hydrocephalus resulting from meningitis was very high among our study population as compared to the report from Norway (60) (37% vs 2%) because Norway is a high-income country with the low rate of childhood infectious diseases. Moreover, the health systems in Norway can quickly treat and better prevent morbidity from infectious diseases as compared to the South African Public health hospitals.

Congenital hydrocephalus accounted for 69.23% of cases in our study population. The findings tend to agree with similar work by Yusuf, Omokanye (9) in Nigeria, in which congenital hydrocephalus constituted 68% of the study sample. However, Jaykar and Patil (61) in their study, reported only 40% of cases been congenital with the remaining being acquired. Jaykar

and Patil (61) study population included all age group unlike the aged 0 – 14 included in our study; also, the small sample may not be representative of the total population. In our study population, congenital hydrocephalus was more common in males than females. This is consistent with previous studies (18, 62). The underlying causes for the difference male to female ratio remain unclear.

South Africa, being a post-apartheid nation, most blacks, especially women, have no formal education and are unemployed, poor financing status, poorly nourished and lack adequacy health information and access to health facilities. With a high predisposition to infections and attendance risk of congenital hydrocephalus.

This study, showed that 23% of the children had an abnormal motor function, 29.38% had impaired vision and 4.02% hearing loss Hoppe-Hirsch, Laroussinie (43) in France, reported an outcome at ten years was evaluated in 129 consecutive children with hydrocephalus without tumour who had shunt placement before two years of age. Motor deficits, visual or auditory deficits, and epilepsy occurred in 60, 25, and 30% of patients, respectively (43). The value of these studies rests in the unselected nature of the patient sample, homogeneity of medical management, and long duration of follow-up. The functional disabilities that are complicated hydrocephalus can be averted if hydrocephalus is diagnosed early and treated appropriately.

4.3. Radiological findings of hydrocephalus

Brain imaging remains the diagnostic investigation of choice. This may show ventricular enlargement and often the aetiology of the hydrocephalus. The images of the patient in our study showed all of the children had ventriculomegaly, while 37.6% transependymal oedema (TEO). The number of patients with ventriculomegaly was expected because all cases recruited, were clinically confirmed hydrocephalus before imaging. Relatively fewer cases had transependymal oedema (TEO) because many of our patients were < 2 years, thus expansion of cranial cavity delayed the appearance of the TEO.

Majority of the patient in this study had imaging findings in keeping with raised intracranial pressure evidence by thinning and elevation of corpus callosum (71.39%), and ballooning of ventricular recesses (96.50%). This not surprising as majoring had non-communicating hydrocephalus with ventricular obstruction coupled with the preponderance of under two years of age. The under two years of age have a relatively immature brain with high compliance.

Communicating hydrocephalus was seen in 32.17% while, non-communicating hydrocephalus comprises the remaining 67.9%. Ofori, Gyamfua (63) in Ghana reported communicating and non-communicating types of hydrocephalus to be 49.0% and 47.8% respectively. The difference in the finding is because of the wide age range of their study population. The cranial imaging may show hydrocephalus that communicating or presence of structural abnormality that might be related to the hydrocephalus.

Imaging revealed that 32.17% of the our patients had communicating hydrocephalus with the cause not known while aqueductal stenosis accounted for 21.68%, Dandy walker syndrome for 15.38%, myelomeningocele (MMC) for 11.19%, intracranial tumours for 6.29%, Arnold Chiari malformation for 6.29%, infection for 4.20%, intraventricular haemorrhage (IVH) for 2.10%, and cysts for 0.70%. The mean age of hydrocephalus in our study for six months, with 79.02% of the cases below two years of age. Bentaleb, Sahel (64), reported post meningitis 27 %, intracranial, tumours 20%, myelomeningoceles 17 %, Dandy-Walker spectrum Malformations 11%, aqueductal stenosis 5 % with the undetermined being 20 %. The mean age of paediatric hydrocephalic in their study was seven years. The difference in the pattern of aetiology is due to the disparity in mean age. This also affirms to the fact that intracranial tumour tends to cause commoner cause of hydrocephalus with increasing age in paediatric cases.

The Evans ratio could not be deployed because most of the patients came with scans done at referral centres.

4.4. Management options of children with hydrocephalus

Surgical treatment is the mainstay of management in the majority of cases of hydrocephalus. Endoscopic third ventriculostomy (ETV) is the preferred treatment of choice, where appropriate. ETV has the advantage of being minimally invasive and having a lower complication rate (65). A ventriculoperitoneal shunt is a popular device used in cerebrospinal fluid diversion, although it associated with a high complication rate. Ventriculoperitoneal shunt was the method used in 77.2% of our patients in diverting cerebrospinal fluid, and this is similar to other studies (9, 66) unlike in our study, 95.83% had relieved of obstruction.

The low rate of ETV interventions in our study could be as a due to recent acquisition of the equipment and slow learning curve. Almost all the cases that underwent ETV CSF diversion were revised. Lack the expertise, especially amongst the doctors in training (registrar) who are

often bestowed with the responsibility of diverting CSF in paediatric with hydrocephalus. Also, non-availability of ETV services after working hours therefore children that requires emergency CSF diversion cannot benefit from ETV intervention. Lastly, some of our hydrocephalus could best be treated using VPS. Complication following surgical intervention occurred in 45.83% of cases. Of these patients, 37.50% had sepsis, and 4.17 each had a subdural haematoma and subdural empyema.

4.5. Length of hospital stay

The median duration of hospital stay was 7 (IQR: 6 - 13) days. The minimum and maximum length of hospital stay were 0- 45 days and about three-quarters (n= 108/142, 78.06%) of the babies were discharged within 14 days of admission. Lam, Srinivasan (65) reported length of stay (LOS) of 18.2 ± 28.5 days. The male sex, the black race, and Medical aid patients had a higher mean total costs than female and reported congenital hydrocephalus length of hospital stay (65).

The median length of hospital stay was lower in babies with congenital hydrocephalus (7, IQR = 5-12 days) compared to babies with acquired hydrocephalus (8, IQR= 6-21 days), P-value = 0.0049. The reason for the statistically significant maybe because of the high proportion of the children having congenital hydrocephalus. An earlier study reported the average length of stay in the management of congenital hydrocephalus as 14 days (65), which is double the length of stay compared to our study.

South Africa, like other developing countries, face an enormous burden of paediatric hydrocephalus due to high birth rates and greater risk of neonatal infections (7). This burden is related to more general global health challenges, including, infectious diseases, maternal and perinatal risk factors, and education gaps. Unique challenges of the treatment of hydrocephalus in the developing world include a preponderance of post infectious hydrocephalus, limited resources, and restricted access to neurosurgical care. In the 21st century, several organisations have established programs that provide hydrocephalus treatment and neurosurgical training in Africa, Central and South America, Haiti, and Southeast Asia. These international efforts have employed various models to achieve the goals of providing safe, sustainable, and cost-effective treatment. The Gauteng government needs to invest most in health education, facilities and workforce to reduce the mortality and morbidity associated with childhood hydrocephalus.

5. CHAPTER FIVE: CONCLUSION

Hydrocephalus is the most common paediatric neurologic condition with significant implications for the patient and society. Lack of consistent epidemiological data has negatively affected the awareness of the disease and promoted disproportionate allocation of resources for the care of patients with hydrocephalus and research. This work has highlighted the presentation and management of paediatric hydrocephalus. It has described the socio-demographic characteristics, clinical and radiological presentations of hydrocephalus.

It also bring to the fore the social and medical factors predisposing to among other sepsis. Congenital hydrocephalus tends to be the common type, clinical features varying with age. The ventriculoperitoneal shunt is the most common device used in cerebrospinal fluid diversion

Neurodevelopmental disorders hydrocephalus cause significant disability and need continuous and expensive medical care (shunting procedures, etc.) (8). Therefore, it is necessary to observe their predisposing and causative factors and associated defects to implement effective preventive measures. In addition, a proper nationwide register for all congenital malformations should be instituted.

Greater commitment from the paediatric neurosurgery community, increased funding, public education, surgeon training, and ongoing surgical innovation will be needed to address the global burden of untreated hydrocephalus meaningfully.

5.1. Recommendation

We recommended that the Gauteng province, RSA should:

1. Health education as regards risk factors, diagnosis and prompt treatment. health campaigns to increase community awareness about hydrocephalus and promote early reporting
2. Ministry of health should increase availability of up to date facilities including ETV in paediatrics neurosurgical units of CMJAH and CHBAH and strengthen them
3. Training of staff and provide facilities especially at the health centres to enhance early diagnosis and prompt referral.
4. Screening or multicentre study to provide prevalence of hydrocephalus, pattern of occurrence, nature, identify causes and associated risk factors and ultimately to prevent or reduce the occurrence of hydrocephalus.
5. Implement a community-based national surveillance system for congenital abnormalities.

6. Further researches are recommended to confirm the extend risk factors such as HIV, and medications contribute to hydrocephalus in our locality pattern

This study provides an insight into the burden, pattern and management of hydrocephalus in Gauteng province, Republic of South Africa (RSA) and suboptimal treatment of children with hydrocephalus.

5.2. Strength of the study

1. It is a prospective study.
2. Site of study being institutions where most children with hydrocephalus receive treated in Gauteng.

5.3. Limitations of the study

1. There was a challenge in recruiting the patients with hydrocephalus that presents as an emergency, especially at night, before intervention due to the dual study site.
2. In older children, the parent may have difficulties remembering detail on the antenatal and neonatal history, a form of information/recall bias.
3. This study was a cross-sectional study with inadequate follow-up. The associations observed in the study may not be assumed to be causal because of the cross-sectional nature of the study.

REFERENCES

1. Nielsen N, Breedt A. Hydrocephalus. In: Cartwright Cathy C, Wallace Donna C, editors. Nursing care of the pediatric neurosurgery patient. Verlag Berlin Heidelberg: Springer; 2007. p. 29-65.
2. Abou-Hamden A, Drake JM. Hydrocephalus. In: Albright A Leland, Pollack Ian F, Adelson P David, editors. Principles and practice of pediatric neurosurgery. New York: Thieme; 2015. p. 89-99.
3. Aronyk KE. The history and classification of hydrocephalus. *Neurosurgery Clinics*. 1993;4(4):599-609.
4. Pople IK. Hydrocephalus and shunts: what the neurologist should know. *Journal of Neurology, Neurosurgery & Psychiatry*. 2002;73(suppl 1):i17-i22.
5. Garton H, Piatt JJ. Hydrocephalus. *Pediatric Clinics of North America*. 2004;51(2):305-25.
6. Warf BC, Alkire BC, Bhai S, Hughes C, Schiff SJ, Vincent JR, Meara JG. Costs and benefits of neurosurgical intervention for infant hydrocephalus in sub-Saharan Africa. *Journal of Neurosurgery: Pediatrics*. 2011;8(5):509-21.
7. Warf BC, Collaboration EANR. Pediatric hydrocephalus in East Africa: prevalence, causes, treatments, and strategies for the future. *World Neurosurgery*. 2010;73(4):296-300.
8. Jeng S, Gupta N, Wrensch M, Zhao S, Wu YW. Prevalence of congenital hydrocephalus in California, 1991-2000. *Pediatric Neurology*. 2011;45(2):67-71.
9. Yusuf AS, Omokanye HK, Adeleke NA, Akanbi RO, Ajiboye SO, Ibrahim HG. Management and outcome of infantile hydrocephalus in a tertiary health institution in Nigeria. *Journal of Neurosciences in Rural Practice*. 2017;8(2):249.
10. Jaiswal A, Jaiswal J. Incidence of hydrocephalus in pediatric age in a tertiary care centre of Chhattisgarh. *Journal of Evolution of Medical and Dental Sciences*. 2015;4(83):14564-71.
11. Adam R, Fisher C, Hakim S, Ojemann R, Sweet W. Symptomatic occult hydrocephalus with "normal" cerebrospinal fluid pressure. *New England Journal of Medicine*. 1965;273:117-26.
12. Abou-Hamden A, Drake JM. Hydrocephalus and Arachnoid Cysts. In: Swaiman Kenneth, Ashwal Stephen, Ferriero Donna, Schor Nina, Finkel Richard, Gropman Andrea, Pearl Phillip, Shevell Michael, editors. Swaiman's pediatric neurology: principles and practice. Edinburgh: Elsevier; 2017. p. 226-32.

13. Schrander-Stumpel C, Fryns J-P. Congenital hydrocephalus: nosology and guidelines for clinical approach and genetic counselling. *European Journal of Pediatrics*. 1998;157(5):355-62.
14. Kalyvas AV, Kalamatianos T, Pantazi M, Lianos GD, Stranjalis G, Alexiou GA. Maternal environmental risk factors for congenital hydrocephalus: a systematic review. *Neurosurgical Focus*. 2016;41(5):E3.
15. Kahn K. Population health in South Africa: dynamics over the past two decades. *Journal of Public Health Policy*. 2011;32(1):S30-S6.
16. Carmichael SL, Shaw GM, Nelson V. Timing of prenatal care initiation and risk of congenital malformations. *Teratology*. 2002;66(6):326-30.
17. Van Landingham M, Nguyen TV, Roberts A, Parent AD, Zhang J. Risk factors of congenital hydrocephalus: a 10 year retrospective study. *Journal of Neurology, Neurosurgery & Psychiatry*. 2009;80(2):213-7.
18. Munch TN, Rasmussen M-LH, Wohlfahrt J, Juhler M, Melbye M. Risk factors for congenital hydrocephalus: a nationwide, register-based, cohort study. *Journal of Neurology, Neurosurgery and Psychiatry*. 2014;85(11):1253-9.
19. Shallat RF, Pawl RP, Jerva MJ. Significance of upward gaze palsy (Parinaud's syndrome) in hydrocephalus due to shunt malfunction. *Journal of Neurosurgery*. 1973;38(6):717-21.
20. Muir RT, Wang S, Warf BC. Global surgery for pediatric hydrocephalus in the developing world: a review of the history, challenges, and future directions. *Neurosurgical Focus*. 2016;41(5):E11.
21. Kartal MG, Algin O. Evaluation of hydrocephalus and other cerebrospinal fluid disorders with MRI: an update. *Insights into Imaging*. 2014;5(4):531-41.
22. Jea A, Bradshaw TJ, Whitehead WE, Curry DJ, Dauser RC, Luerssen TG. The high risks of ventriculoperitoneal shunt procedures for hydrocephalus associated with vein of Galen malformations in childhood: case report and literature review. *Pediatric Neurosurgery*. 2010;46(2):141-5.
23. Alexander E, Davis CH. Macewen's Sign: "The cracked pot sound". *Annual Review of Hydrocephalus*: Springer; 1990. p. 38-9.
24. Mazzola CA, Choudhri AF, Auguste KI, Limbrick DD, Rogido M, Mitchell L, Flannery AM. Pediatric hydrocephalus: systematic literature review and evidence-based guidelines. Part 2: management of posthemorrhagic hydrocephalus in premature infants. *Journal of Neurosurgery: Pediatrics*. 2014;14(Supplement_1):8-23.

25. Haridas A, Tomita T. Hydrocephalus in children: Management and prognosis: Uptodate; 2018 [Available from: <https://www.uptodate.com/contents/hydrocephalus-in-children-management-and-prognosis>].
26. Schneider SJ, Wisoff JS, Epstein FJ. Complications of ventriculoperitoneal shunt procedures or hydrocephalus associated with vein of Galen malformations in childhood. *Neurosurgery*. 1992;30(5):706-8.
27. Bogdahn U, Lau W, Hassel W, Gunreben G, Mertens H, Brawanski A. Continuous-pressure controlled, external ventricular drainage for treatment of acute hydrocephalus—evaluation of risk factors. *Neurosurgery*. 1992;31(5):898-904.
28. Habib HA. Intraoperative precautionary insertion of external ventricular drainage catheters in posterior fossa tumors presenting with hydrocephalus. *Alexandria Journal of Medicine*. 2014;50(4):333-40.
29. Nikas DC, Post AF, Choudhri AF, Mazzola CA, Mitchell L, Flannery AM. Pediatric hydrocephalus: systematic literature review and evidence-based guidelines. Part 10: Change in ventricle size as a measurement of effective treatment of hydrocephalus. *Journal of Neurosurgery: Pediatrics*. 2014;14(Supplement_1):77-81.
30. Kulkarni AV, Riva-Cambrin J, Holubkov R, Browd SR, Cochrane DD, Drake JM, Limbrick DD, Rozzelle CJ, Simon TD, Tamber MS. Endoscopic third ventriculostomy in children: prospective, multicenter results from the Hydrocephalus Clinical Research Network. *Journal of Neurosurgery: Pediatrics*. 2016;18(4):423-9.
31. Siomin V, Cinalli G, Grotenhuis A, Golash A, Oi S, Kothbauer K, Weiner H, Roth J, Beni-Adani L, Pierre-Kahn A. Endoscopic third ventriculostomy in patients with cerebrospinal fluid infection and/or hemorrhage. *Journal of neurosurgery*. 2002;97(3):519-24.
32. Ogiwara H, Dipatri AJ, Alden TD, Bowman RM, Tomita T. Endoscopic third ventriculostomy for obstructive hydrocephalus in children younger than 6 months of age. *Child's Nervous System*. 2010;26(3):343-7.
33. Beni-Adani L, Biani N, Ben-Sirah L, Constantini S. The occurrence of obstructive vs absorptive hydrocephalus in newborns and infants: relevance to treatment choices. *Child's Nervous System*. 2006;22(12):1543-63.
34. Chumas P, Tyagi A, Livingston J. Hydrocephalus—what's new? *Archives of Disease in Childhood-Fetal and Neonatal Edition*. 2001;85(3):F149-F54.
35. Drake JM, Kestle JR, Milner R, Cinalli G, Boop F, Piatt J, Haines S, Schiff SJ, Cochrane DD, Steinbok P. Randomized trial of cerebrospinal fluid shunt valve design in pediatric hydrocephalus. *Neurosurgery*. 1998;43(2):294-303.

36. Langley JM, LeBlanc JC, Drake J, Milner R. Efficacy of antimicrobial prophylaxis in placement of cerebrospinal fluid shunts: meta-analysis. *Clinical Infectious Diseases*. 1993;17(1):98-103.
37. Shang F, Xu Y, Wang N, Cheng W, Chen W, Duan W. Diagnosis and treatment of severe neurosurgical patients with pyogenic ventriculitis caused by gram-negative bacteria. *Neurological Sciences*. 2018;39(1):79-84.
38. Casey A, Kimmings E, Kleinlugtebeld A, Taylor W, Harkness W, Hayward R. The long-term outlook for hydrocephalus in childhood. *Pediatric Neurosurgery*. 1997;27(2):63-70.
39. Klimo P, Van Poppel M, Thompson CJ, Baird LC, Duhaime A-C, Flannery AM. Pediatric hydrocephalus: systematic literature review and evidence-based guidelines. Part 6: Preoperative antibiotics for shunt surgery in children with hydrocephalus: a systematic review and meta-analysis. *Journal of Neurosurgery: Pediatrics*. 2014;14(Supplement_1):44-52.
40. Ratilal B, Costa J, Sampaio C. Antibiotic prophylaxis for surgical introduction of intracranial ventricular shunts: a systematic review. *Journal of Neurosurgery: Pediatrics*. 2008;1(1):48-56.
41. Kahle KT, Kulkarni AV, Limbrick Jr DD, Warf BC. Hydrocephalus in children. *The Lancet*. 2016;387(10020):788-99.
42. Bouras T, Sgouros S. Complications of endoscopic third ventriculostomy. *World Neurosurgery*. 2013;79(2):e9-S. e12.
43. Hoppe-Hirsch E, Laroussinie F, Brunet L, Sainte-Rose C, Renier D, Cinalli G, Zerah M, Pierre-Kahn A. Late outcome of the surgical treatment of hydrocephalus. *Child's Nervous System*. 1998;14(3):97-9.
44. Bourgeois M, Sainte-Rose C, Cinalli G, Maixner W, Malucci C, Zerah M, Pierre-Kahn A, Renier D, Hoppe-Hirsch E, Aicardi J. Epilepsy in children with shunted hydrocephalus. *Journal of Neurosurgery*. 1999;90(2):274-81.
45. Brookshire BL, Fletcher JM, Bohan TP, Landry SH, Davidson KC, Francis DJ. Verbal and nonverbal skill discrepancies in children with hydrocephalus: a five-year longitudinal follow-up. *Journal of Pediatric Psychology*. 1995;20(6):785-800.
46. Pappas A, Adams-Chapman I, Shankaran S, McDonald SA, Stoll BJ, Laptook AR, Carlo WA, Van Meurs KP, Hintz SR, Carlson MD. Neurodevelopmental and behavioral outcomes in extremely premature neonates with ventriculomegaly in the absence of periventricular-intraventricular hemorrhage. *JAMA Pediatrics*. 2018;172(1):32-42.
47. Jaraj D, Rabiei K, Marlow T, Jensen C, Skoog I, Wikkelsø C. Prevalence of idiopathic normal-pressure hydrocephalus. *Neurology*. 2014;82(16):1449-54.

48. Tasbasi FY. The Experienced Problems of Mothers Having Children with Hydrocephalus: A Qualitative Study. *International Journal of Caring Sciences*. 2015;8(2):435.
49. Daniel W. *Biostatistics: A Foundation for analysis in the health sciences*, 7th edR Wiley. New York. 1999.
50. Garne E, Loane M, Addor M-C, Boyd PA, Barisic I, Dolk H. Congenital hydrocephalus—prevalence, prenatal diagnosis and outcome of pregnancy in four European regions. *European Journal of Paediatric Neurology*. 2010;14(2):150-5.
51. Breuning-Broers JM, Deurloo JA, Gooskens RH, Verkerk PH. At what age is hydrocephalus detected, and what is the role of head circumference measurements? *The European Journal of Public Health*. 2013;24(1):32-4.
52. Musau CK, Nganga HN, Mbuthia NK. Management and functional outcome of childhood hydrocephalus at the Kenyatta national hospital, Nairobi. *African Journal of Neurological Sciences*. 2015;34(1):26-35.
53. Saidu SA, Maaji SM, Nzeh DA, Shehu BB, Ismail NJ. Sonographic pattern of hydrocephalus among the under five children in Sokoto North Western Nigeria. *Sahel Medical Journal*. 2015;18(4):172.
54. Stein SC, Feldman JG, Apfel S, Kohl SG, Glenn Casey A. The epidemiology of congenital hydrocephalus. *Pediatric Neurosurgery*. 1981;8(4):253-62.
55. Subramanian S, Chen JT, Rehkopf DH, Waterman PD, Krieger N. Comparing individual-and area-based socioeconomic measures for the surveillance of health disparities: a multilevel analysis of Massachusetts births, 1989–1991. *American Journal of Epidemiology*. 2006;164(9):823-34.
56. Kumar R, Singh V, Kumar MV. Shunt revision in hydrocephalus. *The Indian Journal of Pediatrics*. 2005;72(10):843-7.
57. El Awad ME, Al-Barki AA. Infantile hydrocephalus in southern Saudi Arabia. *Journal of Family & Community Medicine*. 1997;4(2):71.
58. Ali M, Abdelaal M. Epidemiological study of Congenital Hydrocephalus in Sohag Governorate. *The Egyptian Journal of Community Medicine*. 2015;33(2):49-55.
59. Shawky RM, Elsayed SM, Zaki ME, El-Din SMN, Kamal FM. Consanguinity and its relevance to clinical genetics. *Egyptian Journal of Medical Human Genetics*. 2013;14(2).
60. Paulsen AH, Lundar T, Lindegaard K-F. Pediatric hydrocephalus: 40-year outcomes in 128 hydrocephalic patients treated with shunts during childhood. Assessment of surgical outcome, work participation, and health-related quality of life. *Journal of Neurosurgery: Pediatrics*. 2015;16(6):633-41.

61. Jaykar RD, Patil SP. Indications of ventriculoperitoneal shunt: a prospective study. *International Surgery Journal*. 2017;4(4):1319-26.
62. Persson E-K, Anderson S, Wiklund L-M, Uvebrant P. Hydrocephalus in children born in 1999–2002: epidemiology, outcome and ophthalmological findings. *Child's Nervous System*. 2007;23(10):1111-8.
63. Ofori EK, Gyamfua AO, Tettey KD, Brakohiapa EK, Angmorte SK, Oblitey J. Hydrocephalus cases in Ghana: a six-year (2007-2012) review of computed tomography (CT) radiological reports. *South African Radiographer*. 2018;56(1):15-9.
64. Bentaleb D, Sahel Z, Elkhadir K, LAOUDIYI D, Chbani K, Salam S, OUZIDANE L, editors. *Imaging of hydrocephalus in childhood* 2019: European Congress of Radiology 2019.
65. Lam SK, Srinivasan VM, Luerssen TG, Pan I-W. Cerebrospinal fluid shunt placement in the pediatric population: a model of hospitalization cost. *Neurosurgical Focus*. 2014;37(5):E5.
66. Wali MB, Alenezi MS, Saad AA, Alanazi AS, Alanazi WA, Al Shakarh MK, Alenezi AT, El-Fetoh NMA, Alanazi OMM, Hussain MA. Congenital Hydrocephalus in Arar, Northern Saudi Arabia. *The Egyptian Journal of Hospital Medicine*. 2018;71(3):2651-5.

APPENDIX ONE: ETHICS APPROVAL



R14/49 Dr C Agbor

HUMAN RESEARCH ETHICS COMMITTEE (MEDICAL) CLEARANCE CERTIFICATE NO. M170805

NAME: Dr C Agbor
(Principal Investigator)
DEPARTMENT: School of Clinical Medicine
Department of Surgery
Division of Neurosurgery
Charlotte Maxeke Johannesburg Academic Hospital

PROJECT TITLE: Presentation and management of paediatric hydrocephalus in two academic hospitals in Gauteng Province, South Africa

DATE CONSIDERED: 25/08/2017

DECISION: Approved unconditionally

CONDITIONS:

SUPERVISOR: Dr J Ouma

APPROVED BY: 
Professor PE Cleaton-Jones, Chairperson, HREC (Medical)

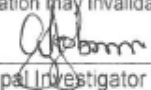
DATE OF APPROVAL: 04/10/2017

This clearance certificate is valid for 5 years from date of approval. Extension may be applied for.

DECLARATION OF INVESTIGATORS

To be completed in duplicate and **ONE COPY** returned to the Research Office Secretary on 3rd floor, Phillip V Tobias Building, Parktown, University of the Witwatersrand, Johannesburg.

I/We fully understand the conditions under which I am/we are authorised to carry out the above-mentioned research and I/we undertake to ensure compliance with these conditions. Should any departure be contemplated from the research protocol as approved, I/we undertake to resubmit to the Committee. I agree to submit a yearly progress report. The date for annual re-certification will be one year after the date of convened meeting where the study was initially reviewed. In this case, the study was initially reviewed in August and will therefore be due in the month of August each year. Unreported changes to the application may invalidate the clearance given by the HREC (Medical).


Principal Investigator Signature

17/10/2017
Date

PLEASE QUOTE THE PROTOCOL NUMBER IN ALL ENQUIRIES

APPENDIX TWO: PERMISSION TO CONDUCT STUDY



GAUTENG PROVINCE

HEALTH
REPUBLIC OF SOUTH AFRICA

MEDICAL ADVISORY COMMITTEE
CHRIS HANI BARAGWANATH ACADEMIC HOSPITAL

PERMISSION TO CONDUCT RESEARCH

Date: 7 Aug 2017

TITLE OF PROJECT: Presentation and management of paediatric hydrocephalus in two academic hospitals in Gauteng Province, South Africa

UNIVERSITY: Witwatersrand

Principal Investigator: C Agbor

Department: Neurosurgery

Supervisor (If relevant): J Ouma


Permission Head Department (where research conducted): Yes


Date of start of proposed study: Aug 2017

Date of completion of data collection: Dec 2019

The Medical Advisory Committee recommends that the said research be conducted at Chris Hani Baragwanath Hospital. The CEO /management of Chris Hani Baragwanath Hospital is accordingly informed and the study is subject to:-

- Permission having been granted by the Human Research Ethics Committee of the University of the Witwatersrand.
- the Hospital will not incur extra costs as a result of the research being conducted on its patients within the hospital
- the MAC will be informed of any serious adverse events as soon as they occur
- permission is granted for the duration of the Ethics Committee approval.


.....
Recommended
(On behalf of the MAC)
Date: 07 August 2017


.....
Approved/Not Approved
Hospital Management
Date: 08/08/17



GAUTENG PROVINCE

HEALTH
REPUBLIC OF SOUTH AFRICA

CHARLOTTE MAXEKE JOHANNESBURG ACADEMIC HOSPITAL

Enquiries:
Ms. N. Mzila
Office of the Clinical Director
Tell: (011) 488-4812
Email: Nolwazi.Mzila@gauteng.gov.za
17 November 2017

GP_201710_030

Dear Dr. C. Agbor

STUDY TITLE: Presentation and Management of Paediatric Hydrocephalus in Two Academic Hospitals in Gauteng, Province South Africa.


Permission is granted for you to conduct the above recruitment activities as described in your request provided:

1. Charlotte Maxeke Johannesburg Academic Hospital will not anyway incur or inherit costs as result of the said study.
2. Your study shall not disrupt services at the study sites.
3. Strict confidentiality shall be observed at all times.
4. Informed consent shall be solicited from patients participating in your study.

Please liaise with the HOD and Unit Manager or sister in charge to agree on the dates and time that would suit all parties.

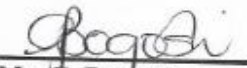
Kindly forward this office with the results of your study on completion of the research.

~~Supported / not supported~~


Dr. M.L. Mofokeng
Clinical Director

DATE: 17/11/2017

Approved/not approved


Ms. G. Bogoshi
Chief Executive Officer

Date: 20.11.2017

APPENDIX THREE: INFORMATION SHEET

Title: Presentation and management of paediatric hydrocephalus at two academic hospitals in Gauteng province, South Africa.

Principal investigator: Dr Cyril Agbor

Gooday Sir/Madam. I am a postgraduate medical doctor conducting research with the above title. This research is part of the educational activities for the award of a professional postgraduate degree (M.Med degree). The study seeks to describe the common symptoms and medical problems encountered by children suffering from hydrocephalus. The type of treatment that is given to them in our environment will also be elicited. . The study hopes to assist the managing doctors and policy makers to know the various risk factors of the disease in our environment.

You are therefore invited to participate in this study because your child has been diagnosed with hydrocephalus. Your participation will be highly invaluable and will not be taken for granted

Study procedures and what the study entails:

Your participation in this study will involve asking you questions about age, your ethnicity, the details of current and relevant past medical history of the child's and other contributing factors. Your child will then be weighed and his/her height and head circumference measured and recorded. The radiological result and treatment that would be offered your child will also be recorded.. All these information will then be collated for all the participants, from where the presentation and management would be obtained.

Risks:

There is no envisaged risk to you and your child in the conduct of this research. All the questions that will be asked are not intrusive and we hope that none of the question would make you feel uncomfortable. However, you are at liberty to decline to answer any question without any untoward consequence for such. Your personal information and that of your child will not be shared with anybody. The information obtained will be stored in a secure computer.

Benefits

Although you will not benefit directly from participating in this study, your participation will make significant contributions to the information and knowledge base of Hydrocephalus in children. This will surely aim at improving how doctors take care of children with this disease.

Participation is voluntary:

Please note that your participation in the study is absolutely voluntary and it will not adversely affect the care your child will receive in the hospital. You are also free to withdraw from the study at any time without any penalty.

Reimbursements:

There is no reimbursement

Confidentiality:

The information that you volunteered to us and receive that extracted from the casefile of your ward will be kept confidential and would not be disclosed to a third party. We will only aggregate all the information of the participants and then analyses. Your information that will not include your name will be available to only the research team, which includes the researcher, his supervisor, biostatistician and occasionally a member of the ethical committee, for the purpose of analysis and verification.

If you have any queries about this study, you may call the researcher Dr Cyril Agbor on 0603224006. Or you may visit the researcher at the Department of Neuro surgery, Charlotte Maxeke Academic Hospital, Park town, Johannesburg.

APPENDIX FOUR: INFORMED CONSENT

Title: Presentations and management of paediatric hydrocephalus in two academic hospitals in Gauteng province, South Africa.

Principal Investigator: Dr Cyril Agbor

I, having read and fully comprehend the information sheet attached, that explained the essence and procedure of the above titled research do volunteer to be a participant in the study.

Participant's name (Please Print)

Participant's Signature /right thumbprint

Date / Time

Project investigator

Date / Time

APPENDIX FIVE: ASSENT CONSENT

Title: Presentations and management of pediatric hydrocephalus in two academic hospitals in Gauteng province, South Africa.

Principal Investigator: Dr Cyril Agbor

SIGNATURE OF CHILD

I _____ (*Name of child*) have read the information sheet about the study and I do agree to participate in the above named research study.

Child's Signature Date

Signature of Parent / Guardian Date

Investigator Signature Date

APPENDIX SIX: QUESTIONNAIRE

Title: Presentation and management of paediatric hydrocephalus in two academic hospitals in Gauteng province, South Africa.

Date.... Time

Venue of history taking clinic () ward () others ()

Hospital CMJAH () / CHBAH ()

Referral source.....

Date of admission..... Date of discharge

A. HISTORY

A1. DEMOGRAPHIC CHARACTERISTICS

Name (Initials)... Hospital Number

Age ... () Sex – Male () Female ()....

Race..... Blacks (), Coloureds (), Indians/Asians (), Whites (), others (), (please specify)

Highest educational attainment - Nil education () Primary school () High school () Tertiary education ()

Father's occupation

Mother's occupation

A2. HISTORY OF ILLNESS

	Yes	No	Duration
Children < 2 years			
• rapid increase in head circumference			
• abnormal eyes gaze			
• seizures			
• extreme fussiness			
• vomiting			
• excessive sleepiness			
• poor feeding			
• low muscle tone and strength			
Children > 2`years			
• headaches			

•muscle spasms			
•delayed growth			
•trouble eating			
•extreme sleepiness			
•irritability			
•loss of coordination			
•loss of bladder control			
•larger than normal head			
•trouble staying awake or waking up			
•vomiting or nausea			
•seizures			
•problems concentrating			
Related Diseases & Conditions			
•Meningitis			
•Brain Tumour			
• Seizure (Epilepsy)			
• Spina Bifida and Anencephaly (Neural Tube Defects			
• Other Birth Defects (Specify)			

A3. PRENATAL AND BIRTH HISTORY

Antenatal history:-		
	Yes	No
▪Maternal Diabetes mellitus		
▪Did the mother have fever during pregnancy that lasted for 1 week?		
Did the mother have skin rashes during pregnancy		
▪Exposure to radiation		
▪Apart from blood enhancing (haematinics) drugs, did you take any other drug?	Yes () If yes, how old was the pregnancy when the drug was taken? What was the name of the drug?	No ()
Was the antenatal care uneventful NB: Uneventful antenatal care means there was no adverse event during antenatal care as recorded from the response above.	Yes ()	No ()
Natal history:-		
•Gestational age at delivery.		
•Place of delivery.		

•Mode of delivery: -		
Prolong Rupture of Membrane		
Normal vaginal ()	Assisted vaginal ()	Caesareans section()
•Resuscitation	Yes	No
•Birth measurements:-		
Weight () Kg	Length () cm	Head Circumference () cm
•Maternal risk factors for sepsis (maternal UTI, maternal fever...)		
A4. NEONATAL HISTORY		
Past medical history (Previous History - Medical & Surgical) :-		
• Major illnesses		
• Recurrent infections		
• Exposure to HIV		
• Exposure to TB		
• Operations		
• Traumatic Brain		
A5. IMMUNIZATION HISTORY		
Vaccination History		
A6. DEVELOPMENTAL HISTORY		
Physical growth;		
Weight ()	Height ()	Head Circumference ()
Developmental mile stones.		
A7. SOCIAL HISTORY (Social history of family - environmental risks);		
Smoking at home	Yes ()	No ()
A8. FAMILY HISTORY.		
Consanguinity	Yes ()	No ()
Any similar condition in any member(s) of the family	Yes ()	No ()

B. PHYSICAL EXAMINATION

B1. ANTHROPOMETRIC MEASUREMENT (Measure or plot on growth chart)			
	WEIGHT (kg)	HEIGHT (cm)	HEAD CIRCUMFERENCE (cm)
Measure < 2 years.			
Plot > 2 years (10-25th percentile) (50th percentile) (50th percentile)
B2. HEAD			
General shape			
Normal ()	Dolichocephaly ()	Brachycephaly ()	Plagiocephaly ()
Sutures			
	Presence	Yes ()	No ()
	overriding	Yes ()	No ()
Fontanelle(s)			
	Size		
	Sunken	Flat	Bulging & Tension
Scalp vein	Not dilated	Dilated	
B3. VITALS SIGNS			
Temp (0 C).....	Pulse (beats/min).....	BP (mmHg).....	Respiratory Rate (cycles/min).....

B4. NEUROLOGICAL EXAM

Motor function			
•Reflexes	Decrease ()	Normal ()	Increase ()
•Muscle strength	Decrease ()	Normal ()	Increase ()
•Muscle tone	Decrease ()	Normal ()	Increase ()
Sensory status			
•Sense of touch	Decrease ()	Normal ()	Increase ()
Eye			
•Vision	Normal ()	Blur ()	Abnormal ()
•Gaze	Up	Down	Lateral
• eye movement	Normal ()	Abnormal ()	
Ear			
•Hearing	Decrease ()	Normal ()	Increase ()
Movement status			
•Coordination	Normal ()	Abnormal ()	
•Balance	Normal ()	Abnormal ()	
Psychiatric condition			
• Mental status	Normal ()	Abnormal ()	

• Mood	decrease ()	Normal ()	Increase
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C. INVESTIGATIONS

CT Scan		
	Yes	No
•Ventriculomegaly		
•Trans ependymal oedema		
•Thinned and elevation of the corpus callosum		
•Fenestration of the septum pellucidum		
•Depression of the fornix		
•Inferior displacement of the floor of third ventricle		
•Ballooning of the recesses		
•Sella turcica erosion		
•Cortical thickness		
•Calcifications/Mass		
•Holoprosencephaly		
•Dandy walker		
•Arnold Chiari Malformation		
•Agenesis corpus callosum		
•blood in ventricles		
•Others (Specify)		

D. DIAGNOSIS

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E. TREATMENT

Observation	Yes	No	Specify
Medical	Yes	No	Type
Surgical	Yes	No	
Cerebrospinal fluid diversion	Yes	No	
	Yes	No	Complications
•Ventriculo-peritoneal (V-P) shunt			
•Ventriculo-subgaleal shunt			
•External Ventricular Drain (EVD)			
Endoscopy ;			

• Endovascular Third Ventriculostomy (ETV)			
•Endoscopic pellucidostomy			
Relief of obstruction			
	Yes	No	Procedure & complication