## The 10 Major Subjects in Hydrocephalus Research Fields

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| Subject I. | Definition and Terminology of Hydrocephalus |
| Subject II. | Classification of Hydrocephalus |
| Subject III. | Pathophysiology 1. Cerebrospinal Fluid (CSF) Physiology |
| | Pathophysiology 2. Intracranial Pressure (ICP) Physiology |
| | Pathophysiology 3. Miscellaneous |
| Subject IV. | Hydrocephalus Chronology |
| Subject V. | Specific Forms of Hydrocephalus |
| 1. Pathogenic Concepts | 1) Congenital Hydrocephalus |
| | 2) Acquired Hydrocephalus |
| | 3) Idiopathic |
| 2. Pathophysiological Concepts | 1) Intracranial Pressure (ICP) |
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| | 3) Miscellaneous |
| 3. Chronological Concepts | 1) Phase |
| | 2) Progression |
| 4. Miscellaneous | |

### Subject VI. Associated Congenital Anomalies/ Syndrome and Underlying Conditions

### Subject VII. Diagnostic Procedures for Hydrocephalus

### Subject VIII. Treatment Modalities in Hydrocephalus

### Subject IX. Experimental Hydrocephalus and Invention

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2. Diagnostic and Therapeutic Methodology and Invention
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### Subject X. Ethics & Moral/ Society/ Education in Hydrocephalus Medicine and Science

1. Medico-ethics/ Medico-social/ Medico-legal/ Medico-economical Issue
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3. Miscellaneous
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As a medical condition requiring considered research and diverse approaches to management, Hydrocephalus has challenged clinicians since ancient times. The emphasis has remained at defining aetiology, in the understandable belief that appropriate management follows an accurate recognition of the underlying pathophysiology. This led to the introduction of shunting procedures in the early 1960s, followed by an ever increasing (and ever more costly) array of shunting devices aimed at correcting every feasible variety of the condition. Hence, we now have a multitude of variable pressure shunts, that can be adjusted and their flow rates altered, all aimed at mimicking nature and countering the perceived pathophysiology.

This has been followed by approaches to “enter, visualise and mechanically correct” the pathology using endoscopic techniques. Debates abound as to the merits and demerits of the options available to the clinician for treating the child, adolescent and adult afflicted with this condition which, without treatment is generally a death sentence, and for those who survive following a spontaneous “arrest” of their hydrocephalus, mean a life of severe mental and physical handicap, adjoing their family and carers in a life of solitude.

With rates of incidence quoted between 0.9 to 2 per 1000 live births, a live birth rate in the developed world of less than 1 per cent per annum, and with facilities for prenatal diagnosis available to the majority of its population, combined with the afore-mentioned array of devices and ability “to enter, look and correct” the problem, the dilemma of treating this condition would seem to have been resolved. And indeed for a minority of the planets children, this may indeed be true.

Alas, but is it indeed true for the majority?

Africa, in particular its Sub-Saharan region, represents the show case for the majority of the planets children who live in the countries of the emerging world across the globe, in South America, Eastern Europe, the parts of the Far East, amongst many other similar regions. With rates of Hydrocephalus of (and possibly above) 2 per 1000 live births, population growth rates nearly 4 fold that of developed nations, inadequate antenatal diagnosis, high rates of perinatal infections, the numbers of children afflicted becomes mind boggling. In the East, Central, and Southern African region of just over 300 million, an estimated 15,000 children are born annually with hydrocephalus, with an equivalent number in the Western Sub-Saharan region. Of these, even in the most advanced Sub-Saharan settings, only a quarter of the total will receive treatment in time. Upto a quarter of these treated cases, will fall victim to a complication of shunting procedures.

It is this realization that begins to awaken the moral conscience of those amongst us who treat this condition. There has got to be a way to ensure that the array of treatment options, of every modality from shunting, to endoscopic approaches, and beyond to research, becomes focused in the regions of the globe where the condition is maximal in its ferocity.

Yet all is not lost! Indeed there is hope!!

The emergence of organisations and groups dedicated to Hydrocephalus, and the beginning of a scientific journal dedicated to a condition afflicting this particular section of humanity lends comfort that a collective will exists to overcome. as best as we can, this hitherto unacknowledged epidemic. The Journal of Hydrocephalus is welcome evidence of this collective will, spanning all corners of this globe, providing hope that we can, and shall join, in the collective effort to reduce the burden created by this treatable condition of hydrocephalus

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Continental Editorial Advisory Board [Africa]
Journal of Hydrocephalus
Neural Tube Defects in the Developing World: Achieving Closure
A hope for the use of Folic Acid Fortification in the reduction of the high incidence Spina Bifida and associated Hydrocephalus worldwide

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Abstract

Neural Tube Defects (NTDs) are amongst the most serious birth malformations worldwide, with an estimated 300,000 births occurring annually 19. The magnitude of these defects in developing regions such as Sub Saharan Africa (SSA) is unknown as detailed prospective analyses of the incidences of these defects in most developing countries have not yet been conducted 1. However, the results from small hospital based studies, carried out within some African countries have recorded incidences as high as 7/1000 births 1. This present data on NTDs in SSA may only represent the ‘tip of the iceberg’ as it has been suggested that in many African countries, the majority of malformed children born outside hospitals are neglected or concealed and often not reported 17.

Moreover the socio-economic consequences of such anomalies are devastating especially in developing countries where the risk factors for such conditions are elevated and resources for health care treatment and management are limited 2. The most ethical treatment of NTDs would be prevention. NTDs have been associated with folate deficiency at the time of conception, and many countries have recommended the periconceptual supplementation for women planning their pregnancies. However these recommendations have not been effective in reducing the overall occurrence of these defects. Thus some developed countries, such as the USA and Canada among others, have focused on an alternative solution: the fortification of food with folic acid. This has proved to be successful in the countries in which it has been implemented. However, with the exception of South Africa, the use of folic acid in the prevention of NTDs is still calling out for adoption in SSA.

NTDs are not high on the agenda in developing countries mainly because policy makers in these countries are not aware of the problems associated with these preventable defects and the affordable and cost-effective solutions available to reduce their occurrence. This paper has used the Kingdon’s three stream policy model to try examine how these birth defects can be brought onto the agenda in developing countries, particularly those within SSA.

Key Words: Neural Tube Defects • Hydrocephalus • Spina Bifida • Prevention • Folic Acid Fortification • Sub-Saharan Africa
I. Methodology

Information for this paper was obtained from a wide variety of sources, including books, peer-reviewed journals, news articles and websites. Literature searches were conducted through library searches, library catalogues, and electronic databases including the Web of Science, PubMed, Google Scholar, and Jstor. The preferred source of information was peer-reviewed journals. However the availability of literature on the extent of the problem of NTDs in SSA was limited, so secondary searches were made on global statistics and studies conducted in the developed world.

II. Introduction

NTDs are among the most common and serious malformations detected at birth. These defects are potentially life-threatening conditions and often result in severe disability. They occur in the first twenty eight days after conception due to a failure of the fetal neural tube to close and include spina bifida, anencephaly and encephaloceles.

The most common type of NTD is spina bifida which results from the failure of the spinal cord, vertebral column and surrounding skin to fuse. The term 'spina bifida' covers a variety of defects whose severity ranges from mild to severe. They include spina bifida occulta, meningocele, myelomeningocele (spina bifida cystica-SBC) and myeloschisis (Figure 1A). The World Health Organisation (WHO) estimates that spina bifida causes 0.4% of the burden of disease among children aged 0-4 years and 0.02% among children aged 5-14 years.

Approximately 80% - 90% of children with spina bifida develop hydrocephalus (Figure 1C). Hydrocephalus without appropriate treatment can lead to irreversible brain damage, eyesight problems and mental retardation.

A complex interaction between diet, genetics and environmental factors have been implicated in the aetiology of NTDs. However, a substantial amount of evidence suggests that the failure of the neural tube to close is associated with a deficiency in folate. In order to reduce the risk for NTDs, it is recommended that women capable of becoming pregnant consume 400 µg of folic acid daily from vitamin supplements, fortified foods, or both, in addition to their diet. The link between the incidence of NTDs and nutrition creates a great tool for primary prevention and many industrialised countries have adopted policies aimed at reducing the incidence and burden of these birth defects through the use of folic acid. This is not the case with majority of the countries in the developing world mainly because critical gaps in understanding about the prevention, treatment and management of children affected with NTDs exist in these countries. In order to achieve a comprehensive solution to the problem of NTDs in countries, such as those within SSA, the issue needs to be brought onto the government health policy agenda of these countries. Kingdon (1984) proposed that for an issue to be brought onto the health policy agenda of governments, a ‘window of opportunity’ needs to emerge whereby three streams of processes - namely a problem stream, a solution stream and a politics stream – must come together.

III. The Problem Stream

The magnitude of NTDs in developing countries, such as those within SSA, is unknown as detailed prospective analyses of the incidences of these defects have not yet been conducted. However, the results from small hospital based studies, carried out within some African countries have recorded incidences as high as 7/1000 births. The current statistics on the epidemiology, prevalence and incidence of NTDs in SSA represents a
lack of definitive data. A vast majority of the information has been derived from small hospital-based studies in developing countries or extrapolated from the statistics present in developed countries. Thus present data on NTDs in SSA is subject to underestimation. This present data may only represent the ‘tip of the iceberg’ as it has been suggested that in many African countries, the majority of malformed children born outside hospitals are neglected or concealed and often not reported. Indeed, both the magnitude and burden of these defects in SSA, are believed to be severe, as the risk factors for such conditions in these countries are elevated and resources for health care are limited.

Current evidence suggests that the failure of the neural tube to close is strongly associated with a deficiency in folate in the peri-conceptual period. The lack of access to nutritious foods, high rates of poverty, harmful dietary practices and high incidences of infectious diseases are some of the factors that lead to micronutrient malnutrition. These factors are found more commonly in underprivileged populations in SSA than in most other parts of the world. Furthermore, it is estimated that two in three African women do not have access to essential services, such as family planning clinics and health centres where they may be offered dietary advice and provided with nutrient supplements. Thus, folate deficiency among women of child-bearing age and the subsequent risk of an NTD affected pregnancy is likely to be high among the majority of rural and urban poor African populations.

Indeed, NTDs pose many challenges when considering the early treatment and management of these defects in SSA. Early surgical treatment, often within the first 24 hours, is vital to increase the chances of survival and reduce the risk of severe disability amongst affected newborns. This is often not possible in SSA where there is a high likelihood of late presentation, as majority of the families of low socio-economic status go through great difficulties to bring their children in for treatment. Thus, folate deficiency among women of child-bearing age and the subsequent risk of an NTD affected pregnancy is likely to be high among the majority of rural and urban poor African populations.

As demonstrated, the overall socio-economic burden of these defects on impoverished African populations is large as management and rehabilitation facilities for affected individuals are lacking. There is an urgent need to focus on reducing the incidence of these defects. The reduction of these birth defects can not only assist in the reduction of child mortality, but can also address the problem of the severe life long disabilities that are a great burden in resource poor settings in SSA.

IV. The Solution Stream

The most ethical treatment of NTDs would be prevention. Indeed, the link between the incidence of NTDs and nutrition creates an ideal tool for primary prevention and many countries have adopted policies aimed at reducing the incidence and burden of NTDs through the use of folic acid. One such policy is the recommendation of peri-conceptual folic acid supplementation for all women planning their pregnancy. Folic acid supplements need to be taken at least one month before conception and during the first three months of pregnancy. However many studies have shown that these recommendations have not been effective in reducing the occurrence of these defects due to a high proportion of unplanned pregnancies, a lack of awareness of the protective effects of folic acid supplementation and poor compliance.

Some developed countries have thus focused on an alternative solution: the mandatory fortification of food with folic acid. Food fortification is defined as “the practice of deliberately increasing the content of an essential micronutrient in a food, so as to improve nutritional quality, and provide a public health benefit with minimal risk to health.” There are many advantages to fortifying food with folic acid. Fortified food is widely distributed, and thus has the potential to reach a large percentage of the population, including all women of child-bearing age before they become pregnant. In addition, fortification of a staple food does not require food compliance or a change in existing food patterns and therefore can be delivered to large populations without drastic changes in food consumption patterns and food habits. The maxim that ‘prevention is better than cure’ is confirmed once again in developed countries, such as the USA, that have demonstrated that primary prevention of NTDs through the fortification of food is both practical and cost-effective as by reducing the incidence of NTDs, the overall costs of care are also reduced.

However, despite important advances in the knowledge
regarding prevention and treatment of NTDs, it is estimated that only 10% of the estimated 300,000 preventable cases of NTDs, such as spina bifida and anencephaly, have been prevented through fortification. This is mainly attributable to the fact that currently, fewer than 40 countries, most of which are developed nations, have adopted fortification of food with folic acid as a preventative intervention.  

V. The Politics Stream

In order to achieve a comprehensive solution to the problem of NTDs in SSA, and other developing countries, the issue needs to be brought onto the government policy agenda of these countries. Arguably the most powerful actor in agenda setting is the government, as it has control over the policy and legislative processes. Only when policy makers believe that a problem is of public health importance and that a failure to act can result in more disturbing consequences, do they give it serious consideration. Within SSA, NTDs have not yet been placed high on the agenda because policy makers are largely unaware of the extent of the problems associated with these grave albeit preventable defects, their socio-economic effects and the availability of affordable, cost-effective solutions to reduce their occurrence. This predominantly results from the lack of knowledge and research in this field.

VI. Conclusion

The full potential of folic acid in the prevention of NTDs has not been reached in many regions of the world, especially in developing countries, and despite the existence of many cost effective interventions, preventable NTDs still occur. There is agreement that policy and practice should be based on the best available evidence applicable in a particular setting. However, in low-income countries, there is a lack of knowledge about which interventions work which in turn hinders effective development of policy. Only when research and knowledge gaps on the issues around NTDs in developing regions, such as SSA, are addressed can there be effective translation of knowledge into action. Thus, national investment into micronutrient research, research into the epidemiology of these defects in developing countries, culture and country specific dietary consumption data together with detailed research into the planning of fortification strategies needs to be undertaken.

It is imperative to bring to the attention of senior policy makers in developing countries the preventative strategies, such as the mandatory fortification of food with folic acid, which can reduce the burden of these defects on their populations. In addition, media attention, influential individuals, lobbying, health statistics and the dissemination of research results, all have a role to play in the opening of this ‘window of opportunity’ and gaining the attention of policy makers. Consequently effective collaborations between researchers, policy makers and specialists in the field need to be established and a concerted effort to reduce these defects needs to be made.

Therein lies the hope of a solution to the huge burden of NTDs, including anencephalyencephaloceles, spina bifida and associated hydrocephalus in the developing world, particularly those within SSA.

References

Using “catheter à fentes” in child’s hydrocephalus:  
A prospective study about 197 cases

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Abstract

[Subject] An experimental study of some differential pressure valves rate, led the authors to build a ventriculoperitoneal shunt: the” catheter à fentes”. The authors assessed the results of the hydrocephalus treatment by this shunt.

[Methods] A prospective study of childhood hydrocephalus was conducted in Niamey national hospital – Niger from January 2006 to July 2009. Surgical procedure was the classic right frontal ventriculoperitoneal shunt. The model of shunt used was “catheter à fentes”. The length of the catheter was 100 cm corresponding to the medium pressure. The fellow up was 3 months.

[Results] The study concern 197 infants aged between 1 to 18 months. 57% were female. The hydrocephalus was post infectious for 48.22% of patients. The mean age was 8.53 months. The mean head circumference was 47.533 cm. According to Lezine score, 76.54% of patients had important psychomotor deficiency. The mean intracranial pressure before operation was 16.277 cm H$_2$O. The psychomotor score was significantly improved for all patients after operation. The mean head circumference reduction was 3.877 cm. Hydrodynamics complications occurred in 7.61% of patients at 3 months after operation.

[Conclusion] The clinical evolution of hydrocephalus infants operated with the catheter à fentes is satisfactory. The cost of the “catheter à fentes being 2 dollars US offer the opportunity to developing countries for the treatment of hydrocephalus.

Key Words: Hydrocephalus • meningitis • congenital • shunts • catheter à fentes • Niamey • Niger
I. Introduction

From the hydrocephalus description in Ebers papyrus by the Egyptians (1900 BC) to this day through Dandy (1886 - 1946), the treatment of hydrocephalus underwent several adaptations. History of the treatment of hydrocephalus is similar to the development of civilization history.

After the cranial punctures, the treatment became medical (thyroid extract, diuretics, irradiation), then, medico surgical\(^1\).

The use of a ventriculoperitoneal catheter (simple shunt) for the treatment of hydrocephalus goes back to 1914 by Heile\(^1\). It was a simple catheter without valve or slits; in 1927 lumborenal shunts were used. Over drainage problems leaded hydrodynamic engineers Donald Matson and Franck In graham in 1946\(^2\) to create a unidirectional valve. Lasten, the differential pressure valve was born and the use of the silicone catheter was popularised.

Other types of flow controlled shunt, different from the differential pressure valves, were developed to try to remedy the over drainage problem\(^3\). The introduction of the antisiphon system\(^4, 5, 6\) in the making of different valves did not totally solve the problem of over drainage. In fact, in a lying posture the hydrostatic pressure of the cerebrospinal fluid returns homogeneously in the cerebrospinal axis for a 15 cm H\(_2\)O values. In the erect posture there is a pressure gradient toward the medullar cone; at the lumbar area the pressure is approximately 50 cm H\(_2\)O, which gradually decreases to zero at the lateral ventricles. The valves don’t adapt perfectly to these physiological variations; moreover, when they are shut the absence of flow favours the adherence of proteins on the catheter’s inside and bacterial colonization. Julio Soleto and col.\(^7\) invented an open shunt system with a constant flow proportional to the daily CSF output. The resistance to the discharge is exercised by the peritoneal catheter internal diameter, which is reduced to 0.51 mm instead of the standard 1.1 to 1.2 mm. This model while being efficient makes the shunts affordable to developing countries.

The catheter with slits that we used in this study for the treatment of the hydrocephalus is an open system with a constant flow and in conformity with Poiseuille low\(^8, 9\). We act on the length of a catheter fitted with slits at its distal extremity to have an effect on the fluid flow. The initial experimental study aimed to compare the rate obtained with a simple silastic tube with slits at its blind distal extremity with the rates of SOPHISA*, CODMAN*, and BEVERLY* valves. The flow obtained with this 1.1 mm internal diameter catheter varies according to the size of the slits, the type of the slits, the lay out of the slits, and the length of the catheter. (Figures 1, 2, 3). This experimental study helped to demonstrate that these various valves, while having the same opening pressure, do not have the same rates. Furthermore, it helped to define catheter models offering a rate close to these valves, especially the SOPHISA one. Thus, by extending by 35 cm from the \(\Delta P\) (differential pressure between the proximal and the distal reservoir), a catheter fitted with the two 3 mm long slits gives a rate close to the

FIG. 1 experimental schema for “catheter à fentes” flows measurements.

FIG. 2 Distal extremity of the catheter with slits.
low pressure SOPHYSA* valve (Figure 4); an extension of 45 cm from the ΔP of this same type of catheter gives a rate close to the medium pressure SOPHYSA* valve (Figure 5); and an extension of the same type of catheter from the ΔP of 65 cm gives a rate close to the high pressure SOPHYSA* valve (Figure 6).

The “catheter à fentes” with extension of 65 cm from ΔP (total length 100 cm) were used for 197 hydrocephalus infants.

II. Materials and methods

It was a prospective study from January 2006 to July 2009 conducted in the neurosurgery department of the national hospital of Niamey (Niger); were registered history of pregnancy delivery and neonatal periods for all infants. Progression of the head circumference size from the born to operation was registered for all infants. Gesell and Lezine score were used for psychomotor evaluation.

Courbes comparatives: 2 fentes de 3 mm (basse pression)

![Graph showing comparative curves of low pressure valves with catheter à fentes after extension 35 cm and 65 cm.]

FIG. 4 Comparative curves of low pressure valves with catheter à fentes after extension 35 cm and 65 cm.

Courbes comparatives: 2 fentes de 3 mm (moyenne pression)

![Graph showing comparative curves of medium pressure valves pressure with catheter à fentes after extension of 45 cm and 75 cm. The flow of the catheter with slits (45mm extension) is close to that of the valves.]

FIG. 5 Comparative curves of medium pressure valves pressure with catheter à fentes after extension of 45 cm and 75 cm. The flow of the catheter with slits (45mm extension) is close to that of the valves.
Hydrocephalus children with QD lower than 60 were not included. Diagnosis of the hydrocephalus and evaluation of the shunt efficiency were based on Evans index measure on the brain CT scan before the operation and one and three months later. The length of the “catheter à fentes” was 100 cm for all patients corresponding to the SOPHYSA* medium flow rate. The hydrodynamics’ complications were evaluated by the appreciation of the clinical and Evans index evolution. The management of hydrodynamics complications was lengthening of 10 cm the abdominal catheter for over drainage complication or shortening of 10 cm the catheter for under drainage complication. Post operative evaluations included cranial size and psychomotor evolutions.

**Shunt placement procedure.**

The procedure is a right frontal or occipital ventriculoperitoneal shunting. The measure of the ventricular pressure is done at the time of the ventricular puncture. The two 3 mm slits on the 2 cm distal blinded extremity were made in surgical period. The ventricle catheter is linked to the peritoneal catheter through a connector and fixed to the temporal aponevrosis to avoid migrations. The breast feeding resumes immediately the day after the operation in the arms of the mothers who will continue carrying their children on their backs as usual after the hospital to facilitate the siphoning effect.

### III. Results

These infants were mostly from rural areas far away from the national hospital of Niamey and their families’ living conditions are precarious. The study group was made up of 99 females (50.25%) and 98 males (49.74%), whose mean age was 8.53 months ranging from 1 to 18 months. For 59.55% of patients the duration symptoms was superior to 3 months. The hydrocephalus was post infectious for 48.22% (95 cases), associated with spina bifida and brain anomalies in 47.71% of cases (94 cases), related to hemorrhage in 3.55%, (3 cases) associated with tumor in 1.01% of cases (2 cases). The cranial circumference was superior to 2DS in all cases. It was ranged between 39 to 55 cm and the mean was 47.533 cm. According to Lezine score, 76.54% of patients had important psychomotor deficiency (60 ≤ QD ≤ 80). Hydrocephalus was tetra ventricular for 85.78% (169 cases) of cases and tri ventricular for 14.21% of patients (28 cases). The intracranial pressure ranged 10 to 25 cm of water for 186 patients (94.41%). The mean intracranial pressure before operation was 16.277 cm H$_2$O. The Geisel Lezine psychomotor score was significantly improved for all patients after operation ($p ≤ 0.05$). (Illustrations 1, 2). 118 patients with marked ventriculomegaly (Evans index ≥ 0.4) and 79 patients with moderate ventriculomegaly (Evans index ranged from 0.35 to 0.40) were reported. The means Evans index before surgery was 0.412. The study reported hydrodynamics complications before 1 month post surgery for 13 infants and 2 cases of subdural hematoma at 2 and 3 months after surgery. According to clinical signs and CT scan Evans index control in the 3 months follow up, over drainage (reduction of the Evans index ≥ 0.1) was reported for 8 patients, low drainage (reduction of the Evan Index ≤ 0.05) for 5 patients and obstruction for 2 patients. For the rest of patients systemic CT scan controls were performed between 1 to 3 months post surgery with measurements of Evans index. The mean reduction of Evans index was 0.0853 ranged from 0.06 to 0.1 (Figure 7). The mean cranial size reduction was 3.016 cm. The management
of hydrodynamics complications (7.61%) (15 cases) was uneventful. The study showed that the ventricular pressure ranged from 10 cm H\(_2\)O to 25 cm H\(_2\)O for 186 patients (94.41%); from 25 to 33 cm H\(_2\)O for 11 patients. The mean ventricular pressure was 18.824 cm H\(_2\)O.

The Geisel-Lezine psychomotor score \(^17\) was evaluated at 3 months post surgery for all patients and was significantly improved for all patients (\(p \leq 0.05\)). Patients having a QD superior than 60 gain an average of 10 points in 3 months while the infants with a pre operative QD higher than 80 gains 10 points in one month. The main complications (over drainage, and infections) were encountered during the first operative month.

Hydrodynamics complications were encountered during the first month (7.61%) (15 cases). Thus, we report 6 cases of over drainage, 9 cases of lower drainage obstruction. In case of hydrodynamics complications, starting again the abdominal incision with the extension (over drainage) or the shortening (lower drainage) the catheter solves the rate problem in all cases. The average hospitalisation duration was 12 days.

The infection rate was 6.59% (13 cases) and the death rate was 1.01% for infections.

IV. Discussion

In treating hydrocephalus, two major valve groups are used: there are the differential pressure valves or standard valves and the flow pressure valves \(^1, 2, 3, 4, 5, 6\). They differ on over drainage long-term risk, which would more important for the standard valves \(^13, 14\). A recent study by Soleto and col. \(^7\) reports satisfactory results with the use of open ventricular shunt with the adult. It’s an open catheter which the distal extremity has 0.51 mm internal diameter. The catheter with slits functions like an open catheter with slits at its distal extremity.

In this clinical study we analysed the result of the use of the catheter with slits in infant’s hydrocephalus. The analysis of these results concerned intracranial hypertension signs, cranial perimeter reduction, psychomotor improvement and various complications.

In the series we reported 7.61% of hydrodynamics complications (Obstruction, lower drainage and over drainage), after 3 months. Saint Rose, Hoffman and Drake \(^13, 14\) reported in a study concerning 1700 patients treated with conventional valves, 30% mechanical complications before one year. Oregon and Bierbaeur \(^14\) quoted in the same study found similar results after 1 year. These complications partly due to over drainage during the passage to vertical position.

Intracranial hypertension signs disappeared in all cases by the first postoperative week. This confirms most authors’ results \(^1, 7, 9, 14, 15, 16\). Infection was present in 6.59% of cases with 1.01% of deaths from ventriculitis in this study. Beverly \(^17\) reports 15% of infection. According to the studies the infection varies between 2-3% and 29%. This series concerns 1 to 18 months old infants. In
a randomised multicentric study concerning 345 patients James Drake and col. reports that the mechanical and infections complications apart from the valves type are more frequent before 6 months. This study concluded also that the infection risk is age dependent.

Our study is characterized by the duration of the symptoms’ evolution. For 59.55% of patients the duration symptoms was superior to 3 months in this study. This had repercussions on the psychomotor status, the preoperative cranial perimeter and the type of hydrocephalus. Thus, the postoperative psychomotor improvement is significant only for patients having a pre-operative QD higher than 60. This confirms Derek. A. Bruce’s conclusions on the postoperative psychomotor recovery in paediatric neurosurgery. This study recommended precocious (before 3 month evolution) surgery for psychomotor status improvement.

The mean head circumference reduction was 3.877 cm 3 months after operation. The reduction of the cranial perimeter is proportional to the efficiency of the drainage. This is reported by several authors. In this study the cranial circumference remained in post operative above 2SD in almost the whole cases due to the pre operative macrocranie.

This study showed the profile of hydrocephalus in Niamey: The mean age was 7.53 months, the mean cranial size was 47.533, the mean QD was 71.784, and the mean preoperative Evans index was 0.412. These results means, long evolution of hydrocephalus with adult cranial size and intra ventricular pressure slightly elevated. For management of these types of hydrocephalus medium pressure or high open pressure valves must be used (100 cm length of “catheter à fentes”). These types of hydrocephalus can be considered as child’s chronic hydrocephalus. The 3 months post operative results showed: mean reduction of Evans index was 0.0853, mean cranial size reduction was 3.016 cm, and mean QD gain was 13.321 points. The use of catheter à fentes is efficient in the treatment of hydrocephalus.

V. Conclusion

The catheter with slits is efficient is efficient in the treatment of infants hydrocephalus. Its modest cost makes it affordable to developing countries hence allowing a precocious treatment of the hydrocephalus.

Acknowledgements

Pr Maitrot, Pr Christian Sainte Rose, Pr Pierre Kehrli, Dr Patrick Boyer.

References

Endoscopic third ventriculostomy in infants up to the age of six months: is it effective?

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Abstract

Main problem: Endoscopic third ventriculostomy (ETV) is considered nowadays as the method of choice in the treatment of obstructive hydrocephalus. However, its effectiveness in babies remains controversial. Our aim is to assess the effectiveness of ETV in infants up to the age of 6 months and try to determine the predictive factors of failure.

Methods: We studied retrospectively a series of 18 infants with a mean age of 87 days treated by ETV for obstructive hydrocephalus. The main etiology was congenital aqueductal stenosis (78%). The follow up ranged from 5 to 60 months. ETV was considered successful when no shunt was needed.

Results: ETV was successful in 7 patients (38%). The highest rate of success with respect to the etiology was noticed in congenital aqueductal stenosis (42%). ETV failed in all cases of post-infectious aqueductal stenosis and of Chiari malformation. Infection is the most predictive factor of failure.

Conclusion: ETV is effective in more than one third of cases of obstructive hydrocephalus in infants. Its outcome depends on the etiology of hydrocephalus and not on the age of the infant.

Key Words: Obstructive hydrocephalus, Endoscopic third ventriculostomy, Infants, Outcome

I. Introduction

Endoscopic third ventriculostomy (ETV) is considered at the present time as the method of choice in the treatment of obstructive hydrocephalus [1]. Its use is increasing and it reveals a favorable outcome for most of adult patients with few complications compared to ventriculo-peritoneal shunt [2]. However, its effectiveness in babies remains controversial. Through this study we will attempt to assess the effectiveness of ETV for hydrocephalus in infants up to the age of 6 months and try to determine predictive factors of failure.

II. Materials and methods

Between June 1998 and December 2009, 18 infants up to the age of 6 months bearing obstructive hydrocephalus were treated endoscopically in our department. This study included 11 boys and 7 girls. The age of patients was between 17 and 180 days with an average of 87 days.
Computed tomography scans (CT scan) were carried out in all infants and magnetic resonance imaging (MRI) in only 7 cases. None of our patients have had an internal shunt inserted previously. All ETV procedures were performed by the same technique using rigid endoscope (KARL STORZ) and ventriculostomy was obtained using coagulation electrode.

The etiology of hydrocephalus was congenital aqueductal stenosis in 14 cases, post-infectious in 2 cases, Chiari malformation in 1 case and a vascular malformation (aneurysmal torcular dilation) in 1 case.

The follow up ranged from 5 to 60 months, during which clinical assessment was based on head circumference and frontal bulging evolution and neuro-imaging studies considered the ventricular size as the main criterion of survey. ETV was considered successful when no shunt was needed in patients definitively.

III. Results

The overall success rate of ETV in our study was 38% (7 patients). Considering the rate of success with respect to the etiology of hydrocephalus, congenital aqueductal stenosis was the most predictive state of success counting up to 42% (6/14). ETV was also successful in the case of huge aneurysmal torcular dilation compressing the fourth ventricle. However, it failed in the 2 cases of post-infectious aqueductal stenosis and in the case of Chiari malformation. The results are summarized in Table 1.

Coming to the rate of success according to the age of infants at the time of the procedure, we noticed one successful case out of three up to the age of one month, 3 cases out of 7 at 2 months, a success in the only two cases of 3 months-aged infants and one case out of two at the age of 4 months. On the other hand, no success was noticed in the ages of 5 and 6 months. Age distribution at the time of ETV and the success of the procedure is presented in Figure 1 and correlation between etiology and rate of succeed is demonstrated in Figure 2.

Post-operative complications were sub-dural hygroma in 3 cases, aseptic hyperthermia in 2 cases, meningitis in 1 case which was successfully treated by antibiotics and cerebro-spinal fluid leak in 1 case treated conservatively. There was no mortality after ETV procedures.

IV. Discussion

In the literature, ETV success rate is lower in the group of infants up to the age of 6 months than in adult patients and by comparing conclusions of several studies, we note an important variability of ETV success rate. According to recent studies, ETV effectiveness in children of this age shows a success rate ranging from 32 to 67%. [3, 4, 5, 6]. Our results much very well this data.

Several factors are influencing the success or failure of ETV. The most important of them are age and etiology of hydrocephalus, but previous ventriculo-peritoneal shunt, head circumference, anatomical factors and low gradient in cerebrospinal fluid resorption in newborns or immature CSF absorption capacity in young infants are also described as predictive factors. [7]

Many authors consider age of young patients as the most important factor in ETV failure [3, 8] and according to Koch and Wagner in their study and meta-analysis of other publications; the worst results of ETV are obtained in infants up to the age of 3 months [9]. However this idea is not supported by all authors [10, 11]. Cinalli et al., for instance, showed that there is no difference in success rate of ETV in children under 6 months and older children with aqueductal stenosis [10]. Moreover, our

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Success</th>
<th>Failure</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopatic Aq S</td>
<td>6 (42%)</td>
<td>8 (58%)</td>
<td>14</td>
</tr>
<tr>
<td>Post infec. Aq S</td>
<td>0</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Vascular malf.</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Chiari</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Total (%)</td>
<td>7 (38%)</td>
<td>11 (62%)</td>
<td>18</td>
</tr>
</tbody>
</table>

FIG. 1 Bar graph showing the correlation between the age of patients at the time of ETV and the success rate.

FIG. 2 Bar graph showing the correlation between etiology and the success rate.
results show that there is no statistically significant correlation between age and the rate of ETV success in young infants, as we noticed, on the contrary, a better response in very low aged infants than in elder ones. This suggests that the success of ETV depends on the etiology of hydrocephalus rather than on the age. In deed, the very young infants in our series, who responded very well to ETV, harbored mostly a congenital aqueductal stenosis whereas all the elder infants, aged between 5 and 6 months, presented with a post-infectious AS or a chiari malformation. According to literature, the etiology of hydrocephalus represents an important predictive factor of ETV success. All studies are agreed that congenital or idiopathic aqueductal stenosis is the best indication for ETV. For Faggin et al the success rate of ETV in infant up to the age of 6 months was 88% [3] and for Buxton et al. it was about 55% [6]. The reason of failure of ETV in infantile period according to these authors was that certain of idiopathic aqueductal stenoses may not be pure AS. Patients may have had a slight intra-ventricular hemorrhage which might cause an aqueductal stenosis but also arachnoiditis and impairment of CSF resorption. MRI and cine-flow MRI may be helpful, by studying the anatomy of ventricles and the kinetic of CSF, to guide the treatment choice and evaluate correctly the results.

Post-hemorrhagic and post-infectious aqueductal stenoses have generally poor results with ETV even if in few studies ETVs were successful [12, 13]. For the other etiologies (Dandy Walker malformation, Chiari malformation, and Spinal dysraphism) results are controversial. All similar cases in our series were followed by failure.

In case of failure of ETV procedure, Cine flow MRI might be useful to determine the exact cause and precise the site of obstruction. Balthasar et al. recommend to repeat ETV [12], but many authors think that repeating ETV might be dangerous and that it would be better to pass directly to a ventriculo-peritoneal shunt (VP shunt) [11, 14, 15]. In our current practice we always adopted the latter attitude in such cases.

V. Conclusion

Despite the small number of patients in our series and according to our results, ETV should be recommended as the first therapeutic option in obstructive hydrocephalus even in very young infants, because it offers the opportunity to avoid VP shunt complications. The rate of ETV success is directly related to the etiology of hydrocephalus and not to the age of infants itself. A better understanding of the physiopathology of hydrocephalus in new born and in infants may lead to a more rational selection of good candidates for this procedure.

References
Complications of Ventriculo-Peritoneal Shunt Insertion seen at the Kenyatta National Hospital, Kenya, East Africa

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Abstract

A prospective study involving 76 patients operated upon for non-neoplastic hydrocephalus by a ventriculo-peritoneal (V-P) shunt procedure at the Kenyatta National Hospital, Nairobi, Kenya, over a period of 6 months, between 1st September 2001 and 28th February 2002 is reported.

The age range was one month to 54 years, with a male to female ratio of 1.5:1

The commonest cause of Hydrocephalus was congenital (72.4%).

23 patients (30.3%) suffered a shunt related complication.

Shunt infection was the commonest complication - 15 patients (19.7%), followed by shunt blockage in 7 patients (9.2%); shunt migration and prolapse through the anal orifice occurred in 1 patient (1.3%).

The average time interval between diagnosis of hydrocephalus and surgery for insertion of V-P shunt was less than 2 months in 36.8% of patients, 2-4 months in 32.9% of patients, 4-6 months in 15.8% patients, 6-12 months in 6.6% patients and greater than 12 months in 7.9% of patients

Variables assessed for co-relation between surgery and complications included age, duration of symptoms, length of surgical procedure, use of prophylactic antibiotics and type of shunt. Of these variables only one factor, that of shunt type, revealed a co-relation with a higher complication rate (p < 0.05). The reason for this was not clear.

The mortality rate in patients who developed a shunt complication was 30.4% with all seven deaths occurring in the group of 15 patients who developed a shunt infection. This yielded an overall mortality rate of 9.2%. When mortality was assessed for patients developing a shunt infection, the mortality rate was 46.6%

Key Words: Ventriculo-peritoneal shunt, Complications, Hydrocephalus, Africa
I. Introduction

Modern shunting procedures were introduced in the early 1950’s, with the ventriculo-cardiac shunt described by Nulsen and Spitz (68) being amongst the earliest procedure for diversion of CSF from the ventricular cavity, for relief of hydrocephalus. The Spitz-Holter valve, invented by John Holter in 1956 to save his son (68) led to the procedure gaining popularity.

Prior to 1954, when silastic material was first used for ventriculo-atrial shunts (75), other materials including rubber, portex vinyl, and polyethylene tubing were used (44, 97). These materials led to foreign body reactions and a high rate of complications from obstruction (23). Ventriculo-peritoneal shunts gained favour over V-A shunts due to relative ease of insertion, less severe complications, and greater ease during shunt revisions (7, 47, 56).

Literature is abound with various complications of V-P shunting procedures and include:

1. Shunt infection
   Infection rates have been reported across a wide range, from 2.4% to 40% with most series reporting rates between 10%- 20% (57, 66, 88, 96). Most infections occur at the time of surgery, from the patients own flora (9, 31), with the most common organism being coagulase negative staphylococci (9, 31, 77, 81, 88), which have the capability to colonise shunts due to an extracellular mucoid substance, forming micro-colonies on the luminal surface of the shunt catheters (9, 10). A second group, not having the capability to adhere and colonise shunts includes staph epidermidis and staphylococcus aureus (20), whilst a third group includes a wide variety of organisms (17).

2. Mechanical blockage/malfunction
   This more commonly occurs at the distal (peritoneal) end rather than the ventricular end (62). Kast et al (50), however found 50% proximal malfunction, 14% distal and 10% attributable to the valve itself. Disconnection has been reported as a cause in 15% of shunt malfunctions, with the occipitally placed shunts at higher risk of dislocation that frontally placed shunts (3).

3. Shunt Migration
   Shunt and erosion through the bowel, presenting as prolapse through the rectum, ventriculitis or peritonitis has been reported (102). Extrusion through the mouth, following a bout of vomiting can similarly occur (Figure 1). Perforation of other organs include the vagina (63), bladder (38), gallbladder (65), the liver (65), through the umbilicus (2), and as a scrotal mass (72, 95).

4. Pneumocephalus secondary to colonic perforation has been reported (91).

5. Slit ventricles
   A clinical challenge, collapsed or “slit ventricles” present with low pressure headaches, worse on standing, and improving on lying supine usually occur in a setting of a disconnected distal shunt valve and a patent fibrous tract allowing overdrainage of CSF (84).

6. CSF ascites
   The peritoneum appears unable to absorb the CSF adequately, presenting with abdominal swelling and ascites (15).

7. Abdominal psuedocyst
   First reported by Harsh (42). Histological examination shows a thickened fibrous tissue with inflammatory cells on the inner wall, and the outer wall formed by matted bowel wall. Infection and multiple shunt revisions are implicated in its aetiology (42).

8. Miscellaneous
   Several other infrequent complications have been reported, including:
   a) Expanding septum pallucidum (14)
   b) Spontaneous bacterial peritonitis (32)
   c) Hydrothorax (28)
   d) Intrahepatic cyst (39)
   e) Epidural haematoma (74)
f) Intraperenchymal pericatheter cysts (46)
g) Spread of tuberculous meningitis into multiple disseminated abscesses (100)

II. Materials and methods

All patients with non-tumour hydrocephalus undergoing treatment via a first V-P shunt procedure at the Kenyatta National Hospital between September 2001 and February 2002 were included in the study. The patients were followed up for a further period of three months after surgery.

All patients recruited in the first two months did not receive an intravenous antibiotic at time of induction of anaesthesia. While patients in the third month received Ceftriaxone 250 mg received the antibiotic at induction. The choice of antibiotic was based on its broad spectrum cover and availability.

Variables studied further included time interval between diagnosis and surgery for insertion of the shunt, type of shunt, type of hydrocephalus, presence of spinal dysraphism and length of surgical procedure.

All shunts were dipped and cleansed in gentamycin (80 mg in 100 ml normal saline solution). A standard cleaning and draping technique was used for all the patients.

Intra-operative data was collected in the operating room. Post-operative review was carried out the following morning and then at 48 hours post-operatively. A follow up review at 2 weeks and at three months was documented.

The final outcome was based on the following criterion:

GOOD: Improved on treatment and discharged within 10 days
FAIR: Improved on treatment and discharged from 11th to 20th day
POOR: Patient requiring inpatient care as inpatient at 30 days
DEAD: Patient died while under treatment for shunt related complication during the study period, but not in the immediate post-operative period

Data was analysed using the SPSS statistical package. Statistical significance was determined using the Chi-square test and a $p$ value of $<0.05$ considered significant.

III. Results

1. Sex distribution

There were 46 males (60.5%) and 30 females (39.5%), a male to female ratio of 1.5:1.

![Sex Distribution of patients with non-tumour hydrocephalus.](image)

2. Age distribution versus development of complications

Age range was one month to 54 years.
- 35 patients (46.1%) were below the age of 6 months,
- 54 patients (71.1%) were below the age of 12 months
- 16 patients were between 13 – 60 months
- 6 patients were above 60 months

The peak age of V-P shunting in this study was 3-4 months. There was no significant relationship between the age of the patient and development of complications. However children below the age of 2 months seemed to have a higher rate of complications, although this did not reach significant levels.

<table>
<thead>
<tr>
<th>Age (months)</th>
<th>Pts with complications (%)</th>
<th>Pts without Complications (%)</th>
<th>Total Pts</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;2</td>
<td>5 (41.7)</td>
<td>7 (58.3)</td>
<td>12</td>
</tr>
<tr>
<td>3-4</td>
<td>4 (28.6)</td>
<td>10 (71.4)</td>
<td>14</td>
</tr>
<tr>
<td>5-6</td>
<td>3 (33.3)</td>
<td>6 (66.7)</td>
<td>9</td>
</tr>
<tr>
<td>7-8</td>
<td>2 (22.2)</td>
<td>7 (77.8)</td>
<td>9</td>
</tr>
<tr>
<td>9-10</td>
<td>1 (20.0)</td>
<td>4 (80.0)</td>
<td>5</td>
</tr>
<tr>
<td>11-12</td>
<td>1 (20.0)</td>
<td>4 (80.0)</td>
<td>5</td>
</tr>
<tr>
<td>13-24</td>
<td>1 (20.0)</td>
<td>4 (80.0)</td>
<td>5</td>
</tr>
<tr>
<td>25-36</td>
<td>3 (60.0)</td>
<td>2 (40.0)</td>
<td>5</td>
</tr>
<tr>
<td>37-60</td>
<td>2 (33.3)</td>
<td>4 (66.7)</td>
<td>6</td>
</tr>
<tr>
<td>&gt;60</td>
<td>1 (16.7)</td>
<td>5 (83.3)</td>
<td>6</td>
</tr>
<tr>
<td>Total</td>
<td>23 (30.3)</td>
<td>53 (69.7)</td>
<td>76</td>
</tr>
</tbody>
</table>

3. Aetiology of non-tumoural hydrocephalus

CT scans could not be obtained in 55 of the 76 patients, due to financial reasons. In the patient group where CT...
scans were available Aqueductal stenosis was diagnosed in 14 patients, and an Arnold Chiari malformation was diagnosed in one patient. On clinical grounds, however, Congenital hydrocephalus was diagnosed in a further 40 patients. Congenital hydrocephalus was diagnosed, therefore, in 72.4% of the cases seen in this study.

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Aetiology of Hydrocephalus.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type of hydrocephalus</td>
<td>Aetiology</td>
</tr>
<tr>
<td>Acquired</td>
<td>Post meningitic</td>
</tr>
<tr>
<td></td>
<td>Post traumatic</td>
</tr>
<tr>
<td>Total</td>
<td></td>
</tr>
<tr>
<td>Congenital</td>
<td>Aqueductal stenosis</td>
</tr>
<tr>
<td></td>
<td>Arnold Chiari</td>
</tr>
<tr>
<td></td>
<td>Unspecified</td>
</tr>
<tr>
<td>Total</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td></td>
</tr>
</tbody>
</table>

4. Role of time interval between onset of hydrocephalus and shunt insertion

Patients in this study, as in most developing countries, have a considerable delay between onset of symptoms and being diagnosed to have hydrocephalus. There was further delay before they finally sought treatment and undergo shunting. Table 3 outlines the interval delay between onset of symptoms and shunt insertion. The majority of patients (85.5%) had a shunt placed within a period of 6 months from onset of their symptoms. Whilst this delay results in cognitive complications, and affects their mental development, the complications evaluated by this study were not affected by the delay.

<table>
<thead>
<tr>
<th>Table 3</th>
<th>Time interval between onset of Hydrocephalus and shunt insertion.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Interval</td>
<td>Pts with complications (%)</td>
</tr>
<tr>
<td>&lt;2</td>
<td>12 (42.9)</td>
</tr>
<tr>
<td>2-4</td>
<td>5 (25.0)</td>
</tr>
<tr>
<td>4-6</td>
<td>3 (25.0)</td>
</tr>
<tr>
<td>7-12</td>
<td>1 (20.0)</td>
</tr>
<tr>
<td>&gt;12</td>
<td>2 (33.3)</td>
</tr>
<tr>
<td>Total</td>
<td>23 (30.3)</td>
</tr>
</tbody>
</table>

5. Time interval between shunt insertion and onset of complications

The majority of the patients (17 of 23 i.e 74%) developing complications presented within one month of shunt insertion. A further 21.7% presented in the subsequent 4 weeks (within 8 weeks) and 1 patient (4.3%) presenting after a period of 8 weeks.

<table>
<thead>
<tr>
<th>Table 4</th>
<th>Time interval – shunt insertion to onset of complications.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Time interval (weeks)</td>
<td>Episodes of complications</td>
</tr>
<tr>
<td>0-2</td>
<td>9</td>
</tr>
<tr>
<td>2-4</td>
<td>8</td>
</tr>
<tr>
<td>4-8</td>
<td>5</td>
</tr>
<tr>
<td>&gt;8</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>23</td>
</tr>
</tbody>
</table>

6. Complications of v-p shunt insertion

The rate of complications in this study was 30.3% during the 3 month follow up period.

Shunt infections accounted for 65.2% of the complications. Diagnosis of shunt infection was made on clinical grounds. Laboratory diagnosis was available in only a few patients, and was reflected by raised white cell counts and positive CSF culture results.

Shunt malfunction/blockage was diagnosed on clinical grounds and accounted for 30.5% of the complications.

One patient presented with the shunt protruding from the anal passage. It was noted to be draining well with the patient showing no sign of infection or of shunt blockage.

<table>
<thead>
<tr>
<th>Table 5</th>
<th>Type of complications of V-P Shunt.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type of complication</td>
<td>Number of patients</td>
</tr>
<tr>
<td>Shunt Infection</td>
<td>15</td>
</tr>
<tr>
<td>Shunt Blockage</td>
<td>7</td>
</tr>
<tr>
<td>Shunt Migration</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>23</td>
</tr>
</tbody>
</table>

7. Symptoms and signs of shunt infection

Table 6 illustrates the presenting clinical features of shunt infection in this study. Pyrexia and irritability were the most common presenting feature of shunt infection.
8. Role of prophylactic antibiotics

Twenty seven patients were randomly selected to receive a stat dose of Ceftriaxone at induction of anaesthesia. 250 mg of a generic preparation was administered intravenously. Of these 27 patients, 3 patients developed shunt infection and 1 developed a blockage related to shunt infection, a rate of 14.8%.

Forty nine patients did not receive antibiotic prophylaxis. Of these 12 developed shunt infection and 6 developed shunt blockage related to shunt infection, a rate of 36.7%.

The use of antibiotic prophylaxis appeared to reduce the risk of shunt infection and infection related blockage. However, the difference was not statistically significant ($p = 0.160$).

9. Signs of local shunt infection

Table 8 shows the episodes of clinical manifestation of infection.

10. Association between length of procedure and shunt infection

The length of surgical procedure was defined as the period between making the skin incision to the time when the last skin suture was applied. 10 out of 34 patients (29.4%), whose surgery lasted less than 30 minutes developed shunt infection, while 11 out of 39 patients (28.2%), whose surgery lasted 30-60 minutes developed shunt infection, yielding no statistical difference in the rates of infection ($p = 0.870$).

11. Association between status of surgeon and development of complication

Fifty one (67.1%) of patients underwent surgery by a Senior House Officer (SHO), while Registrars undertaking neurosurgical training, operated on 24 (29%) of patients. One patient, operated by a consultant neurosurgeon, developed a shunt blockage. The rates of infection, blockage and shunt migration are shown in Table 8 below:

Status of the operating surgeon did not reveal a significant relationship with development of shunt infection or complication.

12. Association between shunt type and development of complications

Table 10 below outlines the relationship between type of shunt used and development of a shunt complication. The Chhabra shunt (developed in India), was the most commonly used shunt, being used in 61 (80.2%) of the 76 patients. The Pudenz-Heyer, Cordis Hakim and Elekta shunts were used in 5, 9, and 1 patient respectively. The infection rate was unduly higher in the later group (non Chhabra type shunts), the difference reaching statistical significance. However it must be emphasized that the number of these non-Chhabra type shunts was small, and the study design was not structured to effectively compare, and reliably comment, on the association between type of shunt and development of complication.

### Table 6  Symptoms and signs of shunt infection.

<table>
<thead>
<tr>
<th>Symptom and Sign</th>
<th>Number of Episodes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pyrexia</td>
<td>13</td>
</tr>
<tr>
<td>Irritability</td>
<td>11</td>
</tr>
<tr>
<td>Convulsion</td>
<td>6</td>
</tr>
<tr>
<td>Vomiting</td>
<td>3</td>
</tr>
<tr>
<td>Dehydration</td>
<td>4</td>
</tr>
</tbody>
</table>

### Table 7  Antibiotic prophylaxis versus risk of infection.

<table>
<thead>
<tr>
<th>Prophylactic antibiotics</th>
<th>Shunt infection occurred</th>
<th>No shunt infection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prophylaxis given</td>
<td>3/12 (14.8%)</td>
<td>85.2%</td>
</tr>
<tr>
<td>Prophylaxis not given</td>
<td>18/49 (36.7%)</td>
<td>63.3%</td>
</tr>
</tbody>
</table>

### Table 8  Presenting Clinical Sign

<table>
<thead>
<tr>
<th>Presenting Clinical Sign</th>
<th>Episodes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wound Sepsis</td>
<td>4</td>
</tr>
<tr>
<td>Csf Leakage</td>
<td>2</td>
</tr>
<tr>
<td>Exposed Shunt</td>
<td>3</td>
</tr>
<tr>
<td>Shunt Tract Inflammation</td>
<td>2</td>
</tr>
</tbody>
</table>

### Table 9  Association between status of surgeon and complication.

<table>
<thead>
<tr>
<th>Surgeons status</th>
<th>Patients with complications</th>
<th>Total number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Infection</td>
<td>Blockage</td>
</tr>
<tr>
<td>Consultant</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Registrar</td>
<td>7</td>
<td>4</td>
</tr>
<tr>
<td>SHO</td>
<td>8</td>
<td>2</td>
</tr>
<tr>
<td>Total (%)</td>
<td>15/19.8</td>
<td>7/9.2</td>
</tr>
</tbody>
</table>
13. Presence of neural tube defects (ntd) and shunt infection

Twenty two patients had an associated NTD (Spina Bifida or Occipital encephalocele), of whom five (22.7%) developed a shunt infection. Fifty four patients did not have an associated NTD; of this group 10 (18.5%) developed a shunt infection, as shown in Table 11.

14. Final outcome in patients managed for shunt complications:

Fifteen out of a total of twenty three patients (65.3%) who developed shunt related complications were successfully treated and discharged, while treatment of one patient was still on-going as an inpatient beyond thirty days after the shunt procedure. Seven of the twenty three patients (30.4%) were readmitted for treatment of their shunt complication, but succumbed and died, an overall mortality of 30.4%.

Within the shunt infection related group shunt, the mortality was 46.6%; shunt infection proved to be a particularly grave life threatening risk following shunt surgery. There was no mortality in the group that presented with shunt blockage or shunt migration. The results a tabulated in Table 11.

### Table 10  Shunt type and development of complications.

<table>
<thead>
<tr>
<th>Type of shunt</th>
<th>Pts with complications (%)</th>
<th>Pts without complications (%)</th>
<th>Total number of patients</th>
<th>% of total number operated</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chaberra</td>
<td>13 (21.3%)</td>
<td>48 (78.7%)</td>
<td>61</td>
<td>80.2%</td>
</tr>
<tr>
<td>Pudenz-Heyer</td>
<td>3 (60.0%)</td>
<td>2 (40.0%)</td>
<td>5</td>
<td>6.7%</td>
</tr>
<tr>
<td>Cordis-Hakim</td>
<td>6 (66.7%)</td>
<td>3 (33.3%)</td>
<td>9</td>
<td>11.8%</td>
</tr>
<tr>
<td>Elekta</td>
<td>1 (100%)</td>
<td>0</td>
<td>1</td>
<td>1.3%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>23 (30.3%)</strong></td>
<td><strong>53 (69.7%)</strong></td>
<td><strong>76</strong></td>
<td><strong>100%</strong></td>
</tr>
</tbody>
</table>

### Table 11  Presence of Neural Tube Defect in Congenital Hydrocephalus and association with shunt infection.

<table>
<thead>
<tr>
<th>Presence of NTD</th>
<th>Number of patients</th>
<th>Number of shunt infections</th>
<th>% of Total number in study</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present</td>
<td>22 (28.9%)</td>
<td>5 (33.3%)</td>
<td>22.7%</td>
</tr>
<tr>
<td>Absent</td>
<td>54 (71.1%)</td>
<td>10 (66.7%)</td>
<td>18.5%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>76</strong></td>
<td><strong>15</strong></td>
<td></td>
</tr>
</tbody>
</table>

### Table 12  Final outcome of patients who developed shunt complications.

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Number of patients in group of complication</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infection</td>
<td>4</td>
<td>21.7%</td>
</tr>
<tr>
<td>Blockage</td>
<td>3</td>
<td>43.6%</td>
</tr>
<tr>
<td>Migration</td>
<td>1</td>
<td>4.3%</td>
</tr>
<tr>
<td>Dead</td>
<td>7</td>
<td>30.4%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>15</strong></td>
<td><strong>100%</strong></td>
</tr>
</tbody>
</table>

IV. Discussion

This prospective study of 76 patients with non-tumoural Hydrocephalus treated with insertion of a Ventriculo-peritoneal shunt at the largest public health facility in the East, Central and Southern African region in Sub-Saharan Africa highlights the challenges faced by clinicians involved in the care of this common condition in this region.

Shunt complications occurred in 30.3% of patients. Shunt infections accounted for 19.8% and shunt blockage in 9.2%. Bayton found that shunt malfunction within 3-4 months of insertion was particularly indicative of shunt infection (9, 10). He proposed that colonized shunts develop blockage or malfunction due to formation of local adhesions and fibrinous deposits often with encystment by the greater omentum following discharge of micro-organisms into the peritoneal cavity (9, 10).

The infection rate in this study is considerably high; however this rate falls within the range of 0.0% to 38% reported by other authors (77). The aetiology of this high rate is likely to be multifactorial, with the hospital and operating room environment playing a role, as high infection rates have been reported in other operations at the same hospital (60). The study by Mukasa (60), noted that additional reasons such as poor nutritional status of the child, theatre overuse, overcrowding in the wards and poor aseptic protocols and long waiting times may be contributing factors for such high rates for surgical procedures at the institution.

The male to female ratio of 1:5:1 is similar to that reported in other studies, males predominance in children presenting with hydrocephalus. Although unconfirmed, this may reflect a greater concern for a male child in certain cultures. Rather than a predeliction for the male by the disease.
The age range was one month to 54 years, with over 70% aged below 12 months. The peak age at time of shunt insertion was 3-4 months. This is due to the aetiology being congenital hydrocephalus, a finding reported by other authors (66, 77). There were, however, 3 adults whose hydrocephalus occurred following head trauma.

Children below the age of two months had a greater propensity to develop shunt complication, with 41.7% of children below 2 months developing a shunt complication, compared to children between 3-12 months (26.2%). This was not statistically significant.

Studies evaluating time interval between onset of hydrocephalus and shunt insertion is not commonly reported in literature. This is possibly because, western studies do not encounter this issue, with the facilities being available treat the child very soon after diagnosis. Most children in this study were operated after a delay of 1-4 months after diagnosis. Several reasons contribute to such a delay. Poor diagnostic facilities in the peripheral regions of the country, the families inability to afford transport costs from peripheral regions to the National hospital in the capital, financial difficulty to purchase the shunt when, as is common, these are unavailable at the hospital , and finally, the parents reluctance to consent to the shunt procedure.

Interval between onset of hydrocephalus and first insertion of the shunt, however did not reveal an association with an increased risk of developing a complication (p = 0.0698).

Congenital hydrocephalus was the commonest type in this study. The diagnosis of a congenital cause was based on C-T scanning or ultrasonography in fifteen patients, and on clinical findings in forty patients. Of the fifteen patients with congenital hydrocephalus, fourteen were due to aqueductal stenosis, and one due to Arnold Chiari malformation. Eighteen patients (23.7%) had post-meningitic hydrocephalus, a diagnosis found to be more common in our study, possibly due to delay in effective treatment of meningitis.

This study also addressed the risk of developing shunt infection in relation to the aetiology of hydrocephalus. Statistically no association was found between the acquired and the congenital type in the development of shunt complication (p = 0.71). Other studies have, similarly, found no such association (40, 81, 88). Presence of Neural Tube Defects (NTD’s, including spina bifida / myelo-meningocele, encephalocele) did not increase the risk of developing a shunt complication in this study (p = 0.670). A previous study by Davis et al (20) similarly reported that aetiology of hydrocephalus, age of patient, and presence of an open neural tube defect was not associated with development of shunt infections.

The time interval from shunt insertion to development of shunt infection ranged from 2 days to 3 months (mean of 24 days) in this study. Seventeen out of the twenty-three episodes of shunt complications presented in the first month (73.9%). Many studies report the time interval between shunt insertion and onset of infection to range from a few days to several years (33, 69). Schoenbaum (88) reported 78% of infections presenting within the first four months.

Due to inability to pay towards the cost of a CT Scan, the majority of patients were diagnosed on clinical grounds, without radiographic evidence. Only 43% of patients in this study could afford a C-T scan or ultrasonography.

All the patients with a diagnosis of shunt infection had presented with fever. It was the commonest presenting symptom. Of the general features of shunt infection, fever is seen by many as the major indicator of shunt infection (69, 77, 88).

Inflammation along the shunt tract was seen in two of our patients with shunt infection. Schoenbaum (88) has suggested that the finding of erythema overlying the shunt tubing is pathognomonic of infection.

Wound breakdown and exposure of the shunt system was seen in three cases of shunt infection. In one case it had actually dislodged from the abdominal site.

V-P shunt malfunction/blockage has been reported, by some authors, to be a sign of shunt infection (9, 31, 77, 69), especially in the initial 3-4 months after insertion. The signs of malfunction include headache, seizure, bulging of anterior fontanelle, excessive rate of head growth and backing up of CSF along shunt tract, among others. In our study the commonest features of malfunction were tense/bulging anterior fontanelle and vomiting. Twelve patients had bulging anterior fontanelle, five (5) of who were diagnosed as shunt infection due to presence of fever. Quigley (77) found that 12% of patients with shunt infection presented with features of shunt malfunction.
Meningitis (evidenced by neck stiffness, Kernig’s positive sign, or isolation of bacteria from the CSF) has been reported in many series (30, 33, 69). Features of meningitis were seen in 6 of our cases of shunt infections (30%). George (33) found a very high rate of meningitis in his series, with 74.3% of infections presenting with meningitis.

Peritonitis resulting from infected CSF containing bacteria being discharged into the peritoneal cavity, has been reported by some workers (9, 31). In this study peritonitis was a feature in one case of shunt infection.

One patient presented with features of intestinal obstruction, and one with features of ventriculitis.

Shunt migration has been mentioned in many studies. Gichuhi (34) noted a shunt migration rate of 5.2% in his study. The catheter tip may erode through the bowel wall, causing extrusion of the catheter through the anus, along with a risk of ventriculitis, or peritonitis. In our study one case presented with a prolapsed shunt from the anus on defecation. The patient, however, had no clinical manifestation of shunt infection or malfunction.

Specimens of CSF were obtained for bacteriological analysis in all 15 patients with a clinical diagnosis of shunt infection. The CSF was obtained either by a ventricular tap or during shunt revision in the operating room. Only 4 cultures were positive. Two specimens grew Staphylococcus aureus while, of the other two, one grew Coagulase negative Staphylococcus and one grew a mixture growth of Proteus and Enterobacteriaceae. CSF aspiration from the shunt system for bacteriology was not carried out in this study. It has been established that Coagulase negative staphylococcus albus or staphylococcus epidermidis) are the commonest organisms involved in V-P shunt infections (9, 31, 69, 40) with staphylococcus aureus being reported as the second most common organism isolated in culture (9, 31, 40).

Several other factors that may influence shunt complication were also studied. Some of these factors include length of surgery, type of V-P shunt used, seniority of the surgeon, and peri-operative prophylactic antibiotic.

Length of surgery was calculated for each patient from the time of first incision to the last sutures, in minutes. The rationale was that the longer the length of surgery the higher the chance of complications. It was noted from this study that among the 15 patients with shunt infection, 7 had an operating time of between 15-30 min, while 7 had an operating time between 31-60 min. Only 1 out of the fifteen (15) patients, had an operating time of greater than 60 min. Statistically there was no correlation between length of surgery and rate of shunt infection ($p = 0.870$). A study done by Shutleff et al (92) assessing the procedure of ventriculo-aurecistomies, showed no correlation between length of surgery and rate of shunt infection.

The role of status (reflecting seniority) of the surgeon was assessed by this study. The Senior House Officer (SHO) is the postgraduate resident in general surgery at the Kenyatta National Hospital. SHOs are required to rotate for a period of 3 months in the neurosurgical unit. 52 patients (68.4%) in this study were operated on by an SHO; of these, 8 (15.4%) developed shunt infection. A Registrar is a qualified general surgeon, undertaking neurosurgical training within the neurosurgical unit, prior to proceeding for formal training at an accredited centre for a period of training. The Registrars operated on 23 (30.3%) of the patients in this study, out of which seven (30.4%) developed infection. A consultant operated on one patient. Comparing the outcome between the Registrars and SHO’s did not show any statistical significance ($p = 0.130$).

Some reports (81, 92) have shown no differences in infection rates between surgeons of greater versus and lesser experienced surgeons (example consultant neurosurgeon and registrars) while other reports (33, 71) showed inexperience of the surgeon as a risk factor in shunt infection. Another study (77) indicated that differences between surgeons and shunt infection may reflect an individual surgeon being prone to have high infection rates.

The type of shunt used for the procedure was also. Patients operated at Kenyatta National Hospital for shunt surgery, at the time, had to purchase the shunt themselves. The least expensive available shunt system is the Chabbra (Indian made) shunt (cost approximately, US $ 50). The Chabbra shunt was used in 61 patients (80.2%) in this study, of which 13 patients (21.3%) developed shunt complications. The remaining 15 patients (19.8%) in this study were shunted using the Pudenz-Heyer and the Cordis-Hakim shunts, out of which 10 (66.7%) developed shunt complications. This was statistically significant ($p = 0.0003$). These shunts are sometimes available at the Kenyatta National Hospital as donations from visiting surgeons from western countries. As they are often hand carried, protocols of secured aseptic transportation are
more likely to be breeched. The shunt system may indeed not necessarily be to blame. Other studies (90, 92) have shown no such co-relation between the type of shunt used and complications that developed.

There was, at the time of study, no standardized protocol at the Kenyatta National Hospital on the use of prophylactic antibiotics in shunt insertion surgery. A generic formulation of Ceftriaxone was used in this study, and patients randomly chosen to receive an intravenous dose of 250 mg of antibiotic during induction of anesthesia. A total of 27 patients (35.5%) received the antibiotic, out of which 3 (11.1%) developed shunt infection. Out of 49 patients who did not receive antibiotics 12 develop shunt infection, an infection rate of 24.5%. Of the 15 patients who developed shunt infection, 3 (20%) had been given prophylactic antibiotic while 12 (80%) had not received prophylaxis. Statistically no significant reduction in shunt infection was found using the antibiotic ($p = 0.160$).

Different treatment modalities are used in the management of shunt complications. All the patients with a diagnosis of shunt infection in this study were commenced on intravenous antibiotics for a total duration of 7-14 days. Many workers have recommended the use of intravenous antibiotics in the management of shunt infection (1, 49). In our study, 6 patients improved on intravenous administration of antibiotic alone without any surgical intervention. Many workers have reported unfavourable results in treatment of shunt infection with antibiotics alone without removal of the shunt (49, 88, 93). Although this option was used in some of our infections, it is to be discouraged. No patients in this study was managed with intraventricular use of antibiotics.

Nine (9) of the patients with shunt complications were managed by externalization of the distal end of the shunt. The externalized end was connected to a modified urine collection bag or, in some patients, a large syringe, and the CSF allowed to drain out. These patients were also administered antibiotics intravenously. Upon resolution of the pyrexia and improvement of clinical condition, the patients were returned to the operating room (OR) for a shunt revision.

External ventricular drainage, as has been cited by James (49), requires constant and experienced nursing, facilities to monitor and replace fluids, electrolytes and proteins. There is also the added risk of super-infection.

Seventeen patients with shunt complications had to be taken to the OR for revision of the shunt. One patient whose shunt was removed, did not undergo a shunt revision, as the clinical signs were suggestive of an arrested hydrocephalus.

It is now established that the best option in the treatment of any CSF shunt infection is immediate removal of the infected shunt (9, 69, 77, 78, 88) combined with antibiotic administration intravenously (9, 49, 88) and intraventricularly (9, 49) to control infection before a new shunt is reinserted. External ventricular drainage is advised in the intervening period (9, 49, 77, 88) to avoid complications that may arise from raised Intracranial pressures.

Outcome of patients managed for complications was also evaluated in this study. Fifteen of the twenty-three patients (65.3%) managed for complications in the study improved within 7-14 days and were discharged for follow up in the clinic. One patient was still in the ward being managed for shunt infection at the end of the 3-month period of follow up. Seven patients died as a result of the shunt infection. None of the patients being managed for shunt malfunction died. An overall mortality of 30.4% was therefore observed in the group of patients who developed a shunt complication in this study, with mortality occurring exclusively in the group of patients who developed shunt infection or systemic complication, related to the shunt infection.

V. Conclusion

In conclusion this study observed that at the Kenyatta National Hospital, which is a leading tertiary referral hospital in the East, Central and Southern African region of Sub-Saharan Africa, shunt complication rates, as well as the mortality associated with such complications, are notably high, with shunt infection being the commonest morbidity associated with ventriculo-peritoneal shunting.

The study highlights the commonest cause of hydrocephalus as being congenital, thus re-iterating the need to focus on factors that contribute to high incidence of congenital hydrocephalus, not only in this hospital but, indeed, in this region of Sub-Saharan Africa as a whole.

Ventriculo-peritoneal shunting had been, until the time of the study, the only method available at the Kenyatta National Hospital for treatment of Hydrocephalus. The study seeks to encourage the development and promotion of neuroendoscopic management of hydrocephalus, which has now commenced at this hospital and several other...
hospitals in the region. It is a technique that promises a significantly more refined and scientific approach for hydrocephalus, a condition that can have devastating outcomes if not managed well, yet has the potential of exceptionally gratifying outcomes if the correct management approach is offered in timely fashion.

References


HYDROCEPHALUS ESSAY

“Health of Children Worldwide, Seeking God’s Guidance!”

Shizuo Oi, M.D., Ph.D.
HYDROCEPHALUS ESSAY

“Health of Children Worldwide, Seeking God’s Guidance!”

Part I-1
Offering a helping hand to the hydrocephalic children of Kenya!
Report on implemented lectures and live-surgery hands-on seminars

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I. Mission

December 6, 2007, 6:30 p.m. Singapore Airlines Flight No. SA 182 landed at Nairobi airport, after departing from Singapore and stopping over at Johannesburg, South Africa. One leg of the flight took seven hours, another 10 hours, and the final leg took 21 hours, clocking up a total of 21 hours to reach Nairobi. It certainly was a long flight. The town was a very large, dark-brown wasteland with little greenery. I saw several giraffes in the distance below the airplane. The town was sponsoring a domestic academic conference, which had as its theme, as I understood in Tokyo, “the evolution of neuroendoscopic surgery from a global perspective and the issues and ambitions held by this country.” The event also included a worldwide satellite relay of a live symposium (photograph 1). I was not fatigued, and my stamina had been restored, since I had spent most of the ten hours it took to reach Johannesburg sleeping. The first step of my Kenyan activity started here.

I made yet another transfer and with the scenery of the mountaintop of Kilimanjaro visible to my right (photograph 2), I headed for Mombasa, the town where hydrocephalic children were waiting.

The atmosphere of Mombasa’s airport was as brutal as Nairobi’s airport. The exit area of the airport was awfully crowded with people welcoming passengers.

There were people of various races. I was uneasy, since I had heard that public order was not good. An African approached me. “Professor Oi...?” he asked. I finally heaved a sigh of relief and went on to arrive at the
“Sarova Whitesands Hotel,” where the conference was to take place.

At the hotel, Dr. Mahmood Qureshi welcomed me with a full-faced smile. Dr. Qureshi is the head of the Central African Neurosurgery Institute. “In Kenya, it is impossible to introduce expensive shunt treatments. For this reason, there exist extremely miserable hydrocephalic children who are untreated, and their number reaches up to 1,200 every year. I would like you to teach the technology of neuroendoscopic surgery, which has yet to spread in this land. Last year, finally in 2006, the first successful neuroendoscopic surgery was carried out. And what was used there was none other than the Oi Handy Pro endoscope, which you had developed in Germany,” said the person who had eagerly invited me.

I’d like to relate what happened in May of this year (2007). I was appointed as the Chairman of the Board of Directors of the International Federation of Neuroendoscopy, and attended the Fourth International Endoscopic Conference held by the federation in France (Versailles, May 9, 2009 through May 12, 2009) (photograph 3). On this occasion, Dr. Qureshi requested me to offer my cooperation and guidance for promoting hydrocephalic treatment activities in Kenya.

The conference covered up to 50 countries, and about 400 participants came from various countries. After finishing my lecture as the chairman, two African neurosurgeons began to talk to me. They were Kenya’s Dr. Qureshi and his staff member. He said, “Even the Central African countries had joined this federation, and furthermore, for the sake of hydrocephalic children who remain untreated, I would like to see the practice of neuroendoscopic surgery to spread.”

For hydrocephalic research workers like myself who engaged in discussions of advanced medicines as representatives of developed nations, and engaged in the competition of coming up with the better research results, this was a new aspect of the conference with perspectives that completely differed from what we had been accustomed to. It is known all over the world that many children born with the affliction of hydrocephalus continue to go about their daily existence without receiving treatment to this day in Africa and are essentially left untreated. I heard such an opinion as, “Medically speaking, even if African countries participate in the conference, they do not necessarily progress academically.” It is my intention to reflect the fruits of the latest hydrocephalic studies in the medical scenes the world over…I asserted that this is the aim of the federation, and the members of the board of directors
On December 7, 2007, which was the next day after my arrival in Kenya, at 8am, The College of Surgeons of East, Central, & Southern Africa (COSECSA) conference was held along with the 58th Academic Meeting of The Association of Surgeons of East Africa (ASEA) at the Savora Whitesands Hotel. My keynote address was titled, Advanced Hydrocephalus Research and Hydrocephalus Treatment All Over the World Has Begun. I began the speech by saying, “I am truly honored to have been invited in Kenya to attend the academic gathering of The College of Surgeons of East, Central, & Southern Africa. As a researcher investigating hydrocephalus, and as a pediatric neurosurgeon and as a Japanese, I came to the land of Kenya with the hope of helping to save even just one more child afflicted with hydrocephalus. At this moment, I am filled with this sense of mission,” I said. (Photographs 4, 5)

II. An African class for developing manual neuroendoscopic surgical skills

In the academic gathering of the federation of the greatest surgeons representing East Africa, Central Africa, and South Africa, I turned out to finish a special lecture as the first Japanese doing so, and delivered a keynote address related to hydrocephalus, which nobody in the world had touched upon. However, I was not necessarily satisfied. I was not certain whether my audience had truly understood the nature of hydrocephalus, an intractable illness, and how we should be thinking about its treatment. “Will this be an opportunity to save many hydrocephalic children in Africa who remain unable to receive treatment...?” The figure and facial expression of a Kenyan mother embracing her baby and seeking help flashed in my mind. No matter how intense the appeal, reality cannot be improved with just words alone.

After wrapping up my special lecture, I immediately headed towards the assembly hall where the “Oi Handy Pro Hands-on and Hydrocephalus Course” was all set up. There, I found prepared six units of the Oi Handy Pro, the latest neuroendoscope I had constructed during my research at the International Neuroscience Institute (INI), where I held multiple posts.

While in the former century, the fiberscope was miniaturized and came to be applied for use within the brain, this prevented doctors from carrying out sterilization (autoclave high-temperature heat sterilization) to prevent the contraction of the Creutzfeldt–Jakob disease (a kind of prion disease such as mad cow disease), which can be caused from the instrument’s repeated use within the brain. For this reason, the use of the fiberscope remains banned in advanced nations. As a solution to this, I spent seven years in Germany completing a neuroendoscope model in 2003 that established an operation channel with the world’s narrowest diameter and the highest image resolution, making use of a rigid mirror lens that reproduces a higher picture quality than the fiber lens. In 2005, I reported to the Journal of Neuroscience on the extremely delicate freehand surgical technique required to operate the Oi Handy Pro. (Photograph 6) Since then, over 200 units came to be used in various countries, saving many hydrocephalic children in the process.

In the assembly hall where the “Oi Handy Pro Hands-on and Hydrocephalus Course” was to take place, 20 applicants were chosen from ten African countries to participate. (Photograph 7) Judging from the common sense of the Japanese medical situation today, this is surprising. However, in Africa the number of physicians is absolutely miniscule, not to mention their ratio to the population. For example, neurosurgeons in Kenya amount to ten, in neighboring Uganda, three, in adjacent Tanzania, Kampala, Addis Ababa, and Cameroon, there are only one to two in each of them. This is the reality. This handful of neurosurgeons is fighting against encephalic disorders for 237 million people in 10 countries. What’s more, half of them are children. On this earth, there are 2,200 million children and among them 1,900 million live in developing nations. Every year,
illnesses claim the lives of ten million children under the age of five.

The neurosurgery specialists who participated in this “Oi Handy Pro Hands-on Course” are representatives who have single-handedly been undertaking cerebropathic treatment in each of the ten countries. If I instruct here properly the surgical technology of the neuroendoscope, I could help realize a breakthrough for the treatment of hydrocephalus in African countries. All the participants received my guidance eagerly. Their manual skills were not bad at all. And their mastery of new skills was quite fast! At this point, I concretely affirmed for the first time here the magnitude of the position in which I found myself.

In African countries there is an enormous number of patients, but hospitals are limited in that they only accommodate one through three patients. So I wondered what would happen if there were to be a rush of people afflicted by cerebropathic ailments. Wouldn’t these hospitals fail to cope? However, Dr. Qureshi answered, “That will not be the case. We will form a team comprised of 8 Kenyan neurosurgery specialists and two anesthesiologists who specialize in pediatric anesthesia. With a ‘Mobile Endoscopy System’ kit equipped with an “Oi Handy Pro,” they will make rounds to various hospitals and carry out treatment.” (Photograph 7, bottom).

III. Facing the challenge of operating on Kenyan children, whose heads have reached gigantic proportions

After presenting the special lecture at The College of Surgeons of East, Central and Southern Africa (COSECSA), and conducting a technical guidance class on the manual surgical skills for neurosurgeons representing ten countries, I finally left for a hospital in Mombasa, where the hydrocephalus children of Kenya were waiting. The people of the town were all poverty-stricken. I saw people pushing bicycle-drawn carts loaded with foodstuffs and belongings, and people after people, covered in clouds of dust, running street stalls. There, the hot tropical sunlight mercilessly glared down. Covered with perspiration, the skin of the people shined black. Still, everyone was working hard. I arrived at the hospital on the itinerary and exchanged greetings with the director-doctor and went to examine the child patients at once. At that moment, before I knew it, I was at a loss for words. The size of the head approached 70 cm, and the eyes had sunk due to the setting sun phenomena, and the gigantic head appeared to be hanging over and covering the small face, which was under strain. Why did it reach this extent...?! (Photograph 8).

I wondered how many children such as this could I possibly treat within two days? I carefully examined the conditions and the findings evident through the encephalic images. The cerebral forms of these children were already turned into aqueous spaces, and their cerebral parenchyma had, for the most part, become paper-thin... I thought that five children appeared to be suited to receive neuroendoscopic surgery. The ages of the infants ranged from four and half months to ten months, and their head...
girths (the maximum cephalic boundary length) ranged from 47 cm through 67 cm. An adult’s head girth reaches up to a little more than 50 cm. It is just unimaginable just how large the head is in relation to the small body. In the eyes of the mother who was embracing the child, the eyes that were seeking help, I was reminded of my own mission very strongly—I stand here in Kenya for this child, I affirmed. (Photograph 8).

I changed into my surgical gown and began to sterilize by washing my hands. But the reality was that there was only one big disinfectant soap available for this purpose. About 50-odd years ago, during my childhood, I had observed the surgeries conducted by my father. The soap that I was looking at the time recalled to my mind the figure of my father of those days when he used to carry out hand-wash sterilization. The skills of the two female anesthesiologists were great. They managed to position the large head well and skillfully entered into intratracheal intubate ventilation. With an indication from Dr. Qureshi, assistants involved in the surgeries of the five children, on whom I would be operating, began to prepare themselves in pairs. The first surgery was to be conducted on a seven-month old girl, whose head girth was 47 cm, but even at this size, it was still the smallest. Since this was the first surgery, I was to demonstrate all the procedures myself. “In the juvenile example, in the case of establishing a slight bar hole, you must not use an anterior fontanel. Also for the bar hole, you should apply manual skills that help to establish bone frap. To produce less of a pool of cerebrospinal fluid below dura matter, edema, and the skin, you should pack the passage of the neuroendoscope. To this end...” and so and so forth. Consequently, I went on demonstrate the operational methods of the “Oi Handy Pro.” (Photograph 9)

When I went inside the brain of the first hydrocephalic child, firstly, I was surprised by what I had never seen until then. There was a high-protein lump within the encephalic chamber (the cerebral ventricle), due to being strikingly affected by the flow (the circulation) of the water (cerebrospinal fluid) inside the gigantically expanded brain of the child. I soon opened up, all the way, the washing mouth of the “Oi Handy Pro,” and went on to wash inside the cerebral ventricle, using several liters of artificial cerebrospinal fluid, whose method of preparation I had just instructed. One hour passed, but the structure of the cerebral chamber remained opaque. So I installed a cerebral ventricle drainage, and made plans to repeat the washing every day from now on, then made plans for a shunt operation, and thereby ended my first Kenyan surgery case. I came to Kenya with the mission to transfer the technology of neuroendoscopic surgery to improve the treatment of hydrocephalus in Africa, where it is not possible to purchase expensive, up-to-date shunts and apply clinically. However, at that moment, my shoulders suddenly dropped in disappointment...

Regarding hydrocephalus, I was convinced that I understood most things about it, but my facial expression had become fierce. Simply put, I had never detected such atrocious surgical findings of hydrocephalus, neither in the USA nor Germany even, not to mention Japan! When the anesthesia for the second case began and I was about to sterilize by hand-wash again, I saw my face in the mirror used for checking the appearance of surgeons. I realized how stern looking I had become with all these thoughts running across mind. (Photograph 10). I was feeling that I stood no chance against the challenge of this gigantic condition of hydrocephalus.

I began pursuing studies in hydrocephalus in 1977, thirty years ago from today, which traces back to my days of residency in Northwestern University in Chicago,
USA. In those days, in the Children’s Memorial Hospital, which used to be the world’s top pediatric hospital, I was being taught hydrocephalus and pediatric neurosurgery by Professor Anthony J. Raimondi, who is said to be the father of modern pediatric neurosurgery. In the course of one of his classes, I came across the chance to examine a fourteen-year old child whose state of hydrocephalus had resulted in a head girth of over 110 cm. This child, when born, was already profoundly afflicted with the condition, and was told by another hospital that the lifespan would perhaps last only two years. For this reason, the child was left untreated, but grew up to the age of fourteen. The child’s inborn hydrocephalus constantly progressed and would manifest in time as a gigantic head. However, back then, there was absolutely no knowledge to inform what was going on inside the brain of this child, what types of morphological changes and transformations in the property of the spinal fluid took place. While remembering all this, I remained unable to understand why the water (cerebrospinal fluid) in this child’s brain had become such a sticky, lump of a high level of albumin. ‘Was there an infection?’ I wondered.

The second Kenyan hydrocephalic child had the largest head girth expansion among the five children. This child was a girl whose head girth had reached 67 cm in a matter of four and a half months. The anterior fontanel (the boneless, dented part found at the center of a baby’s head) was largely opened up and was under strain. Even diagnostic imaging showed that thinning of the cerebrum had occurred, giving it the look of paper. However, the cerebellum and brainstem parts were recognized as normal. The thalami on both sides seemed to appear projected and were seen inside the huge ventricular enlargement, which formed from the unification of the lateral ventricles found on both sides. They had become remarkably enlarged after an emptiness was formed. (Photograph 11) However, I was not able to grasp at all the structures, the images indicated morphological changes inside the brain that could have only been caused by an extremely terrible state of hydrocephalus. I saw that the thalami of both sides had swollen to a large extent and was seen projected outward inside the cerebral ventricle, which had become unified. Even the midline structure was entirely destroyed, and the central wall, septum, and the roof of the third ventricle even were not visible. I could observe from the top that the fornices on both sides joined at the midline. (Photograph 12) However, identifying the foramen of Monro and the third ventricle was difficult. This was an extremely difficult neuroendoscopic surgery presentation to make.

When it comes to performing a surgery, regardless of whether it is neuroendoscopic or not, I am mindful of enforcing an operation in the most safest way possible. I believe no surgeon should overextend themselves or take risks. No one’s life should ever be entrusted to the spirit of your own adventure. “…These seen in the thalamus are probably the mammillary bodies of both sides.” “… The fornices of both sides running below them appear to be coming out from there…” The Kenyan neurosurgeons had studied these areas well. However, without compromising, I went on to operate the “Oi Handy Pro,” the world’s narrowest, rigid endoscope to probe deeper between the greatly protruding peaks of what were thought to be the thalami. Then, there appeared in the slit, closed cerebral aqueducts, the mammillary bodies of both sides, and a spread across the monitor screen. There was no opacity in the cerebrospinal fluid. Strictly speaking, the water was clean. While I was able to grasp all the structures, the images indicated morphological changes inside the brain that could have only been caused by an extremely terrible state of hydrocephalus. I saw that the thalami of both sides had swollen to a large extent and was seen projected outward inside the cerebral ventricle, which had become unified. Even the midline structure was entirely destroyed, and the central wall, septum, and the roof of the third ventricle even were not visible. I could observe from the top that the fornices on both sides joined at the midline. (Photograph 12) However, identifying the foramen of Monro and the third ventricle was difficult. This was an extremely difficult neuroendoscopic surgery presentation to make.

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The bottom of the third ventricle was confirmed without a doubt. It had turned flat and remarkably narrow. Still, I was able to neatly open it up and the Liliequist membrane. Yay! Ole!! All staff members, visitors, and surgical assistants present in the operating room let out shouts of joy. A hydrocephalic child in Kenya was saved with the neuroendoscope.

The next morning, on that day, the remaining third case was scheduled. The memory of yesterday’s success with the difficult second case still lingered in my mind. I exchanged congratulations with each one in the team, and noticed how cheerful everyone looked. The child of the third case for the surgery today was a five-month old girl with a head girth of 63 cm. This was a significant state of hydrocephalus. It was a merge between holoprosencephaly and the Dandy-Walker syndrome. Previously, she was treated with shunts, but this was discontinued due to an infection. Still, her head turned out to become gigantic and the hydrocephalus continued to worsen. (Photograph 13)

The team moved more swiftly than the previous day, and their tempo was good. The neuroendoscope was immediately inserted into the huge head. What we then saw took us by surprise again! On the cerebral ventricle wall seen in the depths of the cerebrospinal fluid, I saw innumerable amounts of small, round grains stuck onto it. They appeared to be like bacteria that had grown on a culture medium and had spread out, crowding the area. This time as well, no matter how many times I washed, the normal structure of the cerebral ventricle would not appear and the murky cerebrospinal fluid did not become clean. Without curing the infection, I could not proceed to the next step.

The fourth case was a ten-month old boy and his head girth was 58 cm. In terms of the visual data, there was a blockage in the cerebral aqueduct, and a huge partition was visible in a huge cerebral ventricle, and on an extremely flattened out cerebral parenchyma, several places had accumulations of cerebrospinal fluid. They looked like ponds. However, the fluid this time was absolutely colorless and transparent, helping to reproduce a beautiful picture of the cerebral ventricle on the monitor screen. There was a winning chance here! In the gigantic cerebral ventricle, the midline structure, as expected, had largely collapsed and the third ventricle was pushed away into a far deeper place. However, the original shape of the foramen of Monro had been retained, with the fornix running side by side with it. I made the endoscope enter below the right fornix and reach the bottom of the third ventricle. The structure here was also narrow, but the mammillary body and the roto dip were recognizable. In accordance with the shape, opening up the third ventricle was easy, but the Liliequist membrane was tough. I grasped a part with minute forceps and excised it, and while opening up the forceps, I enlarged the fenestration area. The passage to the basilar cisterna was opened, and in this way, the second child found relief.

IV. The New Pathologic Physiology of Hydrocephalus I observed in Kenya

The last and fifth child was the case with a head girth of 65 cm at just 10 months age. The extremely stretched foniculus anterior was extremely large. The fluid within the brain erupted the moment a needle was inserted, and it appeared to be a clean cerebrospinal fluid. But in fact that was just a supernatant portion of CFS, and deep within the ventricles existed the highly viscous, opaque, semi-fluid liquid containing high protein. We tried to wash off with three or four liters of artificial cerebrospinal fluid, but that didn’t change a thing. It was as if we were pouring water into solid concrete. Observing the otherworldly scene through the monitor screen of the neuroendoscopy, I felt helpless again. Why does such a substance exist within a brain that is devoid of any tumors or infectious diseases? But momentarily it occurred to me that this finding was common with the first case and it might be “Froin’s sign,” which had unquestionably developed within the ventricles. Is this the last destination of hydrocephalus? No one has ever proved the existence of “Froin’s sign” under endoscopy or direct observation. Physiologically, human beings naturally need cerebrospinal fluid and its circulation. I knew that reducing its production by cauterizing the choroid plexus was meaningless for treating hydrocephalus, but moreover, I did not know that decreased circulation of the cerebrospinal fluid would
result in the accumulation of protein, causing extremely strong disorders to the brain!

In 2000, we reported cases of an adult hydrocephalus physiopathology in which patients showed no symptoms throughout the growth, including during childhood, despite the extremely large volume of cerebral ventricles with macrocephaly. However, those symptoms appeared with an increased intracranial pressure in adulthood for some reason. We named this disease as Long-Standing Overt Ventriculomegaly in Adults (LOVA) and this term is now commonly used to represent a type of hydrocephalus. I discussed feverishly with Dr. Qureshi the pathological conditions of hydrocephalus in the five children in Kenya. LOVA was also an extremely difficult pathological condition of hydrocephalus to treat at the time when shunt operations were prevalent. Then with the advent of neuroendoscopy, its treatment saw a remarkable advancement. I have also learned a new finding of hydrocephalus here at Kenya. Dr. Qureshi and I named this disease as Overt Infantile Ventriculomegaly (OIV). This pathology is even more difficult to treat, and even with neuroendoscopic surgery, a cure has not been found. This pathology has brought up many more problems for the study of hydrocephalus treatment.

After listening to my passionate talk on the pathological conditions of this type of hydrocephalus, Dr. Qureshi inquired if neurosurgeons in Kenya could study in Tokyo. Neurosurgeons in Kenya who treat dozens of children with the type of hydrocephalus that triggers the development of huge head sizes understand the academic significance of such cases, and are showing interest in the study of the disease. This was another significant outcome obtained from the visit to Kenya. As I had seen the surgeries of those 5 children with hydrocephalus, the fundamental operative techniques of the Kenyan neurosurgeons were pretty good and I believed that they would soon master neuroendoscopic surgery techniques. The more important issue here was judgment concerning indications and the ways of neuroendoscopic surgery based upon characteristics of extremely various pathological conditions of hydrocephalus and cerebrospinal fluid circulation. In the neighboring country, Uganda, it was reported that surgeons there had resorted to burning the choroid plexus with the neuroendoscope to stop the progression of hydrocephalus because they couldn’t purchase expensive shunts. This was found problematic and received criticism within Africa as well. I hope budding researchers of hydrocephalus in Kenya will exercise leadership in academic societies by one day arguing with confidence about the harmfulness of stopping cerebrospinal fluid circulation, which would adversely and considerably affect the development of the brain.

After completing operations on five cases of pediatric hydrocephalus, Dr. Qureshi said that my visit to Kenya this time would become a great starting point toward the development of neurosurgery in Kenya (Photos 14, 15). Later we had dinner together in a restaurant on a big boat by a huge river that runs through Mombasa (Photo 16). Asked to make a speech, I said, “I believe each member of the neurosurgeon team in Kenya has learned many things including techniques of neuroendoscopic surgery through the operations carried out on the five children with hydrocephalus. Also, as a researcher of hydrocephalus, I have learned a new pathology of the disease. In other words, I have found that another form of hydrocephalus exists here in Kenya that cannot be overcome with our current research capabilities and treatment technologies. But as Dr. Qureshi has said, “This is our first great step forward.” We always learn what is needed to make a refreshing start from a sense of failure and helplessness. A light of advancement shines within a failure. There’s no stronger power than the will to overcome difficulties. We are revived by such situations.” After I said these last words, rounds of applause and cheers from the Kenyan neurosurgeons spilled out of the boat and resounded across the large estuary of the river.
It took as long as 48 hours to fly back from Kenya to Japan, and I had to switch flights at Johannesburg in South Africa on the way from Mombasa due to changes in the schedule. I decided to stay at a first-rate hotel, considering the poor state of security in the country. In Johannesburg I saw gorgeous buildings and people reveling at casinos. The scenes were far removed from the smiles of those who were desperately trying to save children’s lives amid the poor living conditions in Kenya. They were as far removed as heaven from earth. They were, in fact, fitting for Moses to appear and show God’s wrath by hurling flaming stone tablets of the Ten Commandments. The next morning, while observing the African continent grow more distant from the window of the airplane that departed Johannesburg (Photo 17), I couldn’t let go of the image of the mother’s face holding her baby with the huge hydrocephalic head in her arms. I shall return, by all means, I thought to myself. And the next time, if I can save as many as just one more child, I will be grateful. “May the hand of rescue touch the hydrocephalic children of Kenya!” This is my prayer.

Epilogue

Human beings cannot have the hands of god. Any surgeon who is under the misunderstanding that this is possible is guilty of terrible arrogance. But human beings do have the ears and heart to listen to God’s voice, and thereby serve as god’s hands and feet.

(December 11th, 2007. On board flight SQ012 bound for Japan from Africa)
Prenatal Diagnosis of Fetal Hydrocephalus
Part 1: Holoprosencephaly
Case Illustration: Alobar versus Semilobar type of Holoprosencephaly in Prenatal Differential Diagnosis

History:
A 36-year-old healthy woman in 28 weeks gestation, gravida 3, para 2. She was attended to our outpatient clinic because ultrasonography at 23 weeks gestation revealed a monoventricle. The baby has had a normal growth. At 26 weeks of gestation fetal MRI revealed holoprosencephaly either alobar or semilobar type, and ultrasound monoventricle findings were confirmed also with this imaging study. The parents do not have history of familiar or personal genetic disorders neither underlying diseases. The other two children are normal and healthy. The mother has not been exposed to any known teratology agent during her pregnancy. Actually the pregnancy develops normal.

Neuroimaging: Fetal MRI

MV: monoventricle and DC: dorsal sac (cyst) in our opinion

QUESTION

What is your diagnosis, alobar vs. semilobar type of holoprosencephaly?
“My Opinion”

[ ] Fax: 0081-3-3235-9377 [ ] e-mail: shizambroi@aol.com
HYDROCEPHALUS On-line Journal Concensus Conference [HCOLJCC]
on HCOLJCC No._________

[ ] I agree! [ ] I disagree!

What is your diagnosis on the fetal MR images?
[ ] Alobar Holoprosencephaly [ ] Semilobar Holoprosencephaly [ ] Other

Name: __________________________, M.D.
Institute: ______________________ City: __________, Country_________

[ ] I permit the above opinion and comment to be published with my name and
institute/country in “Journal of Hydrocephalus”
[ ] e-mail address: ________________@______________

Comment


I. INTRODUCTION

Holoprosencephaly (HPE) is a severe brain malformation, caused by abnormal cleavage of the prosencephalon in the fifth week of embryologic development. HPE represents a failure of the forebrain into 2 hemispheres. The presentation rate is in 1 of 250 miscarriages and in 1 of 10,000-16,000 live births. These figures place it as the most common human malformation of the forebrain and associated to face defects. According to the severity, the brain defect is often associated with characteristic dysmorphic facies which are formed secondarily to the brain malformations.

Moreover, the fusion of the cerebral hemisphere is
associated with a lot of midline anomalies, such as absence of a septum pellucidum and corpus callosum; even the less severe of forms of HPE show fusion of the thalamus, cingulum and caudate nuclei. The hypothalamus and pituitary may be also affected. Midline facial anomalies ranging from synophthalmia, cyclopia and proboscis to mild hypotelorism, can be present. Ethmocephaly, cebocephaly and cleft lip are also a possible finding\textsuperscript{5, 23, 24}.

The neurodevelopmental defects in HPE are variable; therefore HPE has been classified into 3 main types on the basis of severity: alobar, semilobar and lobar. Alobar is the most severe, the almost complete lack of separation or fusion of the cerebral hemispheres; it is accompanied by a monoventricle, the single ventricle is described in MR imaging like a horse-shape appearance\textsuperscript{7}, which often communicates with a dorsal cyst; and facial complex deformities, such as cyclopia, are common. Semi-lobar HPE type, often shows a lack of separation of the hemispheres anteriorly, but may show separation posteriorly; moreover, the ventricle formation is affected because there are no frontal horns formation, whereas the occipital horns are partially formed and the third ventricle is very small; midline structures such as the corpus callosum and septum pellucidum are partly formed or disappear when the interhemispheric fissure is absent, the thalami may be partially or completely fused; and facial deformities rang from none to minimal\textsuperscript{7, 10, 21}. In the lobar form only minor changes may be seen, for example, the anterior falx cerebri and septum pellucidum are usually complete; the ventricle system do not suffer many anomalies just the frontal lobes and horns are hypoplastic; and facial malformations are rare or absent\textsuperscript{6, 7, 10, 20, 24}.

The etiology of HPE is heterogeneous and complex, including genetic and environmental causes and their interactions. Between environmental causes, the HPE has been associated with maternal exposure to retinoic acid and toxins, poorly controlled maternal diabetes and infectious diseases (rubella, cytomegalovirus infection) \textsuperscript{17}. Among the genetic causes associated are chromosomal abnormalities (trisomies 13 and 18), triploidy, deletions, duplications, rearrangements of at least 12 gene loci and mendelian inheritance \textsuperscript{4, 11, 17, 24}.

The prognosis of alobar HPE is poor, only 50% of patients with alobar HPE will survive to 4 or 5 months of age, and only 20 to 30% will survive by 12 months of age\textsuperscript{2}. On the other hand, 50% of the patients with semilobar or lobar types of HPE survive beyond 12 months\textsuperscript{9}. The early diagnosis of fetal malformations, especially those of the central nervous system is very important, both because of the poor outcome and because of the emotional distress caused by the accompanying brain and facial malformation\textsuperscript{11}.

II. Case Illustration
A healthy 36-year-old woman (gravida 3, para 2) visited our outpatient clinic at 28 weeks gestation, because ultrasonography at 23 weeks gestation had revealed a monoventricle. The fetus had shown normal growth. At 26 weeks gestation MR imaging of the fetus revealed HPE of either the alobar or the semi-lobar type, and confirmed the as well ultrasound findings of a monoventricle. The parents had no history of familial or personal genetic disorders or underlying diseases. Their other 2 children were healthy. The mother had not been exposed to any known teratogen during her pregnancy.

MR imaging showed that the spine and cranium were well conformed. A single posterior ventricle was identified. The interhemispheric separation was not evident; moreover the thalami were fused, and the septum pellucidum and corpus callosum were absent (Fig. 1). On coronal MR imaging, craniofacial malformations were not apparent (Fig. 2-B). The MR imaging findings were not entirely consistent with either alobar or semilobar HPE, but were intermediate between the 2 types. In particular, discordance between the severity of brain and facial abnormalities was evident.

III. Discussion
The spectrum of facial and brain anomalies associated with HPE can be recognized in utero by means of neuroradiologic imaging, reason why they become in our primary tools for early diagnosis, besides they are non invasive techniques. A special attention should be paid to the facial morphology in aspects so simple like the interorbital distances; and also to the anatomical variations of brain, like the ventricular configuration, it is important to notice the presence or absence of the interhemispheric fissure\textsuperscript{10}. Even though the ultrasonography gives the initial diagnostic impression, MR imaging is the most useful method for examining patients with HPE because of its superior definition of the corpus callosum, septum pellucidum, thalami and ventricle system\textsuperscript{7, 9, 15}. Thus MR imaging allows the clear identification of the anatomical features of brain abnormalities and the differential diagnosis of HPE (Table 1).

The abnormalities of brain in the present case are more consistent with the alobar type. But the facial findings suggest a less severe abnormality of the brain, as in the
semi-lobar type. The altered forebrain appears to be related directly to the mesoderm precordial mesenchymal tissue; this tissue is normally the responsible for the division of forebrain and the development of the midface.\textsuperscript{12} Patterson\textsuperscript{17} (2002) in the paper “The face predicts the brain; the image predicts its function” also notes the close relationship to the development of both the brain and face, and suggest that the spectrum of facial dysmorphism broadly reflects the severity of the brain malformation. The phenotype is directly proportional to the separation and integrity of the brain, most of the time\textsuperscript{17}. Multiple author Barkovich\textsuperscript{1} (2000), Blaas\textsuperscript{3} et al. (2002), Patterson\textsuperscript{17} (2002), have proposed that only the less-severe forms of HPE do not have major facial anomalies, supporting the idea that “the face predicts the brain”. Furthermore, Van Gool S et al. (1990)\textsuperscript{22} have described a patient who have alobar HPE, diabetes insipidus and coloboma, but had no craniofacial abnormalities; results of an examination at birth were unremarkable, an alobar HPE was not diagnosed until the age of 2 months, when the infant was hospitalized with endocrine problems related to hypophyseal dysfunction, which is common in HPE\textsuperscript{24}.

Therefore, the case we described and Van Gool\textsuperscript{21} case suggest that a broad range of phenotypes exist in HPE. This phenotypic variability could be associated to the etiological heterogeneity. The joint action of genetics and environmental factors could be responsible for the variables phenotypes found, and the apparent discrepancies between the developing brain and face. However, the mechanism by which environmental and genetic interaction lead to phenotypic variability remains unclear. Therefore, the notion that “the face predicts the brains”, does not always hold true, like we were thinking. It is important to identify in prenatal life the variations of spectrum in the types of HPE, for make a more accurate diagnosis.

Clinical manifestations in HPE include mental retardation; developmental delay; hypogonadism; seizure; hypotonia or hypertonia; motor, endocrine, and autonomic dysfunction, among others problems\textsuperscript{9, 10}. Treatment is usually supportive; although the prognosis

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**TABLE 1** Shows the MR imaging features of semi-lobar and alobar HPE and the findings of the present case. (Adapted from Grossman RI et al., 2005)

<table>
<thead>
<tr>
<th>Feature</th>
<th>Semilobar</th>
<th>Alobar</th>
<th>Case</th>
</tr>
</thead>
<tbody>
<tr>
<td>Facial deformities</td>
<td>None</td>
<td>Moderate</td>
<td>None</td>
</tr>
<tr>
<td>Fark posteriorly</td>
<td>Absent</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Thalami</td>
<td>Partially</td>
<td>Fused</td>
<td>Fused</td>
</tr>
<tr>
<td>Interhemispheric fissure</td>
<td>Present</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>Dorsal cyst</td>
<td>Absent</td>
<td>Present</td>
<td>Present</td>
</tr>
<tr>
<td>Frontal horns</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>Septum pellucidum</td>
<td>Absent</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>Splenium</td>
<td>Posterior</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>Third ventricle</td>
<td>Small</td>
<td>Absent</td>
<td>Absent</td>
</tr>
<tr>
<td>Occipital horns</td>
<td>Partially</td>
<td>Absent</td>
<td>Absent</td>
</tr>
</tbody>
</table>

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**FIG. 1** Holoprosencephaly. A. Monoventricle (MV) and dorsal cyst (DC) are distinguished in sagittal MR imaging, with no corpus callosum. B. Single posterior monoventricle and thalamus fused. A dorsal cyst can be recognized.

**FIG. 2** A. Monoventricle (MV) in coronal MRI. B. On coronal MR imaging no craniofacial malformations are evident. Not even hypotelorism can be identified. An outline of the ventricular division can be observed in midline (arrow).
of HPE depends on the type of HPE and the extent of facial abnormalities, it is generally extremely poor. We must concentrate our initial efforts on providing a comprehensive and multidisciplinary care of the delivery.

The combination of severe HPE of the alobar type, with an apparently normal facies, which is usually associated with less severe forms of HPE\(^\text{18}\), suggest the deployed of a new view of the etiology and variability of the malformation. These findings stressed the importance of the prenatal diagnosis, not only in order to classify the HPE, moreover to give a broad understanding of the variability of the clinical phenotype, complications and survival. A complete analysis of the information is necessary.

IV. Conclusion

We have described a case of severe alobar HPE accompanied by less severe facial anomalies. The main objective of the prenatal diagnosis in case of HPE, is to identify the type and spectrum, so that its particular complications and prognosis can be understood at birth. The best neuroimaging tool for identifying the characteristics of each type is MR imaging, and the information provided facilitates a multidisciplinary approach to treatment. Moreover, prenatal diagnosis allows parent to better understand the nature of their child disease and its poor prognosis, and to prepare psychologically before delivery.

The diagnostic In utero should be extended to all medical practices to provide comprehensive and timely attention, even in cases that seem bleak as this, this provides the certainty of a diagnosis and the knowledge of identify what we are facing. We need to treat patient, not diseases, taking use of the classifications but observing the particularity of each case.

References