



**A RETROSPECTIVE STUDY ON THE INCIDENCE OF OMPHALOCELE AND
GASTROSCHISIS IN NIGERIAN INFANTS USING UPTH AND BMSH AS STUDY
POPULATION**

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ABSTRACT

Introduction: Omphalocele and gastroschisis are both birth defects of the abdominal wall at which there is protrusion of the intestines and other abdominal viscera through a defect at the navel or adjacent to it. In omphalocele such protrusion is entirely covered by a sac unlike that of gastroschisis. There is paucity of information of the incidence or prevalence of omphalocele and gastroschisis in the Nigerian population. **Materials and Methods:** A retrospective study with purposive convenient sampling technique was used for the study. This study was carried out with 3,615 infants to determine the incidence of omphalocele and gastroschisis in Nigerian infants using the University of Port Harcourt Teaching Hospital (UPTH) and Braithwaite Memorial Hospital (BMH) as study population within a 3 year period (2005-2007). Results & Discussions: 6 subjects had omphalocele while 4 subjects had gastroschisis with a percentage incidence of 0.16% and 0.11% respectively, with a combined percentage incidence of 0.27% for both defects. **Conclusion:** It has been observed that despite the low incidence of omphalocele and gastroschisis in Nigerian infants, none of the affected neonates survived. There was a combined incidence of 0.27% among neonates for a period of 3 years which may eventually increase with time.

KEYWORDS: Incidence, Omphalocele, Gastroschisis, UPTH, BMSH.

INTRODUCTION

Omphalocele and gastroschisis which are abdominal wall defects are among the most frequently encountered congenital anomalies in pediatric surgery.^[1] The combined incidence of these anomalies is 1 out of 2000 (1/2,000) births, hence, a pediatric surgeon can expect to see twice as many babies with abdominal wall defects as esophageal atresia/tracheoesophageal fistula.^[1]

An Omphalocele is an abnormality that occurs before birth as a fetus is forming in its mother's uterus. Some of the abdominal organs protrude through an opening, in the abdominal muscles in the umbilical cord. A translucent membrane composed of *Amnion and Peritoneum* covers the protruding organs.^[2]

Omphalocele may be small, with only a portion of intestine protruding outside the abdominal cavity, or large, with most of the abdominal organs (including intestine, liver and spleen) present outside the abdominal cavity. The abdominal cavity itself may be small due to underdevelopment during pregnancy.^[4]

Gastroschisis also known as schistocoelia is a congenital feature of the anterior abdominal wall not involving the

site of the insertion of the umbilical cord and usually accompanied by the protrusion of the small intestine and part of the large intestine.^[5]

Gastroschisis results from a defect lateral to the median plane of the anterior abdominal wall. The linear defect permits the extrusion of the abdominal viscera without involving the umbilical cord. The viscera protrude into the amniotic cavity and are bathed by the amniotic fluid.^[6] Moore Persaud reveals that the defect usually occurs on the right side lateral to the umbilicus and is more common in males than females. The anomaly results from incomplete closure of the lateral folds during the first week of pregnancy. He further reveals that unlike other abdominal wall defects, gastroschisis is not associated with chromosome disorder.^[6]

Growth and Development of Omphalocele and Gastroschisis

During the 4th to 5th week of development, the flat embryonic disk folds in four directions and /or planes: cephalic, caudal, and right and left lateral. Each fold converges at the site of the umbilicus, thus obliterating the extraembryonic coelom. The lateral fold forms the lateral portions of the abdominal wall and the cephalic

and caudal folds make the epigastrium and hypogastrium responding to rapid growth of the intestines and liver also occur at this time.^[7]

During the 6th week of development (or eight weeks from the last menstrual period), the abdominal cavity temporarily becomes too small to accommodate all of its contents, resulting in protrusion of the intestines into the residual extraembryonic coelom at the base of the umbilical cord. This temporary herniation called physiologic midgut herniation (PMH) is sonographically evident between the 9th to 11th post-menstrual weeks. Reduction of this herniation occurs by the 12th post-menstrual week; beyond the 12th week a midgut herniation is no longer physiological.^[8]

A simple midline omphalocele develops if the extra-embryonic gut fails to return to the abdominal cavity and remains covered by the two layer amniotic-peritoneal layer into which the umbilicus inserts.^[9]

Physical Characteristics of Omphalocele

According to Suita Shong^[10] the following are the physical characteristics of omphalocele:

- i. In babies with omphalocele, the abdominal wall defect is 4-12cm, and the defect may be central, epigastric, or hypogastric.^[10]
- ii. Although the ease of accomplishing surgical reduction and repairs are correlated with the size of the abdominal wall defect, a small omphalocele is no guarantee of an uncomplicated clinical course. Associated genetic syndromes involving multiple organ systems, or abnormalities of the intestines, such the association of ileal atresia and a patent omphalomesenteric duct, are potential problems.^[10]
- iii. With a large omphalocele, dystocia may occur and result in injury to the baby's liver; hence, cesarean delivery may be indicated.
- iv. The omphalocele sac is usually intact, though it may be ruptured in 10-20% of cases. Rupture may occur in-utero or during or after delivery.^[10]
- v. Babies with the beckwith-wiedeman syndrome (that is exomphalos, macroglossia, gigantism); have large, rounded facial features, hypoglycemia from hyperplasia of the pancreatic islet cells, and visceromegaly. They may have genitourinary abnormalities and they are at risk for wilms tumours, liver (hepatoblastoma), and adreno cortical neoplasms.^[10]
- vi. Peutalogy of Cantrell describes an epigastric omphalocele associated with a cleft sternum and anterior diaphragmatic hernia, cardiac defects (such as ectopia cordis, ventricular septal defect (VSD) and an absent pericardium.^[10]
- vii. Babies with giant omphalocele have large, central abdominal wall defects. The liver is entirely contained in the omphalocele sac. The abdominal and thoracic cavities are small and underdeveloped. Restrictive lung disease and pulmonary hypoplasia

usually are associated with the diminutive thoracic cavity operative closure.^[10]



Figure 1: The physical characteristics of Omphalocele.^[9]

Physical Characteristics of Gastroschisis

- i. The defect is fairly uniform in size (< 5 vertical opening) and location (to the right of the umbilical cord).
- ii. The amount of inflammation, edema and turgor of the intestines, as well as the size of the abdominal cavity, determines whether reduction of the extruded intestines and closure of the abdominal wall can be accomplished. Inflammation may so distort the appearance of the bowel that it becomes difficult to determine if associated intestinal atresia is present.
- iii. Closure of the abdominal wall defect when the intestines are inflamed requires their temporary placement in a silo to allow the inflammation to resolve. As the intestine softens and becomes pliable, reduction can be accomplished. Correction of the associated intestinal atresia is best delayed until several weeks after the initial repair.
- iv. Intestinal dysfunction takes 4-6 weeks to several months to normalize.
- v. If gastroschisis is defined antenatally, serial sonography is indicated to assess intestinal integrity and amniocentesis to monitor lung maturity.



Figure 2: The physical characteristics of gastroschisis. ^[9]

Causes of Omphalocele and Gastroschisis

Causes of omphalocele and gastroschisis are not really known. However, Steps that usually take place in the development of the abdominal organs and muscles did not occur properly.^[11]

Omphalocele and gastroschisis are not caused by anything the mother went through during the pregnancy. According to James Glasser ^[1], the following can be considered as the causative factors of omphalocele and gastroschisis:

- Factors associated with high-risk pregnancies, such as maternal illness and infection, drug-use, smoking and genetic abnormalities, can be associated with birth of babies with omphalocele or gastroschisis. These factors contribute to placental insufficiency and birth of premature or small-for-gestational-age (SGA) babies, in whom gastroschisis and omphalocele are most common.
- Folic acid deficiency, hypoxia and salicylates have caused laboratory rats to develop abdominal wall defects, but the significance of these experiments is conjectural. Elevation of maternal serum alpha-fetoprotein (MSAFP) levels certainly warrants investigation with high resolution sonography to determine if any structural abnormalities are associated with an omphalocele, amniocentesis is indicated to check for an associated genetic abnormality.^[1]
- Polyhydramnios suggest fetal intestinal atresia, and this possibility should be investigated with ultrasonography. Such information ideally prompts referral to a tertiary care facility where the infant can receive expedition's specialty care.

Risk for Developing an Omphalocele

When an omphalocele is isolated (no other defects are present), the risk for it to happen in future pregnancies is 1% (one percent).^[11] There are some families that have been reported to have an omphalocele inherited as an autosomal dominant or x-linked recessive trait. In these cases, the chance for re-occurrence would be higher.^[11] Many babies born with an omphalocele also have other

abnormalities. The chance for re-occurrence depends upon the underlying disorders.^[11]

Diagnosis of Omphalocele and Gastroschisis

Omphalocele and gastroschisis can often be detected on fetal ultrasound in the second and third trimesters of pregnancy. A fetal echocardiogram (ultrasound of the heart) may also be done to check for heart abnormalities before the baby is born.^[12]

Studies carried out on the incidences of omphalocele and gastroschisis have shown that these abdominal wall defects have contributed to the increasing death of the neonates.^[13-20]

There is paucity of information of the incidence or prevalence of omphalocele and gastroschisis in Nigerian population.

Hence, this study was carried out to determine the incidence of omphalocele and gastroschisis in Nigerian Infants using the University of Port Harcourt Teaching Hospital (UPTH) and Braithwaite Memorial Hospital (BMH) as study population of 2005-2007: To know the survival rate of affected infants, the incidences of omphalocele and gastroschisis in Nigerian infants and determining which gender is more commonly involved in omphalocele and gastroschisis.

Ethical Clearance: Ethical clearance was obtained from the ethics committee of the University of Port Harcourt, Nigeria before commencement of the study.

MATERIALS AND METHODOLOGY

Study Design: A retrospective study with purposive convenient sampling technique. The total sample size was 3,615 infants (subjects). The materials used for this study were provided by the Special Care Baby Unit (SCBU) of University of Port Harcourt Teaching Hospital (UPTH) and Braithwaite Memorial Hospital (BMH). They included the following: Register of births, Hospital folders of babies and Foolschap.

Data Collection: The records of infants with omphalocele and gastroschisis in UPTH and BMH for a period of three years (2005-2007) were collected and tabulated. The incidences of each were calculated per year. The frequency of these abdominal wall defects per year was used to calculate the percentage incidence in a particular year using the formula below:

$$\frac{\text{frequency of omphalocele or gastroschisis}}{\text{Total births}} \times \frac{100}{1}$$

Ethical Clearance: Ethical clearance was obtained from the ethics committee of the University of Port Harcourt, Nigeria before commencement of the study.

RESULTS

The result of the data obtained was analyzed using tables.

A total of 6 omphalocele and 4 gastroschisis using the population study of University of Port Harcourt Teaching Hospital (UPTH) and Braithwaite Memorial Hospital (BMH) were obtained for the period of 3 years. A

combined total incidence of 0.27% was observed for the both defects. Of the two body wall defects, omphalocele was observed to be the most commonly occurring defect with the incidence of 0.16%. While gastroschisis had an incidence of 0.11% for the 3 year period of study.

Table 1: Omphalocele distribution in years.

YEAR	2005	2006	2007
Age	3hrs- 1day	3hrs	1 day
Total Birth	1,246	1,374	995
Sex	F ₁ and M ₁	F ₁	M ₃
Frequency	2	1	3
Remark	Died	Died	Died
Percentage Incidence	0.16%	0.07%	0.30%

Table 2: Gastroschisis distribution in years.

Year	2005	2006	2007
Age	4hrs-1day	3hrs	-
Total Birth	1,246	1,374	-
Sex	F ₁ and M ₁	M ₁	-
Frequency	2	1	-
Remark	Died	Died	-
Percentage Incidence	0.24%	0.07%	-

Table 3: Combined distribution of gastroschisis and omphalocele in the three year period (2005-2007).

Year	Total birth	Frequency of Omphalocele	Frequency of Gastroschisis	Incidence
2005	1,246	2	3	0.40%
2006	1,374	1	1	0.14%
2007	995	3	-	0.30%
Total	3,615	6	4	0.27%

In 2005, omphalocele accounted for the percentage incidence of 0.16% while gastroschisis was 0.14%.

In 2006, omphalocele accounted for 0.07% while gastroschisis accounted for 0.07% as well.

However, in 2007, the percentage incidence of omphalocele amounted to 0.30% while gastroschisis had a 0.00%.

DISCUSSION

The incidence of omphalocele and gastroschisis in UPTH and BMH from this study was calculated to be 0.16% for omphalocele and 0.11% for gastroschisis and a combined percentage incidence of 0.27% for the 3 period. These values are low when compared to the increasing figures of 0.48% to 3.16% in gastroschisis from 1980-1993 in 10,000 births in western Australia and 42% out of 127 cases of omphalocele and 57% out of 121 cases of gastroschisis in south Florida, United States of America.^[9,13,14,20]

This research confirms that omphalocele and gastroschisis are not so common in Nigeria with respect to the study population within the 3 years period which accounted for the low combined percentage incidence of 0.27% and agrees with the findings of other authors.^[13-20] However, the differences in the study carried out in UPTH and BMH (Nigeria) and that carried out in other countries of the world could be due to environmental factors.

CONCLUSION

It has been observed that despite the low incidence of omphalocele and gastroschisis in Nigerian infants, none

of the affected neonates survived. There was a combined incidence of 0.27% among neonates for a period of 3 years which may eventually increase with time. We suggest the use of ultrasonography machine for diagnosis to enhance early diagnosis and treatment of these (neonates) patients.

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AUTHOR'S CONTRIBUTIONS

We write to state that all authors have contributed significantly, and that all authors are in agreement with the content of the manuscript. 'Author A' (Paul, Chikwuogwo Wokpeogu) designed the study design, protocol, the write-up and intellectual content. 'Authors B' (Edibamode Ezon-Ebido I.) reviewed the design, protocol, the write-up and managed the literature searches, 'Author C' (Paul, John Nwolim) managed the analyses of the study, wrote the first draft of the manuscript.

CONFLICT OF INTEREST

We write to state that there is no conflict of interest.

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