

Outcome of Fetuses with Anterior Abdominal Wall Defects in A Tertiary Referral Hospital

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Abstract

Introduction: The most common abdominal wall defects are gastroschisis and omphalocele. Gastroschisis is a case of intraabdominal herniation caused by an abdominal wall defect from exposure to amniotic fluid during pregnancy. Omphalocele is a case of intraabdominal herniation covered with a membranous sac on the umbilical cord's base. Gastroschisis occurred in 1/4000 of the of the global birth rate. Prevalence of omphalocele in between 1/3000 and 1/5000 cases of pregnancy. The purpose of the purpose of the research is to present an overview of patients with congenital defects such as Gastroschisis and Omphalocele.

Method: Research design is observational and descriptive. Data obtained from medical records in Hasan Sadikin tertiary referral hospital Bandung. Sample size was obtained by total sampling and conducted in April 2020–April 2023.

Results: The demographics of gastroschisis include male (50%), female (50%), preterm (20%), stillbirth (30%), severe asphyxia (14.29%), moderate asphyxia (57.14%), normal asphyxia (8.57%), newborn mortality (14.29%), and other congenital anomalies (40%). In comparison, the demographics of omphalocele are male (66.67%), female (33.33%), preterm (58.33%), stillbirth (16.67%), severe asphyxia (40%), moderate asphyxia (40%), normal asphyxia (20%), newborn mortality (50%), and other congenital abnormalities (25%). Abdominal wall defects are seldom related with gender.

Conclusion: Abdominal wall defect is a very rare congenital abnormality. This abnormality will require primary abdomen closure surgery to enhance the baby's prognosis. The more other risk factors exist within abdominal wall defect babies, the worse their following prognosis will be. The prognosis for omphalocele is more severe than gastroschisis due to the presence of asphyxia and prematurity.

Keywords: Abdominal wall defect, Gastroschisis, Omphalocele

Luaran Janin dengan Defek Dinding Abdominal Anterior di Rumah Sakit Rujukan Tersier

Abstrak

Pendahuluan: Cacat dinding depan abdomen yang paling banyak terjadi adalah gastroschisis dan omphalocele. Gastroschisis adalah kasus herniasi intraabdomen yang disebabkan oleh cacat dinding perut akibat paparan cairan ketuban selama kehamilan. Omphalocele adalah kasus herniasi intraabdomen yang ditutupi kantung membran di dasar tali pusat. Gastroschisis terjadi pada 1/4000 angka kelahiran global. Prevalensi omfalokel antara 1/3000 dan 1/5000 kasus kehamilan. Tujuan dari penelitian ini adalah untuk menyajikan gambaran pasien dengan kelainan bawaan seperti Gastroschisis dan Omphalocele.

Metode: Desain penelitian adalah observasional dan deskriptif. Data diperoleh dari rekam medis di rumah sakit rujukan tersier Rumah Sakit Hasan Sadikin Bandung. Besar sampel diperoleh dengan cara total sampling dan dilakukan pada bulan April 2020 – April 2023.

Hasil: Demografi gastroschisis meliputi laki-laki (50%), perempuan (50%), prematur (20%), lahir mati (30%), asfiksia berat (14,29%), asfiksia sedang (57,14%), asfiksia normal (8,57%), kematian bayi baru lahir (14,29%), dan kelainan kongenital lainnya (40%). Sebagai perbandingan, demografi omfalokel adalah laki-laki (66,67%), perempuan (33,33%), prematur (58,33%), lahir mati (16,67%), asfiksia berat (40%), asfiksia sedang (40%), asfiksia normal (20). %, kematian bayi baru lahir (50%), dan kelainan bawaan lainnya (25%). Cacat dinding perut jarang berhubungan dengan jenis kelamin.

Kesimpulan: Cacat dinding perut merupakan kelainan bawaan yang sangat tidak biasa. Kelainan ini memerlukan operasi penutupan perut utama untuk meningkatkan peluang hidup bayi. Adanya faktor risiko tambahan pada bayi baru lahir dengan kelainan dinding perut memperburuk prognosinya. Omphalocele sering menyebabkan asfiksia dan prematur, sehingga prognosinya lebih buruk dibandingkan gastroschisis.

Kata Kunci : Cacat dinding depan abdomen, Gastroschisis, Omphalocele

Introduction

A congenital abdominal wall defect is one of the most common congenital defects in humans, with an incidence of 1 out of every 2000 live births. The most common abdominal wall defect conditions found are gastroschisis and omphalocele. These two abnormalities have different characteristics and outcomes. ¹ Gastroschisis is a congenital abdominal wall defect presented with herniation of intraabdominal organs that were exposed to amniotic fluid during pregnancy. Such a condition is the most commonly found congenital defect in pediatric surgery cases, with a prevalence of 4.9 per 10,000 live births. ^{2,3} Prenatal diagnosis provides an opportunity for clinicians to plan labor and perform fetal monitoring strictly. Nowadays, the survival rate of neonates with gastroschisis is more than 90%. ³ However, this condition is related to significantly high morbidities, including intestinal dysfunction, sepsis, and repeated operations, resulting in extended inpatient duration in the hospital along with parenteral nutrition. ^{3,4}

Omphalocele, also called exomphalos, is a congenital malformation as the consequence of an anterior abdominal wall closure defect. ⁵ Omphalocele is usually detected during the prenatal period. This condition was estimated to happen in 1 of 4000 live births and may be related to genetic and structural abnormalities including trisomy 13, 18, and 21; Beckwith-Wiedemann syndrome; and abnormalities of the heart, renal, gastrointestinal, extremities, or central nervous system. ⁶ Newborns with omphalocele are mostly males born by women aged above 35 years old or below 20 years old. Babies with omphalocele are commonly associated with other congenital abnormalities, such as heart abnormalities (32%). ⁷ The survival rate of isolated omphalocele cases (without comorbidities or chromosomal abnormalities) is 96%, and it decreases drastically when the

case is related to other anomalies or abnormal karyotypes. Death (28.7%) usually occurs within the first 28 days of life. ^{7,8}

Neonates with omphalocele in particular have higher rates of death compared to neonates with gastroschisis because of their higher prematurity and asphyxia moment. ⁹ The difference in mortality rate was expected to be related to the high incidence of genetic and heart abnormalities in neonates with omphalocele. Gastroschisis and omphalocele have worse clinical outcomes compared to normal neonates. A multidisciplinary approach is needed in the management of the patient. To avoid complications, adequate support is needed in terms of nutrition, ventilator usage, and surgical or non-surgical care. This may cause an extended in-hospital stay. Furthermore, congenital abnormalities occur simultaneously and also affect the survival rate of babies with gastroschisis and omphalocele. ⁹

Studies about the outcomes of babies with gastroschisis and omphalocele in Indonesia are still limited. For that, the researcher was interested in performing a study about the outcomes of babies with gastroschisis and omphalocele at Dr. Hasan Sadikin Central General Hospital, Bandung, which is a tertiary referral hospital.

Methods

This study collected patients's data on pregnancy with gastroschisis and omphalocele through prenatal USG treated by the Obstetrics and Gynecology Department of Dr. Hasan Sadikin Central General Hospital Bandung and newborns with gastroschisis and omphalocele between April 2020 and April 2023. The study is a descriptive study; the variables include maternal age, gender, history of previous pregnancy, history of genetic abnormalities, intrauterine fetal death condition during pregnancy, labor method within pregnancy with gastroschisis and

omphalocele, gestation age during labor, newborn's perinatal condition, surgical method for the baby, the newborn's condition after surgery, and other accompanying congenital abnormalities. Samples included in this study were counted using the total sampling method.

Results

The study conducted at Dr. Hasan Sadikin Central General Hospital fulfilled the criteria, with 10 individuals suffering from gastroschisis and 12 individuals suffering from omphalocele. The outcomes of babies with gastroschisis or omphalocele that were collected included accompanying congenital abnormalities, small for gestational age, stillbirth, length of the newborn's survival, and presence of worsening in surviving babies.

In Table 1.1, omphalocele was mostly found in male babies, while gastroschisis's occurrence was equal between male and female babies. The table also shows that babies with omphalocele were mostly born

at <37 weeks of gestational age compared to babies with gastroschisis. Labor occurs before 37 weeks in 20% of gastroschisis cases, and premature birth is found in 58.33% of omphalocele cases.

The study revealed a higher proportion of babies with gastroschisis (40%) than those with omphalocele (16.67%) who were small for their gestational age. The majority of stillbirth babies with gastroschisis (30%), compared to omphalocele (16.67%), were born prematurely. In babies with gastroschisis, the APGAR scores of 1-3, 4-6, and 7-10 were respectively 14.29%, 57.14%, and 28.57%. The APGAR scores for babies with omphalocele were 40%, 40%, and 20%, respectively. In babies with gastroschisis, 40% of them had other congenital abnormalities, whereas 25% had omphalocele as well. The accompanying congenital abnormalities within the collected samples included webbed neck, ambiguous genitalia, congenital talipes equinovarus, encephalocele, meningocele, nasal malformation, atresia ani, cyclopia, umbilical cyst, hypoplasia cerebellum, megacisterna magna, brachiencephaly,

Table 1 Presents Gender and Gestational Age in Babies with Gastroschisis and Omphalocele.

Variable		Gastroschisis N (%)	Omphalocele N (%)
Gender	Male	5 (50%)	8 (66.67%)
	Female	5 (50%)	4 (33.33%)
Gestational age	<37 weeks	2 (20%)	7 (58.33%)
	>37 weeks	8 (80%)	5 (41.67%)

Table 2 The Outcomes of Babies with Gastroschisis and Omphalocele

Variable	Score	Gastroschisis N (%)	Omphalocele N (%)
Small for Gestational Age		4 (40%)	2 (16.67%)
Stillbirth		3 (30%)	2 (16.67%)
APGAR	0-3	1 (14.29%)	4 (40%)
	4-6	4 (57.14%)	4 (40%)
	7-10	2 (28.57%)	2 (20%)
Other congenital abnormalities		4 (40%)	3 (25%)

Table 3 The Presentation of Neonatal Mortality after Surgery for Babies with Gastroschisis and Omphalocele

Variable	Gastroschisis N (%)	Omphalocele N (%)
Surviving baby pre-surgery	5 (71.42%)	6 (60%)
Surgery	5 (100%)	5 (83.33%)
Neonatal death post-surgery	3 (60%)	3 (50%)

abnormal facial profile, ascites, and scoliosis.

This study resulted in surgical actions was done in 100% gastroschisis babies of surviving babies (71.42%) and 83.33% omphalocele babies of surviving babies (60%). After surgery, 60 percent of infants with gastroschisis and 50 percent of infants with omphalocele experienced neonatal mortality.

Discussion

According to this study, the majority of cases of omphalocele were observed in male babies, while gastroschisis was equally observed in both genders. The incidence of omphalocele and gastroschisis was mostly found in males compared to females.¹⁰ A previous study by Jwa E. et al. found that the incidence of gastroschisis and omphalocele were more common in females compared to males which was 60.5% for gastroschisis and 58.3% for omphalocele with a ratio between males and females around 1:1.5 in gastroschisis and 1: 4 in omphalocele, but the result was not significant. Other studies related to the relationship between gender and the incidence of gastroschisis and omphalocele were diverse without any significant results.¹¹ Gastroschisis nor omphalocele were rarely related to gender and there is no linked incidence rate between these abnormalities and gender.^{12,13}

A study by Soares et al. found 36.84% of premature births within babies with gastroschisis while Mayer et al. found 23% within babies with omphalocele.^{14,15} The other study done by Hwang P. found premature

births in 57% of gastroschisis babies and 42% of omphalocele babies. The spontaneous gestational termination rate reached 50% in omphaloceles with chromosomal anomalies and 30% for isolated omphaloceles, while the rate was around 30-40% in pregnancies with gastroschisis.¹⁶ Premature births of babies with gastroschisis and omphalocele may be caused by several factors as fetal emergency, intrauterine fetal death, poor fetal prognosis, premature rupture of the membranes, or premature contraction. Premature birth of babies with gastroschisis and omphalocele was usually a result of pregnancy rather than abnormalities in the fetus.^{13,14,16} Small for gestational age in babies with

gastroschisis and omphalocele was 76.9% in a study conducted in San Diego.¹⁷ Another study by Hidaka et al. presented 36.4% of babies with gastroschisis and 48.1% of babies with omphalocele that was small for their gestational age. This incidence was not yet to be surely explained, but the presence of maternal accompanying diseases such as preeclampsia and malnutrition might increase the risk of small for gestational age as the outcomes of babies with gastroschisis or omphalocele.¹⁷ The presence of other congenital malformations and chromosomal abnormalities of the baby also increases the risk of small for gestational age.¹⁸

A study conducted by Sparks et al. resulted in a 4.8% incidence of gastroschisis. In a study conducted in China, the percentage of stillbirths among babies with an omphalocele was 37.4%. The occurrence of stillbirth in babies with gastroschisis or omphalocele is related to other factors associated with

these abnormalities such as liver herniation, malformations, chromosome abnormalities, and genetic syndromes. The possibilities of risk factors affecting the incidence of stillbirth cannot be confirmed when the fetus is still in the womb so additional examinations are needed after labor, although it is still a constraint due to the family's refusal for autopsy in stillbirth babies.²⁰

Babies that survived through the primary abdomen closure surgery to close the abdominal wall's defect were 100% in babies with gastroschisis and 83.33% in babies with omphalocele. Neonatal death occurred in 60% of the babies with gastroschisis and 50% with omphalocele that was operated with primary abdomen closure. This may be caused by the disturbance of the gastrointestinal tract such as obstruction and peristaltic disruption. The most frequent causes of neonatal deaths, particularly in premature babies, were sepsis and respiratory failure in babies.¹⁷

The study conducted by Benjamin et al. found congenital abnormalities accompanying gastroschisis and omphalocele as much as 32% and 80% respectively. Another study by Oluwafemi et al. stated that 26% of gastroschisis and 56% of omphalocele can be accompanied by other congenital abnormalities.¹⁷ The presence of malformation abnormalities, neonatal chromosomal abnormalities, premature birth with gastroschisis and omphalocele increases the risk of mortality in neonatal or stillbirths higher compared to normal neonates depending on how bad the abnormalities are. Gastroschisis and omphalocele are commonly associated with abnormalities in the cardiovascular, gastrointestinal, and urogenital systems.²¹

Conclusion

Abdominal wall defect is a very rare congenital abnormality. This abnormality will require primary abdomen closure surgery to enhance

the baby's prognosis. The more other risk factors exist within abdominal wall defect babies, the worse their following prognosis will be. The prognosis for omphalocele is more severe than gastroschisis due to the presence of asphyxia and prematurity.

Acknowledgement

Great gratitude to the Obstetrics & Gynecology Department, Hasan Sadikin general Hospital, Padjadjaran University

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