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ORIGINAL ARTICLE

MAJOR ABDOMINAL WALL DEFECTS AND OUTCOME OF MANAGEMENT AT A REFERRAL HOSPITAL.

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ABSTRACT

Background*: Gastroschisis and omphalocele are abdominal wall defects that were first described as early as the 16th century. However, it was not until recently that these two conditions were categorized as separate entities. The aim of the study was to determine the occurrence and outcomes of major abdominal wall defects managed at a referral hospital.*
Materials and Methods*: A retrospective review of omphalocele and gastroschisis managed over 4 years at Tikur Anbessa Teaching Hospital, Addis Ababa, Ethiopia, from 1 September 2010 to 31 August 2014.*
Results: *A total of 39 neonates were seen, 24(61.5%) of whom were male (male: female ratio of 1.6:1). Among these, 23 (59%) presented with isolated omphalocele, 9(23.1%) with isolated gastroschisis, 6(15.4%) with omphalocele and another anomaly and 1 had gastroschisis with another anomaly. A total of 13 deaths were recorded, with overall fatality rate of 33%. Neonates with omphalocele had significantly better prognosis (20% fatality rate) than neonates with gastroschisis (70% fatality rate). Preterm cases with abdominal wall defect had 50% fatality in contrast to term neonates for whom fatality rate was 31.3%. The observed fatality rate for low birth weight neonates with abdominal wall defect was 56.3% in contrast to 18.2% for normal birth weight neonates. Preterm birth (prematurity), low birth weight and time elapsed before arrival to the tertiary center were significantly associated with poor outcome.*
Conclusion: *Overall, major abdominal wall defects have high fatality and gastroschisis has a significantly worse outcome than omphalocele.*
Key word*: Abdominal wall defects, gastroschisis, omphalocele, outcomes*

INTRODUCTION

Abdominal wall defects are among the commonest structural developmental defects in man and remain a source of significant morbidity and mortality despite the advances in pediatric surgical care (1-4). Gastroschisis and omphalocele are abdominal wall defects that were first described as early as the 16th century (5). The two conditions were considered identical until Moore and Stokes categorized them into two separate entities in 1953 (6). Their unique pathogenesis and clinical presentations were further emphasized by Duhamel in 1963 (7).

Gastroschisis refers to a full-thickness defect in the paraumbilical area usually just to the right of a normal insertion of the umbilical cord into the body wall. A variable amount of intestine and rarely parts of other abdominal organs are eviscerated outside with no covering membrane (8). An omphalocele is a midline abdominal wall defect of different sizes, with the herniated organs covered by a membrane consisting of peritoneum on the internal surface, amnion on the external surface, and Wharton's jelly between these two layers. The umbilical vessels insert into the membrane and not the body wall (8).

Not much is known about the cause of anterior abdominal wall defects. But early interruption of the fetal omphalomesenteric arterial blood supply is considered as the probable cause for gastroschisis (9, 10). The primary defect is likely to occur during early embryogenesis, associated with major abnormalities and is often with aneuploidy and familial occurrence has been documented (11).

Currently it is thought that omphalocele is a more common condition than gastroschisis at least in the western world, occurring in 1 per 4000 live births compared to 1 per 6000 for gastroschisis (12). Omphaloceles are frequently associated with chromosome abnormalities and other malformations, whereas gastroschisis tends to be an isolated anomaly (13). For this reason, the long-term prognosis for infants with gastroschisis is significantly

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better than that for infants with omphalocele, in whom a 50% to 60% survival rate and frequently chronic medical problems are seen (13).

PATIENTS AND METHODS

A retrospective review was conducted from patient records on all neonates presenting with major abdominal wall defects and managed at pediatric surgical unit, Tikur Anbesa University Referral Hospital in Addis Ababa between1September 2010 to 31 August 2014. Omphalocele was defined as midline abdominal wall defect of variable size, with the herniated viscera covered by a membrane. Gastroschisis was defined as a full-thickness defect in the abdominal wall usually just to the right of a normal insertion of the umbilical cord in the absence of covering membrane.

Data were collected from their clinical records on a pretested format that included demographic information, gestational age and weight at birth, age and parity of the mother, age of the neonate in hours at presentation, principal diagnosis, treatment given and outcome. The collected data were entered, cleaned and analyzed using SPSS version 16 statistical software.

Descriptive analysis with frequencies, proportions and summary statistics was computed for the variables. Both bivariate and multivariate logistic regressions were used to assess the association between dependent and independent variables. Factors associated with outcome at bivariate analysis were identified and the variables with p-value of less than 0.20 were taken to multivariable analysis and the model was built with backward elimination (backward LR). P-values of 0.05 and less were considered statistically significant.

RESULTS

A total of 39 neonates, 24 male and 15 female with a male: female ratio of 1.6:1 were seen during the four years of study. The cases originated mainly from Addis Ababa (53.8%) and Oromia (28.2%) regions (Table 1).

Among the total cases seen in the study period 23 (59%) presented with isolated omphaloceles, 9 (23.1%) had isolated gastroschisis, 6(15.4%) had omphalocele with other anomalies and 1(2.6%) presented with gastroschisis with other anomalies. Structural anomalies associated with omphalocele were ventricular septal defect (2 cases), hypospadias (1 case), inguinal hernia (1 case), undescended testis (1 case) and imperforated anus (1 case). Only one case presented with gastroschisis and jejunal atresia (Table 1).

Table 1. Pattern of presentation of neonatal abdominal wall defects by age, sex and residence, Tikur Anbessa Hospital, Addis Ababa, 2010-2014.

		Principal diagnosis				
		Omphalocele	Gastroschisis	Omphalocele with another anomaly	Gastroschisis with another anomaly	Total
Sex	Male	15	5	4	0	24
	Female	8	4	2	1	15
Total		23	9	6	1	39
Residence	Addis Ababa	14	4	3	0	21
	Oromia	5	3	2	1	11
	Others	4	2	1	0	7
Total		23	9	6	1	39

The highest maternal age-specific prevalence of both gastroschisis and omphalocele was in the 25–29 year age group. 34.8% of cases with omphalocele were born to para I mothers, 30.4% to para II mothers, 26.1% to para III mothers and 8.7% to para IV mothers. Unlike that of omphaloceles, 88.8% of cases with gastroschisis were born to para I mothers.

Among all cases of gastroschisis, 90% were treated by single closure and 10% by staged operation with the initial repair done by local silo bags (intravenous solution bags). Among cases of omphalocele, 55.2% were initially managed by conservative treatment by GV painting, 37.9 % by single closure and the remaining 6.9% were treated by staged operation. Among all cases treated for abdominal wall defect, the fatality rate was 43.3% for those conservatively treated, 25.0% for those treated by single closure and 33.3% for staged operation (Table 2).

A total of 13 deaths were recorded, giving a fatality rate of 33.3%. Neonates with omphalocele had better prognosis than neonates with gastroschisis. Fatality rates for gastroschisis and omphalocele were 70% and 20% respectively. There was no statistically significant difference of fatality by sex (37.5% among male and 26.6% among female neonates).

The neonates who did not survive were cases who were referred to the hospital relatively late with compromised clinical conditions. Among those who came in the first 12 hours, only 28.6% died compared to 33.3% among those who came within 13 to 24 hours and 50% among those who presented later than 48 hours as shown in Figure 1.

Table 2. Outcome of treatment of major abdominal wall defects managed at Tikur Anbessa Hospital, Addis Ababa, 2010-2014 (n=39)

Principal diagnosis	Treatment given						Total
	conservative		single closure		staged operation		
	improved	died	improved	died	improved	died	
Omphalocele	10	3	7	2	1	0	23
Gastroschisis	0	0	2	6	0	1	9
Omphalocele with another anomaly	2	1	2	0	1	0	6
gastroschisis with another anomaly	0	0	1	0	0	0	1
Total	12	4	12	8	2	1	39

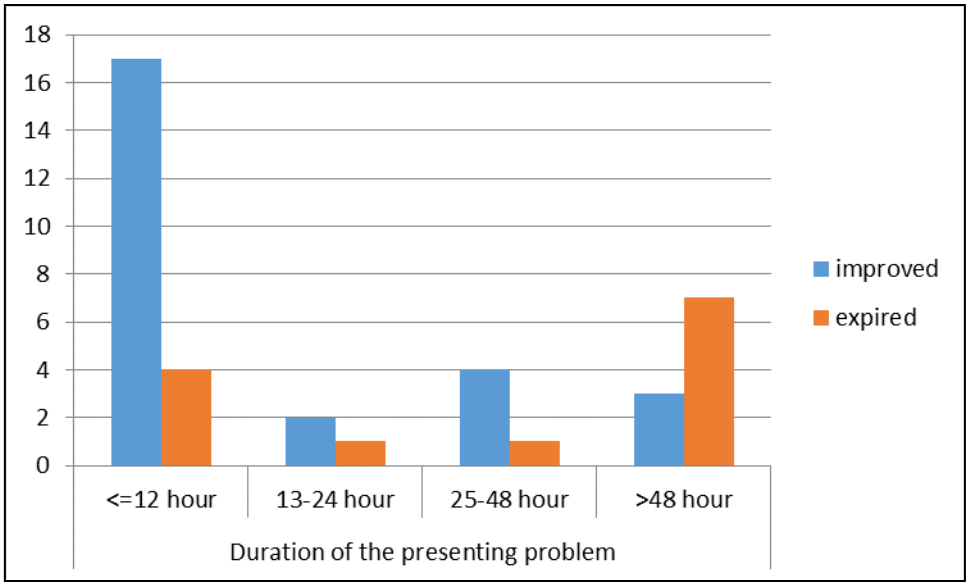

Fig 1. Distribution of treatment outcome by duration of the presenting problem before hospital visit, Tikur Anbessa Specialized Hospital, 2010-2014 (n=39)

With regard to gestational age, among the total cases of abdominal wall defect 15.1% were preterm, 82.1% term and 2.6% post term. 17% of omphaloceles and 11.1% of gastroschisis were associated with prematurity. Preterm neonates with abdominal wall defect experienced 50% fatality in contrast to term neonates with abdominal wall defect for whom fatality was 31.3%.

Low birth weight with abdominal wall defect was associated with 56.3% fatality whereas normal birth weight with abdominal wall defect was associated with only 18.2% fatality. In this study, factors that are significantly associated with outcome in the bivariate analysis at level of P value 0.2 and less were taken into the multivariate logistic regression analysis. Variables such as gestational age, weight of the neonate, principal diagnosis, duration of the presenting problem and treatment given were exported into multivariate logistic regression analysis using backward elimination and it was found that gestational age at birth and duration of the presenting problem remained in the condensed model.

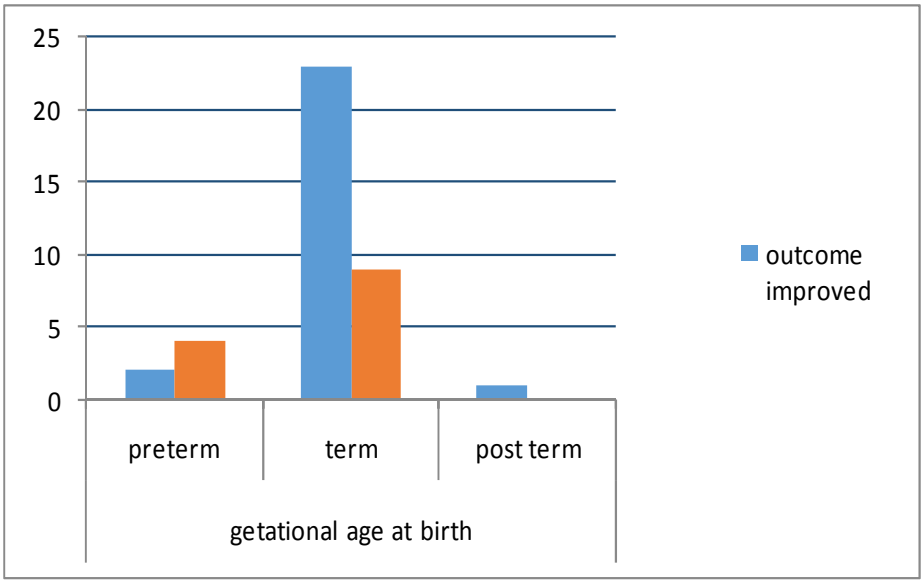

Fig 2. Distribution of outcome of treatment for abdominal wall defect by gestational age of the neonate at birth, Tikur Anbesa Hospital, Addis Ababa; 2010-2014 (n=39)

In order to check the model fitness, Hosmer-Lemshow goodness of fit was applied (P>0.05) and case wise listing of residuals was used to check outliers. Multivariate logistic regression analysis revealed that gestational age at birth and duration of the presenting problem were statistically significant predictors of outcome. There was a statistically significant association between outcome and gestational age at birth. Preterm neonates had higher fatality rate than term neonates. [Adjusted -OR= 124.1; 95 % CI, (3.27, 4.70)].

Similarly, there was a statistically significant association between outcome and duration of the presenting problem. Neonates who arrived later than 48 hours to the hospital had a higher fatality rate compared to neonates who arrived to the hospital within 12 hours [Adjusted-OR= 68.39; 95% CI, (3.84,1.21)] (Table 3).

Table 3. Correlates of outcome of treatment for major abdominal wall defects in neonates, Tikur Anbessa specialized Hospital, 2010-2014 (n=39)

Variable	Category	Outcome		COR (95%C.I.)	AOR (95%C.I.)	P value AOR
		improved n=26	expired n=13			
Duration of the presenting problem	≤12 hour	16	5	1.00	1.00	
	13-24 hour	2	1	1.60(0.11-21.58)	**68.39(3.84 -1.21)****	**0.004**
	25-48 hour	4	1	0.80(0.07-8.91)	3.93(0.17-87.36)	0.38
	>48 hour	4	6	**4.80(0.95-24.4)***	0.71(0.001-4.50)	0.21
Weight of the neonate at birth	Low birth Weight	7	9	1.00	1.00	
	Normal birth Weight	18	4	**0.17(0.04-0.74)***	1.94(0.02-190.0)	0.77
	Macrosomia	1	0	0.00(0.00-0.00)	0.00(0.00-0.00)	1.00
Gestational age at birth	Preterm	2	4	**5.33(0.83-34.34)***	1.00	
	term	24	9	1.00	**124.1(3.27- 4.70)****	**0.009**
Principal diagnosis	Omphalocele	17	6	1.00	1.00	
	Gastroschisis	4	5	**3.54(0.71-17.73)***	8.44(0.44 -161.9)	0.15
	Omphalocele with other anomalies	4	2	1.41(0.20-9.81)	1.11(0.72-17.14)	0.93
	Gastroschisis with other anomalies	1	0	0.00(0.0.00-0.00)	0.001(0.001-0.001)	1.00
Treatment given	Conservative	13	3	1.00	1.00	
	single closure	13	10	0.300(0.67-1.34)	0.096(0.008-1.14)	0.063

*Statistically significant at P< 0.2
**Statistically significant at P< 0.05

DISCUSSION

In this study, the ratio of omphalocele to gastroschisis was 29 to 10 or 3:1, compared to a study done in Tampa, south Florida, where the numbers of cases of omphalocele and gastroschisis were similar i.e. 1:1 ratio (14). A study done in South Africa also shows that the incidence of gastroschisis has increased in relation to omphalocele (15). This may be due to poor recording by less experienced staff.

The percentage of associated anomalies was 26% for omphaloceles versus 10% for gastroschisis unlike in the study done in Tampa, south Florida which revealed a proportion of associated anomalies as high as 73% for omphalocele and 23% for gastroschisis (14). The lower rate of associated anomalies in this study may be explained by a low detection rate because of limited diagnostic facilities.

In this study, the most affected mothers in both gastroschisis and omphalocele were para I mothers aged 25 to 29 years. But unlike in the omphalocele group, there were no mothers of 40 years or more in the gastroschisis group. Boyd et al (16) also found younger mothers in their gastroschisis group. Torfs et al found the incidence of gastroschisis to be highest among mothers under 20 years old, with the incidence declining steeply with increasing maternal age; only 7% of mothers were over 29 years of age in their group.

The reason for the higher occurrence in younger maternal age is unknown (17). The overall fatality rate for major abdominal wall defect in the four years of study was 33.3%. Fatality rate for the gastroschisis group was significantly higher than for the omphalocele group. The fatality rates for omphalocele and gastroschisis were 20% and 70% respectively. In earlier studies, the outcome of gastroschisis was by far better than of omphalocele and it had a good outcome in more than 90% of cases (18). In contrast to this study, the fatality rate for omphalocele and gastroschisis in Tampa area was 22% and 9% respectively. The possible reason for the higher fatality in this study is late referral and arrival of the neonate to hospital after significant clinical deterioration with severe dehydration and sepsis because of the absence of covering membrane in gastroschisis. In this study, preterm (prematurity), late arrival, and low birth weight were significantly associated with poor outcome for both omphalocele and gastroschisis.

Conclusions: In conclusion, this study showed that the number of cases of omphalocele and gastroschisis seen at Tikur Anbesa Teaching Hospital over a 4-year period was 39 with a 3:1 ratio. Para I mothers aged 25-29 years were more frequently affected by both gastroschisis and omphalocele. Over all, major abdominal wall defects had high fatality and specifically, gastroschisis had a significantly worse outcome than omphalocele. Preterm birth (prematurity), low birth weight and time elapsed before arrival to the tertiary center were significantly associated with poor outcome.

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