Frequency of congenital anterior abdominal wall defects in surgically treated infants at Sarajevo region of Bosnia and Herzegovina: A nine-year follow-up

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Abstract

Objectives: Two main and the most common types of congenital abdominal wall defects are omphalocele and gastroschisis. According to the literature, the frequency of omphalocele is generally stable, the rate of gastroschisis has increased lately, and cloacal exstrophy is a very rare birth defect.

Methods: A retrospective study was performed on the basis of clinical records for the period January 2000 to December 2008, and the standard methods of descriptive statistics were performed for the data analysis. Investigation was carried out to obtain the frequency and gender distribution of congenital abdominal wall defects among patients surgically treated in the Clinic for Children’s Surgery, Clinical Center University of Sarajevo, Bosnia and Herzegovina.

Results: A register-based data show that total of 26 abdominal wall defect cases were surgically treated in the investigated period. Out of that number 15 (60%) were male patients, while 11 (40%) were female patients; sex ratio – 1.5:1. Twenty patients had gastroschisis, five omphalocele and one patient were with cloacal exstrophy. Anomalies associated with abdominal wall defects were present in total of six patients (23%), four male (15%) and two female (8%) patients.

Conclusion: The study allows the conclusion that abdominal wall defects are generally rare congenital anomalies of which cloacal exstrophy is the rarest, the gastroschisis is on the rising trend and omphalocele is generally stable.

Keywords: omphalocele, gastroschisis, cloacal exstrophy, frequency

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Introduction

Ventral body wall defects in the thorax or abdomen may involve the heart, abdominal viscera, and urogenital organs. They may be due to a failure of body folding, in which case one or more of the four folds (cephalic, caudal and two lateral) responsible for closing the ventral body wall at the umbilicus fail to progress to that region. Another cause of these defects is incomplete development of body wall structures, including muscle, bone and skin. Two main types of abdominal wall defects are omphalocele and gastroschisis [1]. Their combined incidence is 1 in 2,000 births [2]. The aetiology and incidence of these 2 defects differ [3]; for gastroschisis, teratogens are implicated [4], whereas foetuses with omphalocele also tend to have other anomalies and abnormalities [5, 6, 7]. Omphalocele is herniation of abdominal viscera through an enlarged umbilical ring. The viscera, which may include liver, small and large intestines, stomach, spleen, or bladder, are covered by amnion. The origin of omphalocele is a failure of the bowel to return to the body cavity from its physiological herniation during the 6th to 10th weeks. Omphalocele, which occurs in 2.5/10,000 births, is associated with a high rate of mortality (25%) and severe malformations, such as cardiac anomalies (50%) and neural tube defects (40%). Chromosomal abnormalities are present in approximately 50% of liveborn infants with omphalocele [8]. Gastroschisis is a herniation of abdominal contents through the body wall directly into the amniotic cavity. It occurs lateral to the umbilicus, usually on the right, through a region weakened by regression of the right umbilical vein, which normally disappears. Viscera are not covered by peritoneum or amnion, and the bowel may be damaged by exposure to amniotic fluid. Gastroschisis occurs in 1/10,000 births but is increasing in frequency, especially among young women, and this increase
may be related to cocaine use [1]. Unlike omphalocele,
gastroschisis is not associated with chromosome abnor-
malities or other severe defects. The most serious de-
fect of the ventral abdominal wall is cloacal extrophy
a severe birth defect wherein much of the abdominal
organs (the bladder and intestines) are exposed. It often
causes the splitting of both male and female genitalia
(specifically, the penis and clitoris respectively), and the
anus is occasionally sealed. It is caused by a defect of the
ventral body wall - mesodermal migration is inhibited
and folding fails. Cloacal extrophy is rare birth defect
which occurs in one to 200,000 live births [9]. In our
previous study [10] we investigated the frequency and
sex distribution of the omphalocele and concluded that
our study is not consistent with world-wide trends in
showing the increasing incidence of abdominal wall de-
fects and that more studies should be done to elucidate
this phenomenon. Consequently, the aim of this work
was to obtain the frequency and gender distribution of
all cases of abdominal wall defects which were surgical-
ly treated in the Clinic for Children’s Surgery, Clinical
Center University of Sarajevo, Bosnia and Herzegovina
during the almost same period.

**Materials and methods**

Retrospective study was carried out on the basis of the
clinical records in the Department of Pediatric Surgery
of the Clinical Centre University of Sarajevo, Bosnia
and Herzegovina. From 1st January 2000 to 31st De-
cember 2008, a total of 7029 patients were hospitalized
and out of that number 26 cases (0.37%) were diag-
nosed as some type of abdominal wall defect.
Standard methods of descriptive statistics for the obser-
vational data analysis were performed.

**Table 1. Total number of hospitalized patients and different abdominal wall defect types according to the gender during investigated period**

<table>
<thead>
<tr>
<th>Year</th>
<th>N° of patients</th>
<th>omphalocele</th>
<th>gastroschisis</th>
<th>cloacal extrophy</th>
<th>Σ</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Male</td>
<td>Female</td>
<td></td>
<td>Male</td>
<td>Female</td>
</tr>
<tr>
<td>2000</td>
<td>691</td>
<td>154</td>
<td>845</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>2001</td>
<td>664</td>
<td>145</td>
<td>809</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>2002</td>
<td>683</td>
<td>150</td>
<td>833</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>2003</td>
<td>581</td>
<td>157</td>
<td>738</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>2004</td>
<td>638</td>
<td>150</td>
<td>788</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>2005</td>
<td>615</td>
<td>126</td>
<td>741</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>2006</td>
<td>612</td>
<td>175</td>
<td>787</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>2007</td>
<td>604</td>
<td>165</td>
<td>769</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2008</td>
<td>575</td>
<td>144</td>
<td>719</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Σ</td>
<td>5663</td>
<td>1366</td>
<td>7029</td>
<td>4</td>
<td>2</td>
</tr>
</tbody>
</table>
Results

A total of 7029 patients were hospitalized from 1st January 2000 to 31st December 2008. Significant differences across investigated years were found between males (5663) and females (1366); M:F ratio 4:1; (p<0.00001) (Figure 1).

Out of total 7029 hospitalized patients, 26 (0.37%) were diagnosed as some type of abdominal wall defect and surgically treated (Table 1). There was a slightly higher number of male (15,58%) compared to female patients (11,42%); gender ratio – 1.4:1.

The frequency of particular anterior abdominal wall defects is shown in Figure 2. Nineteen of all twenty-six patients with anterior abdominal wall birth defects had gastroschisis (73%), six had omphalocele (23%) and one (4%) patient had cloacal extrophy.

Each type of anomaly was more prevalent in males compared to females. Male:female ratio for omphalocele was 2:1, gastroschisis 1.7:1 and there was only one case of cloacal extrophy and that was a male patient.

Figure 3. shows the frequency of investigated abdominal wall defect types according to the gender.

Frequency of each type of anomaly was almost uniform throughout the study period in both male and female research group (Table 2).

Anomalies associated with abdominal well defects were present in total of six patients. Figure 4. shows the frequency of different anomalies associated with abdominal wall defects. Meckel’s diverticulum were associated with omphalocele in three cases and in one case of gastroschisis. Intestinal atresia and intestinal duplication were both associated with gastroschisis.

Discussion

Estimated prevalence of gastroschisis (1 in 10 000 births) and omphalocele (2.5 in 10 000) in Western

Table 2. Frequency of abdominal wall defect types during investigated period

<table>
<thead>
<tr>
<th>Year</th>
<th>omphalocele</th>
<th>gastroschisis</th>
<th>cloacal extrophy</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Male Female</td>
<td>Male Female</td>
<td>Male Female</td>
</tr>
<tr>
<td>2000</td>
<td>0.43%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2001</td>
<td>0.15% 0.68%</td>
<td>0.15%</td>
<td></td>
</tr>
<tr>
<td>2002</td>
<td>0.14%</td>
<td>1.33%</td>
<td></td>
</tr>
<tr>
<td>2003</td>
<td>0.34% 0.63%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2004</td>
<td>1.33%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2005</td>
<td>0.16%</td>
<td>0.16%</td>
<td></td>
</tr>
<tr>
<td>2006</td>
<td>0.16%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2007</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2008</td>
<td>0.17% 0.69%</td>
<td>0.69% 1.38%</td>
<td>0.17% 0.01%</td>
</tr>
<tr>
<td>Total</td>
<td>0.08% 0.27%</td>
<td>0.01%</td>
<td></td>
</tr>
</tbody>
</table>

Figure 3. Frequency of different abdominal wall defect types according to the gender

Figure 4. Number of different anomalies associated with abdominal wall defects.
countries [11, 12] are comparable to those of Down’s syndrome (14 in 10 000), neural tube defects (6 in 10 000), congenital heart defects (5 in 10 000), orofacial clefts (11 in 10 000) and limb reduction defects (3 to 8 in 10 000) [13]. Obtaining accurate data on the frequency of abdominal wall defects is complicated by the fact that, in some cases, the pregnancy is ended by elective termination. Consequently, we decided to carry out retrospective study on the basis of the clinical records in the Department of Pediatric Surgery of the Clinical Centre University of Sarajevo, Bosnia and Herzegovina. Our results show that, in the period from 1st January 2000 to 31st December 2008, a total of 7029 patients were hospitalized, and that 5663 male and 1366 female patients. In the same period, a total of 26 (0.37%) abdominal wall defect cases were surgically treated. Out of that number 15 (60%) were male patients, while 11 (40%) were female patients; sex ratio – 1.5:1. Twenty patients had gastroschisis, five omphalocele and one patient were with cloacal exstrophy. Anomalies associated with abdominal wall defects were present in total of six patients (23%), four males (15%) and two females (8%) patients. In our previous study (10) we concluded that omphalocele is a rare congenital anomaly whose frequency doesn’t vary a lot through the years. Although the birth prevalence of omphalocele has remained generally stable over the investigated 20 years [13], reports from Europe, the United States and Japan suggest that the rate of gastrochisis has increased as much as 10-fold over the only one decade [12, 14-16]. Authors concluded that this increase may be partly due to increased detection and ascertainment resulting from increased use of prenatal ultrasonography; however, if these were the only factors, a similar rise in omphalocele rates would be expected. The cause of gastrochisis is multifactorial and seems to involve vascular disruption of the fetal mesenteric vessels [17]. Retrospective analyses of case series of gastrochisis indicate that the risk is greatest for low-income, young mothers who are heavy smokers [18], who are undernourished [19] and who use over-the-counter medications with vasoactive properties (e.g., pseudoephedrine, phenylpropanolamine, ephedrine, methylenedioxymethamphetamine) during early pregnancy [18, 19]. Our study is comparable with retrospectively investigated cases of congenital abdominal wall defects (omphalocele and gastrochisis) of all patients treated at the Clinic of Pediatric Surgery from 1999 to 2003 [20]. In that study, authors evaluated 13 children, 7 with omphalocele (2 female/5 male) and 6 patients with gastrochisis (2 female/4 male). Omphalocele was prenatally detected in 42.8% of fetuses and gastrochisis in 16.7%. Coexisting anomalies were present in 57.1% of patients with omphalocele and in 16.7% of newborns with gastrochisis. Our data are also comparable with those from study in which from August 1987 to August 2004, 129 cases of gastrochisis were treated at the Pediatric Surgical Unit, Department of Surgery, Ratchaburi Hospital. The study included 61 boys and 68 girls. Associated anomalies were present in 13 patients [21]. Our data are also comparable with those from the retrospective study consisted of 27 cases diagnosed with fetal abdominal wall defects between January 2011 and February 2014 in the perinatology outpatient clinic of Celal Bayar University, Manisa, Turkey in which eighteen (66.7%) cases were diagnosed with omphalocele, 6 (22.2%) had gastrochisis, and 3 (11.1%) had limb body wall defects. Regarding the omphalocele cases; 12 (66.6%) cases had isolated omphalocele, whereas 6 of the 18 cases (33.3%) had associated anomalies (22). In our previous study [10] we concluded that our data are not consistent with world-wide trends in showing the increasing incidence of anterior abdominal wall defects and that more studies should be done to elucidate this phenomenon. According to this investigation, we confirm that omphalocele is not, but gastrochisis is on the rising trend, and cloacal exstrophy is a very rare congenital birth defect.

Conclusions
According to the presented research we can conclude that abdominal wall defects are generally rare congenital anomalies of which the cloacal exstrophy is the rarest one, and that gastrochisis shows the rising trend. In contrary, omphalocele was shown as generally stable condition.

Declaration of interest
The authors declare no conflict of interest for this study.
References


