PROGNOSTIC FACTORS FOR CONGENITAL ABDOMINAL WALL DEFECTS

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Abstract

Omphalocele and laparoschisis are one of the most common congenital diseases with serious consequences upon the life and health of the population. Multiple studies, published in international important medical journals have analyzed and presented the factors that may influence the therapeutic outcomes of both diseases. The aim of this study is to assess the main medical, familial, socio-economic prognosis factors with influence over the diagnosis, treatment and post therapeutic outcomes of congenital abdominal wall defects in children. During this period a total of 51 patients with congenital abdominal defects were admitted to our hospital. Postoperative evolution was favorable for 31 patients. Complications occurred in 26 patients, most of them infection related, 3 local and 17 extended. Prolonged bowel obstruction was developed by 13 patients. 20 patients deceased, 15 with laparoschisis (45%) and 5 with omphalocele (28%). The overall mortality rate was 39.2% for both diseases. Predictive factors for complications are: males, low APGAR score, incongruence between the peritoneal cavity and herniated organs, postoperative anemia and/ or thrombocytopenia, renal failure. Unfortunately the mortality is high as well. Predictive factors for an unfavorable outcome being: males, low birth weight, low APGAR score, associated malformations, large defects, renal failure, bronchopulmonary infection and sepsis.

Key words: omphalocele, laparoschisis, prognostic factors, congenital abdominal wall defects

Introduction

Despite of the evident progresses of diagnosis and treatment of congenital lesions in children, omphalocele and laparoschisis remain one of the most common congenital diseases with serious consequences upon the life and health of the population. During the last decades important progress has been made in the field of prenatal diagnosis, progress that allowed us to make a better judgement over the indication for abortion or labor continuing and prepare for the postnatal treatment (1,2). On the other hand the introduction of prosthetic materials in the treatment of these diseases represented a huge step forward to reducing the complications and decreasing mortality (3). Not least the advancement in the field of the new-born intensive care and resuscitation had major influence over the outcome of the treatment. However, the mortality is still high for both these diseases, approximately 10% for laparoschisis and from 20 to 50% for omphalocele (1-3).

Multiple studies, published in international important medical journals have analyzed and presented the factors that may influence the therapeutic outcomes of both diseases. To my knowledge, there has been no such study in Romania for 2 decades. So, we consider that a study like this in the 21th century Romania is necessary and may contribute to the improvement of the treatment of these diseases in our country.

Our intention is to assess the main medical, familial, socio-economic prognosis factors with influence over the diagnosis, treatment and post therapeutic outcomes of congenital abdominal wall defects in children.

Material and method

This study has been carried out at the „Louis Turcanu" Emergency Clinical Hospital for Children in Timisoara. We have reviewed the medical recordings of the patients with congenital abdominal wall defects admitted to our hospital from the 1st of January 1999 until the 31 of August 2015.

We have recorded and reviewed the parameters shown in table 1. We assessed these parameters in relationship with the evolution of each individual case and the outcome of the treatment.

For statistical analysis we used IBM SPSS Statistics v20 for Microsoft Windows. We used t-Student test for numeric variables and Mann-Whitney test for non-numeric variables; p value was set at 0.05.

Results

During this period a total of 51 patients, 29 males and 22 females with congenital abdominal defects were admitted to our hospital. The gestational age ranged from 31 to 41 weeks, mean 36.73 weeks. 21 patients were born premature (less than 36 weeks).

Birth weight ranged from 1250 g to 4060g, meaning 2400g. Birth height ranged from 33cm to 60cm, meaning 46 cm. A total number of 31 patients had the birth weight under 2500 g, and 8 of them under 1500 g.

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Table 1. Assessed parameters.

Pregnancy and birth	Gestational age	APGAR Score
	Weight at delivery	Delivery rank
	Height at delivery	Pregnancy surveillance
	Sex	Type of delivery
Demographic data	Maternal age	Medical history
	Urban/ Rural	
Disease	Type of disease	Size of the defect
	Clinical form	Age on admitted
	Prenatal diagnosis/ on delivery diagnosis	
Associated pathology	Cardiac malformation	Skeletal malformations
	Intestinal malformation	Renal malformations
	Chromosomal anomalies	
Treatment	Concomitant surgical procedures	Re-interventions
	Surgical procedure	Other surgical procedures
	Age at Surgery	
Evolution/ Complications	Intestinal occlusion	Renal failure
	Respiratory infections	Survival rate/ age of decease
	Sepsis	

Prenatal diagnostic was made in 9 cases, in 3 cases there was no medical follow up during the pregnancy. There were 2 breech presentations and C-section was performed in 11 cases.

Out of the total of 51 cases, 33 were of laparoschisis and 18 of omphalocele. The size of the defect ranged from 2-12 cm, meaning 4,7 cm; 2-7 cm, meaning 3,5 cm for laparoschisis and 3-12 cm, meaning 5,6 for omphalocele. The herniated organ was the bowel alone in 39 out of the 51 cases. The liver was out in 3 cases of omphalocele. More than 3 concomitant organs were herniated in 4 cases of omphalocele.

Maternal age varied from 13 to 45 years, the average age being 22.9 years; 14 were under 20, 34 of them were between 20-35 years old and 3 were over 35 years old. Laparoschisis was more common in children from younger mothers and omphalocele is more frequently associated with older mothers (p=0,036).

Associated malformations were present in 27 out of the 51 patients. Chromosomal anomalies had been found in 1 patient, 9 had cardiac malformations, 2 skeletal malformations, 7 with intestinal atresia and 2 with renal malformations. Multiple malformations were present in 2 patients. The omphalocele was associated more frequently with multiple malformations and skeletal malformations. The rest of the malformations were divided equally between the 2 groups of diseases.

The age of the patients at admission into the hospital was between < 1 hour and 72, meaning 6.2 hours. Only in three cases the admission in a neonatal surgical department was not made in the first 24 hours. The age at surgery varied from 5 to 26 hours, meaning 9.4 hours. In only one case the surgical intervention was performed after 24 hours after admission.

Surgery was the first treatment option in 50 out of the 51 patients. Schuster procedure has been performed in 9 cases, Gross procedure in 2 laparoschisis and Fufezan procedure in other 3 omphalocele cases. In the past few years it was possible the use of silastic bags for the progressive reduction of the bowels in the abdominal cavity in 3 cases of laparoschisis and 1 of omphalocele (Table 2).

Table 2. Surgical procedure.

		Disease		
		LAPAROSCHISIS	OMPHALOCELE	Total
SURGICAL PROCEDURE	PRIMAR CLOSE	27	13	40
	SCHUSTER	2	0	2
	GROSS	1	0	1
	FUFEZAN	0	3	3
	SILICON BAG	3	1	4
Total		33	17	50

Concomitant surgical procedures were necessary in 8 patients, 5 intestinal resections and anastomosis for intestinal atresia, 1 resection of Meckel diverticulum and 1 repairing of the bladder exstrophy and 1 omental resection. Reinterventions were necessary for 19 patients, 10 of these had programed for second step Schuster procedure, Fufezan procedure and silastic bag; 2 for the programed delayed repairing of the intestinal atresia. In all there were 51 reinterventions.

Postoperative evolution was considered favorable for 31 patients. Complications occurred in 26 patients, most of them infection related, 3 local and 17 extended. Prolonged bowel obstruction developed in 13 patients. 20 patients deceased, 15 with laparoschisis (45%) and 5 with omphalocele (28%). The overall mortality rate was 39.2% for both diseases.

The statistical analyzes of the main factor influencing the evolution of abdominal wall defects are presented in table 3 and 4 (statistical significant ones with bold).

Table 3. Prognostic factors for complications.

	Mann-Whitney U	Wilcoxon W	Z	p
Sex	894,000	1674,000	-1,884	0,050
Gestational age	813,500	2088,500	-,554	0,579
Birth weight	878,000	2418,000	-1,306	0,192
APGAR	444,500	1572,500	-2,578	0,010
Follow up during pregnancy	273,000	769,000	-,151	0,880
Delivery	1089,000	2742,000	-,234	0,815
Presentation	1110,000	1890,000	-,029	0,977
Mother's age	683,500	1673,500	-,745	0,456
Type of defect	814,500	2467,500	-2,675	0,007
Size of defect	778,500	1306,500	-,358	0,720
Herniated organs	1056,000	1836,000	-,478	0,632
Chromosomal anomalies	1075,500	2728,500	-,561	0,575
Cardiac malformations	981,000	1761,000	-1,411	0,158
Intestinal atresia	1105,500	2758,500	-,078	0,938
Skeletal malformations	1069,500	1849,500	-,547	0,584
Renal malformations	1093,500	2746,500	-,388	0,698
Multiple malformations	1047,000	2700,000	-,953	0,341
Surgical treatment	1090,500	1870,500	-,374	0,709
Age at the surgery	264,000	792,000	-1,058	0,290
Surgical procedure	794,000	1497,000	-1,941	0,052
Surgical re-interventions	818,500	1521,500	-1,500	0,134
Concomitant surgery	908,000	2183,000	-,216	0,829

Discussions

Laparoschisis and omphalocele are of the most serious congenital diseases in terms of distress for the neonate, high mortality and morbidity rate altogether with a high incidence for both diseases. During the period of 12.6 years we have treated 51 cases of congenital abdominal wall defects, of which only 17 originated form Timis County. Of these 7 cases of omphalocele were from Timisoara and 10 patients of laparoschisis were from the rural area of Timis County. Corroborating this data with the general birth rate in our county of 7000/ year (4) we have found an incidence of 1:8400 for laparoschisis and 1:12000 for omphalocele. Our results are slightly different from those in the literature, 1:4000 for laparoschisis respectively 1:5000-10000 for omphalocele (5). The gender distribution of 1:1 for laparoschisis and 1.6:1 for omphalocele is similar with the previous reports (5) and we found a higher rate of mortality and complications in male patients (p=0,04, respectively p=0,050).

Periodic medical visits and appropriate follow-up during pregnancy represent an essential method for prevention and efficient treatment of congenital diseases. The prenatal diagnosis is an imperative condition for planning a good therapeutic conduct. The levels of alpha-fetoprotein and the transvaginal echography may establish the diagnosis since the 10th week of gestation (6). In case of laparoschisis this is of extremely importance since the majority of the specialist recommend that the baby should be delivered prematurely as soon as the fetus becomes viable in order to shorten the exposure of the bowels to the amniotic fluid (7). Unfortunately in our study the prenatal diagnosis was set in less than 20% of the cases. Moreover in 3 cases no medical follow up was carried on during pregnancy. This is a clear indication that there is still a lot to do for the primary care medical assistance in some remote rural areas.

Table 4. Prognostic factors for mortality.

	Mann-Whitney U	Wilcoxon W	Z	p
Sex	846,000	1512,000	-2,056	0,040
Gestational age	660,000	2145,000	-1,631	0,103
Birth weight	586,000	2297,000	-3,404	0,001
APGAR	307,000	1633,000	-3,718	0,000
Follow up during pregnancy	183,500	963,500	-,346	0,729
Delivery	1038,000	1704,000	-,444	0,657
Presentation	1074,000	2904,000	-,118	0,906
Maternal age	555,500	1731,500	-1,936	0,053
Type of defect	876,000	2706,000	-1,864	0,062
Size of defect	760,000	2245,000	-,224	0,822
Herniated organs	873,000	1539,000	-1,810	0,070
Chromosomal anomalies	1080,000	2910,000	,000	1,000
Cardiac malformations	882,000	1548,000	-2,171	0,030
Intestinal atresia	1056,000	2886,000	-,317	0,751
Skeletal malformations	912,000	1578,000	-2,219	0,026
Renal malformations	1056,000	2886,000	-,525	0,600
Multiple malformations	966,000	1632,000	-1,709	0,087
Surgical treatment	972,000	1638,000	-1,949	0,051
Age at the surgery	178,000	958,000	-1,603	0,109
Surgical procedure	705,500	1371,500	-2,786	0,005
Surgical re-interventions	883,500	2368,500	-,649	0,517
Concomitant surgery	853,500	2284,500	-,612	0,540
Intestinal occlusion	986,000	2526,000	-,045	0,964
Renal failure	810,000	1476,000	-2,764	0,006
Sepsis	642,000	1308,000	-3,848	0,000
Wound infection	1074,000	2904,000	-,118	0,906
Total complications	630,000	1296,000	-4,003	0,000

In our study the gestational age varied between 29 and 41 weeks, less than a third (28%) of children being delivered premature. We cannot talk about premature programmed delivery in laparoschisis because the prenatal diagnosis was set in only 17% of these, meaning 9 cases. So the majority of these cases were natural occurred premature birth meaning that they were rather an additional factor for the disease itself. Our statistical analysis did not revealed that. Instead we found out that the low birth weight is an important factor which influences the mortality not only in the case of neonates with congenital wall defects.

APGAR score is the indicator of health in neonates and offers an image upon the future evolution of the patient. Although is not necessary directly related to the underlying disease, a low APGAR score is a strong predictor for a baleful evolution (p=0,010) or for the complications (p=0,010).

The main factors in influencing the therapeutic conduct of congenital abdominal are: the size of the defect and the presence or absence of the associated diseases. Generally the size of the abdominal wall defect is lower in laparoschisis than in omphalocele (7). A possible predictive value and in direct relation with the size of the defect is the number and the type of organs herniated. In most of the cases (62%) the herniated organ was the bowel, followed by the liver. Associated disorders were present in approximately half of the patient and they were a predicating factor for unfavorable outcome. The cardiac malformations are preponderant 11 (32%) for omphalocele and 8 (12%) for laparoschisis. Intestinal atresia was presented in 10 cases of laparoschisis (16%) and 2 cases of omphalocele (5.8%), the percentage being similar to those from literature (1, 2, 3). Statistically speaking we found that the associated malformations in general (p=0.05) and cardiac malformations (p=0.03) had significant influence upon the mortality while the occurrence of postoperative complications had not been influenced by these. So we conclude that the associated malformations, especially the cardiac ones are an important factor in the patient survival with congenital wall defect.

One of the most important factors influencing the outcome of a congenital wall defect is the quality and the precocity of treatment (6). Specific medical measures and treatment planning should begin while the patient with abdominal wall defect is still in utero. After delivery the quality primary care of the neonate appropriate transport condition and the immediately as possible surgical closure of the defect are essential to obtain favorable results. Surgical treatment is the only curative way for these diseases. The choice of surgical procedure depends on the size of the defect, but also on the surgeon's ability and the available options for prosthetic materials. The small number of patients treated by surgical augmentation and/ or staged closure of the abdominal; wall defect does not allow us to

conclude upon the benefits of one procedure. Yet we want to make a special reference to Fufezan procedure which represents in our opinion a cheap and simple way of gradual reintegration of the abdominal organs in the case of patients with omphalocele (8). However, neither the type, nor the number of the surgical procedures influenced significantly the outcomes of the treatment in our study.

The postoperative course of the patients with abdominal wall defect, particularly for those with big defects or associated malformations is marked by high morbidity and mortality rates (9). In our study 60% of the patients had postoperative complications the most prevalent being the infectious ones. The presence of sepsis represented one of the main death causes, 20 out of the 26 patients with sepsis had unfavorable course (p=0.000). Renal failure is the most feared immediately after surgical closure of the defect and is directly related to the capacity of the surgeon to estimate the balance between the capacity of the abdominal cavity and the volume of the herniated organs. Fortunately this complication occurred in a limited number of cases but, also it had a significant influence over the unfavorable course of the case.

Eventually all of the parameters which we found to have predictive value for the course and the outcome of abdominal wall defects can be adjustable and the first step to solve the problem is to find out where it is.

Conclusions

Even though smaller than that reported in literature, both laparoschisis and omphalocele still have a high incidence in the western part of Romania. Primary or delayed closure of the defect shell being the first option, nowadays it is possible in most of the cases. Complication rate is still high with most frequent complications being infection related. Predictive factors for complications are: males, low APGAR score, incongruence between the peritoneal cavity and herniated organs, postoperative anemia and/ or thrombocytopenia, renal failure. Unfortunately the mortality is high as well. Predictive factors for an unfavorable outcome being: males, low birth weight, low APGAR score, associated malformations, large defects, renal failure, bronchopulmonary infection and sepsis.

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