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Management of Omphalocele in the Pediatric Surgery Department of Donka National Hospital

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Abstract

Introduction: Omphalocele is an embryopathy of the first ten weeks of gestation. It corresponds to a defect in the abdominal wall through which viscera contained in a sac constituted by the amniotic membrane and centered by the umbilical cord are exteriorized. The objective of this work was to study the diagnostic modalities and the impact of our therapeutic choices on the outcome of the management. **Material and Methods:** We carried out a prospective study on patients with omphalocele admitted to the pediatric surgery departments of the HND, during a period spread over 4 years, between January 2017 and December 2020. **Results:** we collected 55 files (i.e., 13.7 cases/year). There were 38 boys and 17 girls (sex ratio 2.2) with an average age of 1.9 days. We found 15 cases (27%) of type I and 40 cases (73%) of type II according to the AITKEN classification. Fifteen cases (27%) benefited from surgical treatment and 45 cases (73%) benefited from conservative treatment (Grob). Two surgical methods were used: Primary parietal closure, which was used in 10 cases (66.7%), and the GROSS method, which was used in 5 cases (33.3%). We obtained an improvement of 40 cases (73%) and 15 cases (27%) of death. **Conclusion:** our work reported our experience in the management of omphalocele and the difficulties of postoperative resuscitation.

Keywords

Omphalocele, Tannage, Gross, Embryopathy

1. Introduction

Omphalocele is a congenital anomaly of the abdominal wall, caused by an inability of the abdominal contents to return to their cavity at the 10th week of gestation [1]. It is an early disorder of embryogenesis at the stage of morphogenesis, with inhibition of the somatopleural layer of the lateral folds of the embryo at the level of the umbilical region [2] [3]. It corresponds to a defect in the closure of the umbilical ring with exteriorization of the abdominal viscera covered by a translucent and avascular amniotic membrane, Wharton's jelly next to it and the vessels of the umbilical cord implanted at the apex of the sac [4] [5].

It is one of the most common malformations of the anterior abdominal wall, and its frequency is estimated to be between 1/400 and 1/700 live births [6] [7]. It can be associated with several syndromes, the most common being Beckwith-Wiedemann syndrome [8]. It is often associated with other congenital and chromosomal abnormalities, such as trisomy, cervical cancer and breast cancer [9].

The morbidity and mortality associated with omphalocele depend on factors such as the existence of associated malformations or chromosomal abnormalities, the diameter, the content of the omphalocele and the birth weight [6].

Its diagnosis is most often made prenatally during the obstetric ultrasound scan in the first or second trimester of pregnancy. It appears clinically as an anomaly of the medial wall of variable size and location [10] [11].

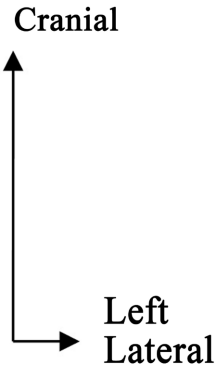
Its treatment is either medical by tanning or surgical using many techniques. These surgical techniques require effective and adapted resuscitation means because of the hemodynamic and respiratory changes and consequences of the reintegration of the loops in an almost always narrow abdominal cavity [4] [12].

In developed countries, the indication for surgical treatment is systematic and the prognosis of this condition is improved thanks to progress in anesthesia and neonatal resuscitation as well as the development of antenatal diagnosis [3] [10].

In developed countries, the therapeutic approach is different and the lethality remains high because of the inadequacy of the means of anesthesia and neonatal resuscitation [3] [10] [12].

The aim of this work was to study the diagnostic modalities and the impact of our therapeutic choices on the outcome of the management.

(A) (B) (C)



(A): hernia in the cord; (B): Omphalocele type I; (C): Omphalocele type II with visible contents.

2. Patients and Methods

Our work is a prospective descriptive study lasting 4 years, from January 1, 2017, to December 31, 2020. It concerned newborns received, diagnosed and hospitalized for omphalocele. All newborns whose omphalocele diagnosis was retained and treated in the department were included in this study. Cases of omphalocele falling within the framework of a malformation complex (OIES syndrome) were excluded.

For each file, the parameters studied were epidemiological, clinical, paraclinical and therapeutic variables.

The data were entered into Excel and analyzed using SPSS software.

3. Results

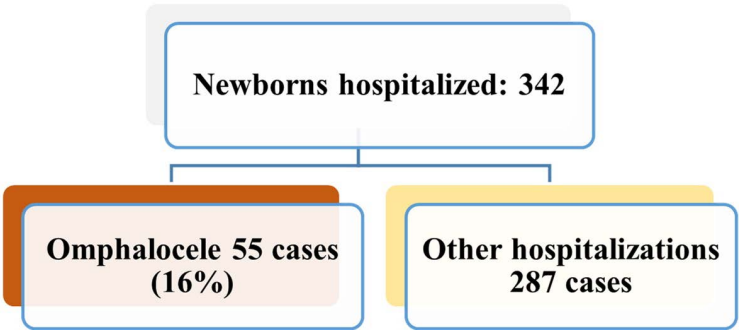


Figure 1. Frequency of omphalocele in relation to hospital admissions.

Table 1. Prevalence of omphalocele.

Year	Newborns hospitalized	Number of omphalocele cases	Percentage
2017	78	20	25.64
2018	94	13	13.82
2019	94	11	11.70
2020	76	11	14.47
Total	342	55	16.00

Annual incidence = 13.7 cases/year.

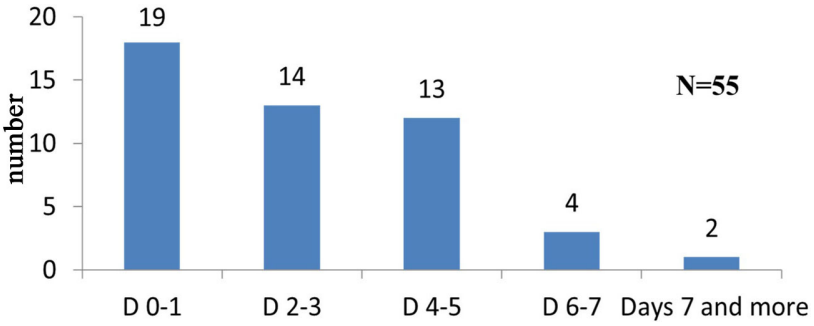


Figure 2. Age distribution of patients.

The mean age was 1.9 days, with extremes of J0 and J10, and a standard deviation of 1.09.

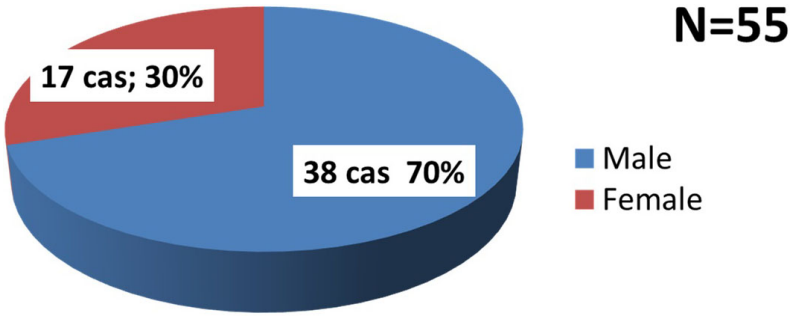
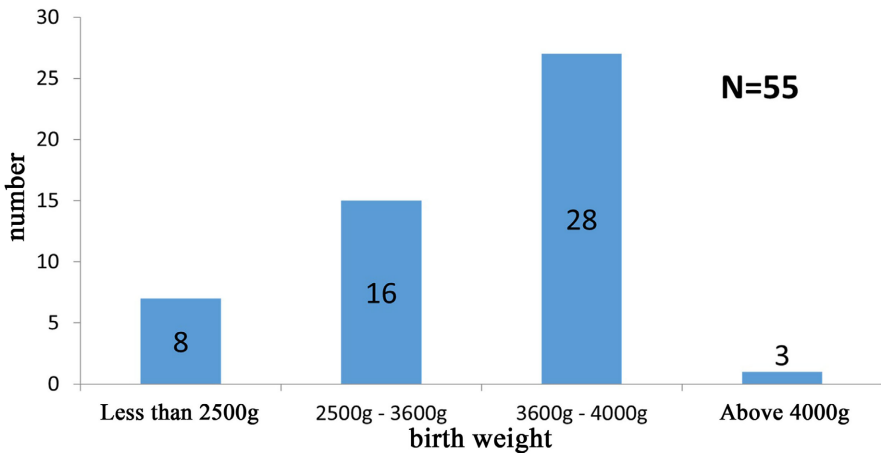


Figure 3. Distribution of patients by gender. Ratio (M/F) = 2.2.



Average weight = 3600 g. Standard deviation = 1.18. Extremes [1800 g – 4200 g].

Figure 4. Distribution of patients by birth weight.

Table 2. Frequency of reasons for consultation.

Patterns	Workforce (N = 55)	Percentage
Swelling in the umbilical region	55	100
Fever	24	43.6
Congenital evisceration	5	9
Dyspnea	5	9

Table 3. Distribution of patients according to omphalocele membrane appearance.

Character of the omphalocele membrane	Workforce	Percentage
Translucent	38	69.30
Opaque	7	12.70
Rompue	5	9
Infected	5	9
Total	55	100

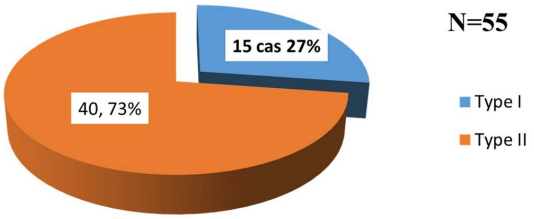


Figure 5. Patient distribution according to AITKEN classification.

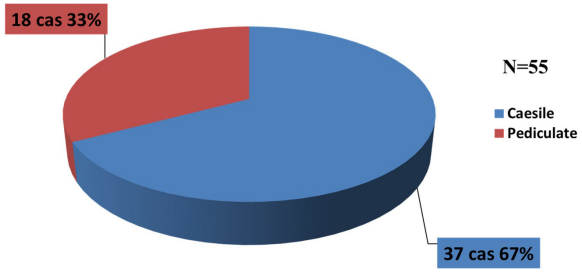


Figure 6. Distribution of patients according to omphalocele shape.

Table 4. Distribution of patients according to associated pathologies.

Neonatal infection	14	25.45
Maternal-fetal infection	12	22.00
Cryptorchidism	5	9.00
Weidman beck withe syndrome	5	9.00
Respiratory distress	3	5.45
Intraventricular collapse	2	3.63
Hydrocele	4	3.63
Inguino-scrotal hernia	2	3.63
Pentalogiy of cantrell	2	3.63
Cleft lip and palate	2	3.63

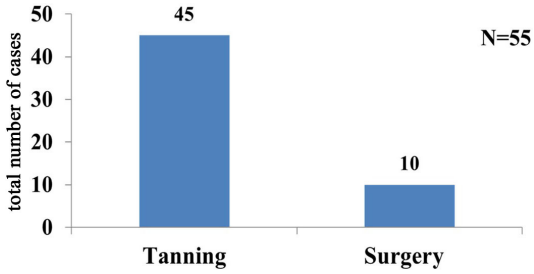


Figure 7. Distribution of patients according to therapeutic option or type of treatment used.

Table 5. Distribution of patients according to evolution of tanning treatment.

Tanning evolution	Workforce (N = 45)	Percentage
Epidermization	34	75.6
Deceased	10	22.2
Evisceration	1	2.2
Total	**45**	**100.00**

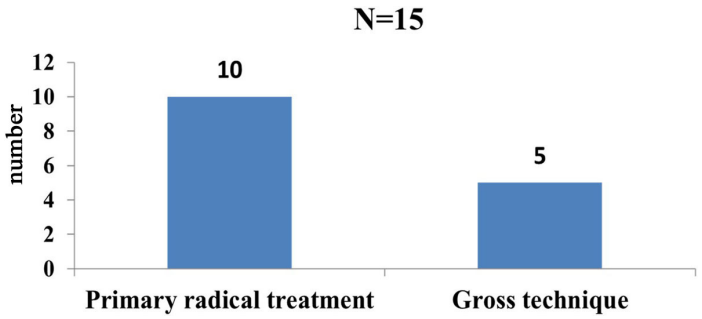


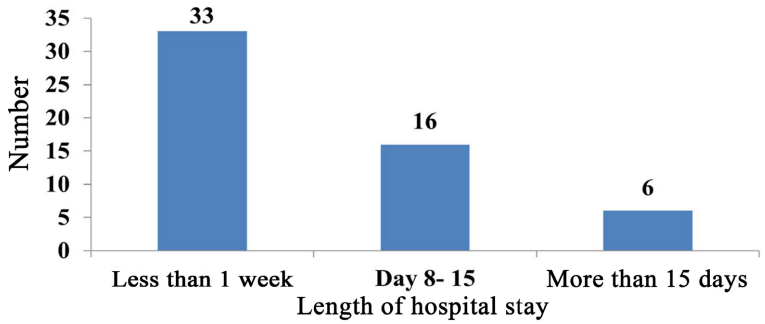
Figure 8. Distribution of patients by type of surgical technique used.

Table 6. Distribution of omphalocele types according to therapeutic method.

Type of omphalocele	Type of treatment		
	Tannage	Chirurgical	
		Radical cure 1st	Gross
Omphalocele type I	5	10	0
Omphalocele type II	40	0	5
Total	45	15	

Table 7. Distribution of patients according to evolution.

Evolution		Workforce	Percentage
Complications	Simples	34	62
	Occlusion by bridles	3	5.45
	Parietal suppuration	2	3.63
	Evisceration	1	1.81
	Suture release	2	3.63
Deceased		15	27
Total		**55**	**100**



Mean duration = 6 days. Standard deviation 1.5. Extremes [3 days – 28 days].

Figure 9. Distribution of patients by length of hospital stay.

Table 8. Summary of bivariate Chi-square analysis of selected mortality factors in relation to death.

Mortality factors	Workforce	Deceased/15	Value of P
Birth weight < 2500 g kg	7	5	**0.032**
Premature	2	2	**0.002**
Transport by taxi	42	5	3.894
Type I	15	05	2.585
Type II	40	03	2.662
Treatment by tanning	45	10	**0.046**
Surgical treatment	10	5	1.563

P < 0.05 is statistically significant.

4. Discussion

Omphalocele is a congenital malformation of the abdominal wall, covered by a membranous sac [13]. The umbilical cord inserts into the apex of this sac, usually containing herniated abdominal contents.

In our study, it represented 16% of hospitalizations in the pediatric surgery department of Donka National Hospital from 2017 to 2020, with an annual incidence of 13.7 cases/year.

This result is significantly higher than that of Andriamanarivo, M.L. *et al.* [12] and Kanté, L. *et al.* [14]. This could be explained by the fact that our department is the only pediatric surgery center in the country.

Table 9. Comparative table of the annual incidence of omphalocele by author.

Authors	Number of cases	Cases/year
Andriamanarivo ML *et al.* [12] (Madagascar)	**32** [**5 years**]	6.4
Kanté L *et al.* [14] (Mali)	20 [5 years]	4
Our series (Guinée)	**55** [4 **years**]	**13.7**

In our series, the age range from J0 to J1 was the most marked with an average of 1.9 days and extremes of J0 and J10. This result is similar to that of Andriamanarivo ML *et al.* [12] who had an average of 1,5. This could be explained by the fact that 80% of our patients were born in a health facility, referred to our department and 64% of our patients come from an urban area.

In our study, a male predominance was found with a ratio = 2.2. This study was similar to those of Kouame BD *et al.* [10], N'Dour O *et al.* [3] and Kanté L *et al.* [14] who reported a male predominance, with respective ratios of 1.2, 1.02 and 1.26.

The pathogenesis of this condition is not fully understood, although various mechanisms have been proposed. But it seems that omphalocele is more common in extreme ages (young or advanced maternal age), in male newborns, in multiple pregnancies, in maternal obesity, in maternal diabetes and in low socio-economic levels [15] [16].

The average weight of our patients was 3600 g with extremes of 1800 g and 4200 g; 08 newborns weighed less than 2500 g. This result is comparable to that of Kouame BD *et al.* [10] an average weight of 3137 g, with 16 cases having a weight <

2500 g. But significantly higher than that of Kamgaing dr'N *et al.* [17] who reported an average weight of 2800 g with extremes of 2500 g and 3000 g.

In the literature, low birth weight (less than 2.5 kg) and prematurity constitute a poor prognostic factor [18]. Indeed, the problems posed by omphalocele are added to those of prematurity. In our study, we found 1 case of prematurity, or 2%. In the literature, the rate of prematurity is higher in omphaloceles integrated into the framework of a polymalformative syndrome than in isolated omphaloceles [19]. In developed countries, screening is systematic allowing to diagnose more than 90% of cases before birth [20], because these congenital anomalies of the abdominal wall are generally detected during pregnancy by ultrasound. But in our study no case of antenatal diagnosis was made.

The predominance of type II of the AITKEN classification in our study is comparable to that of Kouame BD *et al.* [10] who reported 56.2% of type II. This result was different from those of Kanté L *et al.* [13] and N'Dour O *et al.* [3] who had respectively 73.9% and 78.2% of which type I predominated.

The Aitken classification used has a prognostic and therapeutic interest. Type I omphaloceles have a better prognosis and are treated by primary closure. Type II omphaloceles, with a more significant parietal defect, are exposed more to the risks of hypothermia, acidosis and infection and are treated by more complex surgical techniques. They still retain a severe prognosis because the morbidity rate linked to this type is very high.

In our study, omphalocele was associated with medical pathologies in 64% of cases and with congenital malformations in 30% of cases. This result is significantly lower than that of Gaigi SS *et al.* [19], who reported 85% of cases of congenital malformations respectively. But significantly higher than that of Kouame BD *et al.* [10] who reported 16.5% of cases of congenital malformations.

Omphalocele is an embryopathy where associated malformations are very frequent [21]. Associated malformations, in particular congenital heart disease, syndromic omphaloceles and chromosomal aberrations, are the main factors that condition the vital prognosis [22].

The lower rate of congenital malformation in our study could be explained by the fact that 91% of our patients did not perform a malformation assessment. In our study, treatment by tanning was performed in 82% of cases versus 18% of surgery, similar to those of Kouame B.D *et al.* [10] and N'Dour O *et al.* [3] who reported 87.5% and 57.9% of Tanning. This could be explained by the predominance of type II omphalocele in our study, the high frequency of pathologies associated with omphalocele and the insufficiency of the technical platform.

In our study, 2 surgical techniques were used on 15 patients: Simple radical cure: 10 cases (66.7%) and according to Gross: 5 cases (33.3%).

This is the radical cure in one stage with an approximation of the musculoaponeurotic plane, considered by most authors as the method of choice in the treatment of type I omphaloceles according to Aitken [11]. In our series, it was the most used technique, recommended in 10 cases, *i.e.*, a rate of 20%. We noted good

results of 80% for the forms of type I omphalocele [P-value = 0.028], for which we have a low mortality rate of 0.4% [P-value = 2.585].

This result is lower than those of other studies conducted by Kamgaing N *et al.* [17] who had used this technique in 78% of cases. They obtained good results (100%) for the forms of type I omphalocele, for which they had no deaths. This could be explained by the low rate of type I and associated pathologies.

Currently, the indication of surgery is systematic in the treatment of omphaloceles thanks to the progress of anesthesia and neonatal resuscitation as well as the development of antenatal diagnosis [23]. But in our context, we recommend conservative treatment with deferred surgery.

Other therapeutic alternatives exist to minimize the morbidity and mortality related to abdominal hyperpressure. Immediate parietal closure by skin covering according to the Gross technique is simple. Indeed, this technique requires multiple interventions [6].

The most used technique is progressive reintegration by silo (Schuster method) (4). Closure is obtained in about fifteen days (3). However, progressive closure, like the initial closure of the omphalocele, can induce abdominal hyperpressure and, consequently, respiratory distress, a drop in cardiac output, hypotension, risk of intestinal ischemic necrosis and renal suffering. In addition, this technique can be burdened with other complications: septicemia, enterocutaneous fistula, eventration, and parietal infection [8] [17] [18]. In our study, this technique was widely used. It was recommended in 74% of cases, including 5 cases of type II omphalocele that benefited from the Gross technique, *i.e.*, 10%, with a cure in 4 cases [P-value = 0.002]. Kouame, B.D. *et al.* [10] recommended it in 87.5% and 10 cases were operated on according to Gross, *i.e.*, 12.19%. Kanté, L. *et al.* [14] recommended it in 73.87% and 29 cases were operated according to Gross, *i.e.*, 26.12%.

Our choice of this conservative method is linked, on the one hand, to the high rate of type II; on the other hand, it constitutes the therapeutic means for us surgeons that can guarantee a probable cure, especially in our condition of limited neonatal resuscitation.

In total, we have a good result in 73% of cases against 27% of mortality.

The mortality rate in our series is higher than that of Kanté L *et al.* [14] who noted 18% of deaths. But clearly lower than that of N'Dour O *et al.* [3] who had noted 30% of deaths and of Kouame BD *et al.* [10] who had noted 84.6% of deaths.

This high mortality rate in these series could be explained by the high frequency of postoperative complications on the one hand and on the other hand by the high rate of associated malformations. In our study, these deaths were influenced by certain factors such as low birth weight [P-value = 0.022] and Prematurity [P-value = 0.002]. Which is reinforced by the lack of adequate postoperative resuscitation means.

5. Conclusions

Omphalocele is a congenital malformation characterized by evisceration through

a defect in the central anterior abdominal wall covered by a membranous sac and the umbilical cord fixed at its base. It is one of the most common malformations of the anterior abdominal wall, where associated malformations are frequent, particularly in the digestive, cardiac, and urinary areas, which condition the prognosis.

Its expensive management requires a multidisciplinary technical platform. The treatment of giant or type II omphaloceles remains a current problem.

The progressive silo reintegration technique (Schuster) is the most used. It requires the possibility of adequate resuscitation. Despite the progress of resuscitation, this technique is burdened with complications. Prosthetic closure is also used in the treatment of type II omphaloceles. The GROB technique is reserved for giant omphaloceles associated with serious malformations with a poor prognosis.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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