

Giant Omphaloceles: Surgical Management Using The Schuster Technique

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Abstract:

Introduction:

Giant omphaloceles represent a management challenge due to the lack of a standardized therapeutic protocol. Progressive reduction before closure is the only alternative to reduce morbidity and mortality. The objective of this study is to describe the results of the management of giant omphaloceles using Schuster's progressive reduction technique.

Materials and methods: We conducted a descriptive retrospective study of 28 patients with giant omphalocele over a period of 10 years, between January 2012 and December 2021. Of the 28 patients, half (14 patients) benefited from the classical Schuster technique and the other half from a modified Schuster technique.

Results: The sac was ruptured in 4 cases, a rate of 14.28%. A total of 50 associated malformations were recorded in 20 newborns (71.42%), with a predominance of cardiac malformations (42.85%). The mortality rate in our series was 57.14%. Nevertheless, giant omphaloceles treated with the classic Schuster technique had a mortality rate of 85.71% (12 deaths out of 14 patients) while those treated with the modified Schuster technique had a mortality rate of 28.57% (4 deaths out of 14 patients).

Conclusion: Our experience with the use of the modified Schuster technique, the application of honey in the dressings and the conservation of Wharton jelly, has clearly improved the survival rate of our patients in the treatment of giant omphaloceles.

Keywords: Giant omphalocele, Schuster technique, Wharton jelly

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I. Introduction:

Giant omphaloceles represent a management challenge due to the lack of a standardized therapeutic protocol. Primary closure is often impossible, with all the consequences related to the increase in intra-abdominal pressure, in particular abdominal compartment syndrome and consequently death [1]. Gradual reduction before closure is the only alternative to reduce morbidity and mortality. The objective of this study is to describe the results of the management of giant omphaloceles using Schuster's progressive reduction technique.

II. Methods:

We conducted a retrospective study of 28 patients with giant omphalocele, admitted to the pediatric surgery and neonatology departments at the HASSAN II University Hospital in Fez, during a period spread over 10 years, between January 2012 and December 2021. The study included newborns with a clinical diagnosis of giant omphalocele, namely a defect greater than 5 centimeters and/or a hernia of more than 50% of the liver.

The quantitative variables were described by the mean, the standard deviation, the extremes and the qualitative variables were expressed by the frequencies. This analysis was performed using Microsoft Excel 2017.

Surgical technique:

Of the 28 patients, half (14 patients) benefited from the classical Schuster technique and the other half from a modified Schuster technique.

Classic Schuster technique (Figure 1):

Developed by SCHUSTER in 1967 [2], the principle of this technique is achieving progressive closure of the abdominal wall, taking advantage, in particular, of the possibilities of rapid growth of the abdominal cavity in the first days of life. It includes the following times:

- Incision circumscribing the peritoneo-amniotic plate and its excision,

- Checking the abdominal cavity,
- Suture to the two aponeurotic edges of a silastic plate joined at the top and bottom to the contralateral plate,
- Confrontation of the two plates in front of the herniated viscera.

Then, the child is reinstalled in the incubator with maintenance of optimal curarization post-operatively. The application of a sterile dressing and compliance with strict aseptic conditions during subsequent closing times are essential. Every two days, the silo is gradually closed by placing a series of U-shaped points with discharge carefully from the viscera to the abdominal cavity.

After a first series of 4 or 5 successive re-interventions spaced 2 to 4 days apart, we reach a confrontation of the aponeurotic edges in a fortnight at most. The newborn is then taken back to the operating room, the plates are removed and the abdominal wall is closed.



Fig 1 : The classic Schuster technique

Modified Schuster technique (Figure 2):

In our technique we keep the Wharton jelly with a progressive reintegration of the herniated viscera using a synthetic plate, with the application of honey in the dressings.

III. Results:

Almost all of our patients were born at term, only one case of overterm was found. On the other hand, no case of prematurity was found. The prenatal consultation was made in 12 cases (42%) for which an obstetric ultrasound was performed with 8 cases of antenatal diagnosis (28.57%). 7 cases of caesareans were found (25%). The average birth weight was 3074 ± 485 grams, with a minimum of 2300 grams and a maximum of 4500 grams. The sac was ruptured in 4 cases (14.28%). The diameter of the neck was between 5 and 9 centimeters with a hernia of more than 50% of the liver in 20 cases (71.42%). The diameter of the sac was between 8 and 13 centimeters. Associated malformations were systematically sought by clinical examination, radiological examinations and peroperative exploration. The latter could not be possible in half of the patients having benefited from the modified Schuster technique which consists in preserving the Wharton jelly. A total of 50 associated malformations were recorded in 20 newborns (71.42%), with a predominance of cardiac malformations (42.85%). Two associated malformations of Meckel's diverticulum type were noted during the operative exploration of patients treated with the classic Schuster technique. Two specific cases of polymalformative syndrome were noted, namely a case of a Wiedemann-Beckwith syndrome and a PRUNE-BELLY syndrome. Various complications were found postoperatively. They were dominated by respiratory distress (14.28%) and especially infection (50%). The overall mortality rate in our series was 57.14%. All deaths occurred during the initial tapering phase. Nevertheless, giant omphaloceles treated with the classic Schuster technique had a mortality rate of 85.71% (12 deaths out of 14 patients) while those treated with the modified Schuster technique had a mortality rate of 28.57% (4 deaths out of 14 patients).

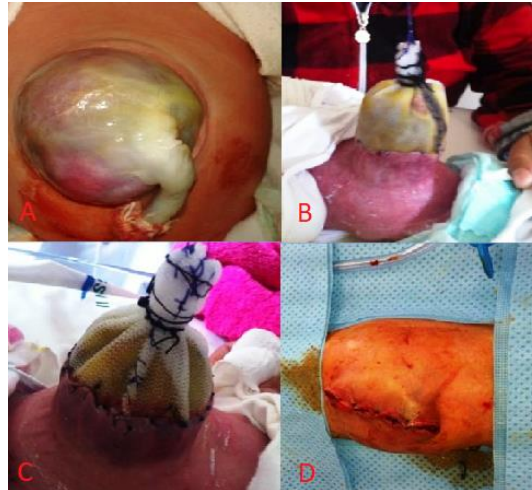


Fig 2 : Modified Schuster technique

- A. Original appearance of giant omphalocele
- B. Use of a synthetic plate sutured to the aponeurosis
- C. Progressive reduction of the viscera with growth of the wall
- D. Secondary closure of the abdominal wall

IV. Discussion:

Antenatal ultrasound makes it possible to ensure the diagnosis of omphalocele, early and effective management of the newborn, and also in some cases the associated multiple malformations [3]. Less than half of our patients (12 out of 28) benefited from an obstetric ultrasound with only 8 prenatal diagnoses out of 12 ultrasounds performed. In the literature, low birth weight (less than 2.5 kg) and prematurity is a factor of poor prognosis [4]. In our series, no case of prematurity was found with 3 cases (10.71%) weighing less than 2.5 kg. The integrity of the omphalocele sac after childbirth determines the urgency of management; its rupture most often occurs after delivery and by vaginal delivery. The caesarean section rate in our series (25%) is low compared to developed countries where the development of antenatal diagnosis makes it possible to pose the obstetrical indication for caesarean section in the event of associated malformations or giant omphaloceles [5]. Rupture of the omphalocele membrane can occur antenatally or postnatally. The route of delivery does not seem to influence mortality or the rupture rate, although a selective caesarean section is not systematically recommended [4]. However, it has been described that vaginal delivery can be a cause of rupture of the omphalocele membrane, especially when the parietal defect is very large; therefore the subject remains debatable [6]. In our series, rupture of the sac was noted in 14.28% of cases, which is in line with the rates described in the literature, namely between 7 and 15% [4]. Omphalocele is an embryopathy in which the associated malformations are very common. The malformations most frequently encountered in the literature are cardiac (20 to 50%), digestive (40%) and genitourinary (40%) [7]. Associated malformations, in particular congenital heart disease, omphaloceles syndromes and chromosomal aberrations, are the main factors that condition the vital prognosis [7]. This is consistent with the data from our study, with a predominance of heart defects (42.85%). Specific cases of polymalformative syndromes have been described associating giant omphalocele with Wiedemann-Beckwith syndrome [8] or PRUNE-BELLY syndrome. [9], which was the case in two of our patients. The reintegration of exteriorized viscera into the insufficiently developed abdominal cavity has hemodynamic and respiratory consequences that must be considered for the surgical decision. Immediate parietal closure by simple parietal approximation is very risky. It can affect the hemodynamic and respiratory status of the newborn [10]. Two main techniques have been described in the literature for the treatment of these particular forms of omphalocele: the first, "paint and wait", consisting of repeated dressings by applying special infectious agents and delayed closure of the parietal defect [11]. The second technique proposed is the progressive reintegration technique with all its variants [2]. This technique was performed in all our patients with conservation of Wharton jelly and application of honey in half of the cases. Preservation of the amniotic membrane has the major drawback of not being able to carry out intraoperative exploration of the abdominal cavity in search of associated malformations, in particular intestinal malrotation which may be complicated by volvulus [12] or Meckel's diverticulum, which is more frequent in small omphaloceles [13]. 2 cases of Meckel's diverticulum were found in our series and the cure was made with a good clinical evolution. Complications of giant omphaloceles are dominated by respiratory distress and infections [4]. These complications can be directly life-threatening, such as infection (50% of our patients), which was responsible for the majority of deaths in our series. With an identical number of cases (14) treated as well by the classic

Schuster technique than that modified by preservation of Wharton jelly and the application of honey, the death rate is lower for the latter (4 out of 14 for the modified Schuster against 12 out of 14 for the classic Schuster). This could be explained by the anti-infectious activity and the acceleration of healing of honey in the dressings [14]. Conservation of the amniotic membrane would also reduce the death rate; the choice of this attitude in the modified Schuster technique should be motivated by the low frequency of malformations associated with intraoperative diagnosis. This amniotic membrane is also a mean of protection against infections and postoperative adhesions [15].

V. Conclusion:

The treatment of giant omphaloceles is still a topical issue. The progressive silo reintegration technique (Schuster) is the most widely used. Despite progress in neonatal resuscitation, this technique is fraught with complications. Our experience with the use of the modified Schuster technique, the application of honey in the dressings and the conservation of Wharton jelly, has clearly improved the survival rate of our patients in the treatment of giant omphaloceles.

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