Considerations on the results of the gastroschisis treatment

- summary -

The newborn with abdominal wall defects is one of the most dramatic situations in medicine, but at the same time it is a challenge for pediatric surgeons. This paper presents fundamental aspects related to one of the most common abdominal wall defects (the gastroschisis) including the prenatal and postnatal care principles and options. Although the gastroschisis is always grouped with the omphalocele, these are two separate and distinct entities with many important differences in terms of pathology and associated conditions, this explaining the differences in therapeutic approach and results. It is essential to understand the similarities and differences between the gastroschisis and omphalocele for the proper management of the newborn.

Although the primary closure of gastroschisis is now possible in many cases various strategies and materials have been described to cover the abdominal viscera for viscera protection and the reduction of heat and liquid loss in newborns with primary irreducible gastroschisis. There have been suggestions to cover the defect with skin allografts, lyophilized dura mater, free corium transplantation and meshed autografts.

Probably the most important therapeutic aspect of treatment for congenital malformations is the abdominal wall in taking care of the case. Given the possibility of antenatal diagnosis discussions were carried out regarding the preferred method of birth but no positive influence in evolution could be proven for a cesarean delivery in order to prevent a mechanical damage of the exteriorized intestinal loops as compared to natural birth (47). But the early identification of possible antenatal complications and of fetal distress that would allow a better determination of the indications for cesarean delivery is important. The birth of these children before the term was proposed in order to limit the intestinal lesions through the prolonged contact with the amniotic fluid but no benefit was demonstrated. It is essential to reduce the time between birth and reintegration of viscera.

Due to fetal echography the diagnosis of such abnormalities can be treated by surgery; it is performed prenatal is a growing number of cases. This allows planning birth in a specialized unit and prenatal counseling of parents regarding prognosis and postoperative outcome.

The therapeutic principles for the correction of gastroschisis remain the same, viscera reintegration and the primary fascial defect closure attempt or the progressive reduction achieved primarily through a silastic net (Silon).

Surgical treatment has evolved a lot since the first therapeutic successes of Grob, Gross and Schuster. There remain cases where the reduction is deemed unsafe, physically impossible due to the viscero-abdominal disproportion or dangerous because of the abdominal compartment syndrome.

The use of umbilical cord as autologous material has proved to be useful in the abdominal wall defects since the early 1970s. As compared to the autologous skin grafts, the isografts and the silastic nets the human umbilical cord was associated with the lowest inflammatory reaction, the lowest incidence of wound dehiscence, a shorter healing time and the highest rate of survival by repairing the abdominal wall defect created experimentally in rats.

The umbilical cord patch material is a "live" from the same patient. The symphysis between the umbilical patch and the defect edge or the subcutaneous tissue and the skin if the defect was extended laterally it heals completely within a few days. On the other hand this natural membrane prevents adhesions between the intestine and the epithelial umbilical patch.

Where it is possible to keep the umbilical cord it is recommended to use this technique because of its availability, its performance with no difficulty and the perfect tolerance of the autologous tissue. The infection rate is low mechanical complications are less frequent.

The work was performed in the Pediatric Surgery Clinic of "S. Marie Curie" Hospital - Bucharest and in Constanta County Hospital managed by Professor Doctor Tica Constantin.

The work includes a statistical clinical study conducted during 2002-2012. The study was conducted on 55 newborns with gastroschisis.

The following parameters were evaluated: the preoperative preparation of the newborn, the gestational age, the weight at birth, other associated malformations, the surgical technique, the hemodynamic tolerance after the per primam closure, the parenteral nutrition, the peri-operative and postoperative intra-abdominal pressure, the mechanical ventilation, the postoperative scar healing, the length of the hospitalization.

The purpose of the work is to present an alternative method to repair the gastroschisis where there is a disproportion between the size of the eviscerated viscera and that of the hypoplastic abdominal cavity, which can cause a significant increase in intra-abdominal pressure. When the complete primary closure was not possible the umbilical cord patch was used. A mesothelial surface is created in contact with the intestine. The use of autologous material such as the umbilical cord has

several advantages including availability, a lower rate of infections and significantly reduced costs.

The remote tracking of these patients showed excellent results.

The 55 cases of gastroschisis were divided as follows: 19 girls and 36 boys.

The gestational age of newborns with these abdominal wall congenital defects was between 33 and 38 weeks.

The weight at birth ranged between 1500 g and 3300 g.

The average age of mothers who gave birth to children with gastroschisis was 25 years (ages between 16 and 37 years old).

From the history of mothers whose children were born with gastroschisis the aspirin use was noted in 37 cases, of ibuprofen in 28 cases, alcohol in 31 cases, cigarettes in 42 cases and drugs in 2 cases.

Out of the 55 cases of gastroschisis 34 cases were diagnosed antenatal due to echography and the diagnosis was confirmed at birth.

Birth was performed in all these cases by cesarean. The remaining 21 cases were unsupervised pregnancies and birth was vaginal.

Among the newborns with gastroschisis 5 cases were associated with intestinal atresia and one case was associated with ventricular septal defect and it died 4 days after surgery because of a subarachnoid hemorrhage.

In 32 cases the per primam abdominal wall closure was attempted when the volume of the abdominal cavity allowed the reintegration of the intestinal loops with no tension, without increasing the intra-abdominal pressure or the ventilatory pressures. The intra-abdominal pressure was monitored intravesical or intragastric, the amount exceeding 20 cm H_2O to avoid the abdominal compartment syndrome. It was necessary to increase the parietal defect for the reintegration of the eviscerated loops in 13 cases.

In 23 of the 55 patients the per primam abdominal closure was not possible (significant intestinal edema in 13 cases, small abdominal cavity in 10 cases) and it was decided to practice the closure with the following variants:

- Umbilical cord patch at the level of the aponeurotic defect in 9 cases;
- Gradually, over several days through a silastic net ("silo") attached to the ends of the aponeurosis în14 cases. This "silo" was closed progressively from top to bottom thus favoring the gradual reintegration of the viscera in the abdomen while the intestinal edema regressed. The average time to complete the closure of the wall was 7 days.

The causes of the per primam abdominal closure failure in 23 patients with gastroschisis out of the 55 patients were the following: significant intestinal edema in 13 cases, namely small abdominal cavity in 10 cases.

The duration of the parenteral nutrition ranged from 7-39 days, especially in those with low gestational age and low weight at birth (1500 g - 1700 g) and in those where the intestinal edema was large.

The duration of the enteral nutrition was less than 31 days in 25 of the newborns with gastroschisis, between 31 - 60 days in 13 of the cases and it was greater than 60 days in the remaining 17 cases.

The duration of the mechanical ventilation ranged between 3 days and 13 days as a result of the increased pressure limiting the diaphragmatic excursions during respiration.

The average length of the hospitalization was 6.6 weeks (range: 23 -135 days).

No infections were registered caused by the umbilical cord patch or the post-surgery wound dehiscence.

Regarding mortality there were 19 deaths of newborns with gastroschisis. One of the deaths is that of a newborn who had a malformation associated with ventricular septal defect. The death occurred 4 days after the surgery by subarachnoid hemorrhage. Other 3 deaths occurred in newborns with gastroschisis who had intestinal atresia as associated malformation. The remaining 15 deaths occurred in newborns that had no other associated malformations. Regarding the mortality of newborns with gastroschisis it seems that depending on their weight, the lower the weight the higher the mortality is, so that 7 of the newborns weighing between 1500 and 2000 g, 5 of those weighing between 2000 and 2500 g, 4 of those weighing between 2500 and 3000 g and only 3 of those weighing at birth between 3000 and 3500 g died. Depending on the type of surgery chosen to treat the gastroschisis the mortality was as follows: out of the 19 deaths 8 occurred in newborns with gastroschisis who underwent the per primam abdominal wall defect closure without a net; out of those who underwent the parietal defect closure with a net 5 have died; in the case of those with progressive closure with Silo bag the number of deaths was 5; out of those who received the umbilical cord patch to close the parietal defect there is only one death.

The primary closure results are more impressive and much better as compared to the procedure with Silon bag or the use of skin flaps, lyophilized dura mater and other techniques to cover the defect and the intestine. The primary fascial closure is possible in 90 % of infants and the mortality rate is about 10%. A tight seal during the primary fascial repair can increase the intra-

abdominal pressure and lift the diaphragm with an increased need for respiratory support. On the other hand the morbidity of assisted mechanical ventilation must also be accepted. In some patients the direct pressure on the reintegrated intestine and the blood perfusion in the superior mesenteric artery diminished in the intestinal wall can cause ileal perforation or diffuse necrosis of the intestine and death. Another intra-surgical problem that can occur during the primary closure is the collapse – the obstruction of the inferior vena cava with severe cyanosis of the legs. In some of these patients another early surgical intervention is necessary and the use of silo bag technique. The morbidity of the silo bag technique is related to sepsis, enteric fistula, prolonged ileus and the need of several surgeries. Silo bag often results in an infection along the suture line. No symphysis can be created between the silo and patient tissues.

The gastroschisis repair method using the umbilical cord patch is a combination of the procedures used in the primary closure and the patch technique. Almost all intestine is reduced into the abdominal cavity except for a few loops, which will be covered with the umbilical cord patch. This technique was performed only in newborns with gastroschisis that could not be closed per primam. Thus the respiratory status compromise, the venous return and the intestine ischemia were avoided. Continuous intra-surgery monitoring of the intravesical pressure was used to prevent abdominal compartment syndrome. The maximum allowed value is 27 cm H_2O.

The umbilical cord patch is a "live" material from the same patient. The symphysis between the umbilical cord and the edge of the defect or of the subcutaneous tissue and skin where the defect was extended laterally heals completely within a few days. No infection occurred in the suture. On the other hand this natural membrane prevented the formation of adhesions between the intestine and the epithelial umbilical cord patch. A ventral hernia is formed which is usually small and easy to correct.

Experience in dealing with omphalocele and gastroschisis treatment allows shaping the following conclusions:

- The omphalocele and gastroschisis are serious congenital defects with still a very high mortality rate in our country, especially the gastroschisis which has an average survival rate still below 50%.
- Although belonging to the same group of abdominal wall malformations called medium celosomia have different embryological origins, specific clinical aspects, completely different prognosis and evolution from one another.

- These two types of abdominal wall malformations are often accompanied by other malformations more or less serious which can have a negative influence on the short-term or long-term prognosis of the patient.
- Associated malformations occur more frequently in the case of the gastroschisis which apparently contradicts the literature but is explained by the fact that the gastroschisis having a higher death rate these malformations were detected necroptically, while in the case of the omphalocele, especially the big ones that were treated conservatively such associated malformations compatible with life went unnoticed.
- The most frequent associated malformations were the digestive tube malformations that accompanied the abdominal wall defects and among these the common mesentery was the most frequent.
- The cardiac malformations have occupied quite a significant place regarding frequency and severity, the atrial septal defect interatrial communication was the most frequent of these.
- Among the genitourinary malformations the most frequent was the hypospadias.
- Prematurity is an element that darkens the prognosis of these malformations.
- A screening of the pregnant women in the gestational weeks 12-20, 32-34 is mandatory in order to assess fetal morphology. Thus these congenital anterior abdominal wall defects can easily be diagnosed and the appropriate therapeutic conduct can be established starting with advising parents by a complex team (obstetrician, neonatologist, pediatric surgeon, psychologist) regarding the prognosis and the possible development of the malformation and planning birth in a tertiary center.
- Amniocentesis should be performed between the 14th and 16th week of gestation only after a previous echography, and this can also be a useful antenatal diagnosis element.
- The dosing of the alpha fetoprotein in the amniotic fluid during the second trimester of pregnancy may be useful for the detection of both the omphalocele and the gastroschisis.
- Planned birth will significantly reduce the intestinal loops edemation and transformation of the intestinal loops especially in gastroschisis facilitating the primary closure.

- Regarding the birth method caesarean section is considered the most useful in improving the survival of these newborns by reducing the intestinal loops trauma.
- A good preoperative preparation of these patients in a neonatal intensive care compartment as well equipped as possible is necessary in order to facilitate where appropriate the primary closure of the wall defect with an intra-abdominal pressure as low as possible.
- If the case of the gastroschisis the weight and appearance of the herniated loops decide whether to carry out a primary or secondary closure.
- The infection rate is low and the mechanical complications are less frequent.
- There is only one drawback: the umbilical cord patch should be built into the operating room.
- The primary closure of the umbilical cord patch was applied successfully in all newborns with gastroschisis and the postoperative intravesical pressure in these newborns didn't exceed 20 mmHg.
- The total parenteral nutrition (TPN) in the postoperative period is very important for newborns with abdominal wall defects especially for newborns with gastroschisis that will not start the enteral nutrition for 14 days.
- Where it is possible to keep the umbilical cord we recommend this technique because of its availability, its performance with no difficulty and the perfect tolerance of the autologous tissue.