NEW THERAPEUTIC AND DIAGNOSTIC APPROACH IN GASTROCHISIS AND GIANT OMPhALOCELE

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ABSTRACT
During the past decade, significant changes have occurred in the management of diagnosis and therapy of the new-born with gastrochisis and omphalocele. Prenatal ultrasonography and measurement of maternal serum alpha-protein have been found to be useful in identifying infants with an abdominal wall defect (17,19).

We report two babies treated in our clinic, one with a fetal gastrochisis and the other one with giant omphalocele, antenatally diagnosed by ultrasound and measurement of maternal serum alpha feto-protein. Both the mothers were advised to undergo a cesarean section at 37-38 week’s gestation at a university maternity but their options were different. One followed the obstetrician’s advice and the baby survived but the other one was born by vaginally delivery, and inspite of all efforts, the baby could not be saved.

Key words: gastrochisis, omphalocele, antenatal diagnosis

Introduction
Prenatal diagnoses of gastrochisis and omphalocele requires adequate antenatal counseling depending upon the gestational age at diagnoses and association of other congenital anomalies (1,4,11). The medical team must discuss the various management options with the parents, which generally include pregnancy termination or delivery at a tertiary level center that has multimodality support available (2,7,16,20). Despite several decades of investigation and a vast array of new therapeutic approaches, the optimal therapeutic strategy for the newborn with a congenital defect of the abdominal wall remains undefined. A unanimous management of the delivery has not been established. For small omphalocele and gastrochisis the baby can be delivered normally at term in a center with pediatric surgeons. Large omphalocele should be delivered by cesarean section at a higher level tertiary center (1,4,18).

Case Reports
Case 1
A 3100 grams female baby, the second borne delivered by cesarean section at 38 week’s gestation at a local hospital. The mother had history of polyhydramnios antenatally and she had undergone serial ultrasound studies once a month since the 20th weeks gestation after the fetus was diagnosed with gastrochisis. She was advised by her obstetrician to borne the child at a hospital with an intensive care nursery and experienced pediatric surgeons. She complied with the doctor’s advice and came to Timisoara. The baby was referred from maternity at the age of 30 minutes, in an infant transport incubator, with the exposed intestines covered with a wet wrap. At the Pediatric Surgical Intensive Care Unit, a nasogastric tube was passed and was electively intubated as she developed respiratory distress syndrome. She was operated one hour after admission. The small intestine was returned easily to the abdomen so a primary repair of the gastrochisis was performed. During the operation and 6 hours postoperatively, the patient was intubated and mechanical ventilated. On the third post operative day, she was given parenteral nutrition and naso-gastric tube feeding was initiated slowly on the fourth post operative day. Unfortunately, on the 6th postoperative day, the child developed abdominal distension, the gastric aspirate increased and the baby did not pass stool, even with enema. Abdominal radiography showed an airless right lower quadrant and a few dilated loops of bowel. We reopened the abdominal cavity in order to reevaluate the situation. No intestinal atresia was found but a couple of intestinal adhesions. We performed adhesiolysis and inserted a drain in the peritoneal cavity. For three more days she was fed by total parenteral nutrition. Once bowel function returned as evidenced

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by passing of a bowel movement, feedings via the naso-gastric tube were slowly initiated with Gesol, while intravenous feeds continued. Progressively the volume of the mother’s milk was increased over the next 3 days and the naso-gastric tube removed. The baby was discharged when she took all her feedings by mouth (mother’s milk) and she had an adequate weight gain.(3200 g)

Case 2

A 3000 g male baby was born vaginally at 38 week’s gestation at a regional maternity centre. The diagnosis of a giant omphalocele was suspected on an antenatal ultrasound study at 28 weeks gestation. The diagnosis was confirmed by repeated ultrasound examination and high levels of the maternal serum alpha-protein. The family was suggested to choose one of the next two options:- termination of the pregnancy, because a cardiac malformation was suspected to be associated; or delivery by cesarean section at 27-28 week’s gestation at a university maternity centre, close to a Pediatric Surgery Clinic Department. The mother refused both the options and the baby was borne vaginally at a regional hospital, 200km far from our hospital. It was a prolongation of labour followed by the rupture of the omphalocele membranes, partial eversion and the contamination of the peritoneal cavity. The baby was transferred from this hospital in poor condition. The baby was diagnosed as Large omphalocele with ruptured membranes, partial eversion, materno-fetal infection and hypothermia.

At the pediatric Surgical Intensive Care Unit, a naso-gastric tube was passed, antibiotics were instituted and hydration was begun intravenously. At about 4 hours after admission he was operated. Intraoperative examination revealed an abdominal wall defect, 8 cm in diameter, with partial rupture of the membranes and eversion of the bowel ,the stomach and the right hepatic lobe. A primary surgical closure was impossible because it could result in increased intraabdominal pressure, so it was decided to use the Fufezan procedure-step1.We excised the borders of the ruptured membranes, restored two thirds of the visceria in the hypoplastic abdominal cavity, under a minimal pressure and sutured the borders of the remaining membranes to the border of the fascial defect.

During the operation and 36 hours after, the patient was intubated and mechanically ventilated.

Postoperatively, the recovery was good. The gastric aspirate diminished and the baby passed stool. Once the bowel function returned, the naso-gastric tube was removed.

After 7 days of life, the child’s condition started to deteriorate despite supportive measures. The baby had sepsis and the blood cultures grew Enterobacter. He died on the 13th day of hospitalisation due to Multiple Systemic Organ Failure. The parents did not give consent for postmortem examination.

Discussion

Gastrochisis and omphalocele are congenital defects of the abdominal wall that occur in approximately 1.75 to 2.5 in 10000 pregnancies (1,5,16). Gastrochisis occurs when there is evisceration of abdominal contents through a paraumbilical defect in the abdominal wall, usually to the right of the umbilical cord. It is less commonly associated with other abnormalities with the exception of intestinal atresia that occurs in about 10 percent cases. No genetic association exists and so, it has a better prognosis. On the other hand, omphalocele, a herniation of abdominal contents into the base of the umbilical cord, is more common and carries an increased risk for concomitant abnormalities and associated poor prognosis. However presence of major organ herniation such as liver and spleen makes the prognosis worse in both gastrochisis and omphalocele (7, 11, 15).

The diagnosis is usually made by antenatal ultrasound. The fetal abdominal cavity can be visualized on ultrasonography by 10 weeks following the mother’s last menstrual period.

Typically, omphalocele and gastrochisis can be diagnosed around the 13 weeks gestation when the normal return of the fetal intestine to the abdominal cavity have already occurred (3).

On ultrasonography, an omphalocele has a smooth outline with an echogenic covering sac, from which the umbilical cord is seen to arise. In contrast, gastrochisis is less smoothly contoured, has no echogenic covering and is seen at some distance from the umbilical cord (7,20).

Maternal alpha-protein levels are significantly elevated over normal levels and is a useful diagnostic test. In many cases, the diagnosis is made by ultrasound following an elevated second trimester maternal serum AFP test. Nowadays, a special management of the fetus with abdominal wall defect is necessary (3, 7, 11).

Termination of the pregnancy might be offered to those mothers who have a fetus with a large omphalocele or a fetus with multiple congenital anomalies. Continuation of the pregnancy needs a close observation and serial fetal sonography for appropriate advice to the parents. The prognosis largely depends upon the size of the defect and the presence or absence of other birth defects (13, 14, 20). The open fetal surgery and fetoscopic fetal intervention can be performed, only for selected cases, but not in our country.

Delivery at a tertiary center might be indicated so that a multidisciplinary team can immediately evaluate and stabilize the baby. The mode of delivery as well as the postnatal treatment approach depend upon the size of the abdominal defect and the degree of liver involvement. If
the defect is small and does not involve the liver, a vaginal delivery might be possible. In case of giant omphalocoele, cesarean delivery is usually required to avoid membrane rupture and liver trauma (13,14,21).

Fetal surgery is now available to selected infants diagnosed prenatally, as an experimental procedure at a few research centers in the world. The future of the fetal surgery is yet to be determined. Its success will depend on accurate identification of the fetus who has a poor prognosis and the demonstration of an increased survival with an in-utero surgery over conventional postnatal therapy for this subgroup of fetuses (3,14,18). Till than we have a long time, but we still believe that in the next few years, in our country, with better medical education and possibility to antenatally detect major congenital malformations with appropriate management of the pregnancy in tertiary centers, we will be able to improve the prognosis of affected infants (4,19).

Conclusions
The benefits of prenatal diagnosis are as follows:
- Prenatal diagnosis determines the outcome of pregnancy
- It is helpful for couples to decide whether to continue the pregnancy
- It indicates possible complications that can arise at birth process
- Prenatal diagnosis is helpful for the management of the remaining weeks of pregnancy
- It prepares the couple for the birth of a child with an abnormality.

REFERENCES
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