CLINICAL STUDY

Prenatal Diagnosis, Associated Findings and Outcome of Fetuses With Abdominal Wall Defects

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Abstract

Objective: To present a retrospective analysis of the prenatal diagnosis and the outcome of fetuses diagnosed with abdominal wall defects at the University Hospital, Department of Obstetrics and Gynecology, Lübeck, Germany.
Materials and Methods: We reviewed the medical records and ultrasound data of cases diagnosed with abdominal wall defects from January 1993 to December 2005. We described the prenatal diagnosis, associated findings and outcome of these patients.
Results: Hundred-and-fourteen patients were diagnosed with abdominal wall defects. Of these, 69 had an omphalocele, 38 presented with gastroschisis and 7 were classified as body stalk syndrome. None of the fetuses with body stalk syndrome survived. Of fetuses with omphalocele, an abnormal karyotype was found in 23 cases, most frequently being trisomy 18 (17 cases). Associated anomalies in fetuses with omphalocele and normal karyotype were found in 26 cases. Of fetuses with gastroschisis, 97.37% had a normal karyotype. The average length of hospitalization was longer for infants with gastroschisis. In fetuses with gastroschisis, serial ultrasound examinations allowed follow-up of the intestinal status and early recognition of intestinal atresia/stenosis through dilation of intra-abdominal intestinal loops.
Discussion: Abdominal wall defects are common fetal anomalies recognizable by ultrasound examinations during pregnancy. The outcome of fetuses with gastroschisis was shown to depend mainly on the intestinal status, while for infants with omphalocele, the associated anomalies were the most important factors in surviving. Fetal karyotyping should be offered in cases with abdominal wall defect. Interdisciplinary prenatal consultation of the neonatologist and the paediatric surgeon will favourably influence the perinatal outcome.

Keywords: fetal abdominal wall defects, omphalocele-gastroschisis-body stalk syndrome, associated fetal anomalies, prenatal diagnosis

Özet

Prenatal Karın Duvarı Defektleri ve İlgili Bulgular Tanımlanan Fetusta Sonuçlar

Amaç: Lübeck (Almanya) Üniversite Hastanesi, Kadın Hastalıkları ve Doğum Bölümü'nde prenatal karın duvarı defekti tanısı konulan fetuslarda sonuçların geriye dönük araştırmasını sunmak.

Materyal ve Metot: Ocak 1993'ten Aralık 2005'e kadar karın duvarı defekti tanısı konulan olayların tıbbi kayıtlarını ve ultrasonografik verilerini inceledik. Prenatal tanıları, birlikte elde edilen bulguları ve fetuslarda alınan sonuçları tanımladık.

Sonuçlar: Karın duvarı defekti tanısı 114 olayda konulmuş olup, 69'unda omfalosel; 38'inde gastroşizis; 7'sinde ise gövde sapı anomalisi saptanmıştır. Gövde sapı sendromu olan fetusların hiçbiri yaşamamıştır. Omfalosel olgularının 23'ünde anormal kromozom yapısı ve sayısı (karyotip) ve bunların 17'sinde 18. kromozomda trizomi bulunmuştur. Normal karyotipli omfalosel olgularının 26'sında birlikte başka anormal bulgular kaydedilmiştir. Gastroşizis olgularının %97.37'sinde kromozom varlığı normal görülmüştür;

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bunların ortalama hastanede tutulma süresi daha uzun olmuştur ve bağırsak durumunu takip için yapılan seri ultrasonoggrafilerle karın içindeki bağırsak ilmeklerinde genişlemelerle bağırsakta tıkanma veya daralmaların tanısı erken konabilmiştir.

Tartışma: Fetusta karın duvarı defektleri bilinen anomaliler olup gebelik sırasında ultrasonografi görüntülerinden tanınırlar. Gastroşizis durumunda fetusta alınacak sonuç öncelikle bağırsak durumuna bağlıdır. Ancak omfaloselde yaşamın sürdürülmesinde en önemli etken birlikte var olan öbür anomalilerdir. Karın duvarı defektinde fetusun karyotiplemesi yapılmalıdır. Neonatalog ve pediyatrik cerrahın prenatal konsültasyonu, doğum sonuçlarını olumlu etkileyecektir.

Anahtar sözcükler: fetusta karın duvarı defekti, omfalosel-gastroşizis-gövde sapı sendromu, birlikte fetal anomaliler; prenatal tanı

Introduction

Abdominal wall defects are common fetal anomalies, with an incidence of approximately 1 in 2000 live births. The major types of these malformations are gastroschisis, omphalocele and bladder and cloacal extrophy. Other less common abdominal wall defects include limb-body wall complex, ectopia cordis and urachal cyst. The incidence, etiology, pathology, and prognosis of these abdominal wall defects differ greatly, and also doing the postpartum management and outcome. With the advent of high resolution ultrasound devices examination of ventral abdominal wall, umbilical cord insertion site, urinary bladder and pelvis in the first or second trimester of pregnancy has become possible. Although the data regarding this pathology are very vast, there are contradictory results regarding the management of fetuses with abdominal wall defects concerning the mode and time of delivery, the intrauterine possibilities of correcting and the ways of postnatal surgical repair.

In this study, we retrospectively reviewed our experience with infants who were diagnosed with abdominal wall defects and to describe the prenatal diagnostic patterns, associated lesions and the short term outcome of fetuses with abdominal wall defects.

Materials and Methods

This is a retrospective study of all fetuses with abdominal wall defects diagnosed sonographically at Department of Perinatal Medicine-Clinic of Obstetrics and Gynecology at the University of Lübeck from January 1993 to December 2005. The following definitions were used to codify a case with abdominal wall defect:

Gastroschisis was defined as an anterior abdominal wall defect located lateral to a normal umbilicus but not involving the umbilical ring. The herniated viscera are often covered by exudate but no membrane covers the intestinal loops floating in amniotic fluid.

Omphalocele was defined as midline abdominal wall defect limited to an open umbilical ring. The intraabdominal organs herniated into the base of the umbilical cord and are covered by an amnioperitoneal membrane.

Body stalk syndrome was defined as a large abdominal wall defect, the fetus being attached to the placenta by sheets of amnion arising from the body wall defect.

The Clinic of Obstetrics and Gynecology Lübeck is a tertiary referral center for prenatal diagnosis and therapy. The ultrasound examinations were performed mainly for the routine first or second trimester screening by obstetricians specialized in perinatal medicine. After a positive diagnosis was made, the patient was followed-up regularly every 2-4 weeks. The following parameters were controlled: fetal biometry and weight, amniotic fluid index, Doppler indexes for uterine arteries and umbilical cord, the aspect of extra- and intra-abdominal intestinal loops. We reviewed the maternal and neonatal medical records. All diagnoses were confirmed at delivery or by the time of pathological examination. The indication of medical abortion was set in cases with chromosomal abnormalities or with severe, life-incompatible fetal malformations. Maternal information gathered included age, gravidity, maternal morbidity, prenatal complications, and associated risk factors. In this study, to evaluate the outcome of children with abdominal wall defects, only those patients who delivered or the pregnancy was terminated in our hospital were included. For these patients, we searched for the following parameters: gestational age at diagnosis and at delivery/medical abortion, birth weight, sex, karyotype, mode of delivery, antenatal and postnatal diagnosis, associated anomalies, and type of closure of abdominal wall defect. Primary neonatal outcomes included death, length of hospitalization, length of mechanical ventilation, length of parenteral nutrition, and postnatal complications. Results were tabulated and statistical analysis was performed using Student *t* test, χ^2, Mann-Whitney test as appropriate (individual measures were analyzed by univariate analyses, and logistic regression was used to identify significant factors for the length of hospitalization). A p value less than 0.05 was considered significant.

Results

There were 114 patients diagnosed with abdominal wall defects. Mean maternal age at delivery was 31.97±5.82 years for patients diagnosed with omphalocele, 24.86±5.39 for patients diagnosed with gastroschisis.

The other maternal parameters and outcome of fetuses diagnosed with abdominal wall defects are illustrated in Table 1.

The fetuses diagnosed with body stalk syndrome who were allowed to develop till term were from multiple pregnancies and the other co-twins were normally developed. One case was a trichorial, triamniotic multiple pregnancy obtained by intracytoplasmatic sperm injection (ICSI). The

Table 1. Maternal data of the studied population

	Omphalocele	Gastroschisis	Body stalk syndrome
No. patients	69	38	7
Maternal age (years±SD)	31.97±5.82	24±5.39	27.71±4.27
Gravidity			
G1	29 (42.03 %)	18 (47.37%)	4 (57.14%)
G>1	40 (59.97 %)	20 (52.63%)	3 (42.86%)
p			
Smoker/Drug consumer	7 (10.14%)	13 (34.21%)	-
p			
ART	5 (7.25%)	2 (5.26%)	2 (28.57%)
Pregnacy complications			
• Multiple pregnancy	3	1	3 (42.86%)
• IUGR	4 (5.8 %)	10 (35.71%)	
• Oligo/ Polyhydramnios	1 (oligo)/5 (poly)	2 (oligo)/3 (poly)	1 (oligo)
Outcome			
• Abortion	47	2	4
Spontaneous	8	2	-
Medical	39	-	4
• Delivered >24 weeks	20	35	3
• Postpartum death	4	3	2
• No records	2	1	-

anomaly was diagnosed during the ultrasound examination performed for the first trimester screening. The fetus had a complete eventration of abdominal content and ectopia cordis and as a consequence a selective fetocide was performed for the malformed fetus. The genetic analysis revealed normal karyotype. The pregnancy evolved without any complication till term. The patient delivered two healthy babies by caesarean section in the 40th week of gestation. For the other 2 cases of body stalk syndrome, the diagnosis was set in the second trimester. In both situations, the patients delivered prematurely, in the 29th and respectively 33rd gestational week. Both affected fetuses died very soon after birth. The 4 cases of body stalk syndrome terminated by abortion were single pregnancy, diagnosed during the 12th and 16th gestational week. In one case, the karyotype showed a mosaic for trisomy of chromosome 2. The parents opted for medical termination of pregnancy due to poor prognosis.

During 1993-2005, 69 fetuses were diagnosed with omphalocele.

Genetic investigations were done in 55 cases. Thirty-two fetuses had normal karyotype (after delivery/abortion, another 12 fetuses had a normal karyotype), and 23 cases had abnormal karyotype (17 cases of trisomy 18, 2 cases of trisomy 13, one case of Turner syndrome, one case was diagnosed with muscular dystrophy (mother was carrier of the chromosomal mutation), 1 case was a 46 XY with balanced translocation between chromosome 14 and 21, and one case was a mosaic between 46 XX and 92 XXXX).

Table 2. Associated anomalies of fetuses diagnosed with omphalocele

	Abnormal karyotype (23 cases)	Normal karyotype <24th week (25 cases)	Normal karyotype ≥24th week (19 cases)
Increased nuchal translucency	7	4	1
Nonimmune hydrops	7	2	-
Cardiac malformations (septal defects, tetralogy of Fallot, ectopia cordis)	8	6	5
Central nervous system anomalies	7	4	-
Other anomalies			
Skeletal	1	3	2
Genitourinary	2	1	-
Facial (cleft lip, micrognathism, proboscis, cyclopia)	3	-	1
Ascites	-	2	1
Intrauterine growth restriction (IUGR)	3	1	3
Single umbilical artery	5	3	3
Cantrell pentalogy	-	3	-
Meckel-Gruber syndrome	-	1	-
Multiple pregnancy - thoracopagus*	1	1	-
Beckwith-Wiedemann syndrome	-	-	1
Oligo/Polyhydramnios	-	3	4

* The fetuses from multiple pregnancies, beside the abdominal wall defect (omphalocele), had also thoracopagus.

Table 3. Perinatal and neonatal outcome of fetuses with omphalocele

	Gestational age at delivery	
	<37 weeks (n=4)	≥37 weeks (n=11)
Mode of delivery		
Vaginal	-	2 (18.18%)
Caesarean section	4	9 (81.82%)
Mean gestational age	30.75±4.76 w	37.45±1.16 w
Fetal gender		
Male	1 (25%)	8 (72.73%)
Female	3 (75%)	3 (27.27%)
Birth weight	1613.75±689.92 g	3040.45±409.97 g
Associated major anomalies	2 (50%)	3 (27.27%)
Polyhydramnios	1 (25%)	2 (18.18%)
IUGR	2 (50%)	1 (9.1%)
Mortality	2 (50%)	2 (18.18%) *p*=0 .01
Mode of closure		
Primary fascial closure	2 (50%)	10 (90.9%)
Secondary fascial closure	1 (25%)	1 (9.1%)
Length of hospitalization	44.75 d	34.45 d *p*=0.86
Postoperative complications		
Bowel obstruction/Necrosis	1 (25%)	2 (18.18%)
Sepsis	1 (25%)	-
Necrotising enterocolitis	1 (25%)	-
Wound dehiscence	-	2 (18.18%)
Cardiovascular	-	1 (9.1%)
Reintervention	1 (25%)	4 (36.36%) *p*=0.01

The diagnosis was made during first (43 cases, 63.24%) or second trimester (23 cases, 33.82%). The average gestational age at diagnosis was 15 weeks. Only two cases were diagnosed in the third trimester. One of the fetuses with trisomy 13 was diagnosed in the 33rd gestational week. In this situation a fetocide was performed after the genetic result. The second case diagnosed in the third trimester (36th gestational week) had a normal karyotype. In 8 cases, the pregnancy evolved in a spontaneous or missed abortion, in 39 cases was indicated medical termination of pregnancy due to poor prognosis, and in 20 cases, the fetus was delivered after the 24th week of gestation.

Associated structural anomalies of fetuses with omphalocele are illustrated in Table 2.

Only 15 fetuses diagnosed with omphalocele were delivered after 24th week of gestation at University Clinic Lübeck and were included in our study regarding perinatal outcome. Table 3 presents main parameters and outcome of fetuses with omphalocele.

The mean gestational age at delivery was 35.67±3.97 weeks, and the birth weight was 2660±805.13 grams. In 5 cases (33.33%), the content of omphalocele was represented by bowel, and for the other 10 cases (66.67%), the omphalocele contained liver with varying amounts of small intestine. In only one case, stomach was also herniated beside liver. The average diameter of abdominal wall defect at delivery was 4.375 cm (2-8 cm). In 12 cases (80%), a primary fascial closure was performed. In 2 cases (13.33%), the omphalocele repair was performed by secondary fascial closure. The premature infant delivered in the 26th week of gestation was not operated due to the poor status. All the other 3 infants who had severe heart malformation (double outlet right ventricle, tetralogy of Fallot, and anomalies of caval veins with pulmonary hypertension and atrial septum defect type II) died. The average length of hospitalization was 38 days, although the difference was not statistically significant for children delivered by caesarean section. Only five children necessitated reintervention, in one case for intestinal

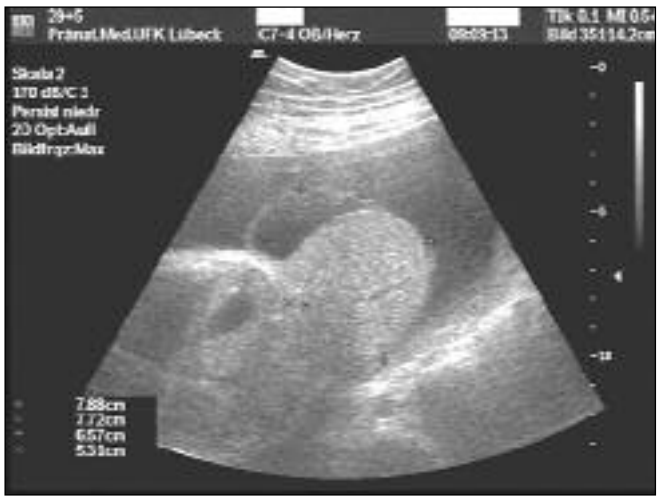


Figure 1. Sonographic image of fetal omphalocele with liver and bowel content at 29±5 gestational weeks.

Table 4. Perinatal and neonatal outcome of fetuses with gastroschisis

	Gestational age	
	<37 w (n=18, 64.29%)	**≥37 w (n=10, 35.71%)**
Birth weight (g)	2024.67±463.08 g	2842.5±813.43 g
Gestational age (mean)	33.67±2.83 w	37.2±0.42 w
Fetal gender		
Male	6 (33.33%)	4 (40%)
Female	12 (66.67%)	6 (60%)
Mode of delivery		
Vaginal delivery	2 (11.11%)	-
Caesarean section	16 (88.89%)	10 (100%)
Associated anomalies/Complications		
Intestinal atresia	5 (27.78%)	-
Oligo/Polyhydramnios	3 (16.67%)	2 (20%)
Meconial stain of amniotic fluid	6 (33.33%)	1 (10%)
Pathological FHR	1	-
Microcephaly	1	-
Heart malformations	1	1 (10%)
Intrauterine growth restriction	7 (38.89%)	3 (30%)
Arthrogryposis congenita	-	1 (10%)
Undescended testis	1	1
Mortality	2 (11.11%)	1 (10%) p=0.04
Mode of closure of abdominal wall defect		
Primary closure	13 (76.47%)	9 (90%)
Secondary closure	4 (23.53%)	1 (10%) p=0.095
Days of ventilation	3.67 d	2.11 d p=0.12
Time to full enteral feed	45.31 d	29.14 d p=0.05
Length of hospitalization	59.8 d	29.11 d p=0.04
Bowel obstruction/Perforation (at delivery)	6 (33.33%)	- p=0.011
Postpartum complications		
Gastrointestinal dysfunction		
Bowel obstruction	8	-
Short bowel syndrome	5	-
Gastroesophageal reflux	2	-
Complications secondary to parenteral nutrition	6	1
Necrotizing enterocolitis	2	1
Sepsis	13 (72.22%)	3 (30%) p=0.022
Arterial hypertension	1	1
Clonic seizure	1	-
Hernia/Eventration	3	2
Reintervention	13 (72.22%)	2 (20%) p=0.018

obstruction due to adhesions, in one case due to necrotising enterocolitis and bowel necrosis, in one case for eventration, and in two cases due to wound dehiscence. Regarding Doppler parameters, only one case presented increased pulsatility and resistance index in umbilical arteries in the 41st week. The outcome of child was good. Figure 1 illustrates the typical sonographic aspect of fetal omphalocele with liver and bowel content.

During January 1993-December 2005, 38 cases of gastroschisis were diagnosed in Department of Perinatal Medicine, University Clinic Lübeck. Of these, only 28 fetuses were delivered alive in our clinic after the 24th week of gestation. The characteristics of these fetuses are presented in Table 4. Four of the fetuses diagnosed with gastroschisis at Department for Perinatal Medicine of Uniklinik Lübeck were delivered in other hospitals and were not included in this study due to the lack of data. Two cases of gastroschisis were spontaneously aborted in the 16th, and respectively in the 19th gestational week. In the first case, beside gastroschisis, there were other anomalies detected: hydrocephalus, ectopia cordis, cleft lip and palate, and clubfoot. The second case was a multiple pregnancy obtained after ICSI. One of the gestational sacks was a blighted ovum, and the other fetus presented with gastroschisis and severe growth restriction at 19 weeks of gestation. The pregnancy ended in spontaneous abortion three days later. Compared to fetuses with omphalocele, only one case of gastroschisis had an abnormal karyotype

(trisomy 18) with other anomalies associated: double-outlet right ventricle, rocker-bottom feet and growth restriction. The pregnancy was terminated in the 33rd week. In two cases of gastroschisis, an intrauterine death occurred in the third trimester, due to placental abruption. In another case of gastroschisis, the pregnancy was terminated in the 26th week of gestation due to the associated anomalies with poor prognosis (severe kyphoscoliosis, hypoplastic lungs and growth restriction). The pregnancy was obtained after ovarian stimulation.

The average gestational age at diagnosis was 22 weeks. Only one case of gastroschisis was diagnosed in the first trimester (13th gestational week) (3.57%), 22 cases were diagnosed in the second trimester (78.57%), and 5 in the third trimester (17.86%). The mean gestational age at delivery for all infants with gastroschisis was 34.9 weeks. Only two children (7.14%) were born vaginally, both being delivered prematurely and in only one term pregnancy a vaginal delivery was attempted which ended in a caesarean section due to pathological fetal heart rate (FHR). In 7 cases, a caesarean section was performed before 37 weeks due to severe bowel dilatation. In all other cases delivery was by elective caesarean section. Associated anomalies occurred infrequently, the most severe were: two cases with congenital heart malformations (atrial septal defect, double outlet right ventricle), one case with microcephaly and one case with arthrogryposis multiplex congenita. Increased pulsatility and resistance indexes in umbilical arteries were present in 8 cases (21.62%), of which in one case the pregnancy ended in stillbirth and in one case, the newborn died postpartum.

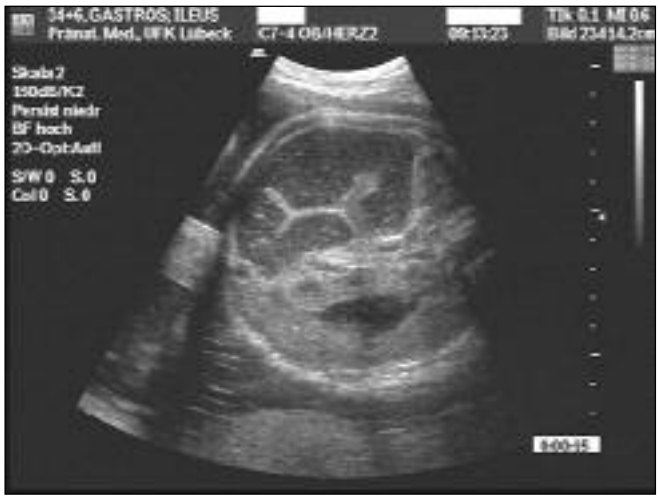
All patients were intubated immediately after birth and 22 (81.48%) infants underwent an attempted primary closure within the first 24 hours after birth. The type of surgical intervention was decided by the pediatric surgeon depending on the size of the abdominal wall defect and the status of herniated bowel after inspection (intestinal atresia, necrosis, perforation necessitating also intestinal resection, stoma and reanastomosis). The average diameter of abdominal wall defect at birth was 3.5 cm (1-7.6 cm), the smaller the defect the more severe intestinal complications were noted. In 71.43% of the patients, the herniated organs included bowel only, whereas the distribution of other organs exposed included stomach (14.29%), bladder (7.14%), gonads and gall bladder (7.14%).

Postoperative complications included 8 (28.57%) cases of bowel obstruction, 7 necessitating reintervention and bowel resection. Short bowel syndrome was noted in 5 infants. Gastroesophageal reflux was observed in 2 children. Complications due to total parenteral nutrition were cholestasis, reactive hepatitis and cholangitis and were noted in 7 (25%) children. There were 16 (57.14%) cases of sepsis, one being lethal. Necrotizing enterocolitis was noted in 3 cases. Arterial hypertension was noted in 2 infants, and clonic seizure in one premature female infant. One child was diagnosed with cystic fibrosis.

The mean interval from delivery to complete oral feeding was 43.69±34.64 days (range 10 to 150 days). The mean hospital stay was 49.58±36.31 days (range 11 to 154 days). The difference of mean hospital stay between children with gastroschisis and children with omphalocele was not statistically significant (for infants with omphalocele the median hospital stay was 38 days; p=0.15, Mann-Whitney test).

Table 4 compares also the outcome of fetuses delivered prematurely and the cases delivered after 37th week of gestation. As mentioned in previous articles (20-23), premature delivery of fetuses with gastroschisis has no beneficial effects regarding the outcome. In fact, premature babies are prone to complications, especially gastrointestinal and infection. In our cases, the indication for premature delivery was due to premature rupture of membranes in 2 cases and in 7 cases was due to the ultrasonographical aspect of herniated organs (dilated intestinal loops with thick wall and "peel").

Three fetuses with gastroschisis died postpartum. In one case, it was a very premature newborn in the 24th week of gestation, delivered due to premature rupture of membrane. The child survived only a few hours and was not operated. The other two cases survived 9, and 2 months respectively. Both children were delivered by caesarean section in the 36th week, and



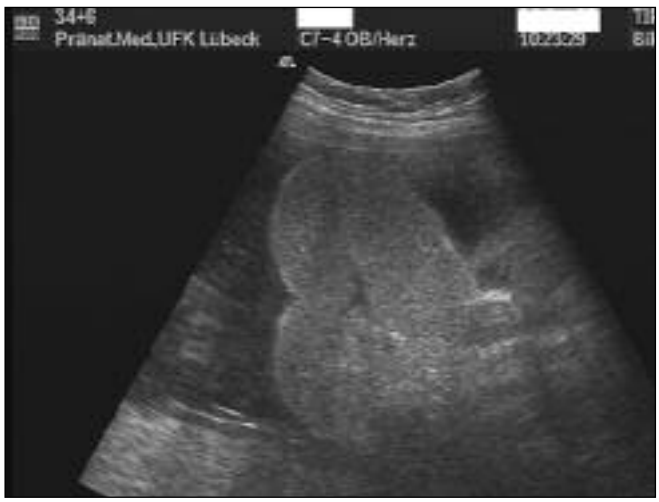


Figure 2 and 3. Sonographic image of fetal gastroschisis with dilated intra-and extraabdominal bowel loops in the third trimester.

respectively in the 38th week of gestation. The first child presented growth restriction (birth weight <5th percentile), and intestinal atresia. Postoperatively, the child developed short-bowel syndrome, nephrocalcinosis and hypertension, and finally sepsis, which was the cause of death. The second child presented oligohydramnios, arthrogryposis multiplex congenita. The child died suddenly due to intestinal obstruction secondary to adhesions which evolved very fast in a generalised peritonitis.

The fetuses were examined every 2 weeks for evidence of bowel obstruction. Dilatation of intra-abdominal bowel was considered as a sign of bowel obstruction and was presented in 8 cases (Figure 2 and 3).

Of these, seven had intestinal obstruction or perforation at birth ($p<0.05$). In 3 cases, the findings were observed before the 34th week of gestation and delivery was recommended. In one case of intestinal atresia, the fetus died intrauterine in the 34th week of gestation.

Discussion

This study compares the outcome of children with abdominal wall defects. Body stalk syndrome is a severe association of anomalies with poor prognosis. In our series, this pathology was noted more often in multiple pregnancies. All of the fetuses diagnosed with body stalk syndrome didn't survive.
Gastroschisis and omphalocele are the most frequent abdominal wall defects (1). While gastroschisis appeared more likely as an isolated defect, omphalocele is more often associated with chromosomal abnormalities (10-30% of cases) and other anomalies (30-70% of cases) (2-7). In our series of patients, gastroschisis was found to be isolated in 81.58% of cases. In only one case (2.63%), the karyotype was abnormal (trisomy 18). Of the 68 fetuses with omphalocele, there were 23 (33.82%) cases with abnormal karyotype (mainly trisomy 18) and other 25 (36.76%) cases with severe anomalies which necessitated termination of pregnancy.

As previous reported gastroschisis is more frequent in younger women, especially teenager pregnancies, while omphalocele appeared to be more frequent at both age extremities (4, 8-11) In our cases, we found the median maternal age for the occurrence of gastroschisis lower than that of omphalocele (24 versus 31; $p<0.0001$).

In gastroschisis, the abdominal wall defect is usually relative small (2-4 cm in diameter), involves all layers of the ventral wall and permits the herniation of small and large bowel through the defect. The size of the defect and the exposure of intestinal loops to amniotic fluid have negative consequences regarding the function of bowel, inducing significant morbidity and mortality to the fetus and newborn (12-14). Stillbirth rates of 12.5% (15) and neonatal mortality rates of up to 7.5% (16) have been reported. It may be associated with atresia/stenosis of the bowel in 10-20% of cases (17). Also, the incidence of intrauterine growth restriction is greater for fetuses with gastroschisis. In our series, we found an incidence of 35.71% of intrauterine growth restriction and 8 cases with severely dilated intra-abdominal intestinal loops (28.57%). The stillbirth rate for our series was 5.88%, and the neonatal mortality rate was 3.57%.

Congenital abdominal wall defects are one of the major causes of neonatal morbidity and mortality. Of paramount importance is distinction of gastroschisis from omphalocele and other abdominal wall defects, since the prognosis and management of these conditions are different (18). Other studies didn't prove any beneficial effect for fetuses with abdominal wall defect delivered by caesarean section (19-22). On the contrary, there were some studies which showed that infants with abdominal wall defect delivered in a hospital with pediatric surgeon available and who were operated in the first 6 hours after birth had a better prognosis than those delivered in a hospital and transported after birth for surgical correction of the defect (23). While for infants with gastroschisis, the condition of bowel at birth affects the postnatal prognosis, for fetuses with omphalocele, the outcome depends mainly on the associated anomalies, especially the cardiac malformations. As a consequence, systematic ultrasound examination including fetal sequential echocardiography is required in case of fetal abdominal wall defect and serial ultrasound scans are useful to follow-up the condition of bowel during gestation (2,23).

Bowel-related abnormalities are the most common and the most clinically important complications related to gastroschisis. Other anomalies occur infrequently to gastroschisis. Only minor cardiovascular malformations and amyoplasia were reported until now (24,25). Characteristic is the nonrotation of small bowel and absence of secondary fixation to the dorsal ventral wall. Intestinal stenosis or atresia is found in 7-30% of cases, secondary to intestinal and mesenteric ischemia. The ischemic lesions result either from compression of the mesenteric vessels by the relatively small abdominal wall defect, or from torsion of the herniated intestinal loops around the mesenteric axis.

As a consequence, the prenatal detection of intestinal obstruction in case of gastroschisis is also of importance, because it predicts neonatal bowel atresia and poorer outcome (17,26). However, we do not know when and how bowel atresia develops in fetuses with gastroschisis. In addition there are no animal models for the study of atresia with gastroschisis. For infants diagnosed with gastroschisis, it was observed that prenatal ultrasonographic evidence of bowel dilatation was significantly associated with postoperative complications (3,17). Near term, the ultrasound aspect of herniated bowel, exposed to amniotic fluid, often shows thick and matted wall, which corresponds at birth to the presence of a fibrinous peel. Complications such bowel atresia, perforation or meconium peritonitis can be identified by ultrasound. The presence of bowel obstruction/atresia is indicated by the progressive intra-abdominal bowel dilation, polyhydramnion and hyperechogenicity of herniated intestinal loops. Optimal perinatal management is necessary and requires immediately available neonatal medical, surgical and anaesthetic expertise.

Unfortunately, there are many limitations to the prenatal assessment of herniated intestine loops. Small bowel diameter varies with gestational age, and it is difficult to differentiate large bowel loops from severely dilated small bowel loops. We observed that the appearance of severly dilated intra-abdominal bowel is very suggestive for presence of intestinal stenosis/atresia. In our series, we found 8 cases with severely dilated intra-abdominal intestinal loops, necessitating immediately delivery of the fetus. In fetuses with gastroschisis, preterm delivery may be advisable in cases with increasing bowel distention, but the risks of prematurity must be weighed against the potential advantage of preterm correction of intestinal status.

A recent work of Nick et al. showed that intra-abdominal bowel dilation in the second trimester of pregnancy predicts neonatal bowel atresia and a poor postnatal outcome (27). They identified 58 fetuses with gastroschisis. Ten of these had intra-abdominal bowel dilation and all had bowel atresia at birth ($p<0.0001$). The authors also observed in 8 cases the presence of intra-abdominal bowel dilation before 25th week of gestation. They concluded that intra-abdominal bowel dilation in the second trimester predicts neonatal bowel atresia in fetuses with gastroschisis.

The use of ultrasonography permits detection of abdominal wall defects early in pregnancy, the follow-up of these fetuses and the decision regarding mode, time and place of delivery. Further work is needed to establish proper parameters for prenatal management of gastroschisis and to clarify whether progressive intestinal damage occurs throughout gestation secondary to atresia. If this would be the case, prenatal intervention may be useful to prevent such ongoing intestinal damage.

In all cases of abdominal wall defects, prenatal diagnosis should be followed by parental counselling by a multidisciplinary team consisting of obstetricians, neonatologists and pediatric surgeons.

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