Case Report

# Report of Three Cases of Unusual Anterior Abdominal Wall Defects in Fallujah General Hospital During the Year 2010

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## Abstract

Abdominal wall defects occur when the normal sequence of the gastro intestinal tract development is interrupted. Gastroschisis represents a herniation of abdominal contents through a paramedian full-thickness abdominal wall fusion defect without involving the umbilical cord. Evisceration usually only contains intestinal loops and has no surrounding membrane unlike omphalocoele. It is unusual for a newborn with gastroschisis to have other serious birth defects. Neonates with gastroschisis have better prognosis than those with omphalocele. Very rarely is gastroschisis associated with herniation of other organs, and their presence makes the prognosis worse.

I report three cases of unusual anterior abdominal wall defects during the year 2010, each having specific associated abnormalities. One was stillborn, and the other two died shortly after delivery.

**Key words:** Abdominal wall defect, gastroschisis, omphalocele, Fallujah, Iraq

## Introduction

The gastrointestinal tract develops between gestational weeks 4 to 16.[1] The embryonic foregut develops into the upper gastrointestinal tract, and the midgut will evolve to form the small intestine and up to two-thirds of the transverse colon. The third embryonic segment, the hindgut, will develop into the remaining transverse, descending, and sigmoid colon, as well as the rectum and anal sphincter. The midgut is a U-shaped structure that herniates into the primordium of the umbilical cord during the sixth week of embryogenesis. It rotates 90° in a counterclockwise direction before returning back to the abdomen at approximately gestational week 10. The midgut then rotates an additional 180° to form the small bowel and transverse colon. Any compromise of this sequence renders the fetus vulnerable to an abdominal wall defect.

Gastroschisis is a full-thickness defect in the abdominal wall that affects structures lying to the right of the umbilical cord insertion.[2] The amount of small bowel and other abdominal organs affected by a gastroschisis defect vary considerably. Unlike other abdominal wall defects, the abdominal structures that herniate in response to a gastroschisis are not covered by a membrane.

Gastroschisis is thought to result from an

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ischemic insult to the developing body wall.[3] Specifically, the right umbilical vein and right omphalomesenteric artery supply the developing gastrointestinal tract until they involute. Disruption of this process is hypothesized to result in ischemia of the abdominal wall and in severe cases, gastroschisis. The defect is usually small in size (< 2.5 cm in diameter).

Omphalocele is postulated to occur around 4 weeks of gestation when the lateral body folds move ventrally and fuse in the midline to form the abdominal wall.[1] Incomplete fusion of these folds is thought to cause a defect allowing protrusion of 1 or more loops of the bowel, sometimes accompanied by the liver or other abdominal organs. In contrast to gastroschisis, an omphalocele defect is covered by a peritoneal membrane on its inner surface, amnion on its outer surface, and an interposing layer of Wharton's jelly.[2] The umbilical cord inserts into this membrane at a location far from the abdominal wall. The defect can vary in size from a few centimeters to most of the anterior abdominal wall.

The incidence of gastroschisis is 0.4 to 3 per 10,000 live births, and the incidence of omphalocele is 1.5 to 3 per 10,000 live births.[4-6] The incidence of gastroschisis appears to be increasing, whereas the incidence of omphalocele appears stable. Factors associated with an increased likelihood of gastroschisis include young maternal age (mother aged 20 years or younger), cigarette smoking, use of illicit drugs, vasoactive over-the-counter drugs and environmental toxins.

Infants with omphalocele and gastroschisis are also at increased risk for other congenital anomalies. Infants with gastroschisis are at increased risk (20%) for gastrointestinal anomalies such as intestinal atresia, malrotation, volvulus or intestinal infarction but are not usually at increased risk for extragastrointestinal defects. They usually have normal karyotype.[3] On the other hand infants with omphalocele are at increased risk (50-70%) for cardiac defects, Meckel's diverticulum, and bladder exstrophy, etc. Up to 10% of omphalocele patients have Beckwith-Wiedemann syndrome resulting in macroglossia, organomegaly, early hypoglycemia, and a greatly increased risk of Wilm's tumor, hepatoblastoma, and neuroblastoma later in childhood. Thirty percent will have chromosomal abnormalities. The size of the omphalocele does not appear to influence the likelihood of additional anomalies.

## Case Reports

*Case 1*

A 3000 gm male was born in March 2010 in Fallujah General Hospital by spontaneous vaginal delivery, to a primigravida, aged 23 years and living in Fallujah city, Alshuhadaa district. She had normal, uneventful pregnancy with no history of hypertension or diabetes, no history of smoking, no exposure to X-ray irradiation, no history of any drug use other than supplemental vitamins and minerals. She is consanguineously married. There was no history of abortion and there was no family history of fetal anomalies. No fetal anomalies were detected on routine prenatal sonography. The baby was full-term and gasping and died within 30 minutes of birth. There was a small right periumbilical defect in the abdominal wall through which the small and large intestine were herniated (Figure 1). There was associated right thoracic cage abnormality (some ribs could not be palpated). No other abnormalities were detected. Hair samples from the mother have been examined for trace minerals in Microtrace Minerals Gmbh Environmental and Clinical Laboratories in Germany and showed high levels of bismuth (8.18 µg /g, acceptable range < 0.27 µg /g) and mercury (6.91 µg /g, acceptable vale <0.60 µg /g). The report is available as a supplementary file in the online version of this article.

**Figure 1. Herniation of small and large intestine through a small right periumbilical wall defect with associated right thoracic cage abnormality (case 1).**

*Case 2*

A 3500 gram male was born in November 2010 at Fallujah General Hospital by spontaneous vaginal delivery to a 23-year old primigravida living in Fallujah, Almuallimeen district. She had no history of diabetes or hypertension, smoking or exposure to X-ray. There was no history of using NSAIDs, aspirin, acetaminophen, pseudoephedrine or other known teratogenic drugs. There was no history of abortion and no family history of fetal anomalies. She gave history of regular antenatal visits. An anterior abdominal wall defect was detected on routine prenatal sonography. The parents were not relatives.

On examination, the infant was full term, gasping on arrival to the neonatal care unit and died within a few minutes of birth. There was an approximately 3 × 4 cm abdominal wall defect involving the full thickness of the abdominal wall slightly up and to the right of the umbilical cord with herniation of the stomach, small and large intestine and the liver. There were no other associated external anomalies (Figure 2).

*Case 3*

A baby was stillborn, weighing about 2500 grams, a product of spontaneous vaginal delivery, January 7, 2010, to a 24 year-old G2 P1 patient from Fallujah. She was not hypertensive or diabetic. She was a non-smoker and had no history of exposure to X-ray irradiation. She had no history of using NSAIDs, aspirin, acetaminophen, pseudoephedrine or other known teratogenic drugs. She gave no history of abortion, and there was no family history of fetal anomalies. No fetal

**Figure 2. Herniation of liver, stomach, small and large intestine through a full thickness abdominal wall defect (case 2).**

anomalies were detected on routine prenatal ultrasound scan done during the 3rd trimester. She is consanguineously married.

On examination, the stillborn showed ambiguous genitalia and a large abdominal wall defect involving the right periumbilical region with herniation of small and large bowel, liver and spleen. There was also associated scoliosis, pelvic abnormality and equinovarus deformity of both feet (Figure 3).

I examined the three cases and photographed them with my cell phone camera. The normally inserted umbilical cords were clamped and cut under my supervision.

## Discussion

Abdominal wall defects can be identified prenatally via ultrasound or measurement of alpha-fetoprotein levels (AFP) within the amniotic fluid.[3] Alpha-fetoprotein is the fetal analog of albumin, and maternal serum AFP (MSAFP) reflects the level of AFP in the amniotic fluid. In gastroschisis, MSAFP is markedly abnormal; up to 9 times the median value whereas on omphalocele MSAFP is elevated by only 4 times the median. A prenatal diagnosis of gastroschisis or omphalocele allows the family time to make preparations for the infant's care after birth.

Gastroschisis is a right paraumbilical defect involving all layers of the abdominal wall. Synonyms used to describe gastroschisis include paraompholocoele, laparoschisis and abdominoschisis.[5] The small bowel always eviscerates through the defect and is, by definition, non-rotated and lacking secondary fixation to the posterior abdominal wall. Skin is

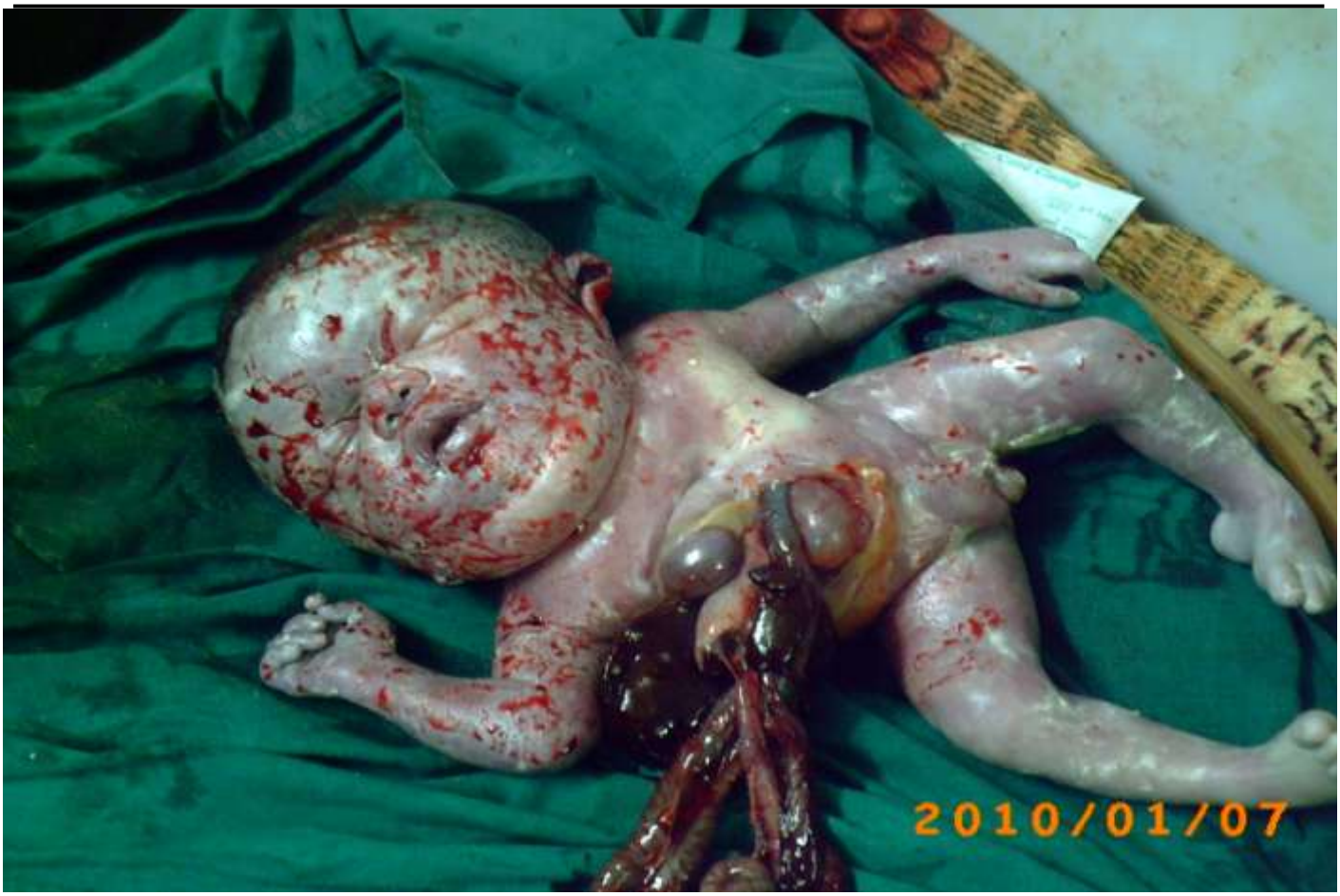


**Figure 3. Stillborn with ambiguous genitalia, scoliosis, pelvic bone abnormalities and equinovarus deformity of both feet associated with herniation of liver, spleen, small and large bowel through an abdominal wall defect (case 3).**

rarely interposed between the defect and the umbilical cord.[6] The loops of bowel are never covered by a membrane; hence, they are directly exposed to the amniotic fluid. Alpha-fetoprotein levels are usually markedly elevated. The loops usually develop a fibrinous coating and are matted together. Other organs that may eviscerate are the large bowel (often), the stomach and portions of the genitourinary system (occasionally). The location on the left side has very rarely been reported. Congenital abnormalities of the other systems are seldom noted.

Gastroschisis is found either incidentally during second trimester ultrasound scan (or during targeted ultrasound scan) or because of elevated MSAFP.[7] The diagnosis can be made with endovaginal sonography as early as 12 weeks, however caution should be taken not to diagnose gastroschisis prior to 11 weeks' gestation due to the normal evisceration of the fetal bowel as part of normal embyogenesis at that time).[8,9] The striking feature in the fetus presenting with gastroschisis is the multiple loops of bowel floating freely in the amniotic fluid. Since gastroschisis is usually not associated with other congenital or chromosomal anomalies, it carries a much better prognosis. Possible causes of death in this group would include complications of the surgical procedure for its repair, associated intestinal atresia, and low birth weight. These fetuses seem to get the most benefit from early prenatal diagnosis since they can be prospectively followed for intrauterine growth restriction and obstruction of gastrointestinal tract, as well delivery in a tertiary care center with an available pediatric surgeon. The size of the defect or the length between the diagnosis and the delivery does not influence the prognosis; thus early delivery does not appear indicated unless there is severe fetal growth restriction or other

complications.[10]

In Fallujah, prenatal ultrasonography is done routinely but not adequately. There are many cases of fetal anomalies that are missed because the equipment quality is poor and the sonographers' training is relatively inadequate. In addition, MSFAP is not done routinely. Karyotyping is not available. Autopsy is generally only performed in medicolegal or police cases, and no autopsy was requested or performed for these three cases. These factors made the diagnosis of these defects difficult.

During the year 2010, 3 cases of unusual abdominal wall defects were reported in Fallujah General Hospital out of 7,314 total births. The incidence was about 4/10,000 births compared to the overall incidence of 0.4-3/10,000 births for gastroschisis and 1.5-3/10.000 live births for omphalocele.

The first two babies died shortly after birth. The third was stillborn. Detailed history from the 3 cases revealed that there were no risk factors and no exposure to known teratogens. The first case was associated with rib cage abnormality in addition to herniation of the small and large bowel. The second case was associated with herniation of liver in addition to the intestine. The third case was associated with ambiguous genitalia, spine, pelvic & lower limb abnormalities in addition to the abnormally large abdominal wall defect. All three case were very unusual, a combination that has not been previously reported in the literature.

Only the mother of the first case had her hair tested for various minerals. That was because at the time of its birth, a research study was conducted in collaboration with the Cancer and Birth Defect Foundation in London in the period between early January and late May 2010 to test for the presence of environmental pollution in Fallujah patients with birth defect. Only the first case was included in that study. The presence of high concentrations of mercury and bismuth[11] may suggest that environmental pollution from mercury, bismuth and perhaps other trace minerals is the cause of the high incidence and the strange presentations of anterior abdominal wall thickness defects in Fallujah patients.

The three cases are unusual. The exact diagnosis of the anomalies is unclear. The herniation of viscera other than bowel and the large size of the defects strongly suggest the diagnosis of omphalocele. On the other hand, the presence of normal umbilical cord insertion suggests gastroschisis. Unfortunately, karyotyping and autopsy were not performed. Karyotypic abnormalities or the presence of other internal organ abnormalities especially of the heart will favor the diagnosis of omphalocele. This diagnosis is especially possible in case 1 where there was a membrane covering the herniated viscera which probably ruptured during delivery.

The fact that these cases are unusual and do not fit the classical criteria of either gastroschisis or omphalocele suggest a different type of abdominal wall defect that may be related to hitherto unknown teratogen that these women were exposed to in this locality in such a short period of time. Mercury and bismuth which were found at unusually high levels in the hair of the only patient out of these three women who was tested could be that teratogen.

## Conclusion

The occurrence of 3 cases of anterior abdominal wall defects within a period of one year (2010) in Fallujah does not appear to be a random event. Since high level of mercury and bismuth were found in live samples taken from parents of our 1st patient, serious consideration should be given to investigate the environment in Fallujah in order to offer logical and urgent solutions for the problem of possible environ-mental teratogen. Since early prenatal diagnosis can lead to better

prognosis, it is important to improve the antenatal care for all pregnant women, paying special attention to high risk pregnancies. Facilities for early and precise diagnosis and treatment should be provided through establishing advanced centers and providing good chances for training the medical staff to diagnose and manage fetal abnormalities cases.

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