

## Duodenal atresia and sudden fetal death.

### Running head: Prenatal diagnosis of fetal duodenal atresia and risk of fetal adverse outcome

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#### ABSTRACT

Duodenal atresia occurs in approximately 1:10.000 live births. The outcome of congenital duodenal obstructions, in terms of mortality rate, has improved over the last decades, mainly attributable to the improvement in the quality of prenatal diagnosis and neonatal intensive care. Nevertheless, several cases of sudden intrauterine fetal death (IUFD) in fetuses with duodenal obstruction have been described in the Literature. We report a case of a fetus with duodenal atresia and sudden IUFD at 32 weeks' gestation and the review of literature. We speculate that the fetal demise was due to a vaso-vagal reaction initiated by esophageal dysmotility. This report is intended to describe the features of duodenal obstruction on prenatal ultrasonography and alert the obstetrician to the possible association with sudden IUFD.

Key Words: Duodenal atresia

#### SOMMARIO

L'atresia duodenale ha un'incidenza di circa 1:10000 nati vivi. L'outcome dei pazienti con ostruzione duodenale, in termini di tasso di mortalità, risulta migliorato nelle ultime decadi, soprattutto grazie al miglioramento della qualità della diagnosi prenatale e dell'assistenza neonatale. Diversi casi di morte intrauterina fetale improvvisa (IUFD) in feti con ostruzione duodenale sono stati descritti in letteratura. Riportiamo il caso di un feto con atresia duodenale e improvvisa IUFD a 32 settimane e la revisione della letteratura. Supponiamo che la morte fetale sia da attribuire ad una reazione vaso-vagale iniziata con alterazione della peristalsi esofagea. Questo report si propone di descrivere i segni ecografici di ostruzione duodenale al fine di allertare l'ostetrico della possibile associazione con la morte improvvisa fetale.

Parole chiave: Atresia duodenale

#### CASE REPORT

A 32-year-old Caucasian woman with a spontaneous physiologic pregnancy and unremarkable obstetric history was referred to our Unit for a third trimester routine scan. A fetal duodenal obstruction was suspected by sonographic examination at 32 weeks' gestation. Detailed fetal survey showed a gastric "double bubble" sign, suggestive for fetal duodenal stenosis or atresia (Figure 1-up), associated with polyhydramnios (deepest vertical pocket of 10 cm) and fetal vomiting-like movements. No other fetal anomalies were detected. The fetal growth was regular and the umbilical artery Doppler-velocimetry was normal (PI 0.90).

After the family had been counseled, a further sonographic examination was performed two days later, showing the fetal demise. A stillborn 1850 g baby-girl was vaginally delivered 4 days later, following pharmacological induction of labor with vaginal prostaglandins. Fetal karyotype on a sample from the neonate skin was found to be normal. Fetal autopsy and histological examination

of the placenta were performed. Atresia of the distal portion of the duodenum, due to failed recanalization of the lumen, was detected 3 cm from the pylorus (Figure 1-down). No pathological



**Figure 1.** Ultrasound: Double Bubble sign (up); Pathology specimen: Atresia of the distal portion of the duodenum (down).

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features of other examined organs were found. Histological examination of the placenta showed normal amniotic membranes and umbilical cord and hypomature villi with hyalinosi.

## DISCUSSION

Duodenal obstruction is recognized as the most common intestinal obstruction occurring in newborns. The overall incidence is 1 in 10,000 live births. It includes duodenal stenosis and atresia. Small intestinal atresia (SIA) is defined as the congenital absence or abnormal narrowing of an intestinal tract (duodenum, jejunum, or ileum) leading to complete obstruction of the lumen. A higher prevalence of SIA in women aged between 20 and 24 years and over 30 years, in twin pregnancies and in male newborns is reported<sup>(1)</sup>. SIA can be associated with chromosomal abnormalities, especially trisomy 21, in about 30% of cases<sup>(2)</sup>, and with anatomic malformations in about 50% of cases, including cardiac defects, cleft lip and palate, others digestive system anomalies, defects of the urinary and musculoskeletal systems<sup>(3)</sup>. The major causes of mortality are believed to be cardiac anomalies and the association of duodenal obstruction with esophageal atresia<sup>(4)</sup>.

Many hypotheses about SIA pathogenesis have been done. Some authors address the defect to the embryologic failure of vacuolization and recanalization of the bowel that is thought to be caused by a defective vascularization occurring before 13 weeks' gestation<sup>(5-7)</sup>. Given the high association with other anomalies, some other authors believe that duodenal obstructions are not caused by vascular or embryogenetic failures but that the etiology might be more complex and as yet unclear.

The sonographic 'double-bubble' sign is

important for the prenatal diagnosis of duodenal obstruction. It is due to the dilatation of the stomach and distension of the first tract of the duodenum and it underlies a connection between stomach and duodenum. The diagnosis of duodenal obstruction usually occurs during the third trimester routine US examination<sup>(8,9)</sup>. When precociously diagnosed, it might represent a fetal condition at high risk for prenatal bradycardia, asphyxia, and fetal death, even without other associated anomalies and with a normal karyotype. Several pathogenetic hypotheses have been postulated to explain this dramatic outcome that, actually, is not well understood, yet. Williamson et al. demonstrated in the rats a correlation between level of fetal serum bile acids and cardiac failure<sup>(10)</sup>. In particular the primary bile acid (taurocholate) can modify Ca<sup>2+</sup> channels and impair the cardiomyocyte contraction. Another hypothesis explaining the correlation between duodenal obstruction and sudden fetal death is that bradycardia or asystole could be caused by a fetal vagal overactivity (VO), due to the distension of the upper gastrointestinal tract (esophagus, stomach, and the first part of the duodenum). According to Brantberg et al., it is also possible that the vasovagal reaction could depend on fetal esophageal dysmotility with vomiting-like movements and a very distended stomach and esophagus<sup>(3)</sup>.

In conclusion, according to the Literature data, the present case shows that when prenatal diagnosis of fetal duodenal obstruction is suspected and the association of gastric "double bubble" sign, polyhydramnios and vomiting-like movements is detected at US examination, sudden IUFD might occur. Further studies are needed for the better understanding of the pathogenetic mechanisms of IUFD and, therefore, to improve fetal monitoring, even in hospital, and outcomes.

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