

# DUODENAL OBSTRUCTION IN PRENATAL ULTRASOUND EXAMINATION - DUODENAL ATRESIA VERSUS ANNULAR PANCREAS - REPORT ON TWO CASES AND LITERATURE REVIEW.



**Authors:**

Klaudia Korecka<sup>1</sup>, Maria Respondek-Liberska<sup>2,3</sup>

1. Paediatric Surgery Clinic at the Upper Silesian Child Health Centre in Katowice, 2. Prenatal Cardiology Department of the Polish Mother's Memorial Hospital 3. Department for the Diagnosis and Prevention of Congenital Defects at the Medical University of Lodz.

PRENAT CARDIO. 2014 JUN;4(2):29-33  
DOI 10.12847/06145

**Abstract**

Obstruction of the duodenum is the most common intestinal obstruction of the fetus and newborn. A typical symptom of duodenal atresia is the double bubble sign. However, in order to diagnose annular pancreas, it is also required to locate a hyperechogenic band around the double bubble. We analysed the possibility of prenatal differential diagnosis of intestinal atresia, such as duodenal atresia and annular pancreas based on the analysis of two cases examined in the 26th week of pregnancy. This material was analysed by two ultrasonographers (one pediatric surgeon).

Conclusions: "Double bubble" symptom in prenatal sonography is typical of high level intestinal obstruction, but it can occur both in classical duodenal atresia, and in the cases of annular pancreas, and in other rare anomalies. Classic prenatal ultrasound examination using 2D option seems to be insufficient for accurate differential diagnosis.

**Key words:** annular pancreas, differential diagnosis, duodenal atresia, prenatal diagnosis

**INTRODUCTION**

Duodenal obstruction is one of the most common types of intestinal obstruction in fetuses and neonates.<sup>1,2</sup> The prenatal diagnosis is more often established. This pathology is usually found in examinations during the second and third trimester<sup>3</sup>.

Based on the analysis of two cases and overview of literature, we analysed the possibility of prenatal differential diagnosis, taking into account duodenal atresia and the occurrence of atresia caused by annular pancreas, in fetuses.

**MATERIALS AND METHODS**

At the Department for the Diagnosis and Prevention of Congenital Defects the ultrasound examination comprises of two parts - genetic ultrasound examination and fetal echocardiogram. Test reports are uniform for all patients. The tests were performed on GE Voluson Expert and then archived using the 4DView and FMaker Pro programs.

Two fetuses prenatally diagnosed at 26 weeks of gestation were selected for the analysis. The fetuses were

**How to Cite this Article:**

Korecka K, Respondek-Liberska M.  
*Duodenal obstruction in prenatal ultrasound examinations - duodenal atresia versus annular pancreas - reports on two cases and literature review.*  
*Prenat Cardio.* 2014 Jun;4(2):29-33



referred to the Department for the Diagnosis and Prevention of Congenital Defects, in order to verify the diagnosis and determine further optimum procedure. The authors of the paper attempted to retrospectively differentiate two different causes of obstruction based on the preserved photographic documentation of the examinations.

**Case 1:**

GI, aged 32, ultrasound examinations at 9, 12, 20 and 24 weeks of gestation, were described as normal. At 26 weeks an irregular image of the abdominal cavity of the fetus was detected. This was the reason for referral to our centre. At 30 weeks the stomach and extended duodenum was revealed with good communication and visible intestinal lumen continuity between them (double bubble). Duodenal obstruction (Photographs 1) was confirmed in the fetus with regular heart structure and function.

Due to polyhydramnion the pregnant woman was qualified for amnioreduction. The karyotype of the fetus was determined (46,XX). No other anomalies were found

Corresponding author: [kkorecka@poczta.onet.pl](mailto:kkorecka@poczta.onet.pl)

Submitted:2014-04-26; accepted: 2014-06-25

in the fetus. Delivery by Cesarean section took place at 38 weeks. A girl weighing 3290g was born and given 10/10 points on the Apgar score. On the second day of life the newborn was operated due to intestinal obstruction. An anastomosis was performed. Post surgery course was without complications. The child was discharged on the 32 day.

**Case 2:**

GI, aged 29, ultrasound examinations at 11, 16 and 21 weeks of gestational age, were described as regular. In the 26<sup>th</sup> week an irregular image of the four heart chambers as well as polyhydramnion were discovered. Ultrasound and echocardiographic examinations in our reference centre at 28 weeks of gestational age revealed a heart defect in the form of a large inlet ventricular septal defect, with no functional changes; cardiovascular efficiency was estimated at 10 points on the CVPS score. Image in the abdominal cavity was described as “double bubble” sign - stomach and another hypoechogenic area behind the stomach with a narrow connection in-between and hiperechogenic structure behind the stomach. Annular pancreas was suspected.

Furthermore, an image of a single umbilical artery (SUA) was obtained. In the 28th week of gestation, amnioreduction was performed and karyotype was determined (46,XX).

Delivery by Cesarean section took place at 33 weeks due to danger of placenta detachment. A girl weighing 1500g was born, receiving 9/10 points on the Apgar score. On the second day of life the neonate was operated on due to obstruction of the gastrointestinal track and prenatal diagnosis was confirmed. She was discharged home after 50 days of hospitalisation - foreseen further cardiological observation and eventual cardio-surgical correction.

The authors of the paper (20 months since the examinations) attempted to “blindly” and retrospectively assign the examination documentation to the described cases. Each made at least one mistake during two attempts.

**DISCUSSION**

The causes of duodenal obstruction may be divided into internal (duodenal blockage, stenosis) and external (pylorus atresia / stenosis, Ladd’s bands, intestinal malrotation, annular pancreas, diaphragmatic hernia)<sup>4</sup>.

Four duodenal obstruction types are distinguished:

- type 1: one or more transverse diaphragms - the most common type,
- type 2: blind ending loops connected by fibrous strings
- type 3: complete separation of the blind-ending loops
- type 4: apple-peel atresia of the small bowel <sup>5</sup>

The most common area in which duodenal obstruction



Photo 1. Typical presentation of duodenal atresia in prenatal ultrasound



Photo 2. “Typical” presentation of duodenal atresia and anular pancreas, however this is the same fetus from photo 1



Photo 3. False positive presentation of duodenal atresia in prenatal ultrasound in case of renal cyst

occurs (80-85%) is its “second area” - distally to Vater’s papilla<sup>6</sup>. Duodenal obstruction occurs at a frequency of 1:5000 - 1:10.000 life births <sup>5,7-10</sup> Annular pancreas is a rare developmental defect - it occurs at a frequency of 1:12,000-1:15,000, <sup>11</sup> which constitutes approximately 5% of duodenal obstruction cases and 1% of intestinal obstruction cases in neonates, (Figure 1.)<sup>8,12,13</sup> In 75% of cases the pancreas ring is partial and in 25% of cases it is complete and surrounds the pancreas in its descending part. Annular pancreas may lead to symptoms directly after birth and then, in 40% of cases, it accompanies

duodenal obstruction, and can manifest itself later or be completely asymptomatic. <sup>4,14</sup>

Embryological diagnosis of duodenal obstruction is possible from the 11th week of gestation, while the duodenal lumen is opened in the 10th week. <sup>15,16</sup> The first prenatal ultrasound diagnosis of DA was established by Houlton, Sutton and Aitken in 1974 - they described 11 cases of the diagnosis of duodenal obstruction <sup>17</sup>. As well as Loveday et al in 1975. <sup>18</sup> The most important ultrasound diagnoses of duodenal obstruction are attributed to Tsukerman - 12th week of gestational age (1993) and Zimmer - 15th week of gestational age (1996). <sup>19</sup> The first prenatal diagnosis of annular pancreas, using 2D technology, was reported in 1982. <sup>17</sup>

Duodenal obstruction diagnosis is established based on the “double bubble” image in transverse scans of the abdominal cavity - it is necessary to show a connection between them. <sup>10</sup> On the other hand, the criteria of the diagnosis of annular pancreas (Photographs 1, 3), apart from those mentioned above, also include the coexistence of a hyperechogenic “band” around the duodenum (its descending part, right at Vater’s papilla) (Photographs 2). Our documentation and retrospective analysis shows how easily mistakes in differential diagnosis, can be made (especially based on the “classic” descriptions in the literature). (Photographs 1,2,3).

In order to avoid false positive diagnoses, it is also important to observe, apart from typical double bubble symptom, the expansion - the duodenum peristaltic wave - for a few minutes, because the peristaltic wave may cause temporary expansion of a narrowed duodenum. <sup>20,21</sup> A connection between the stomach and the duodenal bubble should also be demonstrated in order to rule out other pathologies listed in Table no. 1. <sup>4,22,23</sup>

Diagnosing an obstructed duodenum with a “double bubble” sign as an isolated defect should not pose any difficulty. Difficult cases of accompanying defects have been frequently described. An analysis of such cases shows that duodenal obstruction should be suspected together with the “single bubble” symptom, i.e. a stomach which may be expanded or have regular dimensions but an irregular position.

“Triple bubble” images have also been described

<i>Unusual causes of D-B</i>	<i>Duodenal duplication</i> <i>Distended esophagus</i> <i>Stomach duplication</i> <i>Splenic cyst</i> <i>Renal cyst</i> <i>Hepatic cyst</i> <i>Choledochal cyst</i> <i>Omental cyst</i> <i>Ovarian cyst</i> <i>Diaphragmatic hernia</i>
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Table 1. Other pathologies occurring in the form of “double bubble” <sup>4</sup>

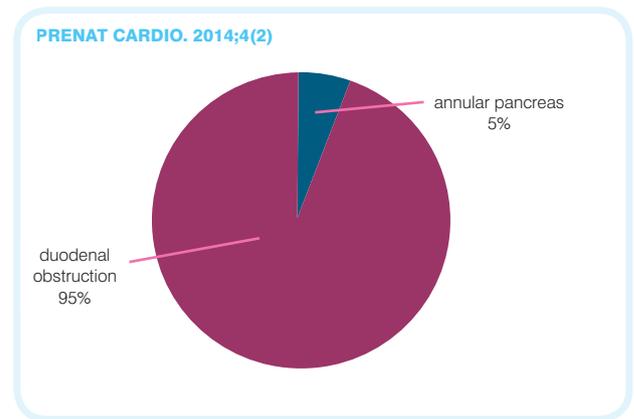


Figure 1. Percentage of annular pancreas in duodenal obstructions in prenatal ultrasound diagnostics. Such images are characteristic for high intestinal obstructions, but the “triple bubble” image has also been found in some annular pancreas cases. <sup>2</sup>

According to reports, the sensitivity and specificity of the annular pancreas diagnoses based on “double bubble” and the band of hyperechogenic ring around the descending duodenum <sup>4,5,24-26</sup> are 100% <sup>4</sup>, which is not corroborated by our observations.

Currently annular pancreas is rarely detected prenatally. Even the literature describes only single cases of this defect. <sup>4</sup> Although 3D technologies make the diagnosis significantly easier. <sup>25</sup>

Postnatal diagnosis (of isolated defects with full duodenal passage blockage) is simple - a classical X-ray photo showing a “double bubble” image is sufficient for diagnosis. A serious suspicion of a defect may be established simply by probing the stomach directly after birth. The surgical procedure consists of cutting out the blockages, anastomosis of the ends of the obstructed duodenum, anastomosis which bypasses the duodenal tissue or the release of narrowing adhesions - connective tissue bands / Ladd’s bands.

This raises the question whether a fetal ultrasonographer should focus on the duodenal pathology while taking into account annular pancreas. Even if the duodenal obstruction was suspected only once, it is a sufficient reason to confirm or rule out accompanying defects and to determine the karyotype of the fetus. The percentage of co-existing additional anomalies is estimated at 40-50% . <sup>10</sup> A separate embryogenesis of duodenal atresia and annular pancreas leads to the conclusion that the aetiology of these defects is different, which entails partial differences in the occurrence of accompanying defects.

In the presented cases, polyhydramnion occurred in both fetuses, annular pancreas was an isolated defect and duodenal obstruction was accompanied by a heart defect (VSD) and SUA as an additional marker of developmental anomalies in the fetus.

The coincidence of anomalies and accompanying defects characteristic for DA and AP is shown in Table no. 2.<sup>3,4,10</sup>

The majority of annular pancreas cases are asymptomatic and there is no systematic screening for this defect in fetal life or after birth. Hence, there is no actual frequency of the occurrence of annular pancreas and it requires further research.<sup>4</sup> The frequency of cases in which annular pancreas causes duodenal obstruction seems more important because these cases may be dangerous for neonates, and is estimated at 40%. It is necessary to conduct multi-centre research aimed at prenatal diagnosis of annular pancreas cases which would make it possible to explore the entire spectrum of this defect.

**CONCLUSIONS**

The “double bubble” symptom in prenatal sonographic examination (easy to detect) is characteristic for high duodenal obstruction, but can also be present in typical obstruction, in the course of annular pancreas and other anomalies.

Precise differential diagnosis in duodenal atresia, its level and causes of obstruction seems to be difficult.

<b>DUODENAL ATRESIA</b>	<b>ANNULAR PANCREAS</b>
Chromosomal anomalies - Down's syndrome	Chromosomal anomalies - Down's syndrome
Intestinal malrotations, common mesentery	Intestinal malrotations Situs viscerum inversus Meckel's diverticulum
Vertebral anomalies	Myelomeningocele
Esophageal atresia Imperforate anus	Esophageal atresia Imperforate anus Communicating bronchopulmonary foregut malformation Gallbladder agenesis
Congenital heart disease	Congenital heart disease
Renal anomalies	
VACTERL association	CHARGE syndrome Cornelia de Lange syndrome
	Myofibromatosis
	Polysplenia syndrome

Table 2. The co-occurrence of congenital defects in cases of duodenal atresia and annular pancreas

7. Fonkalsrud EW, de Lorimer AA, Hays DM. Congenital atresia and stenosis of the duodenum , a review compiled from the members of the surgical section. *Am Acad Pediatr* 1969; 43: 79-83.
8. Sugimoto T, Yamagiwa I, Obata K, Ouchi T, Takahashi R, Suzuki R, Shimazaki Y. Choledochal cyst and duodenal atresia: A rare combination of malformations. *Pediatr Surg Int.* 2004; 20(9): 724-726.
9. Pameijer CR, Hubbard AM, Coleman B, Flake AW. Combined pur esophageal atresia, duodenal atresia, biliary atresia, and pancreatic ductal atresia: Prenatal diagnostic Features and review of the Literature. *J Pediatr Surg* 2000; 35(5): 745-747.
10. Yadav CM, Yadav RP, Krishnanand BR. In utero diagnosis of duodenal atresia. *Journal of Nepal Medical association,* 2001; 41: 271-273.
11. Hulvat MC, Kumar RB, Newman BM, Muraskas Jk. Annular pancreas in identical twin newborns. *J Pediatr Surg* 2006; 41: 19-21.
12. Hemming V, Rankin J. Small intestinal atresia in a defined population: occurrence, prenatal diagnosis and survival. *Prenat Diang* 2007; 27: 1205-1211.
13. Whelan IJ, Hamilton GB. Annular pancreas. *Ann Surg* 1957; 146: 252-262.
14. Ravitch MM: The pancreas in infants and children. *Surg Clin North Am* 1975; 55: 377-385.
15. Sanchez MA. Duodenal atresia. Sanchez [www.thefetus.net/](http://www.thefetus.net/) 2006-05-22-10
16. Ruth BD. Ultrasound evaluation of fetal abdomen. In: Callen's *Ultrasonography in Obstetrics and Gynaecology*, 3rd edn: W. B. Saunders Company 1994; 16: 347-369.
17. Boomsma JH, Weemhoff RA, Polman Ha. Sonographic appearance of annular pancreas in utero: a case report. *Diagn Imaging* 1982; 51: 288-290.
18. Loveday BJ, Barr JA, Aitken J. The intrauterine demonstration of duodenal atresia by ultrasound. *Br J Radiol.* 1975; 48: 1031.
19. Zimmer EZ, Bronshtein M. Early diagnosis of duodenal atresia and possile sonographic pitfalls. *Prenatal diang* 1996;16: 564-566.

**References:**

1. Pariente G, Landau D, Aviram M, Hershkovitz R. PrenPariente G, Landau D, Aviram M, Hershkovitz R. *Prenatal Diagnosis of a Rare Sonographic Appearance of Duodenal Atresia. Report of 2 cases and literature review. J Ultrasound Med* 2012;31:1829-1833
2. Jeng Hsiu Hung, Shu Huei Shen, Tai Wai Chin, Chi Yi Selena Hung. *Letters to the editor . Prenatal diagnosis of double duodenal atresia by ultrasound and magnetic resonance image. Prenat Diagn* 2000; 27: 380-387.
3. Choudhry MS, Rahman N, Boyd P, Lakhoo K. Duodenal atresia: associated anomalies, prenatal diagnosis and outcome. *Pediatr Surg Int* 2009; 25: 727-730.
4. Danovicik R, Jirasek JE, Kucera E, Feyereisl J, Radonak J, Dudas M. *Prenatal diagnosis of annular pancreas: reliability of the double bubble sign with periduodenal hyperechogenic band. Fetal Diagn Ther* 2008; 24:483-490.
5. Cuillier F, Maurel A, Michel JL, Alessandri JL. Duodenal atresia. Cuillier [www.thefetus.net](http://www.thefetus.net/) 2006-06-12-11
6. Ajit SA, Tushar W, Pradip JJ, Prakash V. *Prenatal diagnosis of duodenal atresia by Ultrasound. Indian J Pediatr.* 1991; 58: 145-147.

20. Ritchie CJ, Edward WS, Mack LA, et al. Three dimensional ultrasonic angiography using power-mode Doppler. *Ultrasound Med Biol* 1996; 22: 277.
21. Obata-Yasuoka M, Hamada H, Ohara R, Nakao A, Miyazono Y, Yoshikawa H. Alveolar capillary dysplasia associated with duodenal atresia: ultrasonographic findings of enlarged, highly echogenic lungs and gastric dilatation in a third-trimester fetus. *J Obstet Gynaecol Res* 2011; 37: 937-939.
22. Dewbury KC, Alawihare APR, Birch SJ. Prenatal ultrasound demonstration of a choledochal cyst. *Br J Radiol* 1980; 53: 906.
23. Malone FD, Croinleholme TM, Nores JA, D'Alton ME. Pitfalls of the 'double bubble' sign: a case of congenital duodenal duplication. *Fetal Diagn Ther* 1997; 12: 298-300.
24. Garne E, Loane M, Dolk H, and a Eurocat working group. Gastrointestinal malformation: impact of prenatal diagnosis on gestational age at birth. *Paediatr Perinat Epidemiol* 2007; 21(4): 370-375.
25. Dankovcik R, Muranska S, Kucera E, Jirasek JE, Feyereisl J, Sudak M, Dudas M. Prenatal three-dimensional sonographic findings associated with annular pancreas. *Fetal Diagn Ther* 2010; 27: 57-60.
26. Tsukerman GL, Krapiva GA, Kirillowa IA: First-trimester diagnosis of duodenal stenosis associated with oesophageal atresia. *Prenat Diagn* 1993; 13: 371-376.
27. Lees RF, Alford BA, Brenbridge N, Buschi AJ, Williamson BR. Sonographic appearance of duodenal atresia in utero - case reports. *Am J Roentgenol* 1978; 131: 701-702.

**Conflict of interest:** The authors declare no conflict of interest

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