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Congenital Duodenal Atresia Diagnosis, Treatment, and Influence on Further Development of Patients

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ABSTRACT

Background: Congenital duodenal atresia (CDA) is a common and surgically treated digestive tract anomaly that develops in the early stage of pregnancy. It often coexists with trisomy 21 and other inborn defects. Surgery is the only way of treatment. This study aimed to investigate the relationship of CDA with early diagnosis, course of pregnancy, coexisting congenital defects, and further development of children.

Methods: The data were collected using the medical history and a self-designed survey which consisted of 22 questions about the perinatal interview, coexisting inborn defects, after birth symptoms, time and methods of diagnosis, as well as the treatment outcome.

Results: The surveys were sent to 31 patients who were diagnosed and treated because of CDA in our clinic between 2004 and 2019. According to the collected data, 73.7% of the patients were diagnosed prenatally with the mean time of diagnosis at 28th weeks of gestation. Moreover, 37% of the patients were preborn, and almost half of the patients had low and very low body weight. More than 2/3 of children presented coexisting inborn abnormalities. Among after birth symptoms, the most common ones were abdominal distention and lack of meconium. The majority of children were operated within 24 hours after birth.

Conclusions: Prenatal diagnosis of CDA results in an earlier time of primary operation. Newborns diagnosed antenatally present fewer symptoms after the operation, fewer reoperations, and a shorter time of oral feeding toleration. The coexistence of other congenital defects increases patients' mortality. The important factors that deteriorate the postoperative course are prematurity and low body weight. In order to avoid long-term complications, it is of utmost importance to provide follow-up.

Keywords: Congenital, Duodenal atresia, Double bubble, Neonate, Surgery

Introduction

Congenital malformations of the gastrointestinal tract represent 5%-6% of all inborn defects. One of the most common ones among them is congenital duodenal atresia (CDA) which occurs in 1:2500-10000 of live births (1). Although the underlying cause of duodenal atresia remains unknown, its pathophysiology has been well described. This malformation develops early during gestation due to an abnormal increase in epithelial proliferation or the lack of intestine recanalization. Extremely rare causes of CDA are bands of connective tissue that press on the duodenum and are called Ladd's bands (2). A differential diagnosis of intrinsic and extrinsic reasons for CDA is difficult before and during the operation (3). Typical symptoms of the gastrointestinal tract occlusion appear a couple of hours after the birth. The most common ones include bile vomiting and a scaphoid abdomen (2, 4). There are a few different diagnostic methods of CDA. The most common ones are ultrasonography during prenatal diagnosis and abdomen X-ray when the defect is recognized after the birth. These methods may reveal the characteristics of the duodenal atresia double bubble sign (1, 4, 5). Surgery is considered a golden standard of congenital duodenal atresia treatment. It should be performed as soon as the general state of the patient becomes stable (1, 2). This study aimed

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to investigate the relationship of CDA with early diagnosis, course of pregnancy, coexisting congenital defects, and further development of children.

Methods

This study investigated 31 patients (14 female and 17 male) who were treated surgically in the Department of Pediatric Surgery, Urology, and Traumatology, Poznan University of Medical Sciences, Poznan, western Poland, over 16 years, from 2004 to 2019 on the account of CDA. The basic method of diagnosis was ultrasonography performed by a gynecologist during the second trimester of pregnancy. A plain X-ray of abdomen was also performed to confirm the diagnosis. All patients were treated surgically by the use of "double diamond" duodenoduodenostomy. No patients were excluded from the study, and the study described in the publication consisted of two parts. The first part was a retrospective analysis of medical history, and the second part was a survey. Surveys and questions were prepared along with an appropriate explanation of the study and sent to the patients' parents. In case of doubt, the survey was completed by telephone after contact with the patients' parents. The data regarding age, gender, pregnancy course, perinatal interview, methods of diagnosis, symptoms, and clinical presentation were collected using the patients' medical history. The surveys filled out by the patients' parents were gathered in order to analyze the time of diagnosis, coexistence of inborn defects, surgical process, time of hospitalization, perioperative complications, and long-term outcome of the operation. The analysis results for weight and height in patients treated surgically in the past for CDA were referred to the healthy population based on the percentile grids for the appropriate gender and age. All procedures performed in the studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and the 1964 Helsinki declaration and its later amendments or comparable ethical standards. The study protocol was approved by the Bioethical Commission of Poznan University of Medical Sciences, Poznan, Poland. The obtained data were summarized using Microsoft Excel (Microsoft Inc., Seattle, WA, USA), and they were elaborated using Statistica 10. Descriptive statistics and correlation between variables were performed using the aforementioned program. To examine the relationship between the described variables, the Mann-Whitney's test and the Spearman's rank correlation coefficient were also used in this study. A p-value less than 0.05 was considered statistically significant.

Results

According to the results, pregnancies proceeded without complications in 60% of the mothers. Moreover, 25% of the gravid delivered baby by cesarean section. In the study group, 37% of the patients were born prematurely. The mean time of birth was 36 weeks of gestation and 3 days. In addition, almost half of patients were born with low body weight under 2,500g (mean weight: 2,371.05g). The assessment indicated a higher level of premature and low body weight births in patients with duodenal atresia, compared to the healthy population. The mean time of diagnosis was 28 weeks of gestation. The majority of patients (73.7%) were diagnosed prenatally by the visualization of double bubble sign in ultrasonography. It should be mentioned that 26.3% of the newborns were diagnosed postnatally. In these cases, the CDA was diagnosed using ultrasonography, plain x-rays of the abdomen, and upper gastrointestinal contrast evaluation. The anomalies associated with duodenal obstruction occurred in 68% of the patients, which were Down's syndrome and congenital heart diseases (50%), annular pancreas, neurological and genitourinary defects (10%), anal or esophageal atresia, and intestinal malrotation (5%) in descending order. The afterbirth symptoms in our patients were summarized in Table 1.

Most patients underwent surgery within 24 hours after birth. Among those who were diagnosed prenatally, early surgery was performed in 86% of the patients. The mean time of the operation in a group of patients diagnosed postnatally was the 11th day after birth. Children with a delayed operation were nourished parenterally for 17 days. The mean time of enteral initiation feeding in patients with the early surgical intervention was determined at 14.9 days. Symptoms that were maintained after the surgical treatment were presented in Table 2.

Due to no improvement after the surgery, it was necessary to reoperate 40% of the cases.

Table 1. Afterbirth symptoms in children with congenital duodenal atresia

Afterbirth symptoms	No. of patients; [%]
Lack of meconium	8; [25,8]
Abdominal distention	8; [25,8]
Vomiting	7; [22,5]

Table 2. Symptoms presented after surgery

Symptoms	No. of patients; [%]
Vomiting	3; [9,7]
Abdominal distention	3; [9,7]
Gastroesophageal reflux	2; [6,5]
Belching	2; [6,5]

Table 3. Patients' current general condition

General condition of the patients	No. of patients; [%]
Very good	10; [32,2]
Good	15; [48,4]
Medium	5; [16]
Bad	0; [0]

What was more, the frequency of reoperations in children with other congenital defects of the digestive tract was higher (68%). A small group of children presented gastrointestinal symptoms at the time of the follow-up examination. These symptoms were occasionally mild and did not affect the patients' life and their development. The most common manifestations of gastrointestinal symptoms were occasional constipation (23%), abdominal distention (23%), and vomiting (19%).

According to the questionnaire, most of the patients develop properly both mentally and physically, obviously taking into account the coexisting genetic syndromes. About 83% of the parents evaluated the general condition of their child as good or very good. Precise results are submitted in Table 3.

The height and bodyweight of the vast majority of the patients during control were within a normal range (Figure 1).

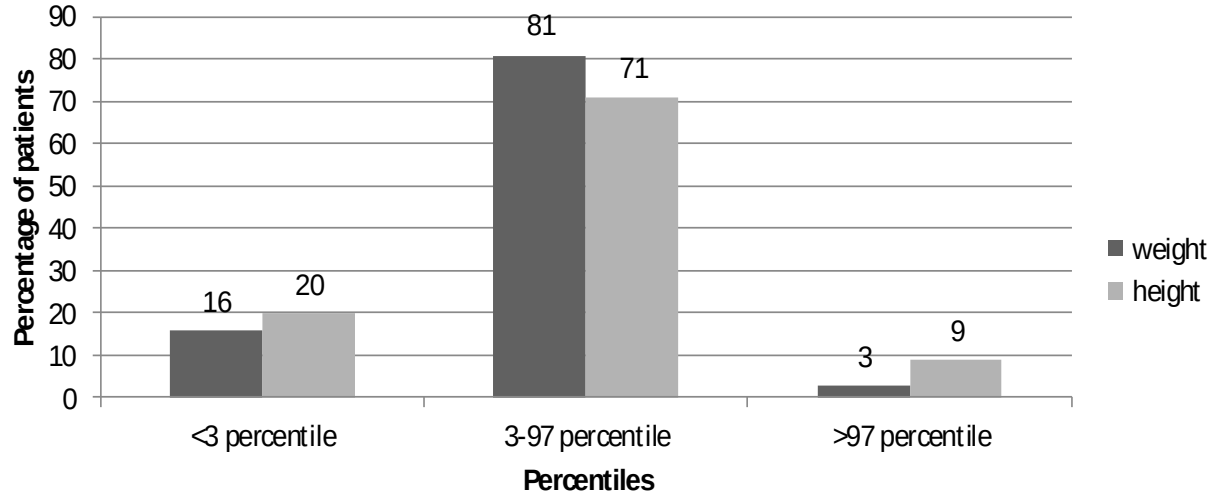

Figure 1. Height and body weight of patients during control

Discussion

The CDA is one of the most common causes of alimentary tract occlusion (6). There are few diagnostic methods for this complication. It is possible to identify duodenal atresia during pregnancy by revealing a double bubble sign in ultrasonography. Obstetricians may take advantage of prenatal ultrasound near 20 weeks of gestational age (4). Examination which confirms the diagnosis in infants is abdominal radiograph, where the dilatation of the proximal duodenum and stomach is present (1). In our study, 73.7% of the patients were diagnosed prenatally by the visualization of double bubble sign in ultrasonography. Moreover, 26.3% of the newborns were diagnosed postnatally using ultrasonography, plain x-rays of the abdomen, and upper gastrointestinal contrast evaluation. There is a correlation between antenatal diagnosis and the earlier time of primary operation. Other authors also indicated that prenatal diagnosis led to sooner operation and reduction in the time of hospitalization; however, it did not change these children's outcome (7-10). A recent publication has indicated that early diagnosis may improve survival rate in the view of metabolic disturbances or dehydration reduction and sooner surgery (11). Our study revealed that the majority of patients underwent the surgery within 24 hours after birth. Among patients who were diagnosed prenatally, early surgery was performed in 86%. More than half of newborns with duodenal atresia present other congenital abnormalities, the most common of which include Down's syndrome, congenital heart diseases, and other gastrointestinal defects. Few retrospective analyses have also shown high levels of concomitant anomalies, especially Down's syndrome (4, 12). It is more probable to overlook

the small intestine obstruction due to the absence of associated abnormalities. Obstetricians are obligated to search for sonographic imaging of duodenal atresia after prenatal Down's syndrome or heart defects revelation (4).

In our study, the anomalies associated with duodenal obstruction occurred in 68% of patients. In 50% of them, they were Down's syndrome and congenital heart diseases. According to the collected data, other congenital defect coexistence correlates with the need for a second surgery. These defects may also deteriorate patients' development and increase the mortality rates (1, 12). The majority of postoperative deaths (80%) are correlated with various congenital disorders (3). In our study, the frequency of reoperations was higher in children with other congenital defects of the digestive tract (68%).

The results of the present study revealed that a significant percentage of newborns with duodenal obstruction were born prematurely (37%) and with low birth weight (47%). The assessment indicates a higher level of premature and of low body weight births in patients with duodenal atresia, compared to a healthy population. Rattan et al. in their study observed 26.3% of preterm births and 2100g of mean birth weight. They also indicated prematurity as a risk factor of duodenal atresia appearance (1). Newborn immaturity is correlated with a longer time of enteral feeding establishment in the postoperative course (13). Long term outcome of the treatment was satisfactory, and the majority of patients in the analyzed group had developed properly and met the standards (their height and body weight lies between 3 and 97 percentile). Only, a minority of the children presented mild gastrointestinal symptoms. However, the prognosis is good, and it is important to follow up check-ups during patients' development due to the risk of late complications occurrence, such as gastroesophageal reflux disease or digestive disorders (10).

Conclusion

According to the results of the present study, prenatal diagnosis of CDA resulted in an earlier time of primary operation. Newborns diagnosed antenatally presented fewer symptoms after the operation, fewer reoperations, and a shorter time of oral feeding toleration. Furthermore, the coexistence of other congenital defects increases patients' mortality. Other important factors that deteriorate postoperative course are prematurity and low body weight. Long-term complications do not appear frequently, and the provision of long-term check-ups is indispensable in order to avoid them.

Ethical issues (plagiarism, informed consent, misconduct, data fabrication and/or falsification, double publication and/or submission, and redundancy) have been completely observed by the authors.

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Conflicts of interest

The authors declare that there is no conflict of interest.

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