Congenital Anomalies of the Duodenum

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Abstract

This is a prospective study that was carried out on 50 patients at the central teaching hospital for children in Baghdad, 20 males and 30 females. Their age ranged from 1-60 days with body weight ranged from 1.8-3.5 Kg., eight patients were premature.

The study revealed atresia in 15 (30%) cases, stenosis in 12 (24%) cases, malrotation in 10 (20%) cases, and annular pancreas in 7 (14%) cases. prenatal ultrasound diagnosed 3 patients. Bilious vomiting was the commonest presenting symptom (42 patients (84%)), associated congenital anomalies were found in 18 (36%) patients. Down syndrome was found in 10 (20%) patients, maternal polyhydramnios was observed in 20 (40%) patients and in one patient there was a family history of previous anomalies. Low birth weight and prematurity significantly affect the outcome, associated anomalies account for most of the morbidity and mortality and early diagnosis result in better outcome.

Objectives (aim of study)

To study cases with congenital anomalies of duodenum and verify the causes, clinical presentation, diagnosis and outcome in infants admitted to the central teaching hospital for children in Baghdad.

التشوهات الخلقية للأثني عشري (العفج)

الخلاصة

هذه دراسة مستقبلية أجريت في مستشفى الطفل المركزي التعليمي في بغداد على خمسين مريضا، 20 ذكر و 30 أنثى، تراوحت أعمارهم بين واحد إلى 60 يوم وتراوحت أوزانهم بين 1.8 - 3.5 كغم ، وكان منهم 8 مرضى خدج.

كشفت الدراسة عن وجود رتق ألاثني عشري في 15 مريض وضيق ألاثني عشري في 12 مريض و تشوه البنكرياس الخلقي في 7 مرضى و إخفاق الاستدارة الكاملة للأمعاء في 15 مريضا كما ظهرت الحزم والأربطة الخلقية في أربع حالات مرضية.

تم تشخيص 3 حالات من انسداد ألاثني عشري الخلقي بواسطة فحص الأمواج الصوتية قبل الولادة في حين كان التقيؤ الصفراوي هو العارض الشائع بين المرضى إذ شكل 45 حالة كما وجدت تشوهات خلقية مصاحبة لدى 15 مريضا وكانت متلازمـــة داون موجودة لدى 15 منهم.

وجدت حالات زيادة الخمط الأمومي لدى 20 مريضا كما ظهرت عيوب خلقية عائلية لدى مريضين آخرين.

إن التشوهات الخلقية المصاحبة والولادة المبكرة تؤثر سلبيا على النتائج وان التشخيص المبكر للحالة يعطي نتائج جيدة

Introduction

Duodenal anomalies can be intrinsic (atresia, stenosis, web) or extrinsic (annular pancreas, malrotation and Ladd's band). It can occur distal or proximal to the ampulla of Vatter. Most commonly distal to the ampulla and there for bilious vomiting is present [1].

Associated polyhydramnios is recorded in up to one half of the cases, with premature delivery in one third [2, 3]. Growth retardation is also common, which may imply that the fetus have been deprived of nutritional

contribution of the swallowed amniotic fluid. Almost 50% of duodenal atresia are associated with some other anomaly (e.g.: cardiac, genitourinary, anorectal, or occasionally, esophageal atresia) [3].

Calder published the first report of duodenal obstruction in 1733 when he described two children with "preternatural confirmation of gut". Both infants died, as did subsequently reported infants with this defect. Scattered report of duodenal obstruction appeared in the European literature over ensuing years. In 1916 the first survival was reported, yet survival in the early 20th century remained rare. Morbidity and mortality significantly improved only over the last 50 years. Progress in pediatric anesthesia and neonatology; combined with improved intra-operative methods and surgical materials; accounts for today's more than 90% survival rate of infants who present with this anomaly [4]. Approximately 24 to 28% of newborn with duodenal atresia or stenosis have Down syndrome. Conversely, approximately 2.5% of patients with Down syndrome have duodenal atresia or stenosis (5). Rotational anomalies occur as a result of an arrest of a normal rotation of the embryonic gut. They are often associated with other gastrointestinal abnormalities, particularly those in which the intestine are located outside the coelomic cavity (e.g.: congenital diaphragmatic hernia or abdominal wall defect. As many as 17% of children with duodenal atresia and 33% of children with jejunoileal atresia may have an associated malrotation *[6,7]*. Three major theories have been proposed to explain the annular pancreas. adherence of the ventral bud to the duodenal wall prior to rotation resulting in its persistence and encirclement of the duodenum (Lecco`s theory) [8]. Persistence and enlargement of the ventral bud (Baldwin`s theory) [9]. Hypertrophy and fusion of the ventral and dorsal bud before rotation of the gut resulting in complete encirclement of duodenum [10].

By prenatal ultrasound most cases of duodenal atresia are detected between 7th and 8th month of intrauterine life, but a normal ultrasound of the fetus with polyhydramnios at that time does not absolutely exclude duodenal obstruction [11].

On a plain abdominal radiograph, duodenal atresia causes a classic double bubble sign [12] (Fig.:1).

Contrast meal is required when there is incomplete obstruction to exclude malrotation and volvulus (Fig.:2, 3, and 4).

Specific studies may be required to evaluate the infant for associated congenital anomalies. in some cases, testing should occur prior to surgery. An Echocardiogram should be performed in infant with duodenal atresia. A preoperative cardiac assessment is most important in patient with Down syndrome. Infant with duodenal and jejunoileal atresia should have an antero-posterior and lateral chest radiograph to detect vertebral anomalies. Renal ultrasonography should be performed in infant with duodenal atresia. infant with duodenal atresia and down syndrome may need a rectal biopsy to exclude Hirschsprung`s disease [13].

Patients and Methods

A prospective study was done on a sample of 50 cases of newborn infants with congenital anomalies of duodenum from august 2007 to December 2008 at the central teaching hospital for children.

Infant age was range from 1- 60 days, their body weight ranged between 1.8 and 3.5 Kg.

Detailed data collected include : age at presentation, gender, presenting features, associated congenital anomalies, family history, clinical examination, investigations (data form).

Results

A series of fifty patients were studied prospectively at the central teaching hospital for children in Baghdad in the period from august 2007 to December 2008.

The series include 30 females (60%) and 20 males (40%). (F:M ratio = 1.5:1).

The number of preterm babies was 8 patients (16%), while that of full term was 42 patients (84%) as shown in table (1).

Table 1 gender, maturity, and polyhydramnios

		Number	%
Gender	female	30	60
	male	20	40
Maturity	Full term	42	84
	Preterm	8	16
polyhydramnios		20	40

Maternal polyhydramnios was observed in 20 (40%) patients.

The causes of duodenal obstruction were; 15 (30%) cases were due to atresia, 12 (24%) cases due to intrinsic web, 10 (20%) cases due to malrotation, 7 (14%) cases due to annular pancreas, 4 (8%) cases due to congenital bands and 2 (4%) cases were due to other anomalies which are multiple

atresia in the small bowel with areas of mesenteric defect as shown in table 2.

Table 2 causes of congenital duodenal obstruction

cause	number	%
Atresia	15	30
Intrinsic web or diaphragm	12	24
Malrotation	10	20
Annular pancreas	7	14
Congenital band	4	8
Other anomalies	2	4

The age at presentation in different pathological types were as follows; all the 15 (100%) cases of duodenal atresia presented in the 1st week of neonatal life , nine cases (75%) of intrinsic duodenal web presented in the 1st week, two cases in the second week , and one case in the third week. For the malrotation of midgut; 4 cases presented in the 1st week, 3 cases in the 2nd week, 2 cases in the 3rd week, and one case in the 4th week. For the annular pancreas; 4 cases of annular pancreas presented in the first week , 1 case in the 2nd week , 1 case in the 3rd week and 1 case in the 4th week. Other congenital anomalies were 2 cases of multiple atresia presented in the 1st week, and congenital bands; 1 case in the 2nd week, 2 cases in the 3rd week, and 1 case in the 4th week as shown in the Table 3.

Table 3 presenting age in different pathological types.

	1st week	2nd week	3rd week	4th week	Total
Atresia	**15 (100%)**	----------	----------	----------	**15**
Intrinsic web	**9 (75%)**	**2 (16.6)%**	**1 (8.3)**	----------	**12**
Malrotation	**4 (40%)**	**3 (30%)**	**2 (20%)**	**1 (10%)**	**10**
Annular pancreas	**4 (57%)**	**1 (14%)**	**1 (14%)**	**1 (14%)**	**7**
Other anomalies	**2 (33.3%)**	**1 (16.6%)**	**2 (33.3%)**	**1 (16.6%)**	**6**
Total	**34 (68%)**	**7 (14%)**	**6 (12%)**	**3 (6%)**	**50**

Associated anomalies were presented in 18 cases (36%) of total cases, distributed as follow; isolated Down syndrome in 20%, this was diagnosed clinically and chromosomal study was not done due to the emergency situation of the cases. Other anomalies were present as follow; GIT anomalies 3 cases (6%), genitourinary 2 (4%), cardiac anomalies 2 (4%), multiple anomalies one case (2%).

Duodenal atresia have the highest percent of associated anomalies (16%), followed by malrotation (6%), annular pancreas (6%) and the least associated anomalies were in the duodenal web (2%) as shown in table 4.

Table 4 associated anomalies

	Atresia	web	malrotation	Annular pancreas	Other anomalies	Total (%)
Down syndrome	**6**	**2**	**1**	**1**	**---**	**10 (20%)**
GIT anomaly	**---**	**----**	**1**	**1**	**1**	**3 (6%)**
Genitourinary	**1**	**-----**	**1**	**---**	**---**	**2 (4%)**
Cardiac	**1**	**---**	**---**	**1**	**---**	**2 (4%)**
Multiple	**---**	**---**	**---**	**---**	**1**	**1 (2%)**
Total	**8 (16%)**	**2 (4%)**	**3 (6%)**	**3 (6%)**	**2 (4%)**	**18 (36%)**

The presenting symptoms and signs were; bile stained vomiting in the majority of cases (42) (84%) cases, dehydration 15 (30%) cases, distension 8 (16%) cases, jaundice in 7 cases (14%) and non bile stained vomiting in cases of pre-ampullary obstruction 2 (4%) cases. table 5

Table 5 presenting symptoms and signs

	Atresia	web	malrotation	Annular pancreas	Other anomalies	Total (%)
Bile stained vomiting	**14**	**11**	**9**	**7**	**1**	**42 (84%)**
Dehydration	**7**	**2**	**3**	**2**	**1**	**15 (30%)**
Abdominal distention	**1**	**---**	**4**	**1**	**2**	**8 (16%)**
Jaundice	**3**	**1**	**2**	**1**	**---**	**7 (14%)**
Non bilious vomiting	**1**	**1**	**---**	**---**	**---**	**2 (4%)**

The imaging investigation used in the study were; erect plain abdominal x-ray in all patients, upper GIT barium study in 25 (50%), prenatal ultrasound in 3 patients and postnatal ultrasound examination in 15 (30%). The double bubble sign on plain abdominal x-ray was seen in all cases , barium study showed blind ended second part of duodenum in all cases of duodenal atresia, and narrowing in the duodenal web, birds beak appearance (complete obstruction) and coiled spring appearance (partial obstruction) in 6 cases of malrotation.

Prenatal ultrasound detects distended stomach and 1st part of duodenum with polyhydramnios in 3 cases and on exploration; 2 were atresia and one case of annular pancreas. Postnatal ultrasound examination done in 8 cases of duodenal atresia, 3 cases of malrotation, 2 cases of annular pancreas, and 2 cases of duodenal web, as shown in table 6.

Table 6 imaging investigations for diagnosis

investigations	Atresia	web	malrotation	Annular pancreas	Other anomalies	Total (%)
Plain x- ray	**15**	**12**	**10**	**7**	**6**	**50**
Barium study	**10**	**4**	**6**	**3**	**2**	**25**
Prenatal US	**2**	**---**	**---**	**1**	**---**	**3**
Postnatal US	**8**	**2**	**3**	**2**	**---**	**15**

Discussion

Congenital duodenal obstruction is relatively common abnormality in the newborn period and may be complete or partial, intrinsic or extrinsic.

Intrinsic atresia or stenosis are relatively common; a population – based study documented that duodenal atresia and stenosis have an incidence of about 1 in 7000 live birth and account for 49% of all small intestinal atresia.

Extrinsic duodenal obstruction may be due to annular pancreas, malrotation and Ladd`s band or as part of multiple congenital anomalies [14].

In our collection of 50 cases of congenital duodenal obstruction, females were relatively more common than males with a female to male ratio of 1.5:1 this is comparable with that mentioned in the literature [15].

Most of the cases were full term infants (84%) and preterm infants formed 16% of the collection. The percentage of the preterm infant is usually higher, where it might reach to 40% [15], the percentage in our study might be less than that mentioned as we do not have the mortality rate of preterm babies and the percentage of referred infants to our department.

The presentation of cases of duodenal obstruction is usually in the early neonatal life.

In our study the time of presentation had relation to the type of pathology, the presentation was earlier in cases proved to be complete obstruction (duodenal atresia) and later in time in cases of incomplete obstruction (web, annular pancreas and malrotation).

All of our 15 cases of duodenal atresia were presented in the first week of life, while cases with annular pancreas and malrotation were distributed over the first four weeks. This is explained by the fact that intrinsic duodenal atresias (when the obstruction is complete) are developmental abnormalities that occur during early development of the foregut, and there is a little difficulty in clinical recognition, but when the obstruction is incomplete, like

congenital intrinsic stenosis which caused by luminal membranes with a crescentic defect or central fenestration of variable size can provide a conduit of sufficient size to postpone symptoms until later in life and the diagnosis may pose considerable difficulty [16].

The relation between the point of obstruction and ampulla of Vatter is important; the site of obstruction is usually either below, at or above the ampulla of Vatter. This mean the bilious vomiting is related to the post-ampulla obstruction, while non bilious vomiting occurs in preampullary obstruction [4]. Bilious vomiting was the main reason for presentation in our series. This formed 84% of our cases. Most series document a predominance of postampullary obstruction, approaching 80% as in study of Fonkelsrud [17].

Dehydration was the second in order regarding the presentation. In this collection, 30% of our cases presented with profound sign of dehydration. This might be a high percentage which can reflect the medical awareness of the medical personnel and parents health education. The degree of the resultant dehydration depends on the duration and severity of vomiting.

Abdominal distension is usually mild and limited to upper abdomen as duodenal obstruction is regarded as high intestinal obstruction. Delayed diagnosis may result in dehydration, hyponatremia, hypochloremia. These facts are mentioned in literatures [18, 19].

Non bilious vomiting is accounting for about 4% of our cases. A European report described preampullary predominance [20].

Jaundice at presentation was rare in our collection 14%. Jaundice if present is rarely due to obstructive causes and is more likely due to dehydration and prematurity.

Congenital duodenal obstruction is commonly associated with other serious congenital anomalies, which account for most of the morbidity and mortality in these patients.

Various reports put the incidence of the associated anomalies in our series 36%. Trisomy 21 (Down`s syndrome) is the most common associated anomaly observed and was present in 20% of our cases. This was diagnosed clinically without ordering chromosomal studies at time of presentation due to the emergency situation of the cases.

In our study, the incidence of down`s syndrome in cases of duodenal atresia was 40% which is similar to those reported in other studies by Alastair [1], Adeyemi [21], and AL-salem [22].

The diagnosis of duodenal atresia is often suggested by prenatal ultrasound. A maternal history of polyhydramnios is common in congenital duodenal obstruction, approaching 75% in one series [19]. A history of maternal polyhydramnios was reported in 20 (40%) of our cases. It is caused by a failure of absorption of amniotic fluid in the distal intestine. Therefore any case of polyhydramnios should be examined carefully to rule out the presence of fetal anomaly.

The dilated stomach and proximal duodenum seen on antenatal ultrasonography are detected between 7^{th} and 8^{th} month of the intrauterine life. These results support the role of fetal swallowing and absorption by fetal GIT in the regulation of amniotic fluid volume [4].

Fetal anomaly had a strong association with prematurity [23]. Both prematurity and maternal polyhydramnios were recorded in other studies; Dalla [2] and Grosfeld [3].

In this study, duodenal atresia and stenosis were the most common causes of duodenal obstruction, while

malrotation of midgut was the 2nd cause which is often caused by a peritoneal band that run from abnormally positioned caecum to the right side of the abdomen crossing the descending part of the duodenum.

The classic presentation of a complete postampullary obstruction includes bilious vomiting within 24 hours of birth in an otherwise stable infant with a non distended abdomen. Plain radiographs of the abdomen typically show the classic double - bubble sign (Fig. 1).

Two distinct gas collections or air fluid levels in the upper abdomen resulting from the markedly dilated stomach and proximal duodenal bulb [16]. If the infant`s stomach has been decompressed by vomiting or previous Nasogastric aspiration, 40-60 ml of air may be injected carefully through the Nasogastric tube and the double – bubble reproduced. Air makes an excellent contrast agent, obviating an upper GI tract contrast study in routine cases [16].

An upright abdominal radiograph using instilled air as a contrast is sufficient to confirm the diagnosis of duodenal obstruction.

Although plain x-ray was done to all patients, it was diagnostic in only 22 (44%) patients.

Prenatal ultrasound examination diagnosed 3 cases (6%). This can influence parent positively in coping with the anomaly and to seek surgical correction relatively earlier than others, which can improve; to some degree; the outcome [24].

Conclusion

Congenital duodenal anomalies are common conditions facing the pediatric surgeon in his life.

1- The most common cause of congenital duodenal anomaly was atresia and stenosis.

2- Prematurity and birth weight has a great importance in determining the outcome of the patient. Low birth weight and extreme prematurity need special care which should be available if good outcome is demanded.

3- Early diagnosis: this can be accomplished if the sonnarist had good clinical awareness and experience in identifying the intrauterine fetal anomalies. Also early diagnosis can permit time for screening and possible early correction of some anomalies and thus helping the patient and parent in getting early surgical advice and avoiding the development of complication.

4- Associated anomalies: it is the first factor influencing the fate of the patient. Absence of other anomalies is considered a good prognostic factor regarding outcome.

5- The most important warning sign to the physician is the bile stained vomiting and it should be considered pathological and surgical until proved otherwise.

Recommendations

1- Careful intrauterine assessment of the fetus by ultrasound examination especially if the mother had polyhydramnios or there are previous congenital anomalies.

2- Any case of bilious vomiting should be considered pathological and surgical until proved otherwise.

3- Any patient with congenital duodenal anomalies should be investigated thoroughly for any associated anomalies as some of them influence the prognosis profoundly.

4- Health education of the community as well as the medical personnel about the early symptoms and signs of neonatal intestinal obstruction.

5- Education of the community about the importance of antenatal care in detecting fetal anomalies.

References

1. Alastair J.W. Millar, Heinz Rod, Sidney Cywes. Intestinal atresia and stenosis chapter 30: pediatric surgery, 4th edition by Elsevier Inc., 2005,416-34.

2. Dalla Vecchia, LK, Grosfeld, JL West, et al. Intestinal atresia and stenosis: a 25-year experience with 277 cases. Arch surg 1998; 133:490.

3. Grosfeld, J.L. Rescorla. Duodenal atresia and stenosis: reassessment of treatment and outcome based on antenatal diagnosis, pathologic variance, and long-term follow-up. World J surg 1993, 17(3):301-9.

4. Casey M Calkins. Duodenal atresia: e medicine January 17, 2003.

5. Epstein, CJ. Down syndrome (trisomy 21). In: Metabolic and Molicular Bases of inherited disease, 8th ed, Scriver, CR, Beaudet, AL, Sly, WS, Valle, D (Eds), McGraw-Hill, New York 2001. P.1223.

6. Smith, El. Malrotation of the intestine. In: Pediatric Surgery, 4th Ed, Welch, KJ, Randolph, JG, Ravich, MR, et al (Eds), Year Book Medical publishers, Chicago 1986. P.882.

7. Glover, DM, Barry, FM. Intestinal obstruction in the newborn. Ann Surg 1949; 130:480.

8. Laughlin, EH, Keown, ME, Jackson, JE. Heterotopic pancreas obstructing the ampulla of Vatter. Arch surg 1983; 118:979.

9. Baldwin, W.A specimen of annular pancreas. Anat. Rec 1910; 4:299.

10. Hill, ID, Lebenthal, E. Congenital abnormalities of the exocrine pancreas, in: pancreas: pathology, Pathobiology and disease 2nd ed, GO, VL, Dimagno, EP, Gardner, JD (Eds), et al, Raven Press, NY 1993. P.1029.

11. Brantberg, A, Blaas, HG, Salvesen, KA, et al. Fetal duodenal obstructions: increased risk of prenatal sudden death. Ultrasound Obstet Gynecol 2002; 20:439.

12. Kimble, RM, Harding, J, Kolbe, A. Additional congenital anomalies in babies with gut atresia or stenosis: when to investigate, and which investigation. Pediatr Surg Int 1997; 12:565.

13. William Adamson. Bowel obstruction in the newborn: e medicine June 15, 2004. (Internet reference).

14. Cragan JD, Martin ML, Moore CA, et al. Descriptive epidemiology of small intestinal atresia in Atlanta, Georgia. Teratology 1993;48:441

15. Irving IM, Rickham pp: duodenal atresia and stenosis; annular pancreas. In Rickham pp, Lister J, Irvin IM(eds): neonatal surgery, 2nd ed. Boston, Butterworths, 1978, p355.

16. David K. Magnuson, Marshall Z. Schwartz; Stomach and Duodenum. In: Oldham, Keith T.;Colombani, Paul M.; Foglia, Robert P.; Skinner, Michael A., Principles and Practice of Pediatric Surgery, 4th ed., 2005, Lippincott Williams & Wilkins, Pennsylvania, Chapter 72.

17. Fonkelsrud Ew, de Lorimier AA, Hays DM. Congenital atresia and stenosis of the duodenum: a review compiled from the members of the Surgical Section of the American Academy of Pediatrics. Pediatrics 1969;43:79.

18. Bailey, P.V., T.F. Tracy, Jr., et. al congenital duodenal obstruction: a 32 years review. J pediatric surgery 1993, 28 (1): 92 - 5.

19. Spigland, N., and S. Yasbeck. Complication associated with surgical

treatment of congenital intrinsic duodenal obstruction. J pediatric surgery, 1990.25(11):1127-30.

20. Schier F, Schier C, Waldschmidt J, et al. Duodenal atresia: experiences with 145 patients. Zentralbl Chir 1990; 115:135.

21. Adeyemi, S.D. Duodenal Obstruction in Nigeria newborns and infants. Scand J Gastroenterology suppl 1986, 124:157-61.

22. Al- Salem, A.h., Khwaja, et al. congenital intrinsic duodenal obstruction: problem in the diagnosis and management. J pediatric surgery 1989, 24(12):1247-9.

23. Jordan J. Weitzman. Logical approach to neonatal intestinal obstruction: On Line pediatric handbook of surgery for medical students 2002. (Internet reference).

24. Arnbjornsson, E. M. Larsson, et al. Transanastomotic feeding tube after an operation for duodenal atresia. Eur J pediatric surgery 2002, 12 (3): 159-62.