

Congenital Duodenal Obstruction; Review of 56 Patients

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ABSTRACT:

BACKGROUND:

The incidence of neonatal duodenal obstruction has been variously estimated as 1 in 10000 to 1 in 40000 live births. Duodenum obstruction may also caused by annular pancreas and band of Ladd in association with malrotation. The detection of double bubble sign on plain radiograph of abdomen lead to suspicion of duodenal obstruction. Duodenoduodenostomy of different technique has been used to bypass duodenal obstruction.

OBJECTIVE:

To study congenital duodenal obstruction (atresia,annular pancreas and malrotation) and to evaluate and analyze the age presentation, x-ray findings and the operative findings in comparison with other studies, with emphases to the operative procedures suitable for each cause.

PATIENTS AND METHODS:

In this study patients were collected prospectively through the period between first of January 2010 to the end of January 2013 in pediatric surgical ward, at Al-Khansa teaching hospital in Mosul/Iraq. Erect abdominal x-ray was done for all patients to assess the radiological findings, while barium meal done selectively. Operative findings were evaluated in relation to the cause of obstruction, site, degree of proximal dilatation, presence of distal obstruction and other associated anomalies.

Surgical procedures were used according to the operative findings which were either diamond duodenoduodenostomy (DDD), side to side duodenoduodenostomy (SSDD), side to side duodenojejunosomy (SSDJ), Ladds procedure and Heinecke-Mikulicz (HM) or combined procedures.

RESULTS:

Fifty six patients were admitted to our center, 31male (55.5%) and 25 female (44.5%). Forty five patients presented in the first 4 days of life (80%), 35 of them (60%) in the first 2 days, while 11 patients (20%) were delayed after the seven day of life, one patient aged one month proved to have fenestrated web. Vomiting was the main presenting symptoms in 50 patients (90%), and was bile stained in 42 patients (85%) and nonbile stained in 8 patients (15%). Erect abdominal x-ray show typical double bubble gases shadow sign in 39 patients (70%), Operative findings at time of exploration shows 36 atresia(67%) including 25 web(type one atresia), type 2 atresia in 8 patients and 3 patients with type 3 atresia. annular pancreas were detected in 11 patients(20%) and 7 patients(13%) with malrotation. Diamond duodenoduodenostomy (DDD) was the procedure of choice in 38 patients (70%).

CONCLUSION:

The double bubble sign in erect abdominal X-ray is still diagnostic for cases with complete obstruction. High index of suspicion in patients with bilious vomiting is important for early referral, Diamond duodenoduodenostomy is very effective technique in most cases with very good post operative results.

KEY WORDS: congenital duodenal obstruction, duodenal atresia, diamond duodenoduodenostomy.

INTRODUCTION:

Congenital obstruction of the duodenum including duodenal atresia and stenosis occurs

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approximately 1-10000 live birth^(1,2,3,4), while annular pancreas is a rare anomaly, and may remain a symptomatic throughout life, but most commonly occur in infancy or early childhood⁽⁵⁾. Duodenal atresia classified into three main types: Type 1 or web with diaphragmatic membrane and sometimes the membrane experienced a wind

sock appearance. Type 2 with short fibrous cord connecting both ends. Type 3 (rare) with mesenteric defect and complete separation of both ends^(2,4).

Partial obstruction of duodenum may also be secondary to band of Ladd in association with malrotation or annular pancreas⁽⁶⁾, or preduodenal portal vein as a rare cause of duodenal obstruction⁽⁷⁾.

Bile stained vomitus in neonates is the typical presentation of duodenal atresia but in few cases the atresia is proximal to the ampulla and the vomitus is free from bile^(8,9), polyhydramnios is positive in 30-50% of cases and about 1/3 of infants have associated down syndrome^(4,8,9).

The detection of double bubble sign on plain radiograph of abdomen lead to suspicion of a number of anomalies, all of which lead to duodenal obstruction⁽¹⁰⁾.

The choice of surgical procedures is largely based on surgeon preference and duodenoduodenostomy has been used to bypass the obstruction⁽¹¹⁾.

In 1977 Kimura performed the diamond shape duodenoduodenalanastomosis (DDD)⁽¹²⁾ and later on transanastomotic feeding tube used after operation for duodenal atresia and obstruction lead to earlier full pre anastomotic feeding⁽¹³⁾, the various types of corrective procedures were similar in final outcome in term of morbidity and total hospitalization time⁽¹⁴⁾.

The surgical management of intestinal malrotation is the classical Ladd's Procedure⁽¹⁵⁾. Annular pancreas successfully corrected by duodenal bypass instead of releasing the annulus around duodenum⁽¹⁶⁾.

PATIENTS AND METHODS:

In this study patients were collected prospectively through the period between first of January 2010 to the end of January 2013 in pediatric surgical ward, at Al-khansaa teaching hospital in Mosul/Iraq; all of our patients were evaluated for polyhydramnios, infection or drugs ingestion in the 1st trimester. The presenting symptom were vomiting ((bilious& non bilious)), mild epigastric distension and in some patients associated with dehydration and Jaundice. Some patients presented primarily because of associated anomalies.

Incidental prenatal diagnosis by ultrasound was the other mode for presentation in few patients; nasogastric tubes were inserted in all neonates to assess the presence of associated esophageal

atresia and as part of preoperative resuscitation measures.

Erect abdominal x-ray were done for all patients and sometimes repeated after aspiration of gastric content, to assess the radiological findings which was either double bubble or single airfluid sign. Other investigations used for assessment includes CBC, chest X-ray, serum electrolytes, ultrasound of abdomen and TSB.

Barium meal was done selectively in patients outside neonatal periods or those with atypical clinical and radiological findings.

All patients were resuscitated with fluid, antibiotics and vitamin K before surgical intervention.

Exploration was done through supraumbilical transverse incision with preservation of umbilical vein. Operative findings were evaluated in relation to the cause of obstruction, site, degree of proximal dilatation, presence of distal obstruction and other associated anomalies.

Different surgical procedures were used according to the operative findings which were either diamond duodenoduodenostomy(DDD), side to side duodenoduodenostomy(SSDD), side to side duodenojejunosomy(SSDJ), Ladd's procedure and Heinecke-Mikulicz (HM) or combined procedures.

Transanastomotic tube was inserted in all patients with anastomosis for drainage and postoperative feedings. All anastomosis were done using single layer interrupted long lasting absorbable suture materials (PDS).

The patients kept for a period of 6-10 days postoperatively, feeding started in the 2nd -3^{ed} postoperative day through transanastomotic tube, with full oral intake in the 6th -8th day. Postoperative complications and mortality were secondary to associated anomalies, delay diagnosis and septicemia and two patients died before surgical intervention. The patients were followed for about 1-2 years.

RESULTS:

Fifty six patients were admitted to our center, 31(55.5%) male and 25 (44.5%) female, 10 of our patients were referred from other cities nearby Mosul city, 80% of them were full term.

The majority of our patients(45) presented in the first 4 days of life (80%) and 34 of them(60%) in the first 2 days, while 11 patients (20%) were delayed after the seven day of life ,one patients aged 2 months proved to have fenestrated web and another patients with annular pancreas. (Table.1.).

CONGENITAL DUODENAL OBSTRUCTION

Table 1: Age presentation.

Age	No.			%	
1-day	23	34	45	60	80
2-day	11				
3-day	5				
4-day	6	11		20	
1-wk	3				
1-month	6		11	20	
2-months	2				
Total	56			100	

Polyhydramnios were positive in 34 of our patient's mother (61%) and only 7 patients (20%) out of 36 patients diagnosed as upper intestinal obstruction on prenatal ultrasound examination in the third trimester.

Vomiting was the main presenting symptom in 50 patients (90%), it was bile stained in 42 patients (85%) and nonbile stained in 8 patients, failure to pass meconium and choking on feeding were the presenting feature in cases

associated with imperforate anus and esophageal atresia.

Erect abdominal x-ray was done to all patients and show typical double bubble gas shadow sign in 39 patients (70%), 15 of them show positive distal gas shadow and 24 show no distal gas shadow, the remaining 17 patients (30%) show single air fluid level with positive distal gas in 11 and no distal gas in 6 patients. (Table .2.).

Table 2: x-ray findings.

Double bubble (39) 70%		Single air- fl (17) 30%		
gas+ 15	_ gas 22	+ gas 11	_ gas 6	
ann. Panc. 5	3	3	-	11
Mal. 3	-	3	1	7
Web 7	11	5	2	25
Artesia -	8	-	3	11
	2 death preop.			2
Total				56

Eight patients underwent Barium meal study including those aged one month and more and it was positive web in 6 patients (5 of them with wind sock deformity), malrotation in one patient

and annular pancreas in the other.

Associated anomalies were 18 including 8 patients with Down syndrome (trisomy 21) (Table .3.).

Table 3: Associated anomalies.

G I T	Renal	Chromosomal	C V S	Total
TEF (2)	Polycys. (1)	Down (8)	VSD (4)	
Imp.anus (2)	Ectopic (1)			
4	2	8	4	18

Operative findings at time of exploration shows 36 atresia (67%) including 25 web, type 2 atresia in 8 patients and 3 patients with type 3 atresia,

annular pancreas was detected in 11 patients (20%) and 7 patients (13%) had malrotation 3 of them with non-ischemic mid gut volvulus. (Table .4.).

Table 4: Operative findings.

Type of obstruction		No.	
Atresia	Type 1(web)	25	67%
	Type 2	8	
	Type 3	3	
Annular pancreas		11	20%
Malrotation		7	13%
Total		54	100%

The degree of duodenal dilatation was found to be mild in 17 patients, moderate in 29 and severe in 8 patients with type 2 and 3 atresia.

The selection of procedure mainly depends on type of the lesion. Diamond duodenoduodenostomy (DDD) was the procedure of choice in 38 patients (70%) including web, type 2 atresia and annular pancreas. In four patients with duodenal web and mild dilatation, Heinecke-Mikulicz duodenoduodenostomy

(HMDD) performed with excision of the web and suturing the longitudinal incision in transverse manner, combined (DDD) and (HMDD) were used in one patient with double duodenal and jejunal atresia.

Side to side duodenoduodenostomy (SSDD) was done in 3 patients with type 3 atresia, Ladd's procedure used to manage 7 patients with malrotation. (Table .5).

Table 5: Operative procedures.

Procedure	No	%
Diamond(DDD)	38	70%
Heinecke-Mikulicz	5	9%
Side-side	3	6%
Ladd	7	13%
Diam+ Heinecke-Mikulicz	1	2%
Total	54	100%

Postoperative complications and mortality were not related to type of procedure. Complications were detected in 4 patients, including prolonged ileus & wound infection (treated conservatively), burst abdomen and adhesive intestinal obstruction (treated by releasing the adhesion surgically).

Mortality rate in this study was about 12% (7 patients), 2 of them died before surgery because of respiratory depression secondary to esophageal atresia, and septicemia secondary to delay diagnosis. Five patients died postoperatively, 3 of them secondary to congenital heart disease and esophageal atresia and 2 patients secondary to sepsis and electrolyte disturbance and delayed referral.

DISCUSSION:

Congenital duodenal obstruction accounting for nearly half of all cases of neonatal intestinal obstruction and affecting boys more frequently than girls (17,18).

Antenatal Ultrasound diagnosis of a partial or complete duodenal obstruction based on detection of the so-called double bubble fluid filled sign can help to determine the prognosis, and to prepare for appropriate treatment

modalities in the postnatal period and surgeons can search for known associated anomalies and plan ahead for an appropriate timing of the operative treatment which may help to decrease the overall morbidity (19,20).

In this study (34) patients their mothers showed polyhydramnios (61%) but only 7 (20%) were diagnosed prenatally as duodenal obstruction by ultrasound, this may be attributed to lack of expert and well trained ultrasonographer and new version of ultrasound specified for neonates. All these patients proved to have complete obstruction (atresia). We suggested that polyhydramnios should raise the suspicion of upper GIT anomalies including complete type of duodenal obstruction.

Despite earlier diagnosis and early surgical intervention, there were no significant differences in outcome comparing to other cases who presented without delay postnatally. Prenatal diagnosis will be valuable in preventing the problems of delay diagnosis which will increase morbidity and mortality as reported by QuingGiang et al and Coheenoverbrecks (18,21).

The age of patients at time of presentation was variable according to the cause of duodenal

obstruction in which patients with complete duodenal obstruction presented during the first few days of life in contrast to those with incomplete or intermittent obstruction⁽²²⁾. This support age presentation in our study, because 80% of our cases presented in the first 3-4 days of life and most of them proof to have complete obstruction intra operatively (duodenal atresia or malrotation with volvulus).

Vomiting (mainly bile stained) is still the main presenting symptoms⁽¹⁸⁾, in our study 85% presented with bile stained vomitus even those who were diagnosed prenatally. In 2 of our patients with esophageal atresia and tracheoesophageal fistula, the diagnosis was made incidentally during assessment for distal gas in stomach; this rare association makes us recommend evaluation for duodenal atresia in every case with esophageal atresia as the approach of management will be different.

The nonbilious vomiting in the remaining 15% contributed mainly to partial obstruction either by fenestrated web which lead to delay in their diagnosis similar to the 2 cases reported by Mehmet M et al⁽²³⁾, or secondary to malrotation with intermittent partial obstruction as described by M. D.Macaias Robles et al⁽²⁴⁾, from the other hand non bilious vomiting may rarely be a result of atresia proximal to the ampulla of Vater^(8,9), but none of our patients have had proximal obstruction.

Children born with congenital intrinsic duodenal obstruction (atresia) has been reported to have a high incidence of down syndrome (trisomy 21) and congenital heart disease⁽²⁵⁾, It has been found that about 30% of duodenal atresia have Down syndrome⁽²⁶⁾. In this study 22% (8 patients) had Down syndrome out of 36 of duodenal atresia and its nearly similar to what reported in most literatures, this slightly lower incidence might be related to under recorded cases of fenestrated web with delay diagnosis and presentation outside the neonatal period as in Smith et al and Lee et al who both reported cases with duodenal obstruction associated with Down syndrome in late childhood^(27,28).

Preoperative plain radiography was sufficient to diagnose duodenal obstruction in all patients except those with malrotation and/or partial

obstruction⁽²²⁾. For the neonates with classical appearance of double bubble sign on erect abdominal x-ray, additional radiological investigation is unnecessary and the surgeon is alerted to plan for surgery since all congenital causes of duodenal obstruction require surgery⁽¹⁰⁾.

In our opinion bilious vomiting and suggestive clinical examination with the presence of double bubble sign were the most dependable for diagnosis of duodenal obstruction and decision making for surgical exploration. Seventy percent (70%) of our patients show the classical double bubble sign and most of them proof to be complete obstructive lesion and this is similar to Quing-Giang et al who found that 68% of his patients demonstrate typical double bubble sign⁽¹⁸⁾.

The presence of single air fluid level is found in 30% of our patients, most of them have partial obstruction with distal gas shadow, in such an equivocal findings contrast study is indicated specially in cases presented after one week or more with a typical signs and symptoms, we believed that proper preparation for erect abdominal X-ray with aspiration of the stomach content and installation of air then repeating the x-ray might give more informative findings and might decrease the need for contrast study which is only used in 8 of our patients to confirm the diagnosis and in 6 of them suggest fenestrated web that proved surgically, in addition to one malrotation and one annular pancreas.

The upper GIT contrast study is the investigation of choice for malrotation presenting non acutely⁽²⁴⁾, 6 neonate out of 7 with malrotation in this study presented with acute intestinal obstruction and 4 of them show non ischemic volvulus while the rest shows single air fluid level with opacity of abdominal gases on erect abdominal x-ray.

Our operative findings regarding the cause of obstruction was mainly duodenal atresia in 67% (of different types) and 20% annular pancreas and only 13% malrotation, this findings is similar to Kadha et al⁽²²⁾ but it's totally different from Quing Jiang et al in which malrotation accounts 54% of his series out 287 cases⁽¹⁸⁾, the large number of cases and the long period of patients collection (more than 9 years) might play a role in this difference. (Table .5.).

Table 6:Operative findings comparative study.

Type of obstruction	Our study (56 pat.)	Kadah(71 pat.)	QuingJiang(287 pat.)
Artesian	67%	69%	23%
Annular pancreas	20%	20%	22%
Malrotation	13%	11%	54%

Our policy in managing every case of duodenal obstruction intra operatively is to pass a nasogastric tube and trying to push it distally to the site of obstruction specially in cases of duodenal web ,annular pancreas and malrotation, at the same time distal duodenum and beginning of jejunum should be irrigated by normal saline down to the ileocecal valve to exclude associated distal obstruction ,by this we decrease the likelihood of reoperation secondary to missed obstruction to about 0%, as none of our patients reoperated because of missed secondary obstruction which is reported by some literatures to be 4-14% , and they found that missed lesion during the initial surgery are still the major

reason for reoperation specially in cases of duodenal web, malrotation and multiple webs ^(30, 31, 32).

By this policy we reported a rare combination of double web, the first web was classical at the second part of duodenum, while the second one found distal to the D-J junction fig (2) which is extremely rare combination as stated by Ganguly S et al ⁽³³⁾. A classical DDD done for the duodenal web, and Heinecke-Mikulicz with excision for the second web as there was great discrepancy between both ends of bowel lumen for the proximal one and no such difference for the distal atresia, this is similar to a case reported by Shilpa Sharma et al ⁽³⁴⁾.



Fig 2: Double web, duodenal and proximal jejunum(pediatric surgery center, Al-Khansaa teaching hospital, Dr. Bassam Alabbasi)

Kimura found that DDD have very low rate of complications, with good long term results than other types of duodenoduodenostomy and drastically reduces the time of postoperative hospitalization ⁽³⁵⁾.

This technique based on transverse incision in the distal end of the proximal duodenum and longitudinal incision in the distal one with double layer anastomosis without gastrostomy ⁽¹²⁾.

In this study we found that Diamond DDD is the procedure of choice for managing 38 of our patients including web, annular pancreas and type 2 artesian table (5), we agree with kadha et al who recommend DDD as standard technique for patients with intrinsic obstruction , annular pancreas and duodenal diaphragm as it prevent any injury to duodenal ampulla or pancreatic ducts ⁽²²⁾, in addition to that we also found that

it's not a lengthy procedure with less bleeding as compared with excision of the web, that's why its preferred by most authors , at the same time it prevents re stenosis and pancreatic fistula⁽³⁶⁾. All these advantages encourage us to use this technique in managing most causes of congenital duodenal obstruction.

The wind sock deformity is rare anomaly and the second part of duodenum is the most common site ⁽³⁷⁾. Five of our patients with delayed presentation proof to have such anomalies with fenestrated web diagnosed by Barium meal, in these cases we found that it's better to do duodenal web excision with Heinecke-Mikulicz anastomosis as this will confirm our diagnosis and avoid faulty proximal duodenoduodenostomy and this is similar to 2 cases reported by Mehmet M et al.⁽²³⁾.

Although proximal massive dilated duodenum may affect anastomosis healing and may cause postoperative functional obstruction and may need tapering duodenoplasty⁽¹⁸⁾, none of our patients with severe proximal duodenal dilatation develop such complication and this might be attributed to wide caliber anastomosis with good drainage achieved by DDD.

There have been several reports indicating that Ladd procedure and duodenoduodenostomy can be performed laparoscopically^(38, 39), but none of our patients treated laparoscopically as we lack the experience and adequate instrumentation, in spite of that we proposed that patient selection for laparoscopy is vital for good results.

We agree with Nasir A et al that high index of suspicion in a neonates with bilious vomiting, rapid diagnosis, and appropriate operative therapy results in favorable outcome in patients with malrotation as its liability to volvulus is a time bomb lying within the abdomen, Ladd procedure is the surgical treatment of choice for all malrotation, with derotation of bowel if volvulus present⁽⁴⁰⁾, fortunately our patients with volvulus were non ischemic and Ladd's procedure was done in all of them without need for resection.

The mortality rate for duodenal obstruction has gradually decreased and the survival rate now exceeds 90%⁽³⁾. Kadha et al reported 21% mortality in his series and he attributed this high rate to gastroduodenal dysfunction and gastric dilatation with its sequel leading to sepsis⁽²²⁾. We reported 12% mortality which is slightly higher than other series like Quing Jiang et al who reported about 6% mortality⁽¹⁸⁾.

The main risk factors for death are cardiac anomalies, prematurity, sepsis, pneumonia and surgical complications like short bowel syndrome, anastomotic leak, and gastroduodenal dysfunction^(28, 3). All our deaths were secondary to associated anomalies and sepsis which is similar to Quing Jiang et al who stated that multiple congenital anomalies and sepsis are still the main risk factors for mortality accounting for nearly 80% for all postoperative deaths⁽¹⁸⁾. In our study we added delay referral as an additional important factor predispose to sepsis secondary to aspiration pneumonia, electrolyte and acid base disturbance which can be avoided or decreased by high index of suspicion, early diagnosis and referral with urgent surgical treatment.

CONCLUSION:

Congenital duodenal obstruction has different etiological predisposition. The double bubble sign in erect abdominal X-ray is still diagnostic for cases with complete obstruction. High index

of suspicion in patients with bilious vomiting and abnormal findings on erect abdominal x-ray is important for early referral, early diagnosis and early intervention with better results and less mortality rate. Careful evaluation for distal bowel by saline irrigation is vital to rule out rare second obstruction. Diamond duodenoduodenostomy is very effective technique in most cases with very good postoperative results even in markedly dilated proximal duodenum.

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