Abnormalities of intestinal rotation and congenital intrinsic duodenal obstruction

Ahmed H. Al-Salem, Mukul R. Kothari, Mohammad M. Oquaish, Suzie S. Khogeer and Mohammed Samir S. Desouky

Background **Congenital duodenal obstruction (CDO) is a unique anomaly with variable presentation depending on the site, type of obstruction, as well as the presence of associated anomalies. This is a review of our experience with 51 infants and children with CDO, stressing on the importance of associated rotational abnormalities of the gut.**

Methods **Over a period of 18 years (January 1993–December 2011), 51 infants and children with CDO were treated at our hospital. Their medical records were reviewed retrospectively for age at diagnosis, sex, gestation, birth weight, clinical features, associated anomalies, method of diagnosis, treatment, and outcome.**

Results **Fifty-one (27 boys and 24 girls) infants and children with intrinsic CDO were treated. Their birth weight ranged from 1.38 to 3.8 kg (mean 2.3 kg). Sixteen (31.4%) were premature. The mean maternal age was 25 years (range 18–40 years). Seven presented at 5 months, 18 days, 3.5 years, 1.8 years, 18 days, 1.5 years, and 2 months of age, respectively. Associated anomalies were observed in 34 (66.7%) of them. Sixteen (31.4%) had Down's syndrome and 11 (21.6%) had congenital heart disease. Nine (17.6%) had rotational abnormalities of the gut, including two with situs inversus. The site of duodenal obstruction was located in the second part of duodenum in 48 (94%) of them. In two, the site of obstruction was in the fourth part of duodenum, whereas in the third it was located in the third part of duodenum. The causes of obstruction were duodenal atresia in 20 (39.2%), without a gap in eight (15.7%) and with a gap in 12 (23.5%), duodenal stenosis in 11 (21.6%), and duodenal diaphragm in 20 (39.2%). In 14 (27.5%) patients, an annular pancreas was observed. The different operative procedures were as follows: duodenoduodenostomy in 26; excision of the duodenal diaphragm and duodenoplasty in 12; Ladd's procedure, excision of the duodenal diaphragm, duodenoplasty, and appendectomy in four; Ladd's procedure, appendectomy, and duodenoduodenostomy in two; and duodenojejunostomy in seven. Two underwent reduction duodenoplasty to decrease the size of the duodenum. Three required reoperations, two because of an anastomotic leak and another because of duodenal dysfunction. Sixteen (31.4%) required total parenteral nutrition. Four died, resulting in an overall survival of 92.2%.**

Conclusion **CDO is classified as intrinsic and extrinsic depending on the cause. Rotational abnormalities including situs inversus are among the rare but interesting anomalies associated with intrinsic CDO. The presence of malrotation or nonrotation with congenital bands does not exclude the presence of an associated intrinsic cause, which should be looked for intraoperatively. This is to obviate the possibility of further subsequent operative treatment.** ***Ann Pediatr Surg*** **9:61–64 © 2013 Annals of Pediatric Surgery.**

Annals of Pediatric Surgery 2013, **9**:61–64

Keywords: congenital duodenal obstruction, duodenal diaphragm, malrotation, situs inversus

Department of Pediatric Surgery, Maternity and Children Hospital, Dammam, Saudi Arabia

Correspondence to Ahmed H. Al-Salem, FRCSI, FACS, FICS, PO Box 61015, Qatif 31911, Saudi Arabia
Tel: +966546102999; fax: +96638630009; e-mail: ahsalsalem@hotmail.com

Received 24 August 2012 **accepted** 23 February 2013

Introduction

Congenital duodenal obstruction (CDO) is one of the relatively common congenital anomalies with an estimated incidence of 1 : 3000 to 1 : 5000 live births [1,2]. Over the years, the prognosis of infants and children with CDO has improved markedly, but several factors still continue to affect the overall outcome including prematurity, a high incidence of associated anomalies, and reoperations [1–6]. The clinical presentation of CDO is variable depending on the site, the type of obstruction, and the presence of associated anomalies. In the majority of cases, the site of obstruction is postampullary, and there is a high association with Down's syndrome. This study is an evaluation of our experience with 51 patients with CDO, stressing on the importance of associated rotational abnormalities of the gut.

Patients and methods

Over a period of 18 years (January 1993–December 2011), a total of 51 patients with intrinsic CDO were treated at our hospital. Their medical records were reviewed retrospectively for age at diagnosis, sex, gestation, birth weight, history of polyhydramnios, presentation, associated anomalies, method of diagnosis, type of treatment, and outcome.

Results

A total of 51 infants and children with intrinsic CDO were treated at our hospital. There were 27 boys and 24 girls. Their birth weight ranged from 1.38 to 3.8 kg (mean 2.3 kg). Sixteen (31.4%) of them were premature. The mean maternal age was 25 years (range 18–40 year). All patients presented at or were referred to our hospital within 2 weeks of life, except seven, who presented at 5 months, 18 days, 3.5 years, 1.8 years, 1.5 years, 18 days, and 2 months of age, respectively. All seven had a duodenal diaphragm with a hole, which was the reason for the delayed presentation, except one, who had duodenal atresia. Three of them presented late, with complete

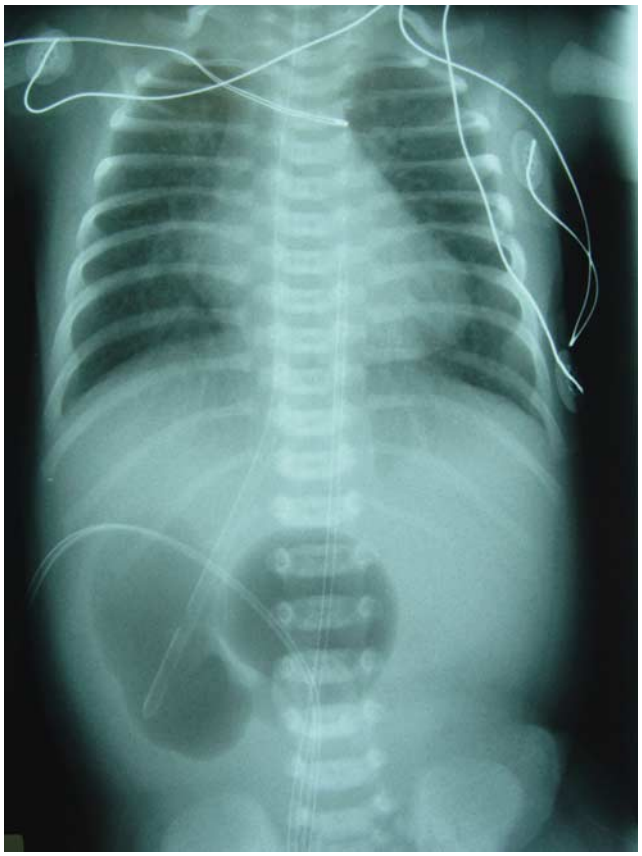
1687-4137 © 2013 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000428236.49123.e0

duodenal obstruction after eating dates with their seeds, which resulted in occlusion of the hole in the duodenal diaphragm. The remaining patients presented with bile-stained vomiting. Upper abdominal distension was seen in 25 (49%) of them and polyhydramnios was reported in 18 (35.3%). Intraoperatively, the site of duodenal obstruction was postampullary in all of them. Associated anomalies were seen in 34 (66.7%) of them (Table 1).

Table 1 Associated anomalies

Associated anomalies	*N* (%)
Down's syndrome	16 (32)
Congenital heart disease	12 (23.5)
Rotational abnormalities of the gut	9 (17.6)
Anorectal malformations	3 (5.9)
Esophageal atresia and tracheoesophageal fistula	1 (1.9)
Hydronephrosis	1 (1.9)
Syndactyly	1 (1.9)
Polydactyly	1 (1.9)
Duplication cyst	1 (1.9)
Meckel's diverticulum	2 (3.9)
Multicystic left kidney	1 (1.9)
Arteriovenous malformations	1 (1.9)
Dysplastic thumb	1 (1.9)
Eventration of right diaphragm	1 (1.9)
Pyloric atresia	2 (3.9)
Congenital leukemia	1 (1.9)
Meconium cyst	1 (1.9)
Jejuna atresia	2 (3.9)

Fig. 1

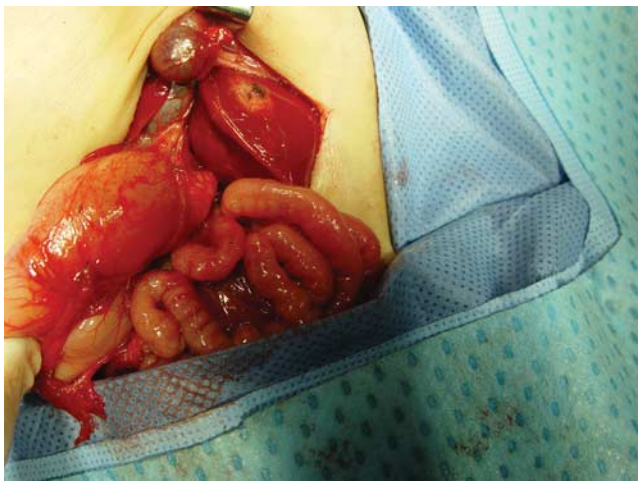


Plain abdominal radiograph showing duodenal obstruction in a patient with situs inversus. Note the reversed double-bubble sign and the absence of gas distally.

Sixteen had Down's syndrome (31.4%) and 12 (23.5%) had congenital heart disease. Nine (17.6%) had rotational abnormalities of the gut, including two with situs inversus. These two patients presented with bile-stained vomiting immediately after birth, and abdominal radiographs showed the classic double-bubble sign, but the stomach was on the right side (Fig. 1). The diagnosis of situs inversus was confirmed by a preoperative abdominal ultrasound. Echocardiography was normal in one patient, and in the other, there were features of Fallot's tetralogy. Intraoperatively, one had duodenal atresia with a gap, whereas the other had a duodenal diaphragm (Fig. 2). In the remaining seven patients, the diagnosis of associated malrotation was made intraoperatively. Five had malrotation and two had nonrotation of the bowel with Ladd's bands; however, in addition, they had intrinsic duodenal obstruction, which was diagnosed intraoperatively. Three of them had duodenal atresia and four had a duodenal diaphragm with a hole. Interestingly, two of our patients had congenital pyloric atresia associated with duodenal atresia in the fourth part of the duodenum. This resulted in a closed duodenal loop with pyloric atresia at one end and duodenal atresia at the other end, with accumulation of biliary and pancreatic secretions. This in turn resulted in duodenal perforation in one of them. In both, the diagnosis of associated duodenal atresia was made intraoperatively. One of our patients had dysmorphic features, esophageal atresia with tracheoesophageal fistula, and hydronephrosis. Intraoperatively, this patient had duodenal atresia, an annular pancreas, and a preduodenal portal vein.

All patients were operated upon, and intraoperatively, the site of duodenal obstruction was located in the second part of duodenum in 48 (94%) of them. In two, the site of obstruction was in the fourth part of duodenum, whereas in the third it was located in the third part of duodenum. The cause of obstruction was duodenal diaphragm in 20 (39.2%), duodenal stenosis in 11 (21.6%), and duodenal atresia in 20 (39.2%). Duodenal atresia was without a gap

Fig. 2



Clinical intraoperative photograph of a patient with situs inversus and congenital duodenal obstruction. Note the liver on the left side and the stomach and dilated duodenum on the right side.

Table 2 The different operative procedures

Operative procedure	Number of patients
Duodenoduodenostomy	26
Excision of the duodenal diaphragm and duodenoplasty	12
Ladd's procedure, excision of the duodenal diaphragm, duodenoplasty, and appendectomy	4
Duodenojejunostomy	7
Ladd's procedure, appendectomy, and duodenoduodenostomy.	2
Gastrostomy	4
Transanastomotic tube	6

in eight (15.7%) patients and with a gap in 12 (23.5%). In 14 (27.5%) patients, an annular pancreas was observed. This was associated with intrinsic duodenal obstruction in the form of duodenal atresia in six and duodenal stenosis in eight. The different operative procedures performed are shown in Table 2. Duodenoduodenostomy was performed in 26 patients; excision of the duodenal diaphragm and duodenoplasty in 12; Ladd's procedure, excision of the duodenal diaphragm, duodenoplasty, and appendectomy in four; Ladd's procedure, appendectomy, and duodenoduodenostomy in two; and duodenojejunostomy in seven. Gastrostomy was performed in four patients, and a transanastomotic tube was used in six. Both these procedures were used initially in the series. Subsequently, none of our patients had gastrostomy or the use of a transanastomotic feeding tube as a part of their operative management. Sixteen (31.4%) required total parenteral nutrition. Two patients underwent reduction duodenoplasty to decrease the size of the duodenum. Three patients required reoperations, two because of an anastomotic leak and another because of duodenal dysfunction. The patient with duodenal dysfunction underwent reduction duodenoplasty and subsequently had a good outcome. Four patients died, resulting in an overall survival of 92.2%. In all four patients, associated anomalies and sepsis were the causes of death.

Discussion

CDO is one of the common congenital anomalies. The exact incidence of CDO in Saudi Arabia is not known. The Maternity and Children Hospital is the main referral hospital in the Eastern Province of Saudi Arabia, with an annual delivery rate of about 10 000–12 000. This gives an estimated incidence of CDO of about 1 : 4000 live births, which is similar to that reported internationally [1–3]. CDO is classified as extrinsic and intrinsic depending on its cause. Extrinsic causes include malrotation with Ladd's bands, a duplication cyst, an annular pancreas, and a preduodenal portal vein. Although annular pancreas is classified as an extrinsic cause of CDO, all our patients with an annular pancreas had an underlying intrinsic cause for the obstruction. There was associated duodenal stenosis in eight patients and atresia in five. Intrinsic causes for CDO include atresia, stenosis, and a duodenal diaphragm with or without a hole. It is this group of patients with a duodenal diaphragm who can remain asymptomatic to present subsequently at an older age or even during adulthood [7,8]. Three of our patients with a duodenal diaphragm presented beyond 1 year of age with complete duodenal obstruction as a result of seeds occluding the hole in the duodenal diaphragm. It is also interesting to note that in all our patients the obstruction was postampullary, and the majority (40%) had a duodenal diaphragm. An interesting report from Taiwan showed racial differences in the clinical characteristics of CDO as their patients had lower incidence of postampullary obstruction and lower incidence of associated Down's syndrome [6]. This is in contrast to our report and reports from Western countries where, in the majority, the site of obstruction is postampullary and associated Down's syndrome was observed in 30–50% of patients [1–4]. There is an association between the incidence of Down's syndrome and maternal age. The incidence of Down's syndrome in our series was 31.4%, which is lower than that reported by Akhtar and Guiney, and the reason for this is the higher proportion of younger mothers in our series when compared with that reported by Akhtar and Guiney [2]. The association of Down's syndrome negatively impacts the management and outcome of patients with CDO [9,10]. Another interesting association in our series was that of CDO with rotational abnormalities of the intestines. In nine (17.6%) of our patients there was an associated rotational abnormality of the intestines. Five had malrotation, two had nonrotation with Ladd's bands, and two had associated situs inversus; these two, in addition, had intrinsic duodenal obstruction, which was diagnosed intraoperatively. Situs inversus is extremely rare and is commonly associated with other cardiac and splenic malformations [11–13]. The diagnosis CDO in the presence of situs inversus can be made on the basis of plain erect abdominal radiographs that show reversed double-bubble signs. However, the diagnosis can be confirmed by barium meal and follow through. The presence of malrotation or nonrotation of the intestines with congenital bands should not exclude the possibility of an associated intrinsic CDO, which should be looked for intraoperatively after Ladd's procedure. This is to obviate the possibility of further subsequent operative therapy. We advocate the addition of an appendectomy to the operative procedures in these patients. This should simplify their future management in case they present with acute abdomen as the possibility of acute appendicitis and confusion associated with it because of its abnormal location are eliminated.

There are reports of successful endoscopic treatment of duodenal diaphragms, but the treatment for CDO is surgical correction, whether this is done through the standard upper abdominal transverse incision, umbilical crease incision, or the more recently developed laparoscopic technique [14–18]. Simple duodenoduodenostomy is the treatment of choice. Kimura *et al.* [19], in 1977, described the diamond-shaped duodenoduodenostomy, which was reported to be superior to simple duodenoduodenostomy in term of early tolerance to feeds. The use of transanastomotic tubes in CDO is still controversial. Recently, there was a report favoring the use of transanastomotic tubes as it shortens the time to full feeds and significantly reduces the need for central venous catheters and total parenteral nutrition [20]. We do not use transanastomotic tubes as our initial

experience with them in six patients was not favorable. They tend to recoil and cause anastomotic disruption. For those with a duodenal diaphragm, treatment involves excision of the diaphragm and duodenoplasty. Care should be taken because of the close proximity of the diaphragm to the ampulla of Vater, and if in doubt, a duodenoduodenostomy is an alternative procedure that can be used. Duodenojejunostomy should be avoided as a procedure to treat CDO. It is less physiological than duodenoduodenostomy and is associated with more complications, but it is a suitable procedure if the obstruction site is in the third or fourth parts of the duodenum. To obviate postoperative functional duodenal obstruction, we advocate reduction duodenoplasty as an initial procedure in those with a megaduodenum. With the recent advances in minimal invasive surgery, laparoscopy, which has been shown to be safe and effective, is likely to become the standard treatment for CDO [17,18].

Conclusion

CDO is one of the common causes of intestinal obstruction in infants and children and continues to present unique management challenges. Rotational abnormalities of the gut including situs inversus are among the rare but interesting anomalies associated with intrinsic congenital duodenal obstruction. The presence of malrotation with congenital bands and congenital duodenal obstruction does not exclude the possibility of an associated intrinsic cause, which should be looked for intraoperatively.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Eustace S, Connoly B, Blake N. Congenital duodenal obstruction: an approach to diagnosis. *Eur J Pediatr Surg* 1993; **3**:267–270.
2 Akhtar J, Guiney EJ. Congenital duodenal obstruction. *Br J Surg* 1992; **79**:133–135.
3 Gavopoulos S, Limas CH, Avtzoglou P, Violaki A, Grigoriadis G, Tryfonas G. Operative and postoperative management of congenital duodenal obstruction: a 10 year experience. *Pediatr Surg Int* 1993; **8**:122–124.
4 Bailey PV, Tracy TF Jr, Connors RH, Mooney DP, Lewis JE, Weber TR. Congenital duodenal obstruction: a 32-year review. *J Pediatr Surg* 1993; **28**:92–95.
5 Anatol TI, Hariharan S. Congenital intrinsic intestinal obstruction in a Caribbean country. *Int Surg* 2009; **94**:212–216.
6 Tsai LY, Hsieh WS, Chen CY, Chou HC, Tsao PN, Hus WM. Distinct clinical characteristics of patients with congenital duodenal obstruction in a medical center in Taiwan. *Pediatr Neonatol* 2010; **51**:343–346.
7 Bhat NA. Congenital duodenal diaphragm and enteroliths: a unique complication. *J Indian Assoc Pediatr Surg* 2009; **14**:226–227.
8 Loh D, Leese T, Anders S. Adult presentation of congenital duodenal diaphragm. *Gastrointest Endosc* 2010; **71**:654–655.
9 Niramis R, Anuntkosol M, Tongsin A, Mahatharadol V. Influence of Down's syndrome on management and outcome of patients with congenital intrinsic duodenal obstruction. *Clin Genet* 2009; **75**:180–184.
10 Cohen-Overbeek TE, Grijseels EW, Niemeijer ND, Hop WC, Wladimmiroff JW, Tibboel D. Isolated or non-isolated duodenal obstruction: perinatal outcome following prenatal or postnatal diagnosis. *Ultrasound Obstet Gynecol* 2008; **32**:784–792.
11 Luchtman M, Golan Y, Heldenberg D, Kessler F. Situs inversus abdominus in association with duodenal obstruction and internal hernia. *Am J Perinatol* 1933; **10**:255–257.
12 Nawaz A, Matta H, Hamchou M, Jacobsz A, Trad O, Al-Salem AH. Situs inversus abdominus in association with congenital duodenal obstruction: a report of two cases and review of literature. *Pediatr Surg Int* 2005; **21**: 589–592.
13 Brown C, Numanoglu A, Rode H, Sidler D. Situs inversus abdominalis and duodenal atresia: a case report and review of the literature. *S Afr J Surg* 2009; **47**:127–130.
14 Barabino A, Gandullia P, Arrigo S, Vignola S, Mattioli G, Grattarola C. Successful endoscopic treatment of a double duodenal web in an infant. *Gastrointest Endosc* 2011; **73**:401–403.
15 Takahashi Y, Tajiri T, Masumoto K, Kinoshita Y, Ieiri S, Matsuura T, *et al.* Umbilical crease incision for duodenal atresia achieves excellent cosmetic results. *Pediatr Surg Int* 2010; **26**:963–966.
16 Kozlov Y, Novogilov V, Yurkov P, Podkamenev A, Weber I, Sirkin N. Keyhole approach for repair of congenital duodenal obstruction. *Eur J Pediatr Surg* 2011; **21**:124–127.
17 Kay S, Yoder S, Rothenberg S. Laparoscopic duodenoduodenostomy in the neonate. *J Pediatr Surg* 2009; **44**:906–908.
18 Riquelme M, Aranda A, Riquelme-Q M, Rodriquez C. Laparoscopic treatment of duodenal obstruction: report on first experience in Latin America. *Eur J Pediatr Surg* 2008; **18**:334–336.
19 Kimura K, Tsugawa C, Ogawa K, Matsumoto Y, Asada S. Diamond-shaped anastomosis for congenital duodenal obstruction. *Arch Surg* 1977; **112**:1262–1263.
20 Hall NJ, Drewett M, Wheeler RA, Griffith DM, Kitteringham LJ, Burge DM. Trans-anastomotic tubes reduce the need for central venous access and parenteral nutrition in infants with congenital duodenal obstruction. *Pediatr Surg Int* 2011; **27**:851–855.