Duodenal Atresia and Stenosis: Long-Term Follow-Up Over 30 Years

By Mauricio A. Escobar, Alan P. Ladd, Jay L. Grosfeld, Karen W. West, Frederick J. Rescorla, L.R. Scherer III, Scott A. Engum, Thomas M. Rouse, and Deborah F. Billmire

Indianapolis, Indiana

Background: Duodenal atresia and stenosis is a frequent cause of congenital, intestinal obstruction. Current operative techniques and contemporary neonatal critical care result in a 5% morbidity and mortality rate, with late complications not uncommon, but unknown to short-term follow-up.

Methods: A retrospective review of patients with duodenal anomalies was performed from 1972 to 2001 at a tertiary, children's hospital to identify late morbidity and mortality.

Results: Duodenal atresia or stenosis was identified in 169 patients. Twenty children required additional abdominal operations after their initial repair with average follow-up of 6 years (range, 1 month to 18 years) including fundoplication (13), operation for complicated peptic ulcer disease (4), and adhesiolysis (4). Sixteen children underwent revision of their initial repair: tapering duodenoplasty or duodenal plication (7), conversion of duodenojejunostomy to duodenoduodenostomy (3), redo duodenojejunostomy (3), redo duodeno-

duodenostomy (2), and conversion of gastrojejunostomy to duodenoduodenostomy (1). There were 10 late deaths (range, 3 months to 14 years) attributable to complex cardiac malformations (5), central nervous system bleeding (1), pneumonia (1), anastomotic leak (1), and multisystem organ failure (2).

Conclusions: Late complications occur in 12% of patients with congenital duodenal anomalies, and the associated late mortality rate is 6%, which is low but not negligible. Follow-up of these patients into adulthood is recommended to identify and address these late occurrences.
J Pediatr Surg 39:867-871. © 2004 Elsevier Inc. All rights reserved.

INDEX WORDS: Duodenal atresia and stenosis, megaduodenum, duodenal dysmotility, tapering duodenoplasty, gastroesophageal reflux disease.

DUODENAL ATRESIA and stenosis is a frequent cause of congenital intestinal obstruction occurring in 1 per 5,000 to 10,000 live births, affecting boys more commonly than girls.[1] More than 50% of affected patients have associated congenital anomalies including pancreatic anomalies; intestinal malrotation; esophageal atresia; Meckel's diverticulum; variants of imperforate anus; congenital heart disease; central nervous system lesions; renal anomalies; and, rarely, biliary tract anomalies.[2,3] Down's syndrome occurs in approximately 30% of patients, polyhydramnios in 33% to 50%, and 45% are premature.[4,5]

In recent years, early postoperative survival rate has improved from 60% to 90%.[5-8] Improved operative techniques and contemporary neonatal intensive care unit (NICU) care result in a very low, early morbidity and mortality rate (5%).[5-8] Postoperatively, most patients are asymptomatic and show normal growth.[5,9-11] A few reports suggest that late complications may occur in 12% to 15% of patients.[5,12-14] The true incidence of long-term complications and mortality after initial repair of duodenal atresia is unknown. The availability of such information would aid the long-term care plan for these patients as they progress into childhood and adolescence. This review attempts to identify patients at risk for late complications that influence long-term management.

MATERIALS AND METHODS

Over a 30-year period (1972 to 2001) 592 infants with intestinal atresia were identified at the Riley Children Hospital, Indianapolis, IN. Patients were identified initially by ICD-9 diagnosis code 751.1 for intestinal atresia. Of these, the patients with the diagnosis of duodenal atresia or stenosis were selected for the occurrence of late morbidity and mortality by a retrospective analysis. IRB approval of the study design was obtained (IRB #303-01).

RESULTS

This study included 169 infants with duodenal atresia or stenosis. There were 89 girls and 80 boys. Sixty-three (37%) patients were premature (gestational age $\leq$ 37 weeks) and 46 (27%) had Down's syndrome. Seventy-seven (46%) babies had associated congenital anomalies including congenital heart disease in 46 cases, esopha-

Table 1. Duodenal Atresia and Stenosis

Late Complications	No. of Patients
Gastroesophageal reflux disease	13
Megaduodenum, motility disorder, +/− duodenogastric reflux	7
Peptic ulcer	4
Adhesive small bowel obstruction	4
Anastomotic dehiscence	2
Anastomotic stricture	1
Choledochal cyst	1
Duodenal blind loop syndrome	1
Missed second mucosal web	1
Gastric outlet obstruction	1

geal atresia in 14, imperforate anus in 6, both esophageal atresia and imperforate anus in 3, renal abnormalities in 8, biliary atresia in 2 (1 with situs inversus and polysplenia), polysplenia alone in 1, pyloric atresia in 1, gastroschisis with colonic atresia in 1, Hirschsprung's disease in 1, and jejunal duplication in 1.

At initial operation, duodenal atresia was noted in 137 instances and duodenal stenosis in 32. One hundred twenty-six babies had a type 1 mucosal atresia, 2 had a fibrous cord connecting 2 atretic ends (type 2), and 9 had a complete separation (a gap) between the 2 atretic ends (type 3). Duodenal stenosis was noted in 32 cases, including 9 with a windsock web containing a small aperture. At operation, 62 patients had annular pancreas, 54 anomalies of intestinal rotation and fixation (malrotation), 5 an anterior portal vein, and 3 a second more distal web. Operative management included duodenoduodenostomy in 143 cases, duodenotomy and web excision in 8, Heineke-Mikulicz type of duodenoplasty in 2, and duodenojejunostomy in 15. One patient had a gastrojejunostomy performed at an outside facility before admission to the Children's Hospital. In patients with duodenoduodenostomy, 87 patients had a side-to-side anastomosis, 52 had a diamond-shaped anastomosis performed (with only 5 being performed before 1992), in 4 the type of anastomosis was unclear, and 2 with a very dilated proximal duodenum had a concurrent antimesenteric, tapering duodenoplasty. A Ladd procedure and appendectomy was performed in all 54 infants who had malrotation.

Twenty children required further abdominal surgical procedures after displaying late complications (Table 1). Thirteen children required fundoplication because of severe gastroesophageal reflux disease (GERD) from 3 months to 7 years postoperatively (average, 5 years). In children with a duodenoduodenostomy, 3 had late complications including distal small bowel perforation treated with resection, duodenal obstruction (stricture) treated with a redo-duodenoduodenostomy, and duodenal perforation treated with repair of the anastomotic leak.

Three of the 15 children who had a duodenojejunostomy required conversion to a duodenoduodenostomy: 1 during resection of a choledochal cyst 13 years postoperatively, 1 for duodenal blind loop syndrome 16 years postoperatively, and the third for a partial small bowel obstruction. One patient had a redo duodenojejunostomy for anastomotic stenosis at 6 weeks of age. One child had an anastomotic leak after duodenojejunostomy requiring revision 3 weeks postoperatively. Another patient required anastomotic revision and tapering duodenoplasty because of recurrent bleeding from a peptic ulcer in a retained distal duodenal mucosal web at 18 years of age. One additional patient underwent takedown of a prior gastrojejunostomy, tapering duodenoplasty with duodenoduodenostomy for a bleeding duodenal ulcer at age 7 years.

Four children with poor weight gain and vomiting required delayed anterior duodenal plication for megaduodenum or duodenal motility disorder (Fig 1). Three required tapering duodenoplasty: 2 (at age 4 and 18 years) for megaduodenum, and 1 (age 14 years) for an
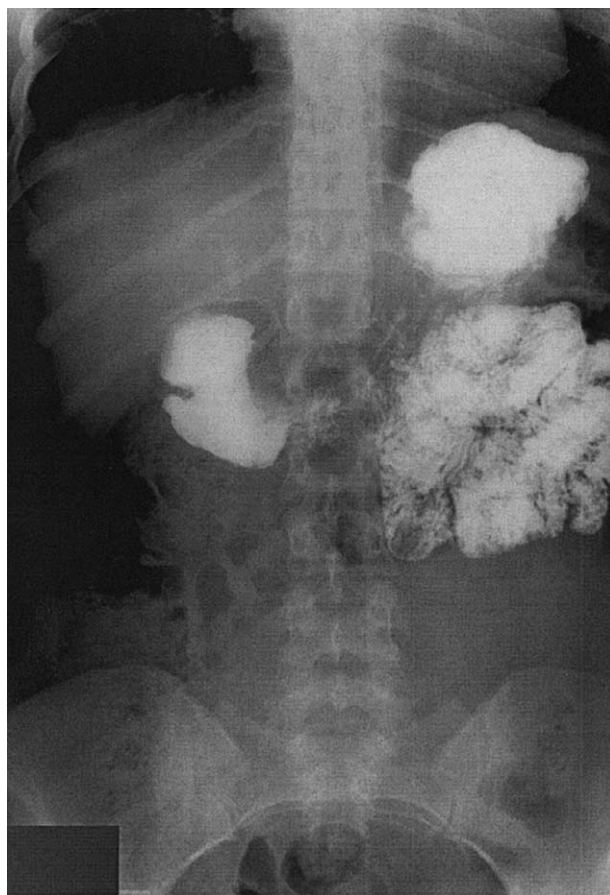

Fig 1. Megaduodenum. Upper GI series shows dilated first and second portion of the duodenum proximal to the anastomosis.

atonic, proximal duodenum. One child that required adhesiolysis at 2 months of age for gastric outlet obstruction had an abdominal wound dehiscence. Three children required suture ligation of a bleeding gastric ulcer. Two patients required enterolysis for adhesive postoperative distal small bowel obstruction at 3 months and 3 years, respectively. One child underwent incisional hernia repair at age 10 months.

Seven (11%) of the 62 patients with annular pancreas required additional operations. They represented 35% (7 of 20) of the patients in the study that required reoperation. These 7 cases included 3 children requiring tapering duodenoplasty, 2 a redo duodenojejunostomy (anastomotic leak [1] and recurrent duodenal bleeding [1]), and 1 revision of his duodenoduodenostomy and Nissen fundoplication for GERD. Another patient with annular pancreas underwent an unsuccessful Kasai procedure for biliary atresia, subsequent orthotopic liver transplantation, and oversewing of a bleeding gastric ulcer. Of the children with annular pancreas, the average age was 6 years at the time of reoperation, and 6 of the 7 children required 2 additional late operations.

One child with Down's syndrome required multiple operations including Nissen fundoplication and exploratory laparotomy for perforated small bowel from adhesions that was unrelated to the primary diagnosis. Five children with late complications had associated midline defects: 2 with imperforate anus (IA), 2 with esophageal atresia and tracheoesophageal fistula (TEF), and 1 with both. One child with IA had a duodenal anastomotic leak and underwent repair at 3 weeks of age. The child with both IA and TEF required a Nissen fundoplication for GERD.

There were 5 (3%) early deaths (<30 days postoperatively) all related to complex cardiac anomalies. There were 10 late deaths occurring from 3 months to 14 years postoperatively. Five deaths were caused by complex cardiac malformations (1 had Down's syndrome), central nervous system (CNS) bleeding after multiple neurosurgical procedures in 1, and aspiration pneumonia after repair of a TEF in 1. One child with Down's syndrome died of complications of duodenal atresia after an anastomotic leak. An additional baby died late after an unsuccessful hepatoportoenterostomy for biliary atresia. One child treated for gastric outlet obstruction died of respiratory failure after takedown of a jejunostomy tube site and small bowel resection.

DISCUSSION

Early mortality data after neonatal repair of congenital defects does not always accurately reflect the long-term outcomes. Early operative mortality after correction of duodenal atresia has been reported as low as 4% to 5%.[2,5]

In our series, the early mortality rate was 3%. Long-term survival rate is excellent at 86% to 90%.[2,5,9,10,11] Children with Down's syndrome or other associated congenital anomalies, especially complex cardiac defects, have a higher long-term mortality rate.[5,15] Five of our 10 late deaths (50%) occurred in patients with complex congenital heart disease, 2 had Down's syndrome, and one child, each, had IA and TEF.

Children with foregut anomalies including those with esophageal atresia and gastroduodenal dysmotility disorders are among the most likely to have gastroesophageal reflux.[16] Dysmotility in small bowel atresia may be related to damaged smooth muscle cells from ischemia,[17] hypoplasia of enteric nerves, and reduced smooth muscle immunoreactivity.[18] The dilated proximal atretic duodenum may be associated with disturbed transit.[18,19]

Before the mid 1970s, duodenojejunostomy was preferred for duodenal atresia and stenosis.[5,20,21] Very rarely a gastrojejunostomy was performed.[22] Duodenojejunostomy was associated with delayed anastomotic function often requiring use of transanastomotic feeding tubes or parenteral nutrition. Spigland and Yazbeck[23] described 6 children that required reoperation; 5 were initially treated with duodenojejunostomy and 1 treated with duodenoduodenostomy. Blind-loop syndrome appears to be more common in patients treated with duodenojejunostomy and may be improved with conversion to a duodenoduodenostomy.[24] In our series, the only case of blind-loop syndrome occurred in a patient with a duodenojejunostomy.

The procedure of choice for duodenal atresia and stenosis is duodenoduodenostomy. The operation has evolved from a side-to-side anastomosis to a proximal transverse to distal longitudinal ("diamond shaped") anastomosis.[5,12,13,20,21] At the time of operative repair, a small, red rubber catheter should be passed distally through the distal enterotomy, because a second mucosal web exists in 1% to 3% of cases, which, if missed, may result in a postoperative obstruction distal to the anastomosis.[5] One patient in this series presented with recurrent bleeding ulcer at a missed second mucosal web with a small aperture 18 years after the primary operation.

Megaduodenum occurred up to 18 years postoperatively. It is associated with poor weight gain, frequent vomiting, abdominal pain, and blind-loop syndrome.[20,21,23] An antimesenteric tapering duodenoplasty may be used during the initial procedure if the proximal duodenum is excessively floppy and distended.[5,25] However, in many infants, the proximal dilatation of the atretic duodenum resolves after relief of obstruction without a concomitant tapering duodenoplasty. Late onset of megaduodenum has also occurred in some infants that had minimal dilatation at the time of the original

repair. It is, therefore, difficult to recommend tapering in all the neonatal cases. Megaduodenum can be managed by either plication or tapering of the dilated atonic proximal duodenum alone[12,13] and possible revision of anastomotic strictures if present.[25]

Patients with annular pancreas had an increased tendency for late complications. Annular pancreas usually is associated with duodenal stenosis.[26] Infants with annular pancreas associated with duodenal obstruction generally are premature or small for gestational age.[27] In this series, 6 of the 7 children with annular pancreas that required reoperation had at least 2 operations up to 18 years after the initial procedure. This population represented 35% of the patients (7 of 20) that required operations for late complications. Moreover, complications of annular pancreas have been described in adults including peptic ulcer disease, gastric outlet obstruction, duodenal obstruction, pancreatitis, and gastric cancer.[28-31] These observations suggest that these patients require follow-up into their adult years.

Among the long-term complications noted in this study were delayed gastric emptying, severe gastroesophageal reflux, bleeding peptic ulcer, megaduodenum, duodenogastric reflux, gastritis, blind-loop syndrome, and intestinal obstruction related to adhesions. Children with GERD requiring fundoplication had surgery at the age of 5 years on average, and 68% of these patients required additional operations. Instances of small bowel obstruction occurred sporadically up to 17 years postoperatively. Children with associated midline anomalies such as TEF or IA had proportionally higher complication rates.

These observations indicate that long-term follow-up is essential for infants treated for duodenal atresia and stenosis, particularly those with associated annular pancreas, GERD, delayed gastric emptying, peptic disorders and megaduodenum. Care guidelines must be developed for long-term follow-up in infants with this condition that may be at risk for late complications. We recommend follow-up throughout childhood and adolescence into adulthood with a seamless transfer of care to general physicians aware of these potential late complications.

REFERENCES

1. Kimura K, Loening-Baucke V: Bilious vomiting in the newborn: Rapid diagnosis of intestinal obstruction. Am Fam Physician 61:2791-2798, 2000

2. Dalla Vecchia LK, Grosfeld JL, West KW, et al: Intestinal atresia and stenosis: A 25-year experience with 277 cases. Arch Surg 133:490-497, 1998

3. Murshed R, Nicholls G, Spitz L: Intrinsic duodenal obstruction: Trends in management and outcome over 45 years (1951-1995) with relevance to prenatal counselling. Br J Obstet Gynaecol 106:1197, 1999

4. Chabra R, Suresh BR, Weinberg G, et al: Duodenal atresia presenting as hematemesis in a premature infant with Down syndrome. J Perinatol 12:25-27, 1992

5. Grosfeld JL, Rescorla FJ: Duodenal atresia and stenosis: Reassessment of treatment and outcome based on antenatal diagnosis, pathologic variance, and long-term follow-up. World J Surg 17:301-309, 1993

6. Mooney D, Lewis JE, Connors RH, et al: Newborn duodenal atresia: An improving outlook. Am J Surg 153:347-349, 1987

7. Wesley JR, Mahour GH: Congenital intrinsic duodenal obstruction: A twenty-five year review. Surgery 82:716-720, 1977

8. Rescorla FJ, Grosfeld JL: Intestinal atresia and stenosis: Analysis of survival in 120 cases. Surgery 98:668-676, 1985

9. Feggetter S: A review of the long-term results of operations for duodenal atresia. Br J Surg 56:68-72, 1969

10. Stauffer U, Irving I: Duodenal atresia and stenosis: Long-term results. Prog Pediatr Surg 10:49-60, 1977

11. Kokkonen ML, Kalima T, Jaaskelainen J, et al: Duodenal atresia: Late follow-up. J Pediatr Surg 23:216-220, 1988

12. Ein SH: The late nonfunctioning duodenal atresia repair—A second look. J Pediatr Surg 35:690-691, 2000

13. Ein SH, Shandling B: The late nonfunctioning duodenal atresia repair. J Pediatr Surg 21:798-801, 1986

14. Okada PJ, Hicks B: Neonatal surgical emergencies. Clin Pediatr Emerg Med 3:3-13, 2002

15. Fonkalsrud EW, DeLorimier 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 43:79-83, 1969

16. Hassall E: Wrap session: Is the Nissen slipping? Can medical treatment replace surgery for severe gastroesophageal reflux disease in children? Am J Gastroenterol 90:1212-1220, 1995

17. Molenaar JC, Tibboel D, van der Kamp AW, et al: Diagnosis of innervation-related motility disorders of the gut and basic aspects of enteric nervous system development. Prog Pediatr Surg 24:173-185, 1989

18. Masumoto K, Suita S, Nada O, et al: Abnormalities of enteric neurons, intestinal pacemaker cells, and smooth muscle in human intestinal atresia. J Pediatr Surg 34:1463-1468, 1999

19. Takahashi A, Tomomasa T, Suzuki N, et al: The relationship between disturbed transit and dilated bowel, and manometric findings of dilated bowel in patients with duodenal atresia and stenosis. J Pediatr Surg 32:1157-1160, 1997

20. Kimura K, Mukohara N, Nishijima E, et al: Diamond-shaped anastomosis for duoduenal atresia: An experience with 44 patients over 15 years. J Pediatr Surg 25:977-979, 1990

21. Weber TR, Lewis JE, Mooney D, et al: Duodenal atresia: A comparison of techniques of repair. J Pediatr Surg 21:1133-1136, 1986

22. Puri P, Sweed Y: Duodenal obstructions, in Puri P (ed): Newborn Surgery. Oxford, UK, Botterworth-Heinemann, 1996, pp 290-297

23. Spigland N, Yazbeck S: Complications associated with surgical treatment of congenital intrinsic duodenal obstruction. J Pediatr Surg 25:1127-1130, 1990

24. Rescorla FJ, Grosfeld JL: Duodenal atresia in infancy and childhood: Improved survival and long-term follow-up. Contemp Surg 33:22-27, 1988

25. Adzick NS, Harrison MR, deLorimier AA: Tapering duodenoplasty for megaduodenum associated with duodenal atresia. J Pediatr Surg 21:311-312, 1986

26. Androulakis J, Colborn GL, Skandalakis PN, et al: Embryologic and anatomic basis of duodenal surgery. Surg Clin North Am 80:171-199, 2000

27. Sencan A, Mir E, Gunsar C, et al: Symptomatic annular pancreas in newborns. Med Sci Monit 8:CR434-7, 2002

28. Kelly SB, Williamson RC: Annular pancreatitis and gastric cancer. HPB Surg 7:69-75, 1993

29. Machado N, Rajan N, Rao BH: Annular pancreas in adults (2 case reports). J Postgrad Med 37(52B):53-55, 1991

30. Urayama S, Kozarek R, Ball T, et al: Presentation and treatment of annular pancreas in an adult population. Am J Gastroenterol 90:995-999, 1995

31. Naylor RG, Juler GL: Congenital causes of duodenal ulcers in adults. Arch Surg 111:658-662, 1976

Discussion

R.J. Touloukian (New Haven, CT): This is a wonderful long-term experience allowing us to review how we should manage new patients with duodenal atresia. We are aware that there is a spectrum of megaduodenum in these patients. Some patients have a larger duodenum at the time of diagnosis than others. Based on your report here, are you able to predict now which patients will require a duodenoplasty based on what their initial films looked like at the time of diagnosis? That might help us in identifying patients who might need a duodenoplasty at the time of the initial procedure.

M. Escobar (response): We identified 2 children in the 169 that had a tapering duodenoplasty for significant proximal dilatation at the initial operation. They did not have late complications. In our series, we found only 4 children that developed late-onset megaduodenum that eventually required either a plication or a tapering duodenoplasty. Others that had moderate proximal duodenal dilatation as neonates had this resolve after duodenoduodenostomy alone. So, at this point, we do not routinely perform a tapering duodenoplasty at the initial operation in the neonatal period unless the proximal duodenum is massively dilated.

S.H. Ein (Toronto, Ontario): A number of years ago, Stephens, Simpson, Shandling, and Ein reported 265 duodenal atresias, and our big problem was that we had 5 patients with dysfunctional duodenum that presented suddenly between 9 months and 24 years with a sudden obstruction. There was no anastomotic stricture, and we pleated them all. So on the basis of what Bob Touloukian said, I don't see how anybody can predict megaduodenum when you look at a dilated proximal duodenum in a newborn with duodenal atresia—they're all dilated, and what I've been doing for the last number of years is to pleat these or plicate them. You have to have a long-term follow-up to see if that's going to make any difference. So far, so good. Just like the person that jumped off the Empire State Building, Bob, when they passed the 52nd floor and you asked how they're doing, "So far, so good," but that's the only way you can do it, I think.

M. Escobar (response): Thank you.

B. Pettit (Atlanta, GA): I'm actually fascinated by the issue of long-term follow-up. My question has 2 parts. For young adults over the age of 18, how many complications did you actually see after the period when they most likely would have been transferred to the care of an general surgeon? And, in the complications that occurred in those over the age of 18, how many were cared for by your group or a pediatric surgery group as opposed to general group? This idea of a seamless transition to the care of an general physician is something we need to deal with as pediatric surgeons. We now have a huge cohort of young adults, in fact, probably some middle-aged adults that were operated on in the 1950s and 1960s. I'm not sure how clear we are when these young adults leave our follow-up care about the future follow-up they'll need. I work with a lot of general surgeons, and they don't remember the details of a lot of these anomalies, and they aren't familiar with the long-term complications that occur. So I'm interested in what happened, was there any experience with people over the age of 18 in your cohort?

M. Escobar (response): As I mentioned in our cohort, the oldest child in our database was 18 years. However, we did have 1 patient present quite recently at 24 years of age with gallstones that was not included. I am not sure if the general surgeons need our help in managing this complication. Whether the child is being followed up by a pediatric surgeon up until 18 years of age or by a primary care physician, pediatrician, or family practitioner, I think that it's our responsibility to discuss these potential problems with the doctor we're transferring care to and educating them concerning the natural history of the condition.