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Selective Management of Gastroschisis According to the Degree of Visceroabdominal Disproportion

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Objective
This study analyzed the factors influencing the postoperative results after the repair of gastroschisis defects during the past 27 years.

Summary Background Data
The clinical results after the repair of gastroschisis abdominal defects have improved appreciably during the past 25 years, with the long-term survival rate in most large children's centers currently being approximately 90%. The improvement in survival has been largely attributed to advances in perioperative care, frequent use of parenteral nutrition, and better techniques of surgical repair.

Methods
Between 1965 and 1992, 84 infants with gastroschisis underwent surgical repair. The management of 52 infants after 1979 was compared with that of 32 during the previous 14 years. Associated anomalies were present in 29%. The average birth weight was 2412 g. In 31%, primary fascial closure was performed. In another 31% with moderate visceroabdominal disproportion (VAD), a silastic chimney was used initially, and complete repair was performed at a second operation. For 25% who had severe VAD, more than two operative reconstructions were necessary. Seven of 52 infants with moderate VAD underwent initial skin-flap closure and secondary repair within 12 days.

Results
Almost all complications (27%) and deaths (4%) occurred in infants with severe VAD and were largely unrelated to associated malformations or birth weight. The length of postoperative mechanical ventilation, need for parenteral nutrition, need for multiple operations, and length of hospitalization were all directly related to the severity of the VAD.

Conclusions
Complete repair of gastroschisis at the initial operation is the optimal goal; however, the severity of VAD has permitted this approach in only one third of patients in this study. Delayed repair with a silastic chimney and one or more reconstructive procedures has provided excellent long-term survival with low morbidity and mortality rates. Although skin-flap closure is no longer used initially,

this technique has been helpful for the residual defect in infants with severe VAD who have had multiple silon chimney repairs (Applied Biomaterial, Silverdale, WA).

Gastroschisis is a congenital herniation of the major portion of the intestine into the amniotic cavity through a small defect 2.5 to 3 cm in diameter immediately to the right of the umbilicus. Because the eviscerated organs are not covered by skin, amnion, or peritoneal membranes, there is prolonged exposure of the eviscerated intestine to the amniotic fluid. Congestion and edema accompanied by matting of the intestinal loops occur, presumably from partial lymphatic and venous obstruction in the mesentery. Thus, infants with gastroschisis defects are more subject to prolonged ileus than are those with omphalocele where the viscera are unexposed.

Until 25 years ago, the mortality rate for infants with gastroschisis exceeded 50%. During subsequent years, the mortality rate has steadily declined to its present level of less than 10% in most major children's centers, largely as a result of improved operative techniques and close attention to the details of pre- and postoperative management, including mechanical ventilation and parenteral nutrition. Successful surgical repair of a gastroschisis defect by primary fascial closure was first reported by Watkins in 1943.[1] In 1948, others recommended repair for omphaloceles and gastroschisis using skin flaps and delayed fascial closure to reduce the hazards from respiratory and vascular compromise caused by elevated intra-abdominal pressure.[2] In 1967, the first prosthetic repair was reported, using a Teflon sac,[3] which was modified in 1969 by others to create a silastic silo, reducing the mortality rate to 25%.[4] The mortality rate was diminished further by stretching the abdominal wall before primary closure.[5] Although many reports recommend one operative repair as being superior to another, our experience indicates that the severity of the intestinal edema and the disparity between the size of the eviscerated intestine and the abdominal cavity will determine which operative technique is best suited for an individual patient.[6] The present report was undertaken to compare the results and complications from the surgical repair of 52 infants with gastroschisis treated between 1980 and 1992, with those of 32 infants whose defects were repaired during the preceding 14 years using primarily the skin-flap technique.

MATERIALS AND METHODS

During the period from January 1980 through December 1992, 52 infants with gastroschisis abdominal wall defects underwent surgical repair at the University of California, Los Angeles (UCLA) Hospital. The average birth weight of the 24 male and 28 female babies was 2412 g. Seven were small for gestational age, and one was a very low birth weight infant (630 g). Thirty-one of the 52 mothers (60%) were 21 years of age or younger. Thirty-eight infants were diagnosed by prenatal ultrasound studies, allowing an opportunity for counseling the parents and planned delivery at a major medical center by a high-risk obstetrician. Cesarean section was not performed routinely, although 24 of the 52 patients were delivered surgically for other indications.

Each of the 33 outborn infants was transferred to UCLA Hospital on an emergency basis after initiation of intravenous hydration, insertion of a nasogastric tube, and placement of the infant in a warm environment and the eviscerated intestine into a plastic bag. The time from delivery to the initial surgical repair of the gastroschisis defect was less than 14 hours in all cases, except for a 630-g infant who was considered too unstable to undergo an operation for the first 24 hours.

The frequency of associated congenital malformations was similar to that observed in the previous review of gastroschisis from our hospital describing 32 patients (Table 1).[6] Each of the 52 infants had incomplete fixation of the bowel mesentery and intestinal malrotation; however, other associated anomalies were present in only 15 patients (29%). Five had cryptorchidism, four had partially obstructing duodenal bands, two had jejunal atresia, one had ileal atresia, and one had colonic atresia. Four infants had urinary tract anomalies, and three had congenital heart defects.

In 16 of the 52 patients (31%), visceroabdominal dis-

Table 1. MALFORMATIONS ASSOCIATED WITH GASTROSCHISIS

Malformation	1965–1979	1980–1992
Malrotation with incomplete small bowel fixation	32	52
Undescended testes	6	5
Urinary tract anomalies	3	4
Duodenal bands (obstructing)	2	4
Congenital heart defect	2	3
Jejunal or ileal atresia	1	3
Colonic atresia	0	1
Bladder exstrophy	1	0
Biliary atresia	1	0

Values are no. of patients.

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proportion (VAD) was sufficiently mild that primary fascial closure could be performed at the initial operation. In 29 infants, the defects were repaired with a silastic chimney, with gradual reduction of the chimney twice daily in the nursery. Complete skin and fascial closure was achieved with one operative reconstruction in 16 of the 29 patients who had mild to moderate VAD, with the mean duration to final fascial closure being 8.8 days. Thirteen infants with severe VAD, whose defects were initially repaired with a silastic chimney, required more than two operations, with six undergoing two or more minor closures of the silastic prosthesis in the neonatal intensive care nursery (Fig. 1). The prosthesis was removed in three patients because of infection with the skin gradually epithelializing over the remaining small area of exposed intestine. In three other patients with complete closure after removal of the prosthesis, mild wound separation occurred. This was allowed to heal by secondary intention. For infants with severe VAD, complete skin closure was achieved a mean of 14.6 days after the initial repair, and complete fascial repair was accomplished a mean of 47 days after birth.

Seven infants with moderate VAD underwent skin-flap closure of the abdominal wall after enlargement of the defect with extension of the opening in the skin and linea alba. Each of the defects was closed with a second laparotomy within 12 days, although three required a prosthetic mesh support, each of which was subsequently removed at a third operation.

A Stamm gastrostomy was performed at the initial operation in 10 of the 52 patients, although it has not been used during the past 8 years. An infant with jejunal atresia and another with colonic atresia underwent cutaneous enterostomy with mucous fistula at the initial operation with reanastomosis of the intestine within 3 months. One patient with jejunal atresia and another with ileal atresia had initial silon chimney repair, placing the atretic intestine into the abdomen without anastomosis or a cutaneous stoma. Within 2 weeks, at the time of silo removal, the atresias were easily repaired, and the abdomen was completely closed, as recommended by others.[7]

Three patients required innovative measures to overcome severe VAD. The silastic chimneys in two patients were removed after two or more reconstructions, and skin-flap closure was performed in one after temporary porcine skin grafting when it became apparent that complete reduction of the viscera and fascial closure would require a lengthy hospitalization. Fascial closure was achieved in both patients at 3.5 and 4 months of age using a Marlex prosthesis (Bard Cardio Surgery Division, Billerica, MA), which was removed approximately 3 months later. In a third patient, the silastic chimney was replaced with a silastic patch at 10 days, followed by three subsequent operations to reduce the size of the prosthesis and eventually remove it by the age of 32 days, at which time skin-flap closure was achieved. Fascial repair was performed 3 months later.

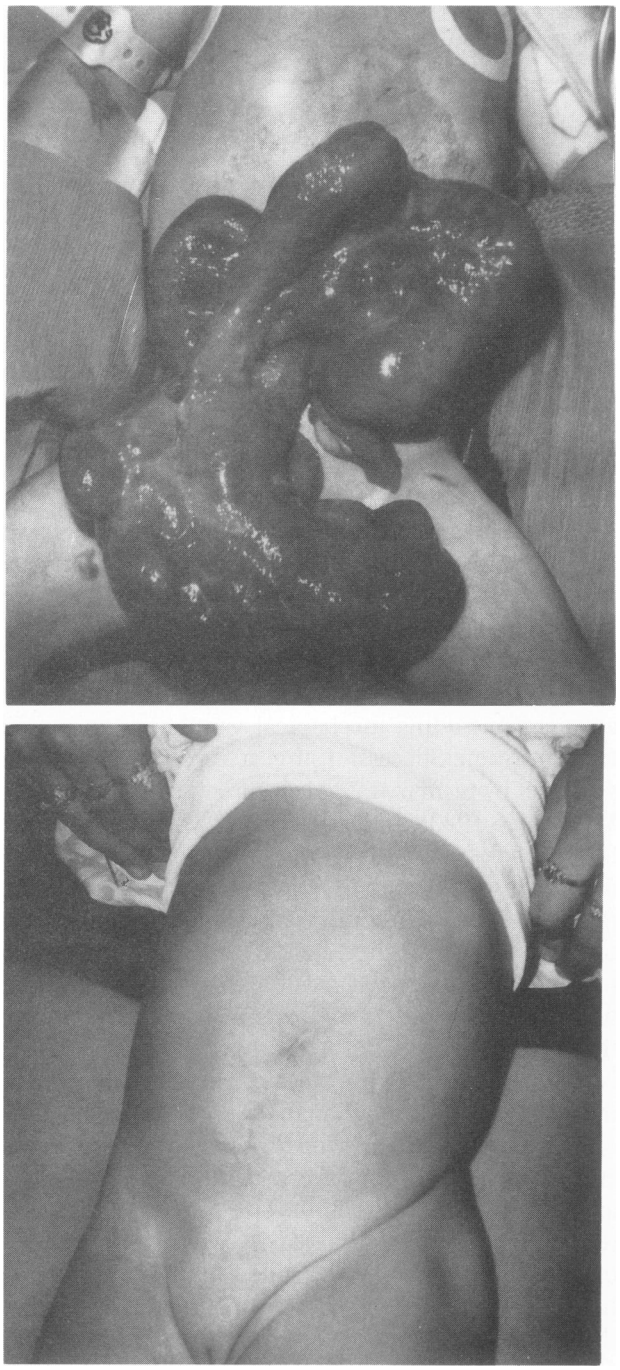

Figure 1. (A) Appearance of gastroschisis defect in 1250-g premature female infant at birth. (B) Abdominal wall of the same patient at age 4 months after an initial repair with a silon chimney and three subsequent reconstructive operations.

The period of mechanical ventilation postoperatively ranged from 4 hours to 21 days (mean, 7.3 days). When

patients experiencing complications were excluded, the average period of ventilation was 3.5 days. Thirteen of the 52 patients were extubated within 8 hours after the operation. The 16 infants who underwent complete abdominal closure at the initial operation had a mean period of mechanical ventilation of 4.8 days, whereas in the 27 infants with silon chimney closure, the mean duration of intubation was 10.4 days. The seven patients with initial skin-flap closure had a mean period of ventilation of 2.8 days.

All patients received parenteral nutrition after surgery until the ileus resolved and complete enteral nutrition was tolerated. The average duration of parenteral nutrition was 33.9 days (range, 5 to 179 days), with three infants receiving intravenous supplementation at home. Percutaneously placed fine silastic central venous catheters inserted through the basilic vein were used in 33 infants, and Broviac single-lumen catheters (Bard Access System, Salt Lake City, UT) were inserted by cut down in the saphenous veins of the 19 patients who required longer periods of parenteral nutrition.

Enteral nutrition with breast milk or formula was first successfully tolerated after a mean of 20 days (range, 5 to 67 days). The length of hospitalization averaged 39.4 days (range, 12 to 161 days). There was no significant difference in time of initial feeding for patients who had a silastic chimney compared with skin-flap repair, although patients with silastic chimney repair had longer periods of hospitalization (43.1 *vs.* 36.2 days) largely as a result of the inclusion of most infants with severe VAD in this group. Infants with initial primary closure began feeding earlier (mean, 15.6 days) and had shorter hospitalizations (mean, 27.6 days).

RESULTS

The complications were similar, although only slightly less frequent, during the recent 12-year period of study compared with those from patients repaired during the previous 14 years when skin-flap closure was used for most patients (Table 2). Fifty of the 52 patients are currently alive 2 months to 12 years after the initial repair (median, 5.2 years). Two patients died during the recent period of study (4%) compared with the 6.2% mortality rate during the previous study period. One infant with severe VAD died 73 days after birth of septic complications resulting from localized small bowel necrosis and perforation with an enterocutaneous fistula subsequent to the initial silon chimney repair. Another infant had a segment of small intestinal necrosis after silon chimney closure and was treated by cutaneous enterostomy and long-term parenteral nutrition. Cholestasis developed with progressive hepatic failure, resulting in cirrhosis with portal hypertension and massive hepatomegaly. An orthotopic liver transplant with a Roux-en-Y choledochojejunostomy was performed at 4½ months of age; however, the patient died 6 weeks later with sepsis and graft failure.

Table 2. COMPLICATIONS FOLLOWING GASTROSCHISIS REPAIR

Malformation	1965–1979	1980–1992
Pneumonia	4	6
Wound separation	3	4
Sepsis	2	4
Small bowel obstruction	2	3
Short bowel syndrome	0	3
Small bowel perforation	0	2*
Liver failure, cholestasis	1*	3*
Necrotizing enterocolitis	1	2
Seizures	1*	0
Small bowel fistula	1	1

Values are no. of patients.
* Indicates one death.

Postoperative complications occurred in 14 of the 52 infants (27.0%), 11 of whom had severe VAD, compared with 37.5% in the earlier series. Six patients experienced one or more episodes of pneumonia, four of whom also had major intestinal complications. Four patients had septic episodes, two from bowel compression with resultant intestinal necrosis and perforation, one from necrotizing enterocolitis, and one from a central venous catheter used for long-term parenteral nutrition. Four patients had superficial wound separation after complete abdominal closure, three of whom had silon chimney repair. Mechanical intestinal obstruction required lysis of adhesions in three patients. Necrotizing enterocolitis occurred in two infants, neither of whom required intestinal resection. In the two infants with necrotic bowels and one infant with jejunal atresia, short bowel syndrome developed, requiring long-term parenteral nutrition. Two patients had parenteral nutrition-induced cholestasis, one of whom required cholecystectomy and the other, a liver transplant. Two patients had persistently symptomatic gastroesophageal reflux and required fundoplication.

There was one remarkable survival of a 630-g female infant with severe VAD born after 24 weeks of gestation. She initially appeared so unstable that repair with a silastic chimney was undertaken only after 24 hours of resuscitation. She subsequently had transient necrotizing enterocolitis, parenteral nutrition-induced cholestasis, and a grade III intraventricular hemorrhage. After 147 days of hospitalization, she was discharged on complete enteral feedings and weighed 3150 g.

Each of the surviving 50 patients is progressing well

with near-normal growth and development. Two of the three children discharged on home parenteral nutrition remained dependent on intravenous support for almost 10 months. Another patient who underwent jejunal resection gradually had near-normal oral feeding patterns, discontinuing intravenous support after 7 months; the 75th weight percentile was achieved by 19 months of age.

Most of the infants who had severe VAD experienced a period of ileus lasting as long as 5 weeks before they could take appreciable volumes of intestinal feedings. The severity of the ileus correlated closely with the degree of intestinal edema and the severity of VAD. Ileus was minimal in patients who underwent primary complete, or skin-flap closure with minimal tension. Emesis in the postoperative period rarely occurred, and no patient experienced aspiration, largely because of the effectiveness of oral gastric tube or gastrostomy decompression. There were no complications related to the gastrostomy, which was removed an average of 7 weeks after surgery in the eight patients in whom it was used. Gastrostomy tubes were removed much earlier than in the first study period.

Thirty of the 32 patients from the early study period are currently alive after 13 to 27 years (mean, 17.4 years) with no long-term complications or limitations from the abdominal wall repair.

DISCUSSION

The present series of patients combined with the preceding review of gastroschisis from the UCLA Hospital provide a breadth of experience from which to make observations during a time of evolving management patterns. As noted three decades ago, the major factors influencing survival after gastroschisis repair continue to be prematurity, the degree of VAD, the presence of additional malformations, and the promptness of operative closure.[8] The majority of infants can be repaired by one or two operations with low risk and an excellent long-term outcome.

The severity of VAD has clearly become the most important factor determining the morbidity and mortality rates, as noted in the present study and by other authors during the past few years; it deserves special attention.[9,10] Attempts to close the abdomen more rapidly may cause pressure necrosis of the intestine, as observed in previous reports[6] and which occurred in one of our patients. However, delay in complete fascial closure for more than 4 months permits the rectus muscles to contract into narrow bands, which subsequently become more difficult to approximate in the midline and cause a permanent narrow waist with an hourglass configuration. More than 25% of infants with gastroschisis have severe VAD, and for these patients, initial repair with a silastic chimney and one or more subsequent staged reconstructive operations during the ensuing few weeks appear to provide the safest method of complete closure. During the past few years, we have increasingly employed a gradual reduction in the size of the abdominal wall prosthesis in the neonatal intensive care nursery using intravenous analgesics and muscle relaxants. For those infants who have been allowed to epithelialize the remaining skin defect over the intestine, it has been remarkably easy to close the muscles and skin surgically 6 to 8 weeks later.

Although skin-flap closure was employed with a good outcome in the majority of infants in the early report from our hospital, there has been an evolution during the past decade toward the use of the silastic chimney technique for almost all infants in whom primary closure was not feasible.[11] Skin-flap closure continues to play a role in managing occasional infants in whom the prosthesis cannot be completely removed after 2 to 3 weeks. Placement of a Marlex prosthesis between the rectus muscles and beneath the skin flaps may permit earlier discharge than might otherwise be possible in selected patients with severe VAD.

There has been considerable improvement in the survival of both low birth weight infants and those with associated anomalies. Complications from concurrent jejunal or ileal atresia have been reduced substantially by delaying anastomosis for 1 to 3 weeks (when intestinal edema has subsided), as in two of our patients. Almost all complications in the present series of patients occurred in infants with severe VAD and were not related to associated malformations or birth weight. The survival of the 630-g infant in the present study was exceptional because she was one of the smallest surviving babies with gastroschisis reported in the literature.

During the past 3 years, we have used continuous epidural analgesia during the postoperative period for most infants with moderate to severe VAD for as long as 7 days. Both the period of mechanical ventilation and respirator support and the time for abdominal wall closure appear to have been reduced as a result of using epidural analgesia. Furthermore, the requirement for pancuronium, fentanyl, and intravenous or muscular analgesic medications appears to have been reduced, with a resultant decrease in both peripheral and pulmonary edema, compared with infants not receiving epidural analgesics, although a randomized trial, or experience with a larger number of patients will be necessary to validate the benefits of epidural anesthesia statistically. We have recorded no complications from this technique.

Despite the continued improvement in surgical techniques for gastroschisis repair, successful management depends as much on the perioperative care as on the surgical repair itself. It is well known that exposure of the fetal intestine to amniotic fluid, combined with compres-

sion of the lymphatic vessels and venous drainage from the gut, produces edema with thickening of all layers of the bowel, resulting in prolonged ileus and delayed absorption of substrates from the lumen.[12] Experimentally produced gastroschisis is associated with a decrease in fetal weight, presumably caused in part by failure of *in utero* intestinal absorption of substrates from the amniotic fluid. This situation may account for the large number of small for gestational age infants with gastroschisis (7 of 52 in the present report).

There has been a progressive trend toward increased use of central venous parenteral nutrition on a routine basis and administering home parenteral nutrition for those with severe VAD or short bowel syndrome to shorten the hospital stay. Because parenteral nutrition increases the risk of catheter sepsis and may occasionally cause cholestasis, particularly in the young infant with an immature liver, enteral nutrition should be initiated as early as considered feasible. The development of cholestatic liver syndrome appears to be variable among neonates and may occasionally lead to rapidly progressive cirrhosis with hepatomegaly and portal hypertension, as in one of our patients. Thus, total parenteral nutrition, although a very helpful adjunct, must be monitored closely because it has the potential of producing serious and even fatal complications.

Although gastrostomy was used routinely in our early clinical experience for infants requiring skin-flap closure, we have rarely used gastrostomy in patients during the past 10 years because the silon chimney technique has been used more frequently. Nonetheless, gastrostomy continues to be helpful in patients with short bowel syndrome or other situations requiring long-term parenteral nutrition.

Although the mortality rate in the present 12-year clinical experience was only 4%, five patients (10%), all with severe VAD, had serious complications requiring bowel resection or prolonged periods of parenteral nutrition. There was surprisingly little difference in the complications between the early and recent groups of patients, despite the newer surgical and perioperative techniques. The mortality rate was 6.2% in the early series and 4% in the recent group. The morbidity rate was 37.5% in the early series and 27% in the present group.

Optimum surgical management of gastroschisis depends on the severity of VAD, which may be difficult to determine by prenatal ultrasound studies. Although controversy exists regarding the decision for Cesarean section to deliver infants with gastroschisis, we observed no complications from vaginal delivery in any of our patients.

Complete repair of gastroschisis at the initial operation is the optimal goal; however, the severity of VAD allowed this approach in only 31% of our patients. Delayed repair with a silastic chimney and one or more reconstructive procedures provided excellent long-term survival with low morbidity and mortality rates. Although primary skin-flap closure with wide opening of the abdominal cavity, as used in our early series, requires a longer time for complete abdominal wall closure than does closure with a silon chimney, the morbidity and mortality rates and the length of hospital stay were remarkably similar. The disadvantages of initial skin-flap closure for infants with moderate VAD include the need for more complex late secondary reconstructions, the large abdominal wall scar, and the tendency toward a permanent wasp-waist body configuration. Although initial skin flap repair is no longer used, this technique was helpful for the residual defect in infants with severe VAD who had two or more previous silon chimney repairs, as in three of our recent patients. This approach may allow earlier cessation of ventilator support and hospital discharge.

References

1. Watkins DE. Gastroschisis. Va Med Month 1943; 78:42—43.
2. Gross RE. A new method of surgical treatment of omphaloceles. Surgery 1948; 24:277–281.
3. Schuster SR. A new method for the staged repair of large omphaloceles. Surg Gynecol Obstet 1967; 125:837–839.
4. Allen RG, Wrenn EL. Silon as a sac in the treatment of omphalocele and gastroschisis. J Pediatr Surg 1969, 4:3–8.
5. Raffensperger JC, Jona JC. Gastroschisis. Surg Gynecol Obstet 1974; 138:230–234.
6. Fonkalsrud EW. Selective repair of gastroschisis based on degree of visceroabdominal disproportion. Ann Surg 1980; 1991:139–144.
7. Shah R, Woolley MM. Gastroschisis and intestinal atresia. J Pediatr Surg 1991; 26:788–790.
8. Moore TC. Gastroschisis with antenatal evisceration of intestines and urinary bladder. Ann Surg 1963; 158:263–268.
9. Harrison MW, Campbell TJ, Campbell JR. Selective management of gastroschisis. Ann Surg 1986; 203:214–218.
10. Caniano DA, Brokaw B, Ginn-Pease ME. An individualized approach to the management of gastroschisis. J Pediatr Surg 1990; 25:297–300.
11. Thompson J, Fonkalsrud EW. Reappraisal of skin flap closure for neonatal gastroschisis. Arch Surg 1976; 111:684–689.
12. Phillips JD, Kelly RE Jr, Fonkalsrud EW, et al. An improved model of experimental gastroschisis in fetal rabbits. J Pediatr Surg 1991; 26:784–787.