Gastroschisis With Antenatal Evisceration of Intestines and Urinary Bladder *

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GASTROSCHISIS is a rare type of congenital evisceration. The evisceration occurs through a congenital, full-thickness defect in the anterior abdominal wall and is present at birth. The malformation is of the abdominal wall rather than of the umbilical cord. The umbilical cord is normal in size and insertion into the abdominal wall. The margins of the defect are rounded and smooth and there is no covering sac or its ruptured remnants. The eviscerated organs, generally small and large intestine, show varying degrees of serosal granularity and thickening due to antenatal and perinatal immersion in amniotic fluid. The term gastroschisis is derived from the Greek word *gastros* meaning belly and the Greek and Latin words *schisma* denoting separation or division. The defect generally is regarded as being due to some abnormality in the development of the somites which are responsible for the integrity of the anterior abdominal wall. The embryologic development of this area is discussed in detail by Wyburn.[33]

In 1953, the author and Stokes [23] presented a classification of congenital eviscerations through anterior abdominal wall defects and stressed the criteria by which the various types of evisceration could be identified. It was emphasized that both omphalocele and intussusception through a persistent omphalomesenteric duct were umbilical cord anomalies whereas gastroschisis was a malformation of the abdominal wall.

With the passage of time and the reporting of a number of additional cases, it now seems appropriate to *revisit* gastroschisis with a review of current published experience and the report of a new case managed initially by a combination of eviscerated mass decortication and a ultilization of the Gross principle of creating a large ventral hernia to accommodate the decorticated viscera followed by subsequent repair of the ventral hernia. Both the small and large intestine and the entire urinary bladder were involved in the evisceration.

Case Report

The patient, a newborn white female infant, was born at the Ball Memorial Hospital on January 31, 1961 and was seen by the author in the delivery room approximately five minutes after birth. There was a massive congenital evisceration of intestine and urinary bladder through an anterior abdominal wall defect. Considerable difficulty had been experienced by the obstetrician in resuscitating the infant. She was taken promptly to the operating room in a heated, oxygen-containing incubator.

She was the first child born to young parents. A maternal great uncle had expired shortly after birth from bleeding at the area of the umbilicus. The family history otherwise was negative for congenital malformations. The pregnancy had been normal. Her birth weight was not obtained due to the urgency for immediate surgical care. She weighed 5 pounds and 6 ounces at 5 days.

Anesthesia was started approximately 20 minutes after birth. General endotracheal anesthesia with a mixture of cyclopropane, nitrous oxide and oxygen was employed. A continuous slow infusion

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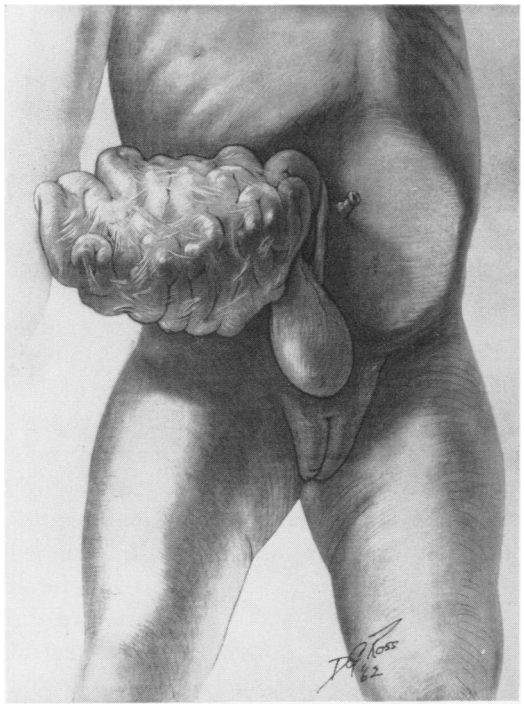

FIG. 1. Appearance of *peal*-covered eviscerated masses of intestine and urinary bladder through extra-umbilical abdominal wall defect at birth. Defect is to right of normal appearing umbilical cord and there is no ruptured sac or its remnants.

of anectine was given for abdominal muscle relaxation through an ankle vein polyethylene tube. The eviscerated masses and the abdominal wall were carefully cleansed with zephiran and sterile drapes were applied.

At this point, careful inspection of the eviscerated masses and the defect established the malformation as a gastroschisis. The evisceration was through an oval defect in the anterior wall to the right of and partially below a normal appearing umbilical cord (Fig. 1). The defect measured approximately 5.0 cm. in its greatest diameter. The margins of the defect were smooth and there was no evidence of a ruptured sac or membrane. There were two separate eviscerated masses. One was an elongated, rigid mass which consisted of the entire large and small intestine from the middle portion of the duodenum to the distal sigmoid colon. The entire discolored mass was of leathery consistency and was imbedded in a relatively thick gelatinous matrix or *peal*. The second prolapsed mass was the entire urinary bladder. It was covered by the same type of thick gelatinous peal. The infant and the eviscerated masses were given a brownish-yellow color by meconium stained amniotic fluid. A portion of the stomach and the uterus and adnexa also had prolapsed through the defect but were not covered by the peal and had a normal appearance. The peritoneal cavity was greatly reduced in size.

The defect was enlarged by incision in the mid-line to the limits of the xiphoid and the pubis and the umbilical cord was excised. It was also readily apparent that even the creation of widely dissected skin flaps as in the Gross operation for omphalocele would not suffice to create a satisfactory enclosure for the firm, elongated, thickened intestinal mass. It was imperative that the mass be both reduced in size and made more pliable to fit into the available space. Complete decortication of both the intestinal and urinary bladder masses accordingly was undertaken. Decortication of the intestinal mass was difficult and tedious. Small punctuate hemorrhagic areas appeared in varying degrees over the bowel serosa after decortication. There was considerable edema in the mesentery and over the intestine under the peal. This edema had helped to exaggerate the size of the prolapsed intestinal mass. Most of the edema was removed during decortication and later by compression. Following decortication, the large and small intestines were essentially normal in appearance (except for the punctate serosal hemorrhages) and normal in pliability. There was nonrotation of the intestines. The appendix was removed. Decortication of the thick peal from the urinary bladder was carried out and, following decortication, the urinary bladder also was found to be normal in appearance (except for the prolapse).

Large skin flaps were freed up by dissection around to the infant's back. A large ventral hernia was created by approximating the skin flaps with interrupted, mattress sutures of 3-0 silk over the decorticated intestines and urinary bladder. The closure was rather snug but the flaps appeared to be viable. Twelve milligrams of anectine were used during the operation.

The postoperative course was quite uneventful as compared with the birth and subsequent operation. Physical examination revealed no other congenital malformations. Blood counts and urine examination were within normal limits. Nasogastric suction and parenteral maintenance were utilized. A meconium stool was passed on the day following operation. Good bowel sounds were heard three days following operation and oral feedings were started one day later. The skin flaps appeared healthy when the sutures were removed 16 days after operation. The infant was released from the hospital one day later weighing 5 pounds and 12 ounces.

She did well during the months following her release from the hospital. Growth and development were normal. She began to sit alone at 5½ months of age and to crawl at 6½ months of age. A compression binder with a bulky mass in front was utilized to reduce the size of the hernia and stimulate growth of the peritoneal cavity. Frequent efforts also were made by the mother to reduce the herniation manually and to maintain the reduction within the rectus muscles for limited periods of time.

At seven months of age the peritoneal cavity appeared of sufficient size to accommodate the viscera in the ventral hernia (Fig. 2). She was admitted to the hospital on August 20. The findings on physical examination were within normal limits except for the presence of the large ventral hernia. The blood counts and urine were normal. On August 21, the hernia sac was opened and the intestines and the urinary bladder were dissected free of the skin flaps. The herniations were reduced and the rectus muscle was approximated in the mid-line from the xiphoid to the pubis with interrupted sutures of 2-0 silk. Redundant portions of the skin flaps were excised and the remaining portions were approximated in the mid-line in layers with 5-0 interrupted silk sutures. Recovery was prompt and the patient was released from the hospital nine days after operation. She had continued to grow and develop normally in the year since the repair of the ventral hernia (Fig. 3). An intravenous pyelogram at 19 months of age was normal. Her mother recently had given birth to a normal infant.

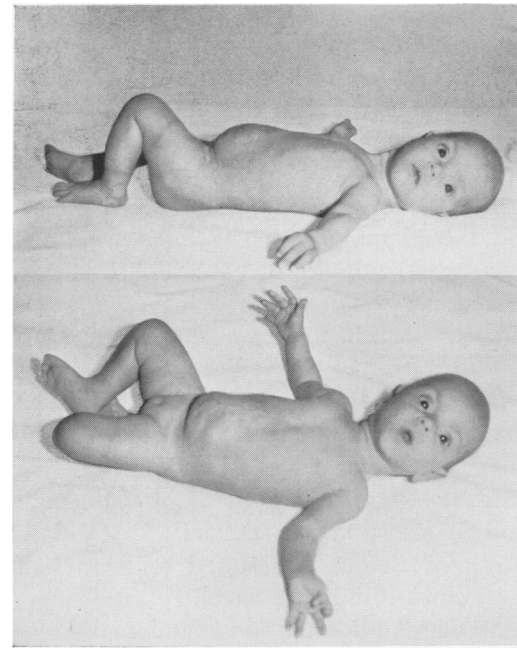

FIG. 2. The infant at 7 months, prior to repair of the ventral hernia.

Discussion

A review of the literature pertaining to gastroschisis, ruptured omphalocele and other abdominal eviscerations has yielded 31 cases which satisfy the diagnostic criteria for classification as gastroschisis. The case of Krauss,[19] which was accepted as a case of gastroschisis by the author and Stokes, has been rejected on *second look* because of insufficient descriptive detail. The extra-umbilical location of the defect, normal appearance and insertion of the umbilical cord and the absence of a sac or its ruptured remnants are the key diagnostic criteria. Such a case of gastroschisis with congenital evisceration through an extraumbilical abdominal wall defect with normal insertion of the umbilical cord was recorded in 1733 by James Calder.[7] The majority of the cases have been reported in recent years.

The sex of the infant was given in 29 of the 32 reported cases of gastroschisis, in-

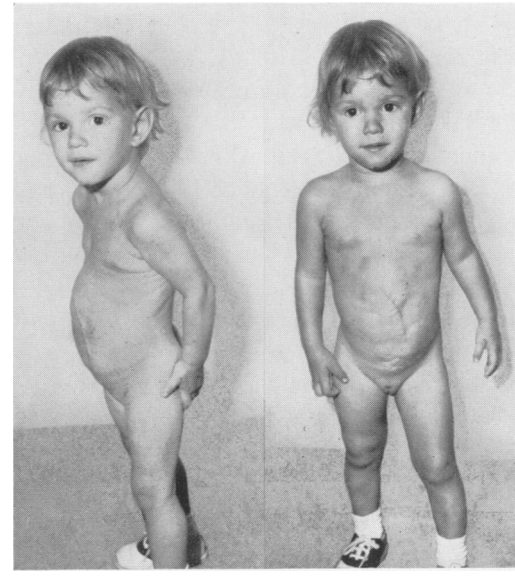

FIG. 3. Appearance of patient one year after repair of ventral hernia.

cluding the case recorded here. Nineteen of the infants were boys and 10 were girls. Information concerning the maturity of the infant at birth also was available in 29 cases. Fourteen of these infants were born prematurely and 15 were full term. The size of the abdominal wall defect has varied considerably. It has ranged from the small, strangulating defect which had produced gangrene of the eviscerated intestines at birth in the case reported by Kieswetter [17] to the xiphoid to pubis defect encountered by Brezin and Mayer.[5] The majority of the defects occurred to the right of the umbilical cord. In 25 of the cases the defect was to the right of the cord and in six it was to the left. In Calder's case, the defect was stated to be a half inch above the intact navel.

Serious additional congenital malformations were encountered in only three of the 31 cases.[3, 23] In all three, the malformation was atresia of the small intestine. Less serious malformations occasionally were found. A Meckel's diverticulum was observed in four cases.[6, 28, 32] The absence of normal intestinal rotation and fixation was commented upon frequently. Some hypoplasia of the right rectus abdominis muscle was found in two cases.[29, 30] A cleft soft palate was encountered in one case.[20] In none of the cases was there a family history of congenital evisceration.

Increasing clinical experience with gastroschisis suggests the occurrence of two major types of this malformation. In one type, the eviscerated mass of intestines is relatively rigid and is surrounded by a thick gelatinous matrix or peal. The peritoneal cavity in these cases is abnormally small. Replacement of the eviscerated bowel and primary closure of the defect is impossible. It is presumed that the evisceration must have been of relatively long standing and, for this reason, this type of case of gastroschisis is referred to as *antenatal.* In other cases the amount of mesentery and bowel serosal reaction is minimal or absent and the size of the peritoneal cavity is normal. In these cases, the eviscerated bowel may be replaced and primary closure carried out with relative ease. It is probable that the evisceration in these cases occurred relatively late in pregnancy, possibly produced by uterine contractions prior to or during active labor. This type of gastroschisis is called *perinatal.* Occasionally, cases of gastroschisis intermediate between these two types may be encountered. A case of this intermediate character was reported by Hardaway.[15] A portion of the eviscerated intestine was edmatous, dark in color and rigid. It was friable and was covered with an exudate. The remainder of the eviscerated bowel was essentially normal in appearance and texture. It was presumed that this normal appearing bowel had eviscerated at a much later date than the rigid, discolored bowel, possibly during labor.

It is of interest to note that there appears to be a relationship between the maturity of the infant at birth and the presence of antenatal or perinatal type of gastroschisis. Twenty-four infants were described in sufficient detail to permit classification into antenatal, intermediate and perinatal groups. Prematurity was encountered with considerably greater frequency in the antenatal group. Ten of the patients are classified as perinatal in type and eight of these were full term with only two being premature. The two patients in the intermediate group were split one and one. Nine of the 13 infants of the antenatal type were premature whereas only three were full term.

The eviscerated organs generally are the small and large intestine. At times, the stomach, the gallbladder and the uterus and adnexa may be seen. In the case of Diller and Travis [10] a portion of the urinary bladder was involved in the evisceration. In the case reported here, the entire urinary bladder was eviscerated and covered with a thick peal similar to that covering the eviscerated large and small intestine.

It is quite apparent that the only chance for survival in these cases lies in the prompt replacement and covering, at least by skin, of the eviscerated organs. Operation was not undertaken in two of the 32 reported cases. Both infants died.[7, 8] Thirteen infants were operated upon but died.[3, 4, 14, 17, 18, 22, 23, 25, 27, 28, 31] Sixteen infants were operated

TABLE 1. *Summary of Clinical Features and Operative Management in Successfully Treated Cases of Gastroschisis*

Author	Year Sex	Birth Wt. (lbs)	Size of Defect (cm.)	Structures Eviscerated	Type	Birth to Operation (hr.)	Operation	Remarks
Watkins[32]	1943 M	5 7/8	2.5	Large and small intestine	Perinatal	0–1	Primary closure	One month follow up. Doing well.
Burgess *et al.*[6]	1951 M	5 11/16	4	Large and small intestine	Intermediate	0–1	Modified primary closure—limited skin flap mobilization. Only skin closed.	Follow up of 15 mo. Doing well. No hernia.
Stowlowski[30]	1952 M	5 3/16	5	Large and small intestine	Perinatal	0–1	Primary closure	Follow up of 5 mo. Doing well.
Brezin and Mayer[5]	1953 M	4 3/8	15	Stomach, small intestine and cecum	Antenatal	2	Creation large ventral hernia	Follow up of 7 mo. Hernia not repaired at time of report.
Stiller *et al.*[29]	1954 F	5 1/2	3	Distal small intestine and cecum	Perinatal	5	Primary closure	Postoperative wound infection with drainage. Follow up 4 mo.
Bell and Brown[2]	1954 F	3 7/8	7	Stomach, small and large intestine	Antenatal	0–1	Creation large ventral hernia	Follow up of 15 mo. Hernia not repaired at time of report.
Hardaway[15]	1954 F	4 1/2	3	Duodenum, small and large intestine	Intermediate	2	Modified primary closure—limited skin flap mobilization. Only skin closed.	Small resulting ventral hernia decreased in size. Hernia had disappeared at 8 mo.
Diller and Travis[10]	1955 F	7 1/8	3	Most of small and large intestine. Part of stomach and urinary bladder.	Perinatal	0–1	Primary closure.	Follow up of 2 mo. Doing well.
Lurie[20]	1955 F	5	3	Two feet of small intestine.	Perinatal	2.5	Primary closure.	Follow up of 14 wk. Doing well.
Gaynor and Diefenbach[12]	1957 M	4 3/4	10	Stomach, duodenum, small and large intestine.	Perinatal	0–1	Primary closure—relaxing incision in rectus muscle.	Follow up of 4 years. Doing well.
Simpson and Caylor[28]	1958 M	6 5/16	5	Stomach, most of small and large intestine.	Antenatal	7	1. Creation large ventral hernia. 2. Closure of ventral hernia.	Hernia repaired at 13 mo. age. Fascial defect 5 cm. at time.
Cook[9]	1959 F	4 5/8	5	Stomach, gall-bladder, small intestine, right colon, uterus and adnexa.	Antenatal	0–1	1. Creation large ventral hernia. 2. Two operations to advance 2nd rectus muscle to skin flaps.	Defect not enlarged at first operation. Enlarged at second operation with rectus mobilization. Follow up 26 mo. Hernia not repaired at time of report.
Ostermann[24]	1960 M	4 3/16	4	Most of small intestine, cecum and ascending colon	Perinatal	0–1	Primary closure	Follow up of 10 mo. Doing well.
Moore	1962 F	5 3/4	5	Stomach, duodenum, small and large intestine, entire urinary bladder, portion of uterus and adnexa.	Antenatal	0–1	1. Decortication of eviscerated masses and creation of large ventral hernia. 2. Closure of ventral hernia.	Hernia repaired at 7 mo. Has done well in 12 mo. following hernia repair.

upon and survived. The clinical experience with fourteen of the 16 surviving infants is summarized in Table 1. The three successfully managed cases reported by Berman [3] were not described in sufficient detail for analysis and are not included in this table. The mortality hazard is difficult to assess from isolated case reports due to a natural tendency to report successful rather than unsuccessful cases.

The major factors influencing operative survival in cases of gastroschisis are the prematurity of the infant, the existence of additional malformations, the promptness of operation and the disparity between the sizes of the eviscerated mass and the peritoneal cavity. Ten of the infants who survived operation were full term and seven were premature. Of those infants who did not survive operation, eight were premature and only four were full term. In nine of the fourteen successfully managed cases of gastroschisis analyzed in Table 1, operation was carried out in the first hour of life. Seven of these 14 cases were of the perinatal type, two were intermediate and five were of the antenatal type. All seven of the perinatal type of cases were managed by replacement of the eviscerated organs and primary closure. In the intermediate group a modified type of primary closure with limited skin flap dissection and closure of the skin only was carried out in both cases. The small ventral hernias which resulted closed spontaneously in both cases and a second operation was not necessary.

The replacement and covering of the eviscerated mass in the antenatal type case is difficult to achieve. The author and Stokes, in 1953, suggested the use of a modification of the Gross operation for omphalocele to create a large ventral hernia by wide dissection of skin flaps. In three of the five patients of the antenatal type of gastroschisis, a large hernia had been created at birth but had not been repaired at the time of the case report.[2, 5, 9] In only the case of Simpson and Caylor [28] had the Gross principle been carried out successfully with creation and closure of a large ventral hernia. In the case reported here, it was immediately apparent that the large, rigid, elongated mass of eviscerated intestines could not possibly be enclosed by application of the Gross principle of creating a large ventral hernia. The massive urinary bladder evisceration created an additional problem. The utilization of total decortication of the eviscerated intestinal and bladder masses made possible their reduction in size and an increase in pliability at the same time. In this way it was possible to cover the decorticated viscera by wide dissection of skin flaps.

The absence of evisceration and the presence of multiple gross malformations has led to the exclusion of the case of Hinton [16] from this study. Cases of successful operative repair of ruptured omphalocele by Fear,[11] Reed,[26] Adams [1] and Maguire [21] have been reviewed and are clearly stated to be cases of umbilical cord, rather than abdominal wall, malformation.

Summary

The literature pertaining to gastroschisis and other congenital eviscerations through the anterior abdominal wall is reviewed. It is emphasized that gastroschisis is an abdominal wall, rather than an umbilical cord, malformation. Thirty-two cases which meet the criteria for classification as cases of gastroschisis have been recorded in the literature.

Approximately one-half of the infants with gastroschisis are born prematurely. Male infants predominate in a two to one ratio. The defect has occurred to the right of the umbilical cord in 80 per cent of the cases. The size of the defect has ranged from 2.5 to 15 cm. Serious additional congenital malformation has been encountered in only three of the 31 cases, including the case recorded here.

The classification of cases of gastroschisis into antenatal, intermediate and perinatal types is discussed. The antenatal type is associated with a higher incidence of prematurity, a considerably greater thickening of the covering of the eviscerated mass and a greater disproportion between the size of the eviscerated mass and the peritoneal cavity.

A case of gastroschisis of the antenatal type with evisceration of the intestines and the urinary bladder is reported. The infant was treated successfully by total decortication of the thick *peal* over the eviscerated intestinal and urinary bladder masses and the creation of a large ventral hernia. The hernia was repaired at seven months of age.

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