Management and Outcomes of Malformations Associated with Intestinal Malrotations in Newborns: About 5 Cases

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ABSTRACT
Bowel malrotation is an abnormality in the rotation of the primary intestinal loop during embryonic life. It is a severe abnormality exposing the patient to fatal complications. We report here a series of five patients presenting digestive malrotation with various malformative associations. The first three cases are multiple intestinal atresia on incomplete common mesentery. They underwent an intestinal anastomosis: end-lateral duodeno-ileal for one and end-to-end jejuno-jejunal for the other two with Ladd procedure. In the latter two cases, the omphalocele was the associated malformation; complications such as rupture of the omphalocele with intestinal perforation for one and occlusion by strangulation at the level of the neck for the other were recorded. Ileal resection removing the perforation with end-to-end ileo-ileal anastomosis was performed for the first, while reduction of strangulation at the origin of the occlusion was sufficient for the second. The LADD procedure was the complementary gesture in all cases. The outcome was favorable in 3 cases; two of the newborns who had jejunal atresia died, one on day 5 in a picture of acute respiratory distress, the other on day 6 in a picture of disseminated intravascular coagulation.

Keywords
Intestinal malrotation, Associated malformation, Management, Outcome.

Introduction
Bowel malrotation is an abnormality in the rotation of the primary intestinal loop during embryonic life. According to the literature, it occurs in 1 case per 500 live births 500 [1].

During embryonic life, the primary midgut undergoes three successive 90° counterclockwise rotations. Once this series of rotations is completed, the digestive tract and its mesos are permanently joined [2]. The interruption of this physiological process leads to an abnormality in the positioning or permanent attachment of the small intestine and the right colon. This anomaly can be isolated or associated with other malformations such as omphalocele, laparoschisis or congenital diaphragmatic hernia [2].

The management of these polymalformative syndromes is difficult in underdeveloped countries where neonatal surgery is still precarious. The aim of this study is to describe the management, and the results of a case of multiple ileal atresia, of two cases of jejunal atresia with a case of spiral bowel, of a case of ruptured omphalocele with necrosis of handle and a case of neonatal occlusion by incarceration of the caeco-appendix in an omphalocele. All these cases presenting an associated intestinal malrotation.

Case 1
This is a female newborn baby received on the 6th day of life for bilious vomiting. Onset dated back to the 2nd day of life with
bilious vomiting in a context of absence of meconium emission since birth. The gestational history showed a well-followed pregnancy, a cesarean delivery at 36 weeks with amenorrhea for acute fetal distress. The adaptation to extrauterine life was good. The physical examination was in good general condition. The abdomen was unincreased with peristaltic undulations, supple, depressible and without a palpable mass. The anus was leaky with an empty rectal bulb. The unprepared thoraco-abdominal x-ray revealed central hydro-aeric images with no distal ventilation. The diagnosis of high neonatal occlusion was made and the indication for surgery was made. Under general anesthesia with orotracheal intubation, a supraumbilical transverse laparotomy is performed. At coeliotomy, we discover multiple ileal atresia (7 atresia areas) with mesenteric defect and an incomplete common mesentery arrangement of the loops. A end-to-side duodenal-ileal anastomosis is performed bypassing the atresia areas and then arranging the slender loops on the right and the colon on the left according to the LADD procedure. The postoperative follow-up was simple with resumption of transit on the 5th postoperative day. After a 6-month follow-up, the evolution was favorable with good weight gain.

Case 2
This is a newborn male received on the 3rd day of life for bilious vomiting and stopping intestinal transit. The newborn had normally emitted meconium on the 1st day of life. The pregnancy was well followed with a delivery by cesarean section at 34 WA + 06 days for acute fetal distress. The adjustment to ectopic life was good. The weight was 1570 grams.

The physical examination revealed poor general condition. The abdomen was normal in size, without rippling, supple, depressible, and massless. The anus was leaky with an empty rectal bulb. The thoraco-abdominal x-ray in a vertical position without preparation objectified central hydro-aeric images. He was diagnosed with high neonatal occlusion and the child had been taken to the operating room on the 4th day of life. By transverse laparotomy under general anesthesia orotracheal intubation, at coeliotomy, multiple jejunal atresias were found (Figure 1) on an incomplete common mesentery. This is preceded by resection of the atretic segments, end-to-end jejuno-jejunal anastomosis and loop arrangement according to the LADD procedure. The operative consequences were simple with a resumption of transit on the 3rd day. The outcome was unfavorable with death on the 5th postoperative day in a picture of disseminated intravascular coagulation.

Case 3
Newborn male received on the 2nd day of life for bilious vomiting. The Onset dated back to 8 hours of life with bilious vomiting. The gestational history noted a poorly followed pregnancy, vaginal delivery at 35 weeks of amenorrhea for acute fetal distress. Adaptation to ectopic life was good with a birth weight of 1800 grams. There was no meconium emission from birth. The physical examination returned to good general condition. The abdomen was unincreased with peristaltic waves, supple, depressible, and without a palpable mass. The anus was leaky with an empty rectal bulb. The thoraco-abdominal x-ray in a vertical position without preparation objectified double bubble images. The diagnosis of high neonatal occlusion was made and the indication for surgery was made. Under general anesthesia with orotracheal intubation, transverse umbilical laparotomy. We discover a cystic duplication of the jejunum on jejunal and ileal spiral atresia, the distal intestine being arranged in a "pig's tail" wound around a nourishing vessel from a right colic artery with loop necrosis downstream (Syndrome Apple Peel), all on an incomplete common mesentery (Figure 2). The jejunal cyst is removed, the spiral loops are removed, the necrotic ileal loops are resected, the end-to-end duodenal-ileal anastomosis and then the loops are arranged according to the LADD procedure. The postoperative follow-up was straightforward with resumption of transit on the 3rd postoperative day. The outcome was unfavorable with death on the 6th postoperative day in a picture of disseminated intravascular coagulation.

Case 4
This is a newborn male received on the 6th day of life for umbilical swelling. The anamnesis found a poorly followed pregnancy,
vaginal delivery at 38 weeks of amenorrhea with good adaptation to extrauterine life. The physical examination revealed poor general condition. In the abdomen, there was an umbilical swelling of 2 cm at the neck covered with Wharton’s jelly broken at its top through which the slender necrotic handles emerge, including an area of sphacele giving way to meconium (Figure 3). The umbilical cord was inserted at the lower pole of the mass. The remainder of the exam does not identify any other visible external defects. The diagnosis of ruptured AITKEN omphalocele type I with necrosis and intestinal perforation was retained and the indication for surgery was given. Under general anesthesia with orotracheal intubation, transverse umbilical laparotomy. At coeliotomy, the loops herniated by the umbilical defect are reduced. The necrotic ileal loops are seen at 50 cm from the ileocecal junction over approximately 20 cm bearing the perforated area. A resection of the necrotic loops is performed followed by an arrangement of the loops in a complete common mesentery according to the LADD procedure. The evolution was simple with resumption of transit on the 3rd postoperative day. Oral feeding with breast milk is well tolerated from the 4th postoperative day. After a follow-up of 6 months, good healing of the operative wound and good somatic growth is noted.

**Figure 3:** Omphalocele type I ruptured with loop necrosis in case 4.

**Case 5**
Newborn female received in the first hour of life for malformation of the anterior abdominal wall. The gestational history noted a well-followed pregnancy and vaginal delivery at 35 WA + 2 days with good adaptation to extrauterine life and a birth weight of 2900 grams. The child had emitted meconium on the 2nd day of life. Clinical examination noted a translucent mass measuring 7 cm by 6 cm in diameter containing loops with the umbilical cord inserted at the top. AITKEN type I omphalocele was diagnosed and modified Grob tanning was undertaken. The course was marked by greenish then fecal vomiting on the 3rd day of life. The diagnosis of neonatal omphalocele occlusion was retained and the indication for surgery was taken. Under general anesthesia with orotracheal intubation, after transverse umbilical laparotomy, a tight neck of 4 cm is observed with a sac containing the ascending colon and the caeco-appendix strangulated at the level of the neck (Figure 4). After reduction, there is an incomplete common mesentery arrangement of the intestines (Figure 5). An appendectomy is performed and then the slender loops are placed on the right and the colon on the left according to the LADD procedure, followed by an omphalocele treatment. The postoperative follow-up was simple with resumption of transit on the 2nd day and a well-tolerated diet from the 4th day. The clinical examination is unremarkable after a follow-up of 6 months.

**Figure 4:** Ascending colon and caeco-appendage in the neck of omphalocele type I in the newborn of case 5.

**Figure 5:** Common mesentery in newborn in case 5.

**Discussion**
The true prevalence of bowel rotation abnormalities is difficult to estimate. Indeed, there is not a well-defined entity but a
malformative continuum of the attachment of the primary intestinal loop that varies from the isolated common mesentery to the anomaly of caecal fixation [2,3]. It is the most common congenital malformation of the small intestine with a sex ratio of 2 in the neonatal period [3]. In the present series, the sex ratio was 1.5.

Between the 5th and 10th week the primary midgut returns to the abdominal cavity and at the same time performs a series of three successive 90° rotations counterclockwise around the axis of the superior mesenteric artery. Once this rotation is completed, the digestive tract and its meso become permanently attached [2]. The interruption of this physiological process leads to an abnormality in the positioning or permanent attachment of the small intestine and the right colon. It is associated with an adhesive peritoneal band called the LADD flange [4]. These intermediate situations are the cause of hail volvulus, a vital emergency [5]. In the cases described we did not object to volvulus but complications of other associated malformations such as jejuno-ileal atresias and omphaloceles.

Atresia result from a defect in vacuolation of the intestinal lumen or from a primary vascular disorder or ante-natal intussusception, while omphalocele results from a lack of reintegration of the primary loops into the abdominal cavity. All these abnormalities that may result from a rotational defect [6] add to the morbidity of this pathology in our context.

In African countries and developing countries like ours, neonatal surgery is still quite lethal because of insufficient resuscitation and anesthesia in this age group [7]. What then does the care of these newborns consist of?

In the first case of our series, it was a neonatal obstruction high type multiple intestinal atresia with provision of the loops in incomplete common mesentery received with a delay on the 6th day of life. Note that these admission delays are frequent in Africa [7]. In this case, we had 7 atresic zones taking all the hail, which made end-to-end anastomoses impossible, hence the decision to have a terminal latero-lateral duodeno-ileal anastomosis bypassing the atresic zones. Despite this delay in admission and the severity of her malformation, the immediate course was simple but distant from delayed weight gain. This could be explained by short bowel syndrome because of the small portion of the remaining small bowel and the lack of parenteral nutrition.

Case 2 of management similar to the first had an unfavorable outcome. This death could be explained by the heavy mortality of neonatal surgery, especially in premature babies. The low birth weight, multiple intestinal atresia, the lack of nutrition and the lack of neonatal resuscitation were lethal for this newborn despite the well-conducted surgery.

In the literature, Apple-Peel syndrome continues to have a severe prognosis with significant morbidity and mortality [8]. This observation was the same in this series where, despite the successful surgical management with resumption of transit, death occurred in a table of disseminated coagulopathy. The association of Apple Peel Syndrome and digestive duplication is exceptional [9]. According to some authors, these types of malformations were mainly the result of a morphogenetic abnormality occurring early in intrauterine life, rather from failure of recanalization or interference with the blood supply in fetal life [10]. End-to-end jejuno-jejunal anastomosis has been the same in several authors in Apple Peel syndrome [11-13].

In the latter two cases, omphalocele was the malformation associated with digestive malrotation. This clinical description has been reported by other authors [2,5]. For one, loop rupture and necrosis required immediate surgical management. In the second case, conservative management had started and it was only the secondary occlusion that required surgery. In these two cases of digestive malrotation on omphalocele, the postoperative evolution was favorable. This could be explained by the good birth weight, the possibility of reintegration of the loops in 1 step, the reduction of the hernia without major resection of the loop. This made it possible to avoid undernutrition postoperatively despite the absence of parenteral nutrition.

Treatment of an abnormal bowel loop rotation is surgical. This intervention, described by LADD, consisting of placing all of the small bowel on the right and the colon on the left, which has become the standard goal in the management of bowel rotation defects, was performed in all of our patients [14]. It allowed a resumption of digestive transit in all our cases.

The course of intestinal malrotations is generally favorable if the diagnosis is early and the management is adequate [15]. The malformative association, especially jejuno ileal atresia, as well as the precariousness of the technical platform aggravates the lethality of this pathology, as evidenced by cases 2 and 3 of our experience.

Conclusion
Bowel rotation abnormalities are responsible for serious complications that are life-threatening if combined with other defects. Knowledge of these rare and potentially serious pathologies is essential for good management. Lethality still remains high (40%) in our work environment where pediatric resuscitation and anesthesia are still underdeveloped.

References


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