

Jejunoleal Atresia: Factors Affecting the Outcome and Long-term Sequelae

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ABSTRACT

Context: Jejunoileal atresia (JIA) is a common abnormality. The outcome is conditioned by several variables. Nutritional problems, and long-term sequelae are described among those who survive. **Aim:** To correlate the type of JIA and its management to the outcome and long-term quality of life. **Settings and Design:** Forty-three cases over a 17-year period (1992–2009). Perinatal data, management, and outcome were extracted from the clinical notes. The cases that had survived were contacted to get information about their present condition. **Materials and Methods:** Morbidity and mortality were matched to maturity, birth weight, mode of diagnosis, type of JIA, associated anomalies, and management and duration of parenteral nutrition. Growth and quality of life in 34 cases were evaluated via a telephone interview at a minimum of one year from surgery. **Statistical analysis:** Fisher test, Linear regression test, Kruskal-Wallis test, Dunn's comparison test. **Results:** Male/Female ratio was 25/18 and median birth weight was 2.644 g. Prenatal diagnosis was recorded in 34%. Six patients (14%) had associated anomalies. Primary surgery was resection and anastomosis in 88% and temporary stoma in 12%. Length of the resected bowel ranged from 3 to 65 cm. Whenever multiple atresia was found, the bowel length was saved by multiple anastomosis. Three dehiscences and three adhesive obstructions required a reoperation. Two patients (4%) died due to a central catheter-related sepsis. Prenatal diagnosis did not influence the outcome and was associated with a higher rate of Cesarean deliveries. Interview, at a median of nine years, showed normal growth in 85%. One case of short bowel syndrome is still on parenteral support at 22 months. **Conclusions:** Preserving bowel length and reducing the recourse to stoma is the key to good outcome and growth. Sequelae are correlated with the type of atresia and length of residual bowel; however, they decrease their severity with time.

Key words:

Intestinal atresia, prenatal diagnosis, surgery

INTRODUCTION

During the last decades, advances in intensive care, surgical techniques, and artificial nutrition have increased the survival of most weak neonates affected by congenital Jejunoileal Atresia (JIA). Surgical choices aim to spare the longest amount of bowel, even through multiple anastomoses. Prenatal imaging allows for early diagnosis and avoids a delay in treatment.^[1] Despite that, protracted bowel dysfunction and the reduction of the absorbing enteral surface, secondary to extensive resection, still call for prolonged medical attention and nutritional support. Quality of life and growth rate of many of these children are seriously affected even many years after surgery.^[2] The aim of the present study is to examine a consecutive series of patients affected by JIA and to identify the main factors affecting survival and long-term quality of life.

MATERIALS AND METHODS

Records of 43 neonates admitted to our unit, between January 1992 and December 2009, for a congenital JIA have been revised. Thirteen cases with an additional digestive tract obstruction (esophageal, duodenal, colonic

or anorectal) were excluded from the present study. Patient demographics, associated anomalies, prenatal diagnosis when recorded, and site and type of obstruction, were extracted. Information on surgical procedures (length and type of resected bowel, temporary stoma, primary resection, type, and number of anastomosis) and postoperative nutritional support (enteral, peripheral or central parenteral), delay of canalization and oral feeding, and length of follow-up, were also collected. Additional information on those who survived was achieved by telephone interviews. Data on growth rate, nutritional habits, and number of bowel movements were collected

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at a minimum of one year from surgery. Mortality and morbidity (in terms of surgical complications and duration of artificial nutrition) were correlated to the patients' demography, associated malformations, site and type of JIA, type of surgery, and the amount and type of resected bowel. The impact of prenatal diagnosis on perinatal management and outcome was also evaluated. Long-term outcome and sequelae, even in terms of growth rate and gastrointestinal function, were considered and correlated to:

- Time from primary surgery;
- Type and site of obstruction;
- Length in centimeters and type of residual bowel.

Data were elaborated by GraphPad InStat software, version 3.10 [Fisher test, Linear regression test, Kruskal-Wallis test (non-parametric ANOVA), Dunn's comparison test]. $P < 0.05$ was considered significant.

RESULTS

The male/female ratio was 25/18. Polyhydramnios was recorded on obstetrical records in 23 cases. Dilated fetal bowel loops, suggestive of JIA, were described by fetal ultrasound examination (US) in 15/43. Mean gestational age (GA) was 36 weeks (range 27–41, SD 3.143) and mean birth weight (BW) was 2.644 g (range 730–4.120, SD 785). Table 1 shows in detail: The perinatal data, time of referral, and mortality among cases with and without prenatal diagnosis (PND). A statistically insignificant, higher Cesarean section rate, and a lower mean BW and GA were recorded among patients with a PND. This occurred more frequently among cases not delivered in other centers (Outborn), without co-located surgical facilities, where planning of postnatal transfer was required. The incidence of associated congenital anomalies was generally low (14%). There were four minor cardiac defects (two ventricular septal defects, one atrial septal defect, and one tricuspid insufficiency) and two cases of cystic fibrosis (CF). Of the 43 atresias, 33 were located in the jejunum, six in the ileum, and four in both the jejunum and the ileum. The type of obstruction, according to Grosfeld's^[3] classification, is illustrated in Figure 1. All the patients were operated after stabilization and rehydration, within three days of admission. The number of multiple atresias varied from two to ten. Tapering of the proximal dilated loop, after primary resection, was done in 29/38 cases to facilitate end-to-end anastomosis. This was performed in two layers

using 5-0 Vicryl sutures. A temporary enteral stoma, after resection of the atresic tract, was required in only five patients. In two of them it was justified by perforation of the dilated bowel and severe peritoneal contamination, which contraindicated primary anastomosis. Another three cases presented an extremely dilated, atonic proximal loop not suitable for an end-to-end anastomosis that was too long to be resected or trimmed. The stoma was closed when upper tract dilatation was reasonably reduced and bowel continuity was restored after a period ranging from one to four months. The total length of the resected bowel ranged from 3–65 cm. Whenever multiple obstructions were found, up to three anastomoses were confectioned, in order to save the bowel length. Trans-anastomotic tubes were never used. Immediately after bowel surgery, total parenteral nutrition (TPN) was required, until the bowel function returned (bowel sounds were detected and stool was produced). Oral feeding could be started at a mean interval of eight days from surgery (range 2–22 days) in all cases that had resection and primary anastomosis. Depending on the severity of the short bowel syndrome, full enteral/oral nutrition could be achieved in a matter of weeks or months, but was sometimes never completely achieved. Patients were given as much enteral/oral nutrition as possible, to facilitate bowel growth and increased absorption of nutrients, and to decrease the deleterious effects of total parenteral nutrition (TPN) on the liver. In our series, a central venous line (CVC) was inserted in 35 cases (81%) and TPN established in 34. TPN was continued for a mean of 22 days (range 3 to 150 days). One case, with a post-resection short bowel syndrome (50 cm), is still on partial parenteral supplementation, at 22 months of age. Dependency on TPN was significantly correlated to the length of the residual bowel [Figure 2]. All other patients could be kept on temporary parenteral support by a peripheral venous line, until the complete oral feeding regime could be re-established. Postoperative complications occurred in nine of the forty-three (20%) patients treated. Three adhesive bowel obstructions and three anastomotic leakages required successful re-operation. Sepsis secondary to central venous catheter (CVC) occurred in three patients and was lethal in two. Table 2 summarizes the associated anomalies, type of surgery, complications, clinical course, and mortality in our series. A higher mortality rate ($P < 0.001$) was significantly associated to low BW and GA and was also

Table 1: Impact of prenatal diagnosis on perinatal data and outcome

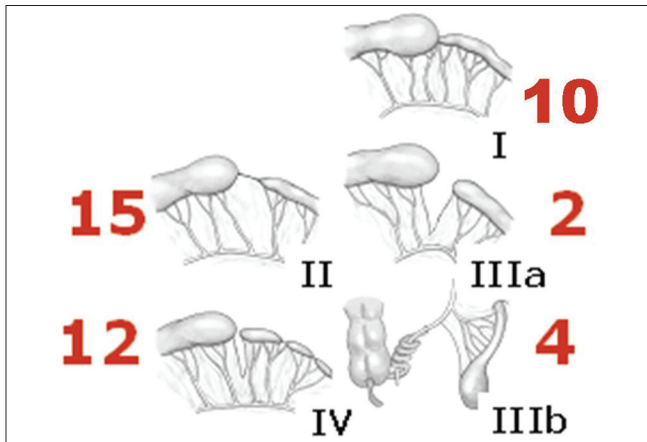
	Mean BW	Mean GA	% Cesarean sections	Time before Referral (hrs)	Mortality
With PND (15)*	2442 (SD 383)	35 (SD 1648)	73	5 (SD 8,912)	0/15
Without DPN (28)**	2763 (SD 917)	36,5 (SD 3615)	46	64,25 (SD 78,572)	2/28
P (Fisher's Test)	0.09 ns	0.05 ns	0.08 ns	0.005	0.4 ns

*Inborn 8 – Outborn 7; **Inborn 3 – Outborn 25

Table 2: Clinical features and course of 43 Jejunoleal obstructions

Type	No	Assoc. Anomalies	Type of Surgery	Tapering (cases)	Complications	Stools passage (mean days)	TPN (mean days)	Mortality
I	10	3	8 Resection & PA 2 Resection & Stoma	–	1 Adhesive Obstr. 1 Anast.leakage 1 Sepsis	10,5	17 (3-29)	–
II	15	2	15 Resection & PA	10	1 Anast.leakage	5	15 (6-90)	–
IIIa	2		2 Resection & PA	2	–	7,5	10 (7-14)	–
IIIb	4		3 Resection & PA 1 Resection & Stoma	3	1 Adhesive Obstr. 1 Sepsis	8	29 (11-40)	1
IV	12	1	10 Resection & PA 2 Resection & Stoma	12	1 Anast.leakage 1 Adhesive Obstr. 1 Sepsis	12,5	36 (7-150)	1
					20%	8 (2-22)	22 (3-150)	4,6%

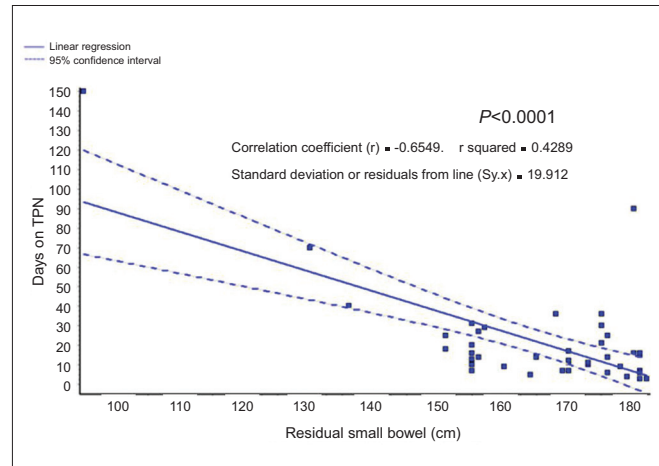
PA – Primary anastomosis

**Figure 1:** Type of obstruction among 43 cases according to Grosfeld's classification

correlated to long-term dependence on TPN and related risk of CVC infection. Among the 41 who survived, only 34 could be contacted at a median time of nine years from surgery (range 1 to 18 years). Three of them (8%) complained of food intolerance and 29 (85%) reported normal bowel function, whereas, two (6%) presented with constipation, and three (9%) had recurrent diarrhea; among these, one still has a short bowel syndrome and needs home IV therapy, nutritional support, at 22 months from surgery. At the interview, our patients' weight growth was above the fiftieth percentile in 19 (55%), above the tenth and below the fiftieth percentile in 11 (32%), and below the tenth percentile in four (13%). These results were significantly correlated to the age at control ($P<0.001$), the length of residual small bowel ($P<0.001$), and the type of atresia ($P<0.01$).

DISCUSSION

Jejunoleal atresia is a common cause of neonatal intestinal obstruction, secondary to intrauterine mesenteric vascular accidents.^[3] Advances in surgical techniques, improved perinatal and postoperative management, and the

**Figure 2:** Residual small bowel length and TPN duration

availability of artificial nutrition have reduced the overall mortality rate to 11–16% in the last few decades.^[2] Among a relevant series of 128 cases of JIA observed along 25 years and published in 1998,^[4] the overall mortality was 16%, which was determined in most of the cases by late complications from supportive measures required after extensive bowel resection performed in multiple JIA or apple peel Atresia. These figures were even more reduced (4%) in our series of 43 patients. This could be due to the recent improvement in nutritional support techniques and the shorter time of observation (17 years), which makes the standard of treatment more homogeneous. Bowel-saving strategies influence the duration of TPN support, which is inversely correlated to residual small bowel length. Prolonged artificial nutritional support exposes patients to an increased risk of complications. CVC infection was responsible for three cases of sepsis in our series; two of them died. Associated anomalies have also been reported to influence the outcome.^[5] In our series, associated anomalies affected 13% of the patients (four minor cardiac defects, two cases of CF), but were never responsible for deaths. CF has been estimated to affect 5 - 24% of the

patients with JIA.^[6] It was found in two of our cases, as the result of an active genetic investigation, as suggested by Stolman *et al.*^[2] JIA, mainly type IIb and IV, and related accidents like volvulus, complicated meconium ileus, and meconium peritonitis, may be responsible for significant intestinal loss with subsequent short bowel syndrome. This is estimated to occur in 43% of the cases.^[7] In our series, at long-term follow-up, only one patient (3%) complained of post surgical malabsorption and is still receiving home parenteral nutritional support at 22 months from surgery. Our results may be explained by:

- Saving maximum small bowel length even by multiple (maximum three) short resections and primary anastomosis, whenever several atresias are present
- Recourse (67%) to extended tailoring of proximal dilated bowel to facilitate primary end-to-end anastomosis
- Limited recourse (11%) to temporary enterostomy.

Prenatal US findings suggestive of a JIA^[8] (hypercogenic bowel, dilated fetal loops, polyhydramnios) were frequent in our series, but had no impact on the outcome. Patients with a PND had a lower median BW and GA. Possible misuse of Cesarean section in prenatally detected cases born in hospitals, without co-located surgical facilities, has been frequently reported, and the negative impact on the outcome of maturity and respiratory status has been hypothesized.^[9-12]

In conclusion, bowel loss and consequent long-term dependence on artificial nutrition are the main factors of morbidity and mortality among patients affected by JIA, and influence the long-term outcome and quality of life. A conservative approach, even in case of minimal intestinal segments, whenever multiple atresias are found, proximal dilated loop tapering and selective recourse to temporary intestinal stoma are mandatory to reduce long-term sequelae.

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