

Intestinal Atresia: A Four-Year Review of Cases in Ikeja-Lagos

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Abstract

Introduction: Intestinal atresia is one of the most common causes of neonatal intestinal obstruction worldwide. The pattern of presentation and management in our institution is reviewed. The objective of the study was to evaluate the pattern of intestinal atresias and stenoses in newborns who presented to our unit. **Materials and Methods:** A retrospective study of patients with intestinal atresias and stenoses who presented between September 2004 and November 2008. The clinical presentation, diagnoses, operative management, post operative care and outcome were obtained from the case notes. **Results:** Thirty cases were seen in that period, M:F; 1:1.5. Eleven (36.7%) were duodenal pathologies and 19 (63.3%) were jejunoileal. The main presenting symptom was bilious vomiting. Overall mortality is 40.9%. Prematurity and delayed presentations are the major contributing factors to mortality. **Conclusion:** Improvement of health care facilities as well as public health education to seek early intervention will improve outcome. Maternal awareness of this condition would lead to prompt seeking of treatment for paediatric patients.

Key words: Intestinal atresia, congenital malformation, total parenteral nutrition

Introduction

Intestinal atresia (IA) remains a well-known cause of neonatal intestinal obstruction worldwide. The pattern of presentation, management and outcome vary with the site of the obstruction and the presence of associated congenital anomalies. In recent times, the management has improved due to advancement in neonatal intensive care, improved preoperative resuscitation, anaesthesia and operating techniques as well as the use of total parenteral nutrition (TPN)^{1,2}. However, the combination of late presentation and lack of neonatal intensive care facilities and TPN have continued to contribute to poor outcome in the developing world^{3,4}. This study reviews the experience and the outcome of the management of neonatal intestinal atresia and stenosis in a teaching hospital in Lagos, Nigeria.

Materials and Methods

This is a retrospective review of cases of intestinal atresia managed in the Paediatric Surgical Unit of the Lagos State University Teaching Hospital between September 2004 and November 2008. Information regarding the biodata, clinical presentation, diagnosis, intervention and outcome of management were

extracted from the patients' case notes as well as theatre and ward records.

On presentation with features of intestinal obstruction, a clinical diagnosis of intestinal atresia was confirmed with plain abdominal x-ray finding of the classical double bubble appearance in duodenal atresia (DA); dilated upper intestine and multiple air-fluid levels in jejunoileal atresia (JIA) with paucity of intestinal gas distally. Upper gastrointestinal contrast studies were done in cases of duodenal atresia with perforated webs to further clarify the diagnosis. All the patients were adequately resuscitated with intravenous fluids and achieved adequate urine output of 2-3ml/kg/hr (1-2 ml/kg), nasogastric tube decompression, correction of electrolyte derangements and broad spectrum antibiotics. They had exploratory laparotomy under general anaesthesia. Complete distal bowel patency was ascertained at surgery using saline flush. Postoperative intravenous fluid, electrolyte and dextrose were given at maintenance rate. In the absence of TPN, NG tube feeding was introduced when there was bowel activity evidenced by bowel motions and significant reduction in NG drainage to less than 20ml/day. Patients were started on 3-5 ml aliquots of feeds, preferably expressed

breast milk, every 2-3 hours, and gradually increased as tolerated by the patients. Symptoms of short bowel syndrome were managed with antimotility drugs for the diarrhea, intravenous fluids and amino acids infusions. Feeding was stopped if vomiting persisted. Post operatively, the patients were discharged home when they tolerated oral feeding and demonstrated sustained weight gain.

Results

During the study period, 30 cases were identified. There were 12 (40%) males and 18 (60%) females (M:F; 1:1.5). The median age at presentation was 7 days (range 2 days – 6 years). Eleven of these patients (36.7%) had duodenal pathologies and 19 (63.3%) had jejunoileal atresias. Of the thirty cases, 22 (73.3%) case-notes were available for review and these were evaluated in this study. Nineteen patients presented in the neonatal period while the remaining three who had perforate duodenal webs presented at 14 months, 16 months and 6 years respectively with failure to thrive. The clinical presentation was in keeping with features of intestinal obstruction (Table 1). Four (18.2%) of the patients were delivered preterm. The history of polyhydramnios in pregnancy was not given or not taken. History of antenatal ultrasound scan was obtained from 12 (54.5%) of the mothers but no record of prenatal ultrasound diagnosis in any of the patients. Other congenital anomalies were seen only in DA patients (Table 2), however, these did not affect the outcome.

At surgery, 7 patients (31.8%) had DA according to the Gray and Skandalakis⁵ classification. Operative findings, treatment and outcome of management are shown in Table 2. One patient had a minor wound infection. Two of the patients with DA were premature. One patient died from respiratory failure complicating prematurity and another died from septicaemia.

The other 15 patients (68.2%) had JIA classified according to Grosfeld's⁶ modification of Louw's⁷ classification of JIA. Two of the patients with JIA were premature. Operative treatment involved resection of the blind bulbous proximal bowel with the atretic portion and anastomosis to the distal unused bowel. Tapering enteroplasty was done for one patient to preserve bowel length. Table 3 shows the clinical characteristics of patients with JIA. Two neonates had double-barrel enterostomies on account of severe peritonitis and eventually died of severe sepsis and malnutrition. One patient had a leak of the anastomosis for which a re-exploration was done but subsequently died. Mortality was recorded in seven (46.7%) patients with JIA.

Overall, 9 patients died giving a mortality rate of 40.9%. Of those that survived, post-operative follow-up period in the outpatient clinic ranged from 6 to 18 months. The patients had significant intestinal adaptation with well-formed stool and progressive weight gain including those who had resection of long length of bowel.

Table 1: Showing various clinical presentations in cases of intestinal atresia

Clinical feature	Frequency (%) DA	JIA
Vomiting	7 (100)	15 (100)
Abdominal distension	2 (28.6)	8 (53.3)
Constipation	1 (14.3)	11 (73.3)
Jaundice	1 (14.3)	3 (20.0)
Prematurity	2 (28.6)	2 (13.3)
Bloody stools	--	2 (13.3)

Table 2: Duodenal atresia – Pattern, management and outcome

TYPE	Associated problems	Procedure	Post-op feeding (days)	Post-op hosp. stay (days)	Outcome
I	Malrotation, Ladd's bands	Duodeno-duodenostomy	4	5	Discharged
I	Down syndrome	Duodenotomy and web excision	4	6	Discharged
I	Annular pancreas, prematurity	Duodeno-jejunostomy	Oral feeding was not established	14	Died Respiratory failure
I	_____	Duodenotomy and web excision	4	8	Discharged
I	_____	Duodenotomy and web excision	4	9	Discharged
III	Midgut malrotation, prematurity	Duodeno-jejunostomy	Oral feeding was not established	20	Died Septicaemia
III	_____	Duodeno-jejunostomy	4	7	Discharged

Table 3: Jejunoileal atresia – Pattern, management and outcome

TYPE	Age (days)	Associated problems			Procedure	Post-op feeding (days)	Post-op stay (days)	Outcome
		Prem.	Volv.	Perf.				
I	5	-	-	-	JIANas	5	10	Discharged
I	7	-	-	x	ICAnas	-	6	Died
I	7	-	-	x	DBE	-	19	Died
II	7	-	-	-	JIANas	4	6	Discharged
II	2	-	-	-	JIANas	5	8	Died
IIIa	3	-	-	-	JIANas	10	17	Discharged
IIIa	11	-	-	x	DBE	-	5	Died
IIIb	3	-	x	x	JIANas	15	31	Discharged
IIIb	5	-	x	x	JIANas	20	29	Discharged
IIIb	7	x	x	x	JIANas	-	7	Died
IV	6	-	-	-	JIANas	6	11	Discharged
IV	7	-	-	-	JIANas	21	40	Discharged
IV	10	-	-	-	JIANas	4	8	Discharged
IV	5	-	x	x	JIANas	-	9	Died
IV	13	x	-	-	JIANas	28	34	Died

Prem. – Prematurity; Volv. – Gangrenous volvulus; Perf. – Bowel perforation with peritonitis; JIANas – Jejunoileal anastomosis, ICAnas – Ileocolic anastomosis; DBE – double barrel enterostomy; post-op – post-operative

Discussion

The management of intestinal atresia remains a challenge in the developing world due to non-availability of facilities for TPN and neonatal intensive care^{3,4}. With this background, late presentation with associated fluid and electrolyte derangement, weight loss and malnutrition aggravate the management problems in our environment. Half of our patients presented after one week and only 5 (16.7%) presented by the age of 3 days. Delayed diagnoses by the primary healthcare giver contributed to the delayed presentation for surgical treatment. This occurred commonly in patients who passed scanty meconium initially and subsequently developed constipation and were managed for neonatal sepsis.

Prenatal diagnoses of intestinal atresia are increasingly being made by routine use of ultrasonography in monitoring of fetal development². The presence of maternal polyhydramnios and distension of the fetal stomach and duodenum by swallowed amniotic fluid are prenatal ultrasound features which are in keeping with duodenal atresia. Diagnosis was postnatal in all of our patients. Prenatal recognition of abdominal congenital anomalies using ultrasound scans is often missed in our environment⁸. Early recognition of the disease with prompt surgical intervention before the onset of severe metabolic complications and sepsis would improve outcome.

Postnatally, patients present with classical clinical features of GI obstruction. In partial obstruction usually

in DA, as seen in 3 of the patients in this series, the diagnosis may be delayed until late infancy or early childhood as symptoms appear when the child's diet becomes more solid and food impaction may occur⁹. Plain abdominal radiographs show the double bubble gas shadow, diagnostic of DA. JIA appear on plain radiographs as proximal dilated bowel loops and multiple air-fluid levels with absent gas shadows in the pelvis. Contrast studies of the upper gastrointestinal tract may be required in cases of partial obstruction and may demonstrate the presence of stenosis or perforate mucosal web². Other congenital anomalies, such as cardiac anomalies and anorectal malformations, are seen more often in association with DA than with JIA² as it occurs early in gestation when other organ systems are also at risk of malformations.

The operative procedure employed would depend on the site and type of the atresia, and the length of the viable residual bowel. Currently, the treatment of DA is most often duodeno-duodenostomy using a side-to-side anastomosis with a proximal transverse and distal longitudinal incisions to make a diamond shaped anastomosis¹⁰. In our series, duodeno-jejunosomy and duodenotomy with web excision was also done. Partial excision of the web is recommended to avoid injury to the ampulla of Vater which should be demonstrated and visualised. A tapering duodenoplasty may be necessary and is a useful technique in the management of the duodenal dysmotility associated with megaduodenum from DA^{11,12}.

In cases of JIA where the length of the remaining bowel is adequate, resection of the proximal dilated bowel is done to avoid functional obstruction and abnormal motility. If the residual bowel is short in length, a proximal tapering enteroplasty is done to preserve bowel length. An end-to-end, where the distal bowel is spatulated or end-to-oblique anastomosis is then done between parts of the bowel with comparable caliber. A temporary enterostomy is performed in the presence of meconium peritonitis resulting from perforation or when there is questionable bowel viability.

The post-operative recovery of patients with IA is hinged on nutritional support. The delay in presentation prolonged the period of fluid and electrolyte correction prior to surgery and consequently, the overall period of malnutrition. Non-availability of facilities for total parenteral nutrition in our environment necessitates early commencement of oral feeding after surgery. Dysmotility associated with IA as well as short bowel syndrome in our patients with JIA resulted in delayed establishment of oral feeding. Parenteral nutrition has been shown to improve the management outcome in short bowel syndrome¹³. However, the lack of the required facility necessitates the introduction of small frequent oral feeds in our patient as soon as bowel activity is demonstrated by negligible nasogastric tube drainage/aspirate and bowel motions. The patients had intravenous administration of dextrose infusion, amino acids infusion when available and electrolytes as required. Antimotility drugs are given when diarrhoea persists. Thus, the JIA patients had a lengthy hospital stay compared with the DA patients.

The overall mortality rate of 40.9% is high and is comparable with findings from studies within Nigeria and in Kenya^{3,4,14}. Factors that contributed to mortality are delayed presentation, peritonitis, prematurity, malnutrition and short bowel syndrome and lack of facility for TPN. These remain the same till date as these factors have not been adequately addressed in the developing world even in healthcare facilities such as ours where health service to the patients in the index age group is free. In developing countries, where most patients pay for health services out-of-pocket, the concept of free healthcare for children goes a long way to provide access to healthcare as many more patients present to the hospitals. However, late presentation for healthcare continues to be a recurrent major factor contributing to high morbidity and mortality in various disease conditions in resource poor countries.

There is need for improvement in our paediatric health care to achieve better results such as provision of facilities for TPN, neonatal and paediatric intensive

care facilities and staff training for skill acquisition in prenatal fetal monitoring, perioperative neonatal care and surgical procedure applied. Education of mothers, during antenatal classes and in immunization clinics, on the recognition and significance of various symptoms in a neonate and positive outcome of early diagnosis and intervention would encourage early postnatal presentation to the surgeon. Furthermore, in our culture where the societal and extended family influence is very strong, a general public enlightenment is important. All neonates with vomiting should be investigated routinely for intestinal obstruction. In our society, some areas of medical care have been known to enjoy rapid development helped by interest of the governing / sponsoring bodies. Thus, paediatric care givers need to play advocacy roles while looking inwards to find ways of improving our practices and the quality of healthcare given to our patients.

In conclusion, late presentation of intestinal atresia patients for definitive intervention is a major contributor to high mortality rate in these patients. In order to improve the outcome in resource poor areas, public health education to achieve early presentation to the specialist surgeon for prompt treatment should be instituted.

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