MANAGEMENT OF PATIENTS WITH JEJUNOILEAL ATRESIA: FACTORS AFFECTING SURVIVAL

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SUMMARY

A total of 75 newborn patients with jejunoileal atresia were studied during 4 year period from August 1993 to July 1997. All the patients were delivered at home with fullterm gestation period spontaneously and were admitted through emergency to the department of Paediatric Surgery PGMI/LRH Peshawar. There were 35 (46.66%) male babies and 40 (53.33%) female babies. Mean birth weight was 2.45 kg with 25 (33.33%) patients less than 2.5kg and 50 (66.66%) patients 2.5-3 Kg. Sixty (80%) patients presented between 2-5 days of age while 15 (20%) presented at the age of more than 5 days. Bilious vomiting was present in all (100%) patients. Abdominal distension in 50 (66.66%) patients. Constipation in 54 (72%) and jaundice in 9 (12%) patients. Maternal history of polyhydramnios was present in 30 (40%) patients. Twenty one (28%) patients were having associated hypothermia. Thirty five out of seventy five patients were having jejunal atresia while 40 had ileal atresia. Type IIIa and II were the most common type of atresia which were found in 30 (40%) & 24 (32%) of patients respectively. Fifty four patients were treated by primary bowel anastomosis and 21 with temporary enterostomies. Thirty (55.55%) patients died postoperatively in the group treated by primary bowel anastomosis while no patient died from the group of patients treated by enterostomies.

Delay in presentation, lack of facilities (intensive newborn care & newborn anaesthesia) were the major problems related with the management of jejunoileal atresia. The over all mortality is 40%. Septicemia (66.66%), aspiration pneumonia (16.66%) and metabolic disturbances (unknown cause - 16.66%) being the major cause of death. Type of atresia (apple peel and multiple atresia), level of atresia (jejunal atresia), associated intra-abdominal complications, other congenital anomalies (cardiac) and the type of treatment offered are the other parameters which affected the survival of these patients.

INTRODUCTION

Jejunoileal atresia is a common congenital abnormality to cause neonatal intestinal obstruction occurring one in 330 to 1500 live births.[1] A number of factors have been found to cause atresia of the foetal small bowel in the intrauterine life including failure of recanalization, genetic, familial, environmental and or ischemic factors.[2-5] Bowel atresia has been found in twins and also in families with certain associated anomalies.[6-10] The occurrence of bowel atresia in association with gastroschisis, omphalocele, volvulus, malrotation, internal hernias, meconium ileus and meconium peritonitis has been well documented in the world literature which explains the vascular

compromise theory as the cause of small bowel atresia.[4,11-13] Most of the neonates with small bowel atresia are under weight because of the impaired absorption of proteins in the amniotic fluid.[14] Impaired absorption of amniotic fluid due to obstructed bowel leads to Polyhydramnios in the mother and distended loops of small bowel which can be seen on prenatal ultrasonography.[9,15,16] The mortality of these patients has markedly decreased with the advent of newborn intensive care units, advances in newborn anaesthesia and with the use of total parenteral nutrition.[17]

In developing countries where the facilities of newborn intensive care unit and newborn anaesthesia are lacking, the mortality of these patients is still unacceptably high.[18] Majority of these neonates are usually otherwise normal, therefore, further improvement in the survival of these patients must be achieved.[17] This study was designed to assess the survival and mortality rate and to study the factors affecting the outcome of treatment in our circumstances.

MATERIAL AND METHODS

A survey study of 75 patients was carried out in a prospective fashion at Paediatric Surgery Unit PGMI/LRH Peshawar from August 1993 to July 1997. A randomised collection of 75 patients with jejunoileal atresia was done and analysed with respect to age at presentation, birth weight, gestational maturity of the baby, pathological type and level of atresia and the result of treatment.

Excluding the other causes of neonatal intestinal obstruction, patients were assessed and associated anomalies were recorded. These patients were operated upon by different grades of Surgeons of Paediatric Surgery Unit PGMI/LRH Peshawar. Most of the patients (50) were operated at casualty operation theatre, while the remaining (25) were operated on elective list after necessary resuscitation. Resuscitation was done in the ward by giving Ringer's Lactate to correct fluid deficit and 1/5th saline in 10% dextrose along with 2-3 meq/kg Potassium chloride as maintenance fluid. Size 10 FR nasogastric tube was put in to decompress the stomach and distended bowel. All the patients were put on prophylactic antibiotics (Cefotaxime, Gentamycin & Metranidazole) along with Vitamin K injection 10 mg preoperatively.

Under endotracheal general anaesthesia, a right transverse upper abdominal incision was used in every patient. Bowel atresia was grouped together according to the classification of Grosfeld et al[9]. Type I atresia consisted of membranous occlusion of the bowel lumen. Type II atresia was the one in which the two atretic ends were connected by a fibrous cord. Type IIIa atresia was the one in which the two atretic ends were separated by a defect in the mesentery. Type IIIb (apple peel) atresia consisted of jejunal atresia with extensive mesenteric defect and the distal small bowel receiving its sole blood supply from the right colic or ileocolic artery coiled around it. Type IV atresia consisted of multiple atresia of the small bowel of any of the above mentioned types from type I to IIIa. At laparotomy, associated intra abdominal complications such as meconium peritonitis, meconium ileus, malrotation, bowel perforation, volvulus and other conditions were recorded and the choice of surgical procedure selected according to the condition of the patient. Primary bowel anastomosis was done in patients either with no associated complications or with minimal complication. Temporary enterosotomies (exteriorization) were done in patients with severe intra-abdominal complications.

Postoperatively every patient was put on total parenteral nutrition during the first half of this study while in the second half TPN was used in selected patients who were kept nil by mouth for more than 7 days. Postoperative course during hospital stay

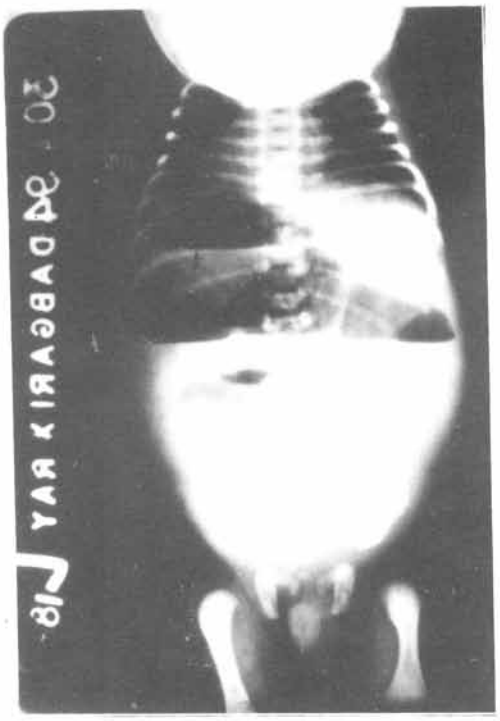

Fig. 1. Small bowel atresia upright film showing multiple air fluid levels.

was analysed for every patient. Mortality and survival rates following emergency surgery were calculated for all the pathological types of atresia and for all the surgical procedures undertaken. Every patient who survived and was discharged to home was followed up for a minimum of six months. The patients with enterostomies who were discharged to home were advised to come for closure of their stomas. Sweat test was performed in every case who was followed up.

RESULTS

A total of 75 patients with jejunoileal atresia were studied. There were 35 (46.66%) male and 40 (53.33%) female neonates. All the patients were the product of full term gestation and spontaneous normal vaginal delivery. The mean birth weight was 2.45 kg with 25 (33.33%) patients of lowbirth weight (< 2.5 kg) and 50 (66.66%) patients between 2.5-3kg. 60 (80%) patients presented between 2-5 days of age while 15 (20%) patients presented at age more than 5 days (table No. 1). There was no patient presenting earlier than 2nd day of life. All the patients presented with features of intestinal obstruction. These included bilious vomiting in 75 (100%) patients, abdominal distension in 50 (66.66%), failure to pass meconium in 54 (72%) and jaundice in 9(12%) patients. Maternal polyhydramnios occurred in 30(40%) patients, 10 with ileal atresia and 20 with jejunal atresia. Hypothermia was present in 21 (28%) patients.

In all patients diagnosis was confirmed by plain roentgenography and Barium contrast enema (Fig 1, 2). Barium enema showed microcolon in 20 (26.66%) patients while malrotation of bowel or perforation was not evident in any case. Blood examinations such as blood complete, blood sugar, blood urea, liver function tests, serum electrolytes and blood gases were done in selected cases showing features of systemic sepsis and metabolic disturbances.

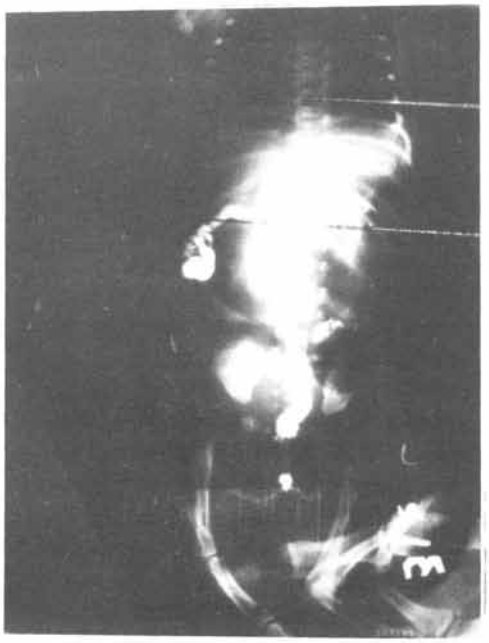

Fig. 2. Barium enema showing microcolon.

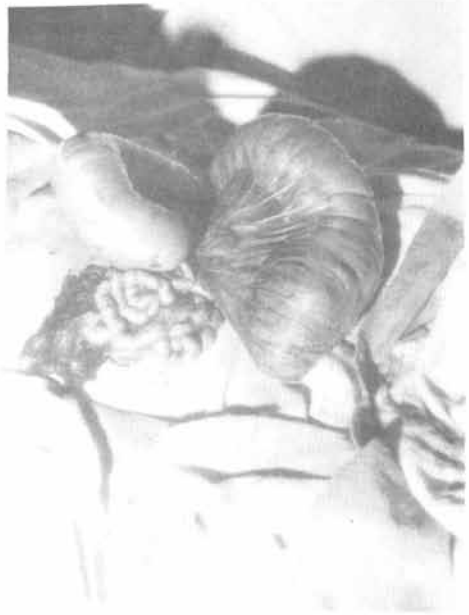

Fig. 3. Jejunoileal atresia showing proximal dilated and distal narrow bowel.

All the patients were assessed preoperatively and after 12-24 hours of resuscitation they were subjected to surgery (Fig 3). At surgery, the type and level of atresia was identified which included 35 (46.66%) jejunal atresia and 40 (53.33%) ileal atresia. The most common types of atresia were type IIIa and type II, which occurred in 30(40%) and 24 (32%) patients respectively. Type I membranous atresia was found in 10(13.33%) patients, type IIIb apple peel atresia in 6(8%) and type IV (multiple) in 5(6.66%) patients. Associated pathological findings were recorded in 62(82.66%) patients which included 24(32%) meconium peritonitis, 12(16%) meconium ileus, 6(8%) malrotation of bowel, 15(20%) volvulus of small bowel and 5(6.66%) gangrenous bowel with perforation.

Twenty one (28%) patients were treated by exteriorization (enterostomies) who has had some of the complications associated with atresia. These include 10 Bishop-koop ileostomies, 6 Santulli ileostomies and 5 double barrel ileostomies. Fifty four (72%) patients were treated by primary bowel anastomosis including 50 simple end to end anastomoses, 2 tapering enteroplasty and 2 multiple anastomoses. Out of 54 primary bowel anastomoses, 41 were with some of the intra-abdominal complications and 13 with out any complication.

The post operative recovery was quick with enterostomies (3-7 days) and delayed (7-15 days) in whom primary bowel anastomoses were performed specifically in the presence of complications. Delayed bowel function (10-20 days) was observed in 5 patients with type IV atresia (multiple atresia) and 2 patients with jejunal atresia in whom tapering entesoplasty was performed as a bowel lengthening procedure. Post operatively every patient was put on triple antibiotic regimen (Cefotaxime, Gentamycin & Metranidazole). Fluid & electrolyte balance was maintained according to the need of the patient. Total parenteral nutrition (TPN) was administered to every patient during the first half of this study but later on it was used in selected cases who were nil orally for more than 7 days.

Mortality was 66.66% in patients with apple peel atesia (type IIIb), 40% in type IV (multiple), 40% in type I, 41.66% in type II and 33.33% in type IIIa (table - II). The most common complication which led to the death of the patients were septicemia 20 (66.66%), aspiration precumonia 5 (16.66%) while in 5(16.66%) patients the cause could not be established (probably severe metabolic disturbances).

Intravenous line sepsis occurred in all patients in whom the line was used for TPN. There was no patient with abdominal wound sepsis, wound dehiscence and anastomotic leak.

TABLE – I

CLINICAL FEATURES AND COMPLICATIONS

FEATURES	No. (% age)
Age at Presentation :	
2-5 days	60(80%)
> 5 days	15(20%)
Sex :Male babies	35(46.66%)
Female babies	40(53.33%)
Weight : < 2.5 kg	25(33.33%)
2.5-3 kg	50(66.66%)
Cause of death :	
Septicemia	20(66.66%)
Aspiration pneumonia	5(16.66%)
Unknown	5(16.66%)

The over all mortality was 40% which was more in jejunal atresia (43%) than ileal atresia (37.5%). Thirty (55.55%) out of fifty four patients who were treated by primary bowel anastomoses died postoperatively while no patient died from the group who were treated by enterostomies. Forty five patients (60%) survived who were discharged to home included 24 primary bowel anastomoses and 21 enterostomies (Table-II).

Thirty patients were seen in the follow up examination including 20 with primary bowel anastomoses and 10 with enterosomies, whose stomas were subsequently closed. Fifteen patients were lost to follow up included 11 patients with ileotstomies and 4 with primary bowel anastomoses. Sweat test was negative in all the patients who were followed up.

DISCUSSION

The aim of study was to identify the factors that affected the survival of patients with jejunoileal atresia. Birth weight, age at

TABLE – II

CHARACTERISTICS AND TYPES OF JEJUNOILEAL ATRESIA AND THE RESULTS OF TREATMENT.

CHARACTERISTICS	NO. (% age)	RESULTS	
		EXPIRED	SURVIVED
Total Patients	75	30(40%)	45(60%)
Level of Atresia :			
Jejunal Atresia	35(46.66%)	15(43%)	20(57%)
Ileal Atresia	40(53.33%)	15(37.5%)	25(62.5%)
Type of Atresia			
Type I	10(13.33%)	4(40%)	6(60%)
Type II	24(32%)	10(41.7%)	14(58.3%)
Type IIIa 30(40%)	10(33.33%)	20(66.66%)	
Type IIIb	6(8%)	4(66.66%)	2(33.33%)
Type IV 5(6.66%)	2(40%)	3(60%)	
Surgical Procedure :			
Primary anastomosis	54(72%)	30(55.55%)	24(44.45%)
Enterostomies	21(28%)	———	21(100%)

presentation, maturity of the baby, type and site of atresia, associated problems and type of treatment offered were the variables correlated with the short term outcome and survival of patients with jejunoileal atresia.

Regarding the cause of jejunoileal atresia it was seen that there was no obvious cause for the development of atresia such as intussusception ,bowel strangulation in a hernia or volvulus in the intrauterine life which is well documented in the literature.[4,6,7,13] The occurrence of jejunoileal atresia was suggested to be due to vascular compromise of the foetal bowel during intrauterine development which was confirmed by the experimental work of low and Barnard in 1955.[7] The hypothesis of failure of recanalization (malformation) was suggested to be due to genetic or exogenous factors leading to bowel atresia.[2,19,20] Although there was no obvious cause resulting into bowel atresia in this study, many of them (62 patients) were associated with one or other of the complications including meconium peritonitis, meconium ileus, malrotation, volvulus and gangrenous bowel with perforation which could be either the result or cause of bowel atresia.

Delay prior to admission was the most common observation found in this study which led to the demise of many patients (30) due to septicemia, aspiration pneurmonia and metabolic disturbances. Majority of these patients presented after the 2nd day of life during which time the patients could have vomited, developing pneumonia, systemic sepsis and metabolic disturbances, due to obstructed bowel. Early diagnosis and prompt treatment of these patients has been stressed upon by many observers[17,21,22]. low birth weight and gestational maturity of the newborn baby also affect the results of surgical treatment but they do not appear to affect the result of treatment in our study. The presence of associated illnesses did affect the survival significantly such as meconium peritonitis and others which occurred in 62 (82.66%) patients. The presence of these complication also affect the result of surgery shown in various studies.[3,10,23]

The type of atresia and operative procedure affected the outcome as all these patients were treated in similar circumstances. Apple peel (type IIIb) atresia had the worst prognosis (mortality 66.66%). Those two (33.33%) apple peel atresia who survived (33.33%) were treated by Santulli ileostomy.

Seashore et al[24]; reports 54% mortality of patients with apple peel atresia which is slightly less than ours. Despite the use of total parenteral nutrition, intensive care of the newborns mortality of patients with apple peel atresia is still 45%. Manning et al[25]; have suggested that spectacular improvement in the survival of these patients can be achieved by the provision of early diagnosis, appropriate surgery and parenteral nutrition. Multiple (type IV) atresia showed 60% mortality (3 out of 5)and difficult to classify them as they involved the bowel from jejunum to terminal ileum. The most proximal atresia was at the level of jejunum so they were regarded as jejunal atresia. Smith and Glasson has reported a 57% mortality for patients with type IV (multiple) atresia.[23]

The operative technique had a direct impact on the survival of these patients. Many observers have suggested that primary bowel anastomosis should be attempted in all cases as temporary enterostomies have resulted into prolonged morbidity and raised mortality of these patients.[17,22,23,26] High morality was observed in this study (55.55%) in the group of patients treated by primary bowel anastomosis (30 died out of 54 patients). Although there was no immediate or early post operative mortality in the group of patients (21) treated by temporary enterostomies, only 10 patients were seen in the follow up visits and later on treated by closure of the stomas.

Fifteen patients were lost to follow up including four with primary bowel anastomosis and eleven with ileostomies. All of the patients (11) with enterostomies who did not come for closure were considered dead, although they were advised to be brought for closure of their ileostomies. In this way a delayed mortality of 52.38% could be speculated in patients treated by ileostomies.

The patients treated in this study were managed in the general ward without the help of intensive newborn care. The common problems which were faced during the management of these patients included delay in presentation, shortage of specialized personnel and lack of facilities. In the first half of this study every neonate with bowel atresia was put on total parenteral nutrition (TPN) but it was observed that many deaths occurred due to unexplained reasons. In the second half of the study TPN was used in selected patients who remained nil orally for more than 7 days with a dramatic decrease in some of the complications related to the use of TPN. However in recent years during some studies the mortality of these patients has decreased to 10%. They believe that it is because of the generous use of TPN, advances in newborn anaesthesia, newborn intensive care facilities and control over sepsis by the generous use of prophylactic antibiotics.[17,25]

Although the overall mortality shown in this study is 40% which is not satisfactory in the days of modern surgery where all the standards of newborn management are fulfilled, but in the developing countries where these facilities are lacking, the results are still encouraging. The prognosis of these patients can be improved by the provision of modified surgical techniques, use of TPN and monitoring of metabolic disturbances. With the exception of compromised patients in the presence of complications, where temporary enterostomies can not be avoided, in all others primary bowel anastomosis is to be preferred. To avoid short bowel syndrome on one hand and temporary enterostomies on the other, closed primary bowel anastomosis and bowel lengthening procedures are recommended and are most appropriate.

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