CLINICOPATHOLOGICAL STUDY OF JEJUNOILEAL ATRESIA

Dissertation submitted for

M.Ch. Degree Examination

PAEDIATRIC SURGERY

Branch V

Institute of Child Health & Hospital for Children

MADRAS MEDICAL COLLEGE

THE TAMIL NADU DR. MGR MEDICAL UNIVERSITY

CHENNAI

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CERTIFICATE

Certified that the dissertation entitled "**Clinicopathological Study of Jejunoileal atresia "** is the original work undertaken by **Dr. M. Balasundaram** under our guidance and supervision, in the Department of Paediatric Surgery, Institute of Child Health and Hospital for Children, Madras Medical College, Chennai-3 during the period of his postgraduation in M.Ch Paediatric Surgery from

2005-2008, in partial fulfillment of the university rules and regulations for the award of M.Ch. degree.

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DECLARATION

I declare that this dissertation entitled "**CLINICOPATHOLOGICAL STUDY OF JEJUNOILEAL ATRESIA"** has been conducted by me at the Institute of Child Health and Hospital for Children. It is submitted in part of fulfillment of the award of the degree of M.Ch. (Pediatric Surgery) for the August 2008 examination to be held under the Tamil Nadu Dr.M.G.R. Medical University, Chennai. This has not been submitted previously by me for the award of any degree or diploma from any other university.

Dr. M. BALASUNDARAM

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INTRODUCTION

Intestinal atresia is one of the most common surgical disease in neonates. Jejunoileal atresia occurs more frequently than duodenal or colonic atresia[1]. It accounts 30% of all cases of neonatal intestinal obstruction. The incidence of jejunoileal atresia varies between 1/300 and 1/3000 live birth. **Ravitch et al**[2] estimated the over all incidence of intestinal atresia at 1 per 2719 live birth. Boys and girls are equally affected. Down Syndrome is most uncommon in babies with jejunoileal atresia compared with duodenal atresia[3].

The intestine proximal to obstruction is usually dilated and hypertrophied and has a cynosed appearance and may have patches of necrotic areas. The peristaltic movement in this segment is subnormal and ineffective.

The distal bowel is unused and worm like, potentially normal in length and function. If the atresia has occured late in intrauterine life the bowel distal to atresia have a near normal calibre.

At the level of atresia, the ganglion of the enteric nervous system are atrophic and hypocellular. These changes are most likely the result of ischemia.

Intestinal dysmotility is an important problem in the post operative management of patients with jejunoileal atresia. The alterations of neural and muscular elements and the extent of histologic changes proximal and distal to atresia may contribute to the postoperative intestinal dysmotility in these cases, but the etiology of this disease is not yet to be understood[4].

The distended proximal bowel produces a significant technical problem for anastomosis and also predisposes to the intestinal dysmotility as it is deficient of muscular and neural elements.

The operative techniques and medical treatments, including nutritional therapy, have led to an improvement in the outcome of patients with intestinal atresia, some problems related to the management of intestinal atresia still remain unresolved.

The post operative intestinal dysmotility is frequently associated with dilatation of the proximal intestinal segment but its etiology is not yet fully understood. Hypoplasia of intramural nerves and pacemaker cells and abnormal musculature in the proximal segment of jejunoileal atresia were accepted as causative factors for intestinal dysmotility[4]

REVIEW OF LITERATURE

In 1684 **Goeller** described the autopsy findings in a neonate with ileal atresia[5].

In 1804 **Voisin** performed enterostomy for intestinal atresia.

In 1888 **Chiari** found intussusception as a cause of atresia secondary to ischemic changes.

In 1889 **Bland sutton**[6] proposed a classification of the types of atresia and postulated that intestinal atresia occurred at the sites of obliterative embryologic events, such as atrophy of vitelline duct.

In 1894 **Wanitschek** unsuccessfully attempted the first resection and anastomosis for intestinal atresia[7].

In 1900 **Tandler**[8] proposed the theory that atresia was related to failure of recanalization (Vacuolization) of Solid cord stage of bowel development.

In 1910 **Johnson**[9] confirmed theory of Tandler.

In 1911, **Fockwens**[10] performed the first successful anastomosis.

In 1912 **Spriggs**[11] suggested that Mechanical accidents, including vascular occlusions might be responsible for these occurrance.

In 1955 **Louw and Barnard**[12] confirmed the role of Late intrauterine mesentric accidents in most of jejunoileal atresias.

In 1996 **Gross and coworkers**[13] reported familial instances of combined duodenal and jejunal atresia.

Guttman and **colleggues**[14] Aigrain and associates[15] **Kimble** and **coworkers**[16] **Peri** and **Fujimoto**[17] and **Fourcede** and **colleagues**[18] described

hereditary instances of multiple intestinal atresias without evidence of vascular insults indicating a malformed process possibly due to an autosomal recessive transmission.

Kilani and associates[19] described a familial pattern of jejunal atresia with renal dysplasia inherited as an autosomal dominant trait.

Rothenbeg and coworkers[20] noted an instance of multiple atresias associated with severe immuno deficiency characteristic by agammaglobulinemia, B Cell deficiency and impaired T cell function.

Alvarez SP and coworkers[21] in their study they noted coexistence of segmental intestinal musculature defect with small bowel atresia.

Masumoto and coworkers[22] suggested hypoplasia of intramural nerves and pacemaker cells and abnormal musculture in the proximal segments of jejunoileal atresia were accepted as causative factors for intestinal dysmotility.

Hemdy et al[23] reported that in the dilated bowel at 2 cc proximal to the atresia the intermuscular ganglion were smaller and less in number and muscle layers were thinner on the antimesentic side when compared with those on the mesentric side and in control specimens.

Shin-Feng-Huang et al[24] in their study of segmental defect of the intestinal musculature of a new born, they suggested the absence of musculature and replacement of fibrosis might have been secondary to an injurious process such as ischemia or inflammation.

Tibboel et al[29] in their study showed the effects of temporary local ischemia and general hypoxia on the development of intestines of chick embryos. In experimental local temporary ischemia, chick embryo suffered intestinal stenosis or atresia. The intestinal loops proximal to the obstruction were dilated and the intestines wall was extremely thin. The structure of both layers of the musculature become abnormal, whereas the intestinal wall was normal distal to the obstruction. This study showed that temporary disruption of the circulation in the mesentric blood vessels during the fetal period leads to atresia or stenosis with structural abnormalities in the muscle coats.

Tepas et al[30] experimentally produced intestinal atresia in fetal lambs by an intrauterine disruption of the mesentric blood supply and showed that the dilatation of the proximal segment induces the involution and lysis of the ganglion cells after an initial hyperplasia of myentric ganglia as the irreversible distension continues.

Pickard et al[31] in their study showed muscle layers were irregular with segmental muscular hypertrophy.

Powell RW[32] in 1990 observed that the absence of musculature in any portion of the intestinal tract, as well as the ganglion cells leads to localized dilatation of the affected segment with obstruction caused by lack of the intestinal peristalsis and when the dilated proximal intestine, which have segmental musculature and neural defect was not adequately resected, the dilatation of that intestinal segment and stasis have been observed.

P. Ramachandran[33] and associates in their study showed significant changes in the three dimensional morphology of the myentric plexus in the atretic bowel in neonates with jejenoileal atresia.

Embryology and Etiology of Jejunoileal Atresia

Various theories put forward to explain the etiopathogenesis of jejunoileal atresias. They are:

1. **Developmental defect**
2. **Inflammatory changes**
3. **Fetal Vascular accidents**

A. **Developmental defect:**

a) **Bland Sutton** in 1889 proposed that intestinal atresias occurred secondary to excessive resportion of the Vitelline defect.

b) **Politzer** opined that disproportionate growth of the gut and inadequate endodermal proliferation led to atresias.

c) **Tandler** classical Failure of recanalization theory was put forward in 1899 and stated that there was a normal phase of epithelial proliferation and occlusion during gut embryogenesis. Failure of vacuolization and recanalization led to atresias**.** **Mount Souris** has subsequently shown that there is no solid stage in the small bowel (except duodenum) as described by **Tandler**[25].

B. **Inflammatory changes:**

Bernstein proposed that Scar formation following perforation secondary to meconium ileus or other causes can cause atresia.

C. **Fetal Vascular accidents:**

Among the three, the most favoured theory is that of intravascular accident with ischemic necrosis of sterile bowel and subsequent resorption of the atretic segment. Jejunal atresia which happens at the later period of intrauterine life thought to be due to intrauterine mesentric vascular accident such as volvulus, intussusception, internal hernias, band, meconium peritonitis and interference with blood supply to a segment of bowel.

Nipping of bowel in abdominal defect like omphalocele and gastroschisis may also result in bowel atresia.

Grossfeld and Cloworthy[26] observed the occurrence of jejunal atresia with infarction of entire midgut in a tight gastroschisis defect. Iatrogenic postpartum ileal atresia owing to umbilical clamping of an occult omphalocele was reported by **Vassy and Boles**[27] **Komuro**[28] described intrauterine intussusception as a cause of intestinal atresia.

Localised nature of vascular accident occuring late in fetal life usually present with low incidence of coexisting abnormalities of extra abdominal organs.

This anomaly usually is not genetically determined although affected monozygotic twins have been described and genetic basis has been established for type 3 and 4 atresia.

PATHOLOGY

The ischemic insult not only causes the morpholgic abnormalities but adversely influence the structure and subsequent function of the remaining proximal and distal bowel. The intestine proximal to obstruction become dilated and hypertrophied and has a cynosed appearance and may have patches of necrotic areas either due to sustained intraluminal pressure or secondary to volvulus.

Sometime perforation may develop antenatally, leading to meconium peritonitis. The perforation usually occurs proximal to the obstruction in the bulbous blind end. Poor vascularity of the terminal end of the proximal bowel has been confirmed by postmortem Barium Sulfate injection studies. It is also believed that this vascular compromise is a postpartum phenonmenon and results Secondary to swallowing of air and progressive distension (Tension gangrene of Rutherford).

Nixon et al showed the peristaltic movement in this segment are subnormal and ineffective. Histological and histochemical abnormalities can be observed upto 20 cms cephaloid to atretic segment.

The distal bowel is unused and wormlike potentially normal in length and function. If the atresia has occurred late intrauterine life, the bowel distal to atresia have a more normal calibre.

At the level of atresia, the ganglion of the enteric nervous system are atrophic and hypocellular with minimal acetylcholinesterase activity. These changes are most likely the result of ischemia.

The discrepancy in diameter between the lumen of proximal and distal bowel may very from 2 to 20 times depending on the completeness of the obstruction and its distance from stomach. The more distal from the stomach,

the dilatation is lesser. Resection of the dilated bulbous proximal end produce better results. The proximal end of distal atretic bowel has been subjected to a similar insult and requires resection at the time of surgical correction of the atresia.

INCIDENCE:

Jejunoileal atresia is seen in 1/360 to 1/3000 live births.

Jejunal atresia 51% (proximal 31%, distal 20%)

Ileal atresia 49% (Proximal 13% distal 36%)

Single atresia - approximately 90%

Multiple atresia 6 - 20%

5% atresia related to internal hernia.

There is no sex predilection.

CLASSIFICATION OF JEJUNOILEAL ATRESIA

Classification systems for jejunoileal Atresia Bland Sutton (1889)

Type I Membranous atresia.

Type II Blind ends connected with fibrous cord with or without mesentric defect.

Type III Blind gut ends separated by a gap with associated mesentric defect.

Martin and Zerella 1976

Type I - Membranous atresia.

Type II - Blind end of gut connected with fibrous cord or separated by a gap.

Type III - Multiple atresia

Type IV - Apple Peel atresia.

Grosfeld Modification of Low classification 1979

Type I Membranous atresia

Type II Blind ends of gut connected by fibrous cord, intact mesentry.

Type IIIa - Blind ends separated by gap and V shaped Mesentric defect.

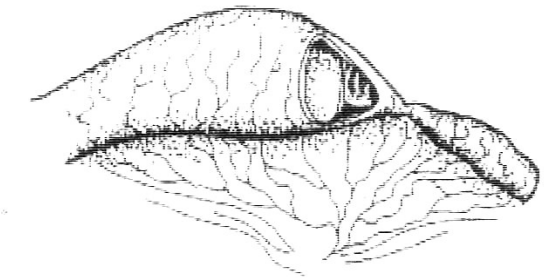
Type IIIb - Apple Peel atresia

Type IV - Multiple atresia.

Type I 20 to 30%

The obstruction is caused by a membrane or web formed by Mucosa and Submucosa. The proximal dilated and distal collapsed bowels have muscular continuity without a mesentric defect. The wind sock deformity occurs as a result of peristalsis forcing the web into the distal intestinal lumen.

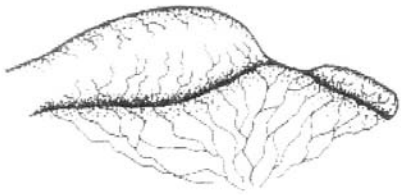
The bowel length is normal.



TYPE 1 ATRESIA
(membrane or web)

Type II 20% to 25% (blind ends joined by Fibrous band)

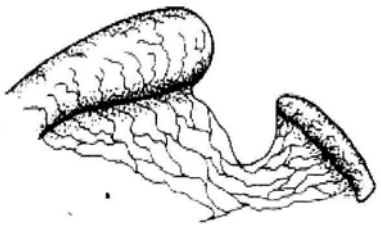
The proximal bowel terminates into bulbous blind end, which is grossly distended and hypertrophied for several cms and aperistaltic. More proximally the distension is less marked and bowel appears normal. Distal bowel is collapsed, two blind ends are joined by a fibrous band. Intestinal mesentry is normal. Small intestine is usually of normal length.



TYPE 2 ATRESIA
(blind ends joined by fibrous band)

Type III Atresia (disconnected blind ends)

Type IIIa (15%) The appearance is similar to that of type 2 but ends are completely separated with a V shaped gap in mesentry. The total bowel length is reduced. cystic Fibrosis commonly associated with this type of atresia.



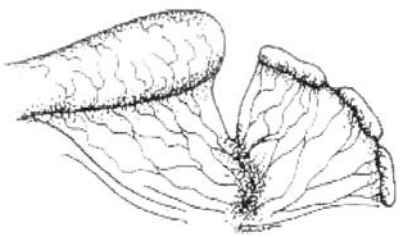
TYPE 3 A
(disconnected blind ends with a V shaped gap in mesentry)

Type IIIB (<5%) (Apple Peel, christmas tree, May pole deformity) Blind ends are disconnected as in Type IIIa, mesentric defect is substantial. This type is a consequent of extensive infarction of mid gut secondary to proximal superior mesentric artery occlusion producing proximal jejunal atresia. The distal ileum remains viable, receiving its blood supply via a precarious collateral from the arterial supply to the right colon, around which the ileum is coiled. Occasionally additional type I or type II atresias are found along the distal blind end. There is always a significant reduction in intestinal length. These babies are usually premature and of low birth weight.

TYPE 3 B
(apple peel, Christmas tree, maypole deformity)

Type IV Atresia 10% -25% (String of Sausage)

Multiple sites of atresia, which could be combination of type I to type III atresia giving the morphological appearance of String sausages. Bowel length is usually reduced. Babies with type IV atresia are often premature.



TYPE 4 ATRESIA (String of sausages)

ASSOCIATED ANAMOLIES

Jejunal atresia is associated with prematurity 30%, gastroschisis 10 to 20% (American Academy of Paediatric surgery). Spinal dysraphism, Down Syndrome are very rare (0.5%) Type 3B is associated with biliary atresia, PUJ obstruction and imperforate anus.

Rare instance of jejunoileal atresia observed to coexist with biliary, duodenal colonic atresia, Hirschsprung's disease and arthrogyposis in mothers who received Ergotamine and caffeine tablets for migraine during pregnancy.

Type 3B has associated anomaly like malrotation and may develop short bowel syndrome. Malrotation is observed in 10%-30% of babies with atresia.

10% babies with atresia have meconium peritonitis and meconium ileus. Meconium ileus must be considered in babies with jejunoileal atresia who have signs of prenatal volvulus, meconium peritonitis, microcolon with pellets of meconium.

CLINICAL FEATURES

PRENATAL DIAGNOSIS:

Polyhydraminios occurs in about 24% of cases with Jejunal atresia. The presence of echogenic dilated bowel loops on maternal ultrasonography after 20 weeks gestation is another indicator of bowel obstruction. Bile stained Liquor and bile acid concentration in amniotic fluid may indicate obstruction distal to the ampulla of vater. All babies with history of polyhydramnios in mother must be evaluated postnatally to rule out atresia.

POSTNATAL CLINICAL PRESENTATION

Bilious vomiting after the birth is the common mode of presentation. It indicates neonatal intestinal obstruction until proved otherwise. Babies with atresia usually develop bilious vomiting on 1st day of life but in 20% of children it may be delayed to 2 or 3 days. Higher the obstruction, earlier is the onset of vomiting and more forcefull and distension may not be marked.

Abdominal distension is always present in children who present later, more so with the lower intestinal atresias, where the distension is generalised. In more proximal atresia the distension is confined to upper abdomen which is relieved by nasogstric tube aspiration. When perforation occurs the distension may be severe and associated with respiratory distress, tenderness, rigidity, edema and erythema of abdominal wall, visible intestinal peristalsis and dilated loops may be felt. Bilious vomiting and intestinal patterning on abdominal wall is also seen in jejunal atresia.

Most of the children with intestinal atresia fail to pass meconium. But 20% babies with jejunal-atresia evacuate normal meconium shortly after birth. When rectal wash is given, grey coloured whitish meconium, occasionally green coloured may be passed. Constipation is usually not absolute. Gross distention with dilated veins over the abdominal wall may indicate the presence of meconium peritonitis. Occasionally ischemic bowel

may present with bleeding per rectum as seen in Type 3B. Dehydration, fever, hyperbilirubinemia and aspiration pneumonia occur with delay in diagnosis.

Few children may present with jaundice (unconjugated hyper bilirubinemia) in 40% of cases with proximal atresia and 20% of cases with distal atresia. Jaundice is due to an elevation of indirect bilirubin caused by the presence of 4B glucuronidase in the neonatal intestinal mucosa. This enzyme unbinds conjugated bilirubin and increases entero-hepatic circulation of unconjugated bilirubin. Conditions like Mucoviscidosis and rotation anomalies of intestine will produce bowel atresia. Type 3B atresia has a familial tendency.

DIFFERENTIAL DIAGNOSIS

Midgut volvulus	Meconium ileus	Duplication cyst
Internal hernia	Iileus due to sepsis	Hypothyroidism
Colonic atresia	Hirschsprung's disease	

INVESTIGATIONS

ANTENATAL ULTRASONOGRAM

The antenatal diagnosis of atresia with intestinal obstruction should be suspected when dilated loops of intestine and polyhydramnios are present.

RADIOGRAPHY OF ABDOMEN

Supine and prone translateral position : Distended small bowel loops and air fluid levels are seen. The level of obstruction may be determined by number of fluid levels. Single large loop of bowel and airfluid level indicate atresia than other causes of intestinal obstruction. More distal the atresia, more distension and greater the number of distended intestinal loops and air fluid levels observed. Triple bubble appearance is seen in proximal jejunal atresia. Absence of gas in the lower abdomen and pelvis is another important sign suggestive of complete obstruction.

Occasionally, intraperitonial calcification of meconium peritonitis may be seen on plain radiograph. Meconium calcification in patients with multiple intestinal atresia produces "string of pearls appearance". Intraluminal calcification (mummification) indicates antenatal volvulus. "Soap bubble appearance" is seen in meconium ileus associated with intestinal atresia. Patients with jejunal atresia have a few gas filled and fluid

filled bowel loops of small intestine but the reminder of the abdomen is gasless. Distal ileal atresias are difficult to differentiate from colonic atresia because haustral markings are rarely seen in neonates.

Prone Translateral view : is useful to distinguish between low small bowel and colonic obstructions.

Contrast Study : Contrast enema is performed to exclude colonic atresia and malrotation. The classical appearance of colon distal to jejunoileal atresia is an unused or microcolon. In ileal atresia the contrast will show a microcolon but no contrast refluxing into the dilated bowel.

TREATMENT

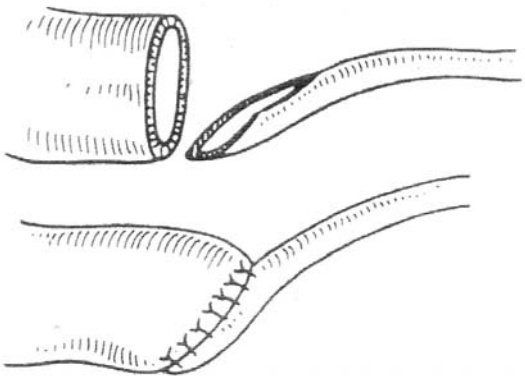
PRE OPERATIVE PREPARATION

- Attention to hypothermia, hypoxemia, hypovolemia, hypoglycaemia and hypoprothrombinemia.
- Baby should be kept under overhead warmer to avoid hypothermia.
- Gastric decompression by infant feeding tube.
- Fluid management (loss replacement followed by maintenance)
- Correction of hematological and biochemical abnormalities

- Prophylactic antibiotics.

OPERATIVE TREATMENT

Baby is taken up for surgery under General anaesthesia with meticulous attention. Abdomen opened through supra umbilical right transverse incision. Clear or yellowish peritoneal fluid is usually present and that should be sent for culture. Identify the type of atresia. Dilated proximal segment is resected and primary anastomosis is done, with distal segment by single layer inverting END-TO-BACK technique with 5-0 silk or vicryl after establishing the patency of distal bowel by injecting saline into distal loop.



End to Back Anastomosis.

Sutures must be tied loose, so that postoperative edema does not cause excessive strangulation of tissues. Proximal dilated ileum is resected to a length of 4-5 inches and the distal bowel for 2-3 inch prior to anastomosis. In high jejunal atresia, it may not be possible to excise the dilated proximal bowel.

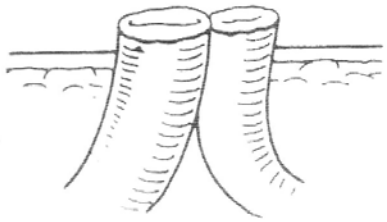
ALTERNATIVE OPTIONS

TAPERING JEJUNOPLASTY on the antimesentric side either by excision and suturing or by staples has been advocated. Since anastomotic leak is common with tapering jejunoplasty IMPBRICATION has been tried with good result. Although the imbrication technique preserves mucosal surface, it has a tendency to break down with repeated dilatation.

In distal ileal atresias, an end to side ileo ascending colon anastomosis is done. Though there is disadvantage of bypassing ileocecal valve.

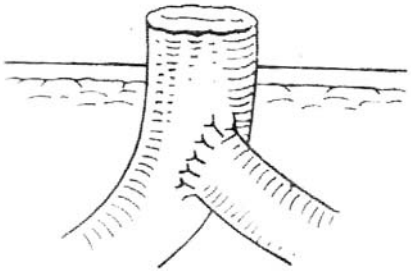
DOUBLE BARREL ILEOSTOMY is done in conditions like atresia complicated by meconium peritonitis, bacterial peritonitis, gangrene bowel, and in low birth weight infants where primary anastomosis is hazardous.

Double Barrel enterostomy later on requires limited target laparotomy to resuture the intestinal continuity by end to end anastomosis.

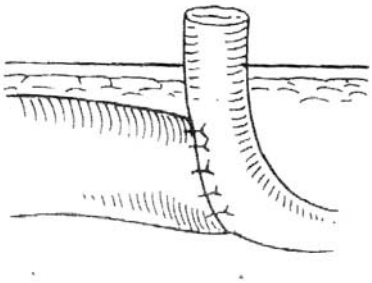


Double Barrel Ileostomy

In ileal atresia with complications exteriorization procedures such as modified mickulicz Double barrel, Bishop-Koop (distal stoma), Santulli - (proximal stoma) and Rehbein anastomotic enterostomies are also done.



Santulli (Proximal Stoma)



Bishop-Koop (Distal Stoma)

In multiple atresias, it is preferable to excise that segment and anastomose the normal bowel, if there is good length of nonatretic bowel between atretic segments. It is preferable to do multiple aanastomosis though there is inherent danger of leak at the anastomotic site. In multiple atresia with diaphragms and a short intestinal length, 17F gum elastic bougie can be used to perforate the diaphragm along the length of the bowel **(By Shish-Kabab technique).**

Extreme care to be taken while anastomosing apple peel syndrome as there is a chance of kink at the site of anastomosis. As far as possible ileocecal valve has to be retained as increased colonization of small bowel has been observed in those in whom ileocecal valve is sacrificed.

In atresia associated with Gastroschisis, the defect is repaired first followed by bowel decompression and parenteral nutrition. Reexploration and anastomosis performed after two to three weeks for the fibrous peel to resolve or primary closure of the defect with proximal enterostomy is done when there is doubt

in viability of bowel. Neonatal small bowel normally measures about 250 cm but minimum of 75 cm is necessary for achieving normal bowel function.

Small bowel transplantation is recommended as an alternative to resection anastomosis to the HMIA group of cases.

POST OPERATIVE CARE

Total or partial parenteral nutrition is necessary in post operative period as there is prolonged postopertive ileus if extensive length of small bowel has been resected. Malabsorption, diarrhoea has to be managed appropriately.

POST OPERATIVE COMPLICATIONS

EARLY COMPLICATIONS

Anastomotic malfunction and leak, lactose intolerance and malabsorption, necrotising entero colitis, TPN related complications, sepsis.

LATE COMPLICATIONS

Adhesive obstruction, motility disorder, shortgut syndrome and its related problems like rickets, osteomalacia, anaemia (megaloblastic, iron deficiency), Gallstones and Renalstones.

OUTCOME

The survival of neonate with an isolated intestinal atresia should be 100%. This success has developed dramatically in the last 40 years with advent of TPN and Neonatal ICU care.

SURVIVAL RATE

Survival rate is more with ileal atresia than jejunal atresia. Distal ileal atresia carry excellent prognosis with 100% cure rate whereas jejunal atresias, carry only 30%-60% survival rate. More than 90% of infant with > 35 cm small bowel with an intact ileocaecal valve after resection will survive. Nearly 50% survival is seen in child with 15 to 25 cm small bowel with an intact ileocaecal valve. Almost 0% survival is seen only in babies with <15 cm bowel, with intact ileocecal valve and in babies with >40cm length bowel without ileocecal valve.

MOBIRDITY & MORTALITY

Nearly 15% of death in Jejunoileal atresias are due to anastomotic leak. Increased mortality is seen in multiple atresias, type 3 B atresia, atresia associated with meconium peritonitis, low birth weight babies and babies who undergo multiple surgeries.

PROFORMA

Name: Age : Sex : IP No. :

Socio Economic Status :

Date & Time of Admission : Informant :

Date of Surgery : Reliability :

Date of Discharge : Nutrition status :

Presentation Family & Siblings:

Abdominal distention -

Not passed meconium	-
Bilious vomiting	-
Visible bowel loops	-
Visible peristalsis	-
Refusal to take feeds	-
Jaundice	-

Antenatal :

Past History :

Family History :

General Examination

Cry	-	Head / Face :
Colour	-	Eyes :
Activity	-	Mouth :
Sclerema	-	Back :
Hydration	-	Genitals :

Cardio Vascular Examination Heart Rate :

Respiratory Examination Respiratory Rate :

Abdominal :

Abdominal wall erythema	-
Addominal wall edema	-
Tenderness	-
Palpable Mass	-

Anus :

Passage of Meconium	-
Rectal Washout	-
Bleeding per rectum	-

Naso gastric aspiration :

Colour :		**Amount (ml)**
Bilious	-	
Non bilious	-	
Bloody	-	

Investigations

Blood

Haemoglobin	-	Total Count	-
Bleeding time	-	Differential count	-
Clotting time	-	Blood Group	-
Platelet	-	Rh typing	-

Serum Electrolytes	- Na / K / HCO3/ Cl2
Serum Bilirubin	-
Serum Creatinine	-
Blood Urea	-

Blood for Enteric and Non Enteric Culture

X-ray Chest and Abdomen:

AP View -

PTL View -

Barium enema -

Gastrographin Study -

Ultra sound Abdomen -

Associated Anomaly :

Provisional diagnosis : ----------

Treatment

Operative Findings:

Type of atresia -

Site of atresia -

Presence of associated anomalies -

Surgical Procedures

Primary Surgery

Additional Surgery

Conservative Management

Post Operative complication

Result : Discharged / Expired / AMA

Post operative follow up:

AIM OF THE STUDY

The aim of this study is to investigate the possible **etiological factors** of Jejunoileal atresia by evaluating the resected specimen **histopathologically**, regarding the histological structure of **intestinal muscular layer and myentric plexuses**.

MATERIALS AND METHODS

For this study, Patients with complaints of Abdominal distension, Bilious vomiting and not passed Meconium were chosen. Patients admitted to the Paediatric Surgical department at ICH & HC, Chennai, Tamil Nadu during the period January 2007 to April 2008 were included in the study.

Totally 20 No. of patients were included for the study. This study is a prospective study. Control tissues of Jejunum and ileum were obtained from 2 patients who underwent small bowel resection for intussusception.

Patients of age group less than one month and either sex were included Informed consent for the study were taken in each case. The nature of surgery is resection and end to back anastanosis. Specimen is taken 5 cm of the proximal dilated segment and 3 cm of the distal segment. Ethical

Committee clearance was obtained from the ethical committee of ICH & HC, Chennai.

A detailed history was taken from the parents and documented in the case sheets.

Exclusion Criteria:

- Duodenal atresia was excluded.
- Colonic atresia was excluded.
- Intestinal obstruction other than due to jejunoileal atresia and its surgical complications were excluded.

Inclusion criteria

- All cases of jejunal and ileal atresias.
- Cases with atresias extending from jejunum to ileum.
- The above cases along with other congenital malformations.
- Cases of jejenoileal atresia who had undergone surgery earlier and presenting with post surgical complication.

A detailed history and physical examination in every case was done.

The following investigations were conducted in every patients before subjecting him/her for surgery.

1. Complete haemogram:

    Haemoglobin%

    Bleeding time

    Clotting time

    White Blood cells - Total count and differential count.

    Packed cell volume

    Platelets count.

2. Biochemical analysis:

    Blood sugar

    Blood urea

    Serum Creatinine

    Serum Electrolytes

    Serum Bilirubin

3. Blood grouping, Rh typing and cross matching

4. Plain Xray Abdomen

5. Ultrasonogram - Abdomen and preparation of patients in case of elective surgery.

Nature of surgery - Laparotomy and Ressection of proximal and distal atretic segment and ends to back anastomosis.

Nature of specimen - Resected portion of atretic bowel with proximal segment of bowel and distal segment of bowel. The specimen is fixed in

formalin fixative to penetrate into the tissues deeply and is preserved for atleast 12 hours for complete fixation.

After fixation specimen (intestine) opened and pinned to the Board. Bits were taken from the atretic segment, from 2 cm and 4 cm proximal to the atretic segment and 1 cm and 2 cm distal to the atretic segment, put in the capsule for processing in different grades of alcohol, then brought to paraffin, embedded in the paraffin block. Sections of 4-5 microns thickness were made and are floated on the 45ºC -50ºC water bath. The sections are then floated onto the slides, then the slides are placed in 62ºC oven or a slide dryer for 30 minutes before staining. All the slides are continuously stained with haemotoxylin and eosin (H&E). Special stains were done like masson, Trichnome stain and the slides were viewed for interpretation. Evidence of ischemic injury ulceration at the site of atresia, of fibrosis and presence of and number of ganglion cells were also noted.

Both mesentric and antimesentric sides of the intestine were examined for ischemic injury, fibrosis etc.

With special reference to mucosa and muscular layer completeness of muscle layer defect in the muscle layer, fibrosis of the muscular layer were looked for.

Also the no. of ganglion cells, morphology of ganglion cells at the atretic site, at 2 cms and 4 cms proximal segments and 1 cm distal segments were studied.

Hematoxylin and Eosin Stain Method:

1. Dewax sections hydrated through graded alcohol and water.
2. Remove fixation pigments if necessary
3. Stain in an alum hematoxylin for 10 minutes until sectious blue.
4. Wash well in tap water for 5 minutes or less.
5. Differentiate in 1% acid alcohol for 5-10 sec.
6. Wash well in tap water until sections are again blue for 5 minutes or less.
7. Stain in 1% Eosin for 10 minutes.
8. Wash in running tap water for 1 to 5 minutes.
9. Dehydrate through alcohols, clean and mount the slide with DPX.

RESULTS:

Nuclei	-	Blue/Black
Cytoplasm	-	Varying shades of pink
Muscle fibres	-	Deep pink/red
R.B.C.	-	Orange/Red
Fibrin	-	Deep pink.

MASSON TRICROME TECHNIQUE (Masson 1929)

Fixation:

Formal sublimate or formal saline.

Stains:

Solution A

Acid fuchsin 0.5 gm.

Glaicial acetic acid 0.5 ml

Distilled water 100 ml.

Solution B

Phosphomolybdic acid 1-0 gm

Distilled water

Solution C

Methyl blue - 2.0 gm
Glacial acetic acid 2.5 ml
Distilled water 100 ml

Method:

1. Dewax sections and bring to water
2. Remove mercury pigment by iodine thiosulphate sequence.
3. Wash in tap water
4. Stain nuclei by celestin blue haemalum method

5. Differentiate with 1% acid alcohols
6. Wash well in tap water
7. Stain in acid fuchsin in solution A for 5 minutes
8. Rinse in distilled water
9. Treat with phosphomolybdic acid Solution B for 5 minutes
10. Drain
11. Stain with methyl blue solution C for 2-5 minutes
12. Rinse in distilled water
13. Treat with 1% acetic acid for 2 minutes
14. Dehydrate through alcohol.
15. Clear in Xylene, Mount in DPX

Result	Nuclei	-	Blue back
	Cytoplasm	-	Red
	Muscle & erythromytes	-	Red
	Collagen	-	Blue

RESULTS

The histologic changes related to small bowel atresia were found to be as follows:

At the atretic segment the intestinal lumen was calcified and there was deficient and ill formed muscular layers. Ganglions cells were absent.

At 2 cm proximal to the atresia, the muscle layers circular and longitudinal were ill formed and fibrosis present. In some of the cases and in

some there was total replacement muscular layer by fibrosis. Ganglion cells were present. They were normal in number and normal in morphology in all the cases.

At 4 cm proximal to the atresia, the muscle layers were well formed in all the cases. Normal ganglion cells were present which were normal in number also.

At distal segment, 1 cm distal to the atresia the muscle layers were thinner than those of the Control groups. Ganglion cells were normal.

DISCUSSION

This study revealed that in neonates with small bowel atresia the musculature of the bowel wall of the proximal dilated segment are abnormal and ganglions are present. Therefore adequate resection seems to be mandatory for the prevention of post operative dysmotility.

In utero vascular insults to the intestine have been shown to cause small bowel atresia in many cases[1]. Many factors have been implicated in the pathogenesis of the muscle coat defects. Tibboel et al[28] studied the effects of temporary local ischemia and general hypoxia on the development of intestines of chick embryos. In experimental local temporary ischemia, chick

embryo suffered intestinal atresia. The intestinal loops proximal to the obstruction were dilated and the intestinal wall was extremely thin. The structure of both layers of the musculature become abnormal, whereas the intestinal wall was normal distal to the obstruction[28] . This study showed that temporary disruption of the circulation in the mesentric blood vessels during the fetal period lead to atresia with structural abnormalities in the muscle coats. Therefore, a casual relationship between, on one hand, vascular accidents in utero on the other, atresia and the absence of muscle coats appears likely.[21]

Tepas et al[30] experimentally produced intestinal atresia in fetal lamps by an intrauterine disruption of the mesentric blood supply and showed that the dilatation of the proximal segment induces the involution and lysis of the ganglion cells after an initial hyperplasia of myenteric ganglia, as the irreversible distension continues. In our study we have observed the presence of segmental musculature defect but the ganglion cells were normal in number and morphology. The abnormalities observed in this study correlate with vascular theory of small bowel atresia, and we agree with the hypothesis that the intestinal muscle defect may be a secondary event to the vascular injury rather than a failure in muscle development.[24]

Hamdy et al[23] reported that in the dilated bowel at 2 cm proximal to the atresia, the intermuscular ganglion were smaller and less number and muscle layers were thinner on the antimesentric side when compared to those on the mesentric side and in control specimens. In our study, the ganglion cells were normal both on the mesentric and antimesentric sides at 2 cm proximal to the atresia. Muscle layers were irregular with segmental muscular hypertrophy.

We have observed that intestinal lumen was filled with calcified material at the atretic segment in few cases. Remnants of the epithelial lining, especially under the effect of bilious component, as well as ischemic changes with high collagen levels of the remnants of the muscle layers do result in calcified changes.

The absence of musculature in any portion of the intestinal tract leads to localised dilatation of the affected segment with obstruction caused by lack of the intestinal peristalsis[31]. When the dilated proximal intestine, which have segmental musculature defect was not adequately resected, the dilatation of this intestinal segment and stasis have observed.

Previous studies have documented that, all histopathologic abnormalities were seen in the antimesentric border of the dilated proximal atretic segment.

Tapering enteroplasty for the management of this dilated segment has been advocated. On the contrary, our observations revealed that histopathologic abnormalities were seen in both the antimesentric side and mesentric side. Therefore insufficient resection and tapering enteroplasty will not prevent the intestinal dysmotility. Besides this, there may be some complications such as anastomotic leakage or obstruction related to the tapering enteroplasty. Muscular defect also present at the proximal end of the distal atretic bowel and it requires resection. We believe it is possible to prevent intestinal dysmotility seen in the postoperative period by doing resection of the dilated segment and distal segment and doing end to back anastomosis.

CONCLUSION

In our studies the proximal segment of atretic intestine showed **structural deficits.**

Ganglion cells were normal.

Defect in the intestinal musculature were prominent but intestinal mucosa was intact.

These abnormalities were seen **both on the antimesentric side and on the mesentric side,** which support **vascular accident** as a causative factor.

When possible **adequate resection** rather than tapering enteroplasty should accompany the repair of intestinal atresia to eliminate the intestinal segment with structural defects.

When this is not feasible sufficient tapering is preferred.

Muscular defect also present at the proximal end of distal atretic bowel and it **requires resection** at the time of surgical correction of atresia.

Adynamic intestinal segment owing to **insufficient resection** may lead to prolonged intestinal dysmotility in the post operative period, which may result in sepsis and death.

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