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MALROTATION IN NEONATES AND CHILDREN: OUTCOME AND EXPERIENCE AT A TERTIARY CARE HOSPITAL

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ABSTRACT

Aim: To study the clinical features, Investigations and Surgical Outcome of Neonates and Children with Intestinal Malrotation.

Materials and Methods: This is a Retrospective study of patients with Intestinal Malrotation admitted and treated from January 2015 to December 2022 (8years). The data pertaining to the age, sex, investigations, associated complications, management, outcome was taken and reviewed.

Results: A total of 38 patients underwent surgery for Malrotation of which 26 were males and 12 were females. Of these 38 patients 30 were neonates and 8 were children.The most common symptom was bilious vomitting in neonates and recurrent pain abdomen in children. Most common investigation done to diagnose intestinal malrotation was upper gastrointestinal contrast study along with Ultrasound abdomen. Of these 38 patients associated volvulus was noted in 29 patients. Of the 38 patients two neonates expired with a mortality rate of 5.3%.

Conclusion: High degree of suspicion is required to diagnose intestinal malrotation in children. In all neonates with bilious vomittings intestinal malrotation must be ruled out. Ultrasound abdomen with colour doppler and upper gastrointestinal contrast study are useful routine diagnostic tools. As associated volvulus with vascular compromise can be detrimental, early diagnosis and prompt management significantly reduces the morbidity and mortality.

Keywords: Malrotaion, Volvulus, Outcome, Ladd's Procedure

INTRODUCTION

Intestinal malrotation is a congenital anomaly resulting from abnormal or incomplete rotation and fixation of the midgut during embryogenesis. It comprises a spectrum of anomalies ranging from nonrotation to reverse rotation. Malrotation was defined as "typical" if the duodenojejunal junction was to the right of the midline and "atypical" if it was on the left side but not ascended to the level of the pylorus. The disease occurs in approximately 1 in 500 births.[1] Up to 75% of patients present in the 1st month of life.[2],[3] The actual incidence of malrotation is difficult to estimate in the older population as most

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people remain asymptomatic for life or present with intermittent colic abdominal pain, chronic vomiting, diarrhea, hematemesis, constipation, and some are recognized during other intraoperative procedures or at autopsy.[4] Sudden onset bilious vomiting is a cardinal sign of intestinal obstruction in neonates, and malrotation with complete midgut volvulus should be a suspected diagnosis until proven otherwise. The presentation can vary from life threatening midgut volvulus to non-specific symptoms like underdevelopment, gastroesophageal reflux, early satiety, and mild abdominal discomfort. Diagnosis is even more difficult in older children, as symptoms are often vague and may appear unrelated to the abdomen[4] due to partial midgut volvulus leading to the obstruction of mesenteric veins and lymphatics and subsequent impaired nutrition support. Recently, Nehra and Goldstein emphasized the need to increase awareness of this entity and its various manifestations at different ages to reduce the time to diagnosis and lead to better outcomes. However, malrotation and associated mortality have been reported at all ages.[4] The purpose of this study is to highlight the clinical features, investigations, treatment outcomes in patients with intestinal malrotation.

MATERIALS AND METHODS

Study Design: Retrospective Study

Study location: Department of Paediatric Surgery, Kurnool Medical College and Government General Hospital, Kurnool, A.P
Study Duration: January 2015 to December 2022
Sample Size: 38 patients
We retrospectively studied the cases of 38 patients who were found to have intestinal malrotation at our Institute from January 2015 to December 2022. All the patients who underwent surgery for Intestinal Malrotation were included in the study and were reviewed for age, sex, investigations, Intraoperative findings, complications, management, outcome was taken and reviewed.

USG with colour doppler and UGI Contrast study were used for confirmation of diagnosis. Once the decision for primary operative treatment was made, the diagnosis of malrotation was confirmed at surgical exploration. Intraoperative diagnosis was confirmed if Ladd's bands were noted to extend across the duodenum and/or the intestinal mesentery was found to be narrowed. Any associated volvulus was noted. All patients found to have malrotation on exploration were treated with Ladd's procedure.
The steps of operative procedure consisted of
1.If volvulus was present derotation(Counter clockwise) was done.
2.Division of obstructing Ladd's bands overlying the secondpart of duodenum.
3.Widening of the duodenocolic isthmus.
4.Straitening of the duodenum.
5.Appendicectomy
6.Bowel Resection if gangrene was present ± ostomy

RESULTS

Out of the 38 patients, 26 were males and 12 were females.(Figure 1) Among the 38patients 30 presented in neonatal age and 8 presented in childhood. Majority of the neonates presented in the first week of birth (22) and the remaining 8 neonates presented after first week of life and before one month of age. The remaining 8 patients presented between one year to 12 years of age. (Figure 2).

The most frequently observed symptoms of malrotation in neonates was Bilious vomiting. One neonate presented with bloody stools and haemetemesis along with history of bilious

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vomiting. In children the most common presenting symptoms were recurrent non specific abdominal pain ± vomiting. (Table 1).
The preoperative diagnostic eval uation included ultrasound abdomen ± doppler study (Figure 3) and upper gastrointestinal study (Figure 4). The median age at surgery was 10.2 days (3 days-12 years). Intraoperatively only malrotation with ladds bands was present in 12 patients(31.6%), Malrotation with ladds bands with volvulus was noted in 24 patients(63.1%) and Malrotation with ladds bands with volvulus with gangrene of small bowel was seen in 2 patient(5.3%) (Table 2).

Operative procedures included derotation of volvulus, correction of malrotation, resection and ostomy in one neonate (Table 3). The most common postoperative complications noted were sepsis (neonates), wound infection ,adhesive obstruction and others as noted (Table 4). None of the patients had recurrent volvulus. Two neonates died. One with intestinal gangrene and sepsis, and the other neonate with pneumonia and sepsis. One child with enterotomy, reversal was done after 3 months and the child did well. The length of hospital stay was 8–27 days. The mean followup was 2 years ranging from 6months to 4years. Majority the survived patients did well during followup. One patient underwent repair of Incisional hernia after 2years.

Table 1: CLINICAL FEATURES

S.No	Symptoms	No(%)
1	Bilious vomitting	34(89.4%)
2	Recurrent pain abdomen	8(21.1%)
3	Failure to thrive	3(7.9%)
4	Intestinal obstruction with constipation	2(5.3%)
5	Abdominal distension	2(5.3%)
6	Blood in stools with Hemetemesis	1(2.6%)

Table 2: OPERATIVE FINDINGS

S.No	Introperative Findings	No(%)
1	Malrotation with Ladds bands	12(31.6%)
2	Malrotation with Ladds bands with volvulus	24(63.1%)
3	Malrotation with Ladds bands with volvulus with gangrene of bowel	3(5.3%)

Table 3: OPERATIVE PROCEDURE

S.No	Procedure	No(%)
1	Ladds procedure	12(31.6%)
2	Derotation of volvulus + Ladds procedure	25(65.8%)
3	Derotation of volvulus + Ladds procedure + resection and Ostomy	1(2.6%)

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Table 4: POSTOPERATIVE COMPLICATIONS

S.No	SYMPTOMS	No(%)
1	Sepsis	18(47.4%)
2	Wound infection	10(26.3%)
3	Postop adhesive Intestinal Obstruction	2(5.3%)
4	Pneumonia	2(5.3%)
5	Incisional hernia	1(2.6%)

Figure 1: SEX INCIDENCE

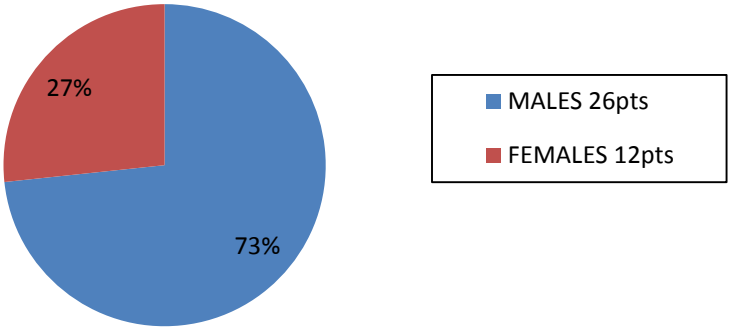

FIGURE 2: AGE OF PRESENTATION

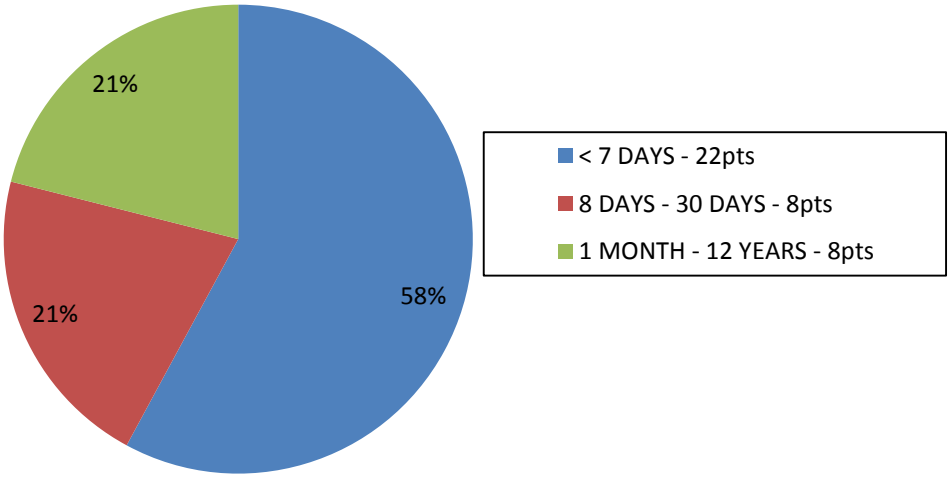

Figure 1: Ultrasound with Colour Doppler showing WHIRLPOOL SIGN in Malrotation with Volvulus

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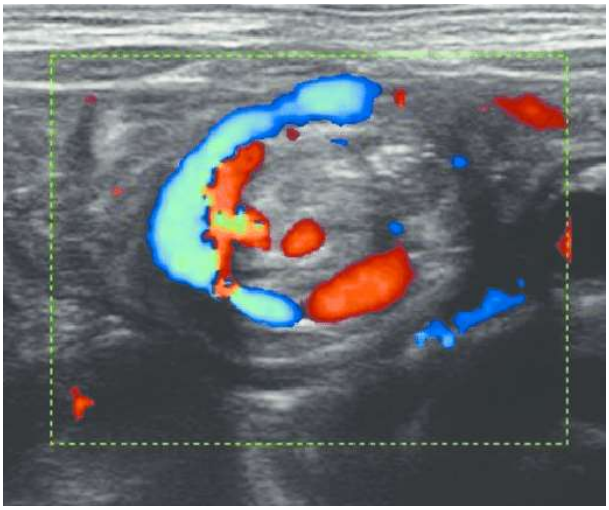

Figure 2: Upper GI Contrast Study

A) UGI Contrast study in Malrotation DuodenoJejunal junction to right of spine

A) UGI Contrast study in Malrotation with volvulus (Cork Screw Sign)

DISCUSSION

Intestinal malrotation is a common neonatal and pediatric surgical disesase with an incidence of 1 in 500 live births [5]. Up to 40% of patients with malrotation present within the first week of life and up to 75% to 85% have been diagnosed by 1 month [6]. In our series 57.9%(22) of patients presented during first week of life and 79%(30) within the neonatal period.

Neonates presented with bilious emesis or other clinical and radiographic evidence of high small-bowel obstruction, which was evident in almost all of patients in our series. Beyond the neonatal period, malrotation may present with bilious vomiting and bowel obstruction more commonly presents with abdominal pain, diarrhea, vomiting, and failure to thrive [7,8]. In our series the most common presenting symptoms in children were abdominal pain and vomitting. The vague nature of these symptoms calls for a high level of clinical suspicion of intestinal malrotation [9,10].

Conventional plain X-ray abdomen is neither sensitive nor specific for the diagnosis of malrotation. It was normal in the majority of cases. Abdominal color doppler may divulge malposition of the SMA, raising the suspicion of gut malrotation [11]. Pacros et al first

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described the 'whirlpool' sign [12]. It includes duodenal dilatation with distal tapering and a fixed midline bowel and mesentery twisted around the SMA axis. The gold standard for the diagnosis of gut malrotation is an upper gastrointestinal contrast study [9]. It demonstrates duodenum and duodenojejunal flexure located to the right of spine.In this series USG colour Doppler diagnosed 63.1%(24), UGI contrast studies diagnosed 92.1%(35) of the patients.

All the patients underwent Standard Ladds Procedure with Appendicectomy. At operation varying degree of volvulus was noted in 68.4%(26)patients. One patient had volvulus with a segment of unviable small bowel so resection and ostomy was performed. One patient had volvulus with extensive gangrene of small bowel for which ladds procedure was done and this patient expired in the postoperative period.

Sepsis was the most common postoperative complication noted follwed by wound infection. The Ladd's procedure is a known risk factor for postoperative adhesion following laparotomy . The raw area after widening predisposes for adhesion. Stauffer et al. reported that 12% of required surgical intervention for bowel obstruction during follow-up[13]. In this study, 5.2% (2) of children developed bowel obstruction. One child was treated conservatively and did well while one patient required surgery(adhesiolysis) [14,15].

Recurrent midgut volvulus is uncommon and has been reported in up to 2-7% of children [16]. In this series, we did not encounter recurrent volvulus. Volvulus is the most feared complication, seen in 60-70 % of neonates with malrotation. Delay in diagnosis may cause strangulation in 15% of cases which leads to intestinal ischemia, intestinal necrosis, septicemia, and short bowel syndrome . We encountered 26 patients(68.4%) with volvulus of which 2pt (7.7%) were strangulated and gangrenous.

The most serious consequence of midgut volvulus is death or loss of significant length of the intestinal tract resulting to a TPN-dependence for life. Bowel necrosis is a major challenge in developing countries where total parenteral nutrition is out of reach and bowel transplantation is not an option [15,16]. If the ischemic bowel is discovered during a laparotomy for volvulus, every effort should be made to preserve bowel length, and if viability is in doubt, a second-look laparotomy should be performed 12-24 hours later without initial resection or with a very conservative resection. In this series one patient had extensive gangrene caused by midgut volvulus and succumbed with overwhelming sepsis.

CONCLUSION

Neonates uniformly presented with bilious emesis and had radiological evidence of malrotation (Doppler USG/UGI contrast studies). The diagnosis of malrotation in children is difficult and a high level of clinical suspicion is required to diagnose due to the vague nature of the symptoms. An ultrasound with colour doppler and UGI Contrast studies can clinch the diagnosis in almost all the cases, These investigations should be used as a routine screening methods. Malrotation with midgut volvulus and intestinal ischemia is a serious complication and is associated with high mortality and morbidity. High level of clinical suspicion directed to early diagnosis and prompt treatment leads to high success rates.

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Conflicts of Interest

There are no conflicts of interest

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