Original Research Article

A study on clinical profile and outcome of malrotation of intestine in children

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ABSTRACT

Background: Malrotation is a congenital disorder featured with abnormal positioning of intestine within the peritoneal cavity. Its diagnosis is established within one year of age in most of the cases. The present study was conducted with the aim to analyse in detail the entire clinical profile of malrotation of intestine in children and to analyse the value of imaging in diagnosing suspected cases of malrotation.

Methods: This prospective study was carried out during the period from September 2010 to January 2013. 70 patients with suspected malrotation based on clinical symptoms and subsequently proven by surgery were included in the study. The surgery was performed based on the clinical signs and symptoms as well as ultrasound and an upper gastrointestinal contrast study by using barium meal follow through (BMFT).

Results: Out of 70 patients in this series, 47 (67.5%) were at the age group of less than one week to one month, 10 (14%) were between one month to one year and 13 (18.5%) were older than year. Male: Female ratio was 4:3. 12 (17%) babies were born to consanguity couple. Bilious vomiting was the most common symptom observed in 61 (87%) patients. After X-ray of abdomen, intussusception was diagnosed in 5 patients, X-ray was suggestive in 35 patients (50%), normal in 16 (23%), double bubble sign noticed in 5 (7%) patients. Out of 70, 51 patients had ultrasound abdomen for malrotation. Among them, 36 patients were noticed with altered SMA/SMV axis, 6 with volvulus, 4 reports were normal and 5 were inconclusive.

Conclusions: The incidence of malrotation in symptomatic children can be identified by ultrasound of abdomen whereas in case of asymptomatic children, it was diagnosed initially by using abdominal X-ray and later confirmed by abdominal ultrasound. Once the diagnosis of malrotation has been established, appropriate surgical approach should be carried out to reduce the risk of mortality in infants and children.

Keywords: Malrotation, Ultrasound of abdomen, X-ray of abdomen

INTRODUCTION

Malrotation is a congenital disorder featured with abnormal positioning of intestine within the peritoneal cavity.1 It is accompanied by abnormal bowel fixation by mesenteric bands or absence of fixation of portions of the bowel, leading to intensified risks of bowel obstruction, acute or chronic volvulus, and bowel necrosis in neonates, infants and older children.

Malrotation is seen approximately one in 500 births.2 It is diagnosed in about 75% of symptomatic cases in newborns and 90% cases in children within first year of life. The most common symptom of malrotation is bilious
vomiting with or without abdominal distention associated with either midgut volvulus or duodenal obstructive bands. Delay in diagnosis and treatment may result in small-bowel necrosis, dependence on total parenteral nutrition and short-gut syndrome. Mortality in affected children in the 1950s and 1960s was approximately 30% but it has markedly decreased, to 3%-5%.

The diagnosis is straight forward in an infant presenting with sudden onset of bilious vomiting i.e. by typical upper gastrointestinal contrast study. Ultrasound of abdomen was done to look for the orientation of mesenteric vessels by an experienced pediatric radiologist and it was an effective screening study in suspicious patients.

The dilemma comes in an asymptomatic patient who was incidentally suspected for malrotation on imaging. Although no clear guidelines exist for managing such asymptomatic patients, they warrant complete evaluation to rule out a narrow mesenteric pedicle that predisposes the patient to the dangerous midgut volvulus.

Role of laparoscopy in managing classical malrotation and its value as a diagnostic modality in equivocal cases has also been discussed in recent literature. The aim of this study was to analyse in detail the entire clinical profile of malrotation of intestine in children admitted in our institute and the value of imaging in diagnosing suspected cases of malrotation.

METHODS

This prospective study was conducted at Institute of child health and hospital for children, Egmore, Chennai, Tamilnadu, india during the period September 2010 to January 2013. 70 patients with suspected malrotation based on clinical symptoms and subsequently proven by surgery were included in the study.

Inclusion criteria

In the present study were neonates, infants and older children with laparotomy proven malrotation within this study period.

Exclusion criteria

In the present study were patients with congenital diaphragmatic hernia, evagination of diaphragm, gastroscisis and omphalocele.

In the present study, malrotation was screened by using ultrasound of abdomen and by radiological contrast study by using barium in symptomatic children. In asymptomatic patients it was accidently diagnosed by X-ray of abdomen. On basis of observations made by the above procedures, appropriate surgical approach was indicated in patients. Follow up was made for every three months till the end of the study.

RESULTS

The study included 70 children after meeting the requirements of selection criteria. Of these patients, 47 (67.5%) were at the age group of less than one week to one month, 10 (14%) were between one month to one year and 13 (18.5%) were older than year. Male: Female ratio was 4:3. Out of 47 neonates, 10 (21%) were preterm babies and 37 (79%) were term babies. Out of 37 term babies 23 (62.2%) were born with weight >2.5 kg, 11 (29.7%) with weight of 2-2.5 kg and 3 (8.1%) with <2 kg. Out of 10 preterm babies, 9 (90%) were born with weight >1.5 kg and one (10%) with <1.5 kg. Out of 70, 12 (17%) babies were born to consanguinity couple. Malrotation was seen majorly in cases of first (43%) and second (46%) born child.

Table 1: Patients characteristics.

| Characteristics                           | No. of patients | Percentage |
|-------------------------------------------|-----------------|------------|
| **Age**                                   |                 |            |
| <1 week                                   | 29              | 41.5       |
| 1 week- 1 month                           | 18              | 26         |
| 1 month- 1 year                           | 10              | 14         |
| >1 year                                   | 13              | 18.5       |
| **Sex**                                   |                 |            |
| Male                                      | 40              | 57         |
| Female                                    | 30              | 43         |
| **Gestational age in neonates**           |                 |            |
| Preterm babies                            | 10              | 21         |
| Term babies                               | 37              | 79         |
| **Birthweight in preterm babies**         |                 |            |
| >1.5 kg                                   | 9               | 90         |
| <1.5 kg                                   | 1               | 10         |
| **Birthweight in term babies**            |                 |            |
| >2.5 kg                                   | 23              | 62.2       |
| 2.25 kg                                   | 11              | 29.7       |
| <2 kg                                     | 3               | 8.1        |
| **Consanguinity**                         |                 |            |
| Non                                       | 58              | 83         |
| II degree                                 | 2               | 3          |
| III degree                                | 10              | 14         |
| **Order of birth**                        |                 |            |
| 1st born child                            | 30              | 43         |
| 2nd born child                            | 32              | 46         |
| 3rd born child                            | 8               | 11         |

Bilious vomiting was the most common symptom observed in 61 (87%) patients. 5 were presented primarily with symptoms of intussusceptions and subsequently diagnosed to have malrotation during surgery.

Duration of symptoms of malrotation in children was presented in Table 3. Among the 47 neonates, 22 of them presented onset of symptoms for one day and 34 patients within 3 days of onset. Symptom duration ranged from 1
day to more than 2 weeks for children of age group one month to one year and older.

Clinical presentations diagnosed by using X-ray of abdomen in detail were summarized in Table 4. Out of the 70 patients, intussusception was diagnosed in 5 patients. Excluding these 5 patients in the rest of the 65 patients, X-ray abdomen was suggestive in 35 patients (50%), normal in 16 (23%), double bubble sign noticed in 5 (7%) patients. All 5 of them were taken directly for laparotomy without further investigations.

Table 2: Symptoms of malrotation.

| Symptoms                                | No. of patients | Percentage (%) |
|-----------------------------------------|-----------------|----------------|
| Bilious vomiting                        | 61              | 87%            |
| Asymptomatic                            | 2               | 2.8%           |
| Failure to thrive                       | 1               | 1.4%           |
| Intussusception                         | 5               | 7.1%           |
| Abdominal distension                    | 3               | 4.2%           |

Table 3: Symptoms of malrotation in children of different age groups.

| Findings                                      | No. of patients | Percentage (%) |
|-----------------------------------------------|-----------------|----------------|
| Neonates N=47                                 |                 |                |
| 1 day                                         | 22              |                |
| 2 days                                        | 8               |                |
| 3 days                                        | 4               |                |
| 4 days-1 week                                 | 7               |                |
| >1 week                                       | 6               |                |
| Infants N = 10                                |                 |                |
| 1 day                                         | 2               |                |
| 2 days                                        | 4               |                |
| 3 days                                        | 2               |                |
| Asymptomatic                                  | 1               |                |
| >1 week                                       | 1               |                |
| >1 year N = 13                                |                 |                |
| 1 day                                         | 3               |                |
| 2 days                                        | 3               |                |
| 3 days-1 week                                 | 3               |                |

Table 4: Clinical presentations diagnosed by using X-ray of abdomen.

| Findings                                      | No. of patients | Percentage (%) |
|-----------------------------------------------|-----------------|----------------|
| Suggestive (n=35)                             |                 |                |
| a. Malrotation with midgut volvulus without gangrene | 25              | 71%            |
| b. Volvulus with gangrene                     | 5               | 14%            |
| c. Malrotation without volvulus               | 5               | 14%            |
| Normal (n=16)                                 |                 |                |
| a. Malrotation without volvulus               | 7               | 44%            |
| b. Malrotation with midgut volvulus           | 8               | 50%            |
| c. Malrotation variant                        | 1               | 6%             |
| Double bubble sign (n=5)                      |                 |                |
| a. Malrotation with volvulus                  | 2               | 40%            |
| b. Malrotation with annular pancreas and intrinsic duodenal obstruction | 2 | 40% |
| c. Malrotation with preduodenal portal vein and duodenal atresia | 1 | 20% |

Table 5: Clinical findings noted with ultrasound of the abdomen.

| Findings                                      | No. of patients (n=51) | Percentage (%) |
|-----------------------------------------------|------------------------|----------------|
| Altered SMA/SMV axis                         | 36                     | 70%            |
| Volvulus                                      | 6                      | 12%            |
| Normal                                        | 4                      | 8%             |
| Inconclusive                                  | 5                      | 10%            |

Table 5 presents the clinical findings of malrotation in children after abdominal ultrasound. Out of 70 patients, 14 patients underwent surgery without any further investigations. 5 of them were primarily admitted for intussusception. 51 patients had ultrasound abdomen for malrotation. Among them, 36 patients were noticed with altered SMA/SMV axis, 6 with volvulus, 4 reports were normal and 5 were inconclusive.

DISCUSSION

Malrotation of the intestine occur when the normal rotational and fixation process was altered at 12th week of gestation after return into the peritoneal cavity. The exact incidence was not known still now. Usually, male preponderance was observed in neonatal presentations at a male: female ratio as 2:1. Similar observations was noticed in our study in which male: female ratio was 4:3.

The common clinical symptoms in newborn infants are intestinal obstruction findings, such as bilious vomiting. In the present study, it was the major symptom observed in 61 (87%) patients. Other symptoms observed in our
series are abdominal distension in 3 children, failure to thrive in one child. Two children had not shown any symptoms. They were diagnosed with malrotation accidentally.

In present study, 10 babies were born before gestational age and 37 were after completion of gestational age. Interference of order of birth in incidence of malrotation was not observed in our study. In our study, majority of the children (83%) with malrotation do not have genetic susceptibility. This was in accordance with Strouse.3

Radiographs are often the first step in the imaging evaluation of pediatric patients with suspected malrotation and are concerned to explore the presence of double bubble sign.12 In the present study, X-ray findings were suggestive in 35 (50%) children, normal in 16 (23%) and double bubble sign was noticed in 5 (7%) children. But in new born, they cannot rule out the incidence of malrotation. In such cases ultrasound of abdomen will be more advantageous. It helps in determining the position of the superior mesenteric vessels and the relationship to the third portion of the duodenum.13 In our series, on ultrasound of abdomen 36 patients were diagnosed with altered SMA/SMV axis, 6 with volvulus, 4 with normal morphology and 5 reports were inconclusive.

CONCLUSION

The incidence of intestinal malrotation was certainly recognized in infants by bilious vomiting in their early age. This can be confirmed by using ultrasound of abdomen. But in asymptomatic cases it was accidently diagnosed by abdominal radiographs and later they are confirmed by the ultrasound of abdomen. Once the diagnosis of malrotation has been established, appropriate surgical approach should be carried out to reduce the risk of mortality in infants and children.

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