Recent Modalities in Management of Intestinal Malrotation

Essay
Submitted for partial fulfillment of Master Degree in general surgery

By
Ahmed Mohie El-Dien Ahmed
M.B.B.Ch, Sohag University

Supervised by
Professor Dr. Ayman Ahmed Al-Baghdady
Professor of Pediatric Surgery
Faculty of Medicine
Ain Shams University

Dr. Hesham Mohamed Abd El-Kader
Assistant professor of Pediatric Surgery
Faculty of Medicine
Ain Shams University

Dr. Amro Abd Alhamed Zaki
Assistant professor of Pediatric Surgery
Faculty of Medicine
Ain Shams University

2015

ACKNOWLEDGMENT

First, I would like to praise and thank **Allah,** the most merciful and beneficial for his help to complete this work.

I am greatly indebted to **Professor Dr. Ayman Ahmed Al-Baghdady, Dr. Hesham Mohamed Abd El-Kader and Dr. Amro Abd Alhamed Zaki** my supervisors, for the guidance and mentorship they rendered to me during the preparation and accomplishment of this dissertation.

I would like to acknowledge all members of the **Pediatric Surgery Department at Ain Shams University** for their assistance and encouragement during all stages of preparation and accomplishment of this dissertation. I would like to thank all Consultants, Specialists, Residents, Registrars and Nurses.

TABLE OF CONTENTS

LIST OF FIGURES

LIST OF ABBREVIATIONS

ACD/MPV	Alveolar Capillary Dysplasia With Misalignment Of Pulmonary Veins
Aptt	Activated Partial Thromboplastin Time
CBC	Complete Blood Count
CT	Computed Tomography
DJJ	Duodenal-Jejunal Junction
IV	Intravenously
MMIH	Megacystis, Microcolon and Intestinal Hypoperistalsis
MRI	Magnetic Resonance Imaging
NG	Nasogastric
NPO	Nothing By Mouth
PICU	Pediatric Intensive Care Unit
PT	Prothrombin Time
SMA	Superior Mesenteric Artery
SMV	Superior Mesenteric Vein
TPN	Total Parenteral Nutrition
UGI	Upper Gastrointestinal Series
US	Ultrasound

INTRODUCTION

Malrotation is the common name for a variety of abnormalities of intestinal rotation and fixation, ranging from a mobile cecum, with the duodenojejunal junction to the right of the spine, to a complete non-rotation with an associated midgut volvulus. Many theories have been put forward regarding its embryological evolution **(Fiets and Vos, 1997).**

Many authors define intestinal malrotation as intestinal non-rotation or incomplete rotation around the superior mesenteric artery. It involves anomalies of intestinal fixation as well. Interruption of typical intestinal rotation and fixation during fetal development can occur at a wide range of locations; this leads to various acute and chronic presentations of disease. The most common type found in pediatric patients is incomplete rotation predisposing to midgut volvulus, which can result in short-bowel syndrome or even death **(Nehra *et al.*, 2007).**

Intestinal malrotation is a congenital anomaly that results from abnormal or incomplete rotation of the midgut during embryonic development. During the second month of embryonic development, the gut undergoes rapid elongation, exceeding the capacity of the abdominal cavity, and herniating out into the extracoelomic umbilical cord. The intestines then return to the abdomen during the third month of development. While returning to the abdominal

cavity, the gut undergoes a counter-clockwise rotation about the axis of the superior mesenteric artery **(John *et al.*, 2002).**

Malrotation was reported prior to the 1900s. During the 20th century, understanding of the embryology and anatomy of malrotation became more complete, along with changes in surgical approaches to the problems. In 1936, William E. Ladd wrote the classic article on treatment of malrotation, and his surgical approach (i.e. Ladd procedure) remains the cornerstone of practice today. Intestinal malrotation occurs at a rate of 1 in 500 live births. Most infants with gastroschisis, omphalocele, or congenital diaphragmatic hernia present with intestinal malrotation. Approximately 50% of patients with duodenal atresia and 33% of patients with jejunoileal atresia have a malrotation as well. In addition, intestinal malrotation occurs in association with Hirschsprung disease, gastroesophageal reflux, intussusception, persistent cloaca, anorectal malformations (imperforate anus), and extrahepatic anomalies **(Nehra *et al.*, 2007).**

Male predominance is observed in neonatal presentations at a male-to-female ratio of 2:1. No sexual predilection is observed in patients older than 1 year. As many as 40% of patients with malrotation present within the first week of life. This condition is diagnosed in 50% of patients by age 1 month and is diagnosed in 75% by age 1 year. The remaining 25% of patients present after age 1 year and into late adulthood; many are recognized intraoperatively during other procedures or at autopsy **(Marcene *et al.*, 2007).**

Younger patients have higher rates of morbidity and mortality. In infants, the mortality rate ranges from 2-24%. The presence of necrotic bowel at surgery increases the mortality rate by 25 times for infants, and the presence of other anomalies increases the risk by 22 times. A report of 25 years' experience demonstrated congenital cardiovascular disease in 27.1% of patients with intestinal malrotation; those patients had a morbidity rate of 61.1% after intestinal malrotation surgery **(Nehra *et al.*, 2007).**

"Malrotation" however, consists of a spectrum of abnormalities of intestinal position and fixation ranging from normal to typical malrotation to complete non-rotation and all variations between Several names have been given to these anatomic variations, including "incomplete rotation" "mixed rotation," "atypical malrotation," and "malrotation variant". Although most cases of malrotation are identified during infancy, atypical symptoms may result in a delay in diagnosis until later childhood or even into adulthood **(Marcene *et al.*, 2007).**

Patients who are referred or evaluated for malrotation undergo an upper gastrointestinal contrast study so this was chosen as a base for the classifications. Because of the variable position of the cecum, sometimes being located normally in the right lower quadrant, upper contrast studies are preferred over contrast enemas as the initial study. Some children also undergo contrast enema or delayed images

after their initial UGI to define the position of the cecum **(John *et al.*, 2002).**

Malrotation, which in its typical form, can be corrected safely and effectively with surgery. However, the clinical presentation and anatomy of malrotation occurs along a wide clinical and anatomic spectrum. The medicolegal dilemma of whether to operate on children who present with atypical symptoms or atypical anatomy is a topic with little data.

Some of these patients clearly have malrotation, are at risk for ischemic volvulus, and require operation. Other patients, however, appear to have anatomy that offers a broad mesentery and normal function but is not textbook “normal.” Traditionally, these patients have undergone operative “repair” for fear of future volvulus and the significant consequences **(John *et al.*, 2002).**

AIM OF THE WORK

Aim of this study is to highlight pathophysiology, types, complications and modalities in management of intestinal malrotation.

CHAPTER (1)

Anatomy, Embryology and Physiology of the Duodenum and the Small Intestine