

# Recent Modalities in Management of Intestinal Malrotation

Essay

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surgery

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# **TABLE OF CONTENTS**

<b>LIST OF FIGURES</b>	<b>I</b>
<b>LIST OF ABBREVIATIONS</b>	<b>V</b>
<b>INTRODUCTION AND AIM OF WORK</b>	<b>a</b>
<b>AIM OF THE WORK</b>	<b>e</b>
<b>CHAPTER 1: Anatomy, Embryology and physiology of the duodenum and the small intestine</b>	<b>1-30</b>
1. Anatomy of the duodenum and small intestine	<b>1</b>
2. Brief embryology of the duodenum and the small intestine	<b>24</b>
<b>CHAPTER 2: Pathophysiology of intestinal malrotation</b>	<b>31-56</b>
1. History	<b>31</b>
2. Definition and mechanism: Abnormal embryology	<b>32</b>
3. Types and etiology	<b>36</b>
4. Epidemiology	<b>41</b>
5. Genetic factors in intestinal malrotation	<b>42</b>
<b>CHAPTER 3: Diagnosis of intestinal malrotation</b>	<b>57-95</b>
1. Presentation	<b>57</b>
2. Associations	<b>60</b>
3. Laboratory workup	<b>62</b>
4. Abdominal radiographs	<b>63</b>

	5. Upper GI series	<b>66</b>
	6. The role of contrast enema	<b>82</b>
	7. Ultrasound	<b>83</b>
	8. CT and Magnetic Resonance Imaging	<b>87</b>
	9. Laparoscopy	<b>87</b>
	10.Special Circumstances	<b>88</b>
	11.Delayed presentation of malrotation and midgut volvulus: imaging findings	<b>89</b>
<b>CHAPTER 4: Management of intestinal malrotation</b>		<b>96-127</b>
	1. Medical care	<b>96</b>
	2. Surgical care	<b>97</b>
<b>Summary</b>		<b>128</b>
<b>References</b>		<b>131</b>
<b>Arabic summary</b>		<b>١</b>

# **LIST OF FIGURES**

<b>1</b>	A, The four parts of the duodenum. B & C, The relations of the duodenum	<b>2</b>
<b>2</b>	The arterial supply of the duodenum	<b>9</b>
<b>3</b>	The superior mesenteric artery and its branches	<b>19</b>
<b>4</b>	Normal intestinal rotation and fixation	<b>28</b>
<b>5</b>	Normal duodenojejunal rotation	<b>30</b>
<b>6</b>	Normal intestinal rotation	<b>30</b>
<b>7</b>	(a) Normal; (b) Malrotation;	<b>35</b>
<b>8</b>	Incomplete Intestinal rotation	<b>38</b>
<b>9</b>	Midgut volvulus in a 1-week-old newborn with bilious emesis from incomplete intestinal rotation	<b>38</b>
<b>10</b>	Intestinal non-rotation	<b>39</b>
<b>11</b>	A toddler presenting with acute onset of bilious emesis for 1 day	<b>40</b>
<b>12</b>	Transverse sections showing schema for development of mesodermal germ layer and gut tube	<b>44</b>
<b>13</b>	Requirement of normal Foxf1 function for lateral plate differentiation and coelom formation	<b>45</b>
<b>14</b>	Model for the directional looping of the gut tube	<b>47</b>
<b>15</b>	Newborn with precipitous fall in hematocrit after delivery	<b>64</b>

<b>16</b>	1-day-old full-term infant with distension and bilious vomiting	<b>65</b>
<b>17</b>	Normal anatomy of the duodenum in a 5-day-old boy	<b>67</b>
<b>18</b>	Incomplete rotation in a 5-month-old girl evaluated for gastroesophageal reflux disease	<b>69</b>
<b>19</b>	Non-rotation of the intestines in a 6-year-old girl who was treated in the past for omphalocele and presented with vomiting	<b>70</b>
<b>20</b>	Midgut volvulus in a 5-day-old boy who presented with bilious emesis. Lateral view shows spiraling (corkscrew sign) of the duodenum. During surgery, 360° midgut volvulus was found	<b>71</b>
<b>21</b>	Midgut volvulus in a 27-day-old boy who presented with progressive emesis that became bilious.	<b>72</b>
<b>22</b>	False-positive diagnosis of intestinal malrotation due to deviation of the duodenum by distended bowel	<b>74</b>
<b>23</b>	Marked deviation of the DJJ by distended colonic loops in a 1-week-old child with Hirschsprung disease	<b>75</b>
<b>24</b>	Deviation of the DJJ by anasojejunal tube mimicking mal-rotation in a 2-month-old boy	<b>76</b>

<b>25</b>	Tortuous duodenum with indeterminate location of the DJJ on the frontal UGIS view (A) in a 4-month-old boy evaluated before percutaneous gastrostomy	<b>77</b>
<b>26</b>	Intermittent distended proximal duodenum in a 6-month-old boy with malrotation and midgut volvulus who presented with bilious emesis	<b>78</b>
<b>27</b>	Non fixated duodenum in a 5-month-old boy who had malrotation and para-duodenal hernia, who presented with emesis	<b>79</b>
<b>28</b>	A wandering duodenum in a 3-month-old boy presenting with feeding difficulties.	<b>79</b>
<b>29</b>	The value of SBFT study in challenging UGI studies. A 56-day-old girl with complex congenital heart disease presented with feeding intolerance.	<b>80</b>
<b>30</b>	Right lower quadrant position of the cecum in a 1-day-old boy with malrotation.	<b>81</b>
<b>31</b>	Normal and abnormal relationships between the SMV and the SMA in ultrasound performed for evaluation of hypertrophic pyloric stenosis in 1-month-old boys	<b>82</b>
<b>32</b>	A second UGIS confirmed normal anatomy of the duodenum in a 56-day-old girl who presented with emesis	<b>84</b>

<b>33</b>	Midgut volvulus diagnosed using US in an 83-day-old boy evaluated for hypertrophic pyloric stenosis	<b>85</b>
<b>34</b>	Malrotation in a 4-month-old girl with heterotaxia. Frontal UGIS view shows the stomach on the right.	<b>86</b>
<b>35</b>	Malabsorption, fortuitous spiraling of the nasogastric tube	<b>92</b>
<b>36</b>	Viral gastroenteritis, pseudo intussusception and abnormal cecal position	<b>93</b>
<b>37</b>	Intractable vomiting and dehydration	<b>95</b>
<b>38</b>	Duodenal obstruction, pseudo SMA syndrome	<b>95</b>
<b>39</b>	This patient had malrotation with midgut volvulus	<b>100</b>
<b>40</b>	The classic Ladd procedure	<b>109</b>
<b>41</b>	Proposed management algorithm.	<b>120</b>
<b>42</b>	Newborn with large gastroschisis and evisceration of the liver, stomach, entire small and large bowel	<b>123</b>



## **LIST OF ABBREVIATIONS**

<b>ACD/MPV</b>	Alveolar Capillary Dysplasia With Misalignment Of Pulmonary Veins
<b>Aptt</b>	Activated Partial Thromboplastin Time
<b>CBC</b>	Complete Blood Count
<b>CT</b>	Computed Tomography
<b>DJJ</b>	Duodenal-Jejunal Junction
<b>IV</b>	Intravenously
<b>MMIH</b>	Megacystis, Microcolon and Intestinal Hypoperistalsis
<b>MRI</b>	Magnetic Resonance Imaging
<b>NG</b>	Nasogastric
<b>NPO</b>	Nothing By Mouth
<b>PICU</b>	Pediatric Intensive Care Unit
<b>PT</b>	Prothrombin Time
<b>SMA</b>	Superior Mesenteric Artery
<b>SMV</b>	Superior Mesenteric Vein
<b>TPN</b>	Total Parenteral Nutrition
<b>UGI</b>	Upper Gastrointestinal Series
<b>US</b>	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 20<sup>th</sup> 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)**

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