The aim of the study. Retrospective analysis of diagnosis, surgical management and final outcome of malrotation in children.

Material and methods. Clinical charts, operative notes and imaging studies of all children operated on for malrotation in years 1985-2005 were studied. Children in whom malrotation was diagnosed incidentally during an operation performed because of other indications were excluded. A detailed analysis of the presentation, signs and symptoms, clinical course, and diagnostic modalities used in affected patients was carried out. Anatomical variants of malrotation and their operative management were evaluated as well.

Results. In the period from 1985 to 2005 forty children with malrotation were treated. There were 25 neonates, 10 infants and 5 children older than 1 year of age. Thirty-five children were referred directly as inpatients from other hospitals. An acute or urgent clinical presentation was noted in 32 children, while in 8 patients as the presentation was subacute or chronic. In all children bile-stained vomiting was the predominant sign. In 21 children, ileus was the referral diagnosis. Only 5 children were admitted with an established diagnosis of malrotation. In six cases duodenal atresia was suspected. Delay in diagnosis of congenital intestinal anomaly was identified in 15 children. In five newborns, the initial diagnosis was enterocolitis. Four other patients, including one infant and three older children, were admitted to the pediatric departments several times before receiving the final diagnosis. In 18 children, the only pre-operative imaging was abdominal plain X-ray. In 22 patients, contrast studies were performed as well. All children underwent operative management. Ladd’s syndrome was diagnosed in 16 patients, and in another 7 children was associated with small bowel torsion. Isolated small bowel torsion was noted in 7 patients. In 6 patients, midgut volvulus with strangulation was found intraoperatively. Two children with typical anatomical features of non-rotation presented with recurrent episodes of ileus. In single cases, mesocolic internal hernia and sigmoid torsion were noted. 5 children died postoperatively, four of whom had midgut volvulus with massive small bowel necrosis.

Conclusions. Malrotation is a rare congenital anomaly that can present with a wide anatomical and clinical spectrum but should be considered in the differential diagnosis for all children, regardless of age, manifesting with bile-stained vomiting. Suspiclon of malrotation seems to be an obvious indication for referral to a tertiary pediatric surgical department where necessary imaging should be undertaken with the participation of an experienced radiologist. Regardless of its clinical presentation, malrotation should be considered an unequivocal indication for surgical treatment due to risk of midgut volvulus with its serious sequelae.

Key words: malrotation, intestine, Ladd’s band, duodenal obstruction

The complex embryology of the midgut, which encompasses developmental stages from a physiological umbilical cord hernia to a final formation of peritoneal fixation, is reflected in a wide spectrum of abnormalities of intestinal rotation and fixation. Between the 4th and 12th weeks of gestation, embryonic midgut undergoes dynamic growth and elongation with rotation around an axis formed by the superior mesenteric artery. Both the duodenojejunal and ileocecal loops rotate through an arch totaling 270 degrees in a counterclockwise direction (1,
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Only few papers dealing with malrotation have been published in the Polish medical literature and this fact prompted the authors to undertake a retrospective analysis of the clinical aspects of this defect in children.

MATERIAL AND METHODS

A retrospective study of the medical charts of all children treated for malrotation in the Department of Pediatric Surgery and Urology between 1985-2005 was carried out. Intraoperative findings of malrotation provided the definitive diagnosis of this congenital anomaly. Only children operated on for malrotation as reason for an operative management were included in the study group. Therefore children in whom this defect was noted incidentally during an operation performed for other reasons or in whom malrotation was an inherent part of pathological spectrum like anterior wall defects or congenital diaphragmatic hernia were excluded from consideration.

Detailed analysis of the clinical symptoms and type of presentation, diagnostic work-up and the results of imaging studies was performed in each case. The details regarding the anatomical features of the defect and the surgical management were extracted from patient’s operative notes. Postoperative complications and treatment outcome were also analysed.

RESULTS

40 children underwent surgery for malrotation in the last 20 years in the University Department of Pediatric Surgery and Urology. The patient’s ages ranged from 1 day of life to 14 years. The number of patients in each age group is shown in tab. 1. Of all patients, 67.5% were newborns, with the highest incidence of malrotation noted within the first days of life. Table 1 presents the occurrence of the clinical variants of malrotation in the studied age groups as well.

The study population included 6 premature infants. In 3 of them, with gestational ages of 33, 36 and 37 weeks midgut volvulus with strangulation was found intraoperatively. In the other 3, delivered at 33, 36 and 37 gestational weeks, torsion of the intestine, Ladd syndrome, and intestinal torsion in association with Ladd’s bands were diagnosed, respectively.
Associated anomalies were noted in 5 children. In 3 newborns, a severe cardiovascular defect was noted; two of these presented with midgut volvulus with strangulation. The third patient, apart from cardiac defect, presented typical morphological features of trisomy 21. In one baby with intestinal torsion a suspicion of Di George syndrome (congenital thymic and parathyroid hypoplasia with immunologic defects) was raised. Recurrent episodes of ileus in the course of non-rotation were noted in an infant with a large open thoraco-lumbar myelo- meningocele.

Acute or sudden clinical manifestation of the defect was noted in 32 children. Analysis of clinical characteristics revealed that bile-stained vomiting was the most common symptom in this group of patients. In 4 of 6 children with midgut volvulus and strangulation, lower gastrointestinal haemorrhage was noted. Recurrent episodes of ileus in the course of non-rotation were noted in an infant with a large open thoraco-lumbar myelo- meningocele.

Table 1. Occurrence of clinical variants of malrotation in relation to age of patients

<table>
<thead>
<tr>
<th>Clinical variant</th>
<th>Age</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>1-7 days</td>
</tr>
<tr>
<td>Ladd syndrome</td>
<td>6</td>
</tr>
<tr>
<td>Ladd syndrome with small bowel</td>
<td>2</td>
</tr>
<tr>
<td>torsion</td>
<td></td>
</tr>
<tr>
<td>Small bowel torsion</td>
<td>4</td>
</tr>
<tr>
<td>Midgut volvulus</td>
<td>2</td>
</tr>
<tr>
<td>Mesocolic hernia</td>
<td></td>
</tr>
<tr>
<td>Sigmoid torsion</td>
<td></td>
</tr>
<tr>
<td>Malrotation with recurrent ileus</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>14</td>
</tr>
</tbody>
</table>

Among children with bowel strangulation as a result of midgut volvulus, the first clinical symptoms occurred on the first day of life in 3 patients. One infant started vomiting on day 6 and his condition significantly deteriorated on day 10. He was admitted to the surgical unit three days later and an emergent operation was performed. A fifth patient with midgut volvulus was referred to the surgical department at age of 29 days after presenting with sudden deterioration of his clinical condition. He had been treated since birth at the neonatal special care unit for sepsis, pneumonia, suspected cardiac anomaly and hyperbilirubinaemia. The last child with midgut volvulus and strangulation, a 2-month–old boy, had presented with 3 episodes of incarcerated inguinal hernia within 2 weeks prior to an episode of sudden deterioration with vomiting. On admission to the surgical unit he presented with sudden cardio-respiratory arrest and required resuscitation. At intubation extensive bilious gastric content was aspirated from the respiratory tract. Intraoperatively, volvulus with strangulation of the small bowel, which had a very narrow mesenteric pedicle, was found.

In 8 patients the clinical manifestation was described as subacute or recurrent. The two oldest patients, a 7-year-old boy with Ladd syndrome and a 14-year-old boy with mesocolic hernia, were admitted to the local hospital on several occasions because of acute episodes of abdominal pain and vomiting prior to referral to the surgical unit. A 13-month-old child who had presented with recurrent episodes of vomiting since birth had been admitted 7 times as an inpatient with a presumptive diagnosis of enteropathy. This patient was diagnosed intraoperatively with intestinal torsion with Ladd’s bands. Two other children with Ladd’s syndrome presented repeatedly with bilious vomiting since the first week of life until a final diagnosis was made on the 22nd day and 5th week of age, respectively. A 1-month-old child vomiting intermittently since day 8 of life and failure of conservative treatment prompted a local pediatrician to refer to a surgical unit with a provisional diagnosis of pyloric stenosis. This
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Plain abdominal X-ray was the only diagnostic study in all 6 children with midgut volvulus and strangulation. In 3 of them, the radiological picture was consistent with high obstruction with two fluid levels in the upper abdomen, in 1 patient the study demonstrated multiple fluid levels, and in 2 only a single fluid level in the upper abdomen associated with a gasless abdomen was noted. In children with Ladd’s syndrome, an abdominal plain film showed either a single or double air bubble in the upper portion and poor aeration or paucity of gas in the distal intestine. In children with intestinal torsion without irreversible bowel ischaemia radiological features included upper abdominal fluid levels indicating ileus in 2 cases and virtually gasless abdomen with a single gastric air bubble in 4 of them.

The diagnosis of malrotation was delayed in 14 children, as indicated by a time lapse between the onset of clinical symptoms and the final diagnosis. Four children had been treated in local hospitals on several occasions due to recurrent abdominal complaints with bilious vomiting prior to final referral to the surgical department, but the diagnostic contrast studies were not performed in any of them. They were found to have intestinal torsion, intestinal torsion with Ladd’s band, Ladd’s syndrome and mesocolic hernia respectively. In another five children, bilious vomiting of variable intensity prompted diagnostic evaluation. This did not show evident abnormality, and temporary clinical improvement caused a delay in recognition of malrotation, which occurred only when the clinical symptoms with vomiting recurred. In two newborns and one infant, the initial clinical diagnosis was necrotizing enterocolitis and conservative treatment commenced during hospitalization in a local neonatal care unit. One child operated on for Ladd’s syndrome had previously been treated twice in a local hospital with diagnoses of enteritis and hypertrophic pylorostenosis. The last patient from this group had first been treated conservatively by his local pediatrician due to recurrent vomiting and subsequently was admitted to a pediatric ward. Finally, he was referred to surgical department with a provisional diagnosis of pyloric stenosis.

All children were subjected to operative treatment. In Ladd’s syndrome treatment consisted of division of the peritoneal bands crossing and obstructing the duodenum, and when
required, in broadening of the mesenteric root through dissection of its anterior layer. In cases of small bowel torsion, the intestine was first untwisted and its viability was assessed immediately after delivery from the abdomen. When no signs of ischaemia were present, a Ladd procedure with widening of the mesenteric root and mobilization of the duodenum was performed. In children with classical midgut volvulus with strangulation, the type of operative intervention depended on the grade and spatial extent of bowel ischaemia and necrosis. In 5 patients, during the first laparotomy the small bowel was untwisted and its viability evaluated. Due to extensive ischaemia and necrosis affecting nearly the whole small bowel, a final decision on resection was postponed to the second-look laparotomy. In one of these babies, three perforations were simply closed and the bowel was placed in an Silastic pouch attached to the margins of the abdominal wound after decompression. In 3 babies, the frankly necrotic intestinal portion of the bowel was resected and the free ends of the remaining intestine were tied up. The second-look laparotomy was performed after 24 hours in 3 cases, and after 48 hours in one patient. One newborn died after the first operation. At repeat laparotomy, extensive necrosis prompted resection of nearly all small bowel loops in 3 of these infants. Short proximal jejunal and distal ileal portions were anastomosed, restoring the continuity of the gastrointestinal tract. One patient had approximately 70 cm of necrotic small bowel resected. In the last patient with midgut volvulus, at first laparotomy a 50-cm long segment of necrotic bowel was resected and double-barrel enterostomy was formed.

In a child with torsion of the sigmoid colon, the sigmoid colon was untwisted and its mesentery carefully widened. Mesocolic hernia was repaired laparoscopically and the procedure consisted of reduction of the small bowel and closure of the hernial neck. Apart from this case, laparoscopy was performed in a 22-day old boy with Ladd’s syndrome.

In 25 children, the postoperative course was uneventful and they were discharged from the surgical department in good condition. Two patients required early repeat laparotomy due to adhesion ileus on the 12th and 18th postoperative days, respectively. One of these patients had the additional postoperative complications DIC and meningitis. Seven children were sent back to their referring departments for further medical treatment.

In 2 children with Ladd’s syndrome and one child with intestinal resection due to volvulus, poor weight gain despite full oral feeding necessitated further medical management. Four newborns were readmitted to their neonatal special care unit after successful recovery from surgical treatment.

In the whole group of 40 children, 5 deaths occurred (12.5%). Four newborns died in the early postoperative period, and all had midgut volvulus with extensive necrosis of the small bowel. One patient, resuscitated on admission to the surgical department, died on the day of the operative procedure. One infant with a complex anomaly of the heart and great vessels died due to circulatory and renal failure on the third postoperative day after a second-look laparo-

<table>
<thead>
<tr>
<th>Variant</th>
<th>Diagnostic study</th>
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<tbody>
<tr>
<td></td>
<td>AxR</td>
</tr>
<tr>
<td>Ladd syndrome</td>
<td>5</td>
</tr>
<tr>
<td>Ladd syndrome with small bowel torsion</td>
<td>1</td>
</tr>
<tr>
<td>Small bowel torsion</td>
<td>2</td>
</tr>
<tr>
<td>Midgut volvulus</td>
<td>6</td>
</tr>
<tr>
<td>Mesocolic hernia</td>
<td>1</td>
</tr>
<tr>
<td>Sigmoid torsion</td>
<td>1</td>
</tr>
<tr>
<td>Malrotation with recurrent ileus</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>17</td>
</tr>
</tbody>
</table>

AxR – abdominal plain-ray
Upper GI – upper GI contrast study
Lower GI – contrast enema
USS – abdominal ultrasound scan

Table 2. Diagnostic studies in children with various types of malrotation
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tomy. In two other cases, death occurred on the 8th and 10th postoperative day due to sepsis and cardio-respiratory failure. The last fatal case refers to the patient with intestinal torsion associated with Ladd’s bands. This was the only child operated on initially in the local pediatric surgical department. This child required two repeat laparotomies due to ileus and enterotomy in the early postoperative period and was subsequently referred to the Department of Pediatric Surgery and Urology. Her condition was very unstable and required very intensive cardio-respiratory support. She died at 45 days of age due to multi-organ failure associated with DIC, after having two further abdominal explorations because of perforation and ileus performed.

DISCUSSION

Malrotation is not a pathology commonly encountered in clinical practice. Although it is found in 0.5-1% of population on autopsy, its incidence among newborns has been estimated as 1:6000 (5). This fact is reflected in the medical literature. Rescorla et al. presented their experience in treatment of 447 children with malrotation and this is the largest group of patients reported to date. However, it should be emphasized that 331 of these patients were affected by abdominal wall defects and congenital diaphragmatic hernia (6). In these babies, abnormal rotation of the intestine is part of the complex congenital pathology and not a separate defect. Most clinical reports dealing with this subject present a series of children, usually ranging from a dozen to several dozens of patients (7-10). The data collected from the pediatric surgical centres in Poland indicate that 107 newborns were treated for malrotation in years 2000-2001. This number indirectly demonstrates the epidemiological aspects, as it does not include infants and older children and refers only to cases that presented with clinical symptoms. Newborns with malrotation were reported from the 29 centres, what clearly indicates that only few patients were treated in each department (11).

In this paper, we undertook a retrospective analysis of diagnostic and surgical management in children in whom malrotation was the principal pathology. For this reason, apart from exclusion of anterior wall defects and diaphragmatic hernia, children with other congenital anomalies of the gastrointestinal tract (duodenal and intestinal atresia) were not analysed. Assuming such restrictive criteria resulted in a low incidence of associated anomalies of 12.5% in the presented series of children. According to the data from literature, this frequency ranges from 7.5% to 62%, but it is again necessary to emphasize that most authors include congenital defects of the gastrointestinal tract in their reports (7, 8, 12, 13).

In the series of patients with malrotation reported herein, there was a clear predominance of newborns (67.5%), and in more than one-third of them the defect was diagnosed within the first week of life. These data confirm that clinical presentation of malrotation usually occurs in the group of the youngest children. According to the literature, between 50% and 82.2% of all cases of malrotation are noted in the neonatal period. When newborns and infants are combined, the incidence of the clinical presentation of this defect reaches 84%-92% (12, 14, 15, 16). One must not omit the fact that malrotation may be diagnosed in older children as well. Therefore, age should not serve as an exclusion criterion for malrotation as a possible cause of abdominal symptoms (10).

Malrotation is a general descriptive term for a wide spectrum of anatomical variants of rotational abnormalities of the intestine and its fixation. From the clinical point of view two sequelae of this congenital anomaly are decisive for its course. Abnormal fixation of the small intestinal mesentery with its pathologically narrow base may result in midgut volvulus and subsequent strangulation. This most serious complication is usually associated with non-rotation or incomplete rotation with the duodenum and duodeno-jejunal junction located to the right of midline and with a high riding ileo-cecal segment. Extrinsic duodenal obstruction is the second important factor affecting a clinical presentation (2, 5, 6). It may result from compression by Ladd’s bands running across the duodenum from the abnormally located cecum to the posterior abdominal wall. Torsion of the small intestine may also be responsible for duodenal obstruction. Other variants of malrotation occur with significantly lower frequency. Mesocolic hernias are the result of abnormal fixation of either the left or the right colon with persistence of the wide mesentery of these segments (2, 5). Sigmoid volvulus has been reported very seldom in children (17).
Normal embryology of intestinal rotation was precisely presented and illustrated by Snyder and Chauffin (2). In 1958, Estrada described in detail all possible anatomical variants of this congenital anomaly in his monograph on malrotation and malfixation (1). A precise determination of the variant of malrotation requires profound knowledge of the embryology of the gastrointestinal tract and very precise assessment of intraoperative anatomy and topography. From the practical point of view, most surgeons use the term “malrotation” exclusively omitting more accurate descriptive terms and focusing on the clinical sequelae of the defect. The authors of the presented paper decided to differentiate variants of malrotation, taking into account the most important anatomical features and their clinical significance. Ladd syndrome and midgut volvulus were noted in most children. The latter variant was further divided into isolated midgut volvulus and midgut volvulus accompanied by Ladd’s bands. Because of their clinical importance, cases of midgut volvulus with strangulation of the bowel were analysed separately.

Analysing the data of the children reported herein, Ladd syndrome appeared to be the most common form of malrotation and was noted in 40% of all cases. Inclusion of the patients with Ladd’s bands and midgut volvulus increases this ratio to 57.5%. Filston et al. noted the Ladd’s bands in 64% of children with malrotation. In the subgroup of patients with midgut volvulus, this incidence was 50% (9). Seashore et al. found the Ladd’s bands in all their patients operated on for midgut volvulus (14). The surgeons from Glasgow reported Ladd’s syndrome in 29% of 34 newborns (8). Review of the relevant literature has shown that most authors reporting their experience in malrotation in children focused predominantly on midgut volvulus. This is without doubt the most important clinical aspect of this anomaly. Midgut volvulus carries a high risk of intestinal strangulation and subsequent ischaemia and necrosis. It is reported in 31.3% to 70.5% of all cases of malrotation in children (7, 9, 12, 14). In the present study it was found in 50% of patients. The highest risk of intestinal torsion was noted accordingly by most authors among newborns and infants affected by malrotation. In children older than 1 year of age this risk was reported as much lower, with a maximum of 14% (5, 16, 18). Spigland et al., analyzing clinical aspects of malrotation in older children, found midgut volvulus in 65% of cases and emphasized that a patient’s age should not be regarded as factor when excluding a complicated course of malrotation (19).

The authors of the presented paper reported intestinal torsion in 2 of five older children, which seems to confirm the aforementioned statement. The clinical presentation of midgut volvulus may be acute, subacute or chronic recurrent. It may be speculated that malrotation is a dynamic pathological condition ranging from a clinically silent entity in the initial phase, when venous and lymphatic intestinal drainage have not been affected, through more advanced stages with venous congestion and potential duodenal or intestinal obstruction. Midgut volvulus with strangulation and arterial ischaemia represents the final stage of intestinal pathology. It may be difficult to differentiate clearly between acute and chronic manifestations of malrotation in some cases. In patients with initially mild or moderate recurrent clinical symptoms, only acute episodes of significant deterioration may serve as a threshold for undertaking diagnostic studies or even referring a patient for urgent surgical management. Two children from the present study manifesting with chronic symptoms since the neonatal period were eventually found to have midgut volvulus with Ladd’s bands at the ages of 1 and 13 months respectively. Midgut volvulus was diagnosed in another 5 children with diagnostic delay taking into account the time lapse between the onset of occurrence of initial clinical symptoms and the surgical procedure. The fact that 5 previously mentioned patients had midgut volvulus associated with Ladd’s band may indicate a specific clinical course of this variant of malrotation. In the initial phase the symptoms of partial duodenal obstruction may predominate but it is only when intestinal torsion ensues that acute presentations occur.

Among 40 children with malrotation, in 6 (12.5%) intestinal necrosis was noted. In three of them surgical management was undertaken within the first days of life. This serious complication was noted only in one infant, who presented with recurrent episodes of inguinal hernia incarceration prior to admission to the surgical department. It may indicate that the potential risk of strangulation is not limited to neonates. The other authors reported midgut volvulus with ischaemia of the affected bowel
in 4% to 18% of their patients (6, 8, 12, 14, 20). Rescorla et al noted intestinal ischaemia in 10 of 18 children with acute presentations of malrotation (6). This is the subgroup of children carrying the worst risk of morbidity and mortality. In a significant number of such patients, extensive resection of necrotic bowel is necessary. Therefore, a staged surgical management with second look procedures has been introduced in many surgical centres. Such operative treatment was undertaken in 4 newborns reported herein with extensive intestinal necrosis. In 3 of them, no significant improvement of bowel viability was noted despite intensive supportive treatment. In the whole group of 40 children 5 deaths occurred and 4 fatal cases were the direct result of midgut volvulus with strangulation. The data from other centers support the notion that this complication of malrotation is responsible for the final outlook. Mortality in malrotation was reported as high as 1.8% to 24.4% (5, 6, 7, 15). Midgut volvulus with bowel ischaemia is characterized by a very acute course in most cases. Such a clinical presentation is a clear indication for urgent surgical management without diagnostic delay. Taking these facts into consideration, it must be emphasized that prompt surgical management in every child with diagnosed malrotation, regardless of its clinical presentation and the patient’s age, seems to be fully justified.

The variable clinical course of malrotation may pose significant diagnostic difficulties. Without doubt, acute symptoms of ileus constitute clear indication for prompt diagnostic studies and urgent operative management. On the other hand, occurrence of less severe symptoms of changing nature may result in delay of establishing the final diagnosis. Analysis of the clinical characteristics of the reported group of 40 children showed clearly that bilious vomiting is the most characteristic symptom of malrotation. This is, however, not pathognomonic for this anomaly. Other authors confirmed this finding in their series. The clinical presentation of abnormal rotation of the bowel depends on the patient’s age as well. The older the child is, the less specific is a clinical course of anomaly (18, 21). This statement, reported by many authors, seems to be justified not by less specific manifestations of malrotation in older children, but rather by its rare incidence in these age groups and even rarer consideration of malrotation in differential diagnosis by physicians. This is confirmed by the fact that 3 of the 5 older children in the presented review were admitted to local hospitals on several occasions prior to referral to a surgical department. On the other hand, study of the clinical course of malrotation in 14 children with noted diagnostic delay, showed that diagnostic pitfalls occurred in neonates as well when their symptoms were attributed to other common neonatal pathologic conditions. In 4 of them an initial working diagnosis of necrotizing enterocolitis was made. In 2 other children presenting with recurrent episodes of vomiting, the suspicion of hypertrophic pyloric stenosis was raised. The fact that only 5 children had malrotation as a referral diagnosis clearly shows that this congenital anomaly is rarely taken into consideration in differential diagnosis by paediatricians or neonatologists.

Diagnostic difficulties in malrotation may be due not only to the variable clinical course of the disease but also to the inconclusive results of imaging studies. In the group of children reported herein, all underwent abdominal X-ray. This study does not allow definitive diagnosis of malrotation but, in association with clinical symptoms, helps to raise suspicion of this anomaly or to provide indications for prompt surgical intervention. Impaired duodenal patency in the course of Ladd’s syndrome or midgut volvulus usually gives the radiological appearance of a double-bubble with paucity of air within the distal part of the gastrointestinal tract. In some patients, mild obstruction of the duodenum may not result in pathologic appearance on plain film. In other patients, a variable course of the disease with various radiological pattern of intestinal aeration on subsequent films may be responsible for inconclusive interpretation and eventually diagnostic delay. Currently, in most centers upper GIT contrast study is regarded as the most important diagnostic modality in children with symptoms of malrotation (5, 14, 22). The patients presenting with unequivocal acute peritoneal symptoms are the exception to this rule. A location of the duodeno-ileal junction to right of midline is a classical finding, but there are other radiological features suggestive of this anomaly (4, 15, 21, 22). On the other hand, an upper contrast study is not associated with 100% accuracy and in some cases may not reveal any abnormality. Its interpretation requires a great
deal of radiological experience as the wide anatomical spectrum of malrotation may produce various radiological features. In the analysed series, this study was initially inconclusive in 4 children. However, their deteriorating clinical symptoms prompted further imaging, which confirmed the rotational abnormality. According to other authors, barium enema showed significantly less sensitivity in detection of malrotation in affected children (8, 21).

Surgical management of malrotation has been well established and is uniformly based on principles elaborated by Ladd (4, 5, 6). In two children with recurrent episodes of ileus, Ladd’s procedure was combined with Malone appendicostomy. In both the final outcome was good. Complete cessation of previously observed abdominal symptoms may be attributed to broadening of the mesenteric base but it seems that appendiceal stomy itself might serve as a factor preventing intestinal torsion through fixation of the caecum to the abdominal wall.

In summary, malrotation is a congenital abnormality with a very wide clinical spectrum. The most common clinical sign is not pathognomonic, but its occurrence in a newborn or older child should always result in its inclusion in a differential diagnosis. Midgut volvulus with strangulation has an urgent clinical course. In all such cases a pediatric surgeon must decide on emergent operative treatment based on clinical symptoms alone, as time plays a major prognostic role and there is no available study to objectively assess small intestinal viability (3, 5, 12). Stable condition of a newborn or infant, presenting with typical signs of high intestinal obstruction, is an indication for precise diagnostic evaluation. Plain abdominal x-ray is, unfortunately, often non-diagnostic or has little value for diagnosis of malrotation, so contrast studies play a major role. Diagnostic contrast studies should be performed only in the presence of an experience radiologist and pediatric surgeon. Fluoroscopic control allows for an objective assessment of all relevant anatomic landmarks of the gastrointestinal tract, and finally for diagnosis of intestinal abnormality. Failure to adhere to these principles may result in misdiagnosis or delay in recognition of malrotation. Lower gastrointestinal tract contrast studies are less accurate and therefore should be carried out in case of diagnostic problems or in children presenting with signs of lower GIT obstruction. In recent years, there have been reports on the usefulness of ultrasonographic studies for preoperative diagnosis of intestinal volvulus through evaluation of the anatomy of the mesenteric vessels (4, 5, 22). Diagnosis of malrotation in a child presenting with associated clinical symptoms, regardless of the patient’s age, should be regarded as a strict indication for operative management. Additionally, recognition of clinically silent malrotation should be followed by operative correction due to the risk of midgut volvulus, which may occur at any age, as its most serious complication.

CONCLUSIONS

1. Malrotation is a rare congenital anomaly which can present with a wide anatomical and clinical spectrum but should be considered in the differential diagnosis for all children, regardless of age, presenting with bile-stained emesis.
2. Suspicion of malrotation appears to be an obvious indication for referral to a tertiary pediatric surgical department where necessary imaging should be undertaken with the participation of an experienced pediatric radiologist and surgeon.
3. Regardless of its clinical presentation, malrotation should be considered as an unequivocal indication for surgical treatment due to risk of midgut volvulus with its serious sequelae.

REFERENCES

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COMMENTARY

Establishing an early diagnosis of intestinal malrotation often creates a problem. Only the children with a symptomatic onset of sickness, which had led to operative treatment, were analyzed.

It was shown that the preoperative diagnosis was often different from the intraoperative findings. These differences with their various clinical courses were consequently analyzed and presented in detail by the Authors. The retrospective analysis of the medical histories of 40 children, operated upon for intestinal malrotation during the last 20 years, demonstrated numerous diagnostic mistakes. These were mainly caused by a lack of experience of pediatricians and neonatologists, as well as pediatric surgeons. There is no doubt that the ability to predict and recognize various symptoms, based on the knowledge of a typical clinical picture, might prevent the development of serious complications, both, early and late – e.g. volvulus, intestinal necrosis, short bowel syndrome, incarceration, subileus. This issue is well illustrated by the fact that only 5 children out of the whole group had a correct diagnosis of an incomplete duodenal obstruction due to Ladd’s syndrome when referred for an operative treatment. The rest of the children had a wide range of preliminary diagnoses, including ileus, necrotizing enterocolitis, intussusceptions, pylorostenosis or enteropathy. These diagnostic difficulties were caused partially by a very dynamic and changing clinical picture, as indicated by the Authors. At the same time, the insufficient knowledge of clinical and morphological pictures of these defects, as well as ineffective use of imaging (in terms of indications, timing and interpretation), were also major contributors to misdiagnosis.

I would like to stress the importance and value of ultrasound examinations, together with a Doppler option, in diagnosing intestinal malrotation in children. Visualizing the incorrect course of the mesenteric artery and vein (opposite directions) might have diagnostic value, as might the location of a duodenum and colon. Ultrasound also enables us to
check the vitality of the bowel. Plain abdominal X-ray, contrast enema and finally a contrast swallow should be the next steps on the diagnostic ladder. Changing this order might make reaching the diagnosis more difficult. It also requires an experienced specialist to avoid misinterpretation, especially in the III-rd type of malrotation dominated mainly by a distressed bowel fixation. Excessive colon mobility or a dysfunction of the colonic-sigmoid junction might lead to chronic constipation with secondary formation of a megacolon and might require a subtotal colectomy in adulthood. The intestinal obstruction caused by the chilaidity Syndrome or sigmoid volvulus are rare entities in children, with a more frequent occurrence in adulthood.

The operative treatment varies and depends on the type of the malformation. In cases of volvulus without bowel necrosis, the bowel should be freed and both the superficial and deep Ladd’s bands connecting the cecum with the posterior abdominal wall and the duodenum should be carefully divided. This maneuver enables the surgeon to free the mesentery, widening its root and relocating the cecum, colon and duodenum appropriately. It is sometimes necessary to free folds and kinks of the ligament of Treitz, most often in cases of type II malrotation. Attempts to create additional points of cecum fixation by creating an appendicostomy or suturing to the peritoneum are controversial, as they may lead to serious complications. Managing the reverse rotation (type III malrotation according to Grobb, with the colon located behind the superior mesenteric artery – a rare entity) is difficult and requires implementing the technique described by Estrada. The application of minimally invasive methods in the diagnostic process and treatment of children with the intestinal malrotation is very useful and should be encouraged.

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