

## RARE CAUSE OF SUDDEN DEATH IN CHILDREN

## UNE CAUSE RARE DE MORT SUBITE CHEZ L'ENFANT

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**Abstract**

We report the case of two children who died of occlusion by volvulus due to primary intestinal malrotation and whose diagnosis was only made at autopsy. These types of observation are particularly rare. Indeed, the literature lists only few fatal cases because of the late and fatal nature of prognosis. Fatal cases are rarely reported after the age of one year. The abnormalities of primary intestinal rotation are exceptionally symptomatic for the big child and the adult. At these ages, the very low incidence of their complications, especially small bowel volvulus, makes this diagnosis rarely done. Surgical treatment is validated and effective but must be done early to avoid bowel obstruction by volvulus. Because of their serious and fatal prognosis, it seems important to us to evoke this diagnosis even for children and adults when facing disturbing and unexplained digestive disorders.

**Key words:** Intestinal malrotation; Autopsy; Death; Volvulus occlusion.

**Résumé**

Nous rapportons le cas de deux enfants décédés suite à une occlusion par volvulus secondaire à une malrotation intestinale primaire dont le diagnostic n'a été posé qu'à l'autopsie. Le décès suite à cette malformation intestinale est particulièrement rare. En effet, la littérature ne répertorie que quelques cas mortels chez les nouveaux nés. Le décès par malrotation intestinale est rarement rapporté après l'âge d'un an. Les anomalies de la rotation intestinale primaire sont exceptionnellement symptomatiques chez le grand enfant et l'adulte. À ces âges, la très faible incidence de leurs complications, en particulier du volvulus de l'intestin grêle, rend ce diagnostic rarement établi. Le traitement chirurgical est validé et efficace mais doit être effectué très rapidement pour éviter l'occlusion intestinale par le volvulus. En raison de son pronostic grave et fatal, il nous semble important d'évoquer ce diagnostic, même chez les enfants plus âgés et les adultes face à des troubles digestifs inexpliqués.

**Mots clés :** Malrotation intestinale ; Autopsie ; Décès ; Occlusion intestinale.

**ملخص**

نقدم في هذا العمل حالنا وفاة لطفلين توفيا بعد انسداد المعوي الدقيق من جراء تشويه خلقي نوع سوء استدارة الأمعاء وقد تم تشخيص سبب الوفاة فقط بعد تشريح الجثة. الموت الناجم عن هذا التشوه المعوي نادر للغاية. أدرجت الدراسات الطبية السابقة عدد قليل من الحالات المميتة لأطفال حديثي الولادة بسبب الطبيعة المتأخرة والمميتة لتشخيص المرض ونتائج خطيرة. نادراً ما يكون هذا التشوه الخلقي سبباً للوفاة بعد سن سنة واحدة. تشوهات دوران الأمعاء الأولية هي أعراض استثنائية عند الطفل الكبير الكهول. خلال هذه الأعمار، نسبة تشخيص مضاعفات تشوه سوء استدارة الأمعاء منخفض للغاية، وبالتالي يكون عادة هذا التشخيص متأخراً أو غير مبرر، الذي يمكن أن يهدد حياة المريض ويكون مميتاً. غالباً ما يتم تشخيص هذه التشوهات في وقت مبكر من حياة الوليد. العلاج الجراحي فعالاً عندما يكون استعجالياً لتجنب انسداد الأمعاء. على الرغم من الطبيعة الاستثنائية لهذا المرض وبسبب نكته الجدي والمميت، يبدو لنا من المهم استحضار هذه الأمثلة حتى لدى الأطفال الكبار والكهول لمواجهة اضطرابات الجهاز الهضمي الخطيرة والمميتة أحياناً إن لم يتم تشخيصها في الوقت المناسب.

**الكلمات المفتاحية:** سوء استدارة الأمعاء ; التشريح ; الموت ; انسداد الأمعاء.

## INTRODUCTION

Intestinal malrotation is an abnormality of rotation of the primitive intestinal loop during de embryonic development. It is always congenital. It occurs in 1 in 500 live births [1] although autopsy studies estimate that it may be as high as 1% of the total population [2]. It is the most frequent congenital malformation of the small intestine. Intestinal malrotation can stay asymptomatic if the bowel movement is possible. This anatomical abnormality is severe as it exposes the patients to a high risk of duodenal obstruction from small bowel volvulus, which also leads to ischemia of the midgut from superior mesenteric artery occlusion [1]. The diagnosis is made before the age of one month in 80% of the cases [3]. In the following report, we describe two uncommon cases of suspect death of two kids whose autopsy revealed a natural death caused by occlusion by volvulus due to primary intestinal malrotation and whose diagnosis was never suspected before their death.

## CASE REPORT

### Case history

Case 1: A 14-month-old boy born full-term of a healthy pregnancy with no family history holds a personal history of multiple consultations in the emergency department for recurring abdominal pain and bloating since the first months of life. The symptoms were attributed to simple abdominal colic and the boy has been discharged with symptomatic treatments. One day before his death, the boy became ill with symptoms including refusal to eat, abdominal pain, minimal bloating and a single episode of green vomiting. A plain abdominal X-ray was made in the emergency department, turning out to be normal. He was once again given symptomatic medicine with a clinical improvement and was discharged. The infant was found dead in his crib the next morning.

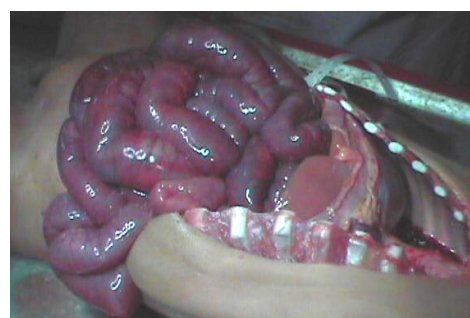
Case 2: It is the case of a two-year-old boy also born full-term of a healthy pregnancy and without family history. He was also brought multiple times to the emergency department for abdominal symptoms and was given symptomatic medicine. He had abdominal pain, diarrhea, green vomiting, and refused to eat one day before his death. The symptoms were attributed to gastroenteritis and the boy was given symptomatic treatment. The evolution of his state was marked by an alteration of his general condition followed by his death after few hours.

### Autopsy findings

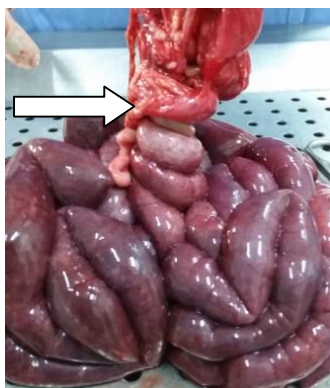
The victims were a 14-month-old and two-year-old eutrophic male infants. Both were slightly dehydrated. No traumatic injuries were found at the external examination. Autopsy findings of the two infants showed a profuse hemoperitoneum and a necrosis aspect of the small intestine and the colon (*Figure 1, 2*). Furthermore, a midgut volvulus in the right upper quadrant was found with a total of four coils containing the duodenal-jejunal junction, the coecum and the appendix (*Figure 3, 4, 5, 6*). There were no other positional abnormalities of the rest of the organs.



**Figure 1:** Case 1: Important hemoperitoneum and necrosis aspect of the intestines and colon.



**Figure 2:** Case 2: Necrosis aspect of the intestines and colon.



**Figure 3: Case 1:** Midgut volvulus in the right upper quadrant. Four coils containing the duodenal-jejunal junction, the caecum and the appendix



**Figure 4: Case 2:** Midgut volvulus in the right upper quadrant. The coils contain the duodenal-jejunal junction, the caecum and the appendix



**Figure 5: Case 1:** The appendix shown after devolvulation of the coils



**Figure 6: Case 2:** the appendix shown after the devolvulation of the coils.

## DISCUSSION

The prevalence of intestinal malrotation at birth is variable according to each author. The prevalence at the adult age is about 0, 2 à 0, 5% and it corresponds to the asymptomatic non-diagnosed forms [1]. The incidence of intestinal malrotation is around 1 out of 500 live births [1]. It is the most frequent congenital malformation of the small intestine [4]. The sex ratio is up to 2/1 for boys in the neonatal period. The prevalence of symptomatic forms is identical in both sexes after the first year of life [7]. The diagnosis is made before the first year of life in 90% of the cases and 80% of the patients are diagnosed in the first month of life [2]. Intestinal rotation occurs from the fourth to the twelfth weeks of gestation [5]. The digestive tube makes a total of three counterclockwise rotations during this period: 90°, 180° and 270° rotations. This sequence of return causes the duodenum and proximal jejunum to be pushed superiorly and to the left posterior to the superior mesenteric artery so that they become fixed in a 270° rotation from their initial position [5]. Fixation of the intestines in this position takes place over the fourth and fifth months of gestation [5]. Intestinal malrotation is the term given to errors of rotation of the midgut around the superior mesenteric artery and the midgut's following fixation in the peritoneal cavity [6].

When the rotation is stopped at 180°, the caecum and the duodeno-jejunal loop are positioned in the upper right quadrant and fixed with an adhesive peritoneal band called Ladd's band. The diagnosis of intestinal malrotation can be done at different ages, the symptoms being different according to the age. In the first month of life and more frequently in the first days, there are acute symptoms such as bilious vomiting, stop of bowel movements after emitting the meconium. Before the age of one year, the patient presents subacute occlusive symptoms. The main symptom is bilious or green vomiting. Between one and fifteen years old, the patient can suffer from chronic abdominal pain, nausea, vomiting, diarrhea, constipation or an acute small bowel obstruction. After that age, if there are abdominal symptoms, the diagnosis is hard and often not mentioned. But in most of the cases, the patients are asymptomatic; therefore, there are a lot of fortuitous diagnoses. The main paraclinical exams are upper Gastro-intestinal contrast study and abdominal doppler ultrasound [8]. If the diagnosis is delayed, a clinical diagnosis of small bowel volvulus is suspected when the patient presents with bilious and green vomiting (97% of the patients [1]), abdominal pain, minor bloating, stop of bowel movements, nausea... [1, 6]. The gold standard for the diagnosis is the upper Gastro-intestinal contrast study. The sensitivity for the diagnosis of malrotation was 96% showing an abnormal location of the duodeno-jejunal flexure and presence of the proximal jejuna loops in the right hemiabdomen [10, 11]. The UGI contrast can also show the characteristic "corkscrew" appearance in volvulus with a sensitivity of 79% [1, 9]. The diagnosis can also be made with a Doppler ultrasound showing a "whirlpool" or "swirl" sign corresponding to a twisting of the superior mesenteric vein and mesentery around the superior mesenteric artery in 83% of the patients [11, 12]. In the case of the one-year-old infant, only plain abdominal X-ray was made, which negative predictive value is very low [7]. When the diagnosis of midgut volvulus is suspected, Ladd's procedure surgery is an emergency and is immediately indicated. It consists in untwisting the intestine, dividing any adhesive bands, and widening the mesentery to result in the bowel being in a "safe" non-rotated position [1]. Both Laparoscopic Ladd and Open Ladd's procedures are the standard surgical approach [13]. The evolution is generally favorable if the diagnosis is made and the patient is managed on time [7].

## CONCLUSION

Intestinal malrotation is an abnormality of rotation of the primitive intestinal loop. It is a severe anomaly as it exposes the patient to lethal complications if the diagnosis is missed; therefore, any clinical symptom reminding the clinician of intestinal malrotation or midgut volvulus must indicate the appropriate investigations especially because there are sensitive and specific paraclinical exams and efficient treatments. This must raise awareness to be more vigilant and to evoke the diagnosis when there are chronic abdominal symptoms in order to avoid the lethal evolution of the complications. The autopsy diagnosis is unfortunately still possible.

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