Scientific Research Publishing

Open Journal of Gastroenterology, 2022, 12, 263-270
https://www.scirp.org/journal/ojgas
ISSN Online: 2163-9469
ISSN Print: 2163-9450

Incomplete Situs Inversus Revealed by Acute Pain of the Right Hypochondrium

Alice Nanelin Guingané[1*], Julien Sawadogo[2], Adjirata Koama[3], Bénilde Marie-Ange Tiemtoré-Kambou[3], Roger Sombié[4], Alain Bougouma[4]

[1]Service d'hépato-gastroentérologie, CHU Bogodogo, Ouagadougou, Burkina Faso
[2]Service de chirurgie générale et digestive du CHU-YO, Ouagadougou, Burkina Faso
[3]Service de radiologie, CHU Bogodogo, Ouagadougou, Burkina Faso
[4]Service d'hépato-gastroentérologie, CHU-YO, Ouagadougou, Burkina Faso
Email: *aliceguingane@yahoo.fr

How to cite this paper: Guingané, A.N., Sawadogo, J., Koama, A., Tiemtoré-Kambou, B.M.-A., Sombié, R. and Bougouma, A. (2022) Incomplete Situs Inversus Revealed by Acute Pain of the Right Hypochondrium. *Open Journal of Gastroenterology*, **12**, 263-270.
https://doi.org/10.4236/ojgas.2022.1210026

Received: August 26, 2022
Accepted: September 26, 2022
Published: September 29, 2022

Abstract

Situs inversus is an abnormal placement of the thoracic and/or abdominal organs that are inverted right/left from normal. It is a rare congenital malformation often discovered in childhood. In adults, it can lead to misdiagnosis. **Clinical case:** A 35-year-old female patient, seen in a hepatogastroenterology consultation for acute spontaneous pain in the right hypochondrium. She had no particular clinical history. A diagnosis of hepatopathy was suspected. Abdominal and pelvic CT scans showed the left liver, stomach and spleen in the right hypochondrium, but with the heart in place, suggesting incomplete situs inversus. The evolution in our patient was spontaneously resolved with analgesics and antispasmodics, which leads us to believe that the volvulus was probably partial and without other complications. **Conclusion:** In developing countries, antenatal diagnosis of situs inversus is rare and is usually made during a pathology that leads the patient to a medical consultation. CT is one of the key paraclinical examinations for its diagnosis as genetic tests are not widely available.

Keywords

Situs Inversus, Abdominal Pain, Hypochondrium, Abdominal CT Scan

1. Introduction

Situs inversus, is also known as heterotaxis, means "orderly" (taxie) "different" (hetero). It is an abnormal placement of thoracic and/or abdominal organs that are inverted right/left in relation to normal. It was first described in humans by Fabricius in 1600 and its incidence is approximately 1 in 10,000 to 1 in 50,000

births per year [1].

It is a rather large group of anomalies as there are multiple possibilities of right/left inversion which can be complete (called situs inversus totalis or situs inversus: all organs normally on the right are on the left and vice versa), or partial (called situs inversus incomplete or situs ambigus: a limited number of organs are inverted, or a normally lateralized organ becomes median). A situs inversus is not in itself a problem, apart from the risk of diagnostic errors [2].

To this day, the etiology of situs inversus remains unclear. However, some authors suggest an autosomal recessive gene [3] [4]. Mutations have been identified in genes of patients with heterotaxy (LEFTY A, ACVR2B, NODAL, CFC1, INVERSINE) but the patients were all heterozygous (never compound heterozygous or homozygous). Autosomal recessive inheritance has only been demonstrated in the genetically modified mouse. This suggests that it is more a question of digenism in humans. Finally, a gene responsible for X-linked inheritance has been identified: it is the ZIC3 (Zinc-finger protein of cerebellum) gene located at Xq26.2.

Although this condition is rare, it should be considered in the presence of any digestive symptomatology. To our knowledge, no case of incomplete situs inversus of incidental discovery in adults has been described in our context. We therefore report the first case of incomplete situs inversus revealed by acute right hypochondrial pain.

2. Observation

A 35-year-old female teacher was seen in a hepatogastroenterology consultation for acute spontaneous pain in the right hypochondrium, radiating to the epigastrium, with torsion and sometimes burning, without any other accompanying signs and evolving for a week. She had no particular clinical history. The clinical examination noted a preserved general condition and pain provoked by palpation of the right hypochondrium. A diagnosis of hepatopathy was suspected. The stool culture and biological examinations results are presented in **Table 1**.

On ultrasound, a macronodular liver was found without the diagnosis of situs inversus. The abdomino-pelvic CT scan of 8 May 2021 invalidated the diagnosis of liver lesions but found an incomplete situs inversus on the common mesentery. Indeed, we noted a heterotaxy syndrome associating a polysplenia, the spleen was located on the right flank with the presence of 5 accessory spleens, all subhepatic (**Figure 1(A)**); an ambiguous situs, the spleen, the right liver, the gallbladder and the pancreas were abnormally located on the right, the stomach was in a right subhepatic position in the right hypochondrium, the heart was well placed on the left as well as the coeco-appendix and the right colon well placed on the right (**Figures 1(A)-(C)**); a pancreatic anomaly of the short pancreas type (**Figure 1(C)**). There was also malrotation of the mesenteric vessels with the superior mesenteric vein on the left and the mesenteric artery. Fifteen days after the onset of the symptomatology the pain had become diffuse throughout the abdomen,

Table 1. Results of stool and biological examinations performed on the patient.

Stool culture	Negative
Biological examinations	**Results**
white blood cells	400,000/mm^3
Hemoglobin	12 g/dl
platelets	155,000/mm^3
AST	43 IU/ml
ALT	47 IU/ml.
HBsAg	Negative
Ac anti HBc total	Negative
Ac anti HCV	Negative
HIV	Negative

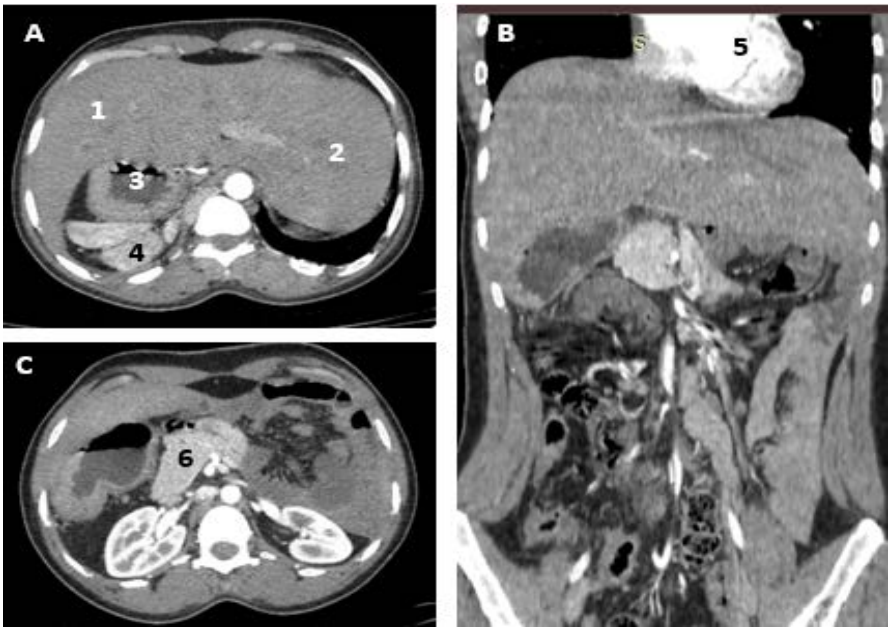

Figure 1. Abdominal-pelvic CT scan at late arterial time, axial sections (A) and (C) and coronal reformation (B). (A) Left liver (1) on the right; right liver (2) on the left; stomach (3) on the right; spleen (4) on the right. (B) Heart (5), located on the left. (C) Short pancreas (6) with head on the left and body on the right.

spasm-like at times, less intense and intermittent. A torsion of the mesentery had been suspected and the re-reading of the abdominal CT images confirmed this diagnosis. A whirl sign was found (**Figure 2(A)** and **Figure 2(C)**) which suggested a diagnosis of small bowel volvulus. There were no signs of associated complications. The small bowel was abutting the left abdominal wall, pushing the sigmoid colon medially (**Figure 2(C)**).

An upper GI fibroscopy performed on June 3, 2021 found no GI lesions other than an anatomical inversion with the greater curvature of the stomach on the

right and the lesser curvature on the left. The pylorus was also located on the left (Figure 3).

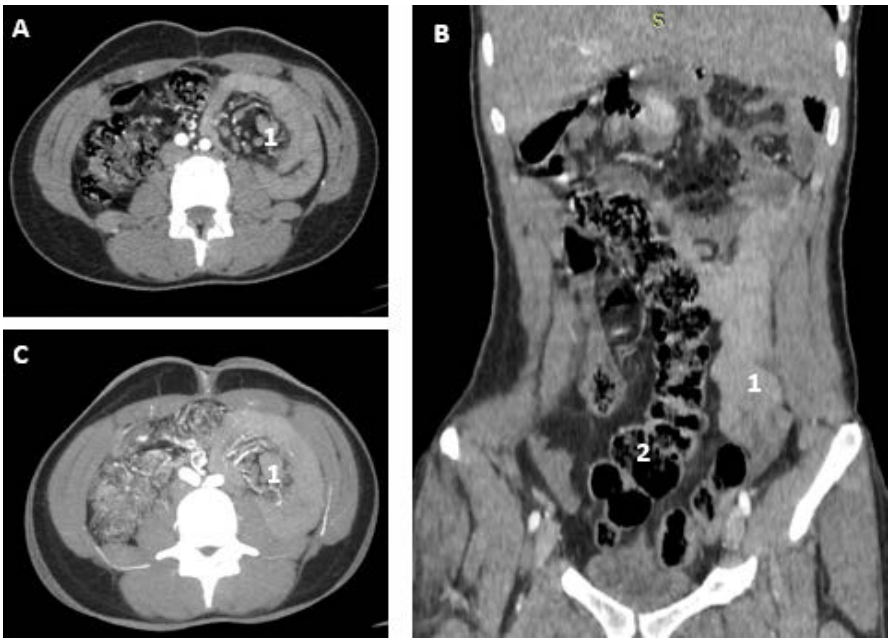

Figure 2. Abdominal-pelvic CT scan, late arterial time (45 s), axial slice (A), coronal reformation (B), MIP axial slice (C). (A) and (C) Whirl sign (1) suggestive of small bowel volvulus. (C) Small intestine (1) attached to the abdominal wall and pushing the sigmoid colon (2) medially.

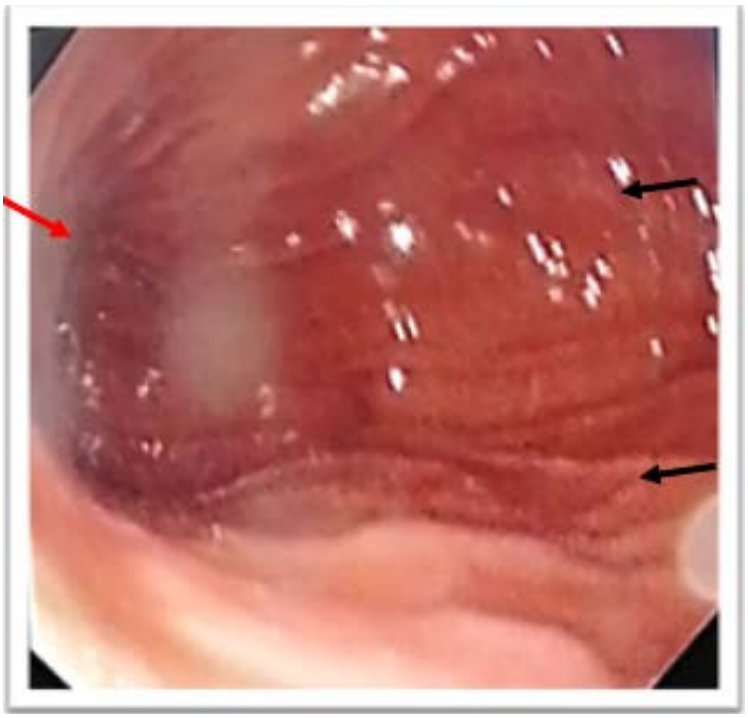

Figure 3. Endoscopic image showing an anatomical inversion with the pylorus on the left (red arrow), the great curvature of the stomach on the right with the fundic folds (black arrows).

The evolution was marked by an improvement of the symptomatology under analgesics and antispasmodics, 21 days after the beginning of the symptomatology. The patient reported no further abdominal pain episodes.

One week after the abdomino-pelvic CT scan, 15 May 2021, on tramadol 50 mg tablet: 1 tablet three times a day combined with spasfon lyoc 80 mg tablets: 2 tablets at the time of pain, to be repeated if necessary after 2 hours and not exceeding 6 per day, the patient presented a complete resolution of the symptomatology. The evolution in our patient was spontaneously resolved with analgesics and antispasmodics, which leads us to believe that the volvulus was probably partial and without other complications. However, close monitoring was instituted during the first week for fear of a resumption of the symptoms; then additional morphological assessment was considered (oesogastroduodenal transit, colonoscopy) in order to search for other malformations. However, due to lack of financial means and in the absence of additional symptoms, the patient preferred to carry out these examinations at a later date. Informed consent obtained from the patient to report this case and anonymity was respected.

3. Discussion

Incomplete situs inversus is a very rare congenital malformation in adults. Its prevalence is estimated to be between 0.2% and 0.5% [5] [6] and the most feared complication is total volvulus of the small intestine.

This malformation can be associated with any type of cardiac malformation, as well as with renal, biliary, midline, etc. anomalies. Asplenia or polyspenia are common. The term asplenia refers to a lateralization defect with a small or absent spleen. This would be a duplication of the right side or right isomerism (the right and left sides being identical to the image of the right side). The term polysplenia refers to a lateralization defect with several small spleens. This would be a left-sided duplication or left isomerism (the right and left sides being identical to the image of the left side) [7]. In our patient we found instead a polysplenia with 5 accessory spleens without any blood flow disorder. Other associated malformations were short pancreas and common mesentery. The common mesentery results from a rotational anomaly of the digestive tract [8]. The fact that this condition is exceptional in adults and that its symptomatology is quite varied is a source of multiple errors and diagnostic and therapeutic delay to the extent that the majority of cases are diagnosed post-mortem [9].

Failure to recognise the presence of situs inversus in a patient beforehand can lead to significant diagnostic errors. When the diagnosis is made prior to any symptomatology, it reduces the risk of diagnostic errors. In our patient, the usual diagnostic approach was compromised by the lack of knowledge of this underlying anomaly. Indeed, chronic liver disease was suggested in view of the frequency of this condition in young subjects presenting with right hypochondrial pain in our setting. Although situs inversus is most often asymptomatic in adults, the association with a malformation such as common mesentery predis-

poses to the occurrence of small bowel volvulus as in our patient.

The diagnosis of small bowel volvulus can be made in an emergency in the presence of an acute intestinal obstruction, or even a state of shock, or in the presence of repeated abdominal pain often associated with transit disorders. This is a dreaded complication of the common mesentery, which is defined as a rotation anomaly of the digestive tract and is very rare in adults [10]. CT is the gold standard for the diagnosis of this condition and its complications.However, the performance of the radiological examination must not delay the therapeutic management.

The "swirl" sign appears to be pathognomonic for most authors [11] and corresponds to torsion of the mesentery. Contrast injection visualises the verticality, or inversion, of the superior mesenteric vessels, with a vein located above or to the left of the artery [12], although this sign is not consistent. The thickness of this swirling mass would be proportional to the degree of rotation of the volvulus, but it is more accurate to assess the degree of rotation by calculating the number of turns made by the mesenteric vessels [13].

In our patient, the presence of this sign led to the diagnosis of small bowel volvulus. However, this volvulus seems partial and not very important. This would explain the spontaneous resolution of the painful crisis without any surgical intervention.

Doppler ultrasonography is of particular interest in this condition.

In adults, it is often hampered by gas and is not always helpful in the diagnosis, but its sensitivity is 86.5%, its specificity 74.7%, its positive predictive value 42.1% and its negative predictive value 96.3% [14]. In our patient, ultrasound did not find a volvulus, so gastric pathology was sought on upper GI fibroscopy.

In addition to CT and ultrasound, the most accessible imaging modality in our context is the unprepared abdominal X-ray (UXR). It is requested as a first-line examination for occlusive syndrome, which is often present in small bowel volvulus [10]. The PSA may show hydro-aerosic levels of the small bowel in relation to a small bowel obstruction. It may not show any specific signs of volvulus on incomplete common mesentery [10].

This examination was not requested in our patient as the occlusive syndrome was not the primary symptom.

When volvulus is suspected, urgent surgical intervention is required to avoid ischaemic necrosis or perforation of the bowel, but also peritonitis, sepsis and death [15] [16] [17]. An exploratory laparotomy can be performed to confirm the diagnosis and guide decision making [18]. Most authors agree that resection is necessary in cases of necrotic bowel [15] [16] [17] [19] [20] [21]. In the absence of necrosis, if the bowel appears oedematous or congested, simple derotation, with or without fixation of the involved small bowel, can be considered. However, this procedure is associated with recurrence of VAS [21]. Published mortality rates vary, but the consensus is 10% - 35% [15] [16] [19] [21]. Patient-specific factors, such as age, comorbidities and general health, play a role in deciding which treatment option to pursue [19]. In our patient, close monitoring

should be instituted.

The course in our patient resolved spontaneously with analgesics and antispasmodics, suggesting that the volvulus was probably partial and without further complications. This type of symptomatology, with intermittent abdominal pain without disturbance of balance, can also be confused with functional bowel disorders among others.

The prognosis of our patient is good, provided that she is closely monitored with immediate management in case of recurrence of the symptomatology. The outcome in case of recurrence depends strongly on the speed of management and the general health.

4. Conclusion

Situs inversus should be diagnosed prenatally or in childhood to avoid misdiagnosis in adults. However, in developing countries, its diagnosis is often incidental. Careful examination of patients should lead to suspicion and confirmation by imaging.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Ouédraogo, N.L.M., Sanogo, K., Traoré, M.S., Simporé, J. and Traoré, S.S. (2020) A Case Report of Management of Gastric Perforation in Situs Inversus Totalis in a 45-Year-Old Adult. *A Case Report. International Journal of Surgery Case Reports*, **74**, 69-72. https://doi.org/10.1016/j.ijscr.2020.07.072

[2] Huh, J.W., Kim, H.R., Cho, S.H., Kim, C.Y., Kim, H.J., Joo, J.K., *et al.* (2010) Laparoscopic Total Mesorectal Excision in a Rectal Cancer Patient with Situs Inversus Totalis. *Journal of Korean Medical Science*, **25**, 790-793. https://doi.org/10.3346/jkms.2010.25.5.790

[3] Postema, M.C., Carrion-Castillo, A., Fisher, S.E., Vingerhoets, G. and Francks, C. (2020) The Genetics of Situs Inversus without Primary Ciliary Dyskinesia. *Scientific Reports*, **10**, 1-11. https://doi.org/10.1038/s41598-020-60589-z

[4] Budhiraja, S., Singh, G., Miglani, H.P. and Mitra, S.K. (2000) Neonatal Intestinal Obstruction with Isolated Levocardia. *Journal of Pediatric Surgery*, **35**, 1115-1116. https://doi.org/10.1053/jpsu.2000.7839

[5] Gamblin, T., Stephens, R., Johnson, R. and Rothwell, M. (2003) Adult Malrotation: A Case Report and Review of the Literature. *Current Surgery*, **60**, 517-520. https://doi.org/10.1016/S0149-7944(03)00030-8

[6] Jarry, J., Razafindratsira, T., Bodin, R., Lepront, D. and Durand-Dastes, F. (2008) À propos d'un cas de mésentère commun complet de l'adulte révélé par une complication occlusive. *La Presse Médicale*, **37**, 1689-1692. https://doi.org/10.1016/j.lpm.2008.05.016

[7] Lee, S.E., Kim, H., Jung, S., Lee, S., Park, K. and Kim, W. (2006) Situs Anomalies and Gastrointestinal Abnormalities. *Journal of Pediatric Surgery*, **41**, 1237-1242. https://doi.org/10.1016/j.jpedsurg.2006.03.045

[8] Peycelon, M. and Kotobi, H. (2012) Complications des anomalies embryologiques de la rotation intestinale: Prise en charge chez l'adulte. *EMC - Techniques chirurgicales - Appareil digestif*, **29**, 1-12.

[9] Coulibaly, M., Boukatta, B., Derkaoui, A., Sbai, H., Ousadden, A. and Kanjaa, N. (2015) Incomplete Small Bowel Volvulus on Common Mesentery—A Severe and Rare Complication in Adults: Report of 1 Case. *The Pan African Medical Journal*, **20**, Article No. 157. https://doi.org/10.11604/pamj.2015.20.157.4182

[10] Alaoui, A., Alami, B., Lamrani, Y.A., Boubou, M. and Maaroufi, M. (2020) Imagerie du volvulus du grêle sur mésentère commun incomplet chez un adulte: Rapport de cas [Small Bowel Volvulus Complicating Common Incomplete Mesentery in an Adult: A Case Report]. *The Pan African Medical Journal*, **37**, Article No. 287. https://doi.org/10.11604/pamj.2020.37.287.17538

[11] Geoghegan, J.G., Maher, M., McNally, O.M. and Kirwan, W.O. (1994) Acute Midgut Volvulus Following Laparoscopic Cholecystectomy. *Surgical Endoscopy*, **8**, 903. https://doi.org/10.1007/BF00843467

[12] Lay, P.S., Tsang, T.K., Caprini, J., Gardner, A., Pollack, J. and Norman, E. (1997) Volvulus of the Small Bowel: An Uncommon Complication after Laparoscopic Cholecystectomy. *Journal of Laparoendoscopic & Advanced Surgical Techniques*, **7**, 59-62. https://doi.org/10.1089/lap.1997.7.59

[13] Vricella, L.A., Barrett, W.L. and Tannebaum, I.R. (1999) Intestinal Obstruction from Midgut Volvulus after Laparoscopic Cholecystectomy: A Report of an Unusual Complication. *Surgical Endoscopy*, **13**, 1234-1235. https://doi.org/10.1007/PL00009628

[14] Orzech, N., Navarro, O.M. and Langer, J.C. (2006) Is Ultrasonography a Good Screening Test for Intestinal Malrotation? *Journal of Pediatric Surgery*, **41**, 1005-1009. https://doi.org/10.1016/j.jpedsurg.2005.12.070

[15] Lepage-Saucier, M., Tang. A., Billiard. J.-S., Murphy-Lavallée, J. and Lepanto, L. (2010) Small and Large Bowel Volvulus: Clues to Early Recognition and Complications. *European Journal of Radiology*, **74**, 60-66. https://doi.org/10.1016/j.ejrad.2009.11.010

[16] Roggo, A. and Ottinger, L.W. (1992) Acute Small Bowel Volvulus in Adults. A Sporadic form of Strangulating Intestinal Obstruction. *Annals of Surgery*, **216**, 135-141. https://doi.org/10.1097/00000658-199208000-00003

[17] Katis, P.G. and Dias, S. (2004) Volvulus: A Rare Twist on Small-Bowel Obstruction. *Canadian Medical Association Journal*, **171**, 728. https://doi.org/10.1503/cmaj.1040662

[18] de Korte, N., Grutters, C.T. and Snellen, J.P. (2008) Small Bowel Volvulus Diagnosed by the CT 'Whirl Sign'. *Journal of Gastrointestinal Surgery*, **12**, 1469-1470. https://doi.org/10.1007/s11605-007-0346-1

[19] Ruiz-Tovar, J., Morales, V., Sanjuanbenito, A., Lobo, E. and Martinez-Molina, E. (2009) Volvulus of the Small Bowel in Adults. *American Surgeon*, **75**, 1179-1182. https://doi.org/10.1177/000313480907501206

[20] Huang, J.-C., Shin, J.-S., Huang, Y.-T., Chao, C.-J., Ho, S.-C., Wu, M.-J., *et al.* (2005) Small Bowel Volvulus among Adults. *Journal of Gastroenterology and Hepatology*, **20**, 1906-1912. https://doi.org/10.1111/j.1440-1746.2005.03945.x

[21] Grasso, E. and Sciolli, L. (2011) Spontaneus Small Bowel Volvulus in an Adult. Case Report and Review of the Literature. *Annali Italiani di Chirurgia*, **82**, 413-416.