JBR–BTR, 2012, 95: 257-260.

COMBINATION OF PARTIAL SITUS INVERSUS, POLYSPLENIA AND ANNULAR PANCREAS WITH DUODENAL OBSTRUCTION AND INTESTINAL MALROTATION

Y. Ben Ahmed[1], S. Ghorbel[1], T. Chouikh[1], F. Nouira[1], H. Louati[2], A. Charieg[1], B. Chaouachi[1]

Situs inversus is a challenge to the physician, both for the diagnostic and for the therapeutic. A combination of partial situs inversus, annular pancreas and polysplenia with bowel malrotation has been reported in a newborn who presented as duodenal obstruction. Situs inversus is rare especially without heart malformation. All the abnormalities in this combination can be explained on the basis of multiple organ malrotation. It also demonstrates the necessity of meticulous investigation and precise description of the anomaly as far as the management of this case is concerned.

Key-word: Infants, newborn.

Situs inversus is a lateralization anomaly that can be associated to other malformations that alter prognosis: isolated cardiac malformations, cardiosplenic syndromes, or Kartagener's syndrome (1).

The location of the thoracic and abdominal viscera in cases with situs inversus has been determined to be everywhere between the normal and the mirror image place. The pathogenesis of heterotaxia has not been clarified yet. Genetic and environmental factors have been reported as possible causes.

We report the case of partial situs inversus, polysplenia and annular pancreas with duodenal obstruction and malrotation. We tried to explain the probable embryogenesis and give detailed description of the anatomy and clinical features of the syndrome.

Case report

An 18-day-old girl, who weighed 2.800 kg at birth, was admitted for non bilious vomiting since the age of 8 days. Parents report a rarefaction of stool. No Antenatal Ultrasounds were available.

Physical examination revealed a normally placed apex beat on the left side of the chest but the liver was palpated in the left upper abdominal quadrant.

The blood tests were normal. Investigation included a plain radiograph of the abdomen, which shows deviation of the nasogastric tube to the right (Fig. 1). That raised the question of inversion of the abdominal viscera. The investigation was completed by upper abdominal opacification (Fig. 2), abdominal ultrasonography and echocardiogram.

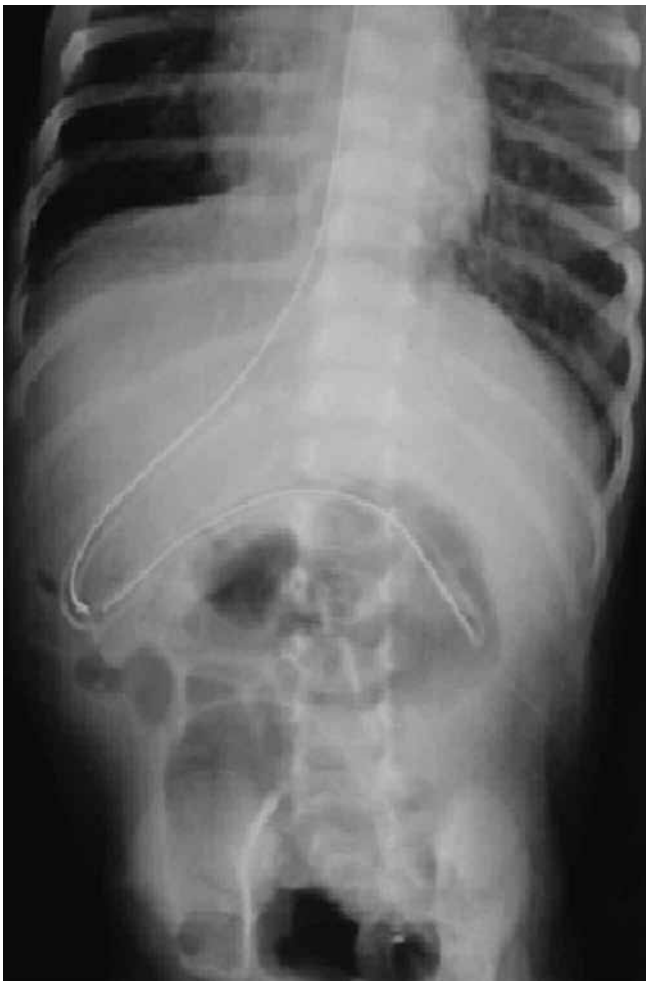

Fig. 1. — Abdominal X-Ray shows deviation to the right of the nasogastric tube.

Abdominal ultrasound was not able to visualize the inferior vena cava that is why a CT scan was performed (Fig. 3). Unfortunately MRI was not available in our center.

The results of the examinations supported the preoperative diagnosis of levocardia associated with inversion of the abdominal viscera

From: Department of 1. Paediatric Surgery B, and 2. Paediatric Radiology, Children Hospital of Tunis, Tunis, Tunisia.
Address for correspondence: Dr T. Chouikh, M.D., Department of Paediatric Surgery B, Children Hospital of Tunis, Place Bab Saadoun Jabary 1007 Tunis, Tunisia.

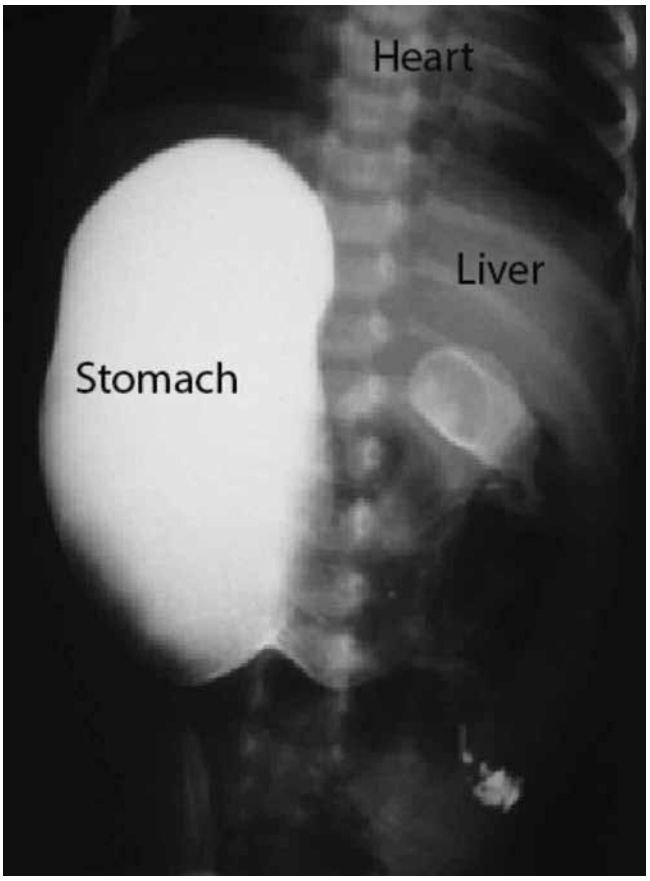

Fig. 2. — Upper abdominal opacification shows stomach in the right area of the abdomen.

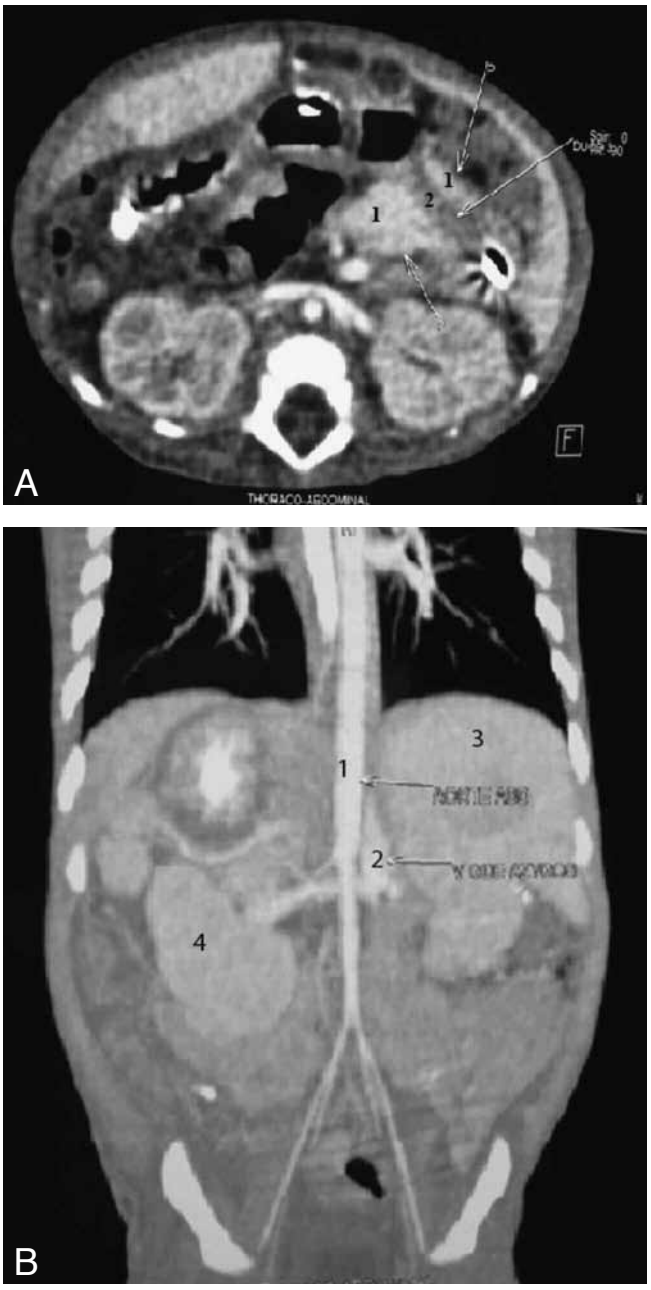

Fig. 3. — A. Abdominal CT: Situs inversus, stenosis of the second portion of duodenum (2) annular pancreas (1).

B: Abdominal CT: interruption of the inferior cave vein in the retrohepatic portion. 1. Aorta, 2. Interrupted inferior vena cava, 3. Liver, 4. Kidney.

with right polysplenia and annular pancreas. Situs inversus was associated to a stenosis of the second portion of duodenum. This part of the duodedum was encircled by the annular pancreas. The inferior cave vein wasn't visible neither on ultrasonography nor on CT, it was interrupted at the retrohepatic portion to be continued into the azygos vein.

The hepatic vein formed a suprahepatic duct that drained into the systemic venous atrium. A bilateral superior vena cava was noticed. The echocardiogram demonstrated that there was a muscular ventricular septal defect and a little mitral insufficiency.

The baby was operated 6 days later, after adequate reanimation.

The intraoperative findings consisted of duodenal obstruction due to an incomplete intraluminal diaphragm of the second duodenum; left-sided liver, right-sided polysplenia and stomach. Multiple spleens were found along the greater curvature of the stomach in the right upper quadrant varying in size from 0.5 to 1.5 cm;

Totally right-sided located colon in position like a mirror image of nonrotation with congenital bands on small bowel;

Abnormal position of the inferior vena cava to the left side was also noted.

After the investigation a longitudinal duodenotomy with diaphragm resection was performed then we close the duodenotomy with a continuous suture helped by a transanastomic gastric tube. LADD's procedure was performed, including division of peritoneal bands and placement of bowel in a state of nonrotation.

Immediately postoperative course was uneventful; the patient tolerated a regular diet on the fifth day and was discharged nine days after surgery. The echocardiogram done at the age of 6 months was normal.

The patient was hospitalized at the age of 7 months for bowel occlusion due to intestinal adhesion.

She underwent surgery after failure of gastric aspiration. There was no need for intestinal resection, Follow-up after this second surgery was normal.

The infant is doing well at age 12 months. She is regularly seen in consultation with a good weight intake and no alimentary or transit problems.

Discussion

Situs inversus viscerum, also termed heterotaxia, is a condition where the position of the organs is a perfect mirror image of their normal physiological position (1). It is divided into total and partial. In total situs inversus the heart is to the right of the midline, the cardiac chamber position is reversed, the aorta courses to the right, there are three left and two right lung lobes, the thoracic duct is on the right, the liver and stomach are transposed, the colon flexures are reversed, and the spleen is on the right. In partial situs inversus, there may be thoracic inversion only, cardiac chamber reversal or abdominal inversion only, and there are frequently syndromes associated with splenic anomalies, annular pancreas, horseshoe kidney, diaphragmatic hernia, or a number of other developmental abnormalities (2).

The true incidence of situs abnormalities is unknown. Some estimates suggest that situs inversus occurs in approximately 1 per 8000 to 25,000 live births (2).

The reported sex distribution of the disease is equal for males and females; when there are associated anomalies of the spleen, boys predominate.

The pathogenesis of heterotaxia has not been clarified yet. Genetic and environmental factors have been reported as possible causes. Von Woelworth reported that some cases could be attributed to early embryonic factors (3). Situs inversus is generally an autosomal recessive genetic condition, although it can be X-linked or found in identical "mirror" twins.

The partial situs inversus being described is probably caused by reversal of location in terms of left and right, of the duodenum, pancreas, liver, stomach, and spleen, as a consequence of disturbances of rotation early in embryonic life (4), precisely between the fifth and sixth gestational week. It is conceivable that a single, or less likely, more than one event occurring at this period can interfere with these rotational activities and cause an absence of rotation of an organ or an incomplete rotation, or perhaps to rotate in the opposite direction. So, anomalies of gut fixation and rotation are not difficult to understand. Various theories have been advanced to explain the formation of the annular pancreas. Perhaps the most popular is that advanced by Lecco, which suggests fixation of the tip of the ventral pancreatic primordium to the duodenum during the 180° rotation of its base to fuse with the dorsal pancreatic bud.

Diversity of structural lesions, and the absence of congenital cardiac malformations suggest a less specific timing of the teratogenic insult than has been postulated in the past, Campbell and Deuchar have suggested that the initial abnormality may be situs inversus which leads to and is responsible for the other malformations (5).

Situs inversus occurs more commonly with dextrocardia. A 3-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia, usually with transposition of the great vessels. Situs inversus with levocardia is rare, and it is almost always associated with congenital heart disease (6). In the absence of congenital heart defects, individuals with situs inversus can lead normal healthy lives.

The functional malformations associated with situs inversus are primarily cardiovascular with severe cardiac malformations such single ventricle, single atrium, pulmonary atresia, ventricular septal defect, transposition of great arteries, truncus arteriosus, mitral stenosis, complete heart block, tricuspid astresia, atrioventricular canal, anomalous pulmonary venous drainage.

Although significant visceral or structural deformities can occur, a review of all cases reported in the literature confirms the presence of significant gastrointestinal pathologies, mainly annular pancreas but also congenital obstruction of the gastrointestinal tract secondary to stenosis or atresia of the duodenum or jejunum or incomplete fixation of the bowel mesentery that can be complicated by a midgut volvulus (7). Preduodenal portal vein which is a rare cause of duodenal obstruction should be borne in mind to avoid inadvertent damage during surgery (8).

The majority of the cases are associated to splenic defect. Polysplenia syndrome is usually characterized by one multilobulated or a number of individual splenules. The most common sites in which extra splenic tissue has been found are the splenic hilum, the splenic pedicle, the peritoneal ligaments and in the tail or head of the pancreas (9).

Venous malformations like bilateral superior venae cavae, the absence of a segment of the inferior vena cava were described. Anomalous of hepatic venous drainage is a frequent finding in cases of levocardia. It has been suggested that the abnormal hepatic veins represent persistent omphalomesenteric veins (10).

So, the combination of annular pancreas and partial situs inversus associated with duodenal obstruction nonrotation is rare. The originality of this case is also due to absence of major cardiac anomalies and the presence of hepatic venous drainage.

Clinical manifestations depend on the individual's specific anatomical anomalies. Severe and complex cardiac abnormalities are likely to be apparent at birth or soon afterward. Individuals without congenital heart disease may present later with conditions such as biliary atresia or volvolus. In persons without clinically morbid abnormalities, these conditions may be discovered during an evaluation for an unrelated problem or complaint when they are adults (2).

Diagnosis of a situs inversus abdominus by physical examination is difficult. Transposition of visceral organs is more difficult to detect on physical examination than dextrocardia, by palpating the liver on the left and the spleen on the right.

In patients with situs inversus viscerum presenting with an acute illness, localization of the abdominal pain can be confusing. Organ reversal can occur without concomitant reversal of the sensory nerve pathway. Non reversal of the pain pathways would explain the occasional case in which symptoms of cholecystitis become manifest in the right upper quadrant or those of appendicitis in the right lower quadrant even though the involved organ is located on the opposite side (11).

Situs abnormalities may be recognized first by using radiography or ultrasonography.

The interruption of the inferior vena cava with azygos or hemiazygos continuation may be suggested after the evaluation of plain chest radiographs showing an absence of an inferior vena cava on the lateral projection and a widening of the

paravertebral pleural reflection in the right or left paravertebral area because of a dilated azygos or hemiazygos vein. A plain radiograph of the abdomen showing a reversed double bubble confirms the diagnosis. Ultrasound and CT scans are the most accurate noninvasive means of diagnosing the abnormal (12).

However, computed tomography (CT) scanning is preferred for the definitive diagnosis of situs inversus. It provides good anatomic details of organ position, cardiac apical position, and great vessel branching and precise the localization of splenules in order to avoid hemorrhage during surgery. Magnetic resonance imaging (MRI) is usually reserved for difficult cases or for patients with associated cardiac anomalies (13).

Moreover prenatal diagnosis of situs anomalies by obstetric ultrasonography was mostly reported in conjunction with congenital heart disease: location of the stomach; liver lateralization; interruption of the Vena Cava associated to abnormal azygos return and visualization of the spleen (14).

The rational approach to a patient with situs inversus abdominus includes the collection of as much preoperative information as possible using sonography and computerized scanning. To prevent incorrect preoperative diagnoses and inappropriate incision placement, such studies are critical to delineate intra-abdominal pathology; but in addition, such information might also provide important data relative to life-threatening associated cardiac disease (1).

Detection of a malformation must lead to search for other associated anomalies.

We consider that any situs anomaly should warrant bowel malrotation exclusion shortly after birth because of their potential association and risk of midgut volvolus.We should also be attentive to an eventual duodenal or jejuna obstruction in case of situs inversus.

Conclusion

Situs inversus is a rare malformation; however it should be suspected and searched through clinical and radiological signs in case of intestinal obstruction. Ultrasound exam and CT are of great help for meticulous investigation and detailed description of the anatomic futures in all the cases of situs inversus. This is of great importance for the management of the patient and useful in case of surgery.

References

1. Cheikhelard A., De Lagausie P., et al.: Situs Inversus and Bowel Malrotation: Contribution of Prenatal Diagnosis and Laparoscopy. *J Pediatr Surg*, 2000, 35: 1217-1219.
2. Winer-Muram H.T.: Adult presentation of heterotaxic syndromes and related complexes. *J Thorac Imaging*, 1995, 10: 43-57.
3. Seung E.L., Hyun-Young K.: Situs anomalies and gastrointestinal abnormalities. *J Pediatr Surg*, 2006, 41: 1237-1242.
4. Tryfonas G.I., Chaidos C., Avtzoglou P.P.: Partial Situs Inversus: Duodenal Obstruction in a Neonate with Isolated Levocardia. *J Pediatr Surg*, 1992, 27: 1584-l586.
5. Debo Adeyemi S.: Combination of Annular Pancreas and Partial Situs Inversus: A Multiple Organ Malrotation Syndrome Associated With Duodenal Obstruction *J Pediatr Surg*, 1988, 23: 188-191.
6. Maldjian P.D., Saric M.: Approach to dextrocardia in adults. *AJR*, 2007, 188: S39-49; quiz S35-38.
7. Mordehai J., Cohen Z.: Preduodenal Portal Vein Causing Duodenal Obstruction Associated With Situs Inversus, Intestinal Malrotation, and Polysplenia: A Case Report. *J Pediatr Surg*, 2002, 37 (4): E5.
8. Koonosuke N., Fumio K.: Digestive Tract Disorders Associated With Asplenia/Polysplenia Syndrome. *J Pediatr Surg*, 1997, 32: 91-99.
9. William J., Futrell S.: Cecal Volvulus Associated with Incomplete Situs Inversus and Splenic Dysgenesis. *Am J Surg*, 1874, 128: 421-426.
10. Paddock R.J., Arensman R.M.: Polysplenia Syndrome: Spectrum of Gastrointestinal Congenital Anomalies. *J Pediatr Surg*, 1982, 17: 563-566.
11. Shlomo S., Egon R., et al.: Levocardia with partial situs inversus, an incidental finding in a 15 year-old boy. *Am Heart J*, 1962, 699-704.
12. Applegate K.E., Goske M.J.: Situs revisited: imaging of the heterotaxy syndrome. *Radiographics*, 1999, 19: 837-852.
13. Silverman N.H.: An ultrasonic approach to the diagnosis of cardiac situs, connections, and malpositions. *Cardiol Clin*, 1983, 1: 473-486.
14. Tonkin I.L., Tonkin A.K.: Visceroatrial situs abnormalities: sonographic and computed tomographic appearance. *AJR*, 1982, 138: 509-515.