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A Rare Cause of Recurrent Constipation With Abdominal Pain and Distension

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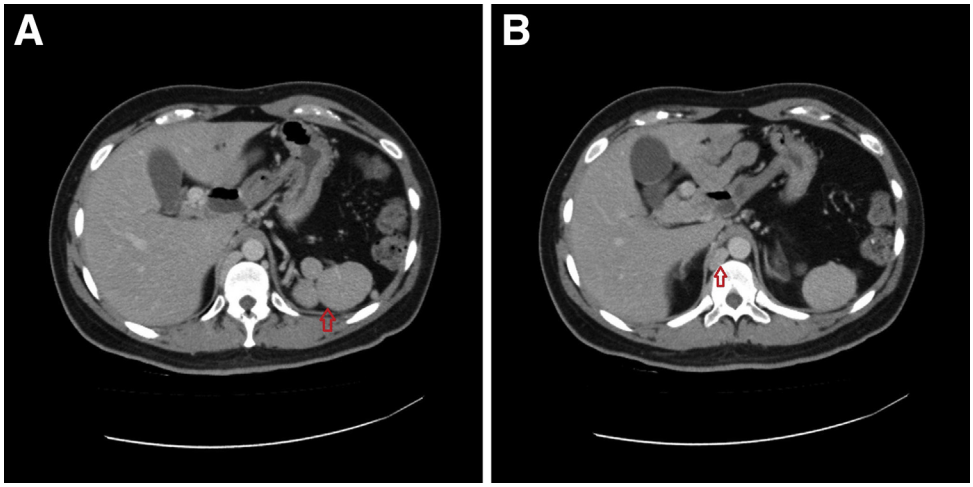

Question: A 38-year-old man presented with a 1-day history of constipation, abdominal distension, and central abdominal pain. He had similar episodes in the past and there was no history of weight loss, vomiting, or bleeding per rectum.

Past medical history was insignificant except for an investigation for infertility. Physical examination showed a mild diffusely tender and distended abdomen. There was no clinically demonstrable visceromegaly and other systemic examinations were also unremarkable. His complete blood count, blood biochemistry and thyroid profile were within normal limit. A computed tomography scan of the abdomen showed unusual findings (Figure *A*, *B*).

Describe the abnormalities on the computed tomography scan. What is the diagnosis?

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Conflicts of interest
The authors disclose no conflicts.

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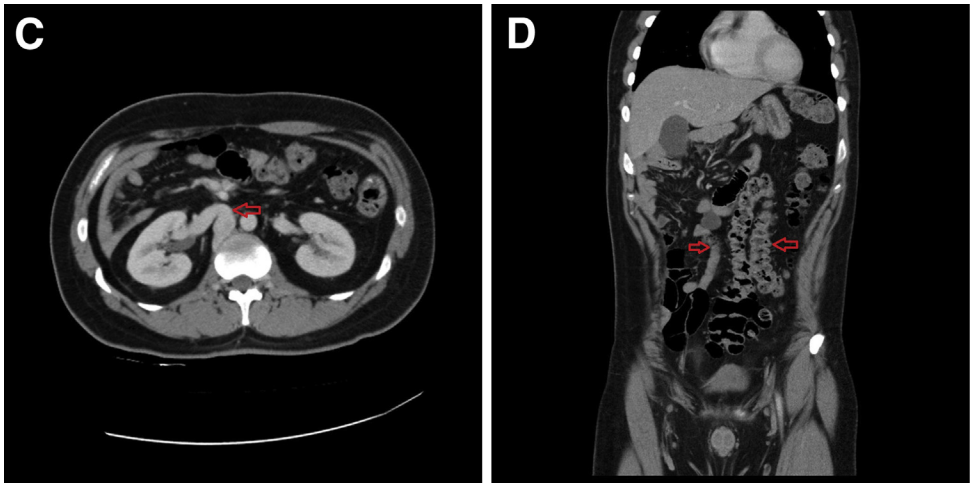
Answer to: Image 5: Visceroatrial Heterotaxy Situs Ambiguous With Polysplenia Syndrome



Postcontrast axial computed tomography scanning showed polysplenia (Figure *A*), interrupted inferior vena cava and dilated hemiazygous vein, dilated hemiazygous vein (Figure *B*) joining the distal inferior vena cava and renal hilum level (Figure *C*). Additionally, a postcontrast coronal reformatted image shows malrotation, left-sided large bowel, and right-sided small bowel loops (Figure *D*). These findings are consistent with visceroatrial heterotaxy situs ambiguous and polysplenia syndrome.

There is no single set of abnormalities that justify the criteria of polysplenia syndrome or situs ambiguous. This complex and controversial entity has no fixed pathognomic features. There is abnormal arrangement of viscera and blood vessels contrast with orderly arrangement in case of situs inversus.[1]

Interrupted inferior vena cava, dilated tortuous azygos vein, nonrotation of the small intestine along with multiple spleens (2-8 in number) are quite frequently seen in this syndrome. Annular pancreas, portal vein abnormalities, a right-sided stomach, and centrally placed liver are also reported with this syndrome. Some other variants of this syndrome are associated cardiopulmonary abnormalities as well as congenital heart diseases.

Human studies have identified several gene mutation notably CFC 1, and SHROOM3 in patients with heterotaxy syndrome.[2] This is not a premalignant condition; however, an association with hepatocellular and rectal carcinoma has been reported.[3]

References

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