

Case Report

**CASE OF HETEROTAXY SYNDROME WITH POLYSPLENIA
AND INTESTINAL MALROTATION**

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ABSTRACT

Heterotaxy syndrome is an abnormal, complex arrangement of heart and visceral organs along right-left axis of the body. Many patients present in early life due to associated congenital heart defects and gut malrotation with midgut volvulus. We are reporting a case of Heterotaxy syndrome with polysplenias and intestinal malrotation and other abnormalities.

Keywords: *Truncated Pancreas, Intestinal Malrotation, Polysplenias, Azygos Continuation of IVC*

CASES

21 years aged male patient presents with recurrent colicky abdominal pain since childhood. He is not a known case of diabetes mellitus, tuberculosis or any medical and surgical problems. His haemoglobin was 10.9gm%, TLC of 9000/mm³, blood urea 45mg/dl and serum creatinine of 0.8. Abdominal sonography showed liver and gallbladder in left side and stomach and multiple spleens in right side. Echocardiography showed dextrocardia with no evidence of any major abnormalities.

Oral and Intravenous contrast enhanced Computed Tomography of thorax and abdomen was done using Seimens CT scanner. Scanogram shows dextrocardia with cardiac apex towards right side and absence of stomach gas shadow (figure 1). Coronal CECT demonstrates cardiac apex and stomach in left side and side to side liver (figure 2). Multiple spleens are seen along greater curvature of stomach in right side with multiple branches of splenic vessels (Figure 3 and 4). Truncated or absence of dorsal part of pancreas noted, gallbladder is seen under liver in left side (figure 4). There is interruption of IVC at the level renal vessels and azygos vessels continuing above diaphragm and drain to Superior vena cava (figure 5 and 8). There is intestinal malrotation with duodenojejunal flexure in right side and ileocaecal junction in left (figure 6). Bilateral bilobed lung is seen with hypoarterial bronchi (Figure 9 and 10).



Figure 1: Scanogram Showing Dextrocardia and Absence of Stomach Fundic Gas Shadow

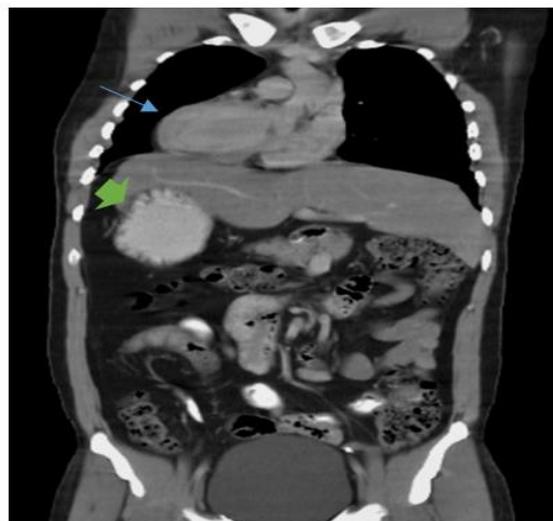


Figure 2: Coronal CECT Demonstrating Cardiac Apex (Arrow) and Stomach in Right Side (Arrow Head)

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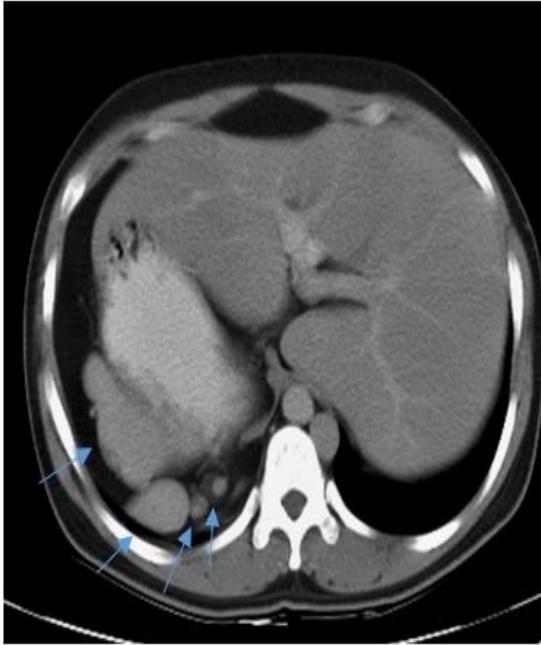


Figure 3: Axial CECT Showing Multiple Foci of Spleen in Right Side and IVC Interrupted Azygos Vein in Left Side

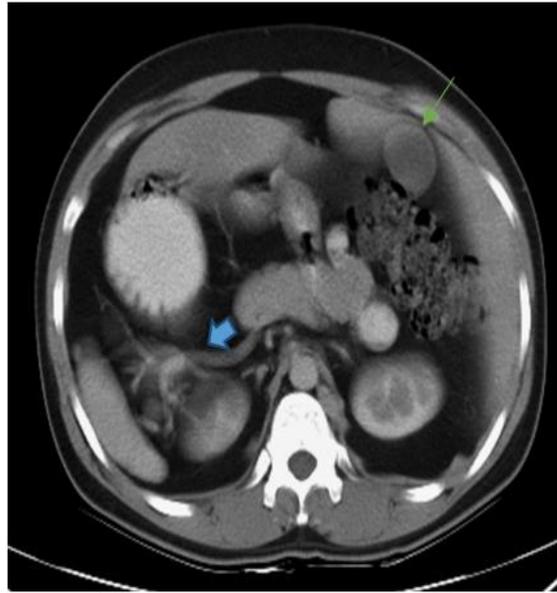


Figure 4: Axial CECT Showing Gallbladder (Arrow) to Left Side and Spleen with Multiple Branching of Splenic Artery in Right Side (Arrow Head); Truncated Pancreas is Seen

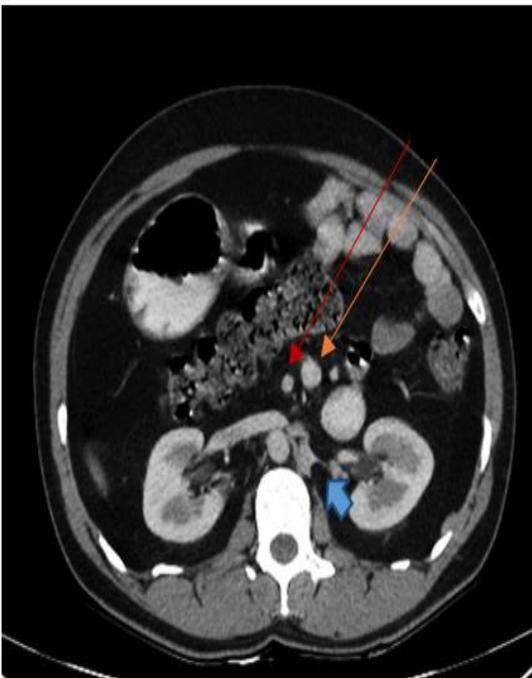


Figure 5: Showing Superior Mesenteric Artery and Vein, Interrupted IVC Continuing as Azygos Vein (Arrow Head)

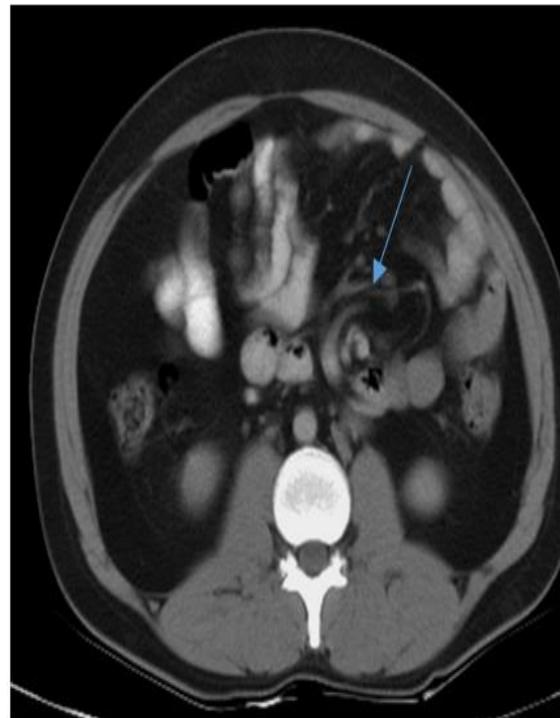


Figure 6: Twisting of Mesentery with Vessels (Arrow)

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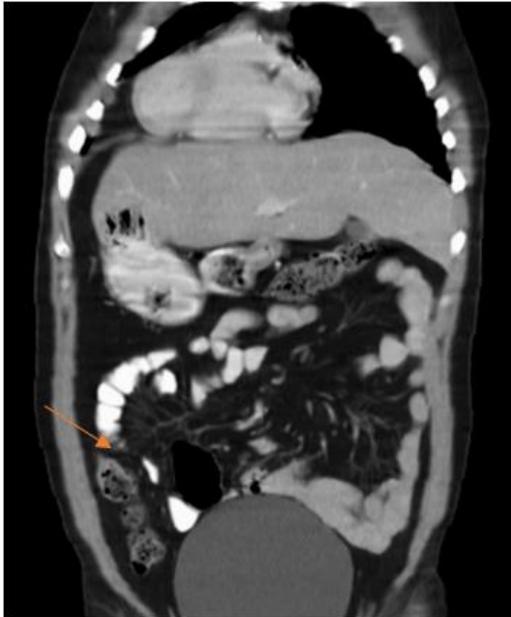


Figure 7: Small Bowel Loops (Arrow) in Right Iliac Region and Large Loops in Left Iliac Fossa



Figure 8: Sagittal CECT Showing Agenesis of Suprarenal IVC and Azygos Vein Seen above Diaphragm (Arrow)

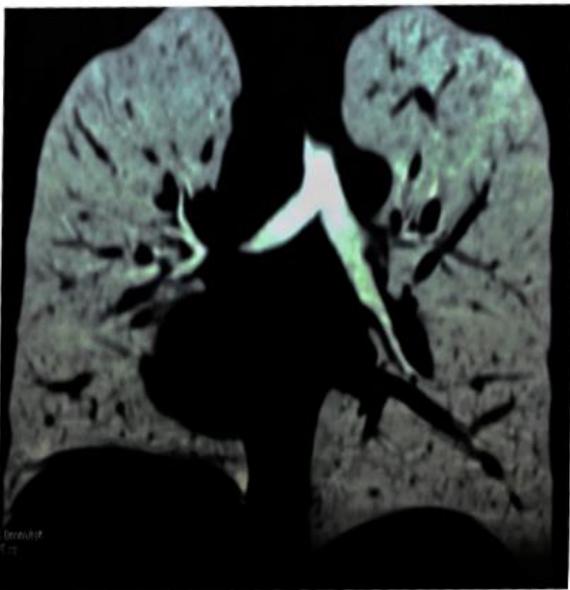


Figure 9: Bilateral Hyperarterial Bronchi

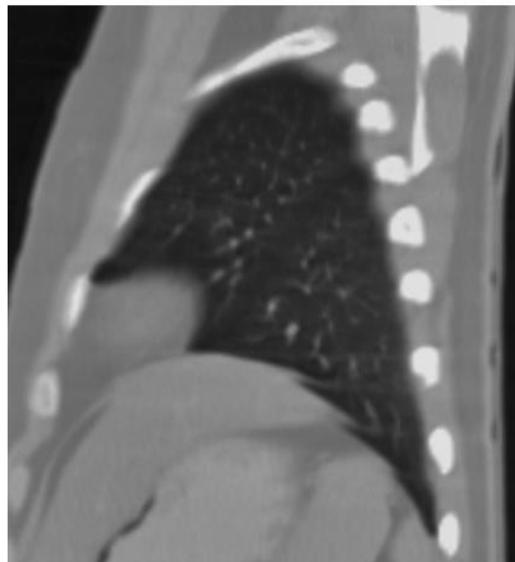


Figure 10: Right Lung Showing Single Oblique Fissure

DISCUSSION

The word heterotaxy is derived from Greek: heteros-meaning other than, and taxis-meaning arrangement (1). It is an abnormality where the internal thoraco-abdominal organs demonstrate abnormal and complex arrangement across the left-right axis of the body. The term situs refers to the arrangement of heart and other viscera relative to midline. Situs solitus is normal usual location of heart and visceral organs with heart, spleen, aorta and bilobed lung located on left and liver, gallbladder,

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inferior vena cava, trilobed lung located in right side of midline. Situs inversus is a complete transposition or mirror-image of heart and viscera. There are two subcategories of situs inversus: situs inversus with dextrocardia (Situs inversustotalis) where cardiac apex, spleen, stomach and aorta are located on right and situs inversus with levocardia where there is mirror image of viscera with left sided cardiac apex.

Heterotaxy syndromes have components of both situs solitus and situs inversus in the same person in variable proportions. The incidence of heterotaxy syndrome is about 1 in 10,000 births with male to female ratio of 2:1 (Applegate *et al.*, 1999; Lin *et al.*, 2000). It can be divided into 2 broad groups: Right atrial isomerism with asplenia where there is bilateral right atrial appendages, bilateral trilobed lungs with eparterial bronchi, and absence of spleen.

Left atrial isomerism with polysplenia have bilateral left atrial appendages, bilateral bilobed lungs with hyperarterial bronchi and multiple splenunculi. They usually present during childhood for congenital heart diseases. Congenital heart diseases is present in 50% to 100% of patients of Heterotaxy Syndrome (Peoples *et al.*, 1983).

Patients with heterotaxy may have different degrees of intestinal rotation anomalies varying from classic malrotation predisposing to volvulus to non rotation (Ditchfield and Hutson, 1998; Dilli *et al.*, 2012). Gut malrotation are seen in 70% to 100% of cases of polysplenia syndrome (Kuvuturu *et al.*, 2008).

The causes of intestinal obstruction in polysplenia syndrome include malrotation with midgut volvulus, intraluminal membrane, annular pancreas, jejunal atresia and preduodenal portal vein (Ruben *et al.*, 1983). Many patients of intestinal malrotation are asymptomatic due to broader mesentery and they come to medical attention for some other reasons.

In our case there is barium filled small bowel loops in right side of abdomen and large bowel loops are seen in left side suggestive of intestinal malrotation. As barium studies may not predict the patients with risk for midgut volvulus, the Ladd's procedure should perhaps be performed in all patients with malrotation (Powell *et al.*, 1989). Moreover, polysplenia syndrome is less associated with congenital heart disease surgery is not contraindicated and elective intervention is always better tolerated than an emergency surgery (Choi *et al.*, 2005; Yu *et al.*, 2009).

Conclusion

Heterotaxy with polysplenia is a complex anomaly which may present as intestinal malrotation. There is increased risk of intestinal obstruction in these patients. Prompt identification and prophylactic Ladd's procedure is performed to prevent complications.

REFERENCES

- Applegate KE, Goske MJ, Pierce G *et al.*, (1999). Situs revisited: imaging of the heterotaxy syndrome. *Radiographics* **19** 837–52.
- Choi M, Borenstein SH, Hornberger L and Langer JC (2005). Heterotaxia syndrome: The role of screening for intestinal rotation abnormalities. *Archives of Disease in Childhood* **90** 813–5.
- Dilli A, Gultekin SS, Ayaz UY, Kaplanglu H and Hekimoglu B (2012). A Rare Variation of the Heterotaxy Syndrome. *Case Reports in Medicine*, Hindawi **2012** Article ID 840453 4.
- Ditchfield MR and Hutson JM (1998). Intestinal rotational abnormalities in polysplenia and asplenia syndromes. *Pediatric Radiology* **28** 303–6.
- Kuvuturu S, Roper W and Stawicki SP (2008). Acute pancreatitis in a patient with situs inversus/polysplenia syndrome: A case report and review of literature. *OPUS 12 Scientist* **2** 10-2.
- Lin AE, Ticho BS, Houde K *et al.*, (2000). Heterotaxy: associated conditions and hospital-based prevalence in newborns. *Genetics in Medicine* **2** 157–72.
- Peoples WM, Moller JH and Edwards JE (1983). Polysplenia: a review of 146 cases. *Pediatric Cardiology* **4** 29–137.
- Powell DM, Othersen HB and Smith CD (1989). Malrotation of the intestines in children: The effect of age on presentation and therapy. *Journal of Pediatric Surgery* **24** 777–80.

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Ruben GD, Templeton JM Jr and Ziegler MM (1983). Situs inversus: the complex inducing neonatal intestinal obstruction. *Journal of Pediatric Surgery* **18** 751-756.

Yu DC, Thiagarajan RR, Laussen PC, Laussen JP, Jaksic T and Weldon CB (2009). Outcomes after Ladd procedure in patients with Heterotaxy syndrome, congenital heart disease and intestinal malrotation. *Journal of Pediatric Surgery* **44** 1089–95.