

SITUS AMBIGUIUS (VISCERAL HETEROTAXIA) Case report

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Abstract

A case of a 3,5 years old boy with situs ambiguus is presented. Among cases of situs anomalies the situs ambiguus implies discordance between the situs of the viscera and situs atria. Asplenia and polysplenia are often associated with these anomalies.

In our case right sided cardia, stomach, and polysplenia, in addition bowel malposition was found.

Keywords: situs ambiguus, polysplenia.

Case report

3,5 years old boy with somatic and mental retardation, hypacusis, strabismus, failure to thrive and constipation was admitted to our unit. Perinatal history was uneventful. From 5 months of age parents became aware of the boy's delayed mental development but medical investigations in another hospital could not find any reason for it. Till present admission the above mentioned complains have existed but he was never examined further.

Besides several, basically normal clinical and laboratory examinations cranial CT was performed with normal findings.

Barium meal and enema (Fig. 1.) revealed several malformation of the gastrointestinal tract including the situs of the stomach with partial obstruction, malposition of small and large bowel. Abdominal ultrasonography could not find the spleen in its normal position, the liver was mildly enlarged and a small gallbladder was visualized. The abdominal US scans were disturbed by large amount of gastric fluid, bowel fluid and gas.

Abdominal CT scans showed the position of abdominal organs clearly and a right sided polysplenia was revealed (Fig. 2.). Bony dysplasia were not discovered.

Discussion

Situs is determined very early in fetal life, but the exact mechanism is not understood. It may be that injury to the dominant side results in anomalies of situs, as has been shown in amphibians [1,2].

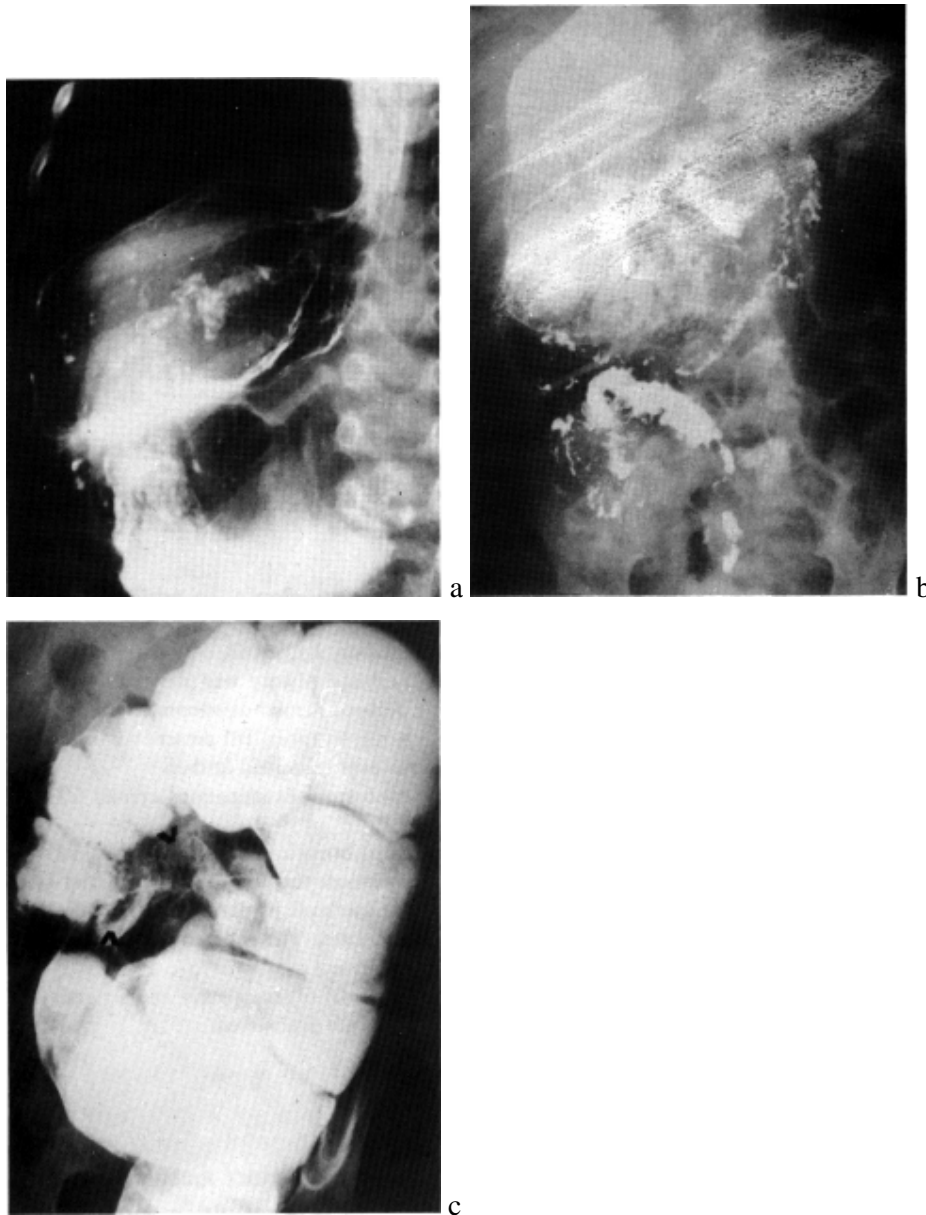


Fig. 1: Barium meal (early phase): right sided cardia and enlarged, distended stomach (a): 15 minutes: enlarged stomach, left sided curvature of the duodenum, right sided proximal small bowel (b). Barium enema. The distended, enlarged colon on the left side, the terminal ileum was visualized in the midline (arrows). Distended small bowel loops were seen on the right side (c).

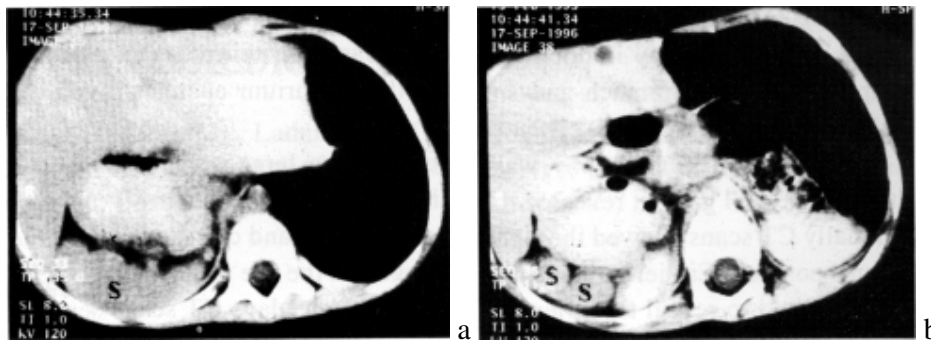


Fig. 2a-b: Abdominal CT scans in different levels showed the multiple spleens on the right side (S).

Abdominal heterotaxia is an anomalous placement or transposition of the viscera and can be of various forms. Situs solitus indicates a normal position of the viscera and cardiac atria. Situs inversus is a mirror image of situs solitus. Situs ambiguus implies discordance between the situs of the viscera and situs of the atria. Asplenia and polysplenia are often associated with anomalies of situs. Several variable forms do exist [3].

For the correct diagnosis abdominal ultrasonography, CT, barium meal and enema all together play important role. In our case barium meal revealed the malposition of the stomach and small bowel and barium enema proved the malrotation of the large bowel.

Abdominal ultrasonography was disturbed by the large amount of gastric fluid and bowel gas but revealed the abnormal position of spleen.

Finally CT scans showed the right sided polysplenia and clarified clearly the organs' position. Earlier published papers suggested the use of technetium 99m sulfur colloid to reveal the splenic anomalies but nowadays CT scans seem to be the gold standard [3,4].

Clinically the bowel anomalies may have caused gastrointestinal complains and delayed somatic development. In case of gastric obstruction surgery may be taken into consideration, in other cases the therapy is conservative. In our case the reason for the mental retardation with hypacusis and strabismus has not been clarified. The associations of central nervous system's developmental disorders has also been published in the literature [3].

References

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