

CASE REPORT

Situs Inversus Totalis with Duodenal Atresia and Preduodenal Portal Vein: A Case Report

Md. Samiul Hasan¹, KM Nurul Ferdous²

Introduction

Situs inversus totalis is the mirror image transposition of abdomino-thoracic viscera. Its association with duodenal atresia is very rare.¹ Preduodenal portal vein (PDPV) itself can cause complete or partial obstruction in patients of situs inversus. Presence of PDPV in association of situs inversus and duodenal atresia is extremely rare.^{1,2} We report a case, in whom combination of these two anomalies were a management challenges.

Case report

A 3 days old preterm female neonate weighing 2.1 Kg presented with bilious vomiting and failure to

pass meconium. Physical examination revealed scaphoid abdomen and dextrocardia. Babygram confirmed situs inversus and duodenal obstruction (Fig 1). Echocardiography revealed with dextrocardia with small ASD and PDA. Laparotomy was planned. On laparotomy stomach was found on right side with type 1 duodenal atresia (Fig 2). Proximal to the atresia there was PDPV, though it was not causing obstruction (Fig 3). An extended duodeno-duodenostomy was made after excising the membrane and bypassing the portal vein, keeping in mind that it may cause obstruction in future. Fortunately the baby recovered well, tolerated oral feed on 6th post operative day (POD) and discharged on 10th POD.



Fig 1 Situs inversus with double bubble sign

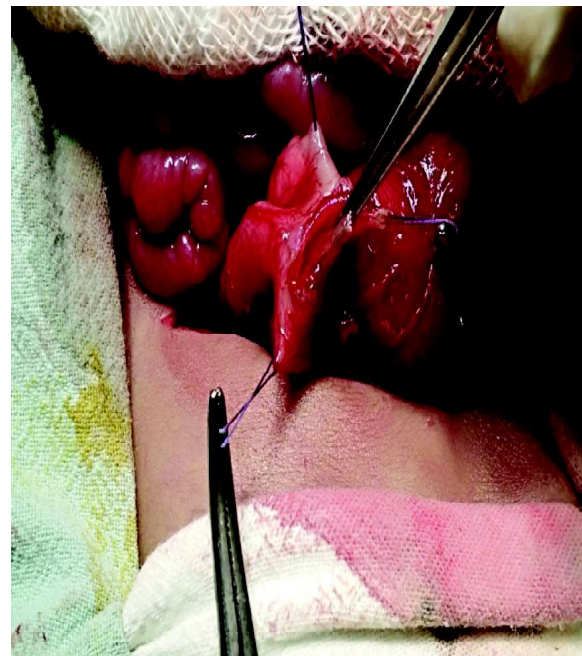


Fig 2 Type 1 duodenal atresia

1. Assistant Professor, Department of Pediatric Surgery, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka.
2. Assistant Professor, Department of Pediatric Surgery, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka.

Correspondence to: Dr. Md. Samiul Hasan, Assistant Professor, Department of Pediatric Surgery, Bangladesh Institute of Child Health (BICH), Dhaka Shishu (Children) Hospital, Dhaka. Cell: 01712886034, E-mail: samiulo45@gmail.com

Received: 02 July 2019; **Accepted:** 05 August 2019

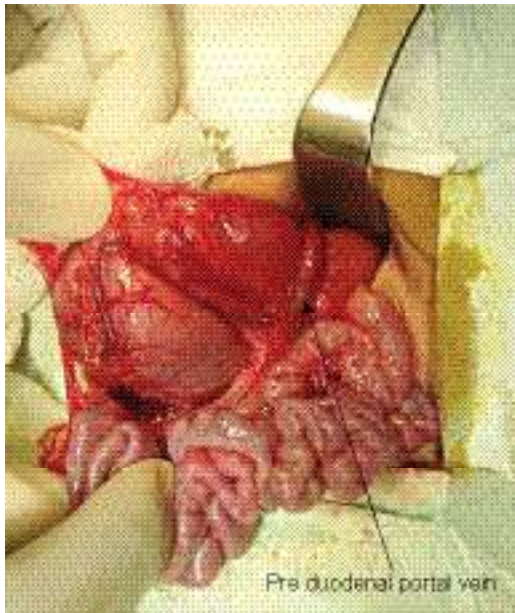


Fig 3 Pre duodenal portal vein causing incomplete obstruction

Discussion

Situs inversus results from complex and multifactorial etiology, with a prevalence of 1 in 10000.³ Incidence of duodenal atresia varies from 1 in 5000 to 1 in 20000 live birth. Association of these two is rare. Around 20 patients have been reported this association.^{3,4} PDPV occur usually in patients with situs inversus in which portal vein passes anterior to the duodenum instead of posterior course. Isolated PDPV rarely causes obstruction.²

Common associated anomalies of situs inversus are splenic and cardiac anomalies. Other associated anomalies are duodenal atresia and PDPV. In our patient, there was cardiac anomaly but spleen was structurally normal.

Duodenal obstruction in patients with situs inversus is usually due to PDPV, atresia or stenosis. Our

patient had both PDPV and atresia in close proximity, which was challenging to correct surgically. The procedure of choice for both conditions is duodenoduodenostomy.^{1,2,5} We performed an extended duodenoduodenostomy to bypass both anomalies. Separate anastomosis would be difficult and risky as both the pathologies were in close proximity.

Survival of these patients largely depends on maturity and associated cardiac anomaly.³ Post operative period of our patient was uneventful like most of the reported cases.^{1,2,4,5}

Conclusion

Combination of duodenal atresia and PDPV in patients with situs inversus is extremely rare but it should be kept in mind during laparotomy because with appropriate treatment outcome is excellent.

References

1. Duncan ND, Trotman H, Seepersaud M, Dundas SE, Thame M, Antoine M. Obstruction of the duodenum by a preduodenal portal vein in situs inversus. *West Indian Med J* 2007;**56**:285-87.
2. D souza F, Nage A, Bendre P. Preduodenal portal vein with situs inversus totalis causing duodenal obstruction. *APSP J Case Rep* 2016;**7**:24.
3. Talabi AO, Sowande OA, Tanimola AG, Adejuyigbe O. Situs inversus in association with duodenal atresia. *Afr J Paediatr Surg* 2013;**10**:275-78.
4. Brown C, Nomanoglu A, Rode H, Sidler D. Situs inversus abdominalis and duodenal atresia. *South African Journal of Surgery* 2009; **47**.
5. Shukla RM, Mukherjee PP, Mukhopadhyay B, Mandal KC. Congenital Duodenal Obstruction with Preduodenal Portal Vein and Situs Inversus Totalis: Report of Two Cases and Literature Review. *Indian J Surg* 2013;**75**:S74-S76.