27

CONGENITAL BILIARY DILATATION AND MALROTATION: AN ASSOCIATION

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Aim of the study: Asymptomatic malrotation is associated with other paediaric surgical diseases on occasion. We attempted to define the incidence of a combination to congenital biliary dilatation (CBD) and discuss the clinical significance of the association.

Methods: Retrospective case note review of all patients admitted to two institutions with a diagnosis of CBD between April 1977 and December 2015. Second surgery for the postoperative intestinal obstruction after abdominal paediatric surgery, including malrotation, was also reviewed.

Main results: Ninety-six patients with CBD were reviewed retrospectively. Five out of the 96 patients (5.2%) had malrotation (4 asymptomatic and 1 symptomatic)(Table). Adhesive intestinal obstruction occurred after excisional surgery in 3 out of 5 (60%) patients after hepaticoduodenostomy. In the case 3, insertion of a long tube could not be achieved beyond the stomach due to a strong curve at the hepaticoduodenal anastomotic site. Although no second surgery was required for intestinal obstruction, long-term postoperative QOL has been disturbed in these patients. The incidence of the postoperative intestinal obstruction was 3/91 (3.3%) in CBD patients without malrotation.

Eleven out of 68 (16.2%) patients with symptomatic malrotation required second surgery for the postoperative intestinal obstruction; adhesiolysis (n=6), intestinal resection (n=3) due to necrosis including one re-midgut volvulus, and gastrojejunostomy (n=2) as a bypass because of severe disturbance of duodenal passage at the duodenojejunal flexure (atypical malrotation). In contrast, the incidence of the second surgery for the postoperative intestinal obstruction was 52/1459 (3.6%) in patients after other pediatric abdominal surgery.

Conclusion: Asymptomatic malrotation was highly associated with patients with congenital biliary dilatation. We have reported that hepaticoduodenostomy should be abandoned because of high postoperative duodenogastric regurgitation (57th and 61st BAPS). In addition, based on this review, hepaticoduodenostomy would be also avoided as a biliary reconstruction considering the high postoperative intestinal obstruction after Ladd's procedure.

Table Congenital biliary dilatation associated with malrotation

		CBD	Malrotation	Initial surgery				Second Surgery		Outcome
Case	**Sex**	**Todani**	**Type**	**Year**	**Age**	**Procedure**	**Postoperative Complication**	**Age**	**Procedure**	
1	female	IV-A	non-rotation	1977	9yo	Ladd	Cholangitis after cystduodenostomy 2yo	13yo	Cyst excision,HJ	Dead 13yo
2	female	Ia	non-rotation	1986	9yo	Cyst excision,HJ/Ladd	none	none	none	Good 29yo
3	female	IV-A	non-rotation	1994	81d	Cyst excision,HD/Ladd	Adhesive intestinal obstruction	16yo	HJH	Good 21yo
4	male	IV-A	imcomplete	1999	11d	Cyst excision,HD/Ladd	Adhesive intestinal obstruction/ DGR	12yo	HJH	Excellent 16yo
5	male	IV-A	imcomplete	2000	5yo	Cyst excision,HD/Ladd	Adhesive intestinal obstruction/ DGR	in plan		Fair 20yo

HD:hepaticoduodenostomy, HJ:hepaticojejunostomy, HJH:hepaticojejunostomy at the hepatic hilum

DGR: duodenogastric regurgitation