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| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Series | Cohort | n | Asymptomatic - not screened |  | Asymptomatic - screened |  | Symptomatic - imaged or immediate surgery |  | Follow-up: conservatively managed | Follow-up: undergoing surgery |
| Chang, 1992 | HS 1980-1991 | 34 | 0 | N/A | 6 | 6 IRA leading to 6 LP | 28 | ?only 4 abnormal UGI, leading to 4 LP | NR | NR |
| Choi, 2005 | HS 1968-2002 | 177 | 143 | 4 developed symptoms and had UGI: 1 normal, 2 non-pathological IRA and 1 malrotation leading to 1 LP | 9 | 3 normal UGI; 1 jejunal obstruction but cardiac co-morbidity precluded surgery; 5 non-pathological IRA; no LP | 11 | 1 normal UGI; 1 hiatal hernia; 2 non-pathological IRA; 7 malrotation leading to 7 LP | Median 114 months | NR |
| Cullis, 2015 | HS 1993-2013 | 92 | 76 | No complications | 13 | 3 normal UGI; 10 IRA leading to 4 LP (1 cardiac death <30 days) | 3 | 1 normal UGI; 2 IRA leading to 1 LP and 1 awaited; no complications | Median 27.2 months | Median 17.8 months |
| Elder, 2014 | LP in infancy or HS 2000-2011 | 99 | 57 | No complications | 40 | 26 UGI showing malrotation leading to 21 LP; 43.5% complication rate and 3 deaths (cardiorespiratory failure) amongst 23 patients undergoing LP (incl. sepsis, respiratory arrest and haemorrhage) | 2 | 2 UGI showing malrotation leading to 2 LP; 43.5% complication rate and 3 deaths (cardiorespiratory failure) amongst 23 patients undergoing LP (incl. sepsis, respiratory arrest and haemorrhage) | Mean 61.2 months | Mean 43.6 months |
| Newman, 2014 | HS 1991-2013 | 34 | <15 | NR | 10 | Of 19 patients undergoing UGI: 4 normal, 4 non-rotation and 11 malrotation (though 1 later reclassified as malrotation), leading to 9 LP and 1 awaited; no complications | 9 | Of 19 patients undergoing UGI: 4 normal, 4 non-rotation and 11 malrotation (though 1 later reclassified as malrotation), leading to 9 LP and 1 awaited; no complications | NR | NR |
| Papillon, 2013 | HS 2003-2011 | 224 | 138 | No complications | 62 | Of 86 patients undergoing UGI: 26 ‘normal’, 40 atypical IRA and 9 malrotation (incl. 2 volvulus), leading to 38 LP with 9 postoperative SBO and 3 wound infections | 24 | Of 86 patients undergoing UGI: 26 ‘normal’, 40 atypical IRA and 9 malrotation (incl. 2 volvulus), leading to 38 LP with 9 postoperative SBO and 3 wound infections | Median 43.7 months | Median 46.7 months |
| Tashjian, 2007 | HS and LP 1984-2004 | 22 | 0 | N/A | 19 | 19 LP performed; of 22 patients undergoing LP, 2 adhesive SBO and 1 recurrent volvulus requiring second laparotomy; no deaths | 3 | 3 LP performed; of 22 patients undergoing LP, 2 adhesive SBO and 1 recurrent volvulus requiring second laparotomy; no deaths | N/A | Mean 108 months |
| Yu, 2009 | HS and LP 1997-2007 | 31 | 0 | N/A | 17  | Details of UGI not discussed, but 17 LP performed with 2 SBO postoperatively; deaths <30 days NR but <3  | 10 | Details of UGI not discussed, but 10 LP performed with 1 SBO postoperatively; deaths <30 days NR but <3 | N/A | NR but median <100 months |
| Zaccagni, 2009 | HS 2002-2007 | 17 | 0 | N/A | 15 | 5 normal UGI and 10 malrotation, leading to 10 LPs with 1 postoperative peritonitis with dehiscence | 2 | 2 abdominal catastrophes leading to 2 deaths | NR | NR |
| Abbreviations: HS: Diagnosis of Heterotaxy Syndrome, n: number of patients with HS in study, IRA: Intestinal Rotational Anomalies, LP: Ladd’s Procedure, SBO: Small Bowel Obstruction, UGI: Upper GI contrast study, NR: not reported adequately. IRA-associated morbidity and mortality are considered complications only. |

Table 3: summarises the findings of series that report the management pathway of patients with regards symptomatology, imaging, surgery and complications.