AB059. P-28. Is cholangiocarcinoma the only concern in western adult patients with choledochal cyst (CC) disease and anomalous pancreaticobiliary junction (APBJ)?

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Background: Choledochal cysts (CC) are congenital bile duct dilatations, intra-and/or extrahepatic, that can be associated with an anomalous pancreaticobiliary junction (APBJ). CCs are thought to predispose patients to an increased risk of cholangiocarcinoma. We hypothesized that the presence of APBJ with CC may lead to other biliary tree malignancies.

Methods: This is a retrospective cohort study including all patients managed at our institution for CC and/or APBJ from 1998 to 2018. CC and APBJ was confirmed with either magnetic resonance cholangiopancreatography or endoscopic retrograde cholangiopancreatography and classified according to the Todani classification. APBJ were classified according to the Komi classification.

Results: A total of 103 patients with CCs were identified with a mean age was 44.53 [18–74] and follow-up for a mean 2.9 [0–16] years. Patients were most commonly identified as Caucasian (n=62), followed by Asian (n=16) and Hispanic (n=10). Sixty-two (60%) of the patients had an APBJ: 46 (45%) P-C type, 13 (13%) C-P type and 3 (3%) complex type. Type I CC were the most common (n=73), followed by type IV CC (n=18), type III (n=5), type V (n=4) and type II (n=3). CC had a mean size of 23 [6–64] mm. Seventy-two (70%) of patients underwent surgery for CCs [53 (51%) with APBJ and 19 (18%) without]. Malignancy was noted in 7 (7%) patients consisting of: 2 cholangiocarcinomas, 2 gallbladder adenocarcinomas, 1 gallbladder neuroendocrine carcinosarcoma, 1 squamous cell carcinoma of bile ducts and 1 intraductal papillary neoplasm 'biliary papillomatosis' with malignant transformation. Six of these patients had type I CCs and APBJ, and type IV without APBJ was found in the remaining patient. Additionally, 2 patients had metaplasia and 4 had hyperplasia, but no malignancy in the surgical specimen. Two patients who presented with advanced malignancy died within a year of diagnosis.

Conclusions: Our results suggest that in patients with CC and APBJ there is an increased risk of cholangiocarcinoma, but other biliary tract malignancies should be considered as well. Long-term follow-up in a larger cohort is necessary to confirm these findings.

Keywords: Anomalous pancreaticobiliary junction (APBJ); choledochal cyst (CC); cholangiocarcinoma

Cite this abstract as: Baison GN, Bonds MM, Rekman JF, Rocha FG, Helton SW, Kozarek RA. Is cholangiocarcinoma the only concern in western adult patients with choledochal cyst (CC) disease and anomalous pancreaticobiliary junction (APBJ)? HepatoBiliary Surg Nutr 2019;8(Suppl 1):AB059. doi: 10.21037/hbsn.2019.AB059

