Non-pancreatic periampullary carcinoma such as ampullary carcinoma (AmpCA), distal cholangiocellular carcinoma (CholCA) and duodenal carcinoma (DuoCA) have a better prognosis than pancreatic head adenocarcinoma (PanCA). This study describes the outcome and parameters, which predict survival of non-pancreatic periampullary carcinoma after resection. Data from 148 consecutive patients with non-pancreatic periampullary carcinomas were recorded prospectively between 1993 and 2005 and analyzed using univariate and multivariate models. One hundred thirty-three of 148 (90%) patients were resected for histologically proven non-pancreatic periampullary carcinomas. R0 resection was achieved for 92% of AmpCA, for 88% of CholCA and for all the DuoCA. The lowest recurrence rate was seen in DuoCA with 18%, followed by AmpCA with 21% and CholCA with 46%. The mean survival time was 60.9 months for AmpCA patients, 42.9 months for CholCA and 45.4 months for DuoCA patients. Five-year survival was 50.5%, 29.9% and 24.5% for AmpCA, CholCA and DuoCA, respectively. Multivariate analysis identified low bilirubin levels (<100 mmol/l), R0 resections and absence of surgical complications to be strong independent predictors of survival (p < 0.05). In AmpCA low tumor stages are also an independent predictor of long-term survival (p < 0.01). For T1/T2 AmpCA the 5-year survival rate was 61%, whereas none of the patients with a T3/T4 tumor survived 5 years. Only T1/T2 ampullary carcinomas have a good prognosis, whereas T3/T4 ampullary tumors show aggressiveness similar to that of pancreatic head adenocarcinomas. Absence of surgical complications determines long-term outcome. Therefore, the combination of a complication-free and radical resection is essential for long-term survival.