Helicobacter pylori infects approximately half of the world’s population, causes peptic ulcer disease, and has been classified by the International Agency for Research on Cancer as a Group 1 (definite) carcinogen, because of evidence linking it to gastric adenocarcinoma. Other Helicobacter species are emerging as enterohepatic pathogens, infecting the intestinal tract and liver of humans and a wide range of animal species. The best studied of this group of enterohepatic Helicobacter species is Helicobacter hepaticus, which was first isolated from laboratory mice with chronic-active hepatitis and hepatocellular carcinoma. Subsequent studies have shown that H. hepaticus is highly prevalent in mouse colonies, causes chronic intestinal inflammation resembling inflammatory bowel disease, as well as intestinal cancer in knockout lines of mice, and causes gallstone formation. The complete genome sequence of H. hepaticus has been determined, and several candidate virulence determinants, including flagella, urease, cytolethal distending toxin, and a pathogenicity island containing type IV secretion system components, have been identified. There is growing evidence that related enterohepatic Helicobacter species, such as Helicobacter bilis, Helicobacter pullorum, and Helicobacter cinaedi are associated with human disease. Therefore, enterohepatic Helicobacter species are emerging as important mucosal pathogens with zoonotic potential that cause chronic inflammation and inflammation-associated cancer.