

INTRODUCTION

Cholangiocarcinoma (CCA), also known as bile duct cancer, is a malignancy arising in the ductular epithelium of the biliary tree. It is classified into three distinct subtypes using the WHO International Classification of Diseases for Oncology (ICD-O) coding system – intrahepatic, extrahepatic perihilar and extrahepatic distal (*Figure 1*):

- Intrahepatic (iCCA, ICD-10 C22.1) arising within the intrahepatic ducts of the liver.
- Extrahepatic (eCCA, ICD-10 C24.0) comprising: perihilar (or hilar) (pCCA) arising at the bifurcation of the main hepatic ducts, and distal (dCCA) arising beyond the cystic duct.

There has been no CCA epidemiological data published for England since 2008. Western studies have shown that the incidence of CCA has been increasing, with differing trends between subtypes, rising rates for iCCA and declining eCCA^{1,2,3}.

The aim of this study was to obtain up-to-date incidence and mortality trends for cholangiocarcinoma in England using data from the National Cancer Registration and Analysis Service (NCRAS).

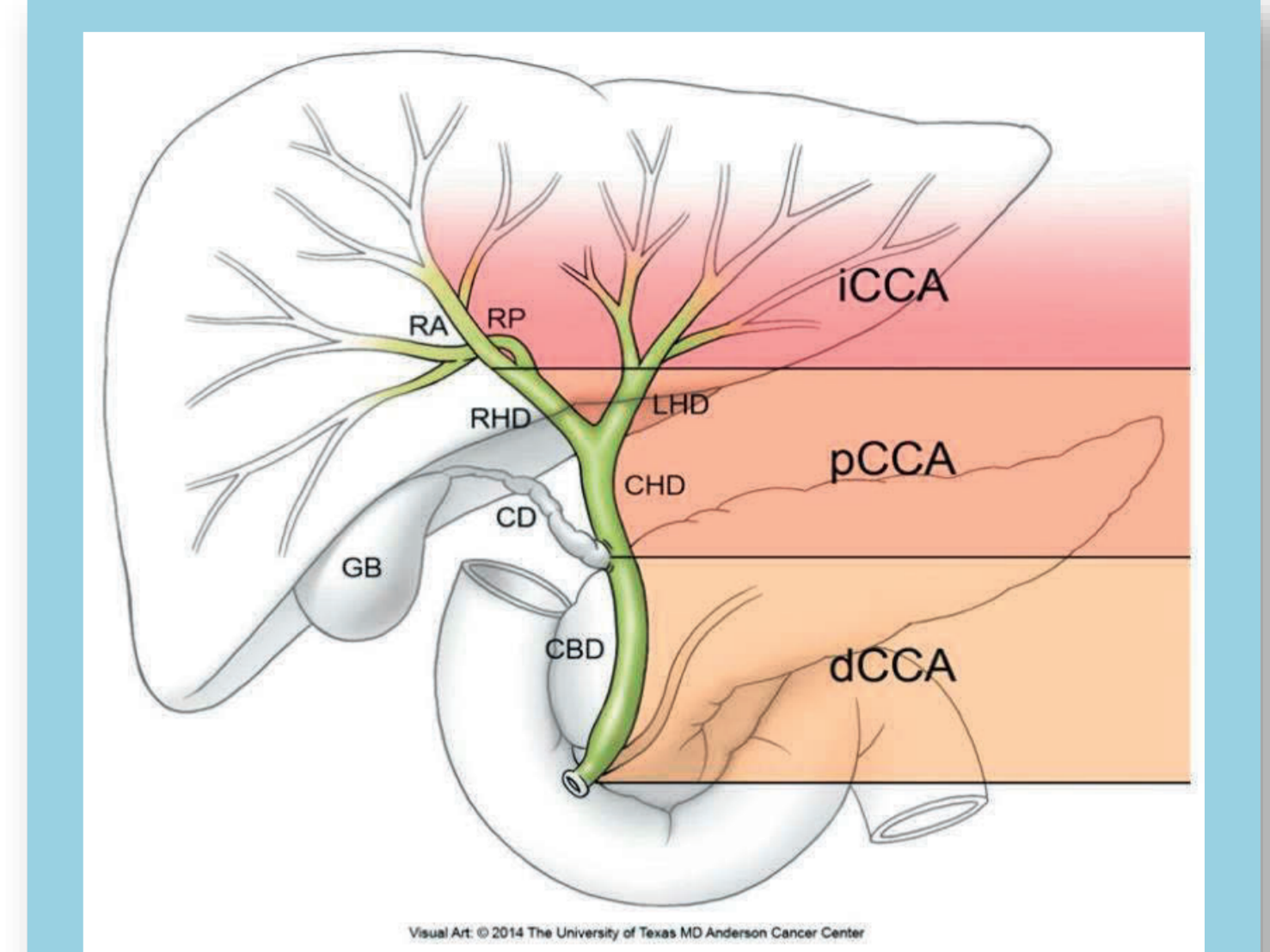


Figure 1. Cholangiocarcinoma subtypes

RA, right anterior segmental duct; RP, right posterior segmental duct; RHD, right hepatic duct; LHD, left hepatic duct; CHD, common hepatic duct; CD, cystic duct; CBD, common bile duct; GB gall bladder

Image from Blechacz B. *Gut Liver* 2017; 11(1): 13–26

METHODS

Cancers diagnosed between 1st January 2001 and the 31st December 2017 in England and coded as ICD-10 C22.1 or C24.0 were included in the study. Annual European directly age-standardised incidence (ASIRs) and mortality (ASMRs) rates per 100,000 population were calculated using data from the NCRAS population-based cancer registry. P-tests for trend were undertaken for incidence and mortality over the time period.

RESULTS

There were 33,585 CCAs diagnosed in England residents between 2001 and 2017, 26,307 (78%) were coded as iCCAs and 7,278 (22%) eCCAs.

Annual ASIRs for CCA overall and by subtype are presented in Figure 2 below. The ASIRs for CCA in England in 2017 for males and females were 4.6 (4.3-4.9) and 4.0 (3.7-4.2), respectively.

Annual ASMRs for CCA overall and by subtype are presented in Figure 3 below. The ASMRs for males and females in 2017 were 4.9 (4.6-5.2) and 4.5 (4.3-4.8), respectively.

The incidence and mortality rates of CCA in England have been steadily increasing over the last 17 years (test for trend <0.001 for both).

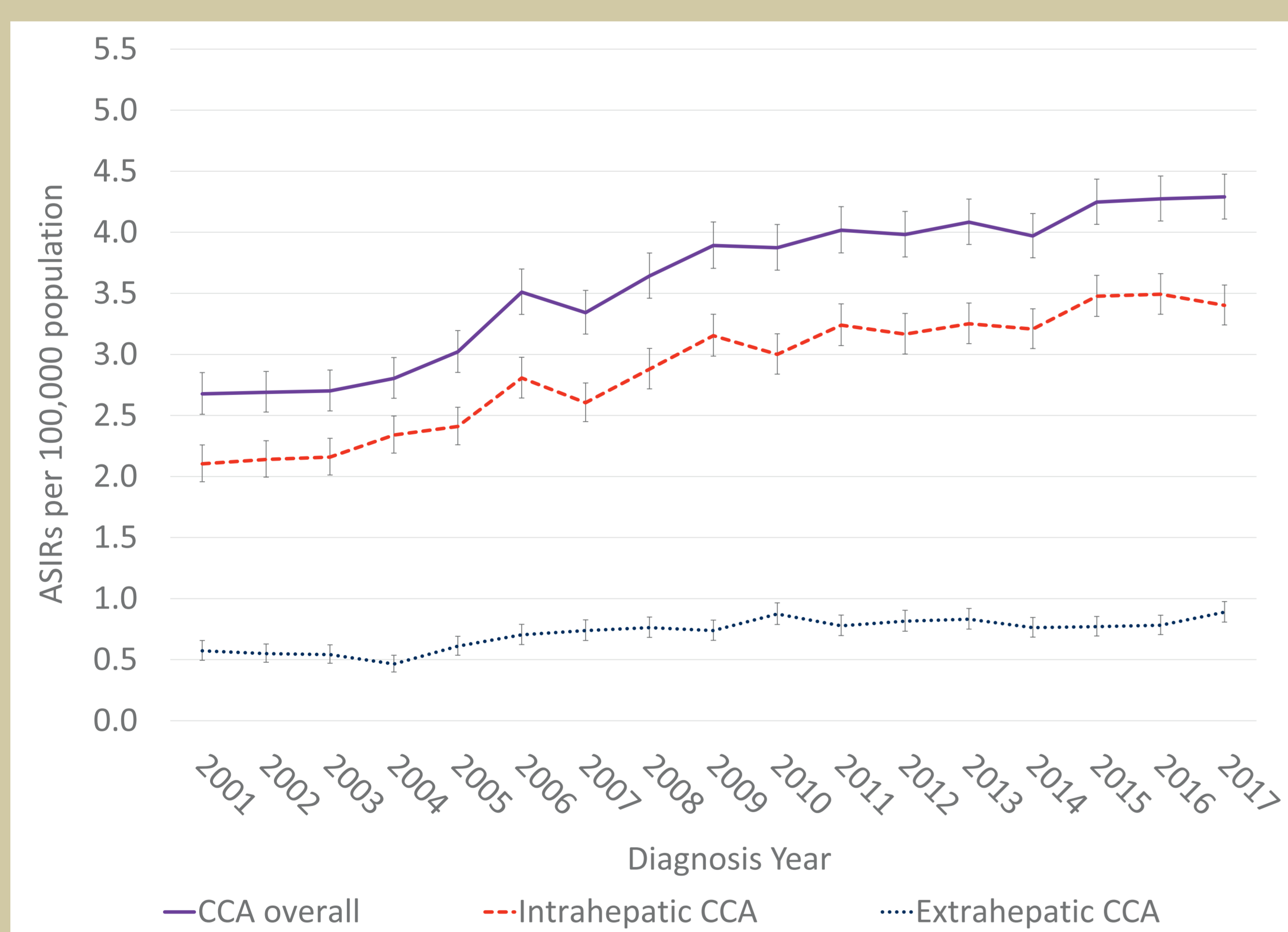


Figure 2. Annual age-standardised incidence rates (ASIRs) for CCA overall, iCCA, and eCCA for persons diagnosed in England between 2001 and 2017.

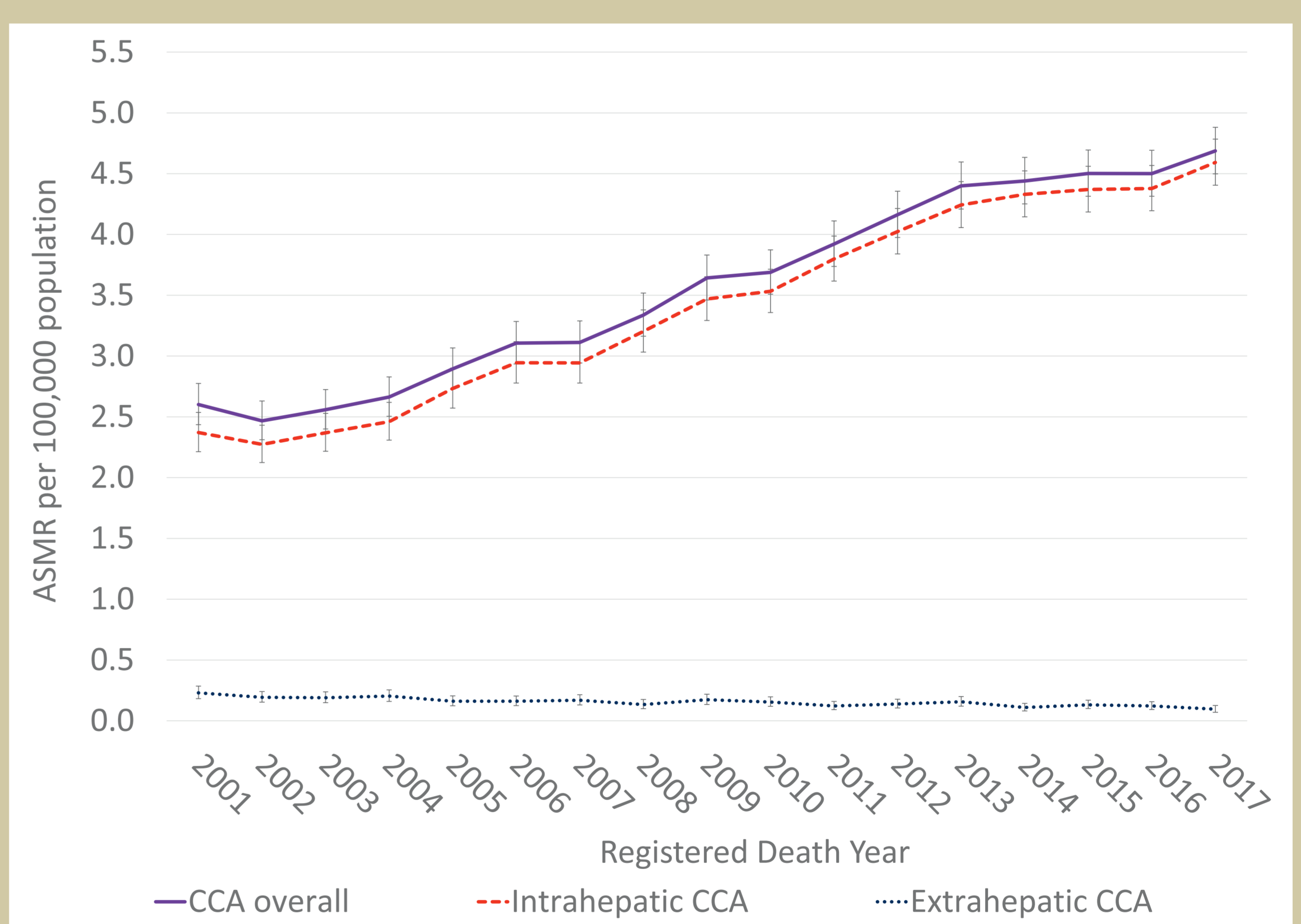


Figure 3. Annual age-standardised mortality rates (ASMRs) for CCA overall, iCCA, and eCCA for persons diagnosed in England between 2001 and 2017.

DISCUSSION

The rising incidence of CCA in England is consistent with findings in the USA² and Canada³. The parallel increase in mortality would suggest that this is a 'true' increase and not just the result of improved diagnostic testing or imaging.

Preliminary analysis of pathology reports received by NCRAS suggests that the incidence of iCCA may still be partially attributable to the misclassification of pCCAs as intrahepatic previously reported¹. The new ICD-11 coding system will rectify this by providing topographical codes for dCCA and pCCA, C24.2 and C24.3, respectively. Further studies are required to determine the true incidence of CCA subtypes by employing ICD-11 coded data.

CONCLUSIONS

- CCA incidence and mortality has been steadily increasing in England over the last 17 years
- Further studies are required to determine true subtype incidence and mortality rates using data coded with ICD-11

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