

Biliary Tract and Liver Complications in Polycystic Kidney Disease

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Supplemental Table 1: Management of biliary tract disease and serious liver complications in patients with polycystic kidney disease treated by Oxford Kidney Unit

Complication	Total number of admissions (number of patients)	Management	Number of Admissions (number of patients)
Biliary tract disease complications			
Biliary sepsis (including cholecystitis and cholangitis)	90 (21)	Antibiotics	90 (21)
Cholelithiasis ± Choledocholithiasis	26 (21)	Ursodeoxycholic acid	2 (2)
		ERCP with;	
		- Stone removal	21 (11)
		- Sphincterotomy	13 (5)
		- Stent	8 (3)
		- Sludge trawled	4 (4)
		Percutaneous cholecystostomy	1 (1)
		Laparoscopic/open cholecystectomy	8 (8)
		Biliary bypass	1 (1)
Serious liver complications			
Infected liver cyst and/or abscess	22 (12)	Antibiotics	22 (22)
		Ultrasound-guided cyst aspiration	3 (3)
Liver cyst de-roofing	4 (4)		
Extrinsic compression from liver cysts	1 (1)	ERCP and stent	1 (1)
Abnormal liver architecture/focal cholangitis	1 (1)	Liver biopsy	1 (1)
Refractory ascites	1 (1)		
Liver hemorrhage	1 (1)	Partial hepatectomy	1 (1)
Liver failure	2 (2)	Liver transplant	2 (2)
Subphrenic abscess	1 (1)	Drainage and antibiotics	1 (1)

ERCP = Endoscopic retrograde cholangiopancreatography

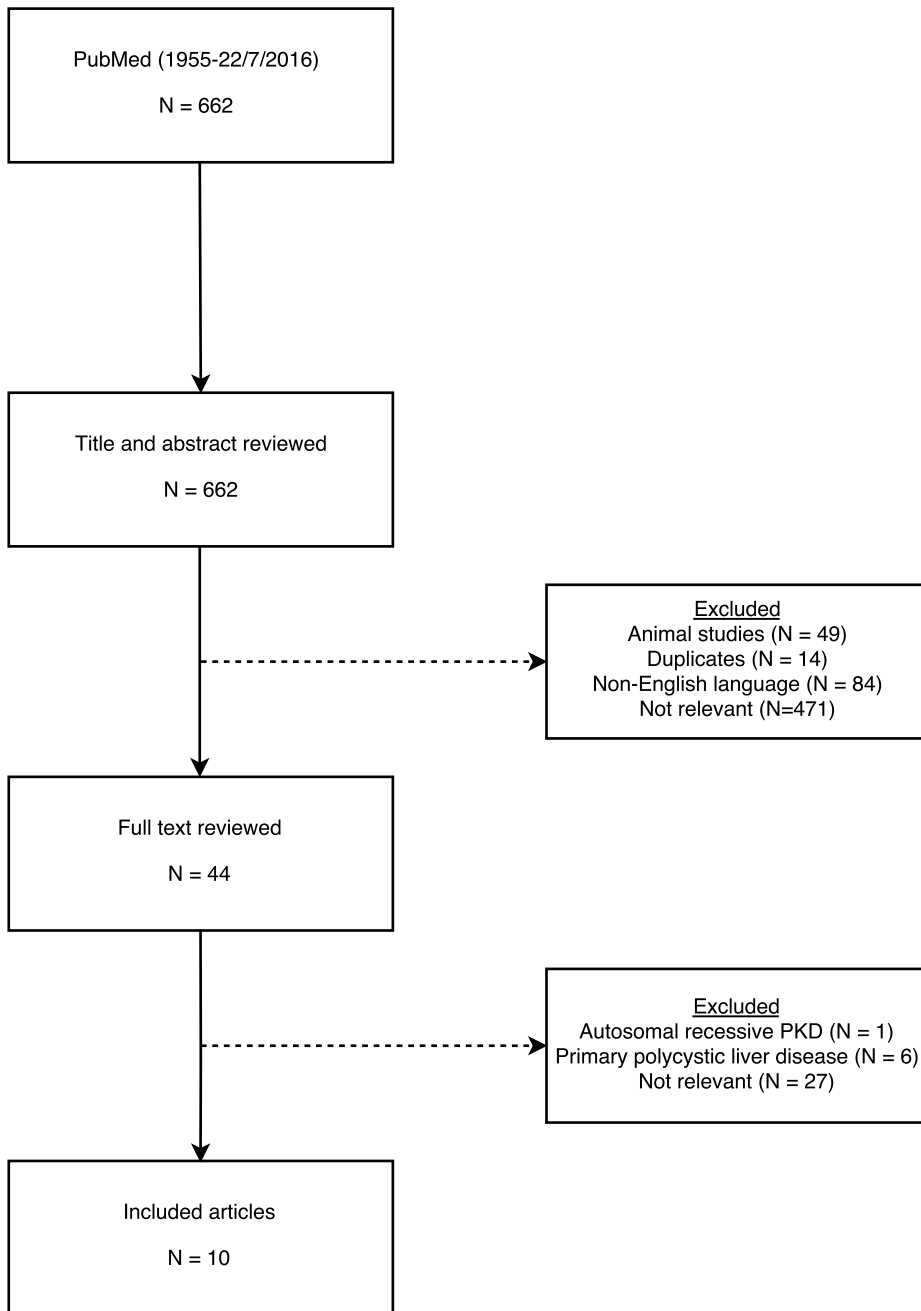
Supplemental Table 2: Diagnostic and procedural codes used to define biliary tract disease, serious liver complications and extra-renal complications

ICD-10 (ICD-9) codes	ICD-10 descriptions	OPCS-4 codes	OPCS-4 descriptions
Biliary tract disease			
K80.0-K80.2 (574.0, 547.1, 574.2, 574.6-574.9, 575)	Gallbladder calculi – with or without cholecystitis	J20.2	Closure of cholecystotomy
K82.1-K82.3 (575.2-575.5)	Hydrops of gallbladder, perforation or fistula	J21.1-J21.3, J24.1-J24.9, J26.1	Open and percutaneous removal of calculus from gallbladder and drainage of gallbladder
K82.8-K82.9 (575.8, 575.9)	Other diseases of the gallbladder	J18.1-J18.9	Partial/Total cholecystectomy and exploration of common bile duct
K80.3-K80.5 (574.3-574.5, 574.6-574.9)	Calculi in bile ducts with/without cholecystitis or cholangitis	J25.8, J25.9, J26.1, J26.8, J26.9	Other gallbladder and calculus procedures
K80.8, K81.0, K81.1, K81.8, K81.9 (575.0, 575.1)	Other cholelithiasis, cholecystitis	J24.1-J24.8	Percutaneous treatments to stones
K83.0 (576.1)	Cholangitis	J41.1, J76.1	Endoscopic retrograde extraction of calculus from bile duct
K83.1-K83.3 (576.2)	Obstruction of bile duct	J33.1-J34.3, J35.1-J35.9, J38.1-J39.1	Sphincterotomy/-plasty
K83.8-K83.9 (576.8, 576.9)	Other diseases of the biliary tract	J48.5-J49.2	Percutaneous drainage of liver/gallbladder, t-tube insertion
K74.3-K74.5 (571.6)	Biliary cirrhosis	J43.3	Endoscopic retrograde cholangiopancreatography and collection of bile
K85 (577.0)	Acute pancreatitis	J42.3	Endoscopic retrograde removal of calculus from pancreatic duct
Serious liver complication			
K74.0-K74.2	Hepatic fibrosis, Sclerosis	J02.1-J02.9	Liver resection
K74.6 (571.5)	Other and unspecified cirrhosis of liver	J08.1, J08.2	Investigation of liver and gallbladder
K76.9 (571.9)	Liver disease, unspecified	Y06.2	Deroofing of cyst of organ
K75.0 (572.0)	Abscess of liver	J08.8, J08.9	Unspecified therapeutic endoscopic operations on liver using laparoscope
Z94.4	Liver transplant status	J01.1-J01.3, J01.5-J01.9	Transplantation of liver
T86.4	Liver transplant failure and rejection		
End-stage renal disease			
Complex algorithm			
Cerebral aneurysm			
I60 (430)	Subarachnoid haemorrhage	L33-L34	Operation on cerebral aneurysm
Other abdominal wall hernia			
K41 (551.0, 552.0, 553.0)	Femoral hernia	T22-T23	Femoral hernia repair
K42 (551.1, 552.1, 553.1)	Umbilical hernia	T24	Umbilical hernia repair
Urinary tract infections			
N10, N13.6, N15.1, N30, N39.0 (590, 595)	Urinary tract infections		
Cardiac valve disease			
I34.0 (424.0)	Mitral valve regurgitation	K25, K30.1	Mitral valve repair
I34.1 (424.0)	Mitral valve prolapse	K26, K30.2	Aortic valve repair
I34.2 (394.0)	Mitral valve stenosis		
I35.1 (424.1)	Aortic valve regurgitation		
I35.0 (424.1)	Aortic valve stenosis		
Breast cancer			
C50 (174-175)			
Diverticular disease			
K57 (562.1)	Diverticular disease		
Inguinal hernia			
K40 (550)	Inguinal hernia	T20-T21	Inguinal hernia repair

ICD-10 (ICD-9) codes	ICD-10 descriptions	OPCS-4 codes	OPCS-4 descriptions
Aortic or other aneurysms I71.1-I71.2 (441.1-441.2)	Thoracic aortic aneurysm	L18-L19, L27-L28	Abdominal aneurysm repair
I71.0 (441.0)	Aortic dissection	L48-49	Iliac artery aneurysm repair
I71.3-I71.6 (441.3-441.9)	Abdominal aortic aneurysm		
I72 (442)	Other aneurysm		
Hiatus hernia and gastroesophageal reflux disease K44, Q40.1, Q79.0 (551.3, 552.3, 553.3, 750.6, 756.6)	Diaphragmatic hernia		
K20-K21 (530.1-530.2)	Gastro-oesophageal reflux disease or oesophagitis		
Renal stones N20-N23 (592, 594)	Urolithiasis		

ICD = International Classification of Disease. OPCS= Office of Population Censuses and Surveys' Classification of Interventions and Procedures.

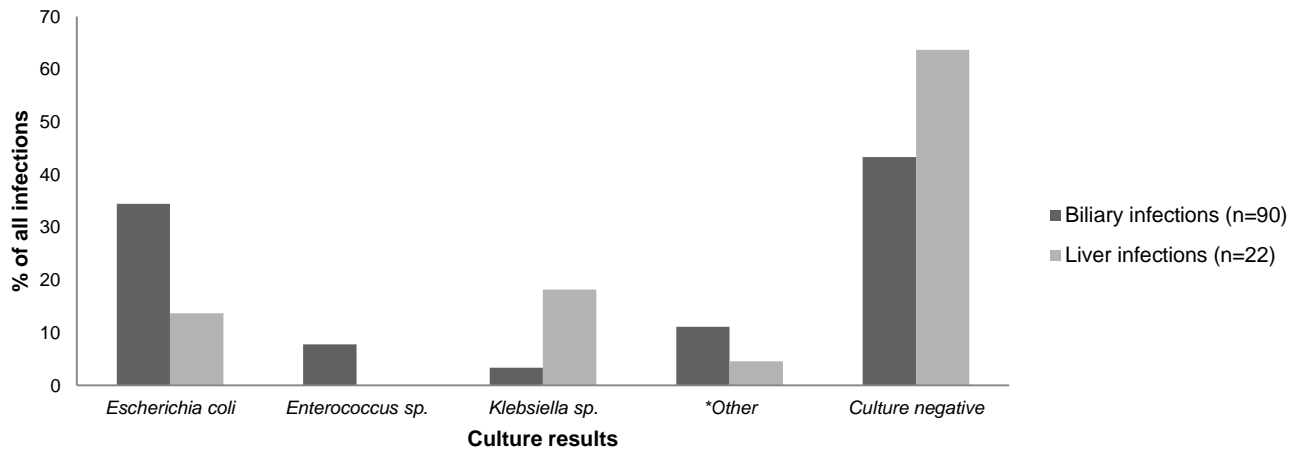
Supplemental Figure 1: PubMed literature search for biliary tract disease and serious liver complications in autosomal dominant polycystic kidney disease



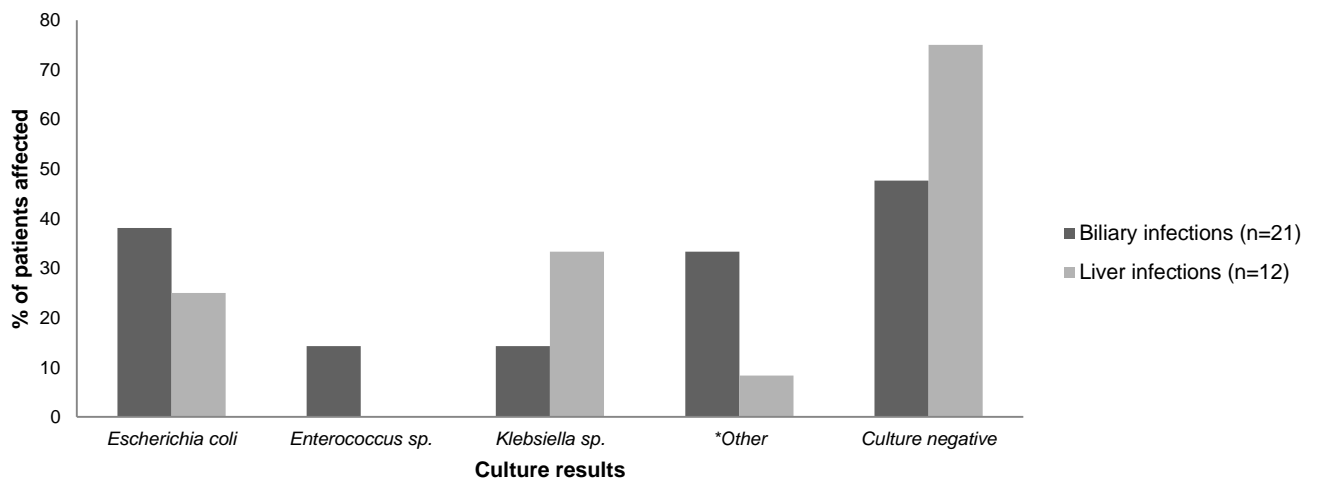
Search terms included polycystic kidney disease (PKD), polycystic liver disease (PLD), hepatic complications, gallstones, cholecystitis, gallbladder and other specific treatments for PLD and PKD.

Supplemental Figure 2: Organisms identified during biliary tract and liver infections in patients with polycystic kidney disease treated by Oxford Kidney Unit by; admissions (panel A), and by patient (panel B)

A



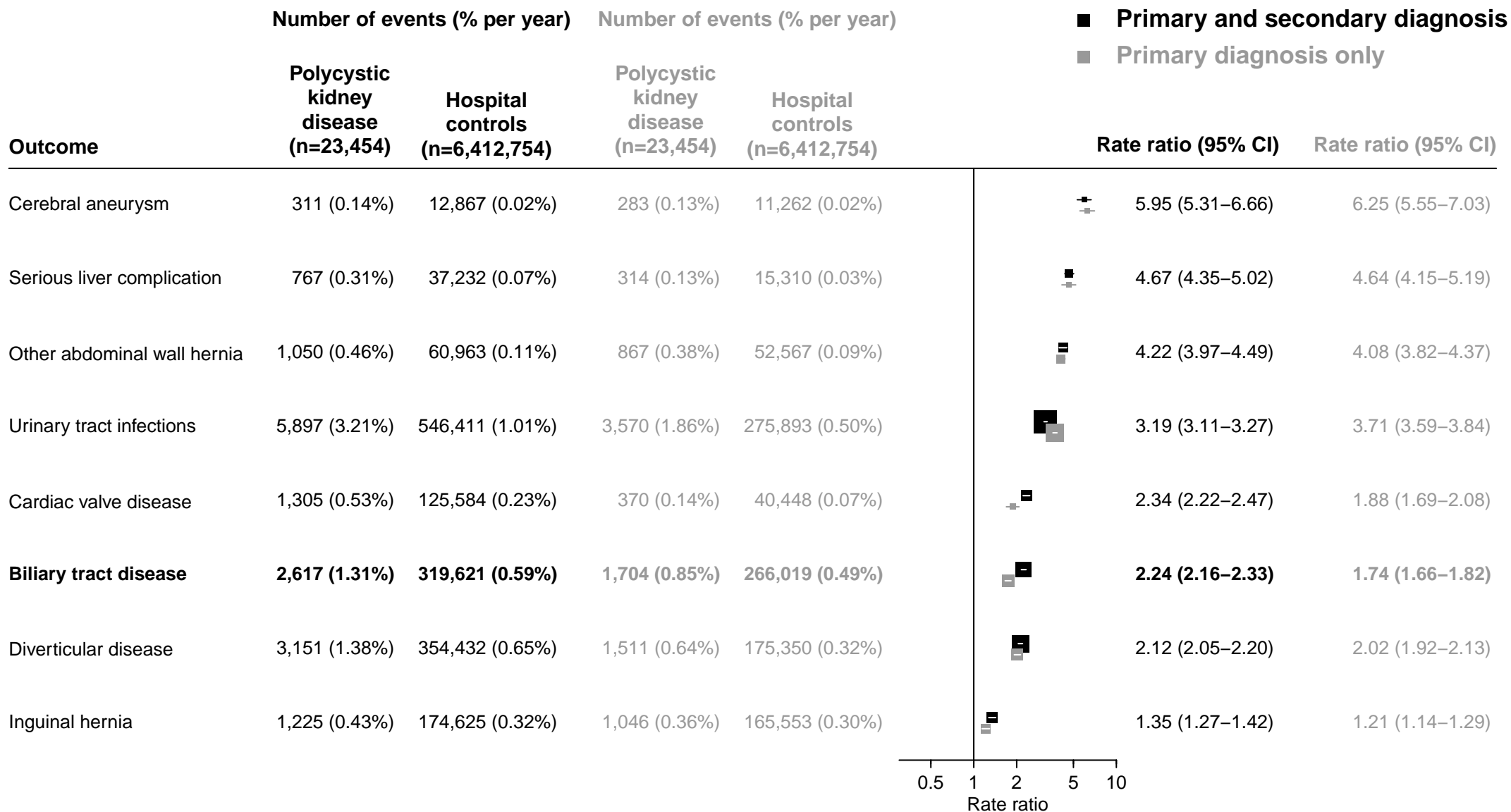
B



* Other infections include: *Pseudomonas aeruginosa* (liver=1, biliary=3), *Serratia marcescens* (biliary=2), extended spectrum beta lactamase (biliary=2), *Enterococcus sp.* (biliary=2), *Proteus sp.* (biliary=1). The 90 biliary infections occurred in a total of 21 patients and the 22 liver infections occurred in a total of 12 patients.

Supplemental Figure 3: Association between polycystic kidney disease and risk of hospitalisation for different disease by diagnostic position in all-England Hospital Episode Statistics 1998–2012 (sensitivity analysis)

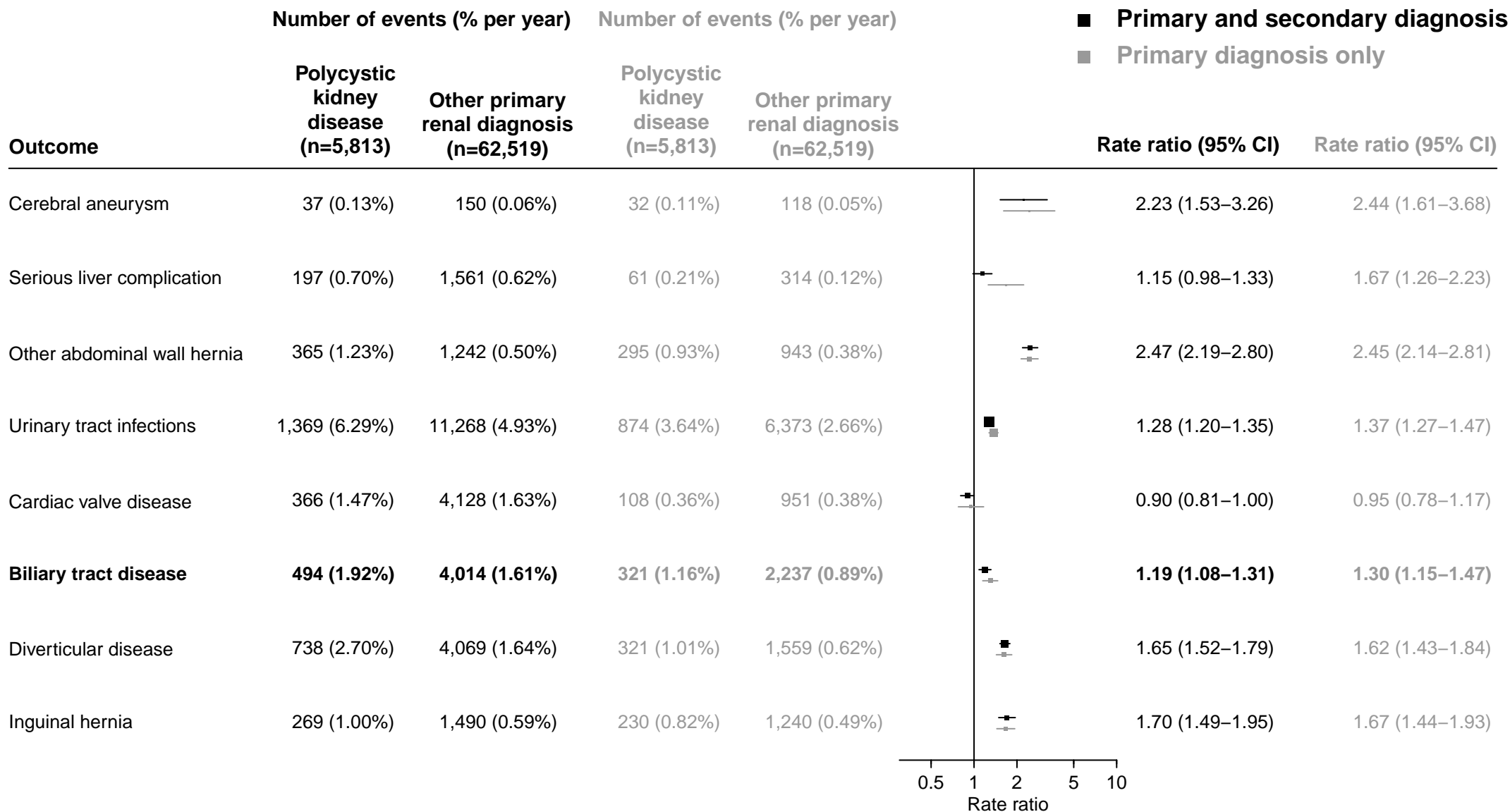
All patients



CI = confidence intervals. Outcomes include admissions with relevant diagnostic codes in either any diagnostic position or primary diagnostic position only, or any relevant procedural codes. Adjusted for age at entry as a continuous variable (using both linear and quadratic terms), sex, ethnicity, quintile of patients' Index of Multiple Deprivation score, region of residence, calendar year of first recorded admission and comorbidities (grouped into vascular, cancer and diabetes).

Supplemental Figure 4: Association between polycystic kidney disease and risk of hospitalisation for different diseases by diagnostic position in all-England Hospital Episode Statistics 1998–2012 (sensitivity analysis)

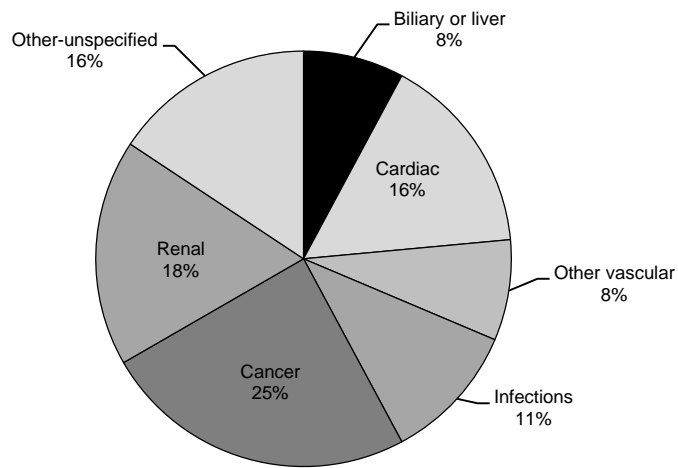
Treated end-stage renal disease patients



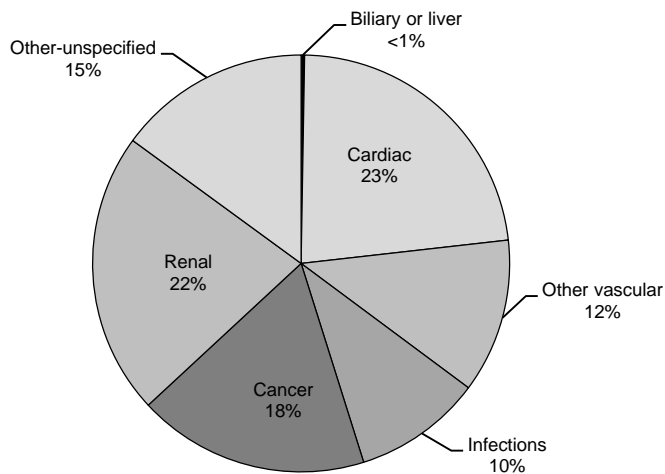
CI = confidence intervals. Outcomes include admissions with relevant diagnostic codes in either any diagnostic position or primary diagnostic position only, or any relevant procedural codes. Adjusted for age at entry as a continuous variable (using both linear and quadratic terms), sex, ethnicity, quintile of patients' Index of Multiple Deprivation score, region of residence, calendar year of renal replacement therapy start and comorbidities (grouped into vascular, cancer and diabetes).

Supplemental Figure 5: Underlying causes of death in all people with polycystic kidney disease by; prior biliary tract or serious liver complication (panel A), or without such complications

A: (1,128 deaths in 3,136 patients)

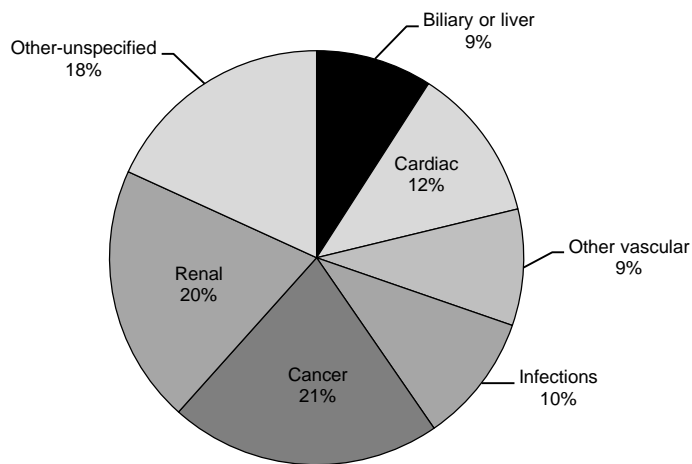


B: (6,192 deaths in 20,318 patients)



Supplemental Figure 6: Underlying causes of death in all people with polycystic kidney disease and prior biliary tract or serious liver complications in; females (panel A), or males (panel B)

A: Females (509 deaths in 1,580 patients)



B: Males (619 deaths in 1,556 patients)

