THE PREVALENCE OF PRIMARY BILIARY CHOLANGITIS (PBC) IS ON THE RISE: A CANADIAN POPULATION-BASED STUDY

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BACKGROUND

- Canada has one of the highest rates of primary biliary cholangitis (PBC) incidence and prevalence worldwide.
- Our group has previously reported the natural history of PBC in the Calgary Health Zone (CHZ), population ~ 1.5 million[1]. Since that report, significant increases in PBC awareness and improved management have occurred.
- Therefore, we aimed to study the impact of these factors on the natural history of PBC in a well-defined Canadian population and evaluate the temporal trends of PBC incidence and prevalence.

METHODS

- We used population-based administrative data (inpatient, ambulatory care, and physician claims) and a previously validated International Classification of Diseases coding algorithm to identify PBC patients in the CHZ between 2003-2018.
- The same methods were used as were implemented in our original study (study period 1996-2002)[1].
- We conducted a validation study on incident cases to confirm the performance of our algorithm in order to identify definite PBC cases (2/3 criteria of: positive anti mitochondrial antibody, cholestatic liver enzyme elevation, and a compatible liver biopsy). PPV of the diagnostic algorithm was 78.2%.
- We used a washout period of 2 years (2003-2004) to identify incident PBC patients. March 31st of each year was chosen as point prevalence.
- Temporal trends were evaluated using generalized linear models assuming a Poisson distribution or a negative binomial distribution if over-dispersion was present. Rates were adjusted to the Canadian population census of 2016.

RESULTS

- 521 incident cases were identified in the CHZ between 2005-2018 (Figure 1.); 78.5% were women. Median age at diagnosis was 55 (IQR 48-65).
- The overall age/sex-adjusted annual incidence of PBC between 2005-2018 was 36.9 per million.
- The incidence changed during the study period (P=0.0162, Figure 2.)

RESULTS

- Age-adjusted incidence rates were 57.0 per million in women compared to 15.7 per million in men (IRR 3.65; 95% CI 2.96-4.49). The highest adjusted incidence was observed among the 60-79 year old age group (IRR 7.92 95% CI 5.95-10.54, Table 1.)
- Age/sex-adjusted point prevalence rates of PBC increased from 253.0 to 479.1 per million between 2005 and 2018 (P<0.001, Figure 3.). Adjusted point prevalence rates were much higher among women than men, 734.0 compared to 153.5 per million (Table 1.)
- The highest prevalence of PBC was among individuals aged over 80 years at 1276.3, compared to 35.3 in the 18—39 year old category (Rate Ratio: 31.12; 95% CI 18.2-52.32, Table 1.)
- After a median follow up of 6.8 years (IQR 3.9-10.6 years; range 1.1 to 15 years), 30 patients underwent liver transplantation (5.76%) and 81 patients died (15.6%). Annual mortality rate was 10.6 per million. Survival was not significantly different between men and women (P=0.5316, Figure 4.) Survival was significantly lower with an older age at diagnosis (P<0.0001, Figure 5.)

Table 1: Incidence Rates and Point Prevalence Rates of PBC (per million population) According to Age and Sex in the CHZ.

	Annual Incidence Rate (2005-2018)				Point Prevalence Rate (2018)			
Age (years)	Women*	Men*	Total	Rate Ratio for Age Category (95% CI)	Women*	Men*	Total	Rate Ratio for Age Category (95% CI)
18-39	16.3	6.5	9.2	1.00 (reference)	52.6	18.3	35.3	1.00 (reference)
40-59	99.3	18.4	45.1	4.86(3.69-6.39)	703.4	99.8	402.0	11.28 (7.03-18.10)
60-79	168.5	43.4	73.9	7.92(5.95-10.54)	1754.1	378.7	1093.4	29.98(18.82-47.74)
80+	47.6	10.1	25.6	2.44(1.34-4.43)	1690.4	593.4	1276.3	31.12(18.52-52.32)
Overall	57.0	15.7	36.9	-	734.0	153.5	479.1	-

Figure 1: Incident cases of PBC in the CHZ between 2005-2018 by age/sex

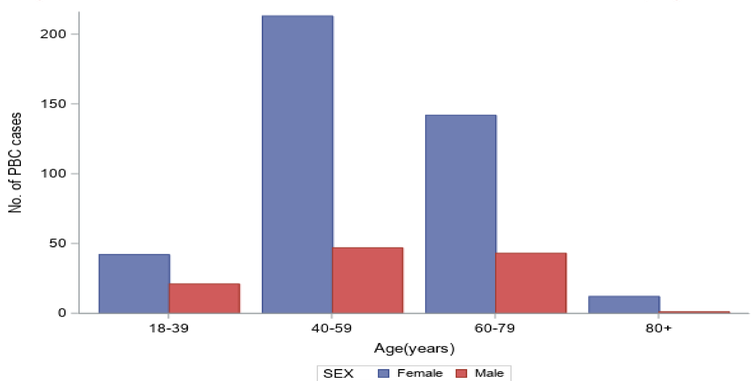


Figure 2: Annual age/sex-adjusted incidence of PBC in the CHZ

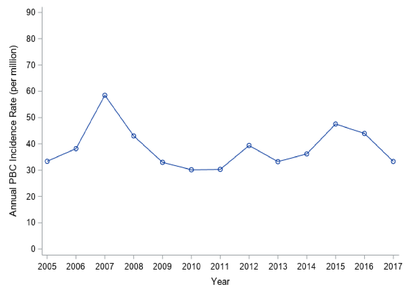


Figure 3: Annual age/sex-adjusted prevalence rates of PBC in the CHZ.

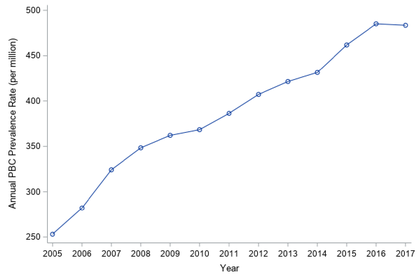


Figure 4: Survival of patients with PBC according to sex at diagnosis

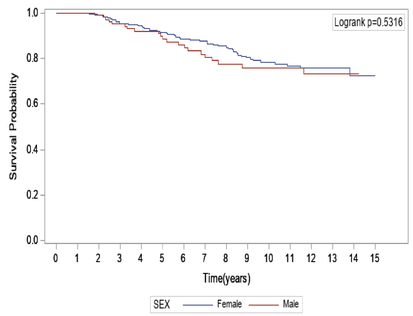
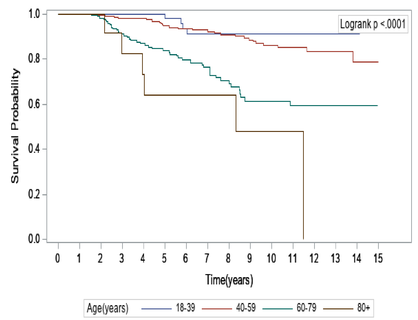


Figure 5: Survival of patients with PBC according to age at diagnosis



CONCLUSIONS

- We report a significant increase in the prevalence of PBC in this Canadian population.
- Prevalence of PBC is highest among the older age group which could reflect better case-identification and management.
- Survival is largely dictated by age at diagnosis and is not impacted significantly by sex.

[1]Myers, Robert P., et al. "Epidemiology and natural history of primary biliary cirrhosis in a Canadian health region: a population-based study." *Hepatology* 50.6 (2009): 1884-1892.