PREVALENCE OF DIAGNOSED PRIMARY BILIARY CHOLANGITIS AND ASSOCIATED CONDITIONS IN THE UNITED STATES: RESULTS FROM A NATIONAL ELECTRONIC PATIENT DATABASE

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Overview

- Primary biliary Cholangitis (PBC) is an autoimmune disease that is associated with a number of autoimmune disorders.
- PBC is associated with liver and non-liver organ co-morbidities that contribute to clinical outcomes.
- Several studies have suggested an increase in the incidence and prevalence of PBC, although this has not been assessed in a large cohort and has not been consistent across all epidemiologic studies.
- We determined the nationwide prevalence of PBC and its association with other autoimmune and non-liver related conditions in the United States (US).

Methods

- Explorys Database.
- Patients (age >18 years) with a diagnosis of PBC between 1999 to present.
- Data were analyzed in patients with/without a diagnosis of PBC.
- The prevalence of associated diseases was compared in in the 2 groups.
- Statistical univariate binary logistic model was performed.

Results

- Of the 70,383,890 individuals in the database, we identified 11,070 cases of PBC, with a prevalence of 15.7/100,000 persons (0.02%). Prevalence was higher in females than males [odds ratio (OR) 32.84; 95% Confidence Interval (CI) 30.50-35.37, p < 0.0001], adults (≥ 65 years) versus adult (18-65 years) (OR) 2.18; 95% CI 2.07-2.30, p < 0.0001) and whites versus non-whites (OR 14.54; 95% CI 13.63-15.51, p < 0.0001) (table1). PBC patients were at higher odds (Table 1) of co-morbidities including ischemic heart disease, cerebrovascular accident, congestive heart failure and sarcopenia when compared to non-PBC patients (p < 0.0001). PBC patients were more likely than non-PBC patients to have primary sclerosing cholangitis (OR 1061.43) autoimmune hepatitis (OR 537.93), Type 1 diabetes mellitus (OR 240.58), ulcerative colitis (OR 15.25), Crohn's disease (OR 7.84), celiac disease (OR 8.99), autoimmune thyroiditis (OR 7.69), systemic lupus erythematosus (OR 14.66) and other autoimmune disorders (table 2).

Table1	With PBC (%) (n=11,070)	Wo PBC (%) (n=70,372,820)	OR (CI)	p-value
Demographics				
AGE 18-65	4,490 (41%)	47940720 (68%)	0.32 (0.31-0.33)	< 0.0001
>65	6,620 (60%)	21,2517,90 (30%)	3.44 (3.31-3.57)	< 0.0001
Male	1,610 (15%)	31,420,380 (45%)	0.21 (0.20-0.22)	< 0.0001
Female	9,390 (85%)	38,453,360 (55%)	4.64 (4.41-4.89)	< 0.0001
Caucasian	8,770 (79%)	37,743,080 (54%)	3.30 (3.15-3.45)	< 0.0001
Comorbidities				
T2DM	2810 (25%)	5,650,420 (8%)	3.90 (3.73-4.07)	< 0.0001
HTN	1950 (18%)	3,493,560 (5%)	4.09 (3.89-4.29)	< 0.0001
HLD	5940 (54%)	11,779,380 (17%)	5.76 (5.55-5.98)	< 0.0001
Tobacco use	2220 (20%)	6,486,950 (9%)	2.47 (2.36-2.59)	< 0.0001
Hypothyroidism	3300 (30%)	4,049,030 (6%)	6.96 (6.68-7.25)	< 0.0001
Ischemic Heart Disease	1240 |(11%)	2,496,690 (4%)	3.43 (3.23-3.64)	< 0.0001
CHF	1070 (10%)	1,834,510 (3%)	4.00 (3.75-4.26)	< 0.0001
Alcohol abuse	320 (3%)	1,090,250 (2%)	1.89 (1.69-2.11)	< 0.0001
Sarcopenia	4910 (44%)	5,480,850 (8%)	9.44 (9.09-9.80)	< 0.0001

Condition	OR	95% CI	p-value
PSC	1061.43	660.91-1704.66	< 0.0001
AIH	507.79	507.79-569.85	< 0.0001
T1DM	240.58	217.08-266.63	< 0.0001
CREST	225.28	196.07-258.84	< 0.0001
AIHA	45.27	36.76-55.75	< 0.0001
Sjogren's Syndrome	44.63	41.42-48.10	< 0.0001
Raynaud's Syndrome	24.81	22.94-26.83	< 0.0001
UC	15.25	13.92-16.71	< 0.0001
CD	7.84	6.99 -8.79	< 0.0001
Celiac Disease	8.99	7.78 -10.37	< 0.0001
SLE	14.66	13.35 -16.09	< 0.0001
ITP	14.74	12.31-17.65	< 0.0001
AIT	7.69	6.82-8.68	< 0.0001
Hashimoto Thyroiditis	7.69	6.71-8.81	< 0.0001
RA	8.74	8.14-9.39	< 0.0001
Psoriasis	5.86	5.29-6.50	< 0.0001
Vitiligo	6.38	4.83-8.43	< 0.0001
MG	5.81	4.06-8.32	< 0.0001

Table 2: Primary Biliary Cholangitis Associated Autoimmune Disorders. Univariate analysis used to calculate OR, OR; odds ratio, CI; confidence interval, PSC; Primary Sclerosing Cholangitis, AIH; Autoimmune Hepatitis, T1DM; Type 1 Diabetes Miletus, AIHA; Autoimmune Hemolytic Anemia, UC; Ulcerative Colitis, CD; Crohn's Disease, SLE; Systemic Lupus Erythematosus, ITP; Idiopathic Thrombocytopenia Purpura, AIT; Autoimmune Thyroiditis, RA; Rheumatoid Arthritis, MG; Myasthenia Gravis.

Discussion

- Our large nationwide analysis study demonstrates higher prevalence of PBC in the US than previously reported, which is likely a result of diagnosis at an early stage and prolonged survival that may be related to improved management.
- The high association between PBC and other autoimmune conditions suggest shared common pathogenesis of immune-mediated destruction of end organs in genetically susceptible individuals.