

# Primary Sclerosing Cholangitis: South Africa

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# **Incidence and Prevalence of Primary Sclerosing Cholangitis: A Meta-analysis of Population-based Studies**

**14 studies on incidence and the 12 for prevalence originated from North America, Asia, Europe, and Oceania**

- Incidence rates: 0.87 (95% CI 0.59-1.29) per 100 000 persons
- Prevalence rates of 13.53 (95% CI 10.20-17.94) per 100 000 persons

**No studies from Africa and Latin America**

# Primary Sclerosing Cholangitis: UCT/GSH Liver Clinic

**Retrospective cross-sectional evaluation: 128 PSC patients:  
30-year period: 1987-2021**

**51% (65) male, overall median age 38 [IQR 23-50] years**

**Ethnic distribution:** White: 50 (39.1%)      Mixed Ancestry: 41 (32%)  
Black African: 29 (22.7 %)      Asian: 8 (6.3%)

**PSC - IBD: 73 (57%)**

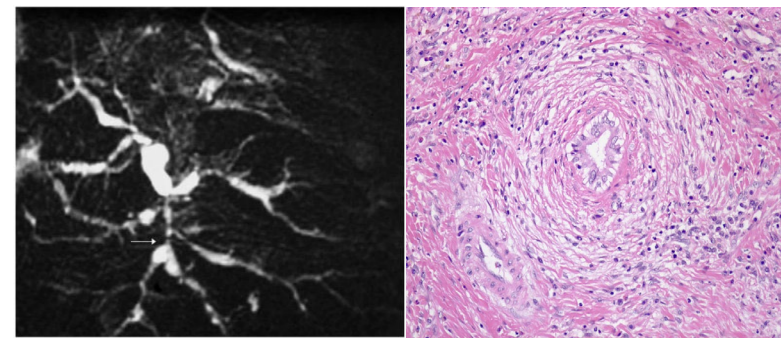
• **UC: 78% (57)      Crohn's disease: 16.4% (12)      IBD-undifferentiated: 5.5% (4)**

**PSC phenotype**

• **Large duct PSC: 88% (113)      Small duct PSC: 12% (15)**

**Diagnosis**

• **MRCP: 82 (64%)      ERCP: 30% (39)      Liver biopsy: 87 (68%)**



# Primary Sclerosing Cholangitis: UCT/GSH Liver Clinic

**Retrospective cross-sectional evaluation: 128 PSC patients:  
30-year period: 1987-2021**

**Presentations: 44% (56) cirrhotic and 41% (23/56) had PSC/AIH overlap**

- Bacterial cholangitis initial presenting feature: 3.9% (5)

**Mean prognostic scores at presentation**

- Amsterdam-Oxford 2017 score:  $2.16 \pm 0.89$
- Mayo score:  $0.78 \pm 1.44$
- MELD score:  $12 \pm 6.25$

**PSC associated malignancies: Clinical follow-up**

- Cholangiocarcinoma: 5% (6)
- HCC: 1% (1)
- Colorectal carcinoma: 3.1% (4)

# Primary Sclerosing Cholangitis: UCT/GSH Liver Clinic

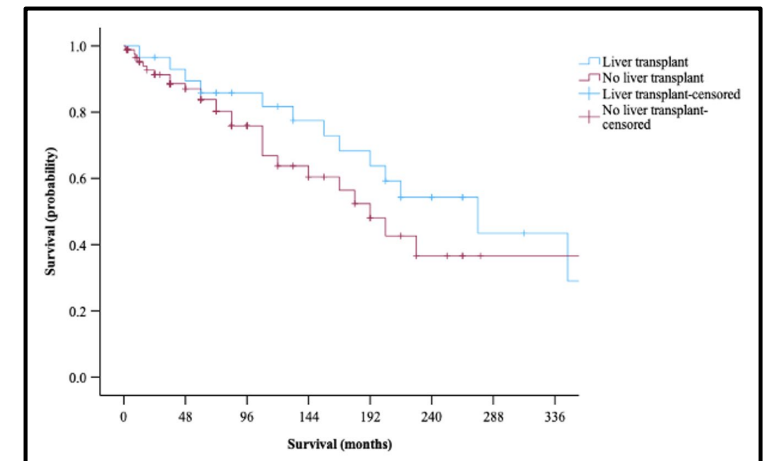
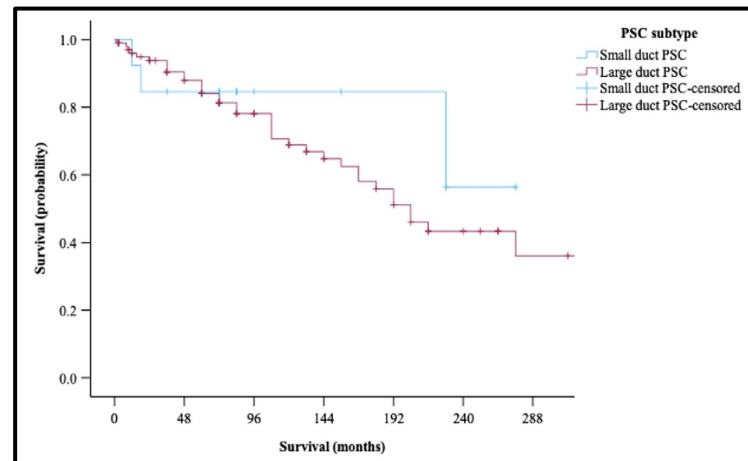
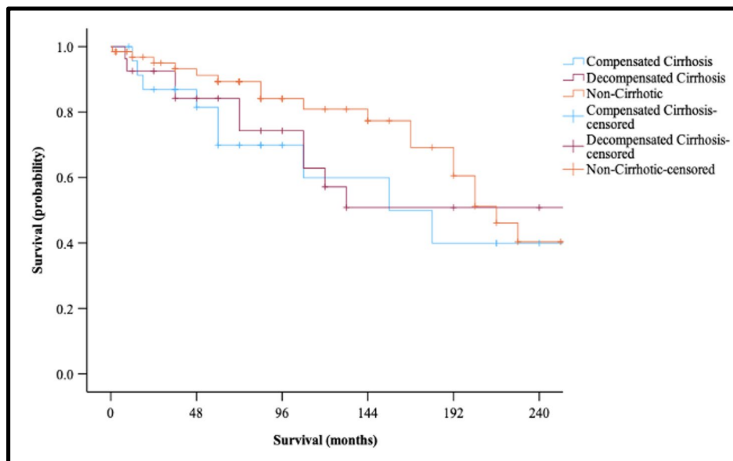
**Retrospective cross-sectional evaluation: PSC: 30-year period**

## Management

- 126/128 patients: UDCA
- PSC/AIH overlap: 92% (35/38): Corticosteroids
- PSC/IBD phenotype, 19% (14/73): Bowel resection

## Outcomes

- **22.7% (29/128) underwent orthotopic liver transplant**
  - 3, 5 & 10-year survival rates: 89.6%, 83% and 67%, respectively
- **Un-transplanted patients: 11% (11/99) lost to follow up; 70.5% (62/88) alive**
  - 29.5% (26/88) died: Liver related mortality: 39% of deaths (10/26)

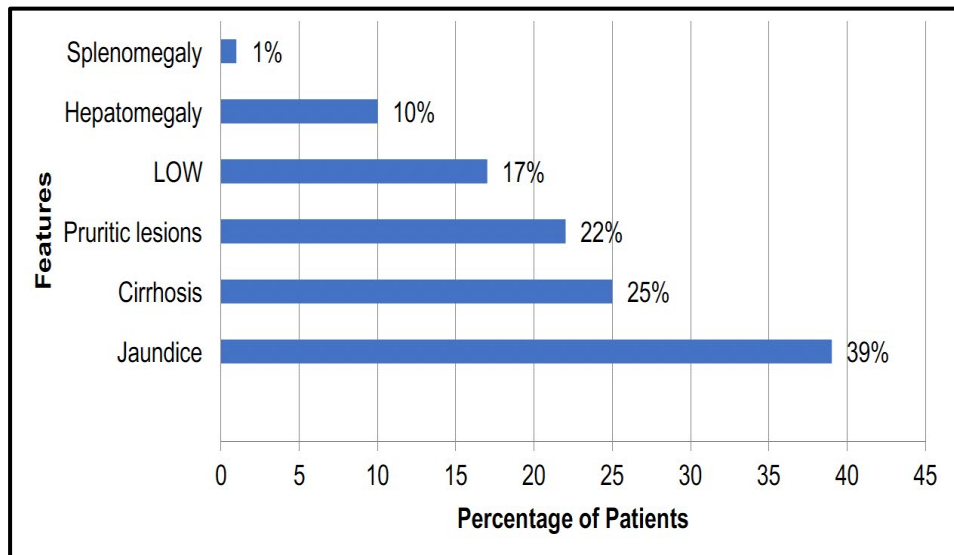


# Primary Sclerosing Cholangitis: SA: Johannesburg

Charlotte Maxeke Johannesburg Academic Hospital: GIT Clinic: 1 Jan 2008 - 31 May 2014

69 PSC patients: 34 Male: Median age: 39 years & 35 Female: Median age: 42 years

- **35 patients with PSC alone:** 2 (3%) developed cholangiocarcinoma during f/u
  - **Ethnicity:** Black African (22) Asian (7) White (4) Mixed Ancestry (1)
- **34 patients with PSC-IBD:** Ulcerative Colitis (20): 56.7% had pancolitis Crohn's disease (4)
  - **Ethnicity:** Black African (25) Asian (2) White (6) Mixed Ancestry (2)
- **Clinical & radiological findings similar in both groups: 39% presented with jaundice**



## No correlation between ethnicity and PSC disease

- HLA-B27 in black African patients may pose a risk for the development of PSC and IBD
- No colorectal carcinoma

# Primary Sclerosing Cholangitis, Crohn's disease and HLA-B27 in Black South African Women: Universitas Hospital, Free State

## 2 Black South African: Crohn's disease, PSC and HLAB27

	Patient 1 Baseline (biopsy)	Under therapy (6 months)	Patient 2 Baseline (biopsy)	Under therapy (6 months)
Age (years)	26		44	
Gender	Female		Female	
Ethnic origin	Black African		Black African	
Language group	Sesotho		Sesotho	
ALP (40-120 U/L)	513	809	497	125
$\gamma$ GT (0-35 U/L)	666	835	300	82
Bilirubin (< 17 mol/L)	11	20	8	5
CRP (mg/L)	20.2	52	7.2	8.4
HLA-B27	Positive		Positive	
ANA	Negative		Negative	
SMA	Positive		Positive	
AMA	Negative		Negative	
c-ANCA	Positive		Positive	
p-ANCA	Negative		Positive	
Magnetic Resonance Cholangiography	Inconclusive		Inconclusive	
Liver histology	PSC		PSC	

# **Primary Sclerosing Cholangitis in SSA**

**Reports on prevalence, presentations and outcomes in SSA are rare**

**UCT/GSH Liver Unit Data: Represents largest PSC cohort reported to date in SSA**

- PSC and AIH/PSC overlap syndromes seen across all ethnic groups
- Lower rate of PSC/IBD observed: 57% vs. globally reported 70-80%
- 44% patients in our cohort had advanced disease at presentation
- Transplant outcomes favourable: Patient survival: 90%, 83% and 67% at 3, 5 and 10 years; respectively

**PSC most likely underdiagnosed in SSA**

- **Requires access to MRCP, ERCP & liver biopsy**

**Visit Sandy Mosenye's Poster 4372: Primary Sclerosing Cholangitis in South Africa-  
A single centre longitudinal experience: 18 November 2024**