**Figure S1**: Number of PSC patients from the first cohort according to disease severity and comorbidity with IBD. LTX, liver transplantation; PSC, primary sclerosing cholangitis; PSC-CD, patients with primary sclerosing cholangitis and Crohn's disease; PSC-UC, patients with primary sclerosing cholangitis and ulcerative colitis; UDCA, ursodeoxycholic acid. Recurrence and no recurrence reports recurrence of PSC after liver transplantation.

52 PSC patients
(38 UDCA)

13 PSC-only
(10 UDCA)

22 PSC-UC
(14 UDCA)

17 PSC-CD
(14 UDCA)

7 Stable
(6 UDCA)

6 LTX
(4 UDCA)

9 Stable
(8 UDCA)

6 Liver cirrhosis
(5 UDCA)

7 LTX
(1 UDCA)

13 Stable
(10 UDCA)

3 Liver cirrhosis
(3 UDCA)

1 LTX
(1 UDCA)

2 recurrence
(2 UDCA)

4 no recurrence
(2 UDCA)

1 recurrence
(0 UDCA)

6 no recurrence
(1 UDCA)

1 recurrence
(1 UDCA)
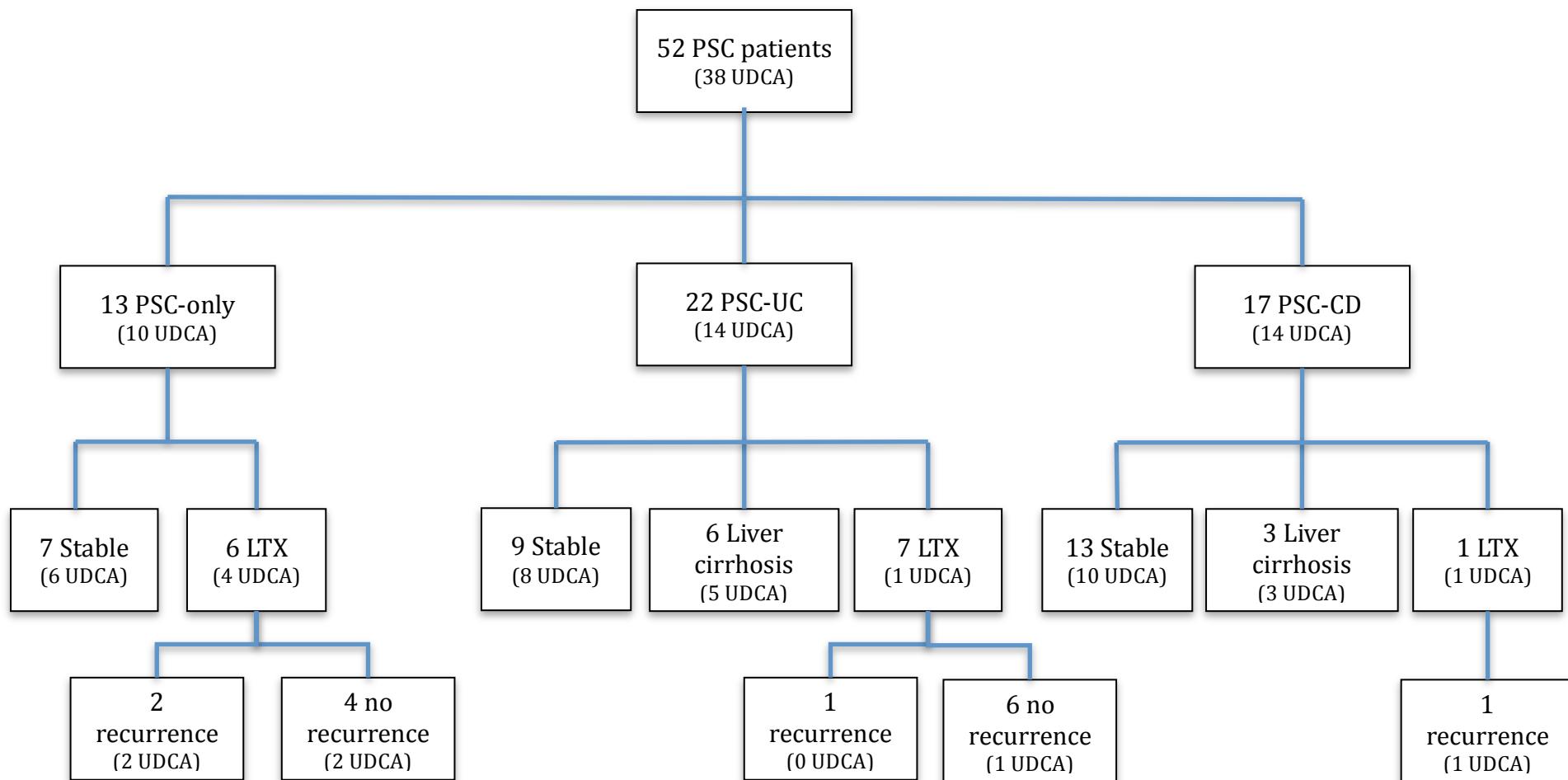
Figure S2: J48 decision tree for PSC diagnosis based on the Enteroccocus, Fusobacterium and Lactobacillus abundances in PSC patients and healthy controls, trained on the first cohort (n=104, Weka 3-6-12). The numbers in the decision tree indicate the number of OTUs needed to make the decision.
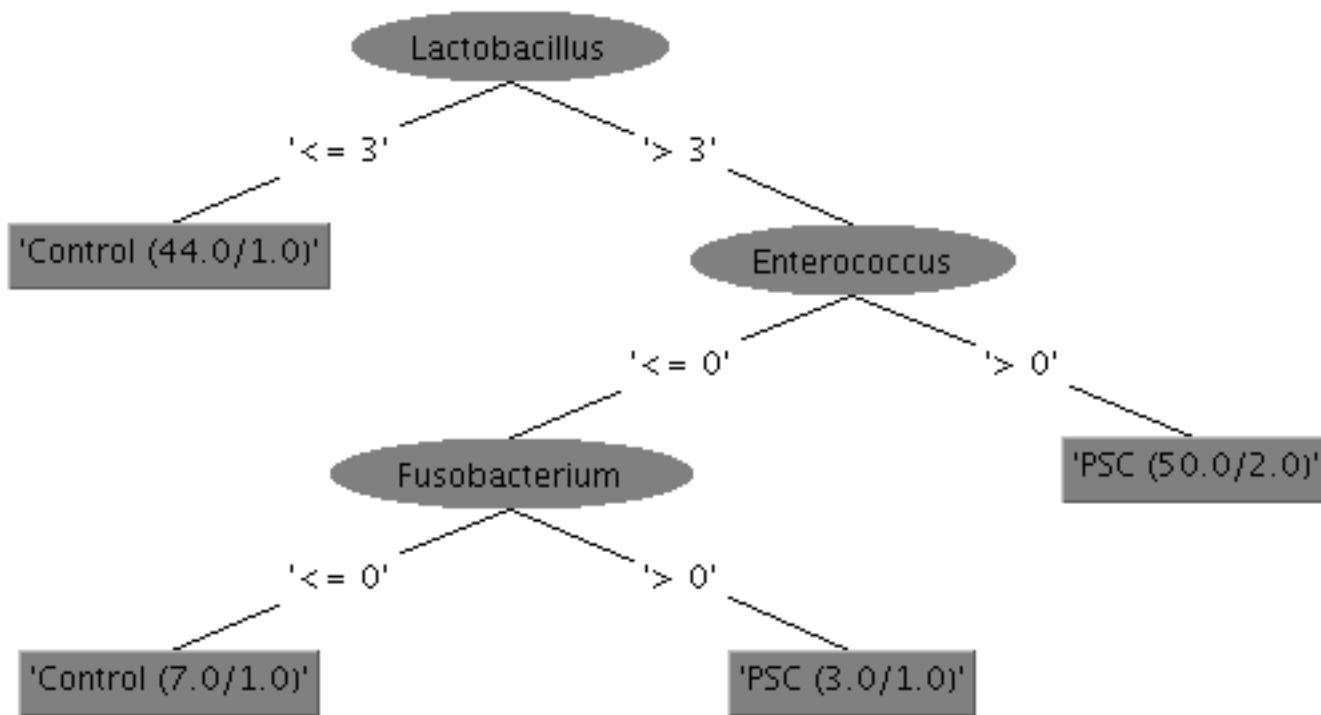
Figure S3a: Genus abundances (log-transformed) according to PSC severity, for the genera significantly associated to PSC.

Enterococcus and PSC severity (n=104)

Fusobacterium and PSC severity (n=104)

Lactobacillus and PSC severity (n=104)
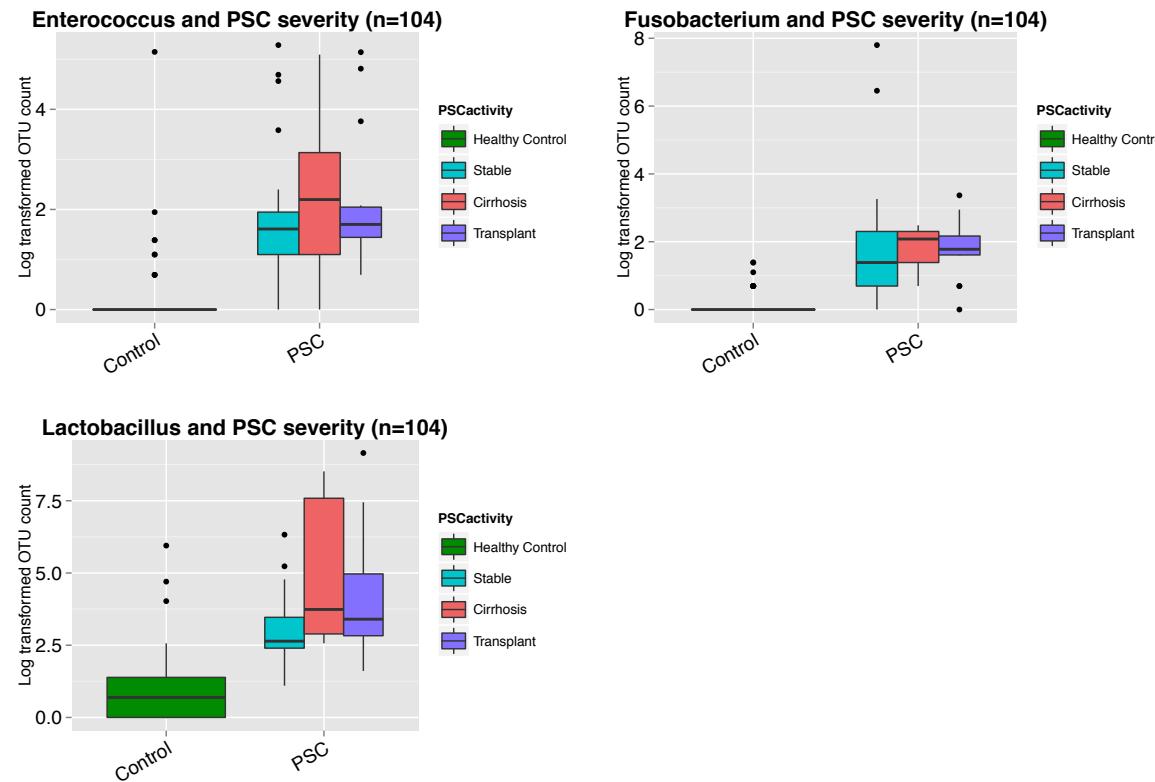
Figure S3b: Genus abundance (log-transformed) according to IBD activity, for the genera significantly associated to PSC.
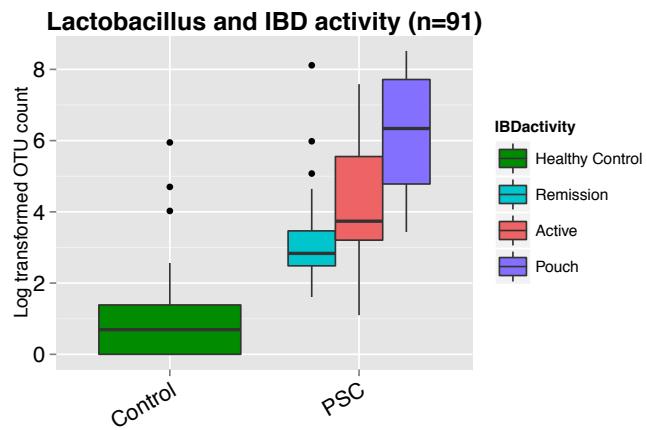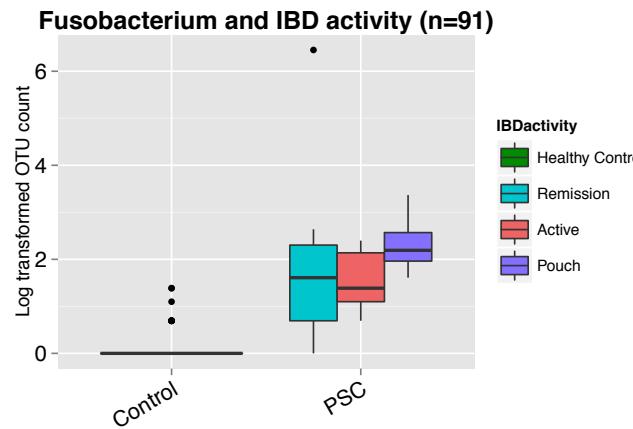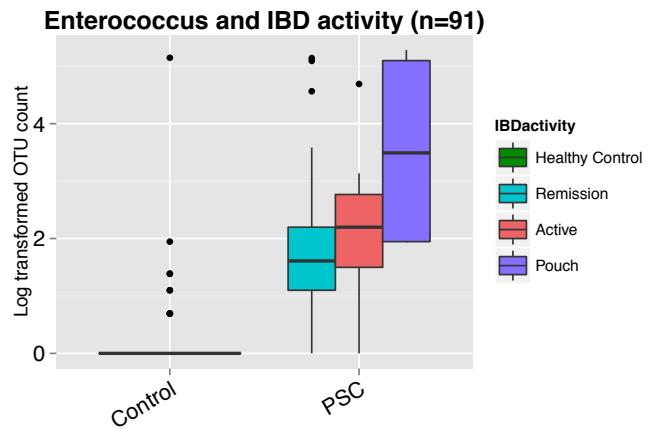
Figure S4: Oligotypes from genus *Enterococcus*

OLIG1	1,267	TG *b*	TACGTAGGTGGCAAGCGTTGTCCGGATTTATTGGGCGTAAAGCGAGCGCAGGCGGTT**T**CTTAAGTCTGATGTGAAAGCCCCCGGCTCAACCGGGGAGGGTCATTGGAAACTGGGA**G**ACTTGAGTGCAGAAGAG
OLIG2	109	TA *b*	TACGTAGGTGGCAAGCGTTGTCCGGATTTATTGGGCGTAAAGCGAGCGCAGGCGGTT**T**CTTAAGTCTGATGTGAAAGCCCCCGGCTCAACCGGGGAGGGTCATTGGAAACTGGGA**A**ACTTGAGTGCAGAAGAG